Mental Health and Behaviour in Childhood Dystonia

Lauren Bates

June 2018

Research submitted in partial fulfilment of the requirements for the degree of Doctor in Clinical Psychology (DClinPsy), Royal Holloway, University of London.

Acknowledgements

I would like to thank Dr Tamsin Owen for proposing this project and helping with the initial stages. Thank you to Dr Jessica Kingston and Dr Sarah Rudebeck who supported me through the majority of this project: Jess with her advice and anticipatory problem-solving skills; and Sarah for providing clinical expertise, supporting with data collection and remaining involved even whilst on maternity leave. Thank you to Dr Alison Blencowe for supervising me through the later stages of data collection. Thank you to Dr Michelle Taylor for your responsiveness and reassurance during the write-up process.

Thank you to the Complex Movement Disorder Service team who were invaluable in identifying and contacting participants for this research. Thank you for your interest in the research and enthusiastic efforts in talking to patients about it.

A huge thank you to every young person and parent/guardian that took part in this research. I was inspired by your courage and achievements despite the difficulties you face; I hope this research will contribute towards improving outcomes and quality of life for young people with dystonia.

Finally, thank you to Alastair and everyone else in my life who has supported and believed in me throughout this whole Doctorate process.

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Executive Summary

Systematic Review

Dystonia is a movement disorder in which "sustained or intermittent involuntary muscle contractions cause twisting or repetitive movements or abnormal postures'' (Sanger, 2003, p. 1509) which impact everyday functioning. The biopsychosocial model suggests that the interrelation of biological, psychological and social factors impact overall health and wellbeing. Studies have shown as many as 66% of adults with dystonia meet criteria for a mental health problem over their lifetime (depression in 25-50%, anxiety in 34-60%). Due to the lack of control groups, and lack of variability in mental health problems explored, it is difficult to establish what the overall likelihood of having mental health difficulties is for adults with dystonia.

This review aimed to examine whether adults with dystonia are more likely to have a mental health problem than adults without dystonia. A literature search was conducted in PubMed and PsycINFO using 'dystonia', 'mental health' and 'control group' search terms. In total, 553 papers were screened against inclusion criteria (i.e., adults with dystonia and a control group, and data specifying numbers of participants scoring above clinical cut-off on a valid and reliable measure of mental health) and exclusion criteria (i.e., no tardive dyskinesia, not clear dystonia, and exclusion of mental health problems), which resulted in 14 papers being reviewed.

Studies were from a range of countries and site types, and included different types of dystonia. Sample sizes were 16-221 dystonia participants and 23-3943 control participants, and participant ages were 20-89 years. Studies used clinical interviews, self-report measures, or a combination of both. Quality assessment carried out using the Newcastle-Ottawa scale found that many studies were of poor quality due to the dystonia sample not being generalisable, no use of matching in case-control designs, and non-blinding of interviewers.

The majority of studies found that people with dystonia (55-64%) were significantly more likely to have a mental health problem than controls (14-37%). There was more consistent evidence for depression, general anxiety and social anxiety among people with dystonia. Some mental health problems were more likely in people with dystonia but were only explored in a small number of studies (e.g., bipolar affective disorder, alcohol problems and bulimia nervosa). There were conflicting results for some mental health problems (e.g., dysthymia, generalised anxiety disorder, obsessive-compulsive disorder, post-traumatic stress disorder, phobias and psychosis). No evidence was found to suggest people with dystonia were more likely to have adjustment disorder or somatization.

This review could access eligible journal articles in full, had no language bias, and used extensive search terms. However, the search was restricted to journal articles, and conducted using just two databases. Studies had different inclusion and exclusion criteria (e.g., use of antidepressants), and some had insufficient sample sizes to provide adequate statistical power or did not do statistical comparisons. This meant that it was not possible to discuss subtypes of dystonia separately. For some

mental health problems, too few studies reported data that reliable conclusions could be drawn from.

