Patient experience of living with benign essential Blepharospasm

Dunlop, Nicola (2022) Patient experience of living with benign essential Blepharospasm. Doctoral thesis, University of West London.

PATIENT EXPERIENCE OF LIVING WITH BENIGN ESSENTIAL BLEPHAROSPASM

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A thesis submitted in partial fulfilment of the requirements of the University of West London
for the degree of Doctorate of Nursing

September 2022

Benign Essential Blepharospasm

Abstract

Background: The fundamental rights of people with benign essential blepharospasm (BEB) to access timely care and treatment is significantly challenged in the United Kingdom and across the world. The condition causes intermittent and sustained spasms, temporary blindness, facial contortion and social stigma which debilitates the lives of those affected. Patient feedback and personal observation as a specialist practitioner within an outpatient clinic suggest the care provided fails to meet individuals' needs at a time of acute relapse.

In the absence of qualitative evidence, a systematic quality of life review was undertaken in relation to BEB care and treatment. Although people with BEB were shown to have poorer quality of life than healthy individuals, quantitative evidence with numerical scales fails to represent the lived experience of BEB. Retrospective to the findings of the current research, a metasynthesis of BEB care and treatment experience was examined through text and opinions from the UK, the United States of America (USA), Australia and Canada dystonia websites. This offered health professionals a model guide on how to improve services.

Aim: The aim of this study is to explore the impact of BEB outpatient treatment and care on patients' lives.

Method: The study data emerged from 10 individual interviews and one focus group discussion. It provided a deeper understanding of BEB lived experience. A lack of knowledge, visual, functional and psychological decline, namely the anxiety experienced from untimely care and treatment in an acute relapse. Qualitative assessment of the patient experience provided the means to give patients a voice to improve the service. This study offers a novel contribution – it bridges the gap between policy, theory and practice.

Outcome: The service has been configured through electronic treatment records, NHS Attend Anywhere video severity assessment to improve access, greater community through focus group discussion, uniform practitioners' training, the introduction of more injectors and clinics to improve geographical disparity. The service is set to address delayed referral, communicating a list of BEB red flags to primary care.

Acknowledgments

All thanks and praise is to God, my heavenly father. I want to thank my husband, Ernest Dunlop, for his patience in keeping me company as I worked on my thesis from dusk until dawn on many occasions. To my daughter, Renee, thank you for always being there each time I requested you read a chapter. To my sons, Ernest Jr and Nicholas, thank you for your understanding. To my mum Venita Rayside, my gratitude to you for always instilling the belief that I can accomplish anything I set out to do.

To the librarians Desa Bokre of the Institute of Ophthalmology and Marc Forster of the University of West London, I am grateful for your assistance in my literature search for this study. Last but not least, thank you to Professor Heather Loveday, who has been instrumental in her guidance and supervision – this research would not have been possible without her support.

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List of Abbreviations

BEB	Benign Essential Blepharospasm
BFMDRS	Burke-Fahn-Marsden dystonia rating scale
BSDI	Blepharospasm Disability Index
BTX	Botulinum toxin
CDQ	Craniocervical Dystonia Questionnaire
DBS	Deep Brain Stimulation
GBI	Glasgow Benefit Industry
GP	General Practitioner
HAD	Hospital Anxiety and Depression Scale
HRQL	Health-Related QoL
JBI	Johanna Briggs Institute
IPQR	Illness Perceptions Questionnaire-Revised
JRS	Jankovic Rating Scale
MMSE	Mini-Mental State Examination
NHS	National Health Service
OSDI	Ocular Surface Disease Index
QoL	Quality of Life
RCT	Randomized Controlled Trial
REC	Research Ethics Committee
SES	Self-Esteem Scale
SF-36	36-item short-form
TRI	Treatment Representations Inventory
UK	United Kingdom
WHOQOL	World Health Organisation Quality of Life
WHOQOL-BREF	World Health Organisation Quality of Life Abbreviated Form
WAIS-III	Wechsler Adults Intelligence Scale

Overview of Thesis

This thesis explores the lived experience of people with BEB and how dystonia services can be remodelled to provide timely access to care. Benign essential blepharospasm is a rare, progressive chronic neurological disorder characterised by involuntary sustained or intermittent muscular contractions of the eyelids and upper facial muscles, which results in closure of the eyelids, abnormal facial expressions, distress and functional blindness from periodic inability to open the eyes and could lead to ideas of suicide (Berardelli et al., 2021). Benign essential blepharospasm prevalence worldwide is 16 to 133 cases per million (Defazio & Livrea, 2002).

There were 7,000 people in the United Kingdom (UK) with the condition in 2016. However, due to symptom variability, the precise number of new yearly BEB cases is unknown. The services for BEB treatment in the UK are primarily restricted to highly specialist ophthalmic or neurology outpatients' clinics. The focus of this study stems from my personal experience as a specialist ophthalmic nurse working in an outpatient setting which provides care and treatment for people with the rare and chronic condition of BEB. Personal observation and patient feedback suggest that patient dissatisfaction and distress occur when an individual experiences an acute exacerbation of the condition before the scheduled three-monthly cyclical Botulinum toxin (BTX) injections, which minimizes the symptoms. Services provided fail to meet their individual needs at the time of acute relapse.

Globally, chronic conditions present a significant challenge for health and well-being and increase the pressure on health services. The psychosocial and economic impact of individuals with chronic conditions is often serious, causing deprivation, social isolation, and mental health issues. The UK has 26 million people living with at least one long-term condition, resulting in a three-fold increase in the cost to the NHS (Office of National Statistics, 2011, Household Census). Furthermore, long-term conditions accounted for 50% of the GP and 64% of the outpatient department (OPD) appointments. The NHS Long-Term Plan (NHS, 2019) includes patient-centred care as a fundamental principle for redesigning outpatient services so that people receive the proper care at the right time.

Throughout the world, policymakers and healthcare practitioners have advocated patient empowerment as a means of addressing the management of long-term conditions and the configuration of services to define their quality (Roberts, 1999; Sanderson and White, 2018; Kayser et al., 2019). In addition, in the UK, the Health and Social Care Act (DOH, 2012; DHSC, 2021) has called for quality improvement and local innovation to deliver the best quality of care to all. People with BEB require prompt diagnosis, treatment, and care, but this is often difficult because of deficiencies in the diagnostic abilities of health professionals due to a lack of awareness and knowledge about BEB, the effect it has on daily life, and how care and treatment are provided. Hence, there is a lack of timely care within primary and secondary care settings.

Benign essential blepharospasm treatment has been studied extensively. However, the study of patient experience has been restricted to quantitative measurements of symptoms, the effectiveness of BTX and quality of life (QoL). Clinical rating scales provide objective measurement, and their main disadvantage is that they do not consider the patient’s perspective of disability and the condition impact on daily life (Wabbels & Roggenkämper, 2012). This study adds to the body of knowledge about the impact of BEB as previous assessments of the patient experience are situated in the realist paradigm or namely quantitative research.

This study offers an original contribution to knowledge and a deeper understanding of the impact of the condition, its treatment, and care on patients' lives. It provides insights that can inform service redesign so that timely treatment and care restores patients as close to their previous health status as possible and addresses the impact that BEB has on individuals' self-esteem and QoL, including the maintenance of psychosocial well-being and physical functioning.

This thesis is comprised of seven chapters:

Chapter 1: The introduction chapter sets the scene of the broader thesis. It defines BEB and outlines the research context of chronic and rare diseases by examining the global, national, and local health services. It describes the symptoms of BEB, highlights the effectiveness of BTX, summarises other existing treatments and critically discusses the QoL literature. Finally, the researcher position, study aims, rationale, and research objectives/questions are outlined.

Chapter 2: The chapter reports a QoL systematic review on the impact of BEB care and treatment. It addresses the goals, expectations, standards, and concerns of people with BEB. This chapter focuses on gender, duration and worsening of the condition, aggravating and relieving factors, visual and functional ability, depression and anxiety, treatment belief and illness perceptions and delays and BTX treatment.

Chapter 3: Presents a systematic review or meta-synthesis of 25 published patients' narratives of personal experiences of BEB in the public domain of the UK, Canada, the United States of America (USA), and Australia. Each narrative describes the diagnostic or treatment experience of people with BEB. The meta-synthesis was undertaken due to the lack of qualitative studies uncovered in a systematic literature review. Utilising the Johanna Briggs Institute (JBI) approach, it situates BEB within the medical literature.

Chapter 4: The method chapter provides presents the rationale for the philosophical and theoretical underpinning of the research and the methodological choices and methods used to undertake the research programme. The chapter is divided into four sections; principles of the theoretical approach used; description and rationale for the chosen methodology; description of the setting and details of how the data were collected and analysed and the final section discusses ethical considerations applied.

Chapter 5: The findings chapter uses dialogue to place the patient at the centre of care firmly. It examines the impact of BEB from diagnosis to treatment and explores the psychological turmoil experienced when seeking care, especially within the outpatient clinic during an acute episode of BEB. It raises the voice of the patients as experts and not just as recipients of care but as co-partners actively seeking to improve their QoL through better-designed healthcare delivery.

Chapter 6: Presents the focus group discussion (FGD) represents participants individually voiced but collective aspirations to improve the specialist BEB clinic. The FGD is representative of Phase 2 of the research and is organised to solicit the participants' combined response to improving their world. The report provides a summary of participants recommendations within the themes generated by the discussion. It addresses the third objective of this study: to inform service improvement or redesign that responds to the needs of BEB patients.

Chapter 7: This chapter discusses the results of the qualitative exploration of the impact of BEB, outpatient treatment, and care on the lives of patients and informs service improvement for the realisation of quality care. It critically discusses the significant findings with the support of known literature in Chapters 1, 2, patient narratives in chapter 3, the findings in chapter 5, NHS quality framework, and IPA. It discusses BEB as a complex constructed interpretation through participants' self-reflection and the researcher's analytical interpretations. It also discusses the participants recommendations and finally outlines the limitations and areas for future research as it concludes in answering the research questions.

Chapter 8: Provides evidence of the researcher's doctoral journey specifically the learning and development achieved while undertaking the professional doctorate course. The research and development framework from the Careers Advisory Centre, 2010, along with (Rolfe, 2001) reflective model, was utilized to explore the doctorate journey. The chapter demonstrates the researcher's knowledge, attributes, behaviour as it presents the impact of the research.

Chapter 1: Introduction

Spasmodic eyelid closure which significantly debilitates the lives of people with BEB could be significantly reduced by greater knowledge and effective organisation of care and treatment. This chapter provides an overview of the condition through BEB presentation, diagnosis, and treatment. It outlines the national and local context and relates BEB to other rare and chronic diseases. Furthermore, it sets out the researcher position. The chapter highlights the knowledge gap found, the aim and objectives of the study as it concludes.

1.1 Benign Essential Blepharospasm

1.1.1 Definition

Benign essential blepharospasm is a rare and progressive neurological disorder characterised by involuntary sustained or intermittent muscular con traction of the eyelids and upper facial muscles. The disorder results in closure of the eyelids, abnormal facial expressions (see figure below), distress, functional blindness, social and emotion consequences (Jinnah et al., 2013; Valls-Solle & Defazio, 2016; NINDS, 2018).

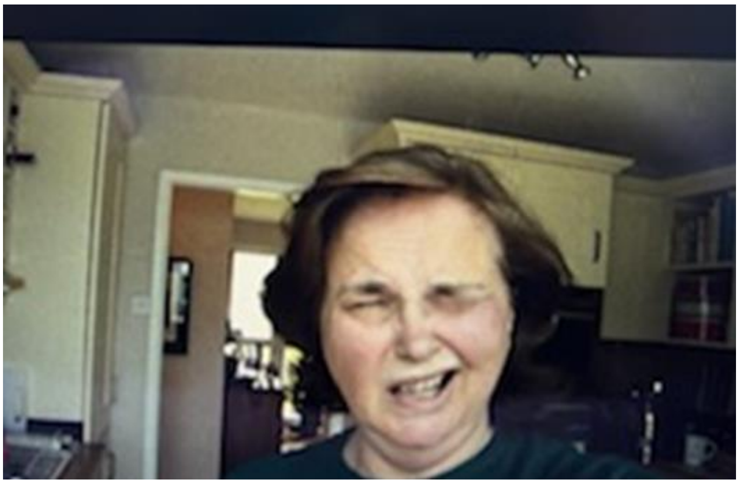


Figure 1: BEB with phenotype Meige (permission gained, see Appendix 1 additional photograph and email contact)

1.1.2 Epidemiology

The scarcity of BEB studies hinders epidemiological knowledge. However, the known prevalence of BEB worldwide is 16 to 133 cases per million (Defazio & Livrea, 2002). It affects the lives of around 7,000 adults in the UK annually (The Dystonia Society, 2016). The condition is more prevalent in women (Coscarelli, 2010), with a ratio of 2.3:1 (Defazio & Livrea, 2002). Women also show more severe effects of the disease (Muller et al., 2011).

The mean age of BEB onset ranges between 50-70 years (Tanner et al., 2013; Defazio et al., 2017), although the true incidence of BEB is uncertain as many people remain undiagnosed. In one study, only 10% of people were accurately diagnosed with BEB during their first encounter with a medical professional. In another study, 60% of BEB patients saw at least five physicians before they received a definitive diagnosis in a period of between one and five years (Wakakura et al., 2018). A study by Rare Disease UK (RDUK, 2016) demonstrated that patients felt they were not given enough information or support on their journey to diagnosis. Not having a diagnosis was a significant barrier to accessing coordinated care and appropriate treatment.

1.1.3 Pathophysiology

Benign essential blepharospasm is a disorder of the basal ganglia of the brain in the area that controls blink co-ordination (Berardelli et al., 1985). Recent evidence from neurophysiology and neuroimaging (see Figure 2) has indicated the involvement of several brain regions (Jiang et al., 2019). This confirms previous findings that BEB is unlikely to result from a single defect in the circuitry (Quartarone et al., 2006; Peterson & Sejnowski, 2017).

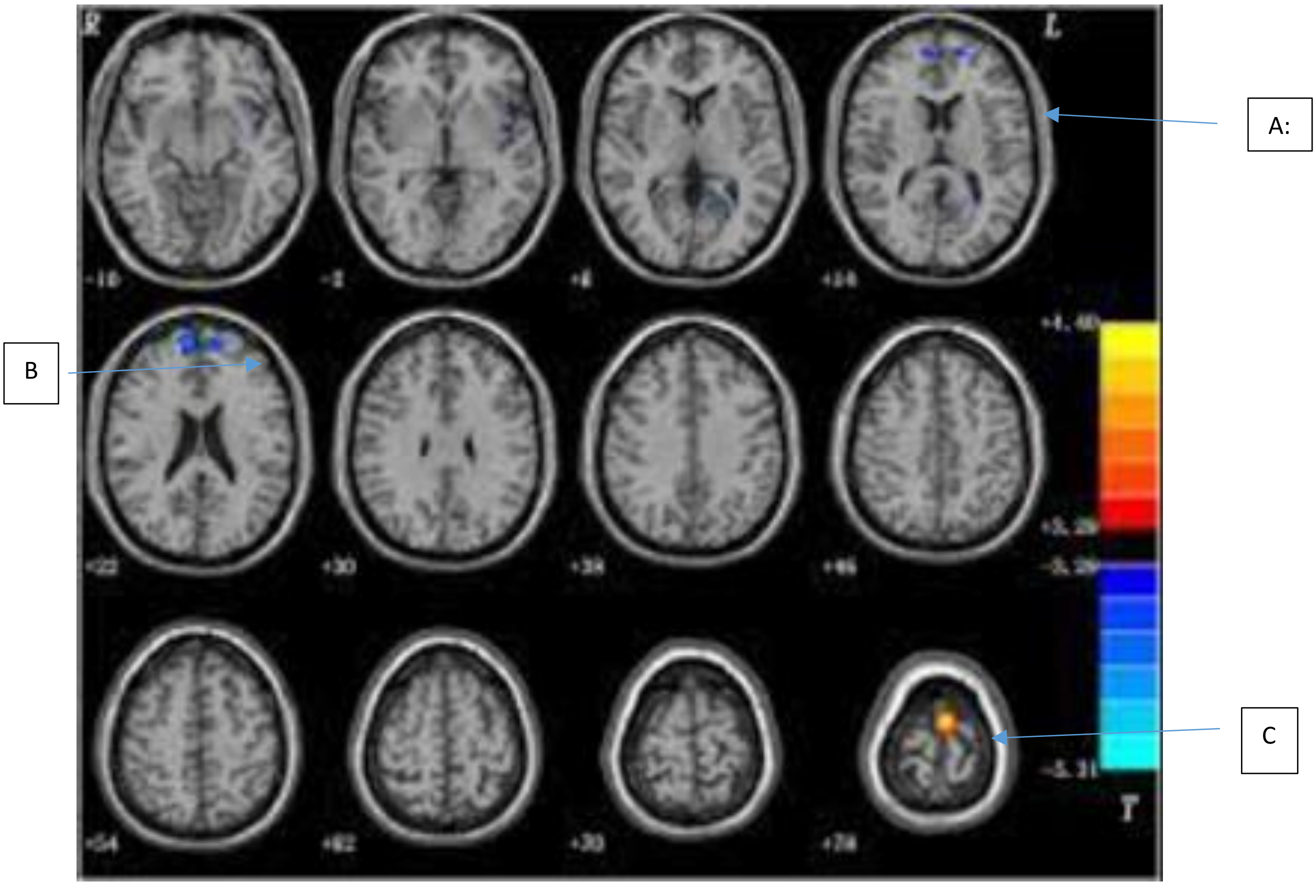


Figure 2: Abnormal neural activity at rest in BEB (Baker et al., 2013)

1.1.4 Risk Factors

Neural imaging of a BEB patient at rest, shows various areas of involvement (A,B&C) and led to the explanation that the cause may be multifactorial. The possible risk cause included numerous environmental factors (Defazio et al., 2017). Various conditions such as neurodegenerative disease, brain lesions, and exposure to neuroleptic medications have been cited as possible factors in the cause of BEB (Grandas et al., 1998). In contrast, Digre (2015) identified genetics as a likely cause of BEB. A study by (Dong et al., 2020) specifically named the gene mutation SYNE1 and CIZ1 as a contributory cause of BEB. Another risk factor identified was structural damage of the eye due to injury or previous infection (Hallet et al., 2008). However, some studies have provided evidence to suggest one or more stressful events example the death of a loved one precede the focal dystonia BEB (Cossu et al., 2016; Roggenkämper & Nüssgens, 1997).

1.1.5 Dystonia classification

The term dystonia encompasses a group of movement disorders (Lewis et al., 2008). People with BEB, like those with other dystonias, experience repetitive movements, contractions and contortions (Gürsoy et al., 2013). The classification of dystonia is determined by the number of body areas involved (see Table 1).

Table 1: Dystonia classification

Focal dystonia	One isolated area of the body involved
Segmental dystonia	It affects two or more close areas
Multifocal dystonia	It affects two or more distant areas
Hemidystonia	It affects half of the body
Generalised dystonia	It affects the whole body

1.1.6 Symptoms of BEB

The symptoms of BEB involve both eyes and comprise both motor and non-motor features (Albanese et al., 2013). Initially, symptoms include bilateral orbicularis oculi muscular contractions, which could be intermittent blinking and/or sustained closure (Berardelli et al., 1985; Defazio, 2017). Sustained contraction of the eyelid can be disabling, and, in a severe case, a person can be rendered functionally blind (Ben Simon and McCann, 2005). The inability of the eyelid to open is due to inhibition of the *levator* *palpebrea superioris* muscles, also known as apraxia of the eyelid opening. A characteristic feature of BEB is sensory tricks or *geste antagoniste* used by patients to relieve their symptoms example, palpating area around the eye, humming, whistling, yawning, coughing and adjusting glasses (Greene & Bressman, 1998).

The progression of the disease to adjacent facial muscles causes contraction and contortion of the *procerus, frontalis, corrugator supercilii* and some lower facial muscles, such as zygomaticus and mentalis (Tolosa & Martí, 1988) (see Figure 3). Facial contortions affect appearance, cause headaches, and lead to impaired concentration and irritability (Gürsoy et al., 2013; Reimer et al., 2005). Lower facial muscle involvement, including the mouth, tongue, and jaw, signals new dystonia known as Meige Syndrome (Hwang & Eftekhari, 2018). Severe mouth twitching can affect speech and eyelid closure, and facial spasms could lead to social anxiety (Streitová & Bareš, 2014).

Patients with BEB also have non-motor symptoms such as psychiatric illness, specifically depression, anxiety, and obsessive-compulsive disorders (Conte et al., 2015; Valls-Sole & Defazio, 2016). In addition, sleep disturbance, cognitive disorders and ocular complaints, such as dry eyes and sensitivity to the sun and bright light, are also symptoms associated with BEB.

There have been debates about whether non-motor symptoms are a part of the condition BEB. A study by (Ferrazzano et al., 2019) concluded that non-motor symptoms represent the condition's clinical spectrum. Non-motor symptoms have also been shown to significantly impact QoL (Hall et al., 2006; Pekmezovic et al., 2009). A detrimental impact is its impact on activities of daily living and perceived stigma (Lawes-Wickwar et al., 2021).

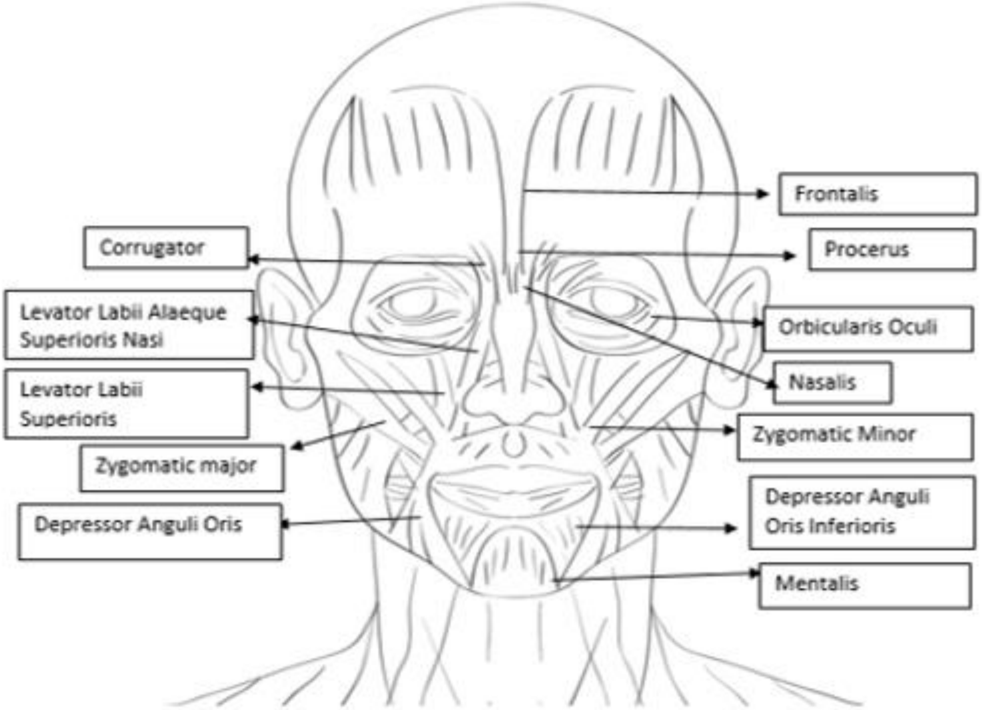


Figure 3: Facial Muscles Frequently Injected with BTX (Google free image modified with labelling)

1.1.7 Diagnosis

Various factors have been identified for the lack of prompt diagnosis. They include the rarity of the BEB, and its relatively low morbidity compared to other neurological conditions (Defazio & Livrea, 2002). A lack of information about the condition is another factor that prevents disease recognition for diagnosis and treatment of BEB (Valls-Solle & Defazio, 2016). Nevertheless, it is essential to consider that the diagnosis of BEB is based on the signs and symptoms and elimination of all possible secondary causes through a thorough neurological evaluation.

Therefore, an entire systematic clinical history and observation of the eyelid, face, and extremities is undertaken. Careful slit-lamp examination is helpful in the detection of ocular causes for the spasm, such as ocular dryness and structural damage (Belinsky et al., 2019). All the above factors can cause a delay in referral but once diagnosed, BEB treatment with BTX is effective.

1.2 Treatment of BEB

1.2.1 Botulinum toxin

Botulinum toxin (BTX) is a neurotoxin that originates from clostridium botulinum, a gram-positive anaerobic bacterium (Sampaio, 2004) that causes food poisoning. Commercially BTX is prepared to produce the required neurotoxin to paralyse muscles (Simpson, 1981). Although botulinum neurotoxins consist of seven different serotypes (A to G), only two strains (A&B) are currently used (Ababneh et al., 2014).

Currently, three formulations of BTX type A are available: onabotulinumtoxinA (Botox; Allergan USA formulation), abobotulinumtoxinA (Dysport; Ipsen-Pharma, UK) formulation, and incobotulinumtoxinA (Xeomin; Merz Pharma, Germany formulation). Two botulinum type B formulation is available; rimabotulinumtoxinB (Myobloc in the USA; Supernus Pharmaceuticals, Inc USA, and Neurobloc in Europe; Sloan Pharma, Switzerland).

Botulinum toxin is considered the treatment of choice for BEB due to its safety and effectiveness (Costa et al., 2004). The relative effectiveness of BTX A in comparison to BTX B in terms of; severity, disability, pain, and safety has shown no difference in multiple level 1 randomised control trials (RCT) undertaken see (Appendix 2). Three randomised control trials recognised BTX superiority over the placebo, normal saline (Fahn et al., 1985, Frueh et al., 1988, Jankovic, 1988). Although placebo-controlled trials continue to be published, Costa and colleagues (Costa et al., 2004) has voiced concern about the ethics of these trials given the volume of evidence for the effectiveness of BTX in the treatment of BEB.

Primarily patients experience efficacy with BTX, but a loss of effectiveness has been reported. Although BTX offers temporary relief, it is not understood why the duration of its treatment efficacy varies (Jinnah et al., 2013). Botulinum toxin effects can be last on average of 3 to 6 months (Calace et al., 2003). However, there is no contraindication to a shorter treatment cycle. In fact, (Jankovic et al., 2011) suggest that 6-week treatment cycles are clinically safe. Therefore, it may be concluded that a shorted treatment plan is an economic decision rather than a QoL.

However, when there is no effect from BTX, it is considered due to the formation of neutralising antibodies (Green et al., 1994). Though highly potent, the drug is neuro-specific with limited diffusion when injected by a skilled practitioner into the affected muscle (Naumann and Jankovic, 2004). Neutralising antibodies can arise if a practitioner lacks dosage knowledge or administers higher doses and booster treatment within the first and second week of initial treatment to a patient (Hellman and Torres-Russotto, 2015).

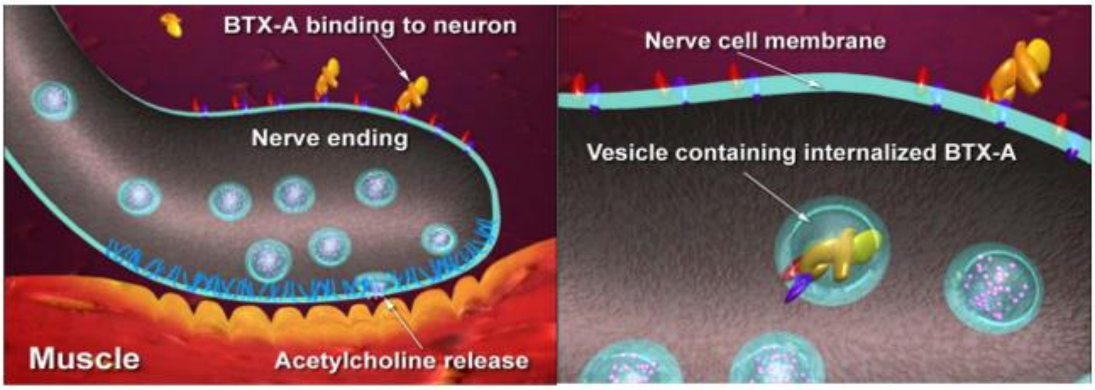
Wang et al. (2014) suggested that the host immune response to the neurotoxin may also contribute to local and systemic side effects. Botulinum toxin may be combined with other pharmacological interventions, emotional therapy, complementary or alternative medicines, spectacles and frames, and sensory tricks to improve effectiveness (see Table 2 below).

Table 2: Other treatments

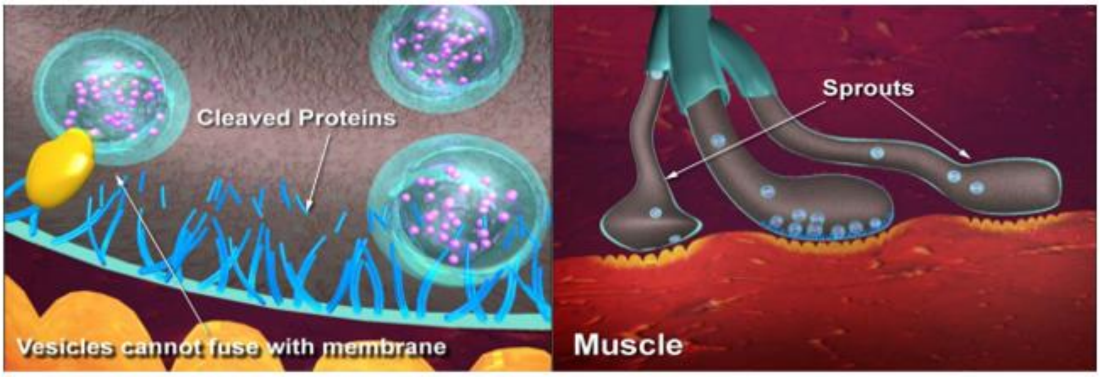
Medical Treatment	**Benefits**
Oral medications	Benzodiazepines, anticholinergics (benztropine and trihexyphenidyl), levodopa, baclofen, VMAT2 inhibitors (tetrabenazine), lithium, valproate, methylphenidate, and zolpidem offer varied effectiveness.
Surgery	Limited orbicularis oculi focus on the pretarsal orbicularis for patients with visual disabling apraxia of eyelid opening, but BTX is still required. Frontalis suspension is helpful in patients for whom BTX injections alone are ineffective.
Deep brain stimulation	Stimulation of the globus pallidus internus and pallidotomy has been described for Meige syndrome by combining BEB and oromandibular spasm.
Physiotherapy	Although evidence is absent, offer physiotherapy according to specific patient needs. Exercise strengthens non-dystonic muscles, ‘retaining’ tone, and stretching tight muscles.
Emotional Therapy	**Benefit**
Counselling	Counselling addresses the experience of diagnostic odyssey – describing their symptoms to multiple doctors over several months or years without getting a precise diagnosis. There is a need for support to cope with associated stigma, depression and anxiety, and BTX's initial treatment trial and error.
Complementary or alternative medicine	**Benefits** Chiropractic therapy, yoga, meditation, and other relaxation or stress reduction programs, acupuncture and dry needling, and others, although there is no evidence to suggest its efficacy.
Sensory Spectacle, Frames, and Tricks	**Benefits**
Sensory Trick Frames	Spectacles with silicone pressure pieces are fitted based on patient advice and help apply periorbital pressure to relieve spasms.
FL-41 Tint Prescribed Spectacles	Improves blink frequency, light sensitivity, and functional limitations in BEB.
Geste antagoniste or sensory tricks	Voluntary maneuver that temporarily reduces the severity of dystonic postures or movements example placing pressure on the temple area, singing, yawning, humming, and whistling.

1.2.2 Mode of action

Botulinum toxin, when injected, acts to prevent muscular contractions (see Figures 4 to 7) by preventing the release of acetylcholine and paralysing the muscle (Dolly, 2003; Silveira-Moriyama et al., 2005). Botulinum toxin works by binding to high-affinity target sites on pre-synaptic cholinergic nerve-endings that cause decreased release of acetylcholine at the neuromuscular junction, thereby inducing partial muscle paralysis. However, the axonal branches sprout retracts, the parent terminal is re-established, and the spasms recommence.



Binding Internalising



Blocking Resprouting of axonal branches

Figure 4: Botulinum toxin Mode of Action (Copyright Allergan Inc., Irvine, CA)

1.2.3 Side Effects

The administration of BTX requires a skilled and knowledgeable professional to reduce the intrinsic risk of systemic side effects, though rare (Naumann et al., 2006), deaths have been reported (Ault, 2008). The administration of BTX subcutaneously or intramuscularly into the affected muscle can cause temporary side effects. Side effects can be immediate and could take 7-10 days and last for several weeks (Calace et al., 2003).

Local side effects include redness, bruising, haematoma, infection, and pain at the injection site. Local side effects may be patient-related example the use of aspirin, anticoagulants, anti-inflammatory drugs, vitamin E, herbal remedies (Blitzer et al., 2000). The practitioner use of small gauge needle that is regularly changed after three to four injections and injecting in the dermis layer also reduce bruising, bleeding, and pain. Pain can be further reduced by the use of a high PH normal saline and topical anaesthetic (Vartanian and Dayan, 2005)

1.2.4 Complications

Poor injection technique, a lack of knowledge of facial and eyelid anatomy and physiology failure to adopt a conservative approach can result in self-limiting but significant ocular side effects (Colosimo et al., 2012). However, meticulous injection technique and knowledge of the eyelid anatomy, specifically staying 2-3cm above the supraorbital margin, avoiding the mid-pupillary line, and aiming the injection needle away from the pre-septal and pre-orbital orbicularis can avoid ptosis or droopy eyelid (King, 2016).

While apraclonidine eye drops can be used to moderately lift the eye to restore some vision, it should be used with caution as it can increase ocular pressure or worsen glaucoma. Injection BTX into the medial two-thirds of the lower eyelid could result in treating the inferior oblique muscle and double vision (Fahn et al., 1985). Other adverse events such as dryness and watering are related to decreased drainage of tears from BTX injected into the medial portion of the eyelid. Administration of an overdose of BTX to the palpebral portion of the lower lid orbicularis oculi may cause poor functionality of the lower eyelid. There is no specific treatment for this complication, and it usually settles within a matter of weeks.

BTX diffusion to adjacent muscles was related to high dosage and improper needle placement. High dose and incorrect placement into the glabella complex may result in paralysis of the face and lower eyelid resulting in lagophthalmus or poor closure of the eyelid, the eyelid rolling inwards (entropion) or outwards (ectropion) (Alimohammadi and Punga, 2017). If an ectropion develops, a prompt ophthalmological referral is recommended to prevent exposure to keratitis or corneal infection and damage (Vartanian and Dayan, 2005).

Therefore, understanding with age an adult anatomy changes and there is namely loose skin and low muscle mass which means there is a need for a lower dosage of BTX is important (Cillino et al., 2010). Higher doses of BTX increase the likelihood of side effects from spreading the toxin to adjacent muscles (Basar & Arici, 2016). A higher dose was found to increase adverse reactions. Therefore, a total dose per session of 80 units per eye is recommended (Bilyk et al., 2018). The diffusion of BTX can also be related to the actions of patients (Vartanian and Dayan, 2005) example placing pressure over the injection site. The adverse event effects can be sight-threatening, last between weeks and months (Walker & Dayan, 2014) reduce an individual QoL and question quality of care. People with BEB like other rare diseases care is challenged by poor identification of the condition and badly managed care (Von Der Lippee, 2017).

1.2.5 Quality of Care and Services for People with BEB

A major concern in the provision of treatment and care for people with BEB has been identified as delayed treatment and the waning effects of BTX which causes dissatisfaction and poor patient experience (Leplow et al., 2017). My experience as a specialist practitioner also suggests that the organisation of timely care is essential to quality treatment and care locally.

The quality of global healthcare is also considered poor when measured against the six dimensions of quality care; safety, effectiveness, person-centredness, timeliness, efficiency, and equity (Abrampah et al., 2018). Although there is insufficient evidence to determine the presence of quality of care since the patient experience of care, friends and family test, and patient-reported outcome measures that influence an individual decision to utilise or avoid a particular service are rarely used.

The first port of call for most people needing healthcare worldwide is primary care and an area where quality care can be determined. Certainly, in the UK, Canada, and Australia, quality care is provided through GPs prompt identification of a condition, referral to specialist care for the treatment of serious diseases (Kostopoulou et al., 2008) within their crucial gatekeeping roles. However, the known diagnostic errors that occur, though multifactorial (Berenson et al., 2014) cause quality of care to be questioned. Specifically, diagnostic errors may be related to missed opportunities, care systems, and cognitive factors (Singh et al., 2017), harming the patient-clinician relationship.

It is not unusual for people with rare diseases to experience a period of limbo, under-diagnosis, misdiagnosis, and delays, as is the case with BEB patients (Lee et al., 2018). In fact, people with rare diseases experience a frustrating, extended journey towards diagnosis, difficulties finding an appropriate sub specialist clinician, and a lack of standard care (Reimann et al., 2007). A significant factor in the challenge of prompt diagnosis is BEB presentation that varies, which can be confusing to clinicians who are not familiar with the condition (Fung et al., 2013). Its rareness also impedes clinical recognition by primary care professionals, preventing patients from gaining access to care and treatment. However, there is also a fundamental lack of BEB knowledge, the impact of the condition on daily life, and how care and treatment are organised in the healthcare environment (Valls-Sole & DeFazio, 2016). A lack of knowledge among healthcare professionals has also been shown to relate to diagnostic mistakes and treatment delays (Kole & Faurisson, 2009; Nutt & Limb, 2011).

Another area found to be deficient is a lack of patient-clinician rapport, which aids communication and patient centred care, which is essential for the promotion of healing and reduction in suffering (Epstein and Street, 2007). People with BEB experience worsening disease over the undiagnosed period of their disease. Shepherd, Tattersall, and Butow (2008) and Dean and Street (2014) have advocated building a trusting relationship to explore the complexity of a disease condition, alleviate anxiety and distress, and enhance patients’ involvement in decisions about their care.

However, the time for a patient-GP appointment is now limited and has been recognised as a possible factor related to delays and misdiagnosis. The insufficiency of time was demonstrated in a survey of 1,000 GPs where a referral did not occur during the patients' first presentation (Burgess et al.,1998; Kantola et al., 2001). Consequently, a reduced appointment duration equals limited consultation time to explore symptomatology, especially for rare diseases, and predisposes patients to delays, misdiagnosis, and frequent re-attendance. Therefore, misdiagnosis causes a loss of trust and suffering, traumatises the patient, and affects future interactions and perceptions of care (Kostopoulou et al., 2008).

Generally, there is a lack of public awareness of rare conditions and health information to highlight these conditions (Dodge & Dion, 2011; Wastfelt et al., 2006) and reduce the psychological challenges delays and misdiagnosis present (Garrino et al., 2015). However, a clinician maybe able to reduce anxiety by being open and honest of diagnostic uncertainty as recommended by the General Medical Council (GMC, 2020). Importantly, more information on rare conditions such as BEB can minimise anxiety, stigma and eradicating discrimination. Research studies suggest that failures in the diagnostic process, in general, result in harm (Berner, 2008; Wachter, 2010).

The European Commission (2017) reported that 27 to 36 million people within its member states lacked efficient treatment and accurate diagnosis for most rare diseases, which represents an enormous unmet medical need and a significant challenge for public health. Qualitative assessment of the patient experience has become essential for assessing the quality of care. It allows healthcare practitioners to gain a deeper understanding by evaluating conditions to improve the treatment, care, and service delivery (Wressle et al., 2006). However, globally, the management of BEB differs under various healthcare systems, as do local factors of cultural impact on patients. The European Parliament and the Council (1999) legislation aims to ensure that lives of people suffering life-threatening or chronically debilitating rare diseases are improved in member states and across Europe.

1.3 National Context

The UK undertook a nationwide consultation toward a UK Strategy for Rare Diseases (DOH, 2013) with approval from health ministers in Scotland, Wales, England, and Northern Ireland. This was an extremely informative exercise to assess whether the NHS was well led with providers and commissioners collaborating in partnership with and for people with a rare disease in the community. The five domains assessed were; empowerment, identification and prevention, diagnosis and early intervention, co-ordination and care, and the role of research.

The UK consultation report (DOH, 2013) demonstrated a lack of accurate, reliable, and up-to-date information vital to making informed choices about their condition. Some 70% of patients do not receive sufficient information. One of the most salient findings was the patient experience of difficulties persuading the medical professionals to believe their symptoms when initially written off as ‘psychological’ or ‘neurotic’.

Only 21% of people received sufficient social support, while 66% of respondents stated their rare disease affected their caring responsibilities and ability to hold paid employment. Similarly, 56% of respondents indicated that their rare condition affected their education and many patients felt isolated. Notably, the ‘patient voice’ was recognised as an essential element, with 48% interested in planning services. A Kings

Fund-commissioned report (Foot, Naylor, and Imison 2010) offered advice on improving care, communication, and greater collaboration between primary and specialist care to prevent delayed referral.

In addition, the UK strategy recommendation seeks to configure the healthcare services to deliver coordinated and expert care to support the needs of patients with rare diseases (Evangelista et al., 2016). The UK Strategies for Rare Diseases 2020 updated its implementation plan for England and the Department of Health and Social Care (DHSC, 2018) and is steadfast to achieving 51 commitments. Specifically, those relevant to people with BEB are improving awareness amongst service providers and others on the effects the condition can have on family, social relationships, and work.

The relevant areas to be addressed are empowering those affected by rare diseases diagnosis by educating professionals, early intervention, co-ordination of care through shared information between GPs, hospital trusts, and specialist hospitals, and increasing the role of research for rare diseases. However, the provision of local care has failed to match the government strategy.

1.4 Local Delivery of Care

The outpatient clinic has both a multidisciplinary team clinic and an expert practitioner stand-alone clinic that provides care and treatment to patients. Expert practitioners provide care and treatment to patients in the BEB clinic, and they belong to a unique group of specialist nurses who provide care to patients with eye dystonia. Although it is difficult to confirm an accurate number of specialist nurses treating dystonia, there are 27 specialist nurses registered on the British Neurotoxin Network. The specialist role is particularly insightful and provides a local understanding of the issues affecting patients UK-wide and their views on how care is organised and delivered.

Although patients become the experts of their condition, educating the medical profession of their treatment duration patient-centred care is hampered by the organisational inability to provide the preferred appointment due to insufficient capacity consistently. Various studies have demonstrated BEB specialist outpatient clinics provide three-monthly cyclical primary recommended injections of BTX to patients (Hallett et al., 2013; Simpson et al., 2008; Jinnah et al., 2013). However, individualised care is lacking as BEB patients often report the treatment wanes before their planned appointment.

Botulinum toxin studies have also shown that the injection only offers temporary relief, with varied treatment duration and efficacy, but it is essential to alleviating distressing symptoms (Colosimo, Tiple, & Berardelli, 2012). This has also been evident in a local randomised control trial which showed untimely BTX treatment and care within outpatient settings means that 70% of patients face a recurrence of symptoms two weeks or more before their scheduled injection (Wickwar et al., 2016).

A particularly distressing report is that untreated BEB is severely disabling and creates anxiety (Jankovic and Orman, 1987; Albanese et al., 2013). It demonstrates that BEB can lead to functional blindness (Jankovic and Orman, 1987; Grandas et al., 1988; Hallet, 2002; Hellman and Torres-Russotto, 2015), which impacts on everyday quality of life, and the emotional and psychological well-being of those affected (Valls-Sole & Defazio, 2016). The impact of BEB on QoL is explored in the systematic review in chapter two.

1.5 The Researcher’s Position and the Context of the Research

From the researcher’s perspective, some patients emotional and psychological well-being impairs their ability to seek treatment immediately. This also affects the efficiency of the service to respond to the needs of patients. In addition, equity to access care is often questioned as only two out of 24 satellite sites offer treatment and care to BEB patients. The geographical differences create a financial burden to patients who face significant fares to travel a long distance to the main site. The service also lacks a patient support group to reduce feelings of isolation.

The chronicity of the condition requires patients with BEB to frequent the service for repeated treatment to avoid acute relapse and adapt to the disease to minimise its physical and psychological impact. The treatment of long-term conditions requires the collaboration of care and a partnership approach. Therefore, understanding the patient-specific duration of BTX treatment enables discussion and agreement on a tailored treatment schedule. However, care co-ordination is hampered if the treatment appointment is poorly timed or the medication prematurely wanes.

Giving BEB patients a more influential voice can readdress local care and inject optimum quality in care. It is essential that BEB patients actively participate in the continuation of changing the previous status quo to ensure that care is aligned with their wishes. This study adds contextual understanding to the local care experienced by patients.

The author and nurse researcher can provide contextual knowledge to this qualitative study. Notably, the researcher can draw upon understanding the physical and psychosocial challenges BEB patients face, their experience, behaviours, interpersonal interactions, and insider knowledge to authenticate the findings. Of importance, the researcher’s perspective is made explicit through the possession of strong subjective values as a nurse and specialist practitioner: holistic care, person-centred treatment, and evidence-based research.

The researcher’s main concerns are with human interactions and emotional elements, such as thoughts, feelings, and perceptions of BEB, care, and treatment in the clinic rather than cause and effect, facts, and measurable findings. The researcher’s ideology is to provide a clear representation of the inductive approach by highlighting the patient voice and respecting the importance of restricting oneself from making any assertions that are not based on valid intuition or sound reflection to create clarity and depth. In addition, the author understands the local healthcare context and understands how access to care and treatment can be perceived.

In contrast, access to healthcare is central to the performance of a health organisation and is often assessed as a measurement tool for quality (Shengelia et al., 2003; Penchansky and Thomas, 1981). It is crucial to understand and remove the barriers to obtaining healthcare resources (Frenk, 1992). Significantly, it is also beneficial to understand how such barriers can impact the experiences of long-term service users, at both the level of the health system and the population (Levesque et al., 2013). Experiential information within specialist care suggests that some ophthalmologists lack the knowledge associated with the level of visual disability on daily task performed by patient (Nelson et al., 1999).

Importantly, there is a moral dimension to untimely care in that it fails to consider the emotional health, well-being, dignity, independence, and social relationships of patients, which all require understanding. Furthermore, the patient experience has become essential for assessing the quality of care. Patient-centeredness, along with safety, effectiveness, equality, and timeliness in care, has been identified as fundamental to healthcare (Van der Eijk et al., 2015).

In addition, qualitative assessment of patient experience helps evaluate the quality of care, allowing healthcare practitioners to understand the condition better, to improve the treatment, care, and service delivery (Wressle et al., 2006). Therefore, this research sought to address the gap between policy, theory, and practice. It directly responds to the need to provide patients with a more influential voice in their care.

1.5.1 Knowledge gaps

An overview of the literature review and examination of the quality of care available globally, nationally, and within the local BEB specialist clinics identified three knowledge gaps:

1) Limited evidence about the lived experience of BEB
2) How the ability to access timely care during an acute episode of BEB impacts the patient experience; and
3) Limited understanding of how services should be configured to be responsive to expressed needs.

1.5.2 Study Aim

This study aims to explore the impact of BEB outpatient treatment and care on patients’ lives.

1.5.3 Study Objectives

- To gain a deeper understanding of the lived experience of BEB;
- To determine the impact of untimely care on BEB patient experiences; and
- To inform service reconfiguration to respond to the needs of BEB patients.

Conclusion

The chapter provided an overview of BEB, its definition, epidemiology, pathophysiology, risk factors, dystonia classification, symptoms, diagnosis, treatment, mode of action, side effects and complications. Several factors have been shown to be associated with people with the rare disease BEB. These include poor quality care and treatment delays, which impact both physical and psychological health. It provided national and local context to the care of people with BEB. Importantly, the chapter acknowledges the researcher's position and the close relationship shared with patients which closely interweaves thoughts and emotions. It also recognises the research gaps; very limited knowledge of the lived experience of BEB, a lack of knowledge on how access to timely care during an acute episode impacts the patient experience and how services should be configured to be responsive to the expressed needs of patients. The literature also recognised the need for patient empowerment and the gap between policy, theory, and local practice. Since BEB literature is largely quantitative, systematic reviews of current literature and patient narratives provide greater understanding of the impact of the condition on QoL.

Chapter 2: Quality of Life Assessment in People with Benign Essential Blepharospasm: A Systematic Review

Abstract

This chapter reports a systematic review of the QoL literature and explores the experiences of people with BEB care and treatment, the condition's impact on QoL and addressed the goals, expectations, standards, and concerns of people with BEB about the impact of BEB on daily life, care and treatment. Despite two previous QoL systematic reviews on dystonia only, ten studies on BEB were included in the reviews (Kuyper et al., 2011; Girach et al., 2019). There is still limited knowledge on what determines poor quality of life in people with BEB. Non-motor symptomology was examined as a primary QoL determinant for BEB (Kuyper et al., 2011). The second review (Girach et al 2019) focused on disease duration, gender, cognitive impairments, and psychiatric problems and the effect of treatment on QoL. This chapter focuses on gender, duration and worsening of the disease, aggravating and relieving factors, visual and functional ability, depression and anxiety, treatment belief and illness perceptions and delays and waning treatment.

2.1 Quality of Life

The World Health Organisation (WHO, 2012) defines QoL as an individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards, and concerns. A recent study by (Lawes-Wickwar et al., 2021) has demonstrated that people with BEB rate their QoL according to how it affects their daily living activities and perceived stigma.

Although previous research has highlighted potential factors impacting on QoL in BEB. A wealth of clinical and psychosocial factors example pain, emotional state, social and family life were also considered profound and associated with appearance concerns, emotional representations, perceived negative consequences of the condition, mood, and dose of BTX. However, factors that may contribute to QoL which have not received attention much attention were explored through an examination of; what aggravates the condition, the effects of motor symptoms example, a periocular spasm that disturb vision and facial spasms, which causes embarrassment, and the non-motor symptoms of psychiatric, mild cognitive, and sensory disturbances and BEB treatment on QoL (Defazio et al., 2017).

2.2 Review Objective

To explore the experiences of people with BEB treatment and care, and the condition's impact on QoL.

2.2.1 Types of Participants

People living with and receiving treatment and care for the condition BEB was included in this review.

2.2.2 Phenomena of Interest

The review considered publications that described the experience of living with BEB and who were seeking or receiving care and treatment.

2.2.3 Inclusion Criteria

Selected studies included was limited to those which considered the care and treatment of people with BEB.

2.2.4 Exclusion Criteria

The present study excluded papers which did not include BEB. Studies without the main purpose of assessing BEB quality of life, treatment and care were excluded.

2.2.5 Types of Studies

This review considered case control, cohort and cross-sectional studies. Although quasi experimental studies were considered no QoL study was identified for inclusion in the selection process.

2.2.6 Search Strategy

A systematic rolling search of the literature was set up on 20th January 2017, using MEDLINE, Embase, Greenfile, PsycINFO, CINAHL Complete, and Google Scholar without a date range to ensure all QoL studies were included. The key concepts, free text search and the Medical Subject Headings (MeSH) terms were used wherever possible. Alternative terms were used to expand the search. Boolean operators “OR” and “AND” were used to combine searches to add further detail (see search strategy in Appendix 3).

2.2.7 Assessment of Methodological Quality

The studies retrieved was assessed by two independent reviewers for methodological validity by using the standardized critical appraisal instrument for case control, cross-sectional and cohort studies (see Appendix 4) from the Joanna Briggs Institute. There was no disagreement between the reviewers. The care and treatment reported were generally multidimensional approaches to assessing the impact of BEB on QoL, and therefore a single intervention was not possible. The methodological quality according to the relevant JBI critical appraisal tool for all papers and the level of effectiveness (Appendix 5A) was documented in the summary of findings table in the (Appendix 5B).

2.2.8 Data Extraction

The data was extracted by using JBI extraction tool for case control, cohort and cross-sectional study see (Appendix 4). The data extracted included specific details about the methodology, method, phenomena of interest, setting, geographical location, cultural content, participant, data analysis and the authors' conclusion.

2.2.9 Data Synthesis

Utilising the JBI critical appraisal tools the findings of studies were pooled and reported in a narrative manner. The findings of each study were rated in according to its level of effectiveness. The study characteristics and results were described and summarised narratively. Selected studies, type of participants, assessment tool and main findings were also summarized.

2.3 Results

The search yielded a provisional sample of 6285 citations. The titles once duplicates were removed. Those that were not relevant, 2691, were eliminated. The full text of 60 papers were reviewed in their entirety to determine eligibility. A total of 41 papers were excluded as the main purpose of the study to assess BEB QoL. The JBI critical appraisal tools was employed see (Appendix 4). A level of effectiveness (Appendix 5A) was assigned to each study included in the summary of QoL findings (Appendix 5B). Excluded studies could also be found at the end of the summary table. A total of 19 studies met the inclusion criteria for this QoL study see (PRISMA diagram p. 38). A total of 1077 persons with BEB, with sample sizes from 9 to 159 was included in the study which showed people with BEB have poorer quality of life than people without the condition.

Numerous measurement tools were employed; Quality of Life in relation to vision was assessment with the 25 items National Eye Institute Visual Function (NEI-VFQ-25) and or the Ocular Surface Disease Index (OSDI). The severity of spasms was assessed with the instrument; Blepharospasm Severity Disability Index (BSDI), Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) and the Jankovic Rating Scale (JRS). The assessment of physical functioning, bodily pain, role limitations due to physical health problems, role limitations due to personal or emotional problems, emotional well-being, social functioning, energy/fatigue, and general health perceptions was undertaken with the instrument 36-Items short form (SF-36) health survey, the World Health Organisation Quality of Life Abbreviated Version (WHOQOL-BREF) contains 26 items which encompass physical, psychological, social relationships and environmental factors overall QoL and health satisfaction.

The Glasgow Benefit Industry (GBI) which assess post-intervention outcomes in three domains: ‘general’ functioning, ‘social support’, and ‘physical’ functioning. The EQ VAS records the patient’s self-rated health on a vertical visual analogue scale. The craniocervical dystonia question (CDQ-24) addressed QoL concerns through patient perception and concerns with cervical dystonia and BEB. The Illness Perception Questionnaire (IPQ-R) measured patient awareness in their condition. The Treatment Representation Inventory (TRI) The 27-item TRI assesses patients’ treatment cognitions, treatment value, treatment concerns, decision satisfaction and cure. The Hospital Anxiety Depression scale (HAD), Beck Depression Inventory and Thai Depression Inventory were utilised to assess the level of depression present. In addition, Self Esteem Scale (SES), self-assessment and other patient questionnaires were used. The tool utilised in the measurement of patient awareness was the Illness Perception Questionnaire (IPQ-R).

The Treatment Representation Inventory (TRI) The 27-item TRI assesses patients' treatment cognitions relating to four subscales; treatment value, treatment concerns, decision satisfaction and cure. The Hospital Anxiety Depression scale (HAD), Beck Depression Inventory and Thai Depression Inventory were utilised to assess the level of depression present. In addition, Self Esteem Scale (SES), self-assessment and other patient questionnaires were used. **Conclusion**: In the evaluation of QOL, all instrument utilised concluded people with the condition BEB had poorer quality of life than healthier individuals. The various tools employed complemented each other and aid in the address of all aspects of QoL.

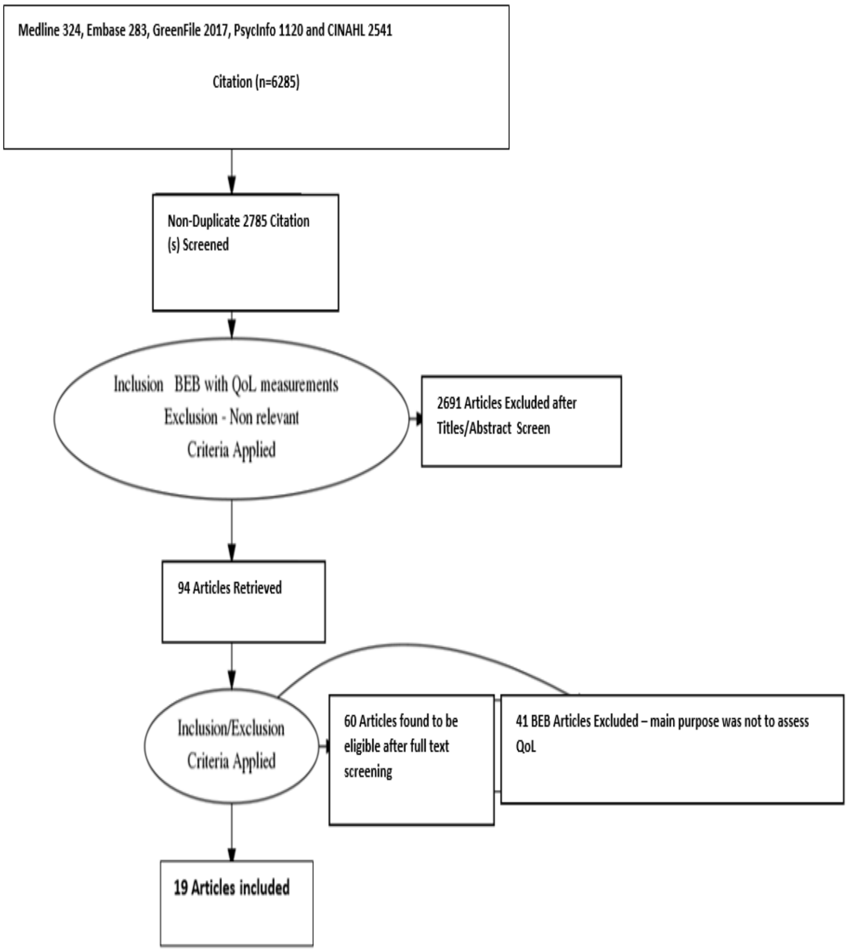


Figure 5: PRISMA Flow Diagram

2.3.1 Narrative QoL Findings of people with BEB

This review explored gender, disease duration, aggravate and relieve BEB, its impact on vision and functional abilities, treatment, care, illness belief and perceptions and the effects of depression and anxiety on peoples QoL. The QoL studies included in the prism diagram (Figure: 5) are narratively described below:

Two systematic reviews were identified within the study design through the application of inclusion and exclusion criteria. Though the reviews were eventually excluded they were used as a source of reference to search for potential studies. Further BEB studies identified were identified through the establish search strategy (Appendix 3).

This study explored gender, disease duration, aggravating and relieving factors, vision and functional disability, treatment and care, the effects of depression and anxiety and patients' perception of BEB illness on their quality of life. The findings below were narratively reported.

2.3.2 Gender, Duration and Worsening Disease

Four studies support this determinant. The first study a longitudinal observational retrospective study (Muller et al., 2002) which utilised SF-36 and depression by the BDI to compare HRQL and level of depression in BEB and cervical dystonia found both patients scored significantly worse in all eight SF-36 domains compared with an age-matched community. Predominantly BEB women experience worst disease than men based on scores of 5 out of 8 domains on the SF-36 form compared to men.

The second study by (Zhang et al., 2010) prospective survey over 3.5 years also assessed severity and the resulting disability using the BFMDRS and SF-36 to measure HRQL. The findings correlated worsening disease in females to their more sensitive nature to facial disfigurement, depression, anxiety, and functional disability. However, Muller et al. (2002) remain the only study to associate a longer duration of BEB with improved QoL due to the adaptive nature of patients to a long-term condition.

The third study (Tang et al., 2018) prospective comparison evaluated 87 patients' QoL, self-esteem, and ocular surface. The study used the WHOQOL-BREF, the SES, and OSDI before treatment on the date of enrolment and again after four weeks of treatment with BXT. Improvements were found in all areas after treatment. The study suggested BEB improvement over time may also be related to the introduction of BTX treatment which reduces spasmodic symptoms, improves vision, functional disability, self-esteem, and overall improves QoL.

The fourth study included a case comparison study by (Streitova and Bares, 2014), utilised a patient self-assessment questionnaire to examine nine patients with BEB and eighteen with hemifacial spasms over a 15–20-years duration. The study found BTX was safe as a long-term therapy and recommended the early introduction of BTX for the benefit of patients and families. It would improve social interaction and leads to less depression which would improve quality of life.

2.3.3 Aggravating and Relieving Factors

One study prospective observational study (Irfan, 2018) was located which provided evidence that stressful events have preceded BEB (Irfan, 2018). A history of acute triggering event causing anxiety or depression was present in 61 cases (96.8%). Secondary triggers causing blepharospasm eye conditions includes meibomian gland dysfunction in 100% cases, dry eyes in 85.71%, chronic Trachoma in 14.28%, concretions in 26.98%, trichiasis in 4.76%, and chronic blepharitis in 12.69%. At least 2 to 3 triggering factors were noted an all cases. An extreme visual and functional disability was present in all cases (100%). The findings of (Irfan, 2018) study supports the determinant depression and anxiety, visual and function ability of people with BEB reduces QoL.

2.3.4 Visual and Functional Ability

Two QoL studies was found to have assessed visual and functional ability as it pertains to people with BEB. Reimer et al., (2005) case control study employed the SF36, the NEI-VFQ and the BDSI demonstrated that HRQL measures were significantly worse in BEB than healthy controls. Although spasmodic symptoms improved after treatment there was still minor functional limitation.

Hall et al., (2006) cross sectional survey which utilised 25-item NEI-VFQ-25 highlighted people with BEB experience ocular pain, difficulties driving distance and undertaking various activities. The study provided evidence of significant reduction in vision targeted QoL in people with BEB in comparison to those without the condition. Both studies were retrospective in nature and concluded people with BEB experience functional limitations and are prone to depression and anxiety.

2.3.5 Depression and Anxiety

Psychiatric symptoms: anxiety and depression have been associated with BEB. Previously mentioned studies Muller et al., 2002; Zhang et al., 2010; Hall et al., 2006; Irfan, 2018; Reimer et al., 2005 have all provided evidence to support the impact of depression and anxiety on BEB. In addition, (Degirmenci et al., 2013) cohort control study employed HAD scale and SF-36 to assess the quality of life. BEB was found to be a key risk factor for anxiety and depression, which worsens quality of life. Although the true origin of depression and anxiety is unknown, some evidence suggests it may be related to how treatment and care have been organised (Hall et al., 2006).

2.3.6 The Effects of BTX Treatment on QoL

Numerous studies have assessed the effects of treatment on QoL. Tucha et al., (2001) case observational study utilised the CDQ24 to evaluate the clinical, demographic, and psychological influences on quality of life. Substantial correlation was found between appearance concerns, emotional representation, and perceived negative repercussions. The study suggest BEB improvement over time may be connected to the start of BTX medication, which lowers spasmodic symptoms, improves eyesight, functional impairment, self-esteem, and overall QoL.

A prospective cohort case control study (Setthawatcharawanich et al., 2011) assessed the impact of treatment BTX on HFS and BEB QoL during long-term treatment with botulinum toxin injections. The study support cognitive impairment in BEB in people with higher educational levels with more knowledge of the condition had better physical and mental health and overall better QoL. The study reviewed 53 patients with hemifacial spasm (HFS) and 32 patients with BEB to assess the condition’s impact on QoL. However, clinical improvement with BTX and physical and mental health were affected in both conditions and overall QoL. Depression and peak improvement after BTX injection were also associated with QoL. In addition, BEB patients were more affected by mobility and activities of daily living than HFS. Though a lack of cognitive processing may account for the educational scores.

The second prospective observational cohort study by (Weiss et al., 2018) assessed disease progression in terms of QoL. The prospective observation cohort study assessed motor symptom improvement, life satisfaction, depressive symptoms, pain and sleep quality. Patients were assessed at the end of a regular three-month period from last injection (Timepoint1) and four weeks after the re-injection of BTX

(Timepoint2). On a group level there was no improvement in HRQL. People who were classed RESPONDERS showed higher impairment in EQ-5D-5L, EQ-VAS, and Beck's Depression Inventory compared to the UNCHANGED (unlike WORSE). Those WORSE showed higher impairment in life satisfaction 'movement disorders' domain.

The third prospective open labelled cohort study by (Hilker et al., 2001) The HRQL was assessed using the EuroQol (EQ-5D) and the short form 36 health survey questionnaire (SF-36). The data confirm that BTX can induce a significant, but temporary amelioration of several aspects of HRQL and furthers the agreement that its benefit out weights its high costs for these severely affected patients.

While a case comparison study by (Streitova & Bares, 2014) use a self-assessment questionnaire a patient questionnaire to evaluate the influence of the treatment on QoL and found BXT to be an effective and safe long-term treatment of these facial spasms. However, the side effects by double vision or eyelid ptosis was frequent. All patients believed that the treatment was safe and effective with a positive impact on their quality of life, especially in social communication. Similarly, (MacAndie & Kemp, 2004) cross-sectional study to determine the impact of treatment on QoL in BEB utilising the Instrument GBI, postal questionnaire. This study demonstrates significant quality of life benefit BTX and justified it continued use. In addition, another cross-sectional study (Lee et al., 2013) reported BTX treatment for BEB to be associated with significant patient-reported improvements in quality-of-life (GBI) and functional ability (BSDI) with a strong positive correlation between both scores.

A prospective comparison study (Tang et al., 2018) Treatment-BTX, QoL, self-esteem, and ocular surface assessed with the WHOQOL-BREF, SES, and OSDI before and after treatment. Improvements were found in all areas after treatment. The study suggested BEB improvement over time may also be related to the introduction of BTX treatment which reduces spasmodic symptoms, improves vision, functional disability, self-esteem, and overall improves QoL.

Leplow et al., 2017 case comparison study utilised a survey to determine BEB treatment satisfaction and emotional responses to a life with a disabling condition. It found BEB satisfaction worsened significantly within the treatment cycle. Despite good overall treatment effects, patients from both groups perceived marked persistence of motor symptoms, restrictions of everyday life functions, and reduced quality of life. Functional amelioration of motor symptoms and emotional well-being were only moderately correlated. About 22% of patients from both groups reported mental disorders or emotional disturbances prior to the onset of treatment.

To assess QoL after BTX treatment, Biuk et al., 2013 prospective study explored whether BTX QoL in BEB. A total of 37 patients with the condition fill the WHOQOL-BREF questionnaire on the day of treatment and 6 weeks later. The study found that participants enjoyed life more (p=0.002), felt physically safer in everyday life (p=0.002), had enough energy in/for everyday life (p= 0.001) and performed everyday activities (p=0.005). In general, patients were more satisfied after treatment and less prone to negative emotions such as bad mood, desperation, anxiety, depression.

A previous case observation study by (Tucha et al., 2001) employed a QoL patient questionnaire. It investigated BEB level of patient satisfaction, in patients who received BTX for 1 year. The study found although injection BXT in BEB is appreciated by the patients, but their well-being is affected by fears and depression.

However (Timlin et al., 2021), retrospective case reviewed utilised the BDSI and JRS instrument to assess the severity and frequency of BEB symptoms provided data on the impact of periocular surgery other than orbicularis stripping. The study found periocular surgery significantly reduce BEB in 83% but 17% experience worsening symptoms.

2.3.7 Treatment Belief and Illness Perceptions

Two cross-sectional studies undertaken by (Lawes-Wickwar et al., 2020; 2021) was included in the review. The first study (Lawes-Wickwar et al., 2020) assessed beliefs about BEB and HFS and treatments utilising IPQ-R, TRI, BSDI and CDQ24. Severity and frequency of spasms were recorded using JRS. The study found participants associated BEB and HFS with a chronic timeline and believed their condition was not caused by psychological factors, risks, or their personality or lifestyle. Participants also demonstrated decision satisfaction related to treatment but had some treatment concerns, emotional well-being, activities of daily living, social/family life, pain, and stigma and overall poorer quality of life.

The second study (Wickwar et al., 2021) evaluated QoL to people’s treatment belief and illness perception in a cross-sectional study that involved 65 patients with BEB. The instrument utilised was an IPQ-R, TRI, the BSDI and CDQ 24, and the JRS to record severity and frequency of spasms. It found people with BEB and HFS reported a detrimental impact on treatment, perception of stigma, mood, and socialising, which correlate with reduced QoL.

2.3.8 Delays and Waning Treatment

A prospective, open labelled cohort study which employed EQ-5D and SF-36 instrument found dissatisfaction, social disability, and impairment in health-related quality of life can be improved with access to treatment. Moreover, significant restoration of self-esteem and mental health means the high cost of BTX should be acceptable (Hilker et al., 2001).

Dissatisfaction was also significant, and distress occurred when BTX wore off before the cyclical three-month scheduled treatment (Wickwar et al., 2021). Overall, the instrument CDQ24 treatment utilised showed satisfaction was recorded as high with BTX but declined before the end of the cycle. The study recommended flexible, individualized treatment intervals to improve treatment satisfaction and outcomes.

2.3.9 Discussion

The objective of this review was to determine the impact of treatment and care on people with BEB. The study reviewed a total of 19 studies inclusive of six cross-sectional studies, six cohort studies and six case control studies. The studies found 96.8% people with BEB had a history of acute triggering event causing anxiety or depression and meibomian gland disease or eyelid inflammation in 100% of people. The latter explains the ocular pain experienced in BEB. In addition, people with BEB have difficulties driving for distance, functional limitation and undertaking various activities and overall poor vision-targeted and HRQL. Although people with BEB was found to be prone to depression and anxiety, the origin of the psychiatric symptoms remains unknown. There is evidence to suggest depression and anxiety may be related to how treatment and care is organised. The study also found dissatisfaction and distress occurred when BTX wore off before the cyclical three-month scheduled treatment and QoL only improves with treatment.

Botulinum toxin was found to temporary ameliorate persistence motor symptoms which restricts everyday life functions and reduce QoL. There is evidence to demonstrate BTX to be a safe as a long-term therapy and people with BEB would benefit from early treatment to improve psychology, spasms, vision, functional ability, self-esteem, appearance issue and perceived negativity fear, depression, social interaction and QoL. However, the frequency of the side effects double vision and eyelid ptosis did not change the belief that BTX is a safe and effective treatment long term treatment. Though peak improvement in BEB and QoL was associated with BTX, periocular surgery was also found to significantly reduced BEB symptoms in 83% of people.

The included studies were reviewed by two reviewers. Two previous systematic reviews undertaken were not specific to BEB, although it encompassed all dystonia. This study in comparison to the other reviews undertook a comprehensive search using five databases, rather than examining PubMed alone. It examined more significant number of studies to provide the final analysis.

Limitations

Three studies did not utilise a standardised instrument to assess QoL.

2.4 Conclusion

This systematic review has reflected research on BEB QoL and increased interest in the advancement of understanding QoL and provided objective outcomes. The findings suggest that people with BEB have a reduced QoL than healthy controls. The reasons underpinning this vary not exclusively to the clinical characteristics of the BEB but other factors such gender, duration and worsening of the disease, aggravating and relieving factors, visual and functional ability, depression and anxiety, the effects of BTX on QoL, treatment belief and illness perception and delays and waning treatment. Inherently, the stigma attached to BEB required people to socially adapt, develop coping mechanisms and seek timely treatment by healthcare professionals to impact their QoL positively. The research highlights many factors that reduce QoL and the numerous studies available with in the field (see summary of findings in Appendix). Although routine BTX treatment will always be required to reduce QoL in people with BEB, the many rating scales fail to express the depth of patients' thoughts, feelings and emotions. However, understanding of BEB can assist in how the treatment and care are organised to achieve improve QoL.

The studies included in the review used multiple QoL measurements. However, outcome measures such as the SF-36 scale, and self-reported questionnaires have the potential for inherent response bias. However, even with multiple QoL measures employed, the study could be questioned on whether the QoL scales genuinely reflect the true nature of BEB. It highlighted the importance of multiple domains in the assessment of QoL in people with BEB but also the need for qualitative evidence to replicate the lived experience of BEB. Since no qualitative studies on BEB was located dystonia patient websites were search for narrative literature.

Chapter 3: Systematic Review of Patient Narratives

Abstract

This chapter reports the findings of a systematic review of narratives or stories from people living with BEB published in the non-peer-reviewed publication, websites, and blogs. The purpose of the review is to synthesize the best available qualitative evidence on the impact of BEB, outpatient treatment and care on the lives of patients. The review was conducted per JBI guidelines for the systematic review of text and opinion (McArthur et al., 2020). The synthesis aims to provide additional insights into the lived experience of BEB patients. The study was undertaken retrospective to analysis and presentation of the current research findings and further informs the discussion in Chapter 6.

3.1 Review Objective

To gain an insight into the experience of individuals with BEB through an examination of personal narratives or stories.

3.1.1 Inclusion Criteria

Textual and opinion reports from the UK, USA, Australia, and Canada were included in personal narratives or stories published in non-peer-reviewed literature, voluntary sector, and support group websites or blogs.

3.1.2 Exclusion Criteria

Reports without a clear conclusion were excluded.

3.1.3 Types of Participants

People living with BEB in the UK, USA, Australia, and Canada.

3.1.4 Phenomena of Interest

The review considered publications that described the experience of living with BEB and who were seeking or receiving care and treatment.

3.1.5 Types of Publication

This review considered text and expert opinion, expert patients' reflections, and personal reports or case studies in the absence of qualitative studies.

3.1.6 Search Strategy

The systematic rolling search of the literature was set up on 20th January 2017, using MEDLINE, Embase, GreenFile, PsycINFO, CINAHL Complete, and Google Scholar without a date range to ensure all QoL studies were included. The key concepts including patient experience, free text search and the Medical Subject Headings (MeSH) terms were used wherever possible. Alternative terms were used to expand the search. Boolean operators "OR" and "AND" were used to combine searches to add further detail above (see search strategy in Appendix 2). Additionally, all QoL study reference list was searched for qualitative literature on BEB treatment and care.

Eight reports were screened but were eliminated; one qualitative study abstract was found but full text could not be located, four systematic reviews and three QoL studies. Four dystonia societies websites were searched, UK – http://www.dystonia.org.uk; Australia – http://www.beb.org.au; Canada–https://www.dystoniacanada.org/;and USA – http://www.blepharospasm.org. A total of 25 text and opinion reports were located. All reports found had clear helpful conclusion and were included. A patient report provided details of the newspaper she read with a narrative account similar in nature to her experience. The *Washington Post* was subsequently searched for the article entitled; Medical mysteries: “I open my laptop, and my eyes snapped shut.” The article was located and included in this review (see appendix).

3.1.7 Assessment of Methodological Quality

A total of 25 textual and expert opinion reports included were peer-reviewed by other BEB patients and medical experts attached to the dystonia society. All narratives/ reports were selected for retrieval following an independent assessment by the researcher and supervisor before inclusion in the review using the inclusion and exclusion criteria.

Each narrative/report was then quality assessed using the standardised critical appraisal Narrative, Opinion, and Text Assessment and Review Instrument (JBI-NOTARI) (McArthur et al., 2020) (Appendix 4). Critical appraisal was essential to establish trustworthiness, in this context referring to the logic of the opinion and its ability to be convincing and to have authority as a source of opinion. A transparent process was used in appraising this textual data and focused on:

a) Assessing the opinion.
b) Identifying the credibility of the source of the opinion.
c) Establishing the motives of the underlying the opinion.
d) Locating alternative opinions that support or question the statement.

There were no disagreements between the two reviewers that required a third reviewer resolution.

3.2 Data Collection

Textual data was extracted from papers included in the review by the researcher, and a 20% sample was checked by the supervisor using the standardized data extraction tool from JBI-NOTARI (Appendix 4). Conclusions were transferred from each included paper to the data extraction instrument to minimize error, provide a historical record of how decisions were made to include conclusions, and become the basis for extraction and synthesis.

Each narrative was read and re-read to extract and state the author's meaning (s). This conclusion was supported by an illustrated text, taken from the report and entered into JBI-NOTARI. To categorize the validity of the conclusions drawn in the narrative, opinion, and text, one of three levels of credibility were assigned to each conclusion. Unequivocal refers to evidence that is beyond a reasonable doubt, including conclusions that are not open to challenge. "Credible evidence" is an opinion that appears reasonable given the current data presented in the paper, and "unsupported evidence" is when conclusions reached are not supported by the data. Each conclusion needs to have a level of credibility assigned to it during the data extraction phase to ensure that the overall findings of the qualitative study were credible.

The findings and illustrations from the data extraction were then assembled to develop a synthesis using a meta-aggregative approach (Lockwood et al., 2015). This involves the aggregation of like conclusions to generate a set of categorical statements to produce a single comprehensive set of synthesised findings that can inform practice.

3.2.1 Data Synthesis and Design

The meta-aggregation presented conclusions and categorised them based on similarity. The textual and opinion reports were collated using JBI's SUMARI program, a web-based software for preparing systematic reviews. Meta-aggregation was the chosen design approach to synthesising the qualitative evidence. The design aims to provide transparency, accountability, reliability, and a synthesised statement referred to as a "line of action" (Hannes and Lockwood, 2011), which is meant to inform the development of care and treatment within the context of the BEB clinic. The underlying premise of meta-aggregation is agnostic of the type of evidence the question requires (Munn et al., 2013; Pearson, 2004; Pearson, Jordon & Munn, 2012). It follows the principles of a systematic review while being sensitive to the research context. This approach provided an integrative review (Flemming, 2007) of the conclusion or summary of each category's experience.

The BEB summaries were graded according to the JBI level of meaningfulness. The summaries and conclusions in this study were then subjected to a meta-synthesis It was synthesised from the actual words of patients. Finally, single comprehensive guidance, derived from the synthesis, was created for evidence-based practice. JBI reviews of this nature are considered rigorous enough to provide a streamlined approach to evidence that can inform healthcare decision-makers (Khangura et al., 2012).

3.2.2 Results

Twenty-five reports were included in this review, and all were textual papers. All of the included papers presented the participants lived experience of BEB and seeking care and treatment for the condition in the UK, USA, Australia, or Canada. Most authors were people with BEB meaning they were expert participants in the healthcare environment. Refer to Appendix 4 for a summary of the included reports. Below is a breakdown of the 25 reports:

- Nine reports focused on Australia;
- two reports focused on Canada;
- Six reports focused on the UK; and
- Eight reports focused on the United States of America.

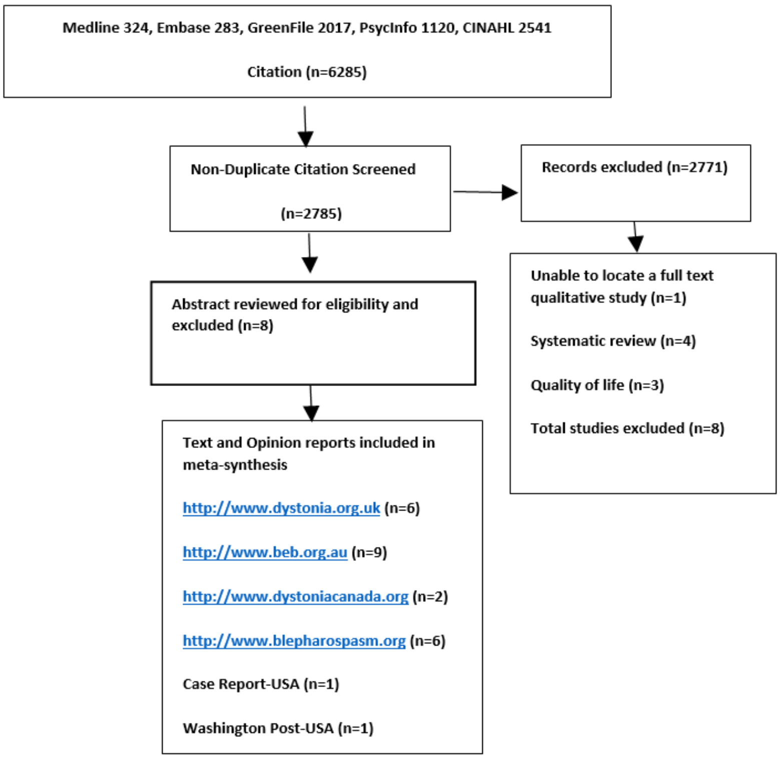


Figure 6: PRISMA Flow Diagram 2

3.2.3 Methodological Quality

The included papers were textual reports from many different dystonia websites, but all opinions were from patients with the lived experience of BEB care and treatment, therefore experts in the area, and deemed of sufficient methodological quality, according to the JBI-NOTARI critical appraisal tool seen in the summary of findings along with JBI assigned level of meaningfulness (Appendix 6B).

3.2.4 Findings of the review

From the 25 included papers, 14 conclusions were extracted and analysed. Conclusions were drawn from the papers and supported by illustrations from the publications. All conclusions were grouped into three categories based on similarity in meaning, and these were subjected to in-depth analysis and review to generate nine synthesised findings. The synthesised findings are as summarized below and then displayed in full tables, incorporating all conclusions and categories into the various syntheses.

A description of the categories was included in the synthesis results and is utilised to support the final synthesised findings. The various countries were not explicitly mentioned since the synthesised results were unvaried, even with differences in a local context. The conclusions presented in the tables have been assigned the following levels of credibility: U = unequivocal, C = credible, US = unsupported. In addition, a level of meaningfulness (Appendix 6) was assigned to each report.

The reported results are presented individually in synthesised findings, listing the categories and displaying each conclusion (and illustration) linked with the generated category and synthesised finding. A visual of the “NOTARI-View” table, which details the relationships between the conclusions, categories, and synthesised findings, is presented for each of the findings. Levels of credibility are also assigned to each conclusion.

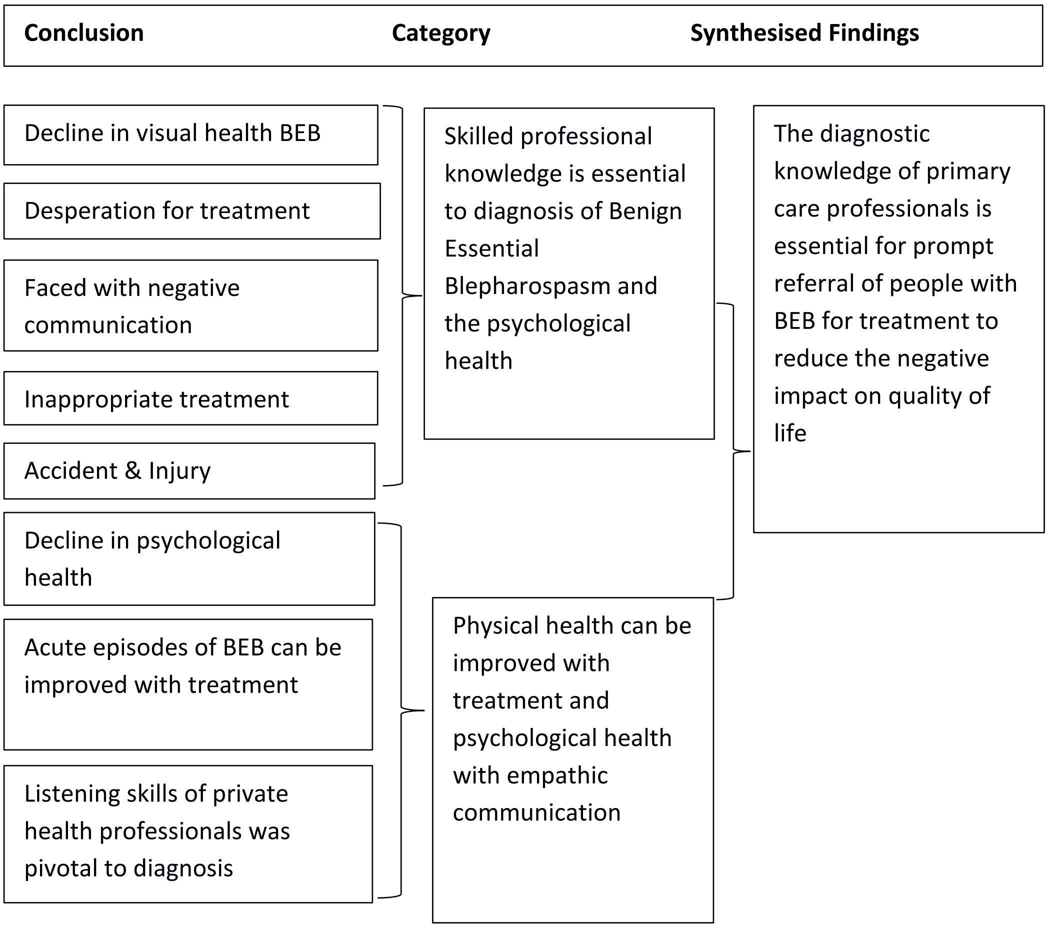
3.3 Synthesis Narrative Results

A total of 3 categories from 14 conclusions was obtained from 25 text and opinion papers, which were synthesised and interpreted to provide two constructs of BEB experiences see new diagram attached and completed summary of findings with excluded papers, which commenced from primary care and followed through to the specialist BEB clinic. Specifically, the syntheses are:

- The diagnostic knowledge of primary care professionals is essential for prompt referral of people with BEB for treatment to reduce the negative impact on QoL; and
- Specialist BEB services should improve access to treatment, which is essential for ameliorating physical symptoms and enhancing patient-clinician rapport, psychological health, and knowledge through shared experiences.

Synthesis 1: The diagnostic knowledge of primary care professionals is essential for prompt referral of people with BEB for treatment to reduce the negative impact on QoL

A total of one category from eight conclusions was developed and synthesised into the vital learning and skilled care essential for health professionals caring for in primary care to reduce the negative impact of the condition on QoL. The category in this group relates to valuing the care of people with BEB through early diagnosis to avoid visual and psychological impact.



Conclusion **Category** **Synthesised Findings**

- Decline in visual health BEB
- Desperation for treatment
- Faced with negative communication
- Inappropriate treatment
- Accident & Injury

Skilled professional knowledge is essential to diagnosis of Benign Essential Blepharospasm and the psychological health

- Decline in psychological health
- Acute episodes of BEB can be improved with treatment
- Listening skills of private health professionals was pivotal to diagnosis

Physical health can be improved with treatment and psychological health with empathic communication

The diagnostic knowledge of primary care professionals is essential for prompt referral of people with BEB for treatment to reduce the negative impact on quality of life

Figure 6: NOTARI-view of Synthesis 1

Conclusion 1: Countries and local services must accept that essential strategies are required to improve the desperate needs of people with BEB to be diagnosed and treated [unequivocal]

The challenge of seeking a diagnosis for BEB and numerous appointments caused people to experience and express *“suffering, desperation to receive an early diagnosis and subsequent early treatment”* (Lori, 2018, p. 5).

Conclusion 2: Healthcare professionals should avoid negative communication and adopt more excellent listening skills to improve their diagnostic abilities [credible]

Many GPs made judgment instead of adopting greater listening skills essential for diagnosis and referral: *"I then went to my doctor to talk to him about it. 'Oh you're stressed.' I emphatically told him I didn't have this problem because I was stressed, I was stressed because I had this problem."* (Case 7, BEB Org, p. 1)

Conclusion 3: Inappropriate treatment in primary care can be eliminated with improved access to specialist care [unequivocal]

People with BEB are often misdiagnosed and inappropriately treated rather than referred immediately to specialist services: *"They diagnosed me with myasthenia and started treating me with a powerful course of steroids."* (Smith, 2011, p. 21).

Conclusion 4: To reduce an acute episode of BEB where visual ability declines, healthcare professionals need to refer people with BEB to treatment [unequivocal]

A decline in visual ability synonymous with an acute episode of the condition impaired daily life and caused those affected to feel: *"Blind, but with perfect vision, That title accurately describes the first five years of my affliction: my vision remained normal, but I couldn't lift my eyelids to see."* (Needle, 2015, p. 568)

Conclusion 5: Acute episodes of BEB and visual decline cause people with BEB to experience accidents and injuries which negatively impact family life [credible]

The accidents and injuries experienced by people with BEB were considered traumatic and negatively impacted family life:

> *"In July 2016, my husband and I went to Phuket for a month holiday as usual, and he had to care for me. Three days before we were due to return home, I tripped over around the pool and fell heavily, snapping my right humerus bone in half. This was a traumatic time. Fortunately, the insurance covered the op and hospital, but I realised then how dangerous it was to have the problem of my eyes constantly blinking and closing shut."* (BEB Org, Case 6)

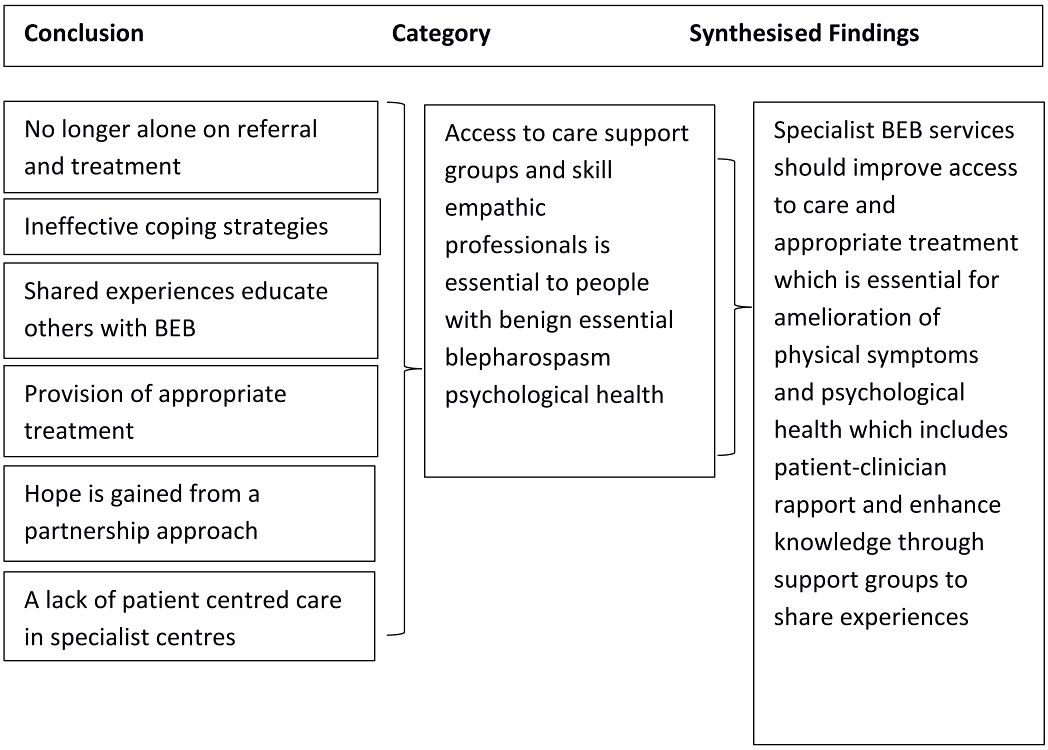
Conclusion 6: A decline in psychological health in BEB can be improved with more effective empathic communication from health professionals [credible]

The lack of empathic communication experienced during interactions with healthcare professionals created further difficulties in coping and a decline in psychological health: *"I was told just wait, it would go away. I was angry, hurt and confused. Why was no one helping me? I was beginning to sink into depression."* (Lori, 2018, p. 5)

Conclusion 7: Private healthcare professionals in countries with public funded system without time constraints were pivotal to diagnosis [credible]

In the absence of answers following repeated consultation within national healthcare systems, people with BEB consulted private healthcare professionals who were not constrained by time but were able to listen to and diagnose the condition: *"I had BUPA via my husband's work. Without private medical insurance I estimate it would have taken me around seven to eight years to find answers."* (Lownsborough, 2011, p. 19)

A widespread lack of BEB knowledge among GPs prevented patients from obtaining a timely and accurate diagnosis and referral to specialist services for diagnosis and treatment. Whether within the publicly funded NHS of the UK and Canada or partially privately funded by Medicare in Australia and USA, patients with BEB faced diagnostic and treatment challenges. Delayed diagnosis and treatment profoundly impacted physical and psychological consequences, predisposing people to negative communication, acute episodes of the condition, accidents, and injury, creating a desperate need for treatment and depression. Countries and local services must accept essential strategies to improve the diagnostic care to expedite treatment for people with BEB. In addition, the provision of skilled professionals in the treatment of the BEB offers an effective means of coping.



Conclusion	Category	Synthesised Findings
No longer alone on referral and treatment Ineffective coping strategies Shared experiences educate others with BEB Provision of appropriate treatment Hope is gained from a partnership approach A lack of patient centred care in specialist centres	Access to care support groups and skill empathic professionals is essential to people with benign essential blepharospasm psychological health	Specialist BEB services should improve access to care and appropriate treatment which is essential for amelioration of physical symptoms and psychological health which includes patient-clinician rapport and enhance knowledge through support groups to share experiences

Figure7: NOTARI-view of Synthesis 2

Synthesis 2: Specialist BEB services should improve access to care and treatment, which is essential for amelioration of physical symptoms and psychological health and enhance knowledge through patient-clinician rapport and support groups shared experiences

A total of one category from five conclusions were developed and synthesised into the critical learning about BEB. Health professionals and service providers can provide patients with timely access and skilled care, and psychological support. The category relates to valuing BEB patients through timely access to treatment by skilled health professionals to ameliorate an acute episode of the condition and collaborate with specialists in the treatment and management of BEB to improve psychological health. In addition, through shared experiences and counselling, the overall quality of care can be improved. The evidence for this appears to be unequivocal**.**

Conclusion 1: It is essential to reverse the depressive mind-set experience during isolation with ongoing psychological support [unequivocal]

The ongoing psychological counselling offered by specialist services on referral enables people with BEB to become aware of the importance of talking therapy and that *“being alone, without others to talk to about my dystonia, was detrimental.”* (Kranz, 2015, p. 1)

Conclusion 2: Provision of skilled treatment by BEB health professionals offers a more effective means of coping than temporary sensory tricks [unequivocal]

Sensory tricks or strategies used by people with BEB to trick the brain only offered a temporary distraction technique. For example, *“chattering away, even if no one was on the other end. But after several weeks, Ecola noticed, her coping strategies were becoming less successful, and a neurologist was sought.”* (Boodman, 2012, p. 5)

Conclusion 3: Access to support groups is crucial for shared experiences, transfer of knowledge, and empathetic understanding [unequivocal]

Sharing experiences within support groups provide education and offer comfort:

> *"I wanted to share my experience with BEB over the past year or so, in the hope that it may offer others some encouragement and a useful way to deal with this disease."* (Sean, BEBFR, Bulletin Board, 2011).

Conclusion 4: Open communication from knowledgeable health professionals gave hope and improved partnerships between patients and clinicians [unequivocal]

There was understanding, hope, and willingness to be patient during the trial period of BTX when clinicians openly communicated, collaborated, and valued patient feedback:

> *"The doctor explained that I'm still in the very early stages of treatment, and it may take a while to get the dosage right. You need to work with your doctor – trial and error – to find an optimal product and dose that suits you, so you need to be patient."* (BEB Case 4, Blepharospasm Australia, p. 1)

Conclusion 5: Access to timely regular treatment is a quality indicator for the delivery of patient-centred care [unequivocal]

People with BEB highly valued patient-centred care, a quality indicator measured by timely access to regular treatment, and would change service provider to achieve the same as described:

> *"This treatment with irregular injections went on for years. It was not until I got on the internet relatively recently and started researching the subject that I discovered there were now more doctors using Botox. I changed doctors twice and got more regular treatment. I finally found a doctor that would treat me when I needed it and not to some one-size-fits-all schedule. I had the injections every eight weeks."* (BEB org, Case 1)

Access issues incorporate many varying factors, from the number of available specialists, clinics, holidays, to the availability of high-quality, trained health staff. Patients with BEB who get a referral to specialist care understood that psychological help was available, and they were no longer alone. The treatment episode offered BTX and an opportunity to discuss BEB symptoms and management approach. The treatment was also seen as a more effective means of coping than sensory tricks.

In addition, one-to-one counselling and support groups provided an accessible means of sharing experiences. Collaborating with clinicians gave BEB patients hope. However, a lack of access or patient-centred care caused BEB patients to change service providers to gain quality care.

Discussion

Spasmodic closure and a decline in vision, creates desperation. However, patients seeking treatment are faced with negative communication, inappropriate treatment, accidents and injury even as their psychological health declines. An acute episode of BEB which also affects psychological health can be improved with BTX treatment. Even though treatment is often delayed by a lack of diagnosis which is hampered by a lack of skill professional knowledge. Therefore, the diagnostic knowledge of primary care professionals is essential for prompt referral of people with BEB for treatment to reduce the negative impact on QoL. Referral to treatment often signals support as coping strategies are often ineffective. Hope is gained from a partnership approach with clinicians. However, a lack of patient centred care can be improved through access to support groups, empathic professionals and specialist BEB services which provide care and treatment to amelioration of physical symptoms and psychological health.

Conclusion

The metasynthesis provided a valuable tool and synthesised 25 text and opinion papers into 3 categories from 14 conclusions. It determined the deep meanings, experiences and perspectives participants shared. Though many previous studies have questioned whether anxiety and depression are causes or consequences of BEB the findings suggest it as a response to the daily limitations and unmet needs. The metasynthesis was undertaken retrospective to the current research results. However, this qualitative approach provided evidence which supports the diagnostic knowledge of primary care professionals is essential for prompt referral of people with BEB for treatment to reduce the negative impact on QoL. The evidence also supports the view that specialist BEB services should improve access to treatment and care, which is essential for ameliorating physical symptoms and enhancing patient-clinician rapport, psychological health and knowledge through shared experiences. This information provides the means to develop services, implement and evaluate health interventions.

Chapter 4: Methods

This chapter presents the rationale for the philosophical and theoretical underpinning of the research and the methodological choices and methods used to undertake the research programme. The chapter is divided into four sections. The first section focuses on the principles of the theoretical approach used in this study. The second section describes the methodology used and provides a rationale for why this was chosen. The third section describes the setting for the research and details of how the data were collected and analysed. The final section discusses the ethical considerations utilised in the study.

4.1 Theoretical Framework

This research explored the lived experiences of people with BEB and the impact of outpatient treatment and care on them. The theoretical framework was informed by a personal theoretical orientation and the need to gain a deeper understanding of a person with BEB's lived experience and the beliefs and values underpinning the researcher's clinical practice as a specialist practitioner in this field. As a specialist practitioner in the NHS, the care and treatment delivered are underpinned by a holistic and person-centred approach which comes from the discipline of nursing; this is further underpinned by the context of quality of care that drives service delivery in the NHS. While the research literature related to treatment and QoL in people with BEB, little is known about the emotional and social well-being of people with the condition (Tucha et al., 2001) and provides little in the way of insight into what it means to live with BEB.

All research is situated within a particular paradigm or 'world view' (Kuhn, 1962). In this study, the researcher's philosophical approach was also influenced by the disciplinary background and personal world view, reflecting an ontological and epistemological position and influencing the choice of methodology and research design. A relativist, interpretivist stance was selected as it provides the opportunity to add to the body of knowledge through the multiple realities of people with BEB and the perspective of the specialist practitioner. Interpretive Phenomenological Analysis (IPA) as a methodological approach fit with the study's objectives. It facilitates the ‘action’ that will need to be taken to deliver high-quality out-patient care that takes account of the physical, emotional, mental, and spiritual well-being of patients. The theoretical framework adopted within this study is summaries below in Figure 9.