The results from this review are generalisable (due to the range of methodologies used, types of dystonia and countries), however there is limited consistency. More research is needed with control groups on a greater range of mental health difficulties. Nonetheless, it is important for clinicians to be aware of the signs for mental health difficulties people with dystonia may have. Screening measures for mental health problems among people with dystonia could be useful in clinical settings.

Empirical Paper

Introduction

Many forms of dystonia begin in childhood; therefore, research with children is needed to identify areas where they may need support. Factors such as not being able to take part in activities and missing school may impact self-esteem and the ability to interact with other young people. Children with movement disorders have been found to have cognitive and behavioural problems, depression, and anxiety. Not much is known about mental health in young people with dystonia, or the factors that make them more or less likely to have a mental health problem.

The research aimed to answer the questions: 1) Are young people with dystonia more likely to have mental health difficulties and behavioural problems

than the general child and adolescent population, and; 2) To what extent are self-concept, severity of movement disorder, pain, and age of onset of dystonia associated with mental health and behavioural problems in young people with dystonia? Young people with dystonia were hypothesised to score significantly higher on measures of emotional and behavioural difficulties than normed controls; and higher levels of severity and pain, younger age of onset and lower self-concept were hypothesised to be significantly associated with higher levels of emotional and behavioural problems in young people with dystonia.

This research was designed to help clinicians identify whether young people with dystonia are likely to need additional support and whether there are identifiable factors associated with mental health problems.

Method

A cross-sectional design (mixed quasi-experimental and within-subjects) was used. Fifty children and adolescents with dystonia and a parent or guardian were recruited from the Evelina London Children's Hospital (Complex Movement Disorders Service, Neurology and Botulinum toxin (Botox) clinics). Young people were aged between 8 and 17 years old. Young people with a life-limiting condition or those who were not able to understand questionnaires or consent for themselves were excluded from the study.

Young people and their parents were met on the day of a medical appointment. Young people completed the Beck Youth Inventories (BYI) a self-report

questionnaire with depression, anxiety, anger, disruptive behaviour and self-concept subscales, and the Strengths and Difficulties Questionnaire (SDQ) a brief, behavioural screening questionnaire with emotional, conduct, hyperactivity, peer relationship and prosocial subscales. Parents completed the SDQ-P, a demographic and clinical questionnaire and the Paediatric Pain Profile (PPP) a behavioural rating scale which measures pain. Gross Motor Function Classification System Expanded and Revised (GMFCS-E&R) scores were collected from medical records.

IBM SPSS Statistics was used for all analyses. One sample z tests were used to compare BYI and SDQ data from the dystonia sample to the population norms. Pearson's product moment correlation coefficients and hierarchical multiple regression analyses were used to identify which factors were significantly associated and provided unique contributions to BYI and SDQ subscales.

Results

Higher levels of anxiety were found for females, participants with learning needs, those who had deep brain stimulation surgery (DBS), and those with moderate/severe pain. High levels of pain were also associated with higher levels of disruptive behaviour, depression, and parent-rated total difficulties. Participants with secondary dystonia had higher levels of anger.

Both children and adolescents with dystonia had higher levels of anxiety (47.9% above clinical cut-off), and lower levels of disruptive behaviour than population norms. Children had lower levels of depression and anger than

population norms, but this was not the same for adolescents (23.4% of the dystonia sample scored above cut-off for depression). For the SDQ, young people with dystonia self-reported higher levels of peer problems and lower levels of conduct problems than population norms. Parents of children with dystonia reported that their child had significantly higher total difficulties, emotional problems, hyperactivity and peer problems, and significantly lower prosocial behaviours than the population norms.

Hierarchical multiple regression analyses revealed that self-concept made a significant unique contribution to explaining anxiety, and type of dystonia and pain each made significant unique contributions to explaining parent-reported total difficulties.