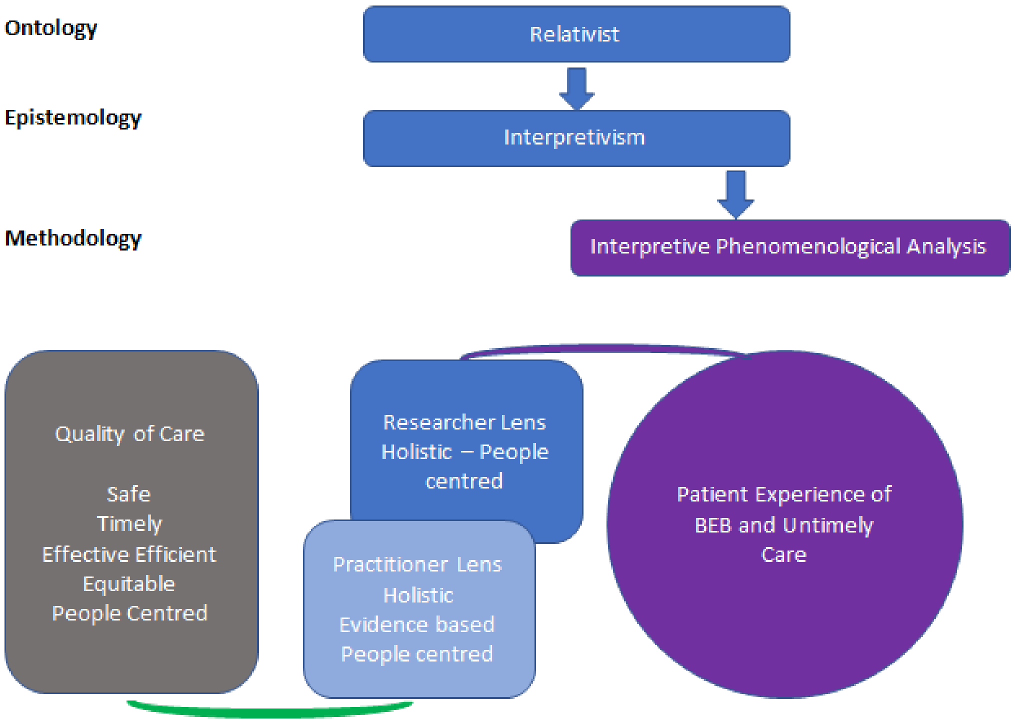


Figure 9: Theoretical Framework

4.2 Philosophical Perspectives Underpinning the Study

4.2.1 Ontology

Ontology is a branch of metaphysics that deals with the nature of being and reality (Guba & Lincoln, 1984; Punch, 1998). The relativist ontological stance of this study was informed by the exploratory research aim, question, elective contextual framework, NHS quality of care framework, and the researcher's world view. It followed deep introspection on the questions such as, "is there a singular or multiple versions of reality?", "What is the nature of reality for people with BEB or what constitutes reality?" and "Are there multiple realities of a lack of patient centred care and untimely care?" This approach addresses the research's main aim: to capture the divergent range of viewpoints and participants' perceptions of their lived BEB experiences.

The ontological stance acknowledged that "the researcher accepts multiple subjective realities" (Hugly and Sayward, 1987 p. 278). It recognises that the reality differs for each participant in this study (Crotty, 1998). Each study participant accessed care and treatment within both primary care and the specialist BEB clinic and framed their care and treatment experience about their reality. In this regard, ontology is human experiences and is socially constructed. Therefore, each participant has their perception of the environment based on previous experiences and interactions.

A relative ontological stance recognises the researcher as a co-creator in the meanings that emerged (Guba and Lincoln, 1989). In addition to collecting the data, the researcher must understand and contextualise the meanings of the multiple views BEB participants provide, reconstructing multiple realities. Whilst BEB patient-participants role was to collaborate and co-produce meanings to create a straightforward experience narrative.

It significantly influenced how the study was approached since a relativist ontology position believes reality is subjective and exists within our thoughts (Denzin and Lincoln, 2005). The objective form of ontology realism was considered but rejected. It assumes that the world contains truths and that knowledge can be reduced to discrete variables that can be tested (Creswell, 2009) and reality can be investigated to gain an objective reality (Crotty, 1998).

It created the rationale for rejecting objective realism as the current research question is wholly qualitative. The recommended solution to improve the quality of care for BEB patients was best answered through multiple views. It helped the researcher to understand multiple truths. A relativist ontological position fits with an interpretivist epistemology. Although ontology overlap with epistemology as identified by (Denzin, 1998; Hughes & Sharrock, 1997), the facts gathered on BEB belong to the ontological reality.

4.2.2 Epistemology

The knowledge gained on that reality is epistemology. The term epistemology describes the nature of the relationship between the knower and what can be known (Guba and Lincoln, 1998). Therefore, the reality was considered only knowable through socially constructed meaning – a truth that is not shared (Ritchie and Lewis, 2003). Interpretivists believe that reality is indirectly constructed based on individual interpretation and subjectivity: the world does not exist independently of our knowledge.

People interpret and make their meaning of events, which are distinctive and cannot be generalized. Therefore, the experiences of people with BEB were considered subjective as a result of their experience rather than based on truth or fact that can be tested or verified. In this study, BEB perception of the experience at the reception of care was explored to identify its meaning and how services could be informed to construct positive experiences.

The epistemological position provided the means to determine which kind of knowledge is legitimate and adequate to acquire and interpret the BEB participants' world. Gray (2013) sought to clarify that the epistemological choice helps a researcher recognise which design will be suited to their chosen ontological choice.

Interpretivism is rooted in the relativist ontological stance, which argues that truth and meaning do not exist in an external world but are created by interaction with the world. It, therefore, follows that, even with the same phenomena, people construct their meanings (Gray, 2013). Within an objective stance, it would be argued that reality exists externally to the researcher and must be investigated through a rigorous scientific inquiry process (Crotty, 1998).

In this study, the real world for participants is the specialist BEB clinic. Within the clinical setting, contextual knowledge can be gained. The philosopher Max Weber has been identified as the main contributor to the interpretive approach for his concept 'verstehen', which means gaining contextual knowledge (Holloway, 1997). Knowledge was based on the assumption of the nature of the relationship between the researcher and participants. Although Guba and Lincoln (1982, 1989) alluded to criticism that knowledge can be obtained regardless of conscious reality.

This study accepts even scientific knowledge has boundaries and limitations and consider contextual knowledge within the naturalised setting of the specialist BEB clinic offers very little confines to the researcher and participants. Notably, the epistemological approach also considered the research question (Wildemuth, 1993) as it explains how useful knowledge was gained. According to (Maynard, 1994) the research question highlights the methods employed and what knowledge should be acquired to ensure a study is adequate and legitimate (Maynard, 1994).

Without contextual evidence, the researcher would have found it impossible to make sense of participants' reality of BEB example severely affected by spasms but unable to access treatment promptly, cancellation of booked appointment when treatment is required. This denotes this study's interpretive element, which argues that truth and knowledge are based on the participants' lived experiences. Therefore, the rationale for this stance was based on the researcher's need to understand the complexities of the phenomena 'BEB lived experience' from the perspective of patients who live it.

The interpretive epistemology was concerned with how the researcher acquired and communicated knowledge and the theory of knowledge, and the justification to broaden the understanding of BEB in ophthalmology. Although Pring (2000) asserts 'truth' or knowledge was a consensus formed by co-constructors whether the lived experiences of the condition were communicated or not, participants' visible symptoms confirm that a real-world of BEB exists.

However, the 'truth' sought was inextricably bound to the subjective meanings of participants' world and the objective negotiation with them during interaction to explore the reality of the phenomena of living with BEB. Therefore, knowledge was individually and socially constructed through personal experiences and perceptions. It arose inductively from situations captured through in-depth, systematic analysis and interpreted by the researcher.

The role was significant for the researcher, who works in partnership with the BEB patient-participants listening to when their treatment wanes and the best period to arrange their follow-up appointment. Assessment of the patient's state of health by asking questions demonstrates the researcher’s role in seeking the ‘truth’ through a collaborative partnership with patients. Furthermore, the research recommendations are grounded in contextual understanding to improve BEB service to deliver high-quality care and treatment for the condition. The researcher was stationed within the context/world of the participants and was, therefore, able to provide a more significant interpretation of the phenomena.

The positive epistemological stance was rejected as most BEB studies have a positivist epistemology. The objectivist-positivist epistemological stance assumes that there is a relationship between the world and its representation and that there is objective truth (Creswell et al., 2011; Willig, 2013), and that it requires rigorous scientific inquiry (Crotty, 1998). It reduces concepts such as QoL and effectiveness of care to statistics to arrive at a singular truth that can be generalised but does not account for individual experience at the interface of the health services. Therefore, such an approach would have provided an inadequate reflection of people’s lived experience since it bracketed or suspended the researcher’s judgment and rendered the study lacking in depth and clarity.

4.3 Methodological Decision

A relativist ontology and an interpretivist epistemology led the researcher to consider which methodological approach provided the best fit with the research questions, aims, and objectives. The focus of the study on the lived experience of people with BEB suggested that a phenomenological approach would be appropriate.

4.3.1 Phenomenology

The method phenomenology has been described as; *“the study of the essence”*, gaining the essential meanings of a phenomenon (Merleau-Ponty, 1962). Phenomenology studies an experience from the perspective of an individual. Phenomenology can be applied to a singular case to understand a system failure or a selective group like BEB patients. However, the ability to make inferences significantly increases when a particular factor recurs. However, when more participants have studied, the qualitative validity of robustness prevents any suggestion related to the entire population.

The essential inductive methods employed in phenomenological studies to gain information and perceptions; interviews, conversations, participant observation, action research, focus meetings, and analysis of personal texts. The current study considered time constraints and utilised good rapport and empathy through interviews and focus group discussion to gain maximum depth within a short period (Oakley, 1981).

4.3.2 Interpretive Phenomenological Analysis

Harding (1987, p. 3) defined IPA as ‘a theory and analysis of how research should proceed’. Interpretive Phenomenological Analysis (IPA) enables exploration of how participants make sense of their personal and social world. It has two primary aims: to look in detail at how someone makes sense of life experience and give a detailed interpretation of the account to understand the experience (Smith et al., 2009). Critics of the IPA methodology have voiced concerns that some IPA studies are not always appropriately conducted, even in a firm philosophical grounding. It has been criticized for being riddled with ambiguities and lacking standardization (Giorgi, 2010).

Another concern is that its methodology gives unsatisfactory recognition of the integral role of language (Willig et al., 2008). However (Smith et al.,2009), even novice researchers like me have disregarded this criticism as the narrative BEB participants provided language links and were the means available for more profound insight to make sense of their experiences.

Importantly this study addressed the concern that it reflected the voices of BEB experiences by providing sufficient interpretation of participants' narratives. IPA was instrumental in discovering why, what, where, when, and how the data was collected and analysed. IPA is well suited to exploring how people perceive and understand significant events in their lives. It is appropriate for this study as it can evoke the essence or meanings of BEB's lived experiences.

The IPA approach of this study explored what their lived experience of BEB means the methodology IPA was carefully chosen as it is closely aligned with the research question and the researcher's personal beliefs, which are heavily vested in caring for and understanding the human condition BEB. It was purpose-fitted to engage and provide deep critical insight for this community of patients whose voices have not been heard. The study questions combined with the scarcity of research on BEB and the nature of suffering patients' experiences present a complex, poorly understood area of health, which also made this methodology the most suitable choice.

Interpretive phenomenological analysis was appropriate for addressing the research aim. It examined how BEB patients made sense of their life experiences and provided a rich, descriptive narrative analytical account within the healthcare context (Smith, Flowers, & Larkin, 2009). Notably, the IPA role was essentially considered an analytical role for its primacy of making sense of the personal experience of the participants (Smith, 2004).

The methodology IPA offers three critical dimensions to this study: ideography, phenomenology, and hermeneutics. Ideography permitted to focus on individual cases and specific aspects of the experience before examining the group's commonality. This approach is essential in creating patterns, and Smith, Flowers, and Larkin (2009) suggested the analysis of and the findings from such an approach can add to nomothetic research.

Phenomenology is suitable for examining lived experiences and the condition of BEB. Since there is no direct access to those lived experiences and interpretation is required to understand the phenomena, this makes it hermeneutic. Phenomenology is often considered doubly hermeneutic, as both the individual and the researcher make sense of the experience (Smith, Flower, Larkin, 2009). Hermeneutics offers the researcher a critical approach to interpreting and connecting thoughts and feelings to what is said.

Without the researcher’s analytical role, the phenomenological essence of BEB would not be uncovered. In practice, the interactive nature of the relationship between the nurse researcher and patient participants is one of closeness developed over many years. Therefore, to authenticate the research findings, the author can draw upon the knowledge and understanding of the suffering faced by BEB patients, their experiences, behaviours, and interpersonal interactions. The aim was to understand participants’ perceptions rather than analysing independent variables that may impact the phenomena. The approach is considered a double hermeneutic, since the patient is trying to make sense of their lived experience, and the researcher is trying to make sense of the patient’s experience.

Two aspects of IPA are well suited to the caring profession: identifying and empathising with participants' perspectives, which is also instrumental in gaining a deep rich analysis of the data (Smith & Osborn, 2009).

Before selecting IPA, the other main qualitative approaches were considered are ethnography and grounded theory (Guba and Lincoln, 1994). Each approach was disregarded due to poor fit and suitability for the current study.

According to Creswell (1998), ethnography is a descriptive interpretation of the phenomena and interpretation of a cultural or social group. It requires the researchers to observe and participate, to provide meanings and interpretation for group behaviours and interactions (Creswell, 1998). Since this study was focused on lived experiences rather than social or cultural issues, ethnography was disregarded.

The second qualitative approach disregarded was action research, a collaborative approach suitable for creating change. It is often used in improvement science. Although the author wishes to inform change, the change process is not the central strategy of this study.

The second qualitative approach considered but was found unsuitable based on its rationale was grounded theory. Strauss and Corbin (1998) asserted that grounded theory aims to move beyond description to formulate or generate theory. The aim of this research was not to generate theory. Therefore, grounded theory was not aligned with this current study, which focuses on the daily lives of patients coping with BEB.

4.4 Methods

This chapter presents the methods used to undertake this qualitative research. It is divided into five sections. The first section describes the rationale of how, why, and which participants were recruited from the outpatient setting. It outlines the ethical considerations and consent procedure employed during the study. The data collection method employed in phase 1, semi-structured interviews, and phase 2, focus group interviews, were discussed. The research transcription method was outlined. It also describes how the data was analysed and addresses the evaluation challenges of qualitative data.

4.4.1 Setting and participants' recruitment

Adult patients diagnosed with BEB for at least 12 months and who attended the specialist BEB nurse-led clinic in an outpatient's department (OPD) in London, England, were approached and invited by the nurse researcher to participate in this research. Although there are few patients with learning difficulties and dementia, 40% of our patients in the outpatient department are non-English speaking and were among those considered vulnerable adults who were not recruited for reasons of mental capacity and the costs associated with translation for a study of this size.

Sampling was conducted following the guidelines of IPA set out by Smith et al. (2009). The sample was homogenous because participants could provide the particulars of their lived experience within the specialist BEB clinic. The final size of the sample was determined iteratively as the interviews progressed and transcripts were analysed. A total of ten participants with the ability to vocalise their lived experience with BEB were given study information and adequate time to consider participation.

4.4.2 Ethical Considerations and Consent Procedure

The present study was reviewed and gained approval from the University of West London College of Nursing, Midwifery and Health Visiting and the University REC: UWL/REC/CNMH-00324, the NHS REC: 18/NW/0388, and Moorfields Eye Hospital Research Board: DUNN1001 before the recruitment of study participants (Appendix 6).

The recruitment processes followed ethical considerations, such as privacy, and potential participants were given patient information regarding the study and sufficient time to decide whether to participate. No follow-up emails or telephone calls were made that could be considered coercion. The information given was well written and unbiased (see Appendix 7), with the description of the study and no pressure to commit. The information addressed patients as participants to remove any misconception that taking place in the study would benefit their treatment.

Consent

Written consent was sought by the research nurse from all patient participants in agreement on their subsequent visit to the OPD. A participant leaflet and consent form provided information on the study, its rationale, the right to change their minds, and how personal data will be protected. Participants were given time to reread the study information while awaiting treatment in the OPD of the nurse-led clinic. The gap between invitation and participation was between eight weeks and three months for the interview and focus group discussion, which took place several months after the initial interview.

Before commencing the interviews, each participant was informed that disclosing any information regarding risk to themselves or others would result in information being shared with the head of the hospital’s risk and safety and with the academic supervisors of the study to determine an appropriate course of action. The hospital counselling service was also open to patients.

The participants were either interviewed at the end of their clinic appointment, or the researcher arranged a convenient date and time to conduct the interview. The researcher gave the participant a chance to read through the information pack again and answer any questions on the interview day. A signed formal written consent form (Appendix 8) was reviewed to ensure participants were still in agreement before the commencement of each interview.

4.4.3 Data Protection and Confidentiality

The data was stored securely per the General Data Protection Regulation (GDPR, 2018), superseded the Data Protection Act (1998). The research findings do not contain participants' personal, identifiable data. Face-to-face narrative interviews were digitally recorded. The digital recording device was locked away in a secure draw, only accessible to the principal researcher. All transcriptions were stored electronically in a password-protected file. Participants' anonymity was protected by using pseudonyms throughout the study's transcription, coding, and reporting to maintain confidentiality.

4.4.4 Data Collection Methods

4.4.4.1 Phase 1: Semi-Structured Interviews

The chosen data collection method for this study was face-to-face, semi-structured interviews. A semi-structured interview is best suited to IPA because of its flexibility and because it invites participants to offer a rich, detailed first-person account of their experiences (Smith et al., 2009).

The qualitative method is seen as a conversation with a purpose and allows researchers to pursue thoughts and ideas. In essence, it offers a richness and depth of understanding that quantitative approaches fail to challenge. It permitted follow-up questions to explore the phenomenon, the lived experience of BEB. This contrasts with other approaches, where surveys and questionnaires provide rigidity that fails to explore the themes generated by participants. The QoL studies included in this study result from a process of condensing and interpretation of primary raw data done by various study authors. Therefore, due to its rigidity, further synthesised cannot be added. However, individual papers can be referred to for detailed discussions and interpretations of the identified domains.

The researcher created the semi-structured interview schedule (see Appendix 9) and was informed by previous research (Smith & Osborn, 2003; Wimpenny & Gass, 2000) and the input of other experts within the BEB specialist clinic. The interview schedule was used to guide the discussions; it was not piloted, but discussions evolved from participants' responses and what each participant felt was essential to talk about (Smith, Flowers, & Larkin, 2009). The interview schedule ensured the focus remained primarily on the lived experience of BEB to address the research aim and objectives while probing deeper into new areas (Coolican, 2004; Kvale, 1996; Robson, 2002) to produce rich data (Smith, Flowers, & Larkin, 2009) relevant to the research questions. The interviews were audio recorded and transcribed. Once the researcher transcribed the individual semi-structured interviews, they were uploaded in Nvivo.

4.4.4.2. Phase 2 – Focus Group Discussion

The chosen data collection method of a focus group discussion (FGD) enabled group interaction. The FGD took place on 31st October 2019 at 10:30hrs at the specialist hospital. The Group was designed to utilise group interaction or capture collective consensus (Kitzinger, 1994) to gain further information about BEB (William & Katz, 2001), specifically how to improve the service. All participants invited to participate consented to undertake Phase 1 and 2 of the study. In that regard, participants were purposely selected in Phase 2 and therefore provided improvement advice. However, only eight BEB participants attended.

The FGD was utilised as a social process, which allows for interaction and explanation in pursuit of consensus and diversity (Barbour & Kitzinger, 1998; Doyle, 2004; Morgan, 1996). It provided the opportunity for participants to validate the data interpretation undertaken in individual interviews and inform the researcher on how services can improve to be more responsive to their needs.

In comparison to individual interviews, it provided an economical means to capture deeper information on how improvement can be realised and non-verbal communication. The FGDs encouraged BEB participants to make connections to various occurrences in the outpatient clinic through discussions that may not occur during individual interviews. The advantage of utilising patient participants as opposed to practitioners is that the entire process is empowering. It places patients at the centre of their own care. The disadvantages are a participant may not honestly express their view if there are opposing views. It also has additional costs such as transport and food and drinks for all stakeholders.

However, intentional, or inadvertent moderator bias can occur. This study sought to eliminate all biases through careful facial expressions, body language, tone, even language that may introduce bias. However, there are unavoidable biases example, the moderator's age, social status, race, and gender which cannot be controlled. As a facilitator of the researcher, neutrality was maintained in dress code, tone, and body language was open, non-persuasive and didn't give opinions while moderating.

Another rationale for undertaking the FGDs was based on the collective response of patients in Phase I of the study to share and gain further knowledge. Although it provided a means of getting their voices heard, the discussion also aimed to change practice. The lead research nurse was invited to support nursing research in the Trust and for the quality improvement led to identifying areas for improvement. The questions asked provided a framework for the discussion, but focus groups were designed to encourage knowledgeable participants to identify the salient issues that affect their quality of care and specifically how the service could improve.

The FGD was used as a qualitative method to bridge the gap between scientific research and local knowledge. It is also understood as participatory research (Bennett et al., 2017; Cornwall & Jewkes, 1995). This FGD provided demonstrable evidence of how the NHS, specifically the BEB clinic, can improve its care through patient-public involvement (PPI). PPI is a government strategy (DOH, 2008) to provide patient-centred care, which empowers patients and fosters quality improvements in healthcare services (Mockford et al., 2012), but such a context has rarely been analysed (Abelson et al., 2007). The moderator/researcher aimed to ensure systemic research was undertaken by collecting in-depth data on the research question: what needs to change to ensure that outpatient services for patients with BEB are responsive to expressed needs?

Box 1 **Sample Focus Group Questions:**

- As a service user, how would you like the service to run?
- What aspects of the service do you think can be improved?
- How would you prefer to book your appointment, by telephone, app, or online, and why?

The method employed by the novice researcher in the FGD was that of facilitation or moderation. Discussions lasted approximately one hour. Phase 1 findings were presented, and participants voted unanimously through a show of hands; it represented their lived experience. Key areas of discussion included how to improve the OPD (see FGD interview schedule, Appendix 10). All discussion was recorded and transcribed verbatim, and participants were assigned pseudonyms to maintain anonymity. The audio of the FGD was recorded and fully transcribed for analysis after being stored in the software package NVivo.

Therefore, the focus group discussion served to determine how to respond to what was learned in Phase 1 of the study to respond to the needs of patients. However, it provided additional reliability since commonalities in the Phase 1 findings were confirmed among participants. Nonetheless, the central area of discussion included how to improve the BEB clinic.

NVivo software (NVivo qualitative data analysis software; QSR International Pvt. Ltd. Version 10, 2014) was used to analyse the data in both phases of the study and will be discussed within this section. The analysis was undertaken in five stages. Both transcripts were read several times, and the researcher listened to the interview recordings again several times. This was done to develop a deep immersion in the data. An overview of the content, thematic analysis (developing a coding scheme), indexing (applying the codes systematically to the data), charting (rearranging the data according to the thematic content to allow comparative analysis), and mapping and interpretation (defining key concepts, delineating the range and nature of phenomena, creating typologies, findings associations, providing explanations and developing strategies).

This approach developed a transcript, and codes which were included in the framework. The coding framework was reviewed, discussed, and revised until it reflected the participants' views and a final version agreed. The code was subsequently grouped. The coding of each theme and subtheme was retained until the final interpretation of the findings was reached. The research transcription is discussed below.

4.4.5 Research Transcription

The researcher transcribed all interviews naturally to represent the real world (Schegloff, 1997) and included notes of expressions. For example, expressions of laughter or pauses were incorporated within the written transcript to give more context to what was being said by both the research participants and the researcher (Lapadat & Lindsay, 1999; Ochs, 1979; Sandelowski, 1994) and to help uncover everyday discourse and the meanings that construct reality (Cameron, 2001). Therefore, it aided in the embodiment of the lived experience of BEB and gave an understanding of the powers of influence on participants' lives (Fairclough, 1993). The interview data were analysed using IPA. The benefit of the researcher transcribing this research is that it helped with the interviewees' quotes and adherence to the research protocol and added credibility, context, and transferability.

4.4.6 Data Analysis

IPA provided a systematic and structural approach to analyse the data (Smith et al., 2009), addressing the experiences encountered by people with BEB. The chosen method of data analysis, IPA, is aligned with the research question, the ontological and epistemological position, theoretical framework and methodology of this study. Although there are various methodological approaches to analysing qualitative data, such as narrative analysis relating to sense-making and discourse analysis relating to how the subject is spoken of. IPA is best suited to this study as it closely examines the personal meaning and sense-making of the specialist BEB clinic where participants share similar experiences.

The analysis deployed followed guidelines provided by Smith, Flowers, and Larkin (2009). This framework consisted of six steps: reading and re-reading of transcripts, initial noting (descriptive, linguistic, and conceptual comments), developing emergent themes, searching for connections across emergent themes for analysis of the findings an example of how initial concepts emerged from each transcript was developed into emerging themes, repeating the process and finally looking for patterns across cases (see Table 4 for a diagrammatic representation of IPA below).

Table 3: IPA analysis using the framework of Smith, Flowers, and Larkin (2009)

Step	**Process**
1	Re-reading the transcription several times to gain a sense of the whole context.
2	Initial noting of three types of comments: participants' descriptive comments to rephrase; linguistic comments, paying attention to words and expressions; and conceptual comments, including the researcher's knowledge of literature and lived experiences.
3	Identification of emerging themes from the comments made previously. Data-reading becomes more focused and interpretive. Further themes created will be rechecked for consistency with the research supervisor.
4	Search for connections across the emerging themes. Themes will be divided between the overarching research questions and grouped into subordinate themes, then further subdivided and organised chronologically to be traced back to the interviewee. A hierarchical node tree will be visually created using NVivo.
5	Step 4 will be repeated, and the subordinate themes will be re organised. Overused or ambiguous descriptions will be removed to prevent the weakening of meanings and exposing fundamental structures. Original data will be returned to ensure hermeneutic dialogue between themes and source, superordinate and subordinate themes.
6	Look for patterns across cases. Superordinate themes across all cases will be developed. Divergence and nuances will be grouped into themes. The data analysis continues throughout the process of writing.

In keeping with the phenomenological approach, the participants were allowed to confirm that their individual transcription represented their words, feelings, and emotions related to their lived experiences. The rationale for participant confirmation was to aid research transparency. The review of participants' interviews demonstrated their collective need to inform service improvement; therefore, a focus group was formulated from the common interest in improving the service and hearing from other people affected by BEB.

Participants were twice allowed to verify the data: once in the individual interview and subsequently in a focus group during Phase 2 of data collection. This verification process fostered participants' feedback and clearly distinguished appearances and essence (van Manen, 1990). The transcription was upload into the computer-assisted qualitative data analysis software package NVivo to add transparency to the data. The rationale for utilising software was to assist in sorting, retrieving, indexing, and handling the large amounts of text generated by transcription (Gibbs, 2002).

4.4.7 Focus Group Data Analysis

Similarly, an inductive data analysis was adopted to examine, organise and analyse the narrative data of the FGD which was grouped into six overarching themes: waiting times, access to service, information and technological challenges, lack of resources, continuity of care, and customer care. (See Figure 11, themes).

4.5 Evaluation of Qualitative Challenges

This study must be considered of high quality. Therefore, standards for interpretive study set out by the researcher and those of Lincoln and Guba (1985) are the assessment of trustworthiness, credibility, transferability, and dependability. In addition, the achievement of the research aim should provide a sufficient basis to judge the authenticity of this study. Although the interpretive challenges are rigorous, this was addressed throughout the process by employing reflexivity (Table 8, Appendix 3) rather than utilising objective standards (Smith, 1993). A reflective diary was used for continuous reflexivity to demonstrate confirmability, trustworthiness, honesty, ethical consideration and openness about decisions made at various study levels. This researcher's approach to the presentation of this qualitative research includes a level of reflexivity (Alvesson & Sköldberg, 2009). See Table 5 below.

Table 4: Levels of reflexivity (Alvesson & Sköldberg, 2009, p. 273)

Aspect/Levels	**Focus**
Interaction with empirical material	Accounts in interviews, situational observations, and other empirical material
Interpretation	Underlying meanings
Critical interpretation	Ideology, power and social reproduction
Reflection on text production and language use	Own text, claims to authority, and selectivity of voices represented in the text

Conceptually, this study sets out the elements of its subjective stance, which shows dependability: a relativist ontological position, interpretive epistemology, the theoretical perspective of phenomenology, the methodology of IPA, and the corresponding semi-structured interviews followed by a focus group of all previously interviewed participants. The insider researcher's positionality sought to demonstrate credibility through contextual knowledge of the BEB condition (Keso et al., 2009). This was critical in determining how decisions were made and how knowledge was perceived and interpreted to influence the study’s findings.

Participants’ reviews of their interview transcripts and collective FGD added credibility to the data and verification that the study is an accurate representation of the patients’ lived experience of BEB. The dissemination of these findings in publications and conference communications to other practitioners, service managers, and researchers will ensure transferability and indicate how health professionals can work with patients to improve care delivery concepts like generalizability.

Furthermore, qualitative research guidelines were followed by the researcher throughout the research process (Yardley, 2008) to improve the validity of the research. The supervisory process was utilized to enhance the credibility of the interpretation of findings plus the consistency and coherency of the analysis (Yardley, 2008). In the form of action learning, the researcher also engaged peer review to enhance both the trustworthiness and quality of data. The researcher had prior experience of BEB and was aware of previous quantitative literature which indicated many difficulties associated with BEB, including the presence of physical and psychological difficulties (Conte et al., 2016; Grandas et al., 1988; Hallett, 2002; Hellman & Torres-Russotto, 2015; Jankovic & Orman, 1987), but she was determined to ensure her openness to all participants’ accounts, including those that might contradict the extant research. The findings of this study are presented in the next chapter.

Conclusion

The chapter presented the rationale for the philosophical and theoretical underpinning of the research, namely the research question and the researcher's disciplinary background as a specialist practitioner. Caring for BEB patients, listening to their thoughts, feelings and emotions during the provision of physical treatment while supporting psychological well-being. The researcher personal world view, which reflects a relativist interpretivist stance provided the opportunity to add to the scarce body of qualitative BEB knowledge through multiple realities of people. Collection of the data took a phenomenological approach and semi-structured interviews, and a focus group discussion was utilised. The data was examined in relation to the six dimensions of quality of care, employing the use of a practitioner and researcher's lens. Interpretive Phenomenological Analysis (IPA) was the chosen methodological approach to facilitate 'action' to deliver high-quality outpatient care that takes account of the physical, emotional, mental, and spiritual well-being of patients. Participants' recruitment, data collection, transcription and analysis were also discussed in detail along with the ethical considerations utilised.

Chapter 5: Findings

5.1 The Patient Experience of Living with BEB

The insider perspective of ten participants' lived experiences were investigated to gain a deeper understanding of the impact of BEB on their daily lives, with particular emphasis on treatment and care within the specialist BEB clinic. This chapter presents Phase 1 of the study, which addressed two of the study objectives: 'to gain a deeper understanding of the lived experience of BEB' and secondly 'to ascertain how an acute episode of the condition BEB affects the patient QoL'.

Five superordinate themes emerged from the data following in-depth analysis of transcribed interviews and field notes: (1) A condition with no name; (2) An incredibly debilitating state; (3) Fleeting normality; (4) Loss of social richness; and (5) A battle for adequate care. Along with 24 subordinate themes, this chapter conveys the interpreted meanings supported by illustration quotes of participants' perspectives ascribed to the lived experience of BEB. Superordinate and supporting subordinate themes are presented in Figure 10.

Individual responses of participants highlighted their need to improve the service. Therefore, Phase 2 of the study was organised to solicit the participants' combined input in a focus group discussion (FGD). It helped address the third objective: to inform service improvement or redesign that responds to the needs of BEB patients and can be found in Chapter 5.

5.1.1 Description of the Participants

The results chapter presents a deeper understanding of BEB captured through ten participants' unique lived experiences. The section also describes the participants, assigned a name for anonymity and a persona in keeping with a phenomenological approach. Participants demographics consisted of nine white and one black participant, and were mainly from graduate professions:

- Delbert, 69-year-old male retired lawyer
- Denise, 65-year-old woman university lecturer
- Egna, 88-year-old retired female teacher
- Judy, 72-year-old retired female magistrate
- Petal, 56-year-old female social worker
- Janey, 65-year-old female retired medical secretary
- Leah, 54-year-old female company director
- Drey, 51-year-old male personal trainer
- Nadia, 55-year-old female music teacher; and
- Jey, 75-year-old male retired bus driver

All participants attended the specialist BEB clinic at a tertiary ophthalmic hospital. There were only three male participants among the invited participants, reflecting the high prevalence of BEB among women.

5.2 Superordinate Theme 1: No knowledge

“No one knew what it was” (Drey). This theme reflects the widespread lack of knowledge among health professionals and participants.

5.2.1 Subordinate Theme 1: Loss of trust in primary care professionals

Most participants concurred that the main barrier to diagnosis and treatment was primary care health professionals’ lack of knowledge of the condition, which caused a loss of trust. Participants conveyed they felt doubted and disbelieved, anger at being misdiagnosed, inappropriately treated. Even their request to be referred to an ophthalmologist or neurologist was refused, and their symptoms were deemed not severe enough to warrant such care.

5.2.1.1 Failure to identify and diagnose apparent symptoms

In the early stages of the condition, participants indicated health professionals missed their symptoms although they were frequently blinking of the condition: *“When I first had the condition, no one knows what it was” (Drey).* Egna reported the symptoms was not uncontrollable but *“blinking very frequently”*, another accompanying problem was ocular discomfort*: “They are just so dry, uncomfortable and sore all the time”* (Nadia). In addition, there was*, “I struggle with glare from the lights”* (Petal). Participants felt desperate for answers and were eventually referred to specialist care, but instead faced treatment delays between six months and two years.

5.2.1.2 Misdiagnoses and delays

Some participants’ tone expressed palpable anger and disbelief from the lack of answers, incorrect diagnoses and delayed treatment, even on reflection. Drey said that *“It took two years for my condition to be diagnosed, two years, which they thought it was dry eyes, and they were treating that.”* The most common condition to be diagnosed instead of BEB was dry eyes.

5.2.1.3 Being doubted, inappropriately treated and refused referral

Some participants explained the frustration they felt like the severity of their condition was doubted, and they lost trust in primary care professionals and sought private healthcare. Janey stated, *"He didn't believe it was serious enough to be referred on anywhere."* In addition, some GPs refused to refer to specialist care, the condition was misdiagnosis and incorrectly treated, as Janey mentioned:

> *"For a year, they thought I had an infection, and I was treated constantly with steroids, and it was only then that I paid for a private appointment, and he diagnosed and wrote to my GP. It was at that point that my GP referred me."*

Similarly, Egna, after failing to find answers, described how she felt she had no other alternative: *"I had to go and see an ophthalmologist privately. He said it was blepharospasm."* Participants' actions confirmed that they lost trust in health professionals in the NHS who failed to recognise the condition, which negatively impacted the expedition of timely specialist care.

5.2.1.4 Private consultation aided referral but not always a diagnosis

Although most participants felt strongly that consultation with private ophthalmologists was instrumental in aiding a referral, a few participants were referred to the specialist BEB clinic but received no clarity regarding the diagnosis. Denise described how, even with private healthcare; a definitive answer remained elusive: *"I went to see a private ophthalmologist who said I should be referred right away. He was pretty sure but did not want to commit to the diagnosis."*

Acutely affected participants explained their long wait for specialist consultation to better understand and address their symptoms. Delbert described his wait without any knowledge of what was wrong: *"Until I got here [specialist BEB clinic], they didn't quite* *know what was wrong with me."* Contrary to other participants, Leah could diagnose her condition by searching the internet and concluded that the search for an accurate diagnosis felt *"like one battle after another".* It resonated with most participants.

5.2.2 Subordinate Theme 2: Ignoring and Minimising BEB

5.2.2.1 Health professionals minimised the condition

Some participants expressed that how health professionals reported their misdiagnosis was considered casual. Participants expressed that dry eye diagnosis was made in a blasé manner, a conclusion they found difficult to accept*: "I was blinking very frequently and he [optician] just said: 'Oh, you've got dry eyes. Put in lubricant"* (Egna). Since there was no change in the eyelid closure, it suggested the medication did not affect spasmodic closure.

5.2.2.2 Participants did not act upon their symptoms until they were noticed by other

A few participants reported their initial failure to seek immediate medical attention and their use of words such as *"I flick a few times"* (Delbert) and *"my eyes close a bit more"* (Denise). It suggested the severity of their symptoms was trivialised. As the symptoms became worse and more apparent to friends and family, the participants were no longer able to ignore the problem: *"I probably had a feeling that something was wrong. One afternoon in July, I think it was when speaking to my sister-in-law, and she thought there was something wrong with my eyes"* (Denise). Delbert also described how he: *"… was working in social care as a lawyer, and one of their specialists for people with sight problems suggested I ask my GP to refer me".* Delbert and Denise both sought medical attention after being advised to do so. In contrast, Nadia received a referred but did not wish to travel.

5.2.2.3 Self-referral was made as the condition worsened

Unlike other participants, although Jey ignored his symptoms initially, he explained: *"I don't know how long ago I was suffering. First, I thought it would go away then I start getting this problem all the time, you know. You're driving; the next thing you know, your eyes close on you. I just walked into the eye hospital."*

Unlike any of the other participants, Jey bypassed primary care and was seen in the specialist ophthalmic accident and emergency*: "They right away told me I had this condition. I had to give up driving and so forth, you know. And then… and they… then they gave me treatment."*

Jey's experience suggests that BEB is not difficult to diagnose by specialists. In addition, the knowledgeable health professional afforded patients much-needed confidence to trust their competence and judgments. The experience also provided a more profound understanding that, without timely treatment, BEB deteriorated into an acute episode of the condition with functional blindness. It also created a psychological decline in participants' moods.

The theme 'no knowledge' represented that lack of knowledge among both participants and health professionals acted as a barrier to timely diagnosis and treatment, leading participants to lose trust in primary care healthcare professionals. In addition, it led to both participants and health professionals trivialising the condition and to the belief that the condition had no significant impact on health. Proactive health-seeking participants experienced feelings of being doubted and disbelief and were angered. For some participants, lay referrals and worsening of the disease provided the impetus for seeking care.

5.3 Superordinate Theme 2: Incredibly debilitating state

“It is incredibly debilitating when it’s bad” (Leah). The theme ‘incredible debilitating state’ is representative of the impact of an acute episode of BEB, which resulted from a lack of treatment. This affected both physical and psychological health.

5.3.1 Subordinate Theme 1: Functional blindness

5.3.1.1 Poor navigation, helplessness, distress and fear

Unanimously, participants described the impact of the feeling of blindness being unable to function, which led to helplessness, distress, and fear as they navigated their life. Drey described, “*I felt helpless*, *bumping into things, so it was affecting my whole daily regime, you know, my whole daily life was dreadful”.* Participants felt helpless and in distress as all aspects of daily life was affected.

5.3.1.2 Functional blindness

A distinctive feature of the severe disease reported by all participants was functional blindness, which acutely rendered participants blind at the most inappropriate time. As Petal described, *“the eyes would start to close while I was walking so it would definitely impact [daily life]”.* Spontaneous closure was debilitating and caused fear among all participants as they were forced to *“stop and really try and get your eyes open again”* (Denise). Some participants sought to provide a label that would best describe them with no definitive answers*. “I think I’d probably describe myself as functionally almost blind”* (Leah). Leah’s definition validated the theoretical knowledge she had gained by searching the internet and reading scientific journals.

This difficulty caused Drey to describe himself as being of *“partial eyesight”*. The term ‘functionally almost blind’ denotes visual failure to any specific task, although, in contrast to ‘true blindness’, all the participants’ vision remained perfect. However, the unpredictable nature of BEB means that moments of perfect vision are suddenly interrupted by the closure of the eyelids*.* The transient state of temporary blindness hardly affords those affected by it the time to learn how to function while suffering.

5.3.1.3 Ocular discomfort

Many participants described feeling frustrated as they experienced ocular discomfort. They struggled to keep their eyes open against the sudden, frequent, and persistent blinking or spasms, glare or sensitivity to light, dryness, and soreness of the eyes, making their lives highly uncomfortable. Janey added further depth to the understanding of the suffering being experienced as she described the enormous impact the condition had on all aspects of a person life: *"It's completely life-changing, and it's not quite like blindness illness, which is terrible, it feels as if somebody has put salt in your eyes"* (Janey). The description provided evidence that BEB is also accompanied by ocular discomfort.

5.3.1.4 Functional blindness causes physical disability

Individually, all participants discussed the presence of the physical disability. All participants expressed, due to an inability to see, that they were physically challenged to watch television, read, drive, cook, use the internet and social media, go to the cinema, read music to sing in the choir, use gym equipment, walk or even sit in the garden. Judy explained her work as a magistrate required her to read a lot, but she found "reading difficult". Petal described the effects of the condition on her abilities: "*Primarily… my ability to drive and things like watching the television."*

5.3.1.5 Limiting activities

As Nadia described, some participants tried to maintain physical abilities: *"Limited how long I can read for and again driving."* Limiting daily activities helped participants to retain some degree of function. Petal added further understanding to the rationale for limitation: *"It has curtailed driving. I can't just jump out into the car and go out for a day* *in the country if I want to."* The use of the term 'can't' suggest an awareness that BEB symptoms restrict capability and prevent normal function.

5.3.2 Subordinate Theme 2: Giving up

5.3.2.1 Frustrated and too tired to function

The impact of functional blindness and unrelenting spasms caused all participants to 'give up', relinquishing most aspects of daily life. Acutely affected and considered day-to-day tasks were considered *"too challenging for me and too tiring"* (Petal). The difficulties experienced caused exhaustion and frustration, and participants felt physically challenged to the extent that they had no other alternative but to give up, as Denise described: *"I was trying to read in bed last night, and I gave up because it was so frustrating"*. All participants relinquished various roles: *"So I gave up work, I gave up exercise, I gave up driving, I gave up cooking pretty much, I gave up socialising, I gave up watching tele, gave up reading"* (Leah).

Similarly, Denise resonated with the feelings of all participants as she described her rationale for needing to give up as related to the avoidance of frustration, especially when *"(reading)… I gave up because it was so frustrating".* Frustration provided some understanding of participants' lowered mood due to physical limitations. Judy explained how multiple aspects of daily life were given up as the condition severely impacted her QoL: *"It's been affected hugely because I've had to give up a lot of the activities that I was involved with, in fact, all of them".* The phase 'giving up', provided acknowledgment or that defeat was conceded as participants felt competence was lacking.

5.3.3 Subordinate theme 3: Disease progression beyond the eyes

5.3.3.1 Disease progression affects facial appearance

Most participants described unrelenting spasms beyond the confines of the eyelid and caused contortions of their facial features, which was fearful and exhausting: *"It was exhausting, I could hardly open my eyes at all, and it was one spasm after the other all day, and I was grimacing with my face with meige"* (Janey). The progression into another facial dystonic condition caused participants' facial expressions to appear grim.

5.3.3.2 Disease progression to the neck challenged mobility and caused fear

Some participants expressed that the progression of the disease to the neck created difficulties walking and caused a fear of falling. To avoid a fall, care was taken while walking: *"In the last few months, my neck seems to have gone down a bit, but I'm always careful"* (Delbert). Judy added depth to this as she explained being unable to walk in a straight line due to her neck dystonia, which she was also diagnosed with: *"I can't walk in a straight line for a start, but I'm not sure that's my eyes. I was also diagnosed with dystonia in my neck".* Egna also expressed being: *"Careful when I am walking because my balance has deteriorated"*. Being unsteady was found to cause extreme difficulties.

5.3.4 Subordinate Theme 4: Activities that worsen and improve BEB

This subordinate theme provides an understanding of activities, and that worsens the condition. In addition, it includes strategies used to improve and help participants to cope temporarily.

5.3.4.1 Activities that worsened the condition

All participants described certain activities that caused deterioration, specifically exercise, eating, walking, and talking. In addition, squinting movements of the eyelids and bright lights also aggravated the symptoms. Worsening of the condition was considered severe spasms that prevented useful vision or closed the eyelids.

5.3.4.2 Avoidance of activities that precipitate the condition

Participants curtailed various pleasurable activities that could precipitate debilitating spasms. Leah described her avoidance of exerting activities: *"I used to be able to go to the gym and work out, and now, any sort of exertion, like running or anything like that, merely makes the face kick-off."* With cognizance, certain activities were avoided to prevent the terribly debilitating symptoms.

Participants described avoiding simple life pleasures to remove the possibility of succumbing to relentless spasms and exhaustion:

> *"Going for a walk, I tend to turn down the offer if my husband says 'shall we go for a walk?' I just say no, because I know actually it is quite exhausting."* (Denise)

Delbert's tone was quite sad as he described avoiding bright light: *"I prefer to sit in the shade rather than sunlight, whereas before it didn't bother me."* This provided the understanding that people would rather compromise pleasure to live a life free of BEB, even though this prevented enjoyment and caused sadness.

5.3.4.3 Ameliorating techniques and coping strategies

Some participants described that concentration, breathing techniques and holding the face as temporary techniques that helped to ameliorate symptoms: *"[walking] By the time you come back, and you have been concentrating really hard on keeping your eyes opened so you don't walk into anything or step off the kerb or something like that"*

(Denise). Similarly, Leah also expressed that: *“I’m battling with my face all the time. I have to really kind of concentrate on keeping my face and breathing and keeping my eyes from screwing up.”* The use of the term battling’ led to some understanding that participants develop counteracting strategies to challenge and resist spasms, especially in public places where their appearance could be perceived negatively. However, such sensory tricks only provided temporary relief.

Other participants described the use of sunglasses, particularly rose-tinted ones, were effective: *“So the rose ones have helped with television and the dark ones I have to wear outside because I am very photophobic”* (Janey). Coping strategies were beneficial in public or unfamiliar places.

5.3.5 Subordinate Theme 5: A decline in psychological health

5.3.5.1 Poor visualization, changes in facial appearance and fear for the future

Being distressed over the persistent nature of the spasmodic closure of the eyelids and new facial appearance caused all participant’s psychological distress. Participants describe the distress felt’ when having difficulties coping with the closure of the eyes in everyday life: *“It has been maddening, my eyes closed, and I missed vital things! I had quite a lot of difficulty in getting my eyes to open and keeping them open”* (Egna).

The use of the word ‘maddening’ indicated the psychological distress felt. In addition, facial contortions also caused some participants distress, as Janey described*: “With the condition, you are pulling faces like you are a mad person.”* Also, the unrelenting nature of spasms and the distressed tone and emphasis placed on ‘all the time’ suggest participants fear for the future*: “I’m having spasms all the time”* (Nadia).

No longer confident in public settings, '*a loss of social richness' was described: "My moods were dreadful. I felt because I didn't have that social richness or confidence. I just walked in, and they checked because nobody was there to help me, you know, so I [was] always on that limit. My social life was affected"* (Drey).

Abnormal spasmodic closure created a loss of confidence but, without support, all participants felt embarrassment each time they were scrutinised in public. Drey's reflections also demonstrated the dilemma all participants faced regarding having the freedom to socialise and needing full-time support while doing so. Although despair and guilt were apparent from the participants' tones, their feeling of gratitude for support was also evident.

5.3.5.2 Grief at the lack of capability

Participants expressed grief and negatively defined themselves due to their inability to undertake day-to-day activities*: "So suddenly I'm a klutz and not capable"* (Janey). The negativity in defining self, give insight to the difficulties participants' felt towards changes in their abilities. All participants expressed grief due to the loss of vision and physical disability as *"everything was a terrible struggle"* (Leah), and the current state of health was reported *as "incredibly debilitating"* (Leah). However, the acknowledgment that everything was a struggle indicated participants' acceptance of psychological defeat.

5.3.5.3 High-stress levels and anxiety

High levels of stress was the conclusion many participants considered as the rationale for being unable to cope with the unknown condition, and Janey described this state*: "You can't switch off the fear and flight ever, and so stress levels are a little bit higher just because of that."* High-stress levels caused the psychological impediment and Petal associated: *"It's probably to do with anxiety."* Some participants reported being unable to feel calm and seeking medical attention. They were then diagnosed with clinical depression.

5.3.5.4 Clinical depression prior to a BEB diagnosis

Many of the participants related that they became anxious and received a diagnosis of clinical depression even before gaining a diagnosis at the specialist eye hospital: *"I'm certainly very anxious a lot of the time, before coming here [the specialist eye hospital]. I went to my GP, and he prescribed antidepressants, and I took those for six months, and they were a lifesaver"* (Judy). The word 'lifesaver' in my interpretation sheds light on participants' belief that without antidepressants, life would not be possible. Both reflections also provided a deeper understanding of the 'debilitating state' of BEB caused and the psychological decline that impacted participants' coping abilities.

Similarly, Nadia explained the closure of the eyelids appeared to close off the social world of the participants*: "You feel, I don't know, left out a bit, you know? I think it's a depressing condition, really. I think it does make you feel depressed at times."* Feelings of depression eventually caused Nadia to become isolated. Most participants persistently pursued a diagnosis to label the condition's existence with daily life severely affected.

5.3.5.5 Clinical depression following BEB diagnosis

Other participants expressed that they became anxious and clinically depressed following the diagnosis of the long-term condition BEB: *"So when it was first diagnosed, it affected my mood very badly, and I actually became clinically depressed and was prescribed antidepressants"* (Leah). Similarly, Petal acknowledges that *"I just couldn't cope"*.

5.3.5.6 A diagnosis of clinical depression required acceptance

Attempts were made to accept the condition and to gain resilience, which was highlighted in some participants' reflection: *"I had some counselling here and elsewhere to try and help me to sort of turn around that thinking really"* (Petal). Some participants needed help accepting the condition, and counselling offered self-awareness to gain emotional clarity. Other participants had therapy: *"I had cognitive behavioural therapy"* (Leah). This involved working with a therapist to change behaviour or thinking patterns around accepting the condition.