Discussion

High levels of anxiety, peer problems, and parent-rated hyperactivity in people with dystonia is consistent with previous literature. Lower levels of depression, anger and disruptive behaviour is inconsistent with previous literature. This may have been impacted by the study's small sample size, the suitability of measures (e.g., the disruptive behaviour scale not being suitable for people with disabilities) and factors such as social desirability. Overall, parent-reports were more consistent with previous literature, which typically has relied on parent-rated measures.

The association between self-concept and anxiety replicates findings from research in cerebral palsy, however self-concept was not associated with depression, anger, or any of the behavioural subscales. Associations between pain and depression and parent-rated total difficulties supports previous literature on children with movement disorders. Contradictory to the hypothesis of the present study, neither age of onset of dystonia nor severity of movement disorder were significantly associated with any of the psychological or behavioural measures. This may be due to the age range being smaller than adult studies, and the measure of severity not capturing fine motor movements or communication.

The study had some methodological limitations. Missing data and the relatively small sample size led to the study being statistically underpowered. Specified age ranges for the population norms also led to small subsamples for these analyses. The conclusions of the present study must therefore be interpreted with caution. A strength of the study is that every child could self-report difficulties using well-validated measures of mental health and behavioural problems. Participants not taking part due to capacity and communication issues and the high number of young people who had DBS limit the generalisability of results.

Tentative suggestions are made, such as mental health screening and risk assessments for dystonia patients, particularly those who may be more likely to have a mental health problem. Targeting self-esteem may help to reduce anxiety, and targeting pain may help reduce anger and behavioural problems. Future research could explore the nature of anxiety, factors associated with depression, and mental

health needs in young people with learning needs and severe communication problems.

Integration, Impact and Dissemination Plan

The Systematic Review informed some decisions for the Empirical study, such as using population norms as a control group, broad inclusion criteria, and focusing on depression and anxiety. The two pieces of research differed on the use of measures. The process of doing the Systematic Review helped me understand about the needs of people with dystonia.

Some of the participants were offered psychological support, and clinicians were encouraged to discuss the scores on measures with patients as part of their routine clinical care. This study will hopefully increase awareness of factors associated with mental health difficulties (e.g., having peer problems, DBS surgery, learning difficulties, low self-esteem and high pain levels). Screening for mental health problems may be a quick and economical way to gain information in routine medical appointments. Identifying needs in young people may protect against difficulties in later life. Play therapy or compassion-focused therapy (CFT) may be useful to improve self-esteem, anxiety, and depression. Cognitive behavioural therapy (CBT) may reduce disability and improve emotional functioning for those who experience pain. It is important that clinicians can access appropriate support for patients.

Future research should use child and adult reported data, and explore development of suitable measures to access children with communication difficulties. Replication in a larger sample is warranted to support the conclusions made by this study.

Personally, I noticed the resilience of the participants and enjoyed witnessing children and their parents communicating with each other about emotional issues. Whilst it was difficult to learn of the challenges that young people and their families face, many young people were achieving things with support.

The main results, factors associated with mental health problems and suggestions for preventing mental health difficulties and accessing support resulting from this research (e.g., developing self-esteem, peer relationships, and managing pain) will be communicated with participants, the Evelina London Children's Hospital and The Dystonia Society.

Are Adults With Dystonia More Likely to Have Mental Health Problems Than Adults Without Dystonia? A Systematic Review of Comparison Studies.

Abstract

There is an extensive literature on mental health problems in adults with dystonia; however, there is less research on how adults with dystonia compare to the general population. This review aimed to examine whether adults with dystonia are more likely to have a mental health problem than adults without dystonia. A search was conducted in PubMed and PsycINFO using 'dystonia', 'mental health' and 'control group' search terms. Data were extracted and quality assessed using the Newcastle-Ottawa scale (Wells et al., 2013). Results are outlined in terms of the methodological quality of studies and data reported on numbers of adults with and without dystonia who have mental health problems. Results showed that studies comparing dystonia and control groups varied greatly in their methodology and quality. Overall, adults with dystonia were found to have depression, anxiety, and social phobia significantly more than controls. The results for other mental health problems were varied. The strengths and weakness of the studies included in this review are discussed. Implications for clinical practice and research are also discussed.