The theme 'incredibly debilitating state' presented the impact of BEB on physical and psychological health, which adversely affected QoL. It created feelings of frustration, helplessness, distress, fear, and anxiety, which led to a diagnosis of clinical depression as participants struggled to see and navigate their world. In addition, the participants grieved their lack of capability and experienced high levels of anxiety amid these challenges – ocular discomfort and eyelid spasms, which progressed to cause facial contortions and neck spasms. Although awareness of worsening activities and sensory tricks to ameliorate severely disabling symptoms provided coping strategies, the relief gained was only temporary. For many participants, a diagnosis of clinical depression was made even before BEB was diagnosed.

5.4 Superordinate Theme 3: Loss of Social Richness

"I didn't have that social richness" (Drey).

The theme 'loss of social richness' is closely aligned with the theme 'an incredibly debilitating state' and represents the social and emotional impact of the condition BEB on participants.

5.4.1 Subordinate Theme 1: Employment and career identity

5.4.1.1 Termination of employment affected career identity

Most participants' words, tone, and inferences suggested they felt significant loss and sadness from being forced to terminate their employment. Drey, a fitness instructor, could not cope with unrelenting spasms and described that his *"working has not been good"* (Drey). Similarly, Judy resigned from her employment: *"I worked as a magistrate in Birmingham for 25 years, and I gave that up because of the amount of reading that was involved with that."* Grief was also felt as Nadia described not being able to do what she loves: *"I used to teach music."*

Leah attempted to remain employed by adopting counteracting strategies to restrain facial spasms. As Leah described: *"By the end of the meeting, I'm often holding my face to keep that side of my face from spasming upwards, so it's anything that involves concentration."* The use of such strategies provided personal protection of body image and temporarily addressed the fear of the spasms being recognised at work. But Leah acknowledged defeat with the words, *"So I gave up work".* However, the change in identity from a fully sighted lawyer or magistrate to being functionally blind and retired, with imposed daily limitations and financial loss, caused sadness as a sense of individuality and career identity was lost.

5.4.2 Subordinate Theme 2: Risk of danger to oneself and others

5.4.2.1 Day-to-day tasks present a danger

All participants described how even the most mundane task presented a danger. Here Janey explains: *"Cooking, if you have a squint… holding a hot casserole, you're trying to remember where the island is to put this down, it's burning your hands now, and if you miss it, it will be all over your feet. It's dangerous, and it ruins my carpets."* [laughs].

The condition causes danger, frustration and affected participants motivation to undertake the simplest task.

5.4.2.2 Danger to oneself and others

All participants carefully considered driving and the possible danger it could cause to themselves and others. However, some participants eliminated the risk, as Petal described: *"I don't feel my reactions are as fast, and that makes me feel less confident and not safe".* Without confidence in the ability to drive safely, most participants ceased driving.

However, a few participants adopted strategies to reduce the danger: *"I'm very conscious not to place myself or anybody else at risk, but driving has been reasonably okay, but I do prefer to drive in sunglasses"* (Delbert). Sunglasses were employed to reduce light triggering spasms, which could cause an accident.

Eradicating all risk was impossible as the simple act of walking or crossing the road could create difficulties, as Jey described*: "I gave up driving. Then, I found crossing the road a bit difficult, so… because you… you've got to cross the road, and all of a sudden, your eyes close on you."* Walking during a spasmodic episode presented an immense challenge to balance and affect safety. All participants resonated with this.

5.4.2.3 Reducing the risk of danger

Denise described how independence and personal safety was maintained: *"I find I am relying on touching things more. I am relying on putting my hands out just because I know where things are. It just makes me do things a little bit longer."* Personal adaptations were made to maintain independence and ensure safety, but the time taken to achieve each task was prolonged.

In contrast, public concern for safety led to being thrown out of the gym, as Janey described: *"The gym threw me out [laughs] because they were frightened that I might fall over, and they'd have an insurance claim."* It provided an understanding of how public scrutiny can lead to embarrassment and feelings of humiliation. Even though Janey's personal perspective is that the gyms' rationale was based not on safety but the fear of litigation, nonetheless, there was less motivation to access social settings without support.

5.4.3 Subordinate Theme 3: Loss of independence and confidence

Poor vision and co-ordination impaired the independence and confidence of most participants, which led to embarrassment, high levels of stress, and anxiety in public places. As Drey explained: *"My social life was affected because everywhere I went, I had to have somebody at my side to support me."* Debilitating spasms caused participants to need support. It also negatively affected their confidence: *"As your confidence goes, so everything else closes in around you"* (Judy).

5.4.3.1 Being dependent in social settings caused anxiety and risked friendships

Janey described real distress in having to ask for help in social settings:

> *"Going out for lunch, crossing the road, would you mind if I hold your arm?"… I sit down, and I can't read the menu, so I have to ask my friend to read the menu, and then because I've become anxious, I can't take in what she's saying. Soon friends don't want to go out with you anymore."*

These experiences highlighted how being dependent on others due to visual impairment in unfamiliar environments caused anxiety and a loss of friendship. In addition, being dependent on others and asking for help, although necessary, created feelings of inadequacy, embarrassment and caused anxiety in public. The reflections also indicate that friends and family members may not be prepared for their new roles as caregivers.

5.4.4 Subordinate Theme 4: Embarrassment

All the participants with the loss of confidence described those two factors caused their embarrassment:

- Eyelid spasms; and
- Visual disability during communication and in public settings.

5.4.4.1 Embarrassment during communication and loneliness in groups

Participants described that they felt embarrassed due to eyelid spasms and visual impairment during communication, and, even in groups, loneliness was felt. As Egna described:

> *"[It was] quite embarrassing if I was speaking to someone and constantly blinking all the time. If I go out with a group of people, I find it difficult to be a part of that because I tend to keep my eyes closed."*

Similarly, Judy explained how communication within a group setting was difficult:

> *"I can't see who is speaking, and strangely enough, if your sight isn't good, I think it affects your hearing as well. So, I think it's quite isolating, and being part of a group is a problem."*

Participants found it challenging and experienced loneliness as they struggled to participate in group conversations.

Even the act of listening triggered severe spasms*: "The whole act of listening to people… so I found that impossibly difficult, so I just basically avoided any sort of* *social setting"* (Leah). Participants were aware it was impossible to engage in conversation without being openly observed or subtly examined and avoided social settings to avoid the embarrassment of eyelid spasms and facial contortions being seen.

5.4.4.2 Embarrassment in social situations

Many participants felt a distinct sense of embarrassment due to eyelid spasms resulting in visual impairment, making them feel socially inept. As Janey described: *"I feel embarrassed [when I] do that, get in the café, and then I fall over somebody's bag on the floor or crashing about. It's not who I am as a person, it's my eyes making me like this"* (Janey). Egna also described the strangeness of the condition and would not go out to eat as *"if I am sitting to eat… then the blepharospasm is bad, which is curious!"* Participants felt embarrassed by their new persona and avoided social interaction. All participants, similarly, to Leah, described that their *"social life became non-existent"* (Leah). This resonated with all participants; the loss of their previous active social life and communal activities with friends was tough to cope with their unintended awkwardness.

5.4.5 Subordinate Theme 5: Isolation and public attitude

5.4.5.1 A lack of public knowledge caused scrutiny

A lack of public knowledge caused participants' facial appearances to be scrutinised in public. Drey described being 'checked out': "I just walked in, and they checked." Abnormal spasmodic closure of the eyes created a loss of confidence, and, without support, all participants felt embarrassed each time they were scrutinised in public. Drey perceived being at the reception of 'strange looks' was because members of the public "just don't understand the sight closure."

Similarly, Denise reflected on her perception of why she was inspected in public: "*You feel it, you feel that they must think 'what is going on, it is strange?'. People commented I was blinking a lot."* Denise's repetition of her words indicated the difficulties experienced while dealing with this aspect of her life. Being examined in a questioning manner or being commented on negatively caused distress and lowered mood.

5.4.5.2 Hiding strategies used in public

Some participants described various strategies to hide the spasm and prevent embarrassment, public scrutiny, and public attitudes towards their differences. Hiding strategies employed included avoiding eye contact, as Drey explained:

> *"When you're looking at people, they tend to look at you, and then you start to look down because you've got this condition, this blepharospasm, with the blinking of the eyes and the closure. It's quite embarrassing, so you tend to turn away from that person whom you're talking to, and so it's not very normal."*

Avoiding eye contact prevented the erosion of participants' confidence, moods, and self-esteem. Similarly, Denise also avoided eye contact: *"I got the hang of how to cope with it, to actually not look at them full-on, or sometimes, actually working by looking at people and focusing on their eyes did work."* Gazing at people entirely caused them to look away, which avoided the perception of scrutiny and helped participants to cope.

Delbert also chose to obscure the eyelid spasms from public view: *"My solution is to try and keep it out of the outside. I put the sunglasses on, I reluctantly put the cap on."* The word 'reluctantly' can be interpreted as people would rather compromise personal distaste to avoid the embarrassment of the public exposure of BEB, as Delbert has done by wearing a hat.

5.4.5.3 A divergent coping strategy to public scrutiny

Jey was the only participant to have a divergent view and explained his coping strategy: *“I just tell everyone that I have an eye problem.”* Anyone making uncharitable comments about his condition was told about it. In contrast Denise chose not to disclose this information although she was aware of the lack of public knowledge: *“I always felt that they were looking at me and thinking ‘well what is wrong? ’ and wondering whether to say ‘is there something wrong with your eyes?*

However, with low confidence, Denise, like most participants, did not dare to speak to strangers about the condition.

5.4.5.4 Isolation

All participants expressed that during their ‘incredible debilitating state,’ they experienced lost social richness and independence, became depressed, and faced embarrassment and public scrutiny, which eventually led to the loss of safety and comfort, hence isolation. As Petal described:

> *“It had actually got to the point where I was virtually housebound for the last couple of months, and I couldn’t go anywhere, and I wouldn’t go anywhere because it was too challenging for me and too tiring. ”*

This experience resonated with all the participants. Leah also highlighted the difficulties of coping to the extent *she “became a recluse. I didn’t want to see anybody, I couldn’t see anybody, everything was a terrible struggle”.* Even Jey, who considered himself easy-going and self-sufficient, could not leave home due to eyelid spasms and described sadly that he *“had to get people to do things for me, you know.”*

5.4.6 Subordinate Theme 6: Diagnosis

5.4.6.1 Diagnosis relief

For all participants, diagnosis provided a name: *“It was blepharospasm”* (Drey). “*Until I got here [specialist eye hospital] no one knew what it was*” (Delbert). A diagnosis validated the BEB’s existence, but more importantly, BEB could be treated to relieve the spasms. In addition, it provided understanding that the ‘incredible debilitation’ which caused self-degradation was neurological rather than psychiatric and did not represent the self-opinion *“I looked mad all day”* (Janey). Abnormal facial features were now understood as physical symptoms of the condition, focally superficial and not indicative of a mental illness.

5.4.6.2 Trauma of the diagnosis

Although most participants experienced relief, the receipt of the long-term diagnosis BEB and how the diagnosis was finally delivered inflicted trauma on a few participants: *“I was 48 when I was first diagnosed, yes, so pretty young, and it hit me like a brick actually, it affected my mood very badly, and I became clinically depressed and was prescribed antidepressants”* (Leah). The expression ‘hit me like a brick’ suggest the impact caused shock because of the way bad news was delivered. Likewise, Egna described how she felt pressured at the time of diagnosis: *“She [the doctor] said, ‘That is what you have got, and eventually you will need injections… it is up to you to wait’, and so that’s what I did”* (Egna). The accounts reveal a lack of care, empathy, and sensitivity, and no time was given to review the decisions or absorb the diagnosis’s impact.

Repeated medical opinion was sought for the correct diagnosis and treatment for more than one year. Despite this, the shock of the diagnosis and its delivery caused Egna to be indecisive about proceeding with the treatment. She was discharged from the specialist BEB clinic and was left her feeling dismissed once again. The experience caused her to lose trust in the clinicians' judgment. Therefore, a new referral to another specialist BEB clinic was sought from her GP.

The theme 'loss of social richness' represented the social difficulties experienced with BEB. Participants lost employment, but career identity became a danger to themselves and others, suffered anxiety in social settings, which led to a loss of independence and confidence and impacted some friendships. These losses resulted in embarrassment. A lack of public knowledge invited scrutiny and, even with strategies designed to hide the spasms, most participants' ability to cope was affected. The latter resulted in their self-isolation. While for many participants, a diagnosis of BEB led to relief, for others, the understanding that BEB was a long-term condition led to them becoming clinically depressed.

5.5 Superordinate Theme 4: Fleeting normality

"It is fairly fleeting" (Denise).

This theme is focused on BTX treatment stability following initial side effects. Improvement in both physical and psychological health provided hopes of a normal life. However, the temporary state of normality was considered fleeting, as well as the optimism gained as the effects of BTX waned.

5.5.1 Subordinate Theme 4: Fear of side effects

5.5.1.1 Side effects caused fear and anxiety

On initial commencement of BTX, most participants experienced improvement, but others did not. Leah communicated she was: *"A bit anxious about whether this is actually how it's going to be for the rest of my life"* (Leah). The experience of no effect from BTX caused participants to feel fear and anxiety for the future.

Similarly, the initial period of dose adjustment and the time the treatment would last cause Petal to feel anxious:

> *"I suppose, early on, when I was first having treatment, and we were trying to find, you know, how many weeks do I need to come and what the right dosage is, I became much more anxious again."*

The use of the word 'again' highlights the previous traumatic experience in primary care still affected participants. The experience of losing trust in NHS professionals' knowledge to care, diagnose and refer them for specialist treatment, which continued to cause fear. The attempts made by health professional to find the correct dose to eliminate all symptoms for a longer duration of time, created anxiety and fear regarding the length of time required to achieve treatment effectiveness.

The side effects experienced by a few participants caused fear of re occurrence. As Delbert explained: *"I really don't want the droop again. It caused me to fall. That was my worst experience, it was just an effect of the treatment and only in one eye."* A droop or ptosis can cause the eyelid to close and not lift or blink for weeks, which creates distress and accidents. However, Delbert showed understanding of treatment effect not being an exact science: *"Obviously there would be… trial and error."* The effect of BTX is not scientifically replicable between people. The latter discussion is important between patients and clinicians during the consent process to ensure the drug is altered per their reported benefit. Though such frequent modification was accepted, it still created anxiety.

5.5.1.2 Side effects and a loss of trust in health professionals

Once again, participants began to experience not only anxiety but a loss of trust in their treating clinician: *"I'm certainly very anxious a lot of the time, and two years ago I was between having injections [at a specialist eye hospital] before coming here"* (Judy). The reflections provided understanding that past trauma caused distrust, which caused her to feel anxious and without adequate knowledge. The doubts and anxiety this caused led to the change in a service provider. The experience challenged participants to forge a trusting relationship with their treating clinicians. Indeed, BTX side effects were seen as another battle for some participants to conquer, and reputable experts/service providers were sought.

5.5.1.3 Ongoing fear and anxiety of BTX side effects

As described by Denise, past side effects continued to cause fear and anxiety during and after the injection procedure: *"I still get nervous."* Delbert explained further that: *"There's also an element of worrying that I'll get a droop."* Nervousness and anxiety remained a constant worry to participants who experienced side effects. The most feared side effect was ptosis or upper eyelid droop, which impaired vision and safety. The fear continued for approximately ten days after the administration of BTX injection: *"If I was going to have a bad reaction, it normally used to come about ten days after I had the injection"* (Egna).

During that period, the participants felt constant anxiety over potential risk, which was worsened by the realization that a side effect could last for two to three weeks. Anxiety and fear remained a constant reminder of the traumatic experience of side effects, although optimum treatment effect was achieved without any adverse reaction.

5.5.2 Subordinate Theme 2: Restored health and positivity with BTX

5.5.2.1 Treatment satisfaction

Most of the participants eventually displayed the capacity to move beyond the negative periods of unstable treatment. All the participants eventually reported that they were satisfied with BTX: *“On my way home from here, I can tell it’s working. It is effective so quickly that it is just like I have been given a prize; it is worth having”* (Denise). The effectiveness of treatment provided hopes for the future and lifted moods. All of the participants’ accounts provided understanding that*: “Treatment here has been fantastic, yeah, like I say, the injections are working now, so that’s built my confidence up because I’m feeling a little bit more comfortable with my eyes, yeah”* (Drey).

5.5.2.2 Reversal of the incredible debilitating state

All participants who achieved optimum treatment expressed improvement in their physical and psychology health: *“With BTX, you are no longer pulling faces like you are a mad person”* (Janey). Without facial and eyelid spasms, participants’ mood improved, and physical abilities were regained. Psychological improvements were conveyed by participants’ description of regaining typical motivation: *“It doesn’t stop me from going out and seeing people anymore the way it used to earlier”* (Leah). Improvement was achieved: *“Once it’s [the treatment] clicked in and it’s at its best, it does make life more comfortable, absolutely”* (Delbert).

BEB was no longer thought to have a severe impact QoL with treatment. Their focus had now shifted to completing treatment and maintaining the quality of their lives. Petal described it *as "In a much better place due to the treatment I have here".* The words, tones, and body language suggest that participants are no longer depressed and have gained perspective – that the condition is treatable rather than life-threatening.

5.5.2.3 QoL and optimism was gained with BTX

All participants experienced improved QoL with BTX: *"Botox gives you the quality of life"* (Drey). Drey's view resonated with all other participants. Petal expounded that her*: "Lived experience now is different from how it was before."* Participants became cheerful and adjusted to their new lives with positive attitudes towards the BEB treatment experiences with treatment effectiveness. Denise also described her ability to cope now*: "At the moment, I can cope with that. It is fairly fleeting."* However, participants also recognised that although the effects were temporary and fleeting, BTX helped them to cope for a while.

With the temporary cessation of visual dysfunction and physical disability, abnormal facial expressions were negated, and participants regained confidence: *"[BTX] actually built my confidence up"* (Drey). With restored confidence, participants were no longer embarrassed or depressed about their facial expressions and were able to resume daily activities and so reintegrate into society: *"I have been able to resume my social life by and large, to the extent that I want"* (Petal).

All participants gained optimism with BTX and transitioned from being symptomatic without treatment to being in a state of anxiety and frustration in the early treatment period and finally being able to live normally. However, optimum treatment effectiveness was temporary and on a cyclical basis. This reaffirmed that participants' mood improved, and positivity was restored as they regained their abilities with BTX treatment and social and career identity.

5.5.2.4 New-found resilience

All participants' reflections provided evidence of a new-found resilience and that the effect of BTX, though short-lived, provided the means to cope: *"To be able to come to the clinic and get regular injections to relieve that is paramount. There's no way I could cope without them"* (Janey). All participants expounded that BTX caused them to regain physical abilities and lifted their moods while engendering hopes, positivity, resilience, and a willingness to fight.

Improvement in psychological health meant the participants began to openly acknowledge the change in their moods and their abilities to cope with the treatment. The condition was now considered marginal in comparison to other conditions, and participants were grateful it was treatable: *"Oh, I wish I hadn't got this, but just appreciate the fact that I have got a condition that is treatable, but I suppose it's marginal compared to other people's problems"* (Denise).

5.5.2.5 New-found understanding of BTX treatment duration

All participants expressed understanding of BTX actions and their individual treatment durations. Jey described that: *"Up to four months is outstanding".* Judy added further understanding: *"If I can come every eight weeks, that would be a huge advantage for me"* (Judy). Participants remained anxious to receive timely treatment to ease the potentially debilitating symptoms, and that feeling resonated with all participants. It was necessary: *"So, at the moment, while I'm on an effective and stable treatment regime that I'm having a very regular intervals, the impact on my life is less than what it was earlier"* (Petal).

5.5.3 Subordinate Theme 3: Restored career identity

With physical abilities restored, a few younger participants – namely Denise, Leah, Drey, Nadia, and Petal – could re-establish professional identities, feel a sense of identity, and recognise their previous selves. The new goal is to prevent a decline in their health, similar to Drey, Leah, Nadia, and Petal described: *"Work full-time on principle because I feel that it may negatively impact my condition".* Although Leah lost her own thriving business, she was able to gain part-time employment by working as a marketing consultant in another firm. Reducing work diminished the need to walk, talk, concentrate, read and use bright screens excessively.

5.5.3.1 Restored physical ability

Without being physically limited, Delbert is once again able to drive, albeit with sunglasses, can watch television, has taken up bowling, and is attempting photography with a digital camera as his flickering has stopped. All participants were extremely satisfied with regaining their abilities to undertake various hobbies. Nadia was once again able to teach music. Jey was happy to cross the road without his eyes closing. After more than a year, Petal only saw a half of a theatre show, but it was considered a significant accomplishment. Leah was again able to see, go for runs, and even take long walks. Drey was able to resume exercise. All the participants believed that the BTX clinic could save lives.

5.5.4 Subordinate Theme 4: BEB clinic is a lifesaver

All participants gained effective treatment and positively considered the specialist BEB clinic and care provided as: *"A lifesaver coming here, there's no doubt about that at all. Once I'm here, the stress level drops. I have great confidence in you."* (Judy) The clinic was seen as a beacon of hope, and treating clinicians were trusted to provide optimum treatment to maintain physical and psychological health.

5.5.4.1 New trust in BTX, the organisation and the treating clinician

All participants' words expressed confidence in the organisation, the clinicians, and the treatment provided: *"It is a great comfort to know that I am coming here and, you know, that… that gives me the confidence to know that, no matter what happens, that I always got such good treatment."* (Egna) Judy explained: *"The treatment obviously that I have, which I feel very privileged to have, and everybody here is exceedingly pleasant and helpful."* It provided understanding that the participants believe they received adequate treatment, which eliminated their physical and psychological disability from trusted and caring professionals.

Continuity of care was also valued, as Leah elaborated:

> *"Well, I think what makes the experience good is that I know the people here care. I know I'm in good hands, you know, I feel safe, and that's important. I've heard lots of people come and say, 'Oh, you know, I am seeing [the nurse consultant], aren't I?' and I think continuity of care is quite a big thing for patients with this condition."*

It showed the participants had developed confidence in the proficiency and experience of the treating clinicians. They appreciate consistently consulting, sharing knowledge, and extended efficacious treatment that reduces anxiety.

5.5.5 Subordinate Theme 5: The return of the 'incredibly debilitating state'

5.5.5.1 The waning effects of BTX

Fast deterioration of the condition caused Delbert to be described*: "As I get towards the end of a cycle, it deteriorates quite quickly."* Denise adds further depth: *"Everything is fine until I notice the effect has worn off, but then quality of life deteriorates considerably."* The loss of treatment effectiveness affected vision, physical abilities, and psychological well-being, which resonated with all participants. Nadia described:

> *"So, once it's getting near the injections wearing off or something, I find it very difficult to read the music because obviously, I'm having spasms all the time, affecting the vision."*

It also provided understanding that the mood also changes with a decline in physical ability. Debilitating symptoms removed the optimism of ever living a normal life.

5.5.5.2 Loss of positivity

The positivity expressed previously during the period BTX was effective. It faded as soon as the treatment wore off. The patients' tone lost optimism: *"I don't get no problem, [for] four months" (Jey).* There is an eagerness to have timely treatment: *"I'm… more anxious about the appointments and sort of fixated on trying to get it at a time that suits me."* (Petal) Timely treatment delivered the means to avoid a relapse in visual function, physical limitations, and loss of employment, social embarrassment, isolation, and depression. Janey's reflection highlighted the low mood experienced when BTX wears off and spasms recur: *"At your most stressful moments and you need it desperately to go away, it won't"*.

This account explained how psychologically stressful an acute episode is and the need for symptom resolution. Similarly, Nadia's words resonated with all participants: *"When it [BTX] is wearing off, I find it very difficult."* Patients reported feedback to prevent untimely scheduled treatment to avoid the difficulties of an 'incredibly debilitating state.

The theme 'fleeting normality' provided a deeper understanding of the participants' treatment experiences with BTX. The initial side effects experienced led to fear and anxiety. Faced with adverse effects, participants again lost trust in the medical professionals administering treatment. Ongoing fear and anxiety, even after treatment stability was established, provides an understanding that the experience of side effects caused trauma. With BTX treatment, satisfaction, health, positivity and career identity were all restored.

The specialist BEB clinic is seen as a lifesaver and treating clinicians were viewed as proficient. Participants became aware of their treatment duration as the effects of the medication wore off, and both the 'incredibly debilitating stated' and the loss of positivity recurs. Participants once again lost positivity as BTX waned. Psychological decline resurfaced, with emotions of fear and anxiety needing to be treated.

5.6 Superordinate Theme 5: A battle for adequate care

This theme represented the impact of untimely care on QoL and participants' battle for adequate care to prevent the negative reversal of physical and psychological health: *"It feeling like a battle just to get adequate care to manage my condition."* (Petal)

5.6.1 Inability to cope during the incredibly debilitating state

Although all participants agreed that when an acute attack happens: *"I think Botox treatment should be available"* (Egna) since they were unable to cope with the 'incredibly debilitating state.' The decline in physical and psychological well-being and its impact on family life challenged participants' ability to cope:

> *"If I have to wait an extra few days or a couple of weeks because of lack of clinic time or holidays or whatever, it interrupts my flow as a patient. Those two weeks are tough to cope with, not only for me but also for my family as I fall over, drop things more, and squint more. It's much more challenging."* (Janey)

The positivity gained during BTX effectiveness is lost as participants' moods change, as Denise signified with the question *“why me.”* Life was considered a struggle and without treatment, as described by Drey:

> *“It would be more efficient if I could have treatment because when you’ve got this condition, and you’re struggling, you know, when you’re on a low, as people will know, you need it, you need it, it’s not good.”*

The participants’ account highlighted a greater understanding of the impact of visual and physical disabilities and the psychological challenges and ‘desperate’ calls by participants for timely treatment to avoid anxiety, depression, social isolation, embarrassment, and social stigma once again.

5.6.2 Subordinate Theme 2: Overbooked clinics, fear, anxiety, and frustration

5.6.2.1 Delayed access and devout attendance

Fear of BTX wearing off and experiencing delayed access to the specialist BEB clinic was a significant concern for participants who adopted a committed approach to attendance, as explained by Leah:

> *“The fear of the botox wears off and then having to wait, you know, potentially a long time until we can get back in and have some more, so that’s what keeps us sticking religiously to our appointments and, you know, I religiously guard it.”*

Similarly, Petal described being: *“Ready and waiting for these appointments because these appointments are hard to get and to get hold of when you need them.”* The fear of not accessing care when required caused participants to demonstrate compliance by never missing a BTX clinic appointment.

5.6.2.2 Scheduling BEB clinic appointments, anxiety and frustration

The process of scheduling an appointment caused anxiety while waiting for clinical consultation, as Petal explained:

> *"It's probably to do with anxiety rather than mood. So, when I was just sitting now, I was thinking, 'Oh, I've come later to the clinic; there are all these people before me. What if I can't get the appointment day that I want when I come out?'"*

A constant state of anxiety over the possibility of not receiving the recommended timely appointment due to a lack of capacity in the specialist BEB clinic was a worry for all participants. Petal's account showed that some participants' anxiety began in the clinic's waiting room

Any failure to receive the correct follow-up BTX appointment caused the participants frustration, anxiety, and fear, as Janey explained:

> *"When the appointment's over, the normal protocol is that I go to the reception and I book the next appointment, and that's the way it's supposed to happen, and that might be ten weeks/12 weeks. It's normally two to three months away. It is quite reassuring going home with a piece of paper that says you have got an appointment on such and such date, but disappointing obviously if it is ten weeks or whatever else."*

To avoid disappointment, the appointment letter was often requested immediately following treatment.

However, the varying clerical booking approach caused frustration. According to Delbert:

> *"I sometimes find it a little bit frustrating when they just want to give you the date and just hope it's the right one because I'm quite anxious to get treatment every three months."*

Participants' use of the word 'hope' indicated that it might not be as predicted. Similarly, Petal expanded on why this caused anxiety: *"That's the trouble when the letter comes to you, it's usually ten days afterward, it's going to be ten days further than normal."* Being left to wait for a letter that was often incorrectly timed prolonged anxiety and often led to disappointment.

5.6.3 Subordinate Theme 3: Desperation for treatment

5.6.3.1 Desperate calls were met by indifference

Being acutely affected, unable to cope with the loss of vision, dependency on others, isolation, embarrassment, guilt, and depressive moods, participants desperately requested BTX treatment sooner than planned, but calls were met by indifference. Janey described being desperate, with calls for treatment, but sometimes being unable to gain an appointment: *"I tried to, but it was tough to get through and to make any extra appointments because there just weren't any."* For others, such conversations proved difficult and caused frustration. Acutely affected, Drey described his interaction with members of the clerical booking team:

> *"I have phoned up for an appointment, yeah, it's just been so hard, too hard, yeah. And I've pleaded with them, but they only can slot me in at a certain time, but then, you know, when I'm desperate on the phone, and they just don't waver."*

Participants' felt they needed to 'plead' and 'beg' for treatment which lowered their mood. Similarly, Nadia described that she became*: "Frustrated, you know… well desperate, you know, I need the treatment, there must be something you can do about* *it. I think like that, really."* Participants' accounts reflect anger at the indifference and frustration at being refused care and treatment without an explanation.

5.6.3.2 A lack of compassion and empathy caused frustration and anger

On reflection, participants showed distress at being refused access to the BTX clinic. Desperate participants 'pleaded' as they battled for adequate care, but instead faced a lack of empathy and compassion, as Jey experienced:

> *"I have cancer, you know. I missed the treatment because I had to go to the hospital for cancer. I had to miss it, and I didn't realise that I had an appointment that day. I had it all around the same time. And I phoned and begged them, and they said 'no, you'll have to wait another six months."*

This account added deeper understanding to unique situations where a lack of compassion was felt during communication with the booking team.

In addition, the refusal for a requested appointment was not delivered in a caring manner, and no rationale for the decision was offered. Like Jey and other participants who refused treatment, Delbert was frustrated and expressed *anger "to be told there's nothing we can do for you"*. Although devastated with the response they received, some participants remained persistent in contacting the hospital multiple times for an earlier appointment.

5.6.3.3 Unresolved complaints

A singular complaint made by Janey to the PALS department did not prompt an earlier appointment:

> *"Awful. We have been to PALS and asked not to complain about the clinic because they are fabulous, they do an excellent job, and the injections are so helpful. It's not to complain. It's just to ask what the problem is? Why aren't there enough appointments? Can we do anything as patients to help this? Nothing was resolved, and eventually, we just fell back into whatever could be offered."*

This account resonated with others and provided understanding that specialist BEB clinics are often overbooked, with no capacity for emergency slots.

5.6.3.4 Overbooked clinics discouraged timely treatment request

A few participants, though desperate, accepted defeat as they considered request for timely treatment would not be granted. Therefore, Egna explained*: "I just grin and bear it."* Although Judy said:

> *"I haven't asked here if I can have an appointment earlier than has been given, but psychologically, it would be a huge boost for me, and it would make it much more manageable, and I would be delighted with that."*

Both Egna and Judy rated their chances of getting an appointment as low as the clinic 'always looks fully booked'. Most participants who contacted the hospital sooner than planned did not gain an earlier appointment.

5.6.3.5 The impact of untimely care and treatment

Most participants commented on the impact of untimely care and treatment and describe it as 'quite unacceptable' and the service as being 'not good' and 'awful'*: "To be suffering for five to six weeks until your injection comes up is quite unacceptable"* (Janey). As Drey explained:

> *"It would be more efficient if I could have the treatment because when you've got this condition, and you're struggling, you know, when you're on a low, as people will know, you need it, you need it, it's not good."*

Participants' use of the words 'suffering' and 'struggle' provided understanding that the service previously accredited with being 'excellent' was now failing. In addition, the lack of timely treatment impacted the health of participants.

5.6.3.6 Assertiveness won the battle for adequate care

A few participants were assertive and won the battle for adequate care. Petal described being *"anxious and probably quite distressed and sort of down in mood because of it feeling like a battle just to get adequate care to manage my condition. I had to be quite assertive".* Undeterred, some participants refused to give up and was rewarded for assertiveness and persistence. Denise was quite distressed and called the hospital numerous times and was finally given a timely appointment. Her account succinctly expresses the benefits of how winning the battle helped to maintain QoL and was synonymous with 'winning a prize'. Unable to cope beyond a week without BTX, Denise described the reduction in QoL and the frustration experienced:

> *"I can manage for a week to get by like this because it does affect the QoL. It does make things harder and frustrating, but the treatment is beneficial. It [is] important to say I did get treatment."*

Assertiveness contributed to the receipt of a timely appointment. However, the fear of having to suffer without treatment was the reality of all participants.

5.6.3.7 Management of BEB clinic was unacceptable

All participants considered the management of the specialist BEB clinic as ‘unacceptable’ and Janey blames funding: *“It falls to funding this condition is very challenging to live with, management just don’t understand.”*

From the participants’ perspective, management did not comprehend or was unaware of the challenges they endured due to the impact of untimely care and treatment. Furthermore, they were willing to undertake any additional discussions, offer recommendations to improve the service. They vowed to do *“anything that can be done to keep the treatment regular and help me”* (Denise). A focus group discussion was organised to capture the collective views of patients on how the service can improve.

The theme provided a deeper understanding of how untimely care and treatment during an acute episode of the condition rendered participants to the return of an ‘incredibly debilitating state’, which they tried to avoid with desperate calls for treatment. Instead, many were denied access to care and treatment and shown very little empathy and compassion, which caused fear, frustration, and anxiety. Only the more assertive participants won the battle for adequate care and treatment. Participants blamed funding and management lack of understanding regarding BEB and offered to help improve the service to improve their care consciously.

Conclusion

The insider perspective of ten participants’ lived experiences offered a deeper understanding of the impact of BEB on their daily lives, with particular emphasis on treatment and care within the specialist BEB clinic. The lived experience of people with BEB in the participants’ words were often impacted by delayed diagnosis due ‘no knowledge’. The delay causes people with the condition to experience ‘incredible debilitation’ visual, causes functional limitation and decreases psychological health. Unable to cope with spasmodic closure of the eye and social perceptions cause ‘a loss of social richness’ as isolation was often sought and a diagnosis of depression was given before BEB. In a few cases, antidepressants, counselling and cognitive behavioural therapy was required. Nonetheless, a diagnosis of BEB offered relief.

A referral to a specialist hospital still presents some challenges which involved coping with the side effects and gaining timely treatment. Botulinum toxin treatment (BTX) was considered essential to improving QoL for people with BEB. All participants demonstrated positivity when discussing the effects of BTX and this extended to the treating clinician and the hospital. However, the effects of BTX are temporary, causing ‘fleeting normality’ as the treatment wears off in many cases before the planned three-monthly cycle. Overbooked clinics prevented a few participants who were extremely affected with spasms from even attempting to gain an appointment. In many cases participants who contacted the hospital felt the need to ‘battle for adequate care’. However, only the more assertive patients were able to win the battle to gain the treatment and care required to ameliorate their physical and psychological symptoms. The diagram below with superordinate and subordinate themes summarises the research findings.

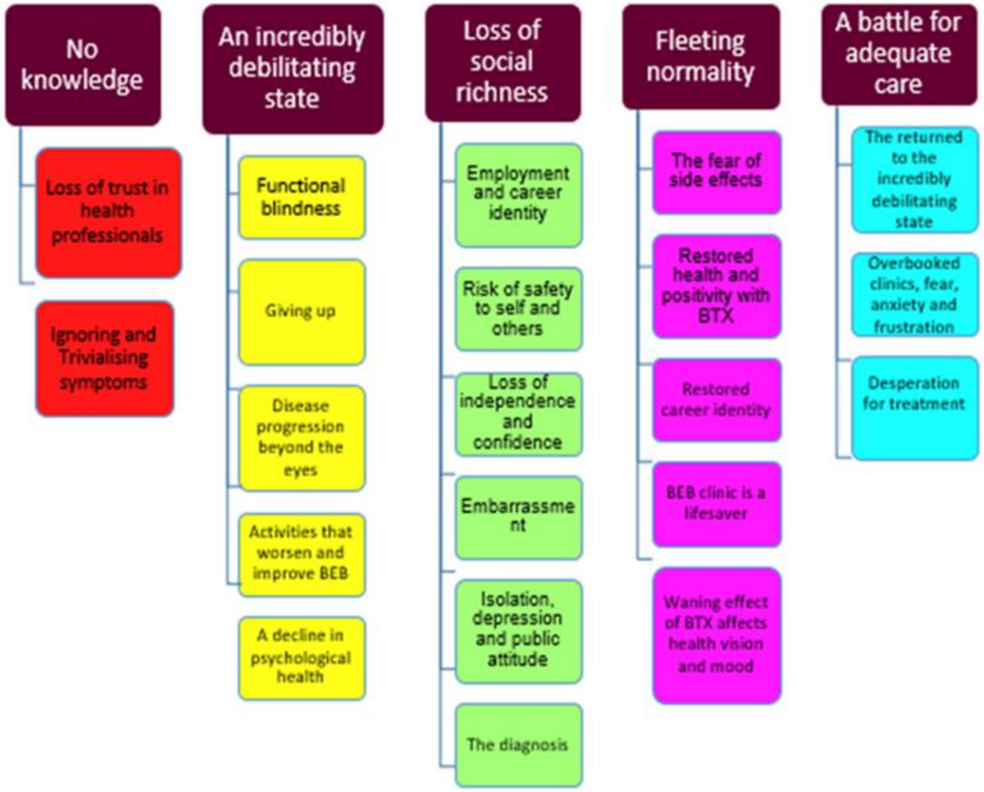


Figure 9: The superordinate and subordinate themes of BEB

Chapter 6: Focus Group Discussion

This chapter presents an FGD that follows all participants individually voiced but collective aspirations to improve the specialist BEB clinic. The FGD is representative of Phase 2 of the research and is organised to solicit the participants' combined response to improving their world. First, the study participants will be described. The report summarizes the recommendations within the themes generated by the discussion. Incidentally, it also addresses the third objective of this study: to inform service improvement or redesign that responds to the needs of BEB patients.

The FGD occurred at a specialist eye centre on 31st October 2019 at 10:30 hrs. All the participants who took part in Phase 1 were invited. However, only eight out of the ten BEB participants attended. In attendance were:

- Denise, 65-year-old female university lecturer
- Egna, 88-year-old retired female teacher
- Judy, 72-year-old retired female magistrate
- Petal, 56-year-old female social worker
- Janey, 65-year-old female retired medical secretary
- Leah, 54-year-old female company director
- Delbert, 69-year-old male retired lawyer; and
- Nadia, 55-year-old female music teacher.

The objective of organising the FGD was to augment the qualitative breadth and depth of understanding of what needs to change to inform OPD improvements through service redesign.

The lead nurse researcher, a doctor, and a quality improvement lead were invited to the FGD. This was an opportunity for them to understand how the quality of care could be improved through research and patients’ feedback. The participants discussed BEB in the context of the research carried out at the BTX clinic, as well as the services of the clinic, which is divided into the leading clinic that runs on Mondays with two doctors and a nurse-led clinic (NLC) and is operationalised by the nurse practitioner/novice researcher on Fridays.

The participants in the group wanted to also talk about access to care, respect, information and communication, physical comfort, and emotional support. Patient feedback aimed to inform quality improvements by increasing the healthcare system's responsiveness to the needs of BEB patients. This FGD highlighted four potential areas for change. Consequently, the full report on the discussion contains the themes generated from a broad discussion on healthcare related to the focus group members’ everyday experiences in the BEB clinic.

A summary presentation of the themes the participants identified and the discussion around them through the FGD process highlighting five areas but only four areas for potential change since the NLC was considered a role model, as shown in Figure 10.

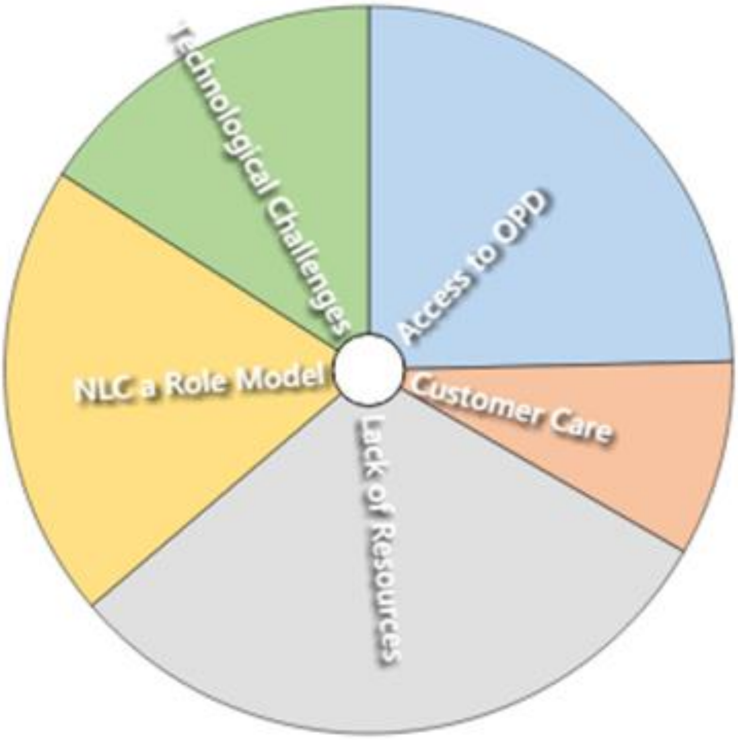


Figure 10: FGD themes

6.1 FGD Findings

6.1.1 Nurse-led clinic (NLC) as an exemplar model

The FGD demonstrated the disparity between the NLC and the leading clinic; for example, as Leah explained: *"I like the nurse-led clinic. It is quick and efficient. Also, I always have the same nurse."*

Along these lines, Petal stated:

> *"It would be nicer if the Monday clinic ran like the Friday clinic, which is, I find, a much more pleasant clinic to come to because, I don't know, maybe there are fewer patients. It seems to run much more smoothly. You're not usually seen too long after your appointment time, if you know, maybe on time as well. So, I would say that the Friday clinic is a good example."*

The NLC was seen in a positive light, and even the body language, smiles, and relaxed manner in which the participants relayed the related information provided affirmation that the clinic was a good role model of care due to its promptness, efficiency, and knowledge regarding continuity of care service that is provided:

> *“It upsets me when they change my appointment for some reason, and I don’t go back to the nurse clinic. The doctor’s Monday clinic is slow, and half the time, they don’t know what’s going on. They just need more clinics. (Nadia)*

> *“I see the nurse a lot. I appreciate that because she is knowledgeable about the condition and my case. I think continuity of care is quite a big thing for patients with this condition.”* (Denise)

The participants resent being placed in the Monday BTX clinic due to long waits and requested more clinics to see patients. In essence, they expected that the BTX clinic should also have well-trained and knowledgeable staff and deliver its services on par with the services delivered by the NLC.

However, the main areas of concern were the following:

- Lack of resources
- Uncertainty about access to OPD treatment and care
- Technological challenges; and
- Inadequate customer care.

6.2 Lack of resources

The participants understood that the greatest challenge to improving the service was healthcare resources or its lack thereof:

> *“So, in an ideal world, it would, you know, if you were having an acute episode, if I were having an acute episode, I would want to be able to come in and see somebody and get some treatment.”* (Petal)

> *“Yes, so probably more money, more clinics, and yes, running a bit like the Friday one. It feels like a better experience, really.”* (Leah)

> *“Well, I presume it falls to funding, and whoever holds the purse strings needs to understand that this is not a little problem that you can put a drop in your eye, and all is well.”* (Janey)

Furthermore, the participants felt strongly, and repeated similar concerns previously voiced in individual interview. It was believed that the onus was on those responsible for the management of the OPD to invest more funding into caring for BEB patients. Some of the participants called for having more outreach clinics to offer BTX closer to home:

> *“Well, certainly more clinics are needed to enable more patients to be seen, and perhaps those who are already being seen to allow them to have a slot when they are desperate when they are in need in an acute period. So, more clinics and possibly even satellite clinics to save people from traveling so far.”* (Janey)

The hospital has many outreach sites. However, there are only two locations that treat BEB patients. There was consensus among the participants that their condition's acute difficulties should prompt immediate treatment in an ‘ideal world’, but they acknowledged the challenge regarding lack of resources. However, there were calls for more funding to develop more clinics to remove capacity issues and provide training to ensure that the patients benefit from the same experiences and standards of the NLC, which runs on Fridays.

All the participants felt that more clinics, rather than the current two, would allow those who call the hospital to have treatment within three to four days in acute situations. Creating multidisciplinary clinics where knowledge can be shared among staff and patients can access psychological support. However, lack of resources was the cause for poor access to OPD care and treatment.

6.3 Access to the outpatient department

All the participants called for better access to treatment and care during acute episodes of the condition to improve declining health:

> *"It would be nice if you could phone up and come in maybe with 3 or 4 days' notice, sort of emergency clinic like you would have at the dentist." (Delbert)*

> *"It would be nice to be able to say, actually, I need the injections now', even if it were within a couple of days, that would be helpful because immediately you feel, 'oh, that is good, something is going to be done about it, and it is okay'. So yes, to be able to ring up and book your appointment."* (Denise)

It was felt that access to the BTX clinic would improve the patients' vision and psychological comfort. However, the abrupt cancellation of the appointment can occur for several reasons. For example, overbooked clinics, shortage of staff, and sickness but the unexplained reason was found to cause distress and difficulties to participants who were unable to return to the NLC as they preferred the services there: *"It really* *upsets me when they change my appointment for some reason, and I don't go back to the nurse clinic."* (Nadia)

A lack of prompt access to treatment and care in the leading BEB clinic on the required day was another concern that caused the participants to deem the clinic less than efficient. The clinical journey commenced with the participants queuing for a prolonged period to check into the BTX clinic. Some participants reported that the wait was tough since they suffered from the poor postural balance due to neck dystonia.

However, a few participants felt that the new self-check-in kiosk machine should eliminate that issue, but the patients need the training to develop trust in the new system: *"The checking-in is a new system that I quite like actually, although there's a degree of lack of confidence."* (Delbert)

In addition, waiting to be called by the doctor could be tiresome due to severe delays. On a few occasions, some of the participants reported being informed that their clinic notes were missing. The choice was either to risk side effects due to the administration of the wrong dose of treatment or to reschedule the appointment hoping that the notes would arrive. The participants have called for addressing the lack of knowledge, poor continuity of care, and the challenge of accessing care.

A lack of holistic care was another area of concern, particularly in cases where the participants had more than one dystonic condition and sought care and treatment in two different secondary care facilities. Nadia goes into this:

"To be honest, I think it should be broadened out, you know. Botox is being used for treating something else. I don't see why one hospital cannot provide the whole service because, you know, it is a linked thing, I think, the chronic migraine."

It was felt that collaborative thinking was necessary to broaden the treatment to include other conditions. For example, all treatments such as migraine, for which an injection is required to be administered on the head, cervical dystonia for which neck injections are required, or the Meige syndrome a phenotype of dystonia where pharyngeal or tongue injections may be necessary could be offered under one roof.

6.4 Recommendations for improvement: the redesign of access to the clinic BEB clinic

Participants asked for the patients to be trained by the staff on the use of the check-in kiosk, which the former wanted to be continued to eliminate long queues and help the patients develop trust in the new system. There was consensus among the participants that the solution to access holistic care and treatment requires more specialists/trained injectors to establish more NLCs. The NLCs can have a multidisciplinary team to meet the demands of the patients concerning accessing care at short notice. It is only if it is armed with the necessary capability that the team can meet the needs of the patients who live with more than one dystonic condition and offer psychological support as required. In addition, it was suggested that more BTX clinics at outreach sites would eradicate the demographic disparity.

To summarise, the participants called for the distribution of funds to be geared towards establishing more BTX clinics and the training of injectors at the established sites, along with outreach or satellite units. In addition, the participants asked for a holistic, multidisciplinary clinic that involves a mental health specialist, such as a counsellor or psychologist, and other dystonic specialists to exchange knowledge and advice to treat more than one dystonic condition as required.

6.5 Technological challenges

The participants were asked how they would prefer to book their appointment, whether through telephone, app, or online, and why. All the participants stated they would prefer a telephone call: *"No, I am not computer savvy or own a smartphone, maybe younger people might like it. A telephone call yes."* (Petal)

Although one participant stated that flexibility would be preferred while communicating with the clerical team regarding appointments. Others, such as Denise, stated they would prefer to continue having an appointment letter:

> *"For me, I wouldn't mind, but this condition affects a lot of older people who would find it difficult to use a computer or an app. As long as there is flexibility to choose, I would be happy."*

Egna also expressed this view: *"No, I would rather have an appointment letter or call than disturb someone."*

Most participants were of the older generation and found technology more challenging. However, younger patients with smartphones or computers may find this means of contacting the hospital more accessible. In addition, leaving the BTX clinic with an appointment letter was reassuring to others. Therefore, various flexible booking approaches should still be available.

6.6 Recommendation for improvement

It was agreed that no one booking approach is convenient, but to have the ability to choose any method whenever emergency access to care is required would cause minor panic and worry. Many participants reported not being technologically savvy, though they prefer to telephone fear of a negative interaction with staff was high.

6.7 Customer care

Telephoning the clerical team caused anxiety as the participants were concerned about being rudely spoken to or a lack of customer care. There was an instance of poor customer care by a clerk, which a participant who was left waiting in the queue reported. The participant's reflection demonstrated that the conversation overheard breached privacy, and there is an apparent lack of customer service and need for improvement in this area. The negative interaction led the participants to state that the doctors and nurses care, but the clerks do not.

> *"I just felt that the receptionist could have said, what clinic do you want, can you go and book your appointment at so and so rather than leave me standing there. And as I say, the poor girl had a problem that I was sitting listening to it which may not be the cleverest thing, bearing in mind that it was about missing notes, so that's not the sort of conversation that should have been going on, I think, in public. A little bit more customer awareness could be better from the reception side, the nurses and doctors care, but the clerks don't."* (Delbert)

In another case, how the clerical team members spoke to the patients in one instant created panic. Each time the need to call the clerical staff arose, the participants became worried*: "I panic and worry whenever that happens and when I call up the clerk, they are often rude"* (Nadia).