Introduction

This review aimed to determine whether adults with dystonia are more likely to have mental health problems than adults without dystonia. Many studies on adults with dystonia have included measures of mental health difficulties; however, they often had relaxed inclusion criteria (where participants had comorbid health conditions), or strict inclusion criteria (focusing on one specific subtype). This makes it difficult to establish what the overall likelihood of having mental health difficulties is for adults with dystonia. In addition, few studies have a control group to which the prevalence of mental health difficulties can be compared. This makes it difficult to ascertain which difficulties are more likely to occur in dystonia as compared to adults without dystonia. This is important to establish to help services supporting people with dystonia gain a better understanding of their mental health needs.

Defining Dystonia

Dystonia is a movement disorder in which "sustained or intermittent involuntary muscle contractions cause twisting or repetitive movements or abnormal postures'' (Sanger, 2003, p. 1509). These movements are typically stereotyped or patterned, twisting, and are initiated or made worse by voluntary action. There is some debate as to what classifies as dystonia and what does not (Frucht, 2013), and historically dystonia has been classified by age at onset, distribution, and aetiology (Albanese et al., 2013). Dystonic movements are "more varied and phenomenologically richer than hyperkinetic movement disorders" (Frucht, 2013, p.

884), and features which indicate dystonia (such as a 'sensory trick', the reduction of symptoms with a slight touch to a specific area) must be explored by a neurologist to reach the correct diagnosis.

Bressman's (2003) distinction between primary and secondary dystonia is typically used in clinical settings. Primary dystonia is caused by genetic abnormalities. Several genes are implicated in dystonia, for example, DYT-11 myoclonus dystonia is a form where movements are more 'jerking' than other types, and is caused by abnormalities in the epsilon-sarcoglycan gene (SGCE). Secondary dystonia describes dystonia-plus (i.e., dystonia co-occurs alongside symptoms of other neurological disorders), degenerative, complex, and acquired forms; these forms are associated with other neurological or metabolic diseases and present with a mixture of neurological features. Secondary dystonias include tardive dyskinesia (due to antipsychotic medication), another movement disorder (i.e., cerebral palsy), or a brain injury (i.e., stroke). Idiopathic dystonia is when the cause is unknown, with no clear genetic cause or comorbid neurological abnormality. The prevalence of primary dystonia has been reported as 16.43 per 100,000 people (Steeves, Day, Dykeman, Jette, & Pringsheim, 2012), however the prevalence of secondary dystonia is unclear.

Dystonia diagnoses are often classified in terms of location (Balint & Bhatia, 2014; Geyer & Bressman, 2006). In focal dystonia, the movements present in one area of the body; for example, as turning of the neck (cervical dystonia), prolonged closing of eyelids (blepharospasm), loss of control of fine hand movements (writer's cramp or arm dystonia) or difficulty speaking (laryngeal dystonia). In segmental

dystonia, movements affect adjacent body areas. In generalised dystonia, all or most of the body is affected. In cranial dystonia (Meige syndrome) two or more areas above the neck are affected, this becomes cranio-cervical dystonia if it also affects the neck.

As there are several subtypes of dystonia; the exact neurological mechanisms are unclear. Abnormalities in the dopaminergic activity of the basal ganglia and in cortico-striato-thalamo-cortical circuitry are implicated in the disease (A. Berardelli et al., 1998; Degirmenci, Oyekcin, Bakar, & Kurklu, 2013; Maia, Cooney, & Peterson, 2008). Treatment includes botulinum toxin (Botox) injections, medication, physiotherapy or deep brain stimulation surgery (DBS; where electrodes are inserted into the brain attached to a batter-powered simulator, which deliver electrical stimulation to areas of the brain associated with dystonia; Mehdorn, 2016).