Although all participants called for better communication and interaction with clerical team members, they requested direct contact with the treating injector to feel better.

Another area that the participants improved was the injector's communication. However, even this area would require investment:

> *"Yeah, by telephoning, keeping in contact, obviously with the patients, asking during their reviews, how they are every time they [attend] come here, keep asking things about how you could do it better, also obviously just keep us updated with all the information that's going on like research."* (Judy)

The communication by participants craved for was geared mainly towards understanding the effectiveness of the treatment before and during its administration. Offering two appointments to the patients, one to the BTX clinic and the other virtual, by telephone or maybe even video, was preferred to assess the patients' symptoms and expedite early care earlier, where necessary. The participants called for the establishment of open communication to relay information on BEB innovation and research.

Yet another area identified for funding was the training of injectors to ensure the delivery of excellent care, including accurate record-keeping and good communication:

> *"The injector has said to me when well where do you have them, and how many do you have? And I'm like, well, I don't know, you should know. I shouldn't be… I can't tell you that. Another time my notes were lost. So, I think training and effective record-keeping so that patients aren't put in a position of being asked things that they shouldn't have to know or don't know."* (Nadia)

Calls from research participants for further training for injectors is essentially a call to invest in people. Training nurses provide a sustainable long-term plan as junior doctors are rotational and do not deliver continuity of care. The creation of a digital infrastructure would provide the technological means of record-keeping, which would negate the offsite storage of paper notes.

The participants were asked to be contacted before visiting the OPD about any missing notes to provide them the choice of being optimally treated with the correct dose of BTX at the correct injection site or about rescheduling the visit when the notes are found. It was felt this was a meaningful conversation that should take place before they leave home for the treatment. Injectors must also be trained to ensure the treating clinicians follow the correct documentation process and know about the required treatment.

In addition, there was a call by the patients for greater engagement with them to ensure that their views were heard. While the FGD on that day provided valuable information, there was a call to establish regular FGDs. Such discussions should aim to hear from the BEB patients about their experiences and concerns and hear from the experts regarding new research and innovation concerning BEB and its treatment.

6.8 Recommendation for Improvement: Customer Care Training for Clerical Staff

The participants called for more respect from the staff and were treated kindly and with empathy. They reiterated the importance of customer care training to provide a new understanding of BEB patients' care. This could be enforced through customer care courses for clerical staff. It would enable patients to be contacted and given information about missing notes, which would save time and money traveling to the hospital.

However, to be able to have a respectful conversation over the phone, rather than feel rudely attacked when already feeling low, would reduce the panic and anxiety felt by the patients when seeking an appointment. There was a recommendation to improve communication by virtual review or telephone to address matters that challenge care, for example, missing notes or to review a patient’s condition and expedite care. The participants also called for a regular FGD to provide information on BEB research and innovation, promoting knowledge and supporting a sense of community.

Conclusion

This focus group discussion presented the lived experience of BEB patients within the clinical setting and offered recommendations to improve the service design. This included a call for more BTX clinics and the training of injectors at the established sites, along with outreach or satellite units to prevent geographical disparity. In addition, the participants asked for a holistic, multidisciplinary clinic that involves a mental health specialist, such as a counsellor or psychologist, and other dystonic specialists to exchange knowledge, offer advice and treat more than one dystonic condition as required.

The participants also asked to be respected by staff and to be treated kindly and with empathy. They reiterated the importance of customer care training to provide a new understanding of BEB patients' care. Participants which to be seen promptly as an emergency dental appointment for the severity of their symptoms to be assessed virtually or by telephone. In addition to communicate missing notes to avoid a wasted costly and regular FGD to provide information on BEB research and innovation and a sense of community.

Chapter 7: Discussion

The purpose of this chapter is to interpret the results of the qualitative exploration of the impact of BEB, outpatient treatment, and care on the lives of patients and to inform service improvement through a redesign. The significant findings are discussed critically with the known literature in Chapters 1, 2, and the knowledge uncovered in Chapter 3's meta-synthesis, Chapter 5 findings, NHS quality framework, and IPA. The use of IPA provides originality. Of critical importance, it marked the move away from studying the condition as a biomedical model of disease and illness that is predictable in recognition of its complex constructed nature made possible through participants' self-reflection and the researcher's analytical interpretations. It also discusses the recommendation improvement for participants in Phase 2 of the study. Finally, the chapter outlines the study's limitations and areas for future research. Importantly, it answers the research questions:

Question 1: How does an acute episode of BEB affect a patient's QoL?
Question 2: How does access to outpatient services during an acute episode of the condition impact the patient experience?
Question 3: What needs to change to ensure outpatient services for patients with BEB are responsive to their expressed needs?

How was Critical Interpretation of the Findings Undertaken?

IPA philosophies underpin the interpretation of the findings: phenomenology, hermeneutics, and ideography, which aided interpretation. Phenomenologically, examining the lived experience of BEB with the researcher's lens of IPA provided a deeper understanding of the condition as meanings were created through participants' self-reflection. The researcher then holistically co-created these, who simultaneously sought to deeply understand the participants' words, thoughts, and actions about an event, making it hermeneutic. However, ideographically the focus on individual experiences, in conjunction with NHS quality of care dimensions, were instrumental to the thorough exploration of the patients' care.

The study fundamentally abides by the founding principles and values of the NHS (DOH, 2016) that closely connect the communities and people it serves, and the staff who work for it, with the organisations who operate within it to ensure high quality and standards. Individually, BEB patients are service users and should be empowered to take more control of their care by being empowered by clinicians to be more involved in the decisions about when and where they receive services.

The experiences of BEB in the clinical setting were also examined with a practitioner's lens utilising the six dimensions of quality healthcare; patient-centred care, the efficiency of service, safety, effectiveness, equity, and timeliness (WHO, 2006). The NHS has clarified what quality means in its definition, that care should be safe, avoiding harm, effectively based on clear evidence that produces benefits. Exploring what quality means to patients, includes a positive experience, caring staff, being treated with dignity, respect, compassion, responsiveness, and patient-centred care.

Sustainable using resources responsibly and efficiently proves fair access to all and according to the population's needs. The service should respond to people's needs and choices, enabling equal partnership in care. It also elaborates that service providers should be well-led, open, and collaborate internally and externally be committed to learning and improving. Lastly, it should be equitable to provide care that does not vary in quality because of a person’s characteristics (DOH, 2016).

Summary of Key Findings

Five superordinate themes emerged from the interpretative analysis, which helped facilitate understanding of questions. The themes were not only based on prevalence but also on the articulacy and immediacy in which the participants’ words exemplify the themes. The first three themes – **‘no knowledge’**, **‘an incredibly debilitating state’** and a **‘loss of social richness’ –** represent the participants’ experience in primary care. Significantly, it captures an acute episode of the condition, which was often overlooked but importantly answered the first research question; how does an acute episode of BEB affect QoL? A lack of knowledge diminished participants' experience, among health professionals.

The following two themes were found to represent the specialist BEB clinic experience – **‘fleeting normality’** and **‘a battle for adequate care’**. In particular, both themes answer the second research question; how does access to outpatient services during an acute episode of the condition impact the patient experience? The themes will each be presented and critically discussed sequentially. The lived experience of people with BEB will be discussed in relation to IPA and the six quality of care dimensions (DOH, 2016).

BEB Experience in Relation to IPA and the Six Dimensions of Quality of Care

Patient-Centred Care and Timeliness

The findings of this study indicate that people with BEB during an acute episode of the condition experience poor patient-centred care and delays while visually, physically, or psychologically being unable to cope and in desperate need of treatment. For most participants, the question 'what is the patient experience of living with BEB during an acute episode of the condition?' evoked their reality and past experiences on lack of respect for their values and preferences, delayed referral, and subsequent treatment in the face of expressed needs. In addition, poor coordinated and integrated care, lack of information, negative communication, and psychological concerns began in primary care and continued in tertiary care. Following the principles of IPA, this study adds contextual understanding in the exploration of the participants' lived experiences.

This study found participants' first experience representing the theme '**no knowledge'** – insurmountable barriers to timely diagnosis and treatment. The concept of 'no knowledge' regarding BEB has been supported throughout the literature with acknowledgment of its rarity and diagnostic difficulties (Wakakura et al., 2018; Fung, 2013). This is a significant disadvantage since neither clinicians nor participants have encountered the disease. The participants suggest that many clinicians have limited knowledge of BEB's symptomology, making diagnosing the condition impossible.

In addition, textbook learning provides a poor substitute for experiential knowledge and vice versa. Participants acknowledged that 'no one knew what it was,' even following multiple clinical visits to numerous doctors and optometrists, misdiagnosis and medication trials still occurred. This caused a loss of trust and further delays in treatment and emotional distress as participants were refused referral to specialist care (Case 7 BEB Org p. 57; Drey p. 90).

Due to a lack of knowledge, a few participants were hopeful that the condition would resolve (Jey, p. 93), leading to delays. Therefore, the fundamental lack of BEB knowledge amongst healthcare professionals and the public significantly impacted access to treatment and ultimate daily life. This supports Valls-Sole & DeFazio's (2016) study, which demonstrates a lack of knowledge affects how care and treatment are organised in the healthcare environment.

A lack of timely care was a significant finding in this study. Lengthy periods of delay and incorrect diagnosis are unmet needs in healthcare (DOH, 2013). Importantly, this is interpreted as a lack of patient-centred care. Participants tried to make sense of why delays took place. The study interpreted the participants' repetitive speech as disbelief and representative of a traumatic experience in primary care: *"It took two years for my condition to be diagnosed, two years, which they thought it was dry eye and they were treating it"* (Drey, p. 90). This experience resonated with many of the participants.

BEB Co-ordination of Care

The most salient experience of BEB was a decline in QoL, representative of **'an incredibly debilitating state'**. Although functional vision and physical and psychological health declined, referral to specialist care was refused, indicating poor coordination of care. It created feelings of frustration, helplessness, distress, fear, and anxiety and led to a diagnosis of clinical depression among MOST of the participants. Participants struggled to navigate their world amid many challenges; ocular discomfort and eyelid spasms, progressive disease which cause facial contortion and neck spasms. Although participants tried to ameliorate the worsening symptoms with sensory tricks example holding around the face and eyes, the relief gained was only temporary. These findings support not only the lack of patient-centred care, but a lack of effective communication and poor coordination of care.

The accounts of participants indicated they tried to understand and rationalise the GPs’ refusal to refer and concluded that it was believed that the condition was not sufficiently challenging to their health (Janey, p. 91)

Similar experiences resonated throughout this study, and previous supporting studies showed the lack of knowledge among health professionals related to diagnostic mistakes and treatment delays (Kole & Faurisson, 2009; Nutt & Limb, 2011).

The study found poor care coordination only occurred in the NHS and not in private healthcare. The study supported Reimann et al. (2007), who showed that people with rare conditions experience a frustrating, extended journey towards diagnosis, have difficulties finding appropriate sub-specialist clinicians, and lack standard care. Adding credence to this is Lee et al. (2018) study, which demonstrated people with BEB experience a period of limbo, under-diagnosis, misdiagnosis, and delays. In addition, the UK strategy for rare diseases such as BEB (DOH, 2013; Valls-Solle & Defazio 2016) reported the lack of accurate diagnosis for rare diseases was due to medical professionals treating conditions they have never heard of.

Significantly, other factors contributing to misdiagnosis are a lack of consultation time in general practice and poor communication. The latter is evident as some participants felt rushed, dismissed, and given a misdiagnosis in a blasé manner; ‘oh, you got dry eyes, just put drops in’ (Egna, p. 92). The study concludes that poor care coordination is also affected by a lack of consultation time to diagnose the condition and poor communication.

Clinical depression is a known factor in determining QoL, a prominent feature among participants in this study. Empathic and responsive quality of care remained lacking for participants, although they were in ‘an incredibly debilitating state’ with significant poor QoL, namely physical and psychological health (Smith, 2011-p. 57; Lori 2018, p.58). Therefore, this finding is supported by numerous studies (Wenzel et al., 2000; Müller et al., 2002; Reimer et al., 2005; Munhoz et al., 2005), which showed the commonality of high levels of psychiatric disorders in people with BEB.

Another significant factor that determines QoL is visual impairment. It affects independence and impedes a range of day-to-day activities. Hall et al. (2016) demonstrated that people with BEB's QoL are reduced due to visual disability when the condition is untreated. Like those in (Grandas et al., 1988; Lee et al., 2018), participants in this study 'give up a range of activities and their social life.

Previous quantitative literature demonstrated that BEB could adversely affect individual physical and psychological health (Muller et al., 2002; Hall et al., 2006; Degrimenci et al., 2013). Through qualitative exploration, this study has uncovered that untimely treatment negatively impacted the patient's QoL in primary and secondary care. The delivery of high-quality healthcare depended on effective and efficient communication, both verbally and written. More importantly, this study suggests a decline in psychological health can be improved with more effective, empathic communication from health professionals (Nadia, p. 139).

The study also suggests GPs are not always open and honest about their lack of knowledge related to BEB. Instead, they demonstrate dismissive tendencies, which devalue the patients' concerns, especially when the expressed need for a referral is ignored. It supports Epstein and Street's (2007) claim that communication and patient-centred care are essential for promoting healing and reduction in suffering.

The poor coordination of care among BEB participants was found to cause functional blindness, which risks participants' safety. Coordination of care depended on sufficient consultation time to diagnose and develop a rapport to ensure effective communication and patient safety.

Ineffective Patient and Clinician Rapport

The study suggests participants developed a lack of trust in primary care professionals due to delays, incorrect treatment, and failure to diagnose obvious symptoms during numerous consultations (BEB Org Case 1, p.62; Janey p.91). In contrast, although the condition was not always diagnosed during an NHS consultation, a referral to treatment was always instigated from attending a private healthcare consultation. Private clinicians may have more time to invest in listening to patients or are more fearful of litigation.

The first step towards treatment is diagnostic communication. The role of the patient is to share their clinical, psychological concerns and psychosocial needs, while GPs occupy an obligatory gatekeeping role and that of a clinician. This dual role requires them to observe, be empathic and listen respectfully, examine, and synthesise the information, and using experiential knowledge, formulate a diagnostic conclusion and treat or refer to specialist care. The GP's dualistic role can be challenging to prioritise. Since GPs' gatekeeping is another factor that may delay referral, referrals cost the GP services considerable expenditure. Nonetheless, inadequacies in the communication process, distrust, or a lack of knowledge can lead to a failed diagnosis. GPs' skills and knowledge within this crucial gatekeeping role will be explored later in the chapter.

Although diagnosing a condition is complex and does not follow a linear process, this does not excuse health professionals' disregard of careful listening skills, which is vital in assessing patients' complaints to evaluate information and undertake a physical and neurological examination to eliminate other sinister pathology. However, a high prevalence of symptoms that lack a medically known biological cause may also contribute to diagnostic confusion.

The excuse of never seeing a particular condition does not absolve them from the responsibility of not referring. It could also be the feelings of ineptitude or bias contributing to the situation. However, a referral for further specialist ophthalmic examination would be considered intuitive. Crucially a lack of patient-clinician rapport was found to delay diagnosis. Essentially poor communication impaired care coordination from primary to secondary care, although referral to the specialist BEB clinic brought relief.

Insufficiency of Diagnostic Time-Impaired Safety

The study demonstrates misdiagnosis, and incorrect treatment created a safety risk and impacted social interaction. The findings of this study suggest while participants contended with the trial and errors of treatment, functional vision deteriorated, and accidents and injuries occurred, and psychological health was affected (Drey, p.94 Nadia, p. 99). The theme **'loss of social richness'** is closely aligned with the theme **'an incredibly debilitating state'** and represents the social and emotional impact of the condition BEB on participants.

The findings of this study suggest safety was impaired due to the lack of timely diagnosis and referral to treatment (BEB Org Case 6, p. 58; Jey, p. 93). Although there were multiple clinical attendances, insufficient diagnostic time reduced the assessment of symptomology (Egna, p. 92; Drey, p.90; Janey p. 91; Lownsborough, 2011 p. 58). It is supported by the King Fund-commissioned report on the quality of GP diagnosis and referral (Foot, Naylor, and Imison, 2010). A survey of 1,000 GPs reported insufficiency of diagnostic time. Similarly, there are apparent failures to diagnose the condition in this study at the first visit. This study supports studies such as (Burgess et al., 1998; Kantola et al., 2001) which highlighted diagnosis and referral did not take place during the patients' first presentation (Lori, 2018 p. 58; Delbert, p.110) The negative impact of misdiagnosis and inappropriate treatment on patients' safety could be interpreted from Janey's experience (p. 92; 107) as it resonated among participants.

The findings suggest a lack of consultation time to explore BEB symptomatology predispose patients to delays, misdiagnoses, and frequent re-attendance. It breached their safety and resulted in an ineffective service where care coordination was poor. Misdiagnosis and errors were found to harm the entire healthcare system (Kostopoulou et al., 2008). Therefore, misdiagnosis caused not only a loss of trust but suffering which traumatised the patient and affected future interactions and perceptions of care.

'An incredible debilitating state' or the period in limbo without treatment led not only to participants' loss of employment and career identity, but there was a risk of danger to themselves and others, and they suffered anxiety in social settings. Participants lost confidence and became embarrassed with facial contortions, poor vision, and mobility issues. Participants sought to come to terms with the impact of the phenomena on both members of the public making sense of interactions with comments such as: "*You feel they must think what is going on is strange"* and *"It's quite embarrassing, so you tend to turn away from that person who you're talking to, so it's not very normal"* (Denise, p. 108; p109)**.**

A lack of public knowledge invited scrutiny. Even with strategies designed to hide the spasms, participants' ability to cope was still affected, and the comfort of self-isolation was sought. While for many participants, a diagnosis of BEB led to relief, for others, the understanding that BEB was a long-term condition caused clinical depression.

The public anxiety, embarrassment, and self-isolation uncovered in this study support others (Garrino et al., 2015; Valls-Sole & Defazio, 2016) who found delays created uncertainty surrounding the treatment course, the future, disease progress and is known to trigger anxiety.

Notably, although various coping strategies were used, for example, the use of rose-tinted glasses. Other strategies include the avoidance of walking and concentrating: *"By the time you come back, and you have been concentrating really hard on keeping your eyes open, so you don't walk into anything or step off the kerb or something like that"* (Denise, p.99). Nonetheless, this study found that participants affected by BEB developed anxiety (Nadia, p. 99), became depressed (Leah, p.101) experienced financial losses (Judy, p.103) due to their inability to work, and even became suicidal as shown in the literature (Hall et al., 2005, 2006; Munhoz et al., 2005; Berardelli et al., 2021).

A few participants' accounts supported a lack of empathic communication during diagnosis and were likened to being '*hit by a brick*'. However, it can be argued that the impact of the diagnosis rather than the actual communication resulted in the trauma. The researcher's interpretation of the participants' accounts within the pretext of this IPA study interpreted the feelings and emotions assigned to the participants' experiences. An example is Petal's reflection in which she stated that she *"everything was too challenging"* in which it was difficult to cope, and Leah's account of her mood being affected to the extent of being clinically depressed and on antidepressants (Leah, p. 109).

Furthermore, how Egna was told to decide whether to await further deterioration or proceed with treatment was controversial since she had already suffered for a lengthy period. A diagnosis of BEB was difficult to cope with and negative communication impacted the diagnostic process. It has been concluded from this study that BEB as a long-term condition could lead to a diagnosis of clinical depression.

Ineffective Co-ordination of Care and Prolonged Suffering

This study found ineffective coordination of care between primary and specialist care prolonged suffering. Participants acutely affected experienced functional blindness, their social life was impaired, and they became depressed. QoL became poor as daily life was affected by poor mobility, embarrassment, and a loss of confidence. Furthermore, the findings indicate that even with sensory tricks and other coping strategies, the comfort of isolation was sought to avoid public scrutiny, as shown in previous literature (Hall et al., 2006). This study suggests that ineffective coordination of care and prolonged suffering may be related to a lack of clarity on which specialist a patient should be referred to. In addition, non-standard descriptions of conditions hampered a coordinated clinical approach.

NICE quality standards (NICE QS198, 2021) recommend that suspected neurological conditions be recognised and referred and that health professionals be aware of local referral pathways for neurological assessment to diagnose suspected dystonia in adults. However, the pathway can be confusing as BEB falls within the domain of both ophthalmology and neurology. In tertiary care, ophthalmic patients undergo a thorough neurological assessment before treatment may not be widely known. Therefore, there may be genuine confusion of who or where to refer, which impedes GPs' promptness even as the condition deteriorates.

The lack of GPs' skills and knowledge to identify and refer BEB patients to specialist care within their crucial gatekeeping roles could be interpreted as a hindrance.

Considering the NHS quality of care BEB participants were exposed to – inequity and a lack of standard care – caused untimely treatment delays, which impacted safety and deterioration. The system was not efficient, and inappropriate treatment caused a risk to safety as care was found to be ineffective. While diagnostic errors were recognised as multifactorial, missed opportunities within the system are also a contributing factor (Singh et al., 2017; Drey p. 90; Janey p. 91). Of critical importance is a general lack of public and healthcare information on rare diseases and specifically BEB (Dodge & Dion, 2011; Drey p. 108; Nadia p.109).

This study supports a King’s Fund report (Foot, Naylor, and Imison, 2010) which recommended that GP diagnosis be improved through good communication and good clinical relationships with specialist care providers to facilitate the exchange of information for learning opportunities and underpin high-quality diagnosis and referral. Good collaborative relationships also make it easier for GPs to seek informal advice, reducing the need for making formal referrals and avoiding duplication of tests. The findings of this study suggest the importance of GPs being aware of BEB more widely. It suggests the need to improve diagnostic knowledge and skills to expedite patients to treatment through shared information between specialists and primary care. The provision of education and resources to aid recognition of BEB would assist in greater awareness and understanding of the need for people with BEB to seek specialist attention and timely treatment.

Botulinum Toxin Treatment Satisfaction, Patient Centred and the Delivery of Timely Care

This study supports the current literature that BTX provides QOL improvement and treatment satisfaction in BEB (Muller et al., 2002; Tucha et al., 2001; Levy et al., 2006). The theme **'fleeting normality'** provides a deeper understanding of the treatment experiences in specialist care. This study found that although the side effects of BTX treatment are temporary and systemically rare (Naumann et al., 2006), initial side effects led to fear, anxiety, and a loss of trust in clinicians.

However, the study found a loss of trust in health professionals in primary care led to feelings of anxiety evident in initial interaction in the specialist clinic. The anxiety also pertains to the possibility of experiencing a side effect from the medication. However, anxiety remained for every episode of care. *"I'm certainly very anxious a lot of the time, and two years ago, I was between having injections [at a specialist eye hospital] before coming here"* (Judy, p.113). Notably, past trauma caused participants to feel anxious and distrust that their treatment would be effectively managed. Without adequate knowledge, doubts and anxiety led to the change in a service provider. However, previous experiences challenged participants' abilities to forge trusting relationships, suggesting that patient-centred care in the form of a partnership would reduce treatment anxiety.

This study supports the systematic review by Colosimo et al. (2012), which showed the patient-reported benefits of BTX range from 0% to 96%. However, all participants in the study treated in the specialist clinic showed significant symptomatic improvement and relief, even in the few instances where initial side effects were experienced. Notably, various studies have shown a three-monthly cyclical injection of BTX in patients with BEB (Hallett et al., 2013; Simpson et al., 2008; Jinnah et al., 2013).

This study supports a local randomised control trial. It showed untimely BTX treatment and care within specialist BEB clinics affected 70% of patients who faced recurrence of symptoms two weeks or more before their scheduled injection (Wickwar et al., 2016) as the injections wore off sooner than the recommended time. Contextual understanding of the needs of patients to be treated effectively when required adds understanding to why this study supports patient-centred care. In addition, it also supports (Colosimo et al., 2012), which shows BTX injection only offers temporary relief, with varied treatment duration and efficacy. However, it is still essential for the alleviation of distressing symptoms.

The study uncovered that BTX treatment improved participants' QoL to a state of almost normality. The positive effect of BTX was found to reduce participants' stress. In addition, concurrence and communication were utilised to prevent a decline in health. The visual, physical, and psychological health improvement felt caused participants to exhibit trust and a positive approach to the service as a whole: *"… a lifesaver coming here, there's no doubt about that at all. Once I'm here, the stress level drops. I have great confidence in you"* (Judy, p.117). However, proactive, individualised reports of the time BTX took to wane did not prevent untimely care. This study concludes that timely treatment depends on the service's ability to schedule the patient-clinician agreed cyclical treatment promptly.

Effective Service Delivery and Timely Treatment

Despite BTX clinical effectiveness and treatment satisfaction, this study found untimely treatment created an acute re occurrence of BEB symptoms and emotional distress, which disturbed patient QOL. The study supports studies (Jankovic and Orman, 1987; Albanese et al., 2013) which show BEB can lead to functional blindness and adds credence by demonstrating the adverse effects of BEB on everyday life, including emotional and psychological well-being (Hellman and Torres-Russotto, 2015; Valls-Sole & Defazio, 2016).

Participants' experiences draw support from (Hall et al., 2006) study, which suggests depression and anxiety are related to how treatment and care have been organised. In addition, it also supports a study by Streitova and Bares (2014), which recommends timely BTX treatment for the benefit of patients and families and to prevent possible personal injury, return to work, and improve social interaction, so avoiding depression.

The findings suggest a lack of effective service delivery results in **'a battle for adequate care'**. Therefore, this study supports the call of Wickwar et al. (2016) to optimise the patient experience and significantly reduce dissatisfaction and distress by treating patients as soon as possible when BTX wears off before the cyclical three-month scheduled treatment. It has been deemed clinically safe to administer BTX on a six-weekly treatment cyclical basis in a study by Jankovic et al. (2011).

It was found that access to care and treatment was denied with very little empathy and compassion, which caused fear, frustration, and anxiety (Petal p.120; Nadia p. 139). Only the more assertive participants won the battle for adequate care and treatment. Participants blamed funding and management for the lack of understanding of the severity of BEB (Janey, p.134).

Nevertheless, the study found that participants effectively treated with BTX could eventually move beyond the negative periods of unstable treatment to become positive. In addition, this study has found the chronicity of the condition requires patients with BEB to frequent the service to avoid acute relapse and adapt to the disease to minimise its physical and psychological impact. The treatment of long-term conditions requires the collaboration of care and a partnership approach. Therefore, understanding the patient-specific duration of BTX treatment enables discussion and agreement on a tailored treatment schedule. However, co-ordination of care is hampered if the treatment appointment is poorly timed or the medication prematurely wanes.

Therefore, this study found participants' demand for the service to deliver an efficient service to respond to the needs of patients was impaired. In addition, the organisational equity is often questioned as there are 20 satellite sites, but only two sites and four nurses provide injection BTX within specialist BEB clinics. The geographical differences create a financial burden on patients who face significant costs to travel a long distance to the main site. The service also lacks a patient support group which patients' belief would offer a sense of community, empower, and reduce feelings of isolation.

Giving BEB patients a greater voice can readdress local care and inject optimum quality in care. It is essential that BEB patients actively participate in the continuation of changing the previous status quo to ensure that care is aligned with their needs. This research sought to address the gap between policy, theory, and practice. It directly responds to the need to provide patients with a greater voice in their care. Nonetheless, it was essential to review the BEB background to identify the knowledge gaps in the literature.

Faced with adverse effects, participants once again lose trust in the medical professionals administering treatment. Ongoing fear and anxiety continued even after treatment stability was established. This provides an understanding of the trauma experienced caused by treatment side effects. With BTX treatment satisfaction, health and positivity were restored, and career identity. The specialist BEB clinic was subsequently seen as a lifesaver and the treating clinician as proficient. The findings support (Shepherd et al., 2008) and (Dean and Street, 2014). They advocated building a trusting relationship to explore the complexity of a condition, alleviate anxiety and distress, and enhance patients' involvement in decisions about their care. Awareness of participants' treatment duration is also needed as the effects of the medication wore off and then returned to an 'incredibly debilitating state'. Participants were found to lose positivity as BTX waned. A decline in psychological health returned with emotions of fear and anxiety that need to be treated – an essential indicator of quality healthcare.

Though BTX provides high levels of treatment satisfaction, the lack of timely treatment negatively impacted participants' QOL in previous and current research studies. Nonetheless, this study found that patients reported that treatment satisfaction was influenced by their expectations, adverse reactions and complications, trust in clinicians, and timely treatment on referral. Participants who expected immediate treatment effectiveness were highly disappointed by the side effects.

The study found that, even in the presence of side effects, participants understood and trusted their clinicians as a possible period of 'trial and error' before BTX optimisation took place. Although side effects such as pain and complications – for example, droop in the eyelid (ptosis) – were found to cause temporary dissatisfaction. However, participants' contention of treatment dissatisfaction was when BTX treatment wore off sooner than planned. But patients who 'religiously attended' or were adherent to a treatment schedule that coincided with timely treatment were most satisfied.

Lack of Equity in Access to the Specialist BEB Clinic

This study found a lack of equity in access to care and appointment management regarding assertive and non-assertive patients. While some individuals ‘begged’ and ‘pleaded’, there was an apparent failure to act, as was reported: “*You’ll have to wait another six months*” (Jey, p.124). In other cases, participants were undeterred, and their persistence was rewarded by prompt treatment, which was likened to ‘*winning a prize*’ (Denise, p.114). Geographical distance also prevented patients from requesting earlier treatment intervention. In addition, the clinic appeared overbooked, which deterred patients from seeking earlier treatment. These participants chose to *‘grin and bear it’*.

Some participants mentioned ‘funding’ and those in management thinking ‘BEB is unimportant. The chronicity of BEB requires older people to have regular costly, and timely treatment to maintain QOL. This study supports the European Commission (2017), which reported that the lack of efficient treatment for most rare diseases represents an enormous unmet medical need and a significant challenge for public health. This study supports the reports, studies, and guidance (Lori 2018, p. 58; Garrino et al., 2015; and GMC, 2020) that show delays create uncertainty surrounding the treatment course, the future, and disease progress which triggers anxiety and psychological decline.

The study also found participants who were unable to access timely treatment became dependent on others for support. The latter feature was seen in studies (Jinnah et al., 2013; Valls-Solle & Defazio) which agree that treatment delays lead to dependency issues. In addition, social contact was avoided to prevent judgment (Janey, p.107) Therefore, this study must support people with rare diseases, specifically those with BEB, in achieving their fundamental human rights to access timely care.

This study offers originality and innovation in its call for timely access to treatment using telemedicine to improve assessment of BEB severity, offer advice and advanced access that would provide the needed empathic care required to reduce untimely appointments.

Focus Group Discussion Recommendations

The overall recommendations of this study were linked to the research literature, study findings and service users' recommendations. However, the study has provided direct feedback from participants to inform service managers, clerical staff, and clinicians what needs to change to ensure the correct response to the needs of patients. This study found that participants felt it was important for health professionals, managers, and the clerical booking team to listen to their views and wishes to improve BEB clinics and quality of care. The recommendations made during the focus group discussion are listed below.

Recommendations

1. The provision of more nurse-led clinics which in contrast to the doctor's clinic offer prompt treatment and efficient care on the day
2. To offer BTX treatment at all satellite sites to eliminate demographic disparity.
3. The continuation of training to patients on the use of the check-in kiosk.
4. Patient to be contacted regarding missing notes before travelling to the outpatients' department.
5. Staff training to treating clinicians to ensure they have adequate knowledge of documenting treatment.
6. To create a holistic multidisciplinary clinic that involves mental health specialists, such as a counsellor or psychologist, and other dystonic specialists

to exchange knowledge and be sufficiently trained to treat more than one dystonic condition as required.

7. To create a sense of community and continuous support through focus groups.
8. To be able to assess patient severity via online and face-to-face assessments.
9. To maintain a telephone service for booking appointments; and
10. Customer care training for clerical staff to enable effective booking and treatment.

Impact of the Study on Practice and Service Delivery

The Need for More Physical and Psychological Support in the Specialist BEB Clinic

This research offers an original contribution to the knowledge of BEB care. It revealed that, without care and treatment, all participants on arrival at the specialist BEB clinic were already in ‘an incredibly debilitating state’ and had ‘lost social richness’. Consequently, they were experiencing visual impairment, which created mobility problems, dependency, financial instability, and clinical depression (Needle 2015; Leah, p.94; Delbert, p.112).

The findings support the BEB literature which demonstrates participants’ physical and psychological health was considerably impaired (Valls-Sole & Defazio, 2016; Lawes-Wickwar et al., 2021). Although most participants expressed relief, the few who found it difficult to cope would benefit from psychological support. Many participants felt a lack of confidence and experienced embarrassment and public scrutiny, leading to self-isolation (Petal, p. 109; Leah, p. 109). The physical debilitation experienced led to the psychological difficulties experienced which are usually accompanying features of BEB (Jey, p. 109; Nadia, p. 101; Kranz, p. 60).

This study also allows some understanding of how and why these emotions occur. The study calls for greater physical and psychological support through multidisciplinary teams working within the service. In addition, one new nurse has been trained and is currently providing additional support to respond to the needs of patient participants for timely treatment and care.

Greater Information and Collaborative Teamwork for BEB patients

Participants in the meta-synthesis and research findings indicated a lack of reliable and up-to-date patient information for people with BEB. They described a lack of knowledge to manage the condition effectively. Even within BEB clinics, patient education and information relevant to the service were unavailable. To effectively manage their condition, some participants became experts by searching the internet – a medium that can sometimes be misleading. The need for reliable and up-to-date knowledge about the condition BEB and its treatment was essential to maintain optimum psychosocial health.

Even so, as the researcher made sense of the participants' self-reflections of the specialist BEB clinical experience, there is a deep understanding that significant relief was felt on referral. Since the adaptive and coping strategies, including sensory tricks and helpful techniques, provided only transitory improvement and were unable to alleviate eyelid and facial spasms for a lengthy period. Instead, participants could not cope and remained anxious, frustrated, and depressed (Lori, 2018 p. 58; BEB org Case 1, p. 62; Leah, p. 94; Petal, p. 94).

Once BTX injections became effective without side effects, participants felt supported, listened to, and no longer felt alone. Importantly, they were hopeful about the future (Leah, p. 114; Delbert, p. 114). The latter finding supports studies that demonstrated BTX increases people's ability to cope with the physical and psychological impact of

BEB (Costa et al., 2004; Jinnah et al., 2013; BEB Org Case 1, p 62). The initial distrust displayed by a few participants who experienced side effects and adverse reactions to the medication subsided once they received adequate treatment. However, participants' subsequent reaction was to develop trust in clinicians, and treatment satisfaction was vocalised (Judy, p. 117; Egna, p. 118; Leah, p. 118) as a partnership approach was developed.

The study suggests the availability of patient information leaflets warning patients of the likely trial and error period would reduce initial treatment dissatisfaction and promote greater teamwork, including with treating clinicians. The use of multidisciplinary specialists, specifically a psychologist, and other dystonic specialists, would provide greater patient support. Patient information for patients with BEB has been introduced within the department.

Telemedicine to Reduce Cyclical Scheduling Anxiety

Though with a decline in the effects of BTX, participants' response reverted to distrust, anxiety, and desperation for treatment. There was hypervigilance among participants, and actions were directed toward avoiding an acute episode of the condition (BEB Org Case 1 p. 62; Petal, p.134 and Janey, p. 134). The research also uncovers anxiety around cyclical scheduling, which indicates undiagnosed post-traumatic stress disorder (PTSD), likely from previous negative healthcare interactions. Participants collaborated in BEB clinical appointment decision-making to prevent the likelihood of a poorly timed appointment.

The research also suggests that the treating clinician and clerical team members should form supportive relationships with patients to provide a more supportive environment to prevent re-traumatisation and suffering as the effects of BTX declines. By actively participating in treatment and clinical appointment decisions, BEB patients demonstrated the ability to be proactive and collaborate with the healthcare team to deliver expert patient experience advice, ensuring the organisation of timely cyclical treatment to avoid suffering. However, acute deterioration of the condition could not be prevented in many patients as the clerical booking system could not support earlier timings. The resultant acute deterioration of BEB negatively affected participants' QOL to a pre-treatment state (Tucha et al., 2001; Hilker et al., 2001; Leplow et al., 2017; Denise, p.126).

The research shows that treatments wore off before the scheduled appointment despite participants' actions. Then the booking team was contacted for access to treatment, but requests were denied.

Participants made numerous unsuccessful calls to 'plead and beg' for an earlier appointment to improve health (Jey, p. 124; Delbert, p. 124 and Drey, p. 123). Even a complaint to the Patient Advice and Liaison Service department made no difference, as overbooked BEB clinics had no capacity for emergency slots. Furthermore, the research found that empathy was lacking on the rare occasion that an appointment was missed due to life-threatening co-morbidities. Some participants' determination and persistence won them the battle for an earlier appointment and the actualisation of adequate care. Due to all of this, this study suggests telemedicine as a means of assessing BEB severity and providing empathic care to people with BEB. In addition, those requiring treatment can be expedited to be seen in person promptly to avoid further physical and psychological deterioration, especially anxiety associated with delayed treatment. Consequential to emergence of Covid-19 and the current study's findings, telemedicine has been introduced within the service.

The Use of Focus Groups to Share Experiences and Knowledge

Finally, the research indicates that the traumatic experience of the BEB journey of patients towards diagnosis, treatment, and care challenged their ability to cope, which can be improved by supportive, empathetic healthcare professionals and organisation of care. Although adaptive strategies are employed without BTX, participants found adjusting to their new reality difficult. Nonetheless, resilience was shown as some participants successfully battle for adequate care (Denise, p. 126; Petal, p. 126). The focus group discussion offered recommendations to ensure healthcare professionals improve contextual difficulties, such as access to care which reduces QOL and challenges coping abilities. The participants’ voices have been raised, and suggestions made from the recommendation of this study. This study suggests future focus group discussions involving patients, clinicians, and clerical management to provide ongoing support to patients in specialist BEB clinics. The nurse-led online focus group commenced November 2021.

Strengths, Limitations and Implications for Future Research

As a practitioner and insider researcher, there is an acknowledgment that the role of care overlaps and can negatively (Hawthorne effect) and positively affect the outcome and interpretation. However, the researchers interpreted the participants' words and meanings unbiased.

This study benefited from a small number of participants with contextual knowledge of specialist BEB clinics. However, its main limitation is a small focus group discussion (FGD) and may not represent all BEB patients’ experiences within the specialist BEB clinical settings. The findings may be taken as indicative and instructive rather than reproducible. The narrative within the findings helps to support the validity of the research of this method. Future community based FGD would be best geared toward PPI (Patient and Public Involvement) to implement and motivate the evaluation of changes within BEB OPD. NHS service improvements have previously found a deficit in local efforts. However, managers will find this paper helpful in informing service redesign and improvements at local levels.

In addition, research can play an essential role in highlighting the shortfall of service and ways to improve it. The study also highlights the importance of seeking patient feedback and that staff who obtain the same should be supported to enable the change required. Essential to change are formal processes for planning and delivering change, covering both minor modifications and more challenging problems, such as the reported issues of patient-clerical communication when seeking access to care. Policymakers and practitioners must be realistic about what can be achieved in the current climate of scarce resources, a commitment to developing patient experience research, and quality improvement to displace other competing priorities. However, this improvement has the full potential of being achieved.

Conclusion

Despite the low prevalence of rare conditions, while some people live and cope well, many depend on medical, psychological, and social support to live with the consequences of their condition. Patients also must take responsibility and adopt a proactive approach to reduce diagnostic delays and expedite care to prevent visual, physical, and psychological decline. Due to the scarcity of information about the condition BEB, this study has called for improved knowledge among health professionals with caregiving roles and throughout society. The communication of BEB red flags to primary care settings may be a way to address the diagnostic knowledge of primary care professional.

The complexities of diagnosing BEB and managing the condition can cause trauma, which healthcare professionals can avoid through collaborative partnerships with patients in primary care and secondary care. This study calls for patients' recommendations to be a reality to inform service improvement. Avoiding untimely access to BEB care and treatment during an acute episode would avoid re-traumatisation and improve health. Holistic, individualised treatment through the organisation of appointments to patient-related treatment duration and the introduction of psycho-education programmes and the continuation of focus group discussions for patients with BEB and their families and friends, can facilitate more supportive, effective, and successful management.

Chapter 8: My Doctoral Journey

This professional doctorate portfolio is structurally different from a PhD due to the course work it entails. This doctorate portfolio provides a deeper understanding of the rationale for undertaking the doctoral study. It utilises the four main domains, 12 sub-domains and 63 descriptors of the careers research and advisory centre vitae (see below diagram) and Rolfe's reflection to explore the researcher's journey and the impact of the study on practice and service delivery.

What?

Rolfe's 2001 Reflective Model has supported the analysis and evaluation of my research journey. My first research project a randomised control trial commenced in 2015 on; patient-initiated botulinum toxin injection Vs planned scheduled injection appointment highlighted the shortcomings of a quantitative study. Not only were patients saying to me that the service was failing to meet their needs when they most required treatment, but I also became aware that the study I was working on failed to capture the voices of patients on their BEB experiences and how to improve the service provided.

So What?

As a consequence, in 2015, I commenced an extremely challenging yet rewarding doctoral journey at West London University. The main reason for undertaking the doctorate study stems from my work as a novice researcher at work. This self-awareness provided the impetus for my actions; reviewing doctorate courses and applying to the university to undertake the current nursing doctorate study. Utilising the four domains of the research development framework; knowledge and intellectual abilities, personal effectiveness, research governance and organisation and engagement, influence, and impact I will critically discuss the impact of doctoral research.

Knowledge and Intellectual Abilities

Without the two years taught programme of the doctoral study, my knowledge, and intellectual abilities to undertake the study would not be possible. Although there were many significant challenges such as working full-time and having a timeline was critical to my success (see appendix). In addition, I the multiplicity of my role as a nurse consultant, a researchers, service manager for my department and an educator, being a mother and a wife, I remained a diligent student. I attended all classes, continuously read and frequenting the West London University library, the Institute of Ophthalmology library and online resources, made valid contributions to class discussions and form supportive bonds such as action learning set to discussion any research dilemmas.

Fundamentally the taught programme was instrumental in providing me with theoretical knowledge on various research methods which inspired me to enquire further by seeking more information to equip my research literacy, practical approach and application to this study. Similar to my thesis, academically successful completion of the taught programme provided me with the needed theoretical knowledge to analyse various literature, synthesise the information, employed critical thinking, evaluate and problem solve to formulate conclusions. Each essay (see example of submitted doctorate work in Appendix 11) was successfully assessed following submission and the feedback was given. The academic feedback increased my knowledge base of what I should or should not include and critically aided my entire study approached. I was able to apply the academic knowledge gained to formulate the qualitative approach most applicable to my research question: what is the lived experience of people with benign essential blepharospasm?

The theoretical knowledge helped me to determine that interpretive phenomenological analysis (IPA) was the approach for this study. Importantly IPA focuses on the exploration of participants‟ experience, understandings, perceptions and views (Reid, Flowers & Larkin, 2005) which was my personal aim for this study. However there was also the understanding that the subjective world of participants was only accessible through the participants' reflection and the analytical process I use as an IPA researcher.

One advantage I had in interpreting the research was my contextual knowledge of the service. However, knowledge base gained during my studies demonstrated that this type of qualitative approach has never been utilised to capture the biological illness experience of BEB, therefore offered the originality required for a doctoral study. Smith and Osborn (2003) has mentioned that IPA would be especially useful when dealing with complex issues, process or novelty which this study addresses. The IPA methodology, method of data collection and wider assessment of its applicability to it theoretical origins and procedures were useful knowledge acquired for this IPA research.

Despite the number of quantitative quality of life studies, the importance IPA or even a qualitative approach to explore the meaning given to the lived experience of BEB has been lacking. The theoretical knowledge acquired permitted exemplary IPA practice of face to face semi-structured interviews and a focus group discussion to employ collaboration is in keeping with an IPA approach. The interview questions were open ended and the schedule was flexible to allow prompts. In accordance with (Smith et al., 1997) such approach is in keeping with IPA and allows disclosure of participants' narrative in a manner they can control. To avoid any complaints regarding the analysis of focus group interview and themes elicited being similar there is acknowledgement by IPA researchers (Swift et al., 2002) that the structure and focus of their interview provides a framework for subsequent analysis. Therefore as an IPA researcher given this information I sought to be open about my role of preconceptions, beliefs and aims throughout the research processes.

The Impact of the Studies So Far

Knowledge and intellectual abilities were demonstrated to highlight the impact of the research on practice example collaborative writing other researchers. The articles written provides clear evidence to link my research and practice whilst highlighting my intellectual abilities:

- Wickwar et al., 2016 The effectiveness and cost-effectiveness of a patient-initiated botulinum toxin treatment model for blepharospasm and hemifacial spasm compared to standard care: Study protocol for a randomised controlled trial
- Wickwar et al., 2017. The illness and treatment beliefs of adults with Blepharospasm and Hemifacial spasm receiving botulinum toxin treatment
- Wickwar et al., 2018 The impact of facial dystonia on quality of life and factors determining quality of life
- Wickwar et al., in 2020, Which factors impact on quality of life for adults with blepharospasm and hemifacial spasm?
- Wickwar et al., 2020. The Illness and Treatment Beliefs of Adults with Blepharospasm and Hemifacial Spasm Receiving Botulinum Toxin Injections: A Cross-Sectional Study.
- Lastly, my written research collaborated articles provided me the confidence to published; Dunlop et al., 2020. A pilot study of a nurse-led adult lacrimal clinic at a tertiary ophthalmic centre.

In addition, my research intellectual abilities was recognised and rewarded during Covid-19 when my organisation was approach to collaborate with Manchester University on the following project:

- Covid-19 staff resilient project (**IRAS 282827**).

I believe my role as the principal researcher of this study was due to my doctoral studies that I was recognised and given the principal researcher role for my Trust. Prior to my appointment I had attended a local research meeting and presented my study and the impact it has made to patient care. Specifically the formulation of a patient focus group and the benefit of patient information leaflets which is appreciated by both staff and patients.

In addition I have presented my research and practice base impact at the following:

- June 2017, World Association of Eye Hospital Meeting China;
- November 2017, the Royal College of Nursing Ophthalmic Conference;
- October 2019 I presented the innovations and improvements my research has made to my clinical role and practice at the Best Practice in Nursing Conference;
- May 2019 UWL Ophthalmic Nursing Students;
- May 2020 UWL Ophthalmic Students online;
- April 2021 University of Manchester online;
- May 2021 UWL Ophthalmic Nursing Students online; and
- November 2020, Ophthalmic Nursing Network (ONN) Webinar on how to write your research.

My dedication to research and patient care have been acknowledged by patients and I have been awarded:

- Moorfields Eye Hospital Patient Choice Award 2018.

In addition, I have also won the Ophthalmic Forum Bid and have presented older people sight-loss early detection and action workshops in all four countries. This project was made possible from the experiential knowledge listening to the narrative of functional blindness.

The enormous contribution my research has made to impact patient care and service delivery was prompted by participants' needs for timely care and treatment. With the emergence of Covid-19, research improvement efforts were expedited. Patients who now have a decline in BTX effectiveness are seen in the nurse-led online telemedicine clinic. Telemedicine provides the technological advancement for BEB patients to be expedited for assessed if a report of waning treatment is made prompting urgent treatment in the specialist BEB clinic. The impact of my study which demonstrated the transfer of the knowledge gainedl was presented:

- March, 2021 at Moorfields Eye Hospital and University College London Conference aptly named; How Technology is Transforming Ophthalmic Nursing.

In addition my research has made me consider not only BEB but all conditions that causes visual impairment.

Now What?

Consequently I am now set to digitally present at the Royal College of Nursing's first ever hybrid congress in September 2021 on sight-loss conditions affecting older people's driving.

In addition, another improvement effort realised through this present research is the establishment of the first BEB online focus group which is set to commence November 2021.

Personal Effectiveness

According to the research and development framework the personal qualities, self-management and career and professional development influences the personal effectiveness of a researcher. The personal qualities I have already identified was my inquisitive nature to undertake an original qualitative study to assess the phenomena the lived experience of BEB. Seeking knowledge to equip myself with the information required to explore the concept and gaining analytical knowledge to analyse the data I immerse myself in, to produce the authentic subjective reality of participants. The study has allow me to demonstrate career and professional development also innovation in research introducing telemedicine to assess BEB patient need for treatment. During my studies I remained focus, dedicated, passionate but was also adaptive in my perseverance and self-management to complete a research which reflects high integrity.

Research Governance and Organisation

What?

One particular example that demonstrates not only my personal effectiveness and research governance and organisation but also my perseverance was an incident that occurred during ethical approval. Ethical approval on reflection was required to ensure my application reflected careful understanding of my role as a researcher. That I had the required ethical thinking to prepared a well-developed protocol which outline my methodology, participants, data collection analysis and ethical consideration. My application as a researchers must satisfy a research ethics committee that my study is ethical, worthwhile and are justified by the anticipated benefits to participants, science, healthcare and society as a whole.

In addition it was important to assured the research committee that I have measures in place to minimise any anticipated risks which can be intrusive to participants and reduce such burdens. Although University ethical approval was granted quite quickly which reflected all research principles were respected. I was unable to gain organisational ethical approval to proceed with the study and therefore I was also unable to apply for NHS ethical approval.

It was important to acquire both NHS and my organisation ethical approval before proceeding with the research that involves patient. Not only for ethical reasons but to protect patients from abuse and to provide indemnity insurance to organisational researchers. Organisational ethics ensure that as a researcher I was aware of the organisational legislation, regulation and protocol including auditing the progress of my study and data storage was in place to manage the organisation risk. However, even though my organisation funded the taught programme of my studies, the research element was not funded. Without research funding I was unable to gain organisational ethical approval to proceed with my studies.

So What?

I did not allow this impromptu set back to impede my progress as a researcher. Instead I worked tirelessly seeking out grant opportunities. I eventually applied to the Burdette Nursing Trust and was given a small of grant. As a result I was given organisational approval and was able to proceed with filling and submitting ethical forms to demonstrate my awareness and dedication to abiding by the research governance set out for NHS staff and patient studies. My IRAS application for HRA and HCRW **ID 235014** and local approval **DUNN1001** was granted (2018). My perseverance was demonstrated as well as my responsiveness to being adaptive and seeking new opportunities through networks to maintain my reputation as a dedicated researcher. Once approval was gained I was able to proceed with delivering the planned research strategy.

Now What?

The research was successfully undertaken as planned with no change to the research protocol. All patient data was managed according to NHS and organisational ethical standards and has produced a study that has engaged participants to provide recommendations for service improvement.

Engagement, Influence and Impact

What?

On completion of the study analysis I have engaged all participants and have reported the findings prior to a focus group discussion. Participants were in agreement that the study reflected their lived experience. A focus group discussion was useful in influencing me as a researcher to share the findings and participants recommendations widely and has influence participants to speak out and form supportive bonds to communicate previous experiences they thought was unique to them.

So What?

The focus group discussion caused participant to become even more vocal if that was at all possible and the following recommendation was made:

- For the specialist BEB clinic to create a greater sense of community with continuous support through focus group.

Actioned: As a researcher participants needs highlighted through the later recommendation influenced me to gain management buy in. The recommendation was actioned with the formulation of the first online BEB focus group November 2021.

A recommendations of the current study which has since been actioned is:

- The provision of more nurse-led clinics which offer quick efficient care.

Actioned: An additional nurse has since been trained to introduce more specialist nurse-led clinic at the main site.

However, while a nurse counsellor remains attached to the clinic the researcher is still influencing the service director and management to incorporate other specialist to ensure equity of care for all patients.

Nonetheless two further recommendations made by patients were that:

- The specialist BEB clinic provides prompt care and treatment; and
- Patients have asked to be contacted regarding missing notes before travelling into the OPD.

Actioned: The researcher is extremely proud of influencing and setting up a nurse-led telemedicine clinic to assess the severity of symptoms in patients with BEB patient. The researcher approached and influenced the local electronic database working group. As a result there is now an innovative online clinical database set to records clinical notes for all patients within the specialist BEB clinic. This negate paper notes becoming loss. The assessment of patient severity via online face to face clinic enables care to be provided and treatment appointments to be expedited. However the telephone appointment system is still being maintained as per the recommendation of many older patients:

- To maintain a telephone service for booking appointments but to also offer the flexibility of more than one booking options.

Another recommendation made by patients:

- Staff training to ensure treating clinicians are knowledgeable and document treatment effectively.

Actioned: The nurse researcher and consultant who provided training to nursing staff locally and nationally since January 2021 is now providing training to all junior doctors to ensure to ensure uniform care and knowledge across the service. In addition patient information on the condition and how the service works has now been made available to patients.

Now What?

However this study continues to influence management to:

- Offer BTX treatment at all MEH outreach sites to eliminate demographic disparity; and
- The continuation of staff training to patients on the use of the check in kiosk.

The final recommendation is as follows:

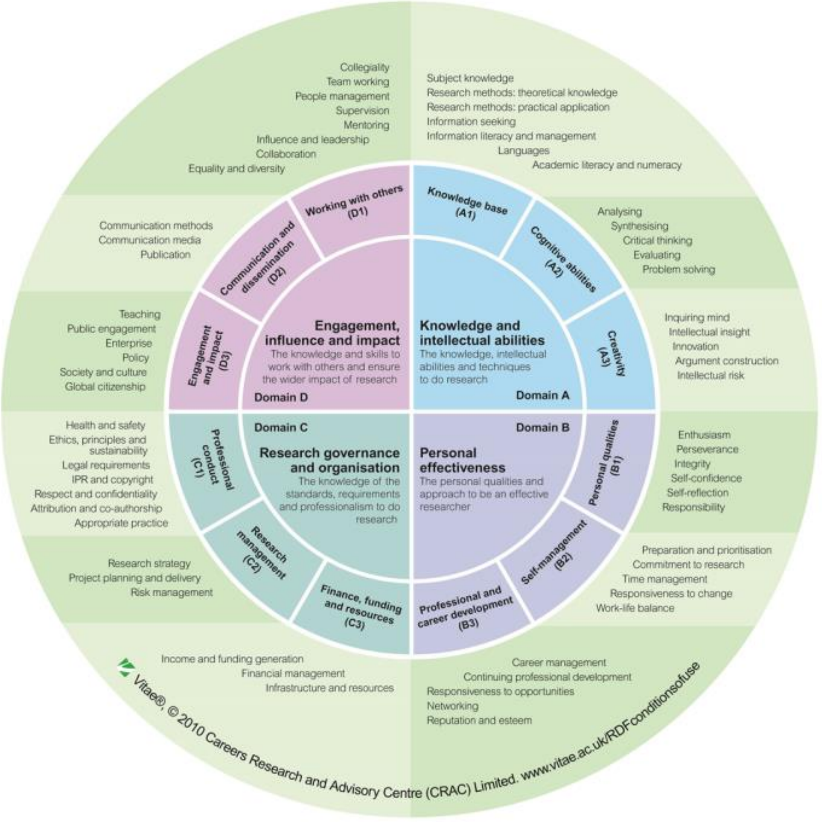
- To create a holistic multidisciplinary clinic which involves mental health specialist such as a counsellor or psychologist, and other dystonic specialist example cervical to exchange knowledge be sufficiently trained to treat more than one dystonic conditions as required.

Although the clerical team has been completely changed since the recommendations of this study were made. Management has brought awareness of patients' recommendations:

- Introducing customer care training for clerical staff to enable for empathy in addressing the concerns of patients.

The practice impact of this study has been highlighted through the engagement and influence of patients, service director and management to achieve the patients' recommendations. The study is yet to be communication to the Burdette Nursing Trust to further highlight the research impact of the study on patient care and service improvement.

- 4 domains
- 12 sub-domains
- 63 descriptors



Figures 12: Domains, sub-domains and descriptors for research development

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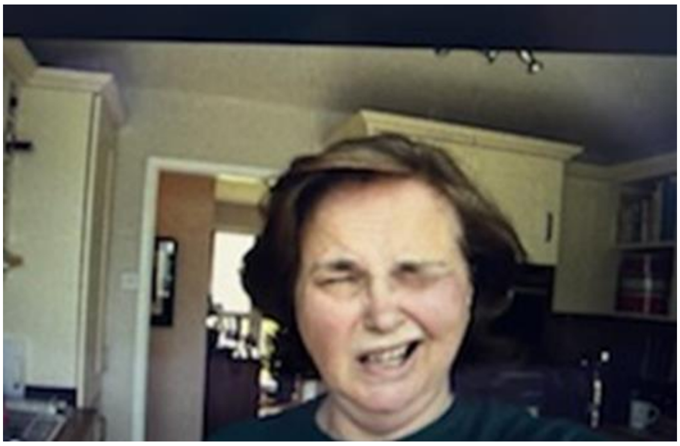
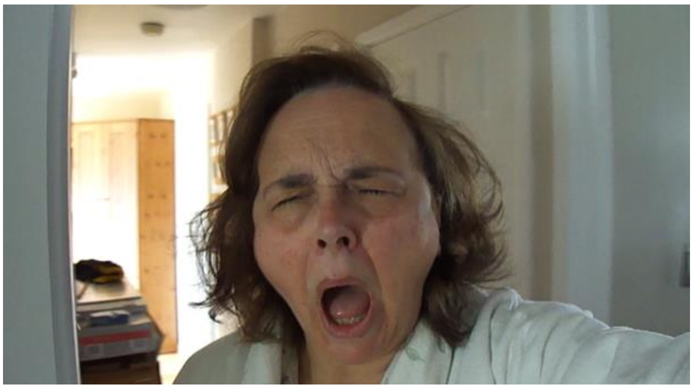
Appendices

Appendix 1





BEB with phenotype Meige (email: jenny_p_sexton@hotmail.com)

Appendix 2: Lists of RCTs associated with BEB

Study	Study Design and Goal	Sample Size and Method	Results
Lungu et al., 2013	Double-blind placebo-controlled randomized trial Formulation: on onabotulinumtoxin or rimabotulinumtoxinB Assessed topical use of acetyl hexapeptide-8 (AH8), competitive SNAP25 inhibitor as new therapy for BEB	n = 24 Injections into Orbicularis Oculi palpebral portion and 3/4 also received in the orbital portion/procerus/corrugator Topical application of AH8 or placebo started on Day 1 of BTX	Time for symptom return to baseline was 3.7 and 3 months in active and placebo groups, respectively AH8 is safe and may help increase the duration of effect
Wabbels et al., 2011	Double-blind, randomized trial Goal: compares incobotulinumtoxinA (Merz, BoNTA) to onabotulinumtoxin A	65 BEB patients received either incobotulinumtoxin A or onabotulinumtoxin A 20–40 U/eye Mean dose was 29 U/eye and 27 U/eye for on a botulinum toxin A and incobotulinumtoxin A	Though there was no significant difference between the two formulations, however, there was a greater tendency to improve at 4 and 8 weeks with onabotulinumtoxin, BDSI score mean change was −0.42 in abotulinumtoxin A and −0.21 in incobotulinumtoxin A No significant differences in side effects
Boyle et al., 2009	Prospective, randomized trial The study looks at differences between low (10 U/mL) and high (100 U/mL) concentrations of BTX A	16 patients 10–30 units per side. The Left and right sides were randomized	62% had equal relief of both sides No difference in efficacy Bruising and redness similar between groups
Truong et al., 2008	The randomized trial, double-blind, placebo-controlled Studied safety and efficacy of abobotulinumtoxin A versus placebo in BEB	10, 23, 25, and 27 patients completed placebo, abobotulinumtoxinA 40, 80, and 120 U/eye, respectively	25%, 87%, 97%, and 94% of patients found placebo, 40 U/eye, 80 U/eye and 120 U/eye to be effective Ptosis occurred in 4%, 13%, 39%, and 58% in placebo, 40 U/eye, 80 U/eye, and 120 U/eye, respectively Blurred vision occurred in 4%, 23%, 19%, and 42% in placebo, 40 U/eye, 80 U/eye, and 120 U/eye, respectively 80 U/eye provided the most efficacy while balancing for adverse effects.
Roggenkamper et al., 2006	Double-blind, randomized trial Assessed NT201(Merz Pharmaceuticals GmbH, Germany) the new formulation of BXT A compared to onabotulinumtoxin A	148 patients received NT201 and 152 patients received onabotulinumtoxin A The mean total dose in NT201 was 39.6 units and Onabotulinumtoxin A 40.8 units	Mean change in JRS was −2.67 and −2.90 for Onabotulinumtoxin A and NT201, respectively. NT201 is safe and efficacious for BSP treatment Ptosis, xerophthalmia, and abnormal vision occurred at 6.1%, 2%, and 1.4% in NT 201 Ptosis, xerophthalmia, and abnormal vision occurred in 4.5%, 0%, and 3.2% in the onabotulinumtoxinA group, respectively
Mezaki et al., 1999	Double-blind trial Assesses the effectiveness of type A versus type F versus combination of A+F	54 patients had five units each of A+F on one side and the other the side had A or F Patients who were randomly given either type A or F on one side and the mixture on the other side	There was no difference between the groups when comparing the AF side The peak effect was similar among the three groups Duration of action of the AF group was less than that of A and more than that of F alone.
Nussgens Z et al.,	Double-blind trial	212 BEB patients received either	OnabotulinumtoxinA and

1997	Studies onabotulinumtoxin A versus abobotulinumtoxin A	onabotulinumtoxin A or Abobotulinumtoxin A the first time and the other BoNT during the second session OnabotulinumtoxinA average dose was 45.4 IU and abobotulinumtoxinA was 182 IU	abobotulinumtoxinA lasted 7.98 ± 3.8 weeks and 8.03 ± 4.6 weeks, respectively Adverse effects such as ptosis, blurred vision, diplopia, hematoma and tearing occurred in 17% and 24.1% of onabotulinumtoxinA and abobotulinumtoxinA, respectively ($p < 0.01$) Bioequivalence of onabotulinumtoxinA: abobotulinumtoxinA is 1:4 in this trial
Fahn et al., 1985	Double-blind a controlled study of BTX for blepharospasm was assessed against a placebo for its effectiveness. Fahn rating scale of movement and disability also utilised	8 patients Electrophysiology	BTX A significantly Reduced evoked Potential contractions at rest and maximum forced contraction versus placebo
Frueh et al., 1988	Randomized, double-blind, placebo-controlled	26 Patient subjective improvement (no scale mentioned)	Similar effects in the lower eyelid with BTX A and placebo
Jankovic et al., 1988	Randomized double-blind placebo-controlled trial (after initial open-label phase) Assessed BTX A for management of various focal dystonia including BEB	22 patients with focal dystonia received either BTX A or saline	All 12 BEB patients had relief of symptoms with BXT, and none who received saline improved. Mean beneficial effect lasted 12.5 week
Sane et al., 2019	Triple masked RCT Assessed efficacy of onabotulinumtoxin A versus Neuronox in BEB	24 patients with BEB were randomized to receive either formulation	Mean duration of improvement was 3.78 months Neuronox and onabotulinumtoxinA were similar in safety and efficacy

Appendix 3: Database Search Strategy

Search Lines	Search Terms Ovid Medline 8th September 2022 (1946-Current)	Search Results
1	**exp blepharospasm**	**1425**
2	**(Blepharospasm or BEB or blepharo facial spasm or benign essential blepharospasm)**	**2454**
3	**1 or 2**	**2454**
4	**exp "quality of life"**	**249478**
5	**(impact or burden or consequence* or coping or experienc*)**	**3046184**
6	**(quality adj5 life).**	**422916**
7	**(daily adj3 living).**	**90784**
8	**4 or 5 or 6 or 7**	**3367176**
9	**3 and 8**	**324**

Search Lines	Search Terms Ovid Embase 8th September 2022 (1946 – Current)	Search Results
1	(MM "Blepharospasm")	0
2	Blepharospasm or BEB or blepharo facial spasm or (benign essential blepharospasm)	2454
3	1 or 2	2454
4	experienc* or (living adj3 with) or impact or burden or consequence* or coping	3092854
5	quality adj5 life OR daily adj3 living	92123
6	4 or 5	3155663
7	3 AND 6	283

Search Lines	Search Terms EBSCO PsycInfo 8th September 2022 (1946 – Current)	Search Results
1	(MM "Blepharospasm")	1
2	Blepharospasm or BEB or blepharo facial spasm or (benign essential blepharospasm)	375
3	1 or 2	2176504
4	experienc* or (living adj3 with) or impact or burden or consequence* or coping	1327164
5	quality adj5 life OR daily adj3 living	130
6	4 or 5	916378
7	emotions quality of life english boolean/phase	742
8	3 AND 6 AND 7	1120

Search Lines	Search Terms EBSCO GreenFile 8th September 2022 (1946 – Current)	Search Results
1	(MM "Blepharospasm")	0
2	Blepharospasm or BEB or blepharo facial spasm or (benign essential blepharospasm)	13
3	1 or 2	397216
4	experienc* or (living adj3 with) or impact or burden or consequence* or coping	204889
5	quality adj5 life OR daily adj3 living	47
6	4 or 5	174538
7	emotions quality of life english boolean/phase	8
7	3 AND 6 AND 7 (references available, abstract available)	2017

Search Lines	**Search Terms EBSCO CINAHL 8th September 2022 (1946- Current)**	**Search Results**
1	(MM "Blepharospasm")	3
2	Blepharospasm or BEB or blepharo facial spasm or (benign essential blepharospasm)	402
3	1 or 2	3907681
4	experienc* or (living adj3 with) or impact or burden or consequence* or coping	1098804
5	quality adj5 life OR daily adj3 living	226
6	4 OR 5	1487224
7	emotions quality of life english boolean/phase	0
8	3 AND 6 AND 7 (references available, abstract available)	2541

Appendix 4: Critical Appraisal of QoL Studies

Critical appraisal of included analytical cross-sectional studies

Citations	Q1	Q2	Q3	Q4	Q5	Q6	Q7	Q8
Hall et al., 2006	Y	Y	Y	Y	Y	Y	Y	Y
MacAndie & Kemp (2004)	Y	Y	Y	Y	Y	Y	Y	Y
Lee et al., 2013	Y	Y	Y	Y	Y	Y	Y	Y
Wickwar et al., 2020	Y	Y	Y	Y	Y	Y	Y	Y
Wickwar et al., 2021	Y	Y	Y	Y	Y	Y	Y	Y
Lee et al., 2013	Y	Y	Y	Y	Y	Y	Y	Y

N, no; N/A, not applicable; U, unclear; Y, yes; JBI critical appraisal checklist for analytical cross-sectional studies Q1. Were the criteria for inclusion in the sample clearly defined? Q2. Were the study subjects and the setting described in detail? Q3. Was the exposure measured in a valid and reliable way? Q4. Were objective, standard criteria used for measurement of the condition? Q5. Were confounding factors identified? Q6. Were strategies to deal with confounding factors stated? Q7. Were the outcomes measured in a valid and reliable way? Q8. Was appropriate statistical analysis used?

Critical appraisal of included cohort studies

Citations	Q1	Q2	Q3	Q4	Q5	Q6	Q7	Q8	Q9	Q10	Q11
Degirmenci et al., 2013	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y
Setthawatcharawanich et al., 2011	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y
Hilker et al., 2001	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y
Weiss et al., 2018	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y
Muller et al., 2002	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y
Zhang et al., (2010)	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y

AIHW, Australian Institute of Health and Welfare; AMANHI, Alliance for Maternal and Newborn Health Improvement. N, No; N/A, not applicable; U, Unclear; Y, Yes; JBI critical appraisal checklist for cohort studies Q1. Were the two groups similar and recruited from the same population? Q2. Were the exposures measured similarly to assign people to both exposed and unexposed groups? Q3. Was the exposure measured in a valid and reliable way? Q4. Were confounding factors identified? Q5. Were strategies to deal with confounding factors stated? Q6. Were the groups/participants free of the outcome at the start of the study (or at the moment of exposure)? Q7. Were the outcomes measured in a valid and reliable way? Q8. Was the follow-up time reported and sufficient to be long enough for outcomes to occur? Q9. Was follow-up complete, and if not, were the reasons to loss to follow-up described and explored? Q10. Were strategies to address incomplete follow-up utilized? Q11. Was appropriate statistical analysis used?

Critical appraisal checklist for included case-control study

Citations	Q1	Q2	Q3	Q4	Q5	Q6	Q7	Q8	Q9	Q10
Timlin et al., 2021	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y
Leplow et al., 2017	Y	Y	Y	Y	Y	Y	Y	N	N	N
Tucha et al., 2001	Y	Y	Y	Y	Y	Y	Y	N	N	N
Streitova & Bares (2014)	Y	Y	Y	Y	Y	Y	Y	N	N	N
Tang et al. (2018)	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y
Reimer et al., 2005	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y

Y, yes; JBI critical appraisal checklist for case-control studiesQ1. Were the groups comparable other than the presence of disease in cases or the absence of disease in controls? Q2. Were cases and controls matched appropriately? Q3. Were the same criteria used for identification of cases and controls? Q4. Was exposure measured in a standard, valid, and reliable way? Q5. Was exposure measured in the same way for cases and controls? Q6. Were confounding factors identified? Q7. Were strategies to deal with confounding factors stated? Q8. Were outcomes assessed in a standard, valid, and reliable way for cases and controls? Q9. Was the exposure period of interest long enough to be meaningful? Q10. Was appropriate statistical analysis used?

Appendix 5a: Level of Evidence of Effectiveness

JBI Levels of Evidence for Effectiveness	Levels
Experimental Designs	**Level 1**
Systematic review of Randomized Controlled Trials (RCTs)	Level 1.a
Systematic review of RCTs and other study designs	Level 1.b
RCT	Level 1.c
Pseud-RCTs	Level 1.d
Quasi-experimental Designs	**Level 2**
Systematic review of quasi-experimental studies	Level 2.a
Systematic review of quasi-experimental and other lower study designs	Level 2.b
Quasi-experimental prospectively controlled study	Level 2.c
Pre-test – post-test or historic/retrospective control group study	Level 2.d
Observational – Analytic Designs	**Level 3**
Systematic review of comparable cohort studies	Level 3.a
Systematic review of comparable cohort and other lower study designs	Level 3.b
Cohort study with control group	Level 3.c
Case – controlled study	Level 3.d
Observational study without a control group	Level 3.e
Observational	**Level 4**
Descriptive Studies	Level 4.a
Systematic review of descriptive studies	Level 4.b
Cross-sectional study	Level 4.c
Case series	Level 4.d
Case study	Level 4.e
Expert Opinion and Bench Research	**Level 5**
Systematic review of expert opinion	Level 5.a
Expert consensus	Level 5.b
Bench research/ single expert opinion	Level 5.c

Appendix 5b: Summary of QoL Findings (Included and Excluded Studies)

Summary of Findings					
No:	**Outcomes**	**Impact**	**No: of participants (n= 1077)**	**Author(s)**	**Evidence of Effectiveness (GRADE)**
1	**Gender & Disease Duration** Assessment tool: 36-item short-form (SF-36)	Women with BEB showed significantly worse scores in 5 out of 8 domains on the SF-36 compared to male patients. Although BTX improved clinical symptoms, health related QoL did not improve. It is the only study to associate a longer duration of BEB with improved QoL due to the adaptive nature of patients with a long-term condition.	89	Muller et al., 2002 Longitudinal cohort study	**Level 3c**
2	**Gender** Assessment of severity and the resulting disability using the Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) and Health-Related Quality of Life (HRQL) measured through the SF-36	The study found that worsening disease in females was associated with to their perceived hypersensitivity to facial disfigurement, depression, anxiety, and functional disability.	13	Zhang et al., (2010) Prospective longitudinal Cohort Study	**Level 3c**
3	**Aggravating and relieving factors** Assessment of functional disability was made subjectively by a Quality of Life questionnaire completed by the patient	Acute triggering event causing anxiety or depression was present in 61 cases (96.8%). Secondary triggers were meibomian gland dysfunction (100% cases), tear film instability (96.8%), dry eyes (85.7%), chronic Trachoma (14.3%), concretions (26.3%), trichiasis (4.8%), and chronic blepharitis (12.7%). At least 2 or 3 triggering factors were noted an all cases.	66	Irfan et al., 2018 Prospective Observational Study	**Level 3e**
4	**Vision and functional disability** Assessed using 25-item National Eye Institute Visual Function Questionnaire (NEI-VFQ-25) were low.	The study found people with BEB experienced a greater reduction in vision-related health-related QoL and were more prone to symptoms of depression symptoms ($p = 0.03$), and generalized anxiety disorder ($p = 0.007$). They reported lower NEI-VFQ-25 composite scores ($p<0.001$), and lower NEI-VFQ-25 subscale scores regarding general vision ($p = 0.03$), ocular pain ($p<0.001$), distance activities ($p= 0.001$), driving ($p<.001$), and all of the vision-specific subscales addressing psychosocial issues. This underscores	159	Hall et al., 2006 Cross sectional survey	**Level 4b**

		the inadequacy of current treatment options for BEB in light of the fact that these patients had been undergoing standard-of-care treatments for some time.			
5	**Vision and functional disability** SF-36 (global HRQOL), the NEI-VFQ (disease-specific HRQL) and the Blepharospasm Rating/Disability Scale	The study found minor functional limitations but identified some symptoms to be improved. In both patient groups global (both SF-36 Component Summaries) and disease-specific (eight of 12 subscales) HRQL were significantly impaired compared with controls. . HRQL were significantly impaired in cases as compared with controls.	31 BEB 21 HFS	Reimer et al., 2005 Retrospective Case Control Study	**Level 2d**
6	**Treatment-BTX** QoL, self-esteem, and ocular surface Assessed with World Health Organisation Quality of Life-Brief Field version questionnaire (WHOQOL-BREF), the Self-Esteem Scale (SES), and the Ocular Surface Disease Index (OSDI) before and treatment.	Improvements were found in all assessed outcomes after treatment. The study suggested that improvement in BEB over time may also be related to the introduction of BTX treatment as this reduces spasmodic symptoms, improves vision, functional disability, self-esteem, and overall improves QoL.	87	Tang et al. (2018) Prospective comparison Study	**Level 3d**
7	**Treatment-BTX** To determine impact on QoL in BEB. Instrument-Glasgow Benefit Inventory, postal questionnaire	This study demonstrates significant quality of life benefit from Botulinum toxin therapy for essential blepharospasm and justifies continued treatment.	44	MacAndie & Kemp (2004) A cross-sectional study	**Level 4b**
8	**Treatment-BTX** Self-assessment and a patient questionnaire to evaluate the influence of the treatment on their quality of life.	BXT-A is an effective and safe long-term treatment of these facial dyskinesias. However side effects in BEB frequency was higher manifested by double vision or eyelid ptosis. All patients believed that the treatment was safe and effective with a positive impact on their quality of life, especially in social communication.	9	Streitova & Bares (2014) Case Comparison Study	**Level 3b**
9	**Treatment-BTX** Glasgow Benefit Inventory (GBI) and Blepharospasm Disability Index (BSDI) scores. Post interventional telephone interview. To assess and compare the change in quality-of-life and function	BTX treatment for blepharospasm was found to be associated with significant patient-reported improvements in quality-of-life (GBI) and functional ability (BSDI) with a strong positive correlation between both scores.	62	Lee et al., 2013 A cross-sectional study	**Level 4b**

	following treatment with BTX				
10	**Treatment -BTX** Quality of life questionnaire completed by patients who had been treated with BTX for 1 year. The purpose of this study was to investigate aspects of quality of life of patients with blepharospasm and level of patient satisfaction with treatment.	BXT injections in BEB is appreciated by the patients, but their well-being is affected by fears of worsening condition and depression	51	Tucha et al., 2001 Case observational study	**Level 4c**
11	**Treatment -BTX** Instrument- EuroQol (EQ-5D-5 L) EQ-VAS, life satisfaction (health and movement disorders domains), Beck's Depression inventory, and sleep quality (One-way ANOVAs, p< 0.05, adjusted for multiple comparisons).	Motor and non-motor signs of blepharospasm including motor symptom improvement, life satisfaction, depressive symptoms, pain and sleep quality was assessed. RESPONDERS showed higher impairment in EQ-5D-5L, EQ-VAS, and Beck's Depression Inventory compared to the UNCHANGED (unlike WORSE). Those WORSE showed higher impairment in life satisfaction 'movement disorders' domain	55	Weiss et al., 2018 Prospective observation cohort	**Level 3d**
12	**Treatment- BTX** The HRQL was assessed using the EuroQol (EQ-5D) and the short form 36 health survey questionnaire (SF-36) at baseline before BTX injections and at two follow up visits after 6 and 12 weeks covering one BTX treatment period with maximum effect size at the first follow up	The data confirms that BTX is able to induce a significant, but temporary amelioration of several aspects of HRQL in both types of focal dystonia. This may substantially contribute to the patients' subjective benefit from the therapy. Moreover, the data provide further arguments to accept high costs of the BoNT/A treatment in these severely handicapped patients, as a consequence of its considerable benefit on quality of life.	20	Hilker et al., 2001 A prospective, open labelled cohort study.	**Level 3d**
13	**Treatment satisfaction and emotional responses to a life** with a disabling condition were investigated through a survey.	BEB satisfaction worsened significantly within the treatment cycle. Despite good overall treatment effects, patients from both groups perceived marked persistence of motor symptoms, restrictions of everyday life functions, and reduced quality of life. Functional amelioration of motor symptoms and emotional well-being were only moderately correlated.	125	Leplow et al., 2017 Case comparison study	**Level 3b**

		About 22% of patients from both groups reported mental disorders or emotional disturbances prior to the onset of the dystonia			
14	**The impact of periocular surgery,** other than orbicularis stripping, on the severity and frequency of blepharospasm symptoms. JRS and BDSI instrument.	Periocular surgery significantly reduced BEB in 83% but 17% experienced worsening symptoms	24	Timlin et al., 2021 Retrospective Case Reviewed	**Level 4d**
15	**Treatment BTX-Impact** of hemifacial spasm (HFS) and blepharospasm (BS) on quality of life (QOL) during long-term treatment with botulinum toxin (BTX) injections. The instrument used include a Disease specific questionnaire and a Thai depression inventory was used.	The disease severity in both patient groups revealed mild functional impairment. In both HFS and BS, depression scores were positively correlated with the HFS-30 scores, while peak improvement and educational level were inversely correlated with the HFS-30 scores. Patients with BS were more affected in Mobility and Activities of Daily Living than HFS patients.	32	Setthawatcharawanich et al., 2011 Cohort with control study	**Level 3c**
16	**Treatment of BTX on QoL**	WHOQOL-BREF questionnaire two (2) times. The first time was on the day of the administration of Botulinum Neurotoxin (i.e. when, according to the opinion of ophthalmologists, the symptoms were most prominent), and a month and a half after	37	Biuk et al., 2013 Prospective non cohort	**Level 3e**
17	**The effects of depression and anxiety.** Anxiety and depression was evaluated with Hospital Anxiety Depression (HAD) scale. Short Form-36 was used to assess the quality of life in both study groups.	Depression and anxiety have a great impact on quality of life of patients with dystonia. Dystonia can be considered as an important risk factor for anxiety and depression which leads to a deterioration in the quality of life of patients.	43	Degirmenci et al., 2013 Cohort with control study	**Level 3c**
18	**Beliefs about BEB and HFS and treatments** were assessed using the Illness Perceptions Questionnaire (IPQ-R) and Treatment Representations Inventory (TRI). Blepharospasm Disability Index (BSDI) and	Participants associated BEB and HFS with a chronic timeline and believed their condition was not caused by psychological factors, risks, or their personality or lifestyle. Participants also demonstrated decision satisfaction related to treatment but had some treatment concerns. 5 domains of QoL were identified, emotional well-	65	Wickwar et al., 2020 Cross-sectional study	**Level 4c**

	Craniocervical Dystonia Questionnaire (CDQ24). Severity and frequency of spasms were recorded using the Jankovic Rating Scale (JRS). Relationships identified using Pearson's product-moment correlation.	being, activities of daily living, social/family life, pain, and stigma. Participant believed frequent blinking (56%) was related to their BEB or HFS. Similar proportions of the sample attributed sore eyes (58%), eye irritation (68%), uncontrollable eye closure (67%) and muscle twitching (67%) to their BEB or HFS. Between 30 and 45% of the sample also attributed sleep difficulties, headaches, fatigue and pain to their condition.			
19	**Clinical, demographical and psychological factor on QoL** was assessed using Craniocervical Dystonia Questionnaire (CDQ24).	CDQ24 showed 80% variance and was significantly associated with appearance concerns, emotional representation, perceived negative consequences of the condition, mood, and dose of BTX. Detrimental impact on activities of daily living and perceived stigma in relation to their condition. Perception and mood were associated with QoL rather demographics and clinical factors	**65**	Wickwar et al., 2021 Cross-sectional study	**Level 4c**
Studies Excluded (no assessment of effectiveness necessary)					
1	**To determine whether the use of alleviating manoeuvres for benign essential blepharospasm or hemifacial spasm correlates with disease severity or botulinum toxin treatment.**	A prospective cross-sectional observational study	**74**	**Kilduff et al., 2016**	
2	**To determine whether botulinum toxin (BtA) is an effective and safe treatment for blepharospasm.**	Retrospective questionnaires	**42**	**Costa et al., 2001**	
3	**To investigate causes, associations, and results of treatment with BEB**	Extensive Questionnaire To investigate causes, associations, and results of treatment with BEB through questionnaire	**1653**	**Anderson et al., 1998**	
4	**To evaluate the characterise patients with benign essential blepharospasm (BEB) by diagnosis, environmental risk factors, and**	Clinical examination and questionnaire.	**240**	**Peckham et al., 2011**	

	family history				
5	**To screen for Cognitive Impairments in Primary Blepharospasm**	Chinese version of the Addenbrooke's Cognitive Examination-Revised and the Mini-Mental State Examination (MMSE) to screen for cognitive impairment in patients with primary blepharospasm and the relationship between performance on the Addenbrooke's Cognitive Examination-Revised and quality of life as measured by the Medical Outcomes Study 36-item Short-Form (SF36)	**68**	**Yang et al., 2016**	
6	**Effectiveness and cost-effectiveness of a patient-initiated botulinum toxin treatment model for blepharospasm and hemifacial spasm compared to standard care: study protocol for a randomised controlled trial**	Primary outcome measures include disease severity (questionnaire), functional disability (questionnaire) and patient satisfaction with care (questionnaire). Secondary outcomes include disease-specific quality of life (questionnaire), mood (questionnaire), illness and treatment perceptions (questionnaire and semi-structured interviews), economic impact (questionnaire) and acceptability (questionnaire and semi-structured interviews).	**65**	**Wickwar et al., 2016**	
7	**To investigate the long-term effectiveness and safety of botulinum neurotoxin A (BoNT-A) treatment in patients with blepharospasm (BEB), hemifacial spasm (HFS), and entropion (EN) and to use for the first time two modified indexes, 'botulin toxin escalation index-U' (BEI-U) and 'botulin toxin escalation index percentage' (BEI-%), in the dose-escalation evaluation.**	In BEB patients, the BEI-% was significantly increased in younger compared with older patients (P=0.008). The most frequent adverse events were upper lid ptosis, diplopia, ecchymosis, and localized bruising. All patients in this multicentre study were followed for at least 10 years and main outcomes were clinical efficacy, duration of relief, BEI-U and BEI-%, and frequency of adverse events.	**83**	**Cillino et al., 2009**	
8	**Sexual Well-Being in Patients with Blepharospasm, Spasmodic Torticollis, and Hemifacial Spasm: A Pilot Study.**	Patients were evaluated just before the periodic injection of botulinum toxin. Sexual functioning was assessed using the Sexual Functioning Inventory, a reduced form of the Golombok Rust Inventory, previously employed in	**30**	**Perozzo et al., 2016**	

		patients with Parkinson's disease. Depression (Beck Depression Inventory) and anxiety (STAI-X1/X2) were also assessed.			
9	**To evaluate the effects of botulinum toxin type A (BTX-A) treatment in patients with benign essential blepharospasm (BEB) by monitoring the ocular surface and ocular higher-order aberrations (HOAs) before and after treatment**	Patients were evaluated for ophthalmoscopic findings, Schirmer I test, tear film break-up time (t-BUT), HOAs, fluctuation index (FI), stability index (SI) using a wavefront aberrometer, 15 subjective symptoms using the Dry Eye-Related Quality of Life Score (DEQS), and complications before and after the treatment.	**38**	**Isshiki et al., 2016**	
10	**The aim of this study was to prospectively follow the spread of dystonia in 132 consecutive patients and to estimate the risk of spread by the site of onset of dystonia.**	In addition to the highest risk for further spread of dystonia, BEB was associated with the fastest rate of spread (the second region affected on average after 1.2 years). Our results demonstrated that the initial site of primary dystonia was relevant for the risk of spread.	**30**	**Svetel et al., 2007**	
11	**To evaluate efficacy and safety of botulinum toxin A injections after more than 10 consecutive years of treatment for benign essential blepharospasm and hemifacial spasm.**	Data collected included diagnoses and patient characteristics, injection dates, doses administered at each visit, response scores, duration of effect, and adverse events. There were no significant differences between genders or between benign essential blepharospasm and hemifacial spasm subgroups. The most common adverse events were ptosis, lagophthalmos and dry eye.	**64**	**Ababneh et al., 2014**	
12	**To describe the incidence, patient demographics, and risk factors associated with benign essential blepharospasm.**	Patients with diagnosis BEB between January 2000 and December 2013 were sampled using the Longitudinal Health Insurance Database 2000. Secondary blepharospasm that may be related to neurological, trauma, and ocular surface disease were excluded. Higher level of urbanization, white-collar work, sleep disorders, mental health diseases, dry eye-related diseases, Parkinsonism, and rosacea are possible risk factors for benign essential blepharospasm.	**1325**	**Sun et al., 2018**	
13	**In our clinical experience, we have observed an increased**	Within our BEB and HFS patient cohort, approximately 15% of patients presented with	**87**	**Khan et al., 2019**	

	prevalence of rosacea in patients who present with BEB and HFS. We investigate our clinical findings with a review of disease pathophysiology and treatment.	rosacea, compared to the general American population prevalence rate of 1.34% ($p < 0.001$). Of the 140 patients reviewed, a total of 21 patients (13 with BEB and 8 with HFS) exhibited rosacea ($p = 0.995$). Further biochemical analysis will likely be necessary to elucidate this potential association.			
14	**To investigate the correlations between motor, non-motor symptoms, and patients' clinical and demographic features.**	In all patients, the severity of blepharospasm was assessed clinically with the Blepharospasm Severity Rating Scale. All the participants underwent a psychiatric, sleep, cognitive, and ocular symptom evaluation. The frequency of psychiatric, sleep, and cognitive disorders and ocular symptoms was higher in blepharospasm patients than in healthy controls. Non-motor symptoms coexisted in the majority of patients and there was no correlation between non-motor and motor symptoms.	**60**	**Ferrazzano et al., 2019**	
15	**The correlations between motor, non-motor symptoms, and patients' clinical and demographic features was investigated.**	The relationship between demographic/clinical variables and spread of dystonia was assessed by Kaplan–Meier survival curves and Cox proportional hazard regression models. The difference in the risk of initial spread by site of onset was partly confounded by age at dystonia onset. Site of and age at dystonia onset, and age at first spread, were not significant predictors of the risk of a second spread.	**124**	**Abbruzzese et al., 2007**	
16	**Evaluation of cognitive performance in a series of patients with BEB and in a group of controls matched by severity of mood symptoms, age, and sex.**	BDI and BAI for subjective measurements Information (WAIS-III) 9.52 (1.04) 13.35 (1.78) 21.85 (26 DF) 0.07 Vocabulary (WAIS-III) 30.79 (2.32) 33.29 (3.22) 20.64 (34 DF) 0.52 Education level (in years) 8 (0.69) 8.23 (0.58) 20.26 (34 DF) 0.8 BDI, Beck depression inventory; BAI, Beck anxiety inventory; WAIS-III, Weschler Adult Intelligence Scale (version III). Depression and anxiety. Severity of BEB was scored using the BDSI disability	**20**	**Aleman et al.,2009**	

		scale. All patients underwent exhaustive neurological examinations, and specifically motor, sensory, visual, and auditory impairment were ruled out. Mild depression noted and visual skills worsen with increasing severity and duration of BEB.			
17	**To investigate symptom relief and neurocognitive change in response to treatment with botulinum toxin (BTX) in a group of patients with PCD/BEB**	patients were age- and educationally matched healthy individuals using tests of memory, sustained attention, span of auditory attention, and perceptual flexibility. The results support the view that executive dysfunction in PCD is secondary to the disrupting effects of the symptoms. Treatment with BTX alleviates the symptoms and, consequently, improves sustained attention.	**9**	**Allam et al., 2007**	
18	**To investigate causes, associations, and results of treatment with blepharospasm, patients extensive questionnaires on BEB and long-term results of treatment with the full myectomy operation, botulinum-A toxin, drug therapy, and help from the Benign Essential Blepharospasm Research Foundation (BEBRF**	The percent of patients improved by the BEBRF was 90%, full myectomy 88%, botulinum-A toxin 86%, and drug therapy 43%. The patient acceptance rate for the BEBRF was 96%, full myectomy 82%, botulinum-A toxin 95%, and drug therapy 57%. All patients should be informed of the support group	**1,653**	**Anderson et al., 1998**	
19	**Evaluation of quality of sleep matched healthy subjects.**	(Pittsburgh Sleep Quality Index, PSQI) and excessive daytime sleepiness (Epworth Sleepiness Scale, ESS) was utilised and the Beck Depression Inventory (BDI) was used for the evaluation of depressive symptomatology. This study suggests assessment and treatment of insomnia-related in BEB complaints should be considered in global management plans	**52**	**Avanzino et al., 2009**	
20	**The assessment of Botulinum toxin for benign essential blepharospasm, hemifacial spasm**	The treatment was effective in all three groups for an average of 3-4 months when symptoms recurred and repeated chemo-denervation with toxin was	**47**	**Carruthers & Stubbs, 1987**	

	and age-related lower eyelid entropion.	needed. The commonest complication was transient ptosis with an overall frequency of 7.8%. This incidence increased to 11.1% with toxin doses higher than 25 units per orbicularis. The treatment was well accepted by the patients, who were subsequently able to return to pre-blepharospasm lifestyles.			
21	**To assess the prevalence of obsessive-compulsive symptoms in patients suffering from BEB and in hemifacial spasm**	Structured Clinical Interview of 13 patients with Blepharospasm and 13 with hemifacial spasm completed the SCL-90-R and the Hamburg Obsession/Compulsion Inventory-Short Form. The findings provide additional support for the hypothesis that obsessive-compulsive symptoms are related to basal ganglia dysfunction.	**13**	**Broocks et al., 1998**	
22	**To compare the clinical characteristics and the long-term outcome of a large series of patients with blepharospasm (BEB) treated with the two most used brands of BTX over the last 15 years.**	Patient case notes were reviewed it showed the doses of Botox (beta = 0.40) and Dysport (beta = 0.16) were significantly increased over time. Side effects occurred in 325 out of 1341 treatments (24.2%): 21.8% of the patients who had received Botox, and in 31.6% of those who had received Dysport (P < 0.01). Both brands are effective and safe in treating blepharospasm; efficacy is long lasting. The differences in outcome and side effects suggest that the drugs should be considered two separate medication.	**128**	**Bentivoglio et al., 2009**	
23	**To compare the performance of patients with BM and HS in the Frontal Assessment Battery (FAB).**	Twenty-two patients with BEB and 29 patients with HFS were submitted to the FAB and the Mini-Mental State Examination (MMSE). FAB scores were compared between the two groups. Correlations between FAB and sociodemographic and clinical variables were calculated. The groups showed no difference in relation to gender, age, length of symptoms, educational level and performance in the MMSE. Executive functioning as assessed by FAB is not altered in BEB in comparison with HFS.	**22**	**Dias et al., 2009**	
24	**To identify risk**	Patients with BEB (n = 159)	**159**	**Hall et al., 2005**	

	factors associated with benign essential blepharospasm (BEB) with reference to hemifacial spasm (HFS). Persons with BEB and HFS experience similar physical symptoms, yet the two disorders have different etiologies.	or HFS (n = 91) were identified from two large neuro-ophthalmology clinics. Demographic, medical, behavioural, and psychological characteristics were obtained from chart review and a telephonic survey questionnaire. Most patients in both groups were retired, white, and female. BEB patients were more than two times as likely to meet the diagnostic criteria for generalized anxiety disorder than HFS patients (odds ratio, 2.13; 95% confidence interval, 1.22-3.72). BEB was significantly more often associated with generalized anxiety disorder			
25	**To analyse the clinical features of benign essential blepharospasm in Korean patients.**	Patients with BEB were evaluated using a clinical examination and questionnaire. The results shows majority of BEB patients are women and non-smokers. In contrast to previous reports though, fatigue and stress were aggravating factors, and the most common relieving factor was rest. No stressful events had immediately preceded the development of blepharospasm in 57.4% of patients.	**101**	**Lee et al., 2018**	
26	**To investigate the response to supramaximal doses of botulinum toxin in patients with refractory blepharospasm.**	Supramaximal dosages were well tolerated. Seven of 8 patients had a prolonged interval between injections relative to that associated with their previous dosing regimen. Four of the patients elected to continue with the new dosage. In select patients with essential blepharospasm who are refractory to standard treatment regimens, increasing the dosage of botulinum toxin above 100 U per session may decrease the interval between injections, improve the patient's quality of life, or both.	**8**	**Levy et al., 2006**	
27	**To determine whether the use of unique customized spectacles provided with modified side arms may be helpful in**	Consecutive patients using sensory tricks were recruited and the disease frequency and severity were assessed with the Jankovic Rating Scale (JRS) and the Blepharospasm Disability Index (BSDI) before and	**9**	**Lorenzano et al., 2019**	

	reducing benign essential blepharospasm (BEB) in patients describing periocular sensory tricks (ST).	after the use of the sensory trick frames (STF). A reduction in the score was noted in both severity (p = 0.0115) and frequency patterns (p = 0.0117) in the JRS in patients using the STF. A significant reduction of the BSDI score was also observed (p = 0.0314). This new device may be helpful in some selected BEB patients who previously responded positively to periocular pressure alleviating manoeuvres.			
28	**To assess sensory tricks in focal dystonia and hemifacial spasm**	Patients underwent a structured interview to assess sensory trick: presence, type and effectiveness. Statistical analysis used the Verisimilitude Reason test and the significance level was set at 5%. Sensory trick was more frequent in the cervical dystonia group (81%) when compared with HFS (38.5%; P=0.004) and writer's cramp (20%; P=0.001), but there was no statistical difference from the frequency in BEB patients (55%; P=0.078). The most common sensory tricks were facial massage (60%) in HFS; to touch the head with the hands (35.3% in cervical dystonia); to touch the upper eyelid with the index finger and thumb (81.8%) in BEB; and to touch the hand with the contralateral hand (100%) in Writer's cramp.	**20 BEB pts**	**Loyola et al., 2013**	
29	**To analyse the association between cranial dystonia and prior head trauma.**	The presence of a history of head trauma and of post-traumatic sequelae (loss of consciousness, bone fractures, scalp/facial wounds) before disease onset was recorded from 177 patients with primary adult-onset cranial dystonia and from 217 controls with primary hemifacial spasm matched by age strata and sex. Differences between groups were assessed by Mann–Whitney U test and Fisher's exact test, and the relationship between prior head trauma and case/control status was analysed by multivariate logistic regression models. No association was found between vault/maxillofacial	**133**	**Martino et al., 2007**	

		trauma and cranial dystonia.			
30	**To characterize patients with benign essential blepharospasm (BEB) by diagnosis, environmental risk factors and family history.**	Patients were evaluated through a clinical examination and questionnaire. BEB was more commonly found in females (2.8:1) and 93% of the patients were Caucasian. Fifty percent had pure BEB, 31% had BEB/Meige's syndrome, and 4% had BEB and eyelid opening apraxia (+/- Meige's syndrome). A minority of patients reported preceding photophobia (25%) or other eye conditions (22%). The majority were non-smokers, had no exposure to anti-emetic or antipsychotic agents, had a normal birth history and had no history of head trauma. Seventy-two percent did report a stressful event immediately prior to the development of symptoms. Treatments reported included botulinum toxin (BoNT), oral medications, surgical procedures and acupuncture. Thirty-two percent of patients reported a family history of focal dystonia and BEB was the most commonly reported.	**240**	**Peckham et al., 2014**	
31	**To characterize satisfaction with current standard-of-care botulinum neurotoxin type A (BoNT/A) treatment for blepharospasm, we performed a cross-sectional, structured survey in subjects with blepharospasm who had received ≥2 BoNT/A cycles.**	Patients were interviewed immediately before re-injection to evaluate treatment satisfaction, time course of treatment effects, preferred injection intervals, Jankovic Rating Scale (JRS), and Blepharospasm Disability Index (BSDI). he most frequent injection interval was 12weeks (46.5% subjects); 30.7% had an interval >12weeks. The main rationale for interval choice was "to maintain treatment efficacy" (44.7%). However, 36.6% reported that treatment effects usually declined within 8weeks; 69.6% within 10weeks. BEB symptoms, such as difficulties to drive and read, re-emerge at the end of a BoNT treatment cycle and that flexible, individualized treatment intervals may improve treatment satisfaction and outcomes.	**114**	**Fezza et al., 2016**	
32	**Efficacy and**	300 patients received study	**300**	**Roggenkamper et al.,**	

	safety of a new Botulinum Toxin Type A free of complexing proteins in the treatment of blepharospasm	medication (intent-to-treat population), and 256 patients completed the study as planned (per-protocol population). At baseline, patients received a single injection of NT 201 or BOTOX (<or=35 units per eye). No significant differences were found between NT 201 and BOTOX for all efficacy and safety variables three weeks after injection. T 201 and the BOTOX group showed a decrease in the Jankovic Rating Scale (JRS) sum score signifying an improvement in the symptoms of blepharospasm during this time period. These data show that NT 201* is an effective and safe treatment for patients suffering from blepharospasm.		**2006**	
33	**To evaluate the characteristics of the essential blepharospasm and hemifacial spasm patients and the feasible treatment with botulinum toxin A.**	Patients were evaluated according to gender, ocular complaint, time of disease, treatment outcome and complications. Age median was 63 years and the mean was 61 years, with no difference regarding sex; 66.66% of the patients had hemifacial spasm and 33.33%, essential blepharospasm. Many patients complained of dry eye associated with involuntary spasm. Botulinum toxin A showed a positive outcome in 91.30% of the treated patients and complications observed after treatment were eyelid ptosis (8.33%) and buccal angle deviation (8.33%).	**34**	**Schellini et al., 2006**	
34	**To explore the efficacy of botulinum toxin type A (BTX-A) therapy in relieving anxiety and depression in patients with hemifacial spasm (HFS) and benign essential blepharospasm (BEB).**	The anxiety and depression status were evaluated by self-rating anxiety scale (SAS) and self-rating depression scale (SDS), respectively, before and after the injection of BTX. Before treatment, the SAS scores of male and female BEB patients were 56.45±8.75 and 60.89±9.11, respectively, and the SDS scores of male and female BEB patients were 57.90±7.93 and 60.12±8.35, respectively. After treatment, the SAS score was 38.17±3.67 and the SDS score was 38.12±4.15, with a significant difference in	**90**	**Dong et al., 2019**	

		before and after treatment scores. In HFS and BEB, especially in female patients, there is an association with anxiety and depression. BTX-A can improve the symptoms of anxiety and depression.			
35	**Study investigated the frequency and types of psychiatric disturbances in 89 consecutive patients with various primary focal dystonias**	Diagnosis was based on the structured clinical interview for DSM-IV, obsessive-compulsive disorder (OCD) was assessed with the Yale-Brown Obsessive-Compulsive scale, anxiety with the Hamilton Rating Scale for Anxiety, the severity of depression with the Beck Depression Inventory. No differences were found in age, dystonia severity, and duration of botulinum toxin treatment between patients with and without psychiatric disturbances. The most common psychiatric features in patients with CD and BEB are depressive disorders.	**28**	**Fabbrini et al., 2010**	
36	**To evaluate the natural history and response to different treatments have been evaluated in 264 patients with blepharospasm.**	The mean age of onset was 55 8 years and there was a female preponderance of 1 8 to 1. Dystonia elsewhere was found in 78% of patients, usually in the cranial cervical region, and appeared to follow a somatotopic progression. A family history of BEB and or dystonia elsewhere in 9-5% of cases, suggesting genetic predisposition. Ocular lesions preceded the onset of blepharospasm in 12- 1% of cases. Response to drugs was inconsistent, although initial improvement was experienced by one fifth in patients treated with anticholinergics. Reoccurrence of symptoms following in 22 out of 27 patients, on average one year after surgery. Botulinum toxin injections were performed in 151 patients. Significant improvement was achieved in 118 cases. Mean duration of benefit was 9-2 weeks. Transient ptosis and diplopia were the commonest side effects.	**264**	**Grandas et al., 1988**	
37	**To determine the frequency of obsessive and compulsive**	Patients were evaluated with structured interview based on the DSM-IV criteria and the Yale-Brown	**30**	**Munhoz et al., 2005**	

	symptoms in patients with BS in comparison with patients with HFS.	scale. Obsessive or compulsive symptoms in 20 (66.6%) patients with BEB and 21 (70%) with HFS. Yale-Brown scale scores for each group were higher among BEB patients: however, differences were not statistically significant. Conclusion: Our study did not show a significant difference in the comparison of the prevalence of obsessive and compulsive symptoms among patients with BEB and HFS.			
38	**Aim of this study was to investigate the long-term results of botulinum toxin A (BoNT-A) injections for the treatment of benign essential blepharospasm (BEB) and to report our experience with (ultra-)long-term treatment with onabotulinumtoxin-A.**	Patients with at least 21 BoNT-A injections were defined as long-term patients. As most of these patients had received onabotulinumtoxin A in at least 95% of injections, detailed analyses of BoNT-A long-term treatment were performed with respect to this homogeneous onabotulinumtoxin A long-term group). In terms of demographics, the two groups compare well, with a slightly higher proportion of women and patients with frontalis-suspension in the Ona Long-Term Group.	**234**	**Bettina Wabbels , Rolf Fimmers and Peter Roggenkämper 2022**	
39	**The purpose of this study was to provide a comprehensive picture of its clinical features including presenting features, motor features, and non-motor features.**	For cases in the literature review and the cohort study, blepharospasm emerged during the 5th decade of life and was more frequent in women. Many presented with non-specific motor symptoms such as increased blinking (51.9%) or non-motor sensory features such as eye soreness or pain (38.7%), photophobia (35.5%), or dry eyes (10.7%). Non-motor psychiatric features were also common including anxiety disorders (34–40%) and depression (21–24%). Among cases presenting with blepharospasm in the Dystonia Coalition cohort, 61% experienced spread of dystonia to other regions, most commonly the oromandibular region and neck. Features associated with spread included severity of blepharospasm,	**10324 systematic literature review** **884 case enrolled cohort study**	**Cho et al., 2022**	

		family history of dystonia, depression, and anxiety.			
40	**The objective of this systematic review is to summarise and compare all the published data regarding benign essential blepharospasm treatment with botulinum toxin.**	Four out of five studies included studies used onabotulinumtoxin A and one study used abobotulinumtoxin A. All studies injected the pretarsal orbicularis occuli muscle. The preseptal orbicularis occuli was injected in four studies, and the preorbital muscle in three studies. The most commonly used method of evaluation was the Jankovic Rating Scale. Adverse events were transient, and dose related. Ptosis was more frequently encountered with the preseptal orbicularis injections.	**854**	**Rayess et al., 2021**	
41	**To describe the efficacy and safety of botulinum toxin A (Botox) in patients with benign essential blepharospasm.**	Retrospective review of operation, injection, and medical records. Botox was a safe and effective treatment in benign essential blepharospasm which required an increased dosage over time. Ptosis was the most common complication and no life-threatening condition was reported. Surgical correction of those with eyelid diseases showed subjective improvement of subsequent Botox treatment.	**29**	**Lai et al., 2020**	

Appendix 6A

Table: JBI LEVELS OF EVIDENCE FOR MEANINGFULNESS

Levels:	Evidence of Meaningfulness
1	Qualitative or mixed-methods systematic review
2	Qualitative or mixed-methods synthesis
3	Single qualitative study
4	Systematic review of expert opinion
5	Expert opinion

APPENDIX 6B: Critical Appraisal for Text and Opinion Summary of Findings with Level of Meaningfulness

Text and Opinion Citation Included	Q1	Q2	Q3	Q4	Q5	Q6	**Comments and Article Conclusion**	Level of Meaningfulness
Smith, C. (2011). Business as usual! *Dystonia Matters!*, *71*.	Y	Y	Y	Y	N	N	Misdiagnosed with dry eyes, then myasthenia prescribed steroids. After two and half years, she was referred to the "boss" and was diagnosed with blepharospasm. The treatment BTX is now effective after having surgery. As she owns her company, she did not lose her job. Now she can walk without difficulties, drive. Found it difficult to join in conversation due to poor eye contact and low confidence, but her friends and family helped. She is no longer dependent on her husband. She is upbeat now.	Reflecton Low: 5
Tredgold, C. (2008). My story: My eyelids fell shut heavily. *Dystonia Matters!*, *59*.	Y	Y	Y	Y	N	N	Denial, blindness, loss of independence. BTX was magical. *"…I began to welcome back the old abilities, the ones we take for granted until we don't have them anymore."* To educate	Reflection Low: 5
Case Study Howard Jenson Dystonia Matters! Issue 68, 2010	Y	Y	Y	Y	N	N	Blindness, misdiagnosis, *"My eyes would close for indeterminate periods, perhaps only seconds, but both scary and potentially dangerous."* *"So far, I have had four sets of injections, and each has had a different response. I don't feel the correct regime has been found yet."* To educate	Reflection Low: 5
Lownsborough, M. (2011). With eyes wide open. *Dystonia Matters!*, *72*.	Y	Y	Y	Y	N	N	The inability to control the eyelids caused an emotionally scarring, two-year journey to find a diagnosis and treatment for her dystonia. In the OPD, finally, an ophthalmologist gave me his usual medication pattern, and when it didn't work, it was decided it was psychological. Desperate for answers, a second opinion was sought, and finally, successful treatment. The frustration I felt caused my whole body to go into spasms—the article is written to raise awareness and encourage others to find answers.	Reflection Low: 5
Blepharospasm Australia. (n.d.). Case histories. Retrieved from: https://www.beb.org.au/managing-beb/case-histories/ "You need to work with your doctor – trial and error – to find the optimal product and dose that suits you, so you need to be patient. There is a lot of information now on the useful Internet, and there are sites like this one where you can find support and can contact	Y	Y	Y	Y	N	N	**1.** Misdiagnosis, lack of knowledge. Poorly organised clinics. *"The injections didn't last that long for me, so by the time the next clinic came round, I was back to square one, and the treatment had to start all over again. It was very frustrating and extremely stressful as I could never depend upon getting the injections when I needed them. I felt the clinics were held to suit the doctor and not the patient."* **2.** Lack of knowledge. I visited an ophthalmologist who said, *"It was just my age 2. The colour of my eyes and skin 3. That I would just have to wear dark glasses 4. That I would just have to learn to live with it"*. I asked my GP for a referral to a specialist eye institute. It was immediately diagnosed. Knowing it wasn't my imagination was sheer relief. Botox was given, but I wasn't happy. So, I moved further away from home again. Now I am happy with my ophthalmologist. We experimented with the dose but to make it last longer, the more significant dose caused the eyelid to drop, so now I make it last 12 weeks, but the last two weeks is complex, and I cannot drive during that time." **3.** Blindness, disability, misdiagnosis and a lack of empathy*; "an ophthalmologist. He told me, as far as he could see, there was nothing wrong with my eyes"* GP referred me to an ophthalmologist. He told me that there was nothing wrong with my eyes as far as he could see.	1-9 Reflection Low: 5

others in your area to compare experiences”.							Back to the GP again, finally referred to another ophthalmologist. In the OPD, I had side effects and bruising over the years. I saw Dr. X yesterday, and today, I am feeling the effects of the injections, but within a few days, the bruising will go away, and my blinking will ease, and I will be able to carry on like usual. “Recently, I had to cancel an appointment and went two weeks over the three-month limit. The difference was striking. My blinking became almost uncontrollable again. To educate **4.** Misdiagnosis and a lack of empathy *“at one stage, I told him I thought I had a neurological problem – he laughed at me and said the sooner I accepted that I had dry eyes and moved on, the better off I would be and would not refer me to a specialist.”* With Botox, *“Wow, I was on my way to normality again.”* After two weeks, I had a follow-up visit with the doctor, and he was happy with my progress. He also strongly suggested that I contact the BEB Support Group. To educate **5.** Disability, misdiagnosis *“By now, I’m getting annoyed with the issue and the lack of attention and support from the medical profession. I’m six months in, and I’m still being told to take different drops!”* Back to the GP. I decide to open up about my depressive episodes, sleepless nights, and occasional suicidal thoughts. I told him I’d tried everything I could. I told him about the apparent lack of concern and knowledge of whatever I was going through. The third ophthalmologist diagnosed me with BEB. I’ve had the first lot of Botox injections, and on about the sixth day, I announced to my family, friends, and colleagues, ‘I CAN SEE!’. Unfortunately, it only lasted about four weeks. I have managed to ride my bike 700kms in a week, raising money for youth suicide and depression. I can also drive my car but only for about four to five weeks after the treatment takes effect. I’m advised It takes a while to get the dosage right, so I was hoping I would eventually get there. *“I now have an interested Specialist willing to work through this with me and the knowledge there is a support group - Blepharospasm Australia, with dedicated people assisting those diagnosed.”* **6.** Misdiagnosis My GP prescribed anti-depressants. I was referred to an ophthalmologist. *“He mentioned my age lots of times and said as you get old, your eyes get dry. He told me to get some eye drops. I tried lots of different types of drops, but none seemed to help.”* I asked to be referred to another. My new ophthalmologist discussed the problem and told me the treatment I would need to keep it under control. He gave me 14 injections using half a dose which he said was a lot, but I needed that many. It was excruciating but undoubtedly worth it. I had to return in 2 weeks for a top-up, but he said he was happy with the result and that I didn’t need a top-up to be back in 2 months. It is a shame it has finally taken so many years to get some relief. *“My sister and I went to the Support Group meeting. It was great to meet fellow sufferers and learn how they coped with the problem.”* To educate **7.** Misdiagnosis *“Oh, you’re stressed*”. I emphatically told him I didn’t have this problem because I was stressed. I was stressed because I had this problem to no avail. I kept getting worse and eventually agreed to go to a therapist. Maybe he was right. On one occasion, she walked through open doors forgetting that I was in direct line to the ajar doors. Slap bang, I went straight into them. There was a lot of crying. I then had lots of surgery, one to lift and realign the muscle above the eye and fix the eyelids. This transformed my sight as it lets more light into my eyes. The ophthalmologist said nothing about me having BEB. I have felt that my eyes have slowly worsened in the last five years. The optician asked me what I was doing about my blepharospasm just out of the blue. I burst into tears.	

							8. At last, a professional had recognised it. I was referred. I have had Botox (about every four months) for about 18 months, and she has also done another lot of surgery on my eyelids. It seemed a long lonely journey. I tried to contact a support group a few years ago but failed. So, to be in touch now is lovely. I want to start a group here in S.A. I would also like to make the medical profession more aware of BEB. *“I would also like to make the medical profession more aware of BEB. Thank you for taking the time to read my story. It means a lot to me to be able to share it.”* **9.** Misdiagnosis, Inadequate *treatment “I was unable to have an appointment for three months, my life was coming to a standstill – some days I considered myself unsafe to drive my car.” “I found the injections had little effect, but he wanted me to wait another six weeks before repeating the treatment. By this time, I had stressed entirely out … spasms were frequent and severe.”* **10.** Misdiagnosis. *“There will be people today going through this trauma.”* I was eventually referred to an ophthalmologist who was able to provide Botox. He gave me my first injections. I have been seeing him ever since and getting on with my life. What is typical is that the first diagnosis is almost always dry eye. Then the unfortunate sufferer has to scratch around till they stumble across somebody who diagnoses the problem accurately as blepharospasm and refers them for effective treatment. It still took a three-step process to find treatment. There will be people today going through this trauma.	
Kranz, R. (2015, Fall). My Dystonia Story. *Dystonia Canada Report*, 6. https://issuu.com/dmrfc/docs/dystonia_report_can_fall2015_oct7_f/8	Y	Y	Y	Y	Y	Y	Misdiagnosis. *“Journey to diagnosis During the previous year and a half, I had visited the optometrist three times complaining of issues with my vision*.” Progressive to head and voice. It was a relief when I was finally diagnosed, but really, it was just the beginning... Learning I wasn’t alone. To educate and offer support	Reflection Low: 5
About Lori. (n.d.). Retrieved from: http://www.oocities.org/lorisgarden/AboutLori.html	Y	Y	Y	N	N	Y	Misdiagnosis. Progressive disease. The journey involved being misdiagnosed by the medical society and dismissed as a psychiatric condition, sinking into depression, isolation, seclusion. One day holding the eye open, I had a car accident. I was misdiagnosed as myasthenia gravis. I became depressed. The psychologist referred me to a neurologist, and he diagnosed me. I cried with joy. He gives me Botox and my life back. Botox worked well after surgery. It only lasted 4-6 weeks, but after myectomy surgery, it now lasted for three months. I want to help now; due to the lack of education, there are many people out there suffering, and each time I get a call during my job at the BEBRF, my heart sinks. Hearing their stories takes me back to my experience. There might be others alone and suffering, so there is a need to educate.	Reflection Low: 5
Collin’s Story DMRFC Email: Info@dystoniacanada.org	Y	Y	Y	Y	Y	N	Misdiagnosis and progressive diagnosis neck, voice, writers’ cramp. After seeing over 11 specialists of varying degrees, I was lucky enough to have an appointment with a Neurologist. I tried neurotoxin injections for my blepharospasm, and after a few tries, we had good results! I was now able to drive my car again, which was a blessing in itself. I was treated by an eye specialist who was a leader in using neurotoxin injections. After about six months, I started having trouble with my speech; around three in the afternoon, I was experiencing difficulty with my pronunciation of some words. I have written this to possibly give hope to some of those suffering from this dreadful debilitating neurological disorder. Don't give up, and never stop looking for answers. The	Reflection Low: 5

							power of positive thinking is impressive as to what it can do and lead.	
Sean's Post USA BEBRF: https://www.blepharospasm.org/bulletin-board.html	Y	Y	Y	Y	N	N	Misdiagnosis. I started receiving Botox in January 2015, and the relief was almost immediately 100% effective. It was fantastic! Over the next couple of years, I regularly received the treatments and experimented with the dosages to get the longest-lasting benefit. At first, I had to practice blinking to stop the spasms. I remember counting to 100 blinks on several occasions. Now, I can sense when the spasms are about to occur and start blinking to counteract almost without thought. *"I post this only to offer encouragement to the others afflicted with this horrible disease. I hope this tool will help some others deal with it."*	Reflection Low: 5
Kirk's Post : USA BEBRF https://www.blepharospasm.org/bulletin-board.htm	Y	Y	Y	Y	Y	Y	Misdiagnosis, a lack of empathy. *"The condition almost destroyed my life. The hardest part for me is, none of the doctors seem interested in helping"* Botox isn't helping. I have a desire to help people with this horrible condition, but I need help	Reflection Low: 5
Nicole's Post https://www.blepharospasm.org/bulletin-board.html	Y	Y	Y	Y	N	N	Misdiagnosis. They both refused to use Botox to manage the spasms and preferred I try a course of anti-anxiety meds and Cognitive Behavioural Therapy to rewire my brain. I use Zoloft and Klonopin and have seen definite improvements, but I am far from 100%. I felt stoned half the time from the Klonopin. Need help	Reflection Low: 5
Bob's Post https://www.blepharospasm.org/bulletin-board.html	Y	Y	Y	Y	N	N	It made my life unbearable. I found myself going to bed at 7-8 o'clock just to escape the pain. I tried several things, including injections. Nothing helped. The injections brought some relief, but the side effects were terrible. Control blink therapy is working. To educate	Reflection Low:5
Needle, A. (2015). A struggle to forgive My long battle with dystonia. *Psychiatric Services*, *66*, 568–569. Retrieved from: https://ps.psychiatryonline.org/doi/10.1176/appi.ps.660601	Y	Y	Y	Y	N	N	Lack of information, the need to educate others. *"Informative, not coercive, and I would have felt I had made an informed choice."*	Reflection Low: 5
Boodman S. G. (2012, April 23). Medical mysteries: "I open my laptop, and my eyes snapped shut." *The Washington Post*. Retrieved from: http://www.washingtonpost	Y	Y	Y	Y	Y	Y	Misdiagnosis. The injections made a huge difference, she said, although she says the 14 painful shots, she receives around her eyes every three months *"feel like 14 flu shots."* She is fearful of worsening conditions. Living with adjustments, but it is very little she can't do now. Written to raise awareness	Reflection Low: 5

.com/								
Bluebird's Post http://www.blepharospasm.org/forumarchieves/beb4/87094.html	Y	Y	Y	N	N	Y	Misdiagnosis, functional blindness. Since I was diagnosed, I have had eyelid surgery or myomectomy now. It works well. Botox wears off in 8 weeks. Written to give hopes	Reflection Low: 5
Indiana's Post http://www.blepharospasm.org/forumarchieves/beb4/87094.html	Y	Y	Y	N	N	Y	BEB is a condition that needs a strong support group. Without experiencing the symptoms is hard to know. Misdiagnosis. Physicians try hard, but they rely on dry eyes, conversion, depression. Botox is terrific but keep researching. Need for information	Reflection Low: 5

Appendix 6C: Excluded Studies at Screening

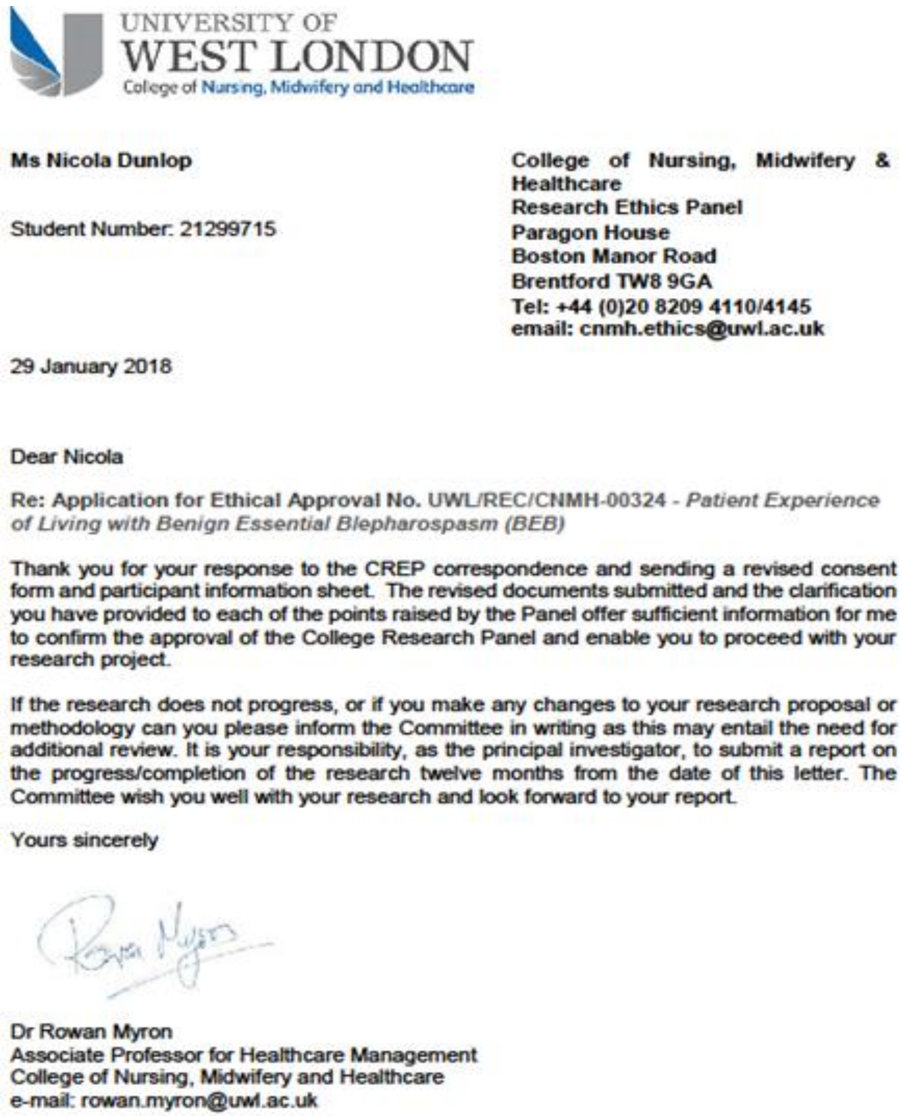
Excluded Studies At Screening								
Kuyper, D. J., Parra, V., Aerts, S., Okun, M. S., & Kluger, B. M. (2011). Nonmotor manifestations of dystonia: A systematic review. *Movement Disorders*, *26*, 1206–1217.	Y	Y	N	Y	Y	Y		Includes both qualitative and quantitative methods
Girach, A., Vinagre Aragon, A. & Zis, P. (2019) Quality of life in idiopathic dystonia: a systematic review. J Neurol 266, 2897-2906.	Y	Y	Y	Y	Y	Y		Included both qualitative and quantitative methods
Hartenstein, E., Tang, N. K. Y., Bernstein, C. J., Nissen, C., Underwood, M. R., & Sandhu, H. K. (2016). Sleep in patients with primary dystonia: A systematic review on the state of research and perspectives. *Sleep Medicine Reviews*, *26*, 95–107	Y	Y	N	Y	Y	Y		Includes both qualitative and quantitative methods
Robbins, C. M. (1996). The experience of women with BEB: A phenomenological study.	Y	Y	N	Y	Y	Y		Unable to locate full text of this study

Silveira-Moriyama, L., Gonçalves, L. R., Chien, H. F., & Barbosa, E. R. (2005). BTX A in the treatment of blepharospasm: 10-year experience. *Arguidos de Neuro-Psiquiatria*, *63*(2a), 221–224	Y	Y	N	Y	Y	Y		Qualitative and quantitative retrospective analysis utilised Columbia Rating
Krack P, & Marion, M. H. (1994). "Apraxia of lid opening," focal eyelid dystonia: Clinical study of 32 patients. *Movement Disorders*, *9*, 610–615.	Y	Y	N	Y	Y	Y		A prospective and retrospective study with qualitative and quantitative and utilises a rating scale
Bernstein, C. J., Ellard, D. R., Davies, G., Hertenstein, E., Tang, N. K. Y., Underwood, M., & Sandhu, H. (2016). Behavioural interventions for people living with adult-onset primary dystonia: A systematic review. *BMC Neurology*, *16*, 1–14.	Y	Y	N	Y	Y	Y		Include qualitative and quantitative risk bias and control cohort
Dressler, D. (2000). Complete secondary BTX therapy failure in blepharospasm. *Journal of Neurology*, *247*, 809–810	Y	Y	N	Y	Y	Y		Qualitative and quantitative and included, the study utilised rating scales.

JBI Critical Appraisal Checklist for Text and Opinion Papers Q1. Is the source of the opinion clearly identified? Q2. Does the source of opinion have standing in the field of expertise? Q3. Are the interests of the relevant population the central focus of the opinion? Q4. Is the stated position the result of an analytical process, and is there logic in the opinion expressed? Q5. Is there reference to the extant literature? Q6. Is there any incongruence with the literature/source logically defended?

https://jbi-global-wiki.refined.site/space/MANUAL/4689783

Appendix 7



UNIVERSITY OF
WEST LONDON
College of Nursing, Midwifery and Healthcare

Ms Nicola Dunlop

Student Number: 21299715

College of Nursing, Midwifery & Healthcare
Research Ethics Panel
Paragon House
Boston Manor Road
Brentford TW8 9GA
Tel: +44 (0)20 8209 4110/4145
email: cnmh.ethics@uwl.ac.uk

29 January 2018

Dear Nicola

Re: Application for Ethical Approval No. UWL/REC/CNMH-00324 - *Patient Experience of Living with Benign Essential Blepharospasm (BEB)*

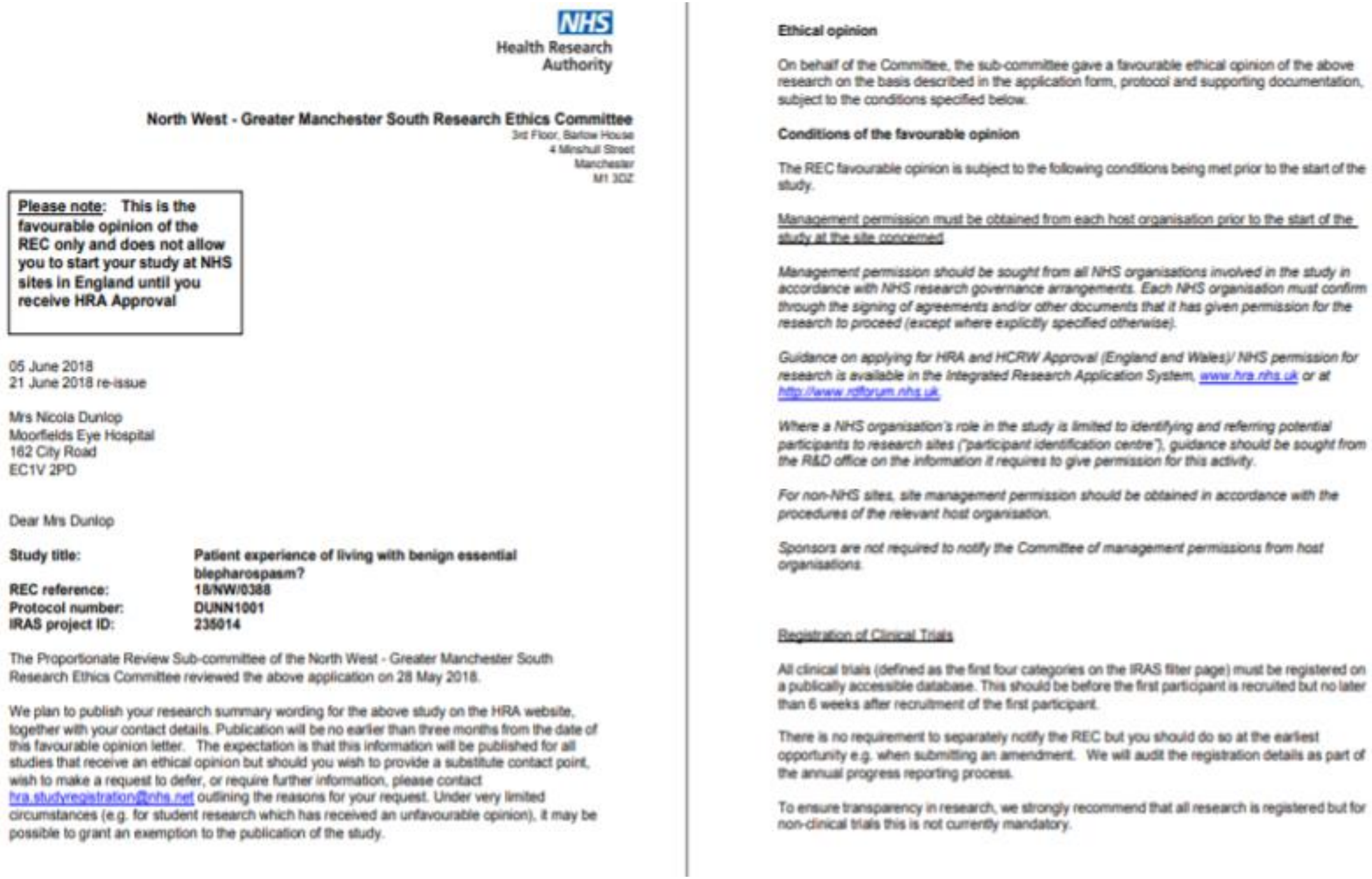
Thank you for your response to the CREP correspondence and sending a revised consent form and participant information sheet. The revised documents submitted and the clarification you have provided to each of the points raised by the Panel offer sufficient information for me to confirm the approval of the College Research Panel and enable you to proceed with your research project.

If the research does not progress, or if you make any changes to your research proposal or methodology can you please inform the Committee in writing as this may entail the need for additional review. It is your responsibility, as the principal investigator, to submit a report on the progress/completion of the research twelve months from the date of this letter. The Committee wish you well with your research and look forward to your report.

Yours sincerely

Rowan Myron

Dr Rowan Myron
Associate Professor for Healthcare Management
College of Nursing, Midwifery and Healthcare
e-mail: rowan.myron@uwl.ac.uk



NHS
Health Research
Authority

North West - Greater Manchester South Research Ethics Committee
3rd Floor, Barlow House
4 Minshull Street
Manchester
M1 3DZ

Please note: This is the favourable opinion of the REC only and does not allow you to start your study at NHS sites in England until you receive HRA Approval

05 June 2018
21 June 2018 re-issue

Mrs Nicola Dunlop
Moorfields Eye Hospital
162 City Road
EC1V 2PD

Dear Mrs Dunlop

Study title:	**Patient experience of living with benign essential blepharospasm?**
REC reference:	**18/NW/0388**
Protocol number:	**DUNN1001**
IRAS project ID:	**235014**

The Proportionate Review Sub-committee of the North West - Greater Manchester South Research Ethics Committee reviewed the above application on 28 May 2018.

We plan to publish your research summary wording for the above study on the HRA website, together with your contact details. Publication will be no earlier than three months from the date of this favourable opinion letter. The expectation is that this information will be published for all studies that receive an ethical opinion but should you wish to provide a substitute contact point, wish to make a request to defer, or require further information, please contact hra.studyregistration@nhs.net outlining the reasons for your request. Under very limited circumstances (e.g. for student research which has received an unfavourable opinion), it may be possible to grant an exemption to the publication of the study.

Ethical opinion

On behalf of the Committee, the sub-committee gave a favourable ethical opinion of the above research on the basis described in the application form, protocol and supporting documentation, subject to the conditions specified below.

Conditions of the favourable opinion

The REC favourable opinion is subject to the following conditions being met prior to the start of the study.

Management permission must be obtained from each host organisation prior to the start of the study at the site concerned.

Management permission should be sought from all NHS organisations involved in the study in accordance with NHS research governance arrangements. Each NHS organisation must confirm through the signing of agreements and/or other documents that it has given permission for the research to proceed (except where explicitly specified otherwise).

Guidance on applying for HRA and HCRW Approval (England and Wales)/ NHS permission for research is available in the Integrated Research Application System, www.hra.nhs.uk or at http://www.rdforum.nhs.uk.

Where a NHS organisation's role in the study is limited to identifying and referring potential participants to research sites ("participant identification centre"), guidance should be sought from the R&D office on the information it requires to give permission for this activity.

For non-NHS sites, site management permission should be obtained in accordance with the procedures of the relevant host organisation.

Sponsors are not required to notify the Committee of management permissions from host organisations.

Registration of Clinical Trials

All clinical trials (defined as the first four categories on the IRAS filter page) must be registered on a publically accessible database. This should be before the first participant is recruited but no later than 6 weeks after recruitment of the first participant.

There is no requirement to separately notify the REC but you should do so at the earliest opportunity e.g. when submitting an amendment. We will audit the registration details as part of the annual progress reporting process.

To ensure transparency in research, we strongly recommend that all research is registered but for non-clinical trials this is not currently mandatory.

If a sponsor wishes to request a deferral for study registration within the required timeframe, they should contact hra.studyregistration@nhs.net. The expectation is that all clinical trials will be registered, however, in exceptional circumstances non registration may be permissible with prior agreement from the HRA. Guidance on where to register is provided on the HRA website.

It is the responsibility of the sponsor to ensure that all the conditions are complied with before the start of the study or its initiation at a particular site (as applicable).

Ethical review of research sites

The favourable opinion applies to all NHS sites taking part in the study, subject to management permission being obtained from the NHS/HSC R&D office prior to the start of the study (see "Conditions of the favourable opinion").

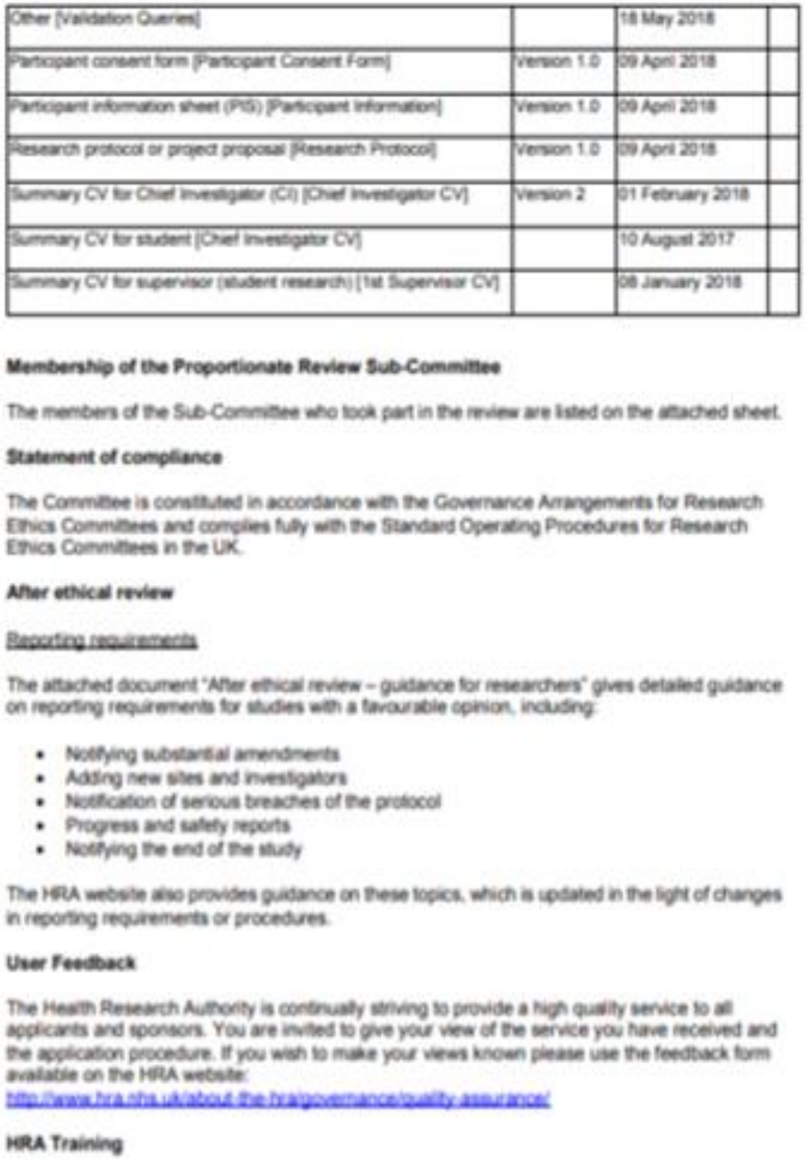
Summary of discussion at the meeting

The Committee considered whether there was a potential for bias and coercion as participants would be identified, approached and interviewed by the same person. After some further discussion with input from the researcher, the Committee was satisfied that this was not a concern.

Approved documents

The documents reviewed and approved were:

Document	*Version*	*Date*	
Confirmation of any other Regulatory Approvals (e.g. CAG) and all correspondence [University Ethics Approval 29.1.18]		29 January 2018	
Covering letter on headed paper [HRA Cover letter]		03 May 2018	
Covering letter on headed paper [Response to Validation Queries]		18 May 2018	
Interview schedules or topic guides for participants [Plan Interview Schedule]	Version 1.0	09 April 2018	
IRAS Application Form [IRAS_Form_09052018]		09 May 2018	
IRAS Checklist XML [Checklist_09052018]		09 May 2018	
Letter from funder [Grant Letter]		20 April 2018	
Letter from sponsor [Sponsor Letter]		11 April 2018	
Other [Student on the degree of Doctor of Nursing Correspondence]			
Other [2nd Supervisor CV]		08 January 2018	
Other [University Conditional Ethics]		15 January 2018	
Other [Supervisor Proportionate Review Support]		16 February 2018	



Other [Validation Queries]		18 May 2018	
Participant consent form [Participant Consent Form]	Version 1.0	09 April 2018	
Participant information sheet (PIS) [Participant Information]	Version 1.0	09 April 2018	
Research protocol or project proposal [Research Protocol]	Version 1.0	09 April 2018	
Summary CV for Chief Investigator (CI) [Chief Investigator CV]	Version 2	01 February 2018	
Summary CV for student [Chief Investigator CV]		10 August 2017	
Summary CV for supervisor (student research) [1st Supervisor CV]		08 January 2018	

Membership of the Proportionate Review Sub-Committee

The members of the Sub-Committee who took part in the review are listed on the attached sheet.

Statement of compliance

The Committee is constituted in accordance with the Governance Arrangements for Research Ethics Committees and complies fully with the Standard Operating Procedures for Research Ethics Committees in the UK.

After ethical review

Reporting requirements

The attached document "After ethical review – guidance for researchers" gives detailed guidance on reporting requirements for studies with a favourable opinion, including:

- Notifying substantial amendments
- Adding new sites and investigators
- Notification of serious breaches of the protocol
- Progress and safety reports
- Notifying the end of the study

The HRA website also provides guidance on these topics, which is updated in the light of changes in reporting requirements or procedures.

User Feedback

The Health Research Authority is continually striving to provide a high quality service to all applicants and sponsors. You are invited to give your view of the service you have received and the application procedure. If you wish to make your views known please use the feedback form available on the HRA website:
http://www.hra.nhs.uk/about-the-hra/governance/quality-assurance/

HRA Training

Moorfields Eye Hospital NHS
NHS Foundation Trust

Date 30th April 2018

Dear Nicola

Project Title: Patient Experience of Living with Benign Essential Blepharospasm

R&D Reference: DUNN1001

Following the review of the following documents:

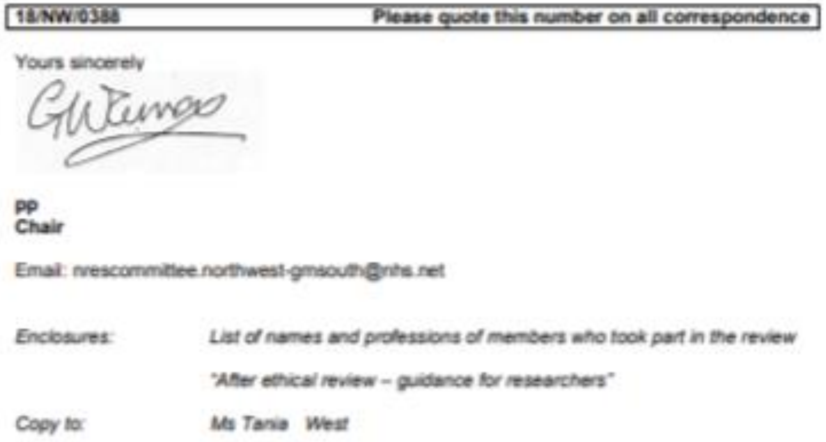
1) Protocol [v1.0_09.04.2018]
2) Other Supporting Documents:
   - Patient Information Leaflet [v1.0_09.04.2018
   - Informed Consent Form (v1.0_09.04.2018]
   - Plan Interview Schedule [v1.0_09.04.2018]
   - University conditional ethics approval [15.01.2017]
   - University chair's action letter [29.01.2018]

Moorfields Eye Hospital NHS Foundation Trust is pleased to offer sponsorship, confirmed by the Trust Research Management Committee (RMC) 11th April 2018. This letter will allow you to state in your Ethics and HRA applications that Moorfields will sponsor your study, subject to the requisite approvals being in place.

Please ensure that all Ethics and HRA opinions/approvals are forwarded to the sponsor, and that the full set of HRA approved study documents are sent to the sponsor once HRA approval is issued.

Please note this letter is **NOT** your management permission letter (confirmation of local capacity and capability). This will be granted following Ethics and HRA approval. You cannot commence your study until you have received your management permission letter as doing so will constitute a serious breach of Good Clinical Practice.

Yours sincerely



18/NW/0388	Please quote this number on all correspondence

Yours sincerely

PP
Chair

Email: nrescommittee.northwest-gmsouth@nhs.net

Enclosures:	*List of names and professions of members who took part in the review*
	"After ethical review – guidance for researchers"
Copy to:	*Ms Tania West*

Appendix 8

UNIVERSITY OF
WEST LONDON

Chief Investigator: Nicola Dunlop

Study Title: Patient experience of living with Benign Essential Blepharospasm

PARTICIPANT INFORMATION

What is the study about?

This study seeks to understand how benign essential blepharospasm, outpatient care, and treatment impact patients' lives.

What is the purpose of the study?

The NHS seeks to improve the quality of care it delivers to patients. Therefore, it is essential to understand how certain conditions affect the daily lives and needs of the patients – our service users. There is very little known about the devastating effects of this condition. This knowledge should be emphasized.

Moorfields Eye Hospital values patients. We need patients to raise their voices about this condition. We want to hear about how your life is affected by this condition and how your condition is affected by how we deliver treatment and care. We would like to hear your views, as this will create more profound insight and understanding of the condition and positively enhance service improvements in the outpatient clinic.

Who is the principal researcher of this study?

Nicola Dunlop is the principal researcher. She is a doctoral student and a nurse consultant working in the outpatient clinic, treating patients affected by this condition.

Do I have to take part?

Participation is voluntary. However, if you decide to take part, you will be required to sign a consent form but can withdraw from the study at any time without giving a reason and without affecting the care you receive.

Why have I been chosen?

You have been chosen because you live with the condition benign essential blepharospasm and can openly communicate the effect of your medical condition in your daily life.

What will I have to do if I decide to take part?

You will be asked to attend a 1-hour interview and focus group discussion in the Research Department at Moorfields Eye Hospital. The interview will occur on the day of your clinic appointment. Once the interview data have been transcribed, you will be given the transcript for comments and verify the interview transcribe is an accurate representation of the interview. You will need to attend a focus group to discuss the study results and offer your feedback to identify everyday experiences amongst participants. This visit will not coincide with your regular clinic appointment; therefore, both you and your escort will be reimbursed for your travel expense.

How will my identity be protected?

If you agree to participate, your identity and data will be protected. All data, including your consent form, interview, and focus group recordings, will be kept in a locked cabinet, and the data will be anonymised. Encrypted data will be placed in a password-protected computer and stored on Moorfields Eye Hospital premises. Your responses will be used to inform the service delivery. The study findings and publication will not contain participants' personally identifiable data.

What happens to the information supplied?

The consent form and digital recording device will be locked away in a secure drawer signed by the principal researcher. Once the anonymised interview and focus group recordings have been transferred to a computer, they will be deleted. Transcription will be stored electronically in a password-protected file. All data will be kept for five years.

Your information will highlight the condition of benign essential blepharospasm, your experience of living with the disease, and the treatment and care you have received. It will also uncover deeper meanings of your perceptions by examining the social, psychological, and contextual factors you encounter in daily life and the outpatient clinic. It will provide a deeper understanding of how health professionals can work with BEB patients to improve care.

Who will review this study?

As a doctoral student, all data collected and analysed will be reviewed by my supervisors Professor Heather Loveday, Dr. Wendy Wigley, and the clinical advisor Dr. Roxanne Crosby-Naowbi, Head of Nursing Research at Moorfields Eye Hospital. Please be aware that the findings of this study will be shared with you, and you will have the opportunity to provide feedback.

What will happen now?

If you are willing, please sign the consent form.

If you require any further information before signing the consent form, please contact Nurse Consultant Nicola Dunlop – telephone 07872414984. Email: nicola.dunlop@moorfields.nhs.uk

Author: Nicola Dunlop

Moorfields Eye Hospital NHS

Foundation Trust

City Road, London EC1V 2PD

Phone: 020 7253 3411

www.moorfields.nhs.uk

Moorfields Direct telephone helpline

Phone: 020 7566 2345

Monday to Friday, 9.00 am to 4.30 pm,

For information and advice on eye conditions and treatments from experienced ophthalmic-trained nurses.

Patient advice and liaison service (PALS)

Phone: 020 7566 2324 or 020 7566 2325

Email: pals@moorfields.nhs.uk

Moorfields’ PALS team provides confidential advice and support to help you sort out any concerns you may have about the care we provide, guiding you through the different services available at Moorfields. The PALS team can also advise you on how to make a complaint.

This study has been funded by Moorfields Eye Hospital & Burdett Trust for Nursing

Thank you for reading this!

Please keep this information sheet for future reference.

Appendix 9

UNIVERSITY OF
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Consent Form

Title of the Study: Patient experience of living with the condition of BEB

Lead Investigator: Nicola Dunlop

I confirm that I have read and understood the information sheet for the above study and had the opportunity to ask questions. Yes ☐ No ☐

I understand that my participation is voluntary and that I am free to withdraw at any time, without giving any reason, without my medical care or legal rights being affected.

Yes ☐ No ☐

I agree to participate in this research by interviewing and attending a focus group.

Yes ☐ No ☐

I wish to be contacted for the interview and focus group meeting. Yes ☐ No ☐

I give consent for the interview and focus group meeting to be recorded.

Yes ☐ No ☐

I give consent for anonymous quotations to be used. Yes ☐ No ☐

I would like the findings of this study to be shared with me Yes ☐ No ☐

Name of Participant	Date	Signature
…………………………………….	…………………	…..…………
Researcher	Date	Signature
…………………………………….	…………………	…..…………

The findings of this study will be shared with you. You will have the opportunity to provide feedback on the outcome, but if you would like any further information about taking part in this study, please contact the Principal Researcher/Adnexal Nurse Consultant – telephone 07872414984. Email: nicola.dunlop@moorfields.nhs.uk

Principal Investigator: Nicola Dunlop

Moorfields Eye Hospital NHS
Foundation Trust
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Moorfields' PALS team provides confidential advice and support to help you sort out any concerns you may have about the care we provide, guiding you through the different services available at the specialist eye hospital.

Appendix 10

Interview Schedule

Notes for Interviewer:

- Please do not use my expertise; allow the interview to be participant-led.
- Ensure the interview brings out participant experience, and beware of personal assumptions.
- Listen for phrases, metaphors, expressive speech, and vocabulary.
- Promptly clarify areas that require elaboration and listen to new information.
- Continue to seek depth until the phenomenon unfolds.

What is it like to live with BEB, and how can an acute episode affect the patient's QoL?

(20 minutes)

- Could you explain the length of time it took for your condition to be diagnosed? (opener)
- Could you describe how your daily life is affected by this condition?
- How would you describe the difficulties you face – opening your eyes to watch TV, read, drive and walk outdoors?
- Describe if and how your interactions with friends or your social life have been affected?
- Did you give up any daily activities to cope with the condition?
- Could you give an example of how the condition affects your mood?

How can the current outpatient service impact people with BEB during an acute episode of the condition? (20 minutes)

- Could you explain your experience – seeking an appointment, waiting for treatment in a clinic, receiving treatment, and booking a routine appointment at the end of the clinic?
- Could you describe what makes your experience suitable or what makes it bad?
- Have you ever contacted the hospital for treatment sooner than planned, and was gaining a new appointment difficult?
- How do you feel during an acute episode when you cannot get an appointment?

What needs to change to ensure that outpatient services for patients with BEB are responsive to expressed needs? (20 minutes)

- Do you feel that the outpatient service should be available during an acute episode of spasms?
- Have you ever reported your concern?
- How would you like the outpatient service to run as a service user?
- What do you think can be done to improve the service?

Appendix 11

FGD Interview Schedule

Notes for Interviewer:

- Do not use my expertise; allow the interview to be participant-led.
- Ensure the interview brings out participant experience, and beware of personal assumptions.
- Listen for phrases, metaphors, expressive speech, and vocabulary.
- Promptly clarify areas that require elaboration and listen to new information.
- Continue to seek depth until the phenomenon unfolds.

How would you like the outpatient clinic to run as a service user? (20 minutes)

- Could you describe what you like about the BEB Clinic? (opener)
- Could you describe what makes your experience suitable or what makes it bad?
- How would you describe the difficulties you experience when timely care is not available?
- Could you describe your experience contacting the hospital for treatment sooner than planned?
- If you could not book a sooner appointment, what explanation were you given?
- What was the outcome if you reported your difficulties booking an appointment?

What method do you think should be used to book an urgent appointment example, telephone, apps, or /other technology, and please explain why? (20 minutes)

Appendix 12

Table 4: Example: Participants' transcript

Emergent Themes	Original Script	Exploratory Comments
	Medical Secretary	
	Researcher: Good morning, JS. This interview is about your lived experience in the outpatient department. So, I want to find out what it is for you to live with BEB during an acute episode of the condition.	
	Researcher: So, could you tell me how long it took you to diagnose the condition?	
Position of a BEB patient **Doubted, challenging to persuade a GP referral** **The deterioration experienced while awaiting a diagnosis** **Delayed Diagnosis and a lack of knowledge**	**Janey:** Speaker 2 Maybe a year, 18 months, something like that, yes. And at first, for a year, they thought I had an infection, and I was treated constantly with steroids. It was only then that I paid for a private appointment, and he diagnosed and wrote to my GP, and it was at that point, my GP referred me to Moorfields because before that, he said it wasn't severe enough to be referred on anywhere. And by then, I could hardly open my eyes at all, and it was one spasm after the other all day, and I was grimacing with my face with a merge. I looked mad all day, and it was exhausting. But this was not serious enough to be referred on, so that's how I got referred to Moorfields.	Description of the length of time it took to be diagnosed. *Factual use of language to describe her relationship with the GP and what he felt about the condition* The financial burden to be referred Description of delayed diagnosis and disease progression *Derogatory comments of self and change in mood from exhaustion*
Psychological decline	**Researcher**: Could you describe how your daily life has been affected by this condition?	
Loss of independence **Physical disability** **Loss of confident**	**Janey:** Well, I think it's truthful to say that it has affected every area of life. I can no longer go anywhere on my own. I can't drive anymore. My eyes just snap shut in the middle of a road if I try to cross the road. I bump into things. I trip over things. I am not the person I was at all, and I need assistance and confidence to do the things I used to do. I can't catch a bus and pop into Sainsbury if I want to or something. So yes, it's entirely life-changing, and it's not a	Description of the impact of the condition on daily life *Blind and physically disabled* *Use of language to describe the loss of*

Blindness **Ocular discomfort** **Botox provides Normality** **Physical limitations of BEB spasm and discomfort**	quiet-like blindness illness that is terrible. It feels as if somebody has put salt in your eyes, and it’s not one eye. Both eyes are always affected if you get something in your eye. So that moment when you have something terrible in your eye, and you're squinting and can't see, and you're rushing to the tap or whatever you can use to wash it out, those moments you can't achieve anything. You can't cook, you can't carry on with what you were doing, you've got to get this thing out of your eye, and that's how it is all the time. It doesn't stop. So, the Botox injections relieve that, you are no longer pulling faces like you are a mad person, it's not as exhausting because of the tight grips, and you get cramp after many of them, and they keep going. It's like running upstairs, and your legs cramp; your face and eyes are tired out with the cramps and grimaces and squints. So, Botox is essential to get towards any regular daily achievement. **Researcher:** Okay. Could you tell me if it affects watching television and reading? I know you've already mentioned driving and walking out. **Janey**: No, I haven't been able to read because of the extreme dry eye and the blepharospasm. It's not just a question of the eye smacking shut now and then, and you have to wait for them to open before you can continue reading. It's because the dense layer, I believe, helps you to focus, and without your viscous layer, no matter how many of these drops you put in, which affects your sinuses because you're pouring this stuff in all day long, no matter how many drops you put in, it's not like your own viscous layer that helps you to focus. Hence, I get prickling like needles going in my eyes which then makes you squint and your eyes stream, and so reading anything but a few lines when you can get your eyes comfortable enough to read is all that's possible. I haven't been able to read a book in 3 years possibly, slightly more, so I can read bits and pieces	*recognition of self* Description of the need for assistance and lack of confidence from blindness and discomfort Factual use of language to describe the physical disability caused and the persistent nature of the condition Description of BEB and the normality BTX provides Description of the ocular discomfort experience from BEB Physically unable to enjoy leisure activities

Coping Strategies are helpful **Loss of friendship** **Embarrassment** **BEB life is a challenge** **Guilt of dependency** **Relinquishing of**	when they are calmer, and I wait for those days to do my filing because I can't even look on the pages what I'm supposed to be filing this under. So yes, it affects reading on the computer and books. **Researcher**: And television? **Janey:** And television, yes. Yes, I often watch the television with my eyes closed and listen to the television and just open them now and again to see who it is, etc., and then they close again because it just, my eyes stream or they prickle—so same situation. **Researcher:** Are the sunglasses helpful? **Janey**: Yes, I do find that. I believe it was Moorfields that suggested these rose coloured glasses. They've been accommodating for the television because the dark glasses mute the colours and the shades of things so much more, so if the screen is a little dark, I can't see what's going on in the dark shades. So, the rose ones have helped with television and the dark ones I have to wear outside because I am very photophobic. **Researcher**: Describe how your interaction with friends or your social life has been affected? **Janey**: Oh, completely. I mean friends, I thought was friends have said, *"Oh, I didn't realise you were going to have to keep changing your glasses to wrap around every time you go outside. Going shopping with you is a nightmare",* and they don't want to be bothered with you anymore. Going out for lunch, crossing the road, *"Would you mind if I hold your arm?".* I feel embarrassed, do that, get in the cafe, and then I fall over somebody's bag on the floor or crashing about. It's not who I am as a person. It's my eyes making me like this. So suddenly I'm a klutz and not capable, and I sit down, and I can't read the menu, so I have to ask my friend to read the menu	Description of the supportive strategies undertaken to cope Description of how BEB impacts friendship Description of disability and embarrassment in public places Derogatory use of language to describe self and anxiety suffered in a social setting Factual descriptions of every aspect of life as a challenge

loved activity	and then because I've become anxious, I can't take in what she's saying. Shall I just remember? I think you must remember something, so I order whatever I've remembered and not what I really want. So, it's challenging, everything you do. Cooking, if you have a squint when you take something from the oven, which mine can last 30 seconds, and you're holding a hot casserole, you're trying to remember where the island is to put this down because it's burning your hands now and if you miss it, it will be all over your feet. It's dangerous, and it ruins my carpets [laughs]. Every area is affected.	Description of Physical limitations, dependency, guilt which cause participants to relinquish loved activities
Anxiety	**Researcher**: Okay, and did you have to give up any daily activity to cope with the condition? **Janey:** Well, I think everything, the gym threw me out [laughs] because they were frightened that I might fall over and they'd have an insurance claim. So, yes, I mean not being able to drive, I can't pop out and see my friends, and I can't go to the gym, and I can't do the things that I did before. (Bryan) my husband takes me everywhere. I used to love singing but to go to the choir, even if my husband (Bryan) drops me off at the door, I have to then go to strange places to perform, and I don't know where I am, I don't know where the steps are, I can't read the book with the words on. So, if this is not a song or a hymn that I know, I can't read it, so I am a pain, so I don't go anymore. I can't think of much I can do as I did before. **Researcher:** JS, could I ask you to give me an example of how the condition affects your mood. **Janey:** Yes, the fear and flight thing is affected because when you have something in your eye, you have the predator thing still in you, the lizard part of the brain that thinks, oh, eyes can't see at the moment, they are streaming, I have got something in my eye, so I need to raise you	*Linguistic use of language to describe the fear and anxiety caused by BEB*
BTX gives		

normality **BTX Improve mood** **BTX improve psychological function** **BTX effects wear off**	and rush you to the tap so that you can see again so that you won't be eaten by something nasty. So, the fear and flight situation means that your heart rate goes up, your breathing goes up, and you don't have any control over that at all. You can't switch it off as a person. So, while you have this condition, it changes you as a person. You become a little more anxious, less confident, more aware of noise. Yes, you are altered as a person because you can't switch off the fear and flight ever, so stress levels are slightly higher just because of that. It is not that you are running around the house screaming in terror. It is just a slight alteration of never being able to switch off, always being slightly anxious, and you notice, well, I notice it when I lay down. It is time to go to sleep, and I notice that my heart rate is more than it used to be, and it takes me longer to switch off to go to sleep. So, I have to tell myself it's okay, calm down, and that just used to happen. You lay down. You want to go to sleep. Very noticeable in quiet moments, yes. So, it affects the mood. **Researcher**: I'm looking at how the outpatient service impacts people with benign essential during an acute episode of the condition. So, could you explain your experience of seeking an appointment, waiting for treatment in the clinic, receiving that treatment, and booking another appointment at the end of the clinic? Speaker 2 Well, finding Moorfields in the first place changed my life because living as I was with these permanent squints was impossible. I understand people used to be put in mental institutions because you look mad grimacing like this. So, coming here and being diagnosed and having the injections means that I can look more regular, and without these tight grimaces, it's life-changing, it is, startlingly so. And so, to come to the clinic and get regular injections to relieve that is paramount. There's no way I could cope	*Description of BEB Clinic as a haven restoring normality* *Description of linguistic use of words to describe the mental affliction* Description of recognising the mental challenge BEB presents and BTX is paramount to coping Description of achieving expert patient knowledge Describing the responsive service required to address the needs of BEB patients Description of achieving expert patient knowledge

FGD Interview **Acute deterioration & untimely care cause suffering** **Clinicians do a**	without them and if I have to wait an extra few days or a couple of weeks because of lack of clinic time or holidays or whatever interrupts my flow as a patient, those two weeks are tough to cope with, not only for me but for my family as I do fall over more, drop things more, squint more. It's much more challenging. **Researcher:** When would you say your treatment is at its peak? **Janey:** Treatment at its peak is when I can get my injections regularly, on time, and also if I have a critical moment when it's just for some reason become unbearable, and that happens sometimes, the ability to be able to ring up and come in and have an additional out of sync injection would be optimum, that would be where it should be. It's unacceptable management. It falls to funding this condition is very challenging to live with, management just don't understand. **Researcher:** Okay. So, your injection usually works best after 2, 3 months, or a few days after the treatment? **Janey:** A few days, two days perhaps for it to settle down, and then it's good. It takes all that away, and I can cope for another 8-10 weeks. **Researcher**: So, when it begins to decline, would that be around what time, eight weeks, six weeks? **Janey**: Probably 8. Yes, you begin to notice the odd thing around eight weeks, and it gradually worsens until the injections wear. It's not terrible then often, but you'd start noticing that you are squinting more, that the sun is affecting you more, the different shades of light. Somebody putting a light on in the room now affects you more. **Researcher**: So, when would you prefer	BTX injections wear off Description of the responsiveness of the service and requirement to address the unbearable spasms experienced during an acute attack of the condition and the needs of BEB patients to be seen in a timely manner Description of the challenge of gaining contact with the hospital during an acute attack of BEB Description of insufficient appointment slots due to overbooked clinics Description of how difficulties experienced when BTX wears off

good job **Lack of BEB Clinics**	your treatment if you had a choice? **Janey:** I think ten weeks is okay generally, with the option that if you have an acute episode and it's unbearable, you could slot in somewhere and be helped. **Researcher:** Have you ever contacted a hospital for treatment earlier than planned? **Janey**: I think I tried to, but it was challenging to get through and make any extra appointments because there weren't any. **Researcher**: Okay. So, did you manage to get hold of them at all? **Janey:** I think I got through to reception, but they're just weren't any appointments available.	Description of the need for more clinics locally and geographically closer to home to address the needs of patients desperate for treatment
Deterioration cause physical and psychological decline	**Researcher:** Was it difficult? **Janey:** Yes, very much so. It was pretty bad, as if I hadn't had any injections, and I didn't understand why they had worn off. Still, I was reassured the next time I went that this sometimes happens and not to worry, but certainly, if that's possible to find more appointments that we could come in, patients could come in on those occasions. They could carry on with their lives, particularly those who have managed to continue working. It must be far worse for them. **Researcher**: Okay. What needs to change to ensure the out-patient service for patients with your condition, BEB, is more responsive towards patient needs? What do you think needs to change? **Janey’**s certainly more clinics to enable more patients to be seen perhaps, and those already being seen have a slot when they are desperate when they need to in an acute period. So, more clinics and possibly even satellite clinics to save	*Description of the linguistic use of the word suffering to describe the effects of untimely treatment* Description of actions taken to address unacceptable service Description of how a patient QoL can be improved with more

	people from travelling so far. **Researcher**: So, do you feel the outpatient service should be available during an acute episode? **Janey**: Definitely. That would be so helpful. The thing is, this doesn't happen all that often, and for many people, it would be once a year maximum, but that once a year to be suffering for 5-6 weeks until your injection comes up is entirely unacceptable. **Researcher**: Have you ever reported your concern about not getting an appointment on time or not accessing the clinic? **Janey**: Awful. We have been to PALS and asked not to complain about the clinic because they are fabulous, they do an excellent job, and the injections are so helpful. It's not to complain. It's to ask what the problem is? Why aren't there enough appointments? Can we do anything as patients to help this? Nothing was resolved, and eventually, we just fell back into whatever could be offered. **Researcher**: As a service user, I think you've touched on this already, but how would you like the outpatient service to run? **Janey:** Well, as it is now, it's perfect except for the fact that there aren't enough slots, there aren't enough clinics, and if that was provided, then more people could be seen, and patients in acute situations can continue to live their lives. **Researcher:** What do you think can improve the service and get more clinics? **Janey**: Well, I presume it falls to funding, and whoever holds the purse strings needs to understand that this is not a minor problem that you can put a drop in your eye, and all is well. It is a life-altering, life taking away problem that is very	clinics Description of the linguistic use of words to describe management lack of understanding and empathy in regards to funding the service and about the condition Description of BEB persistent nature and ability to cause stress Description of the appointment booking system and it is flexible for the less technological patient

	challenging to live with. Adding salt and putting it in the eyes even once and trying to carry on working while the salt is in their eyes and see how that feels and then imagine that you have to live like that. It won't go off as it will for the person that's done this just once. It will be with you forever and every day, whether you like it or not. At your most stressful moments, and you need it desperately to go away, it won't. So, try the salt thing, and it will be understood. This is how you have to live, and I can now understand how challenging that is. Another area is continuity of care. The doctor changes all the time. I prefer the nurse-led clinic **Researcher**: Do you feel booking your appointment by App or online would be a good idea, and why **Janey**: No, I am not computer savvy or own a smartphone. Maybe younger people might like it. A telephone call, yes. **Researcher:** Okay. Thank you.	

Appendix 13: Sample of Submitted Doctorate Work

Contents:

Evaluating Professional Practice: Improving the care and experience of Hemifacial and Blepharospasm Patients

Background

Hemifacial spasm and blepharospasm are conditions that cause sufferers to experience sustained, forced and involuntary closure of the eyelid due to muscular contractions. Both conditions affect the ageing population, and are chronic and debilitating. It significantly impacts the patient's quality of life; sometimes patients are functionally blind. Hemifacial spasms affect one side of the face, whereas blepharospasm affects both eyes and the face. The standard treatment for spasms is botulinum toxin, but its effectiveness varies (Jinnah et al, 2013). A systematic review of patient-reported benefits of botulinum toxin in blepharospasm ranges from no improvement to 96% of patients reporting significant symptomatic relief (Colosimo, Tiple and Berardelli, 2012). Botulinum toxin injections therefore, offer temporary relief but the duration of treatment efficacy varies and no research so far has been able to explain this inconsistency. The standard model of patient care involves routine, scheduled treatment cycles, which is ineffective for many patients.

Rolfe Reflective Model: Challenging issues and Translating Research into Practice

To investigate the effectiveness and cost-effectiveness of a patient-initiated botulinum toxin service a randomised control trial (RCT) was set up. The author employed a reflective model (Rolfe et al, 2001) which asks three simple questions: What? So what? and Now what? To understand the challenging issues, and how research can be translated into practice. The overall aim of the author research to improve the quality and experience of effective care for hemifacial spasm and blepharospasm patients, by improving access in the outpatient clinic.

What?

This startling lack of an effective treatment is the reason for establishing the author’s quality improvement (QI) initiative. Reflection is at the core of both learning and continually evolving practice (Robertson, 2005); to understand and learn from a particular issue. Such reflection suggests the real problem is that standard treatment leads to some patients having unnecessarily frequent treatments, while others experience distressing symptoms whilst awaiting treatment. The condition causes appearance issues, social embarrassment, isolation and depression (Muller et al, 2002; Kraft and Lang, 1988; Reimer et al, 2005). The stressful interval between treatment cycles is also a frequently reported concern. The author has often sympathised with patients but research funding to explore an evidence-based approach for a new innovative treatment model was not available.

So What?

When invited to take part in a RCT that compared patient initiated treatment to standard care, I eagerly accepted as learning whether this research could be translated into practice is of utmost importance. Reflecting in action, I thought this was an opportunity to really make a difference; I am ideally place to influence the trial. My role involved selecting patients, consenting, evaluating the patients' responses to treatment before administering the injection of botulinum toxin. However, I am aware that my close therapeutic relationships with patients are of value to the RCT and my involvement could influence patients' acceptance of the trial. I empathise with patients suffering from this dreadful condition and their call for help resonates strongly with my plans for quality improvement.

Now What?

First, the broader issues were explored; could an innovative approach to service management be beneficial? A systematic review by Whear et al (2013) over three conditions: irritable bowels, breast cancer and rheumatoid arthritis, have shown few differences between the psychological or health-related quality of life between those who initiated their own follow-up appointments compared to those in standard care. However, whilst reducing morbidity and disability, it may reduce unnecessary hospital visits for some patients. Therefore, evidence suggests this is an effective treatment model.

Saving costs is important for the author's organisation, which has Foundation Trust status. According to Monitor (2015), Foundation Trusts have overspent their budget by 52% over the 2014-2015 period. When considering cost in relation to chronic conditions, one needs to understand the NHS's supply and demand issue. Increased expectations along with increase expenditure, creates a rise in general taxation. Although cost is important it should not determine if best practice is introduced. Nevertheless, the introduction of more doctors or nurses is costlier than changing the treatment model to deliver effective care.

The question still remains: how will this knowledge improve patient care beyond the research stage? The author reflected on this but found no immediate answers, so they were sought by reviewing various literature on the topic; why was a RCT instigated to address the concern of delivering effective patient care? Surely the need for evidence-based intervention has led to the call for evidence of the highest quality? One of the most cited articles is the "Hierarchy of Evidence" which placed value on rigorous study designs (Briss, Zaza and Pappaioanou, 2000; Bero and Rennie, 1995) however, the conclusion drawn is that even RCT are unsubstantial frameworks on which to weigh all the information to translate research into practice (Green, 2006; Rychetnik, and Hawe et al, 2002; Pettigrew, 2003). The practice environment also seems to be another area that presents difficulties, with Damschroder et al. (2009) blaming internal and external environments for the failure of the intervention to translate into meaningful patient care outcomes. The external environment was examined under the section; *Now What?* But a suitable framework will be revisited. The internal environment, specifically the outpatient appointment booking system, will be studied to give a broad perspective on the difficulties patients face. A process map allows greater communication of current practice from an unbiased point.

Process Map: Outpatient Booking System

Why research fails to be converted into practice might be due to the fact that some intervention methods do not take into account the diverse circumstances of public health (Green and Mercer, 2001). Understanding the patient journey is crucial to the planning of an improvement initiative in the practice setting. The author requested the help of a colleague whilst shadowing the patient journey and process. The patient was also interviewed about rescheduling an appointment over the phone when symptomatic; the interview and process map was completed within 30 minutes. The map provides an overview of the entire journey a patient takes to book an appointment; it highlights the difficulties patients faced during each step of booking an appointment, in the outpatient department or over the phone (Appendix 2). Such a process map is a powerful model for communicating concerns, involving others and emphasising improvement needs.

Process map of the current booking system highlights in-depth examination of the organisation’s internal process to gain further understanding of bottlenecks, repetition of actions and areas that can be enhanced within the outpatient clinical booking system. Looking at the internal processes through the patients’ eyes revealed time wasted for both staff and patients each time an appointment required booking. A particular disadvantage is that it focuses on an event when it happens but may miss out on details; it does not explain why the patient appointment system is as it is or why things happen the way they do. Kaplan, Robert and Porter (2011) reiterated the identified weaknesses and strengths are limited to users’ understanding; the right process, the right time and resources are not critically examined.

However, the author has considered the process critically and looked at each time an outpatient appointment could be made. Specifically, the appointment booking system and its impact on the quality of care provided to patients, and the likely adverse effect on staff morale. The patient contacts the clerk immediately after treatment for a follow-up appointment and again over the phone when symptomatic. During this worsened period, they are faced with prolonged waiting for a call back; this has been equated to a prison sentence as patients are not only effectively blind, but also house-bound.

Similarly, clerical staff have communicated frustration at being unable to make a booking due to extenuating issues such as insufficient injection staff, an already overbooked session, and having to gain permission from the injector each time a session is unavailable. Further expertise or specialist support is possible but costly. Reflecting on this issue, the botulinum toxin service is mainly nurse-led, and the current compliment of a doctor is due to the RCT. To recruit and train more staff at Band 6 or above requires a cash injection, only to immediately release the staff for 6-9 months to undertake non-medical prescribing. It requires further time to undertake in-house consenting and Botox training in addition to on-the-job training, and even then the clinical decision-making skills needed for independent practice is only acquired with years of experience. Utilising a different treatment model to organise care is more practical.

Critical reflection was used to consider all the relevant factors. It provided analysis of the problem but also a greater understanding of what needs to be done; looking at ones’ practice helps deliver the highest possible standards. However, an argument against reflection is that it can be very individualised or biased; Smith (2008) stated in opposition to reflection that it is: “A critical and rigorous process which pays attention to personal, interpersonal and contextual factors influencing what is said and done or not said and done through research."

Reflection revealed the booking process took two steps each time the clerk was unable to make the requested appointment; telephone booking created a bottle neck. In some instances, patients waited more than two days if the injector was away, before an appointment was made over the phone. Confusion arose over whether the clerk or patient should contact the injector, and there was a general lack of information regarding how to contact the injector for general queries around treatment efficacy and booking another appointment. Examination of this process provides evidence to communicate to stakeholders in support of redesigning and removing elements of repetition that creates delays.

Next, the process mapping information was documented by using an oval for the beginning and end of the process, a rectangle for activities or tasks, a diamond for where a question or decision was made, and arrows to indicate movement (Appendix 3). It was also recognised that process mapping and stakeholder analysis could be combined as the author gathers information for both methodologies simultaneously. To foster more ownership and support for the QI initiative a stakeholder analysis was undertaken.

Stakeholders Views and Analysis

The first step in stakeholder analysis is, identifying the stakeholders and determining their power, influence and interest (Appendix 4); once stakeholders are identified they can be prioritised to be focussed on. This step involves an understanding of the nature and interplay between the individuals and organisations alongside both intentions and behaviours (Brugha and Varvasovszky, 2014). Collaboration creates more buy-in and encourages others to be influenced by key players, however; all players need to be considered as each offers essential analysis for the successful achievement of the research project. Acknowledging, listening, valuing stakeholder views and contribution is a pragmatic approach in influencing the views of others in the achievement of the desired outcome (Appendix 4).

The clerical supervisor when interviewed indicated a neutral position; although it was felt a new system would provide more job satisfaction, general consensus was addressing the lack of injection staff would be a better approach. Another concern raised was the lack of office-bound clerks to answer the increase in telephone calls this new system would generate. And lastly was the need for their views to be taken into consideration, and for them to be informed. Upon reflection of the recent clerical restructuring, I understood the lack of belief that their views are considered. Although doctors, nurses and consultants were all in favour of the new approach for patients initiating their own follow-up treatment, the general manager was not, in fact was fearful of patients being lost to follow-up, complaints and loss of funding. NICE guidelines CG85 (2009) recommended healthcare organisations minimise the risk of avoidable sight loss for patients with established or suspected glaucoma. Similar principles have been cascaded to all patients at risk of temporary or permanent sight loss. Specifically, the guidelines advised: reviewing the levels of hospital-initiated appointment cancellation, identifying and auditing high-risk nonattendance and patients currently awaiting follow-up appointments, ensure compliance with NICE follow-up intervals, and providing patients with information to ensure they can be easily rescheduled when necessary. The manager’s contentious position is that the hospital has a responsibility towards its patients and putting the onus on them to ensure their condition is treated, is risky.

Whilst patients indicated their support for the proposed treatment model, many carers and family members opposed it, as it meant taking time out of their busy schedules to contact the hospital. In some cases, it was felt if the system is not broken, why fix it? On reflection, this is particularly true for patients whose treatment efficacy is stable and routine appointment every three months is perfect. Nonetheless, this innovative treatment approach whilst inclusive, should recognise that patients who are visually impaired, deaf, or have learning disabilities and or multiple hospital appointments are unsuitable. This highlights the potential health inequality for patients who are less affluent, educated or from a lower socio-economic group. Whilst patient led/self-care is empowering and enables patients to cope with their condition, the author believes it should not discriminate but instead provide choice.

The author feels the stakeholder analysis was important to allay the fears of the general manager, patients, families and carers and to keep the clerical staff, nurses and doctors on-side. It was also important to use this new-found knowledge to improve the initiative. All parties were informed that patients will be given a choice to opt-out of this research and treatment model. Self-care does not mean there will be no responsibility towards patient care; but that it develops a more partnered approach. A safety net system will need to be incorporated to ensure no patient is lost to follow-up. The availability of office-bound staff is another area that will require organisation support to ensure a dedicated booking clerk is present at all times. Communication at every stage to gather further information and share the project’s progress has been arranged. An action-effect diagram was formulated to communicate the authors’ research and to highlight the measure’s concept, a source of evidence that is not often linked to QI initiatives. These measures can be tested to determine if the change is indeed a QI. Research timelines were communicated to all stakeholders and the implementation concerns raised will be discussed at future meetings. Stakeholders were asked to consider solutions to issues raised as these will be taken on board.

Action-Effect Method and Diagram to Implement Change

The action-effect method (AEM) is a systematic, structured process to identify and articulate quality improvement initiatives (Reed et al, 2014). It demonstrates best practice but more importantly, communicates how the implementation of a patient-initiated botulinum treatment model could be managed. The AEM involves all the opinions of stakeholders in the achievement of a clear aim; it is a framework methodology which forms the relationship between the intervention, activities, and aims. It hypothesises and provides evidence for a cause-and-effect relationship, and the measure's concepts are a form of evidence used to support the evaluation.

The action-effect diagram (AED) was formulated to communicate the intended research outcome of the service improvement: to improve the quality of care for hemifacial and blepharospasm patients by increasing access to effective botulinum toxin in an outpatient clinic, into achievable objectives. The overall aim of the AED is to reduce variation during the implementation process. The cause-and-effect chains are made up of factors linked by arrows indicating how each action proposed by the author could result in an improvement (Appendix 5).

The objectives are the major contributing factor to achieving the aim, reviewing the current booking system, enhancing the role of the clerk in rapid access booking, augmenting the role of the patient in initiating access, and reducing inappropriate care. Reading the diagram from left to right answers the question: “what changes can we make that will result in an improvement?”, and from right to left: “what are we trying to accomplish?” (Appendix 5). Finding the most appropriate framework will address these questions. The author's uncertainty of the RCT findings automatically being translated into practice have seen the formation of a QI introduced; with the essential elements to translate research into practice.

Now What? – Using the PDSA Reflective Framework

The PDSA (Plan, Do, Study, Act) improvement model (Appendix 5) was chosen as it is most suitable and allows the continuous application of scientific methods to formulate a hypothesis, test that hypothesis and so implement change. The stakeholder analysis and process map already undertaken separately are essential to this model, on which Singh et al (2013) suggested without communication, organisation and participation of all key stakeholders there will be limited success using the PDSA model. To begin the use of this cycle in a QI, the first step is formulating the research question (Appendix 6). However, the PDSA framework is constructed with three key questions and a process for testing ideas:

- What are we trying to accomplish?
- What changes can we make that will result in an improvement?
- How do you know the change is an improvement?

Plan

Research Question

Implementation of hemifacial spasm and blepharospasm care: What impact would a patient-initiated botulinum toxin treatment model have on the provision of effective care, organisation of service, patient experience and team expertise?

Aim Statement

To improve the quality of effective care and experience for hemifacial spasm and blepharospasm patients by improving access in the outpatient clinic.

Objectives

Specific questions to be addressed in this cycle:

- How can the current outpatient booking process be improved?
- What are main determinants of implementation problems?
- What is the impact of the new treatment model on: effective appropriate care, organisation of service, patient experience and staff expertise?
- How can access and quality be improved?

Hypothesis and Achieving Outcome Measures

The quality of care for hemifacial spasm and blepharospasm and general experience will improve if patients have information on how to gain rapid access to the service, on decrease waiting time, if appropriate treatment is available, if staff training and booking protocols are in place.

What Changes can you Make that will Result in an Improvement?

Review the booking system: by demonstrating the faults of the current system and collaborating with doctors, nurses, clerks and managers to review the process map and develop a rapid access booking system and protocols.

Augmenting the role of the patient: by providing a patient information leaflet on the condition, along with injectors' and clerical booking's contact numbers. Creating and providing treatment summary and management will ensure effective follow-up care is provided.

Enhancing the role of the clerk: by providing education on the condition and how to use the rapid access booking system. Providing a more user-friendly booking system, responsive to patient needs, with a safety net to prevent loss to follow-up.

How do you know the Change is an Improvement?

This will be answered by testing the measure's concept (Appendix 3).

Plan for Change, Testing and Intervention

In planning for the change the author has attended numerous meetings with the RCT team and will continue to do so. Help have been sought from a staff member to inform patients about the research and seek their views regarding it. A patient information leaflet was developed and is currently being given to trial patients but can be developed further and replicated for use beyond the trial period.

Testing the Measure's Concepts

Waiting times, cancellations of appointments, patient information on how to access the service, clinical management plan and treatment summary, number of visits per year, patient experience and service recommendations, the establishment of rapid access booking protocol and staff training.

Data Collection

The process map, stakeholder analysis and AED form part of the initial data collection but it is also viewed as an initial action already taken. Current RCT data is also available on patient selection, consent, experience, current time to access the service as well as reasons for not taking part in the trial.

Do

Further Work and Timeline

The recruitment period ends November 2016, and the trial ends August 2017, with the following data being collected:

- Waiting times.
- Average visits per year.
- Cancellations.

The author needs to formulate:

An audit on the patient experience and service recommendation (from January 2017 inclusive, with a total of 50 trial patients).

Rapid access protocol and process map including a safety net to prevent patient loss to follow-up appointments (by August 2016).

Staff briefing and training on the new process (continuously throughout the QI).

Clinical management plan and treatment summary for all patients (by April 2016).

Study

All information collected in the initial stage will be quantified and analysed to determine if improvements have been realised, and any surprises will be noted. Improvement and/or findings will be fed back to all stakeholders.

Act

A decision will be made on whether change can be implemented, then the next steps or cycle will be planned. The research will also be documented and summarised. The author's next steps have been stated in the above timeline but this portfolio is an evolving piece of work, and new ideas that can contribute to achieving the QI can be introduced at any time.

Conclusion

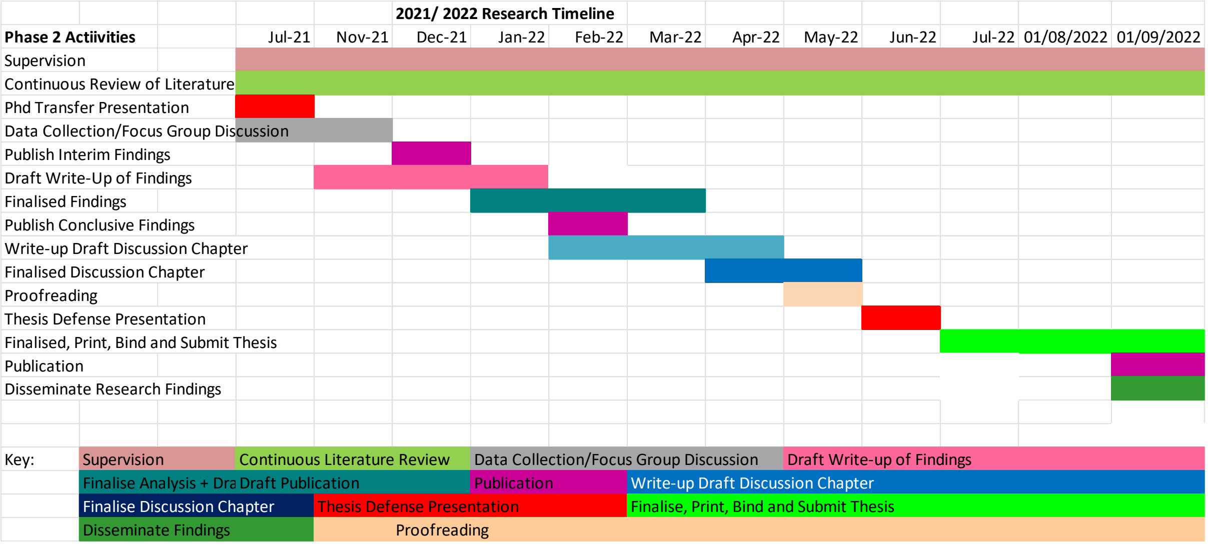
This paper demonstrates critical reflection and evaluation of an area of the authors' practice. The possibility of translating a RCT into the practice was probed but found lacking. By asking questions, the author understood the two main factors required to convert research into practice: the practice environment and a well-structured framework. External evidence suggests patient-initiated services were successful in delivering effective care in other areas, whilst reducing morbidity and cost. Internal scrutiny was undertaken by process mapping the booking system to gain understanding of bottle necks that require improvement. This analysis was essential in determining implementation problems. Similarly, the value of listening and considering the opinions of stakeholders allowed the author to gain new knowledge, concerns were addressed, and patients and family informed choice was exercised by opting out of the patient-initiated treatment. A safety net system will be formulated in the rapid access booking system to ensure patients are not lost to follow-up.

Pragmatically speaking, stage communication to keep an open dialogue with the clerical team and management was instigated to ensure ongoing collaboration. An AED was formulated to communicate the progress of research in its entirety, and to reflect the views of the stakeholders. This approach is aimed at securing further buy-in from all stakeholders, ensuring everyone understands the relationship formed between the intervention activities and aims in the AED that hypothesises the cause-and-effect relationship, and that the outcome measures form evidence to support the evaluation.

The reflective model framework by Rolfe et al. (2011) was initially employed but the PDSA model was eventually selected as it is more appropriate for testing the hypothesis. Planning of the QI was carried out by first obtaining the views of others and communicating the initiative. However, the PDSA model was able to clarify the aim, objectives and hypothesis, identified what changes can create a QI, and tested the measure concepts to determine if the change resulted in a QI. Finally, the author acknowledges there is further work to be done and future timelines have been established. However, this paper forms part of a reflective portfolio and the very use of the PDSA cycle suggests its ongoing nature, subject to change and improvement reflective of evolving ideas geared towards the achievement of the initiative (please main reference list for all reference).

Appendices

Appendix 1

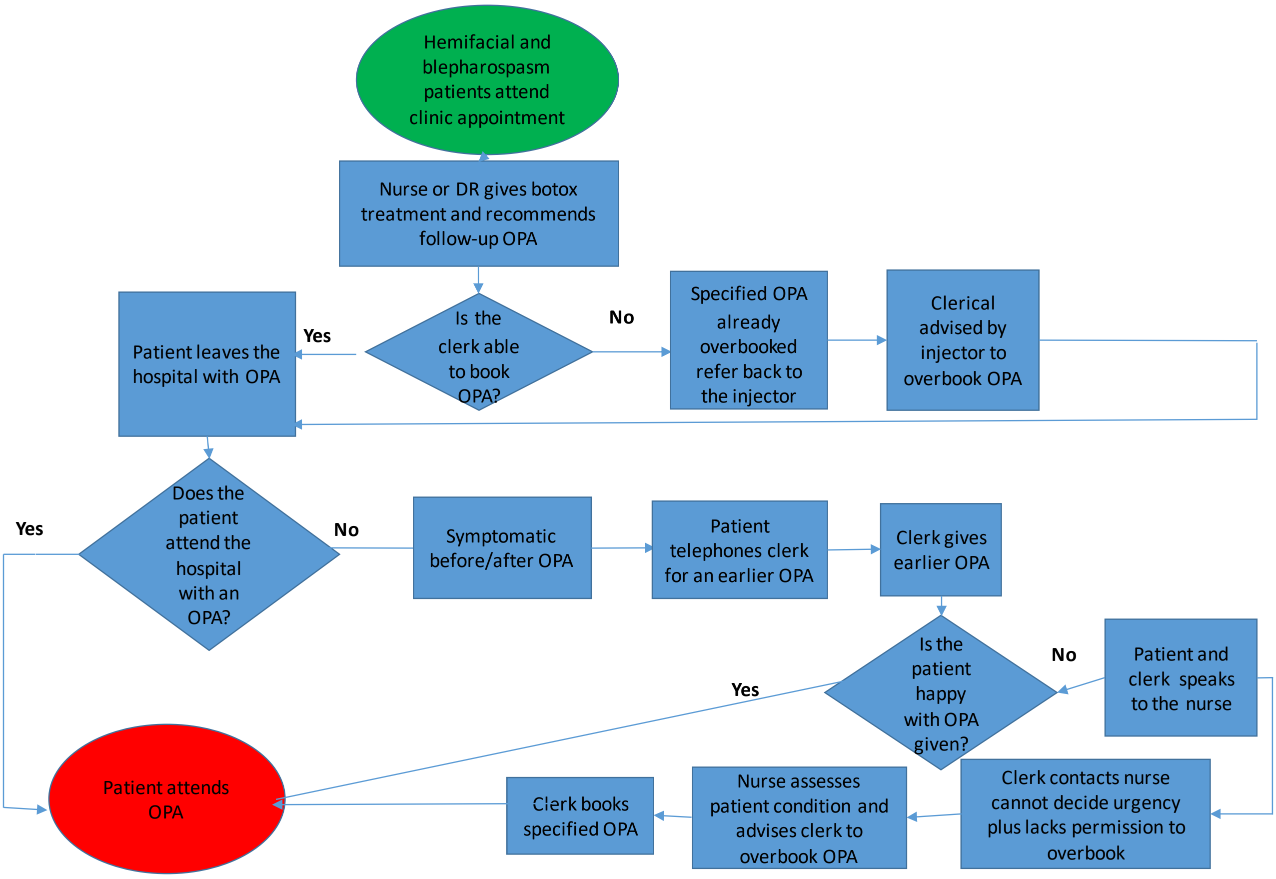


2021/ 2022 Research Timeline

Phase 2 Actiivities	Jul-21	Nov-21	Dec-21	Jan-22	Feb-22	Mar-22	Apr-22	May-22	Jun-22	Jul-22	01/08/2022	01/09/2022
Supervision	■	■	■	■	■	■	■	■	■	■	■	■
Continuous Review of Literature	■	■	■	■	■	■	■	■	■	■	■	■
Phd Transfer Presentation	■											
Data Collection/Focus Group Discussion	■	■										
Publish Interim Findings			■									
Draft Write-Up of Findings		■	■	■								
Finalised Findings				■	■	■						
Publish Conclusive Findings					■							
Write-up Draft Discussion Chapter					■	■	■					
Finalised Discussion Chapter							■	■				
Proofreading								■				
Thesis Defense Presentation									■			
Finalised, Print, Bind and Submit Thesis										■	■	■
Publication												■
Disseminate Research Findings												■

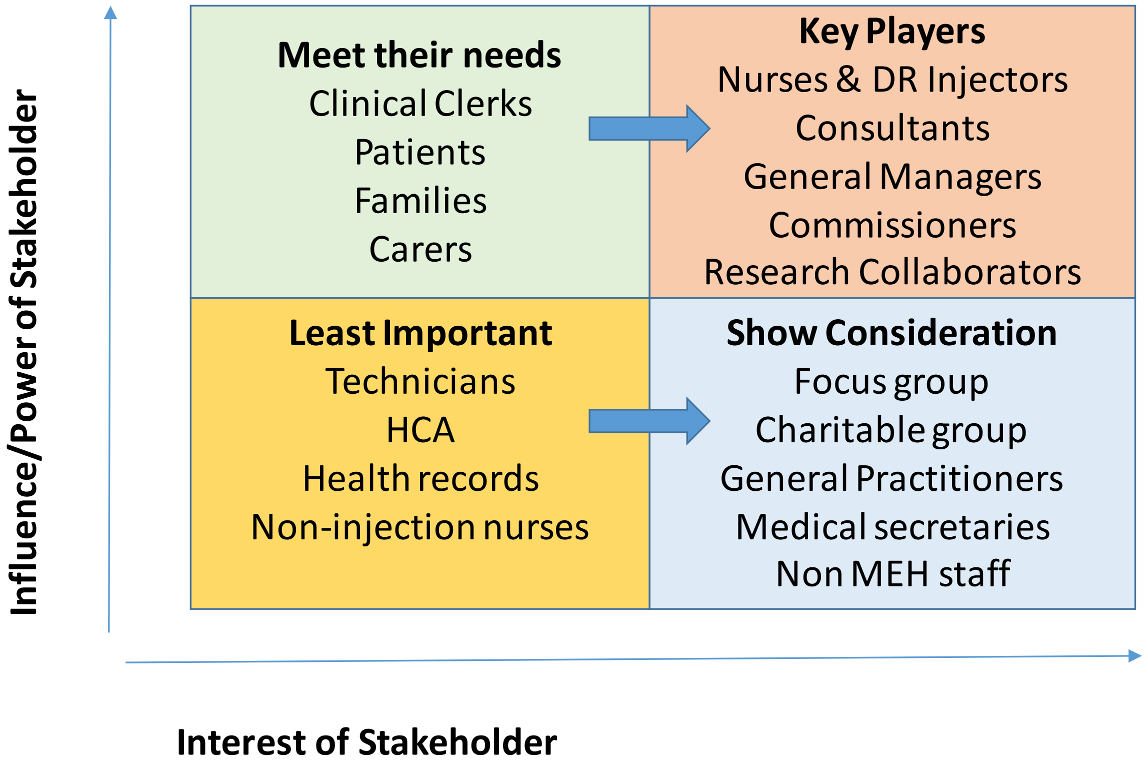
Key:
Supervision
Continuous Literature Review
Data Collection/Focus Group Discussion
Draft Write-up of Findings
Finalise Analysis + Dra
Draft Publication
Publication
Write-up Draft Discussion Chapter
Finalise Discussion Chapter
Thesis Defense Presentation
Finalise, Print, Bind and Submit Thesis
Disseminate Findings
Proofreading

Appendix 2



Process map: Hemifacial and blepharospasm outpatient appointment (OPA) booking system

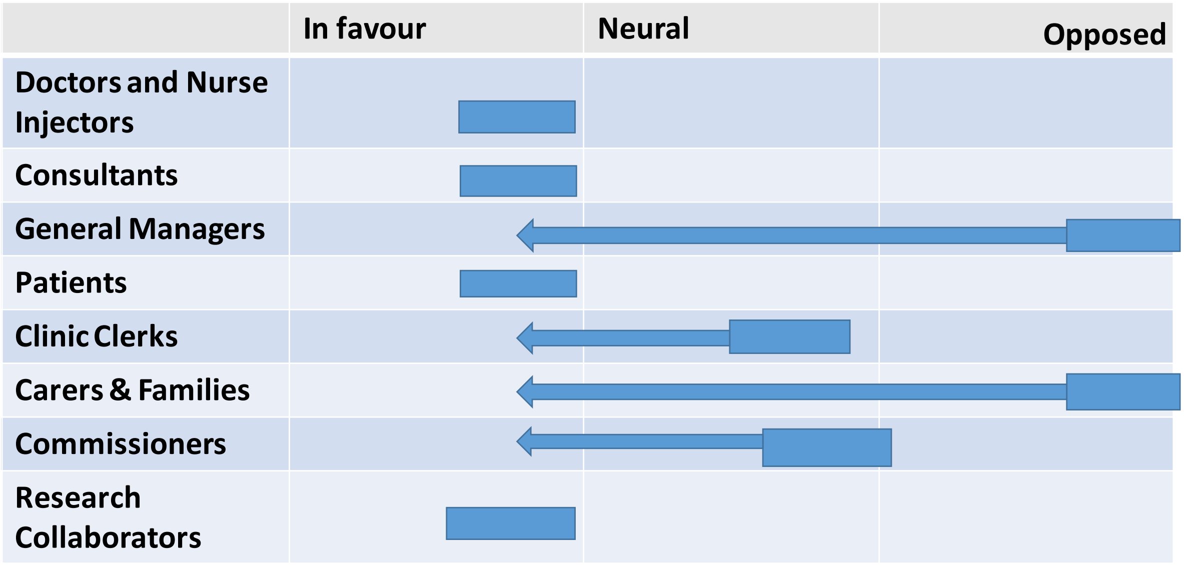
Appendix 3



Stakeholder analysis: patient-initiated botulinum toxin service

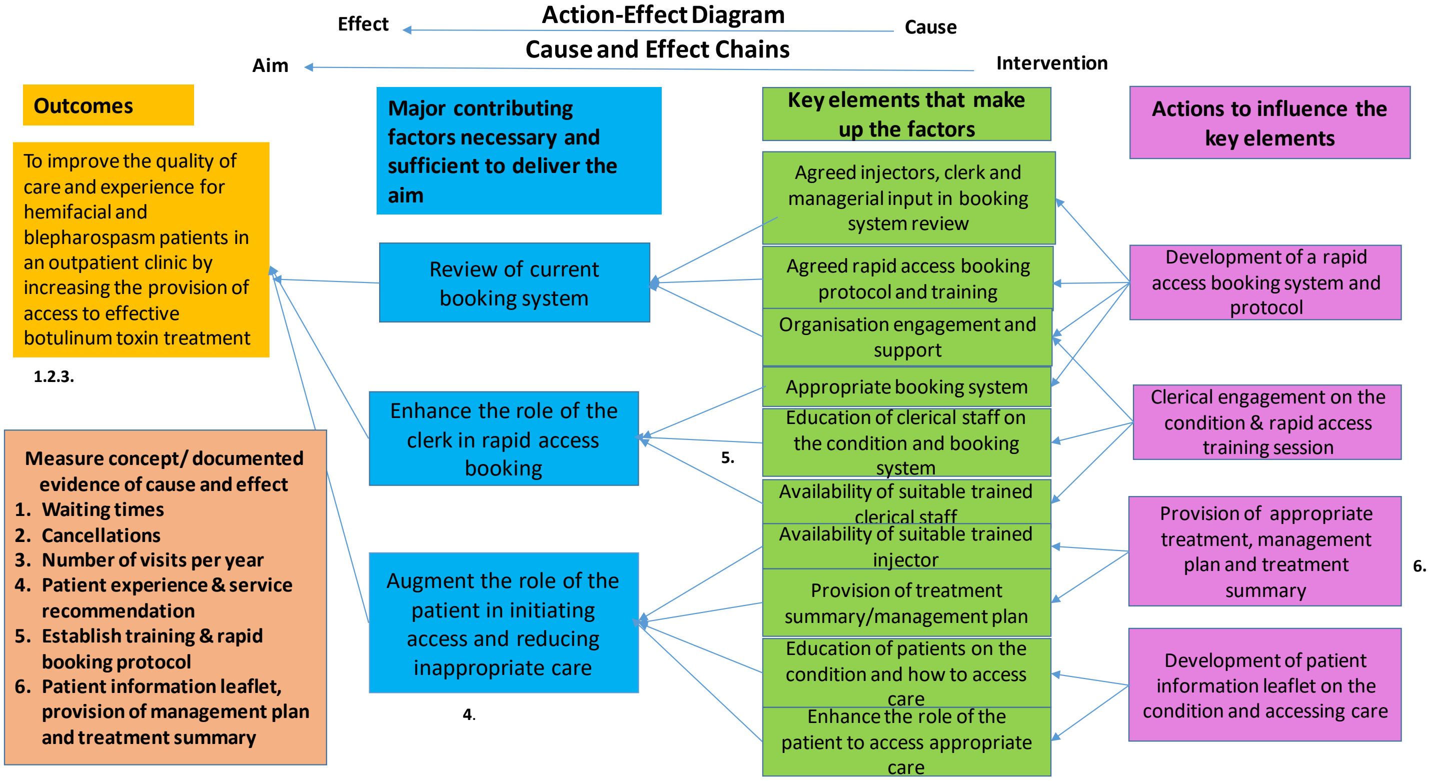
Appendix 4

Stakeholder Position



Stakeholders who need to be influenced in favour of patient-initiated botulinum toxin treatment model

Appendix 5



Appendix 6

PDSA Cycle

Plan

The research seeks to answer; how will the implementation of a patient-initiated botulinum toxin treatment model improve the quality of care for hemifacial and blepharospasm patients in the outpatient setting? However, framing the research under various headings enables the author to continually evolve the nature of the question to make improvements, for example:

Exploratory: What impact does the implementation of a patient-initiated botulinum toxin treatment model for hemifacial and blepharospasm have on the quality of effective care, organisation and services, patient experience and team expertise?

Descriptive: How does the implementation of a patient-initiated botulinum toxin treatment model for hemifacial and blepharospasm improve the provision of care, organisation and services, patient experience and team expertise in comparison to routine scheduled treatment?

Explanatory: An analytical comparison of the effective provision of care; organisation and services, patient experience, and team expertise before and after the implementation of a patient-initiated botulinum treatment for hemifacial and blepharospasm patients in an outpatient setting.

Evaluative: Did the implementation of a patient-initiated botulinum toxin treatment model for hemifacial and blepharospasm impact positively on effective care provision, organisation and services, patient experience and team expertise?

Is asking the right question important? The question asked implies the method of investigation to be used; it is the nucleus of the evaluation and a deciding factor on whether the research is process or outcome-driven. Although the initial research question sought to find out if quality can be improved with the implementation of a patient-initiated botulinum toxin service, it did not specify the areas that would be assessed to determine if quality of care has been achieved. The action of reflecting and changing the question allowed analysis, refocusing, evolution and improvement in order to reframe it as an operationalisation research question. Practically, the research is about assessing the impact of a patient-initiated treatment model on effective care provision, organisation, services, patient experience and team expertise and based on the outcome, will provide access to effective treatment and quality care beyond the RCT period. Following final consideration, the question was reconceptualised to provide a solid plan; the implementation of a hemifacial and blepharospasm patient-initiated botulinum toxin treatment model: what impact does it have on the provision of effective care, organisation of service, patient experience and team expertise?