Most forms of dystonia do not affect a person's lifespan or cognition, however, abnormal muscle contractions in dystonia can cause problems with sitting, walking, talking, eating and sleeping (Hertenstein et al., 2016). Such physical limitations affect the person's ability to carry out everyday activities (Pavone, Burton, & Gaebler-Spira, 2013). Pain is a common complaint and improvements in pain management can improve symptoms of dystonia and quality of sleep (Ramstad, Jahnsen, Skjeldal, & Diseth, 2012). Adults with dystonia may find it difficult to have a job due to uncontrollable movements affecting productivity levels (Molho et al., 2016); making it difficult to achieve independence (Yeo & Sawyer, 2005).

Mental Health Problems Among Adults with Dystonia

Mental health difficulties in adults with dystonia have been well reported. Several studies have found over half of participants to meet criteria for a psychiatric diagnosis (Fabbrini et al., 2011; Moraru et al., 2002; Smit, Kuiper, et al., 2016) with 66% reported to meet criteria for a mental health problem over their lifetime (Wenzel et al., 1998). Mental health problems are strongly linked to poorer quality of life in adults with dystonia, particularly in terms of physical and social functioning (Page, Butler, & Jahanshahi, 2007). As mental health difficulties are associated with poorer health outcomes such as increased risk of cardiovascular disease and diabetes (Simon, 2001), it is important they are identified and addressed.

Studies exploring depression in adults with dystonia have reported 25-50% experiencing clinical levels of depression as measured by diagnostic interviews (Demartini et al., 2017; Moraru et al., 2002; Smit, Kuiper, et al., 2016; Wenzel et al., 1998); whilst one study using the Beck Depression Inventory (BDI) found 30% of 329 participants reported moderate-severe depression (Lewis, Butler, & Jahanshahi, 2008). Mood problems after diagnosis were self-reported by 61% of participants in one study (Comella & Bhatia, 2015).

Anxiety disorders are commonly reported in adults with dystonia; with 34-60% meeting criteria for anxiety disorders on diagnostic interviews (Demartini et al., 2017; Moraru et al., 2002; Smit, Kuiper, et al., 2016; Wenzel et al., 1998). Although few studies report the rates of specific anxiety disorders, dystonia has been associated with increased incidence of obsessive-compulsive disorder (OCD;

Barahona-Corrêa, Bugalho, Guimaraes, & Xavier, 2011; Bihari, Pigott, Hill, & Murphy, 1992), social anxiety (Dias et al., 2011; Moraru et al., 2002), agoraphobia (Smit, Kuiper, et al., 2016), and panic disorder (Wenzel et al., 1998). Panic disorder was reported by 30% of 44 participants; however, as this study included current and lifetime presence, some may not have had panic disorder and dystonia concurrently. Symptoms of anxiety, depression and OCD have been found to be stable across time, with participants scoring highly on screening measures and showing little change after a 5-year interval (Berardelli et al., 2015). Adults with dystonia therefore have mental health needs that are not addressed, potentially due to their physical needs taking priority (Coventry et al., 2011); alternatively, people with dystonia may be receiving treatment for mental health problems, but this is not sufficient for their complex needs.

Despite mounting evidence of an association between dystonia and mental health problems, much of this research is methodologically flawed. Due to the rarity of dystonia, research samples are often small, with broad inclusion criteria which may influence results, such as a wide age range and inclusion of extraneous variables such as medication (Barahona-Corrêa et al., 2011). As dystonia often presents similarly to or alongside other neurological disorders, it is important that a neurologist has confirmed the diagnosis of research participants, to ensure only people with dystonia are included. Additionally, it can be difficult to assess for mental health problems in dystonia as symptoms of dystonia overlap with features of difficulties such as depression (fatigue) or anxiety (feeling shaky). Using self-report measures may result in people scoring highly based on symptoms related to their

dystonia, rather than a mental health difficulty. Studies using clinical interviews could arguably ascertain more valid statistics on people with dystonia who have mental health problems, as their specific symptoms could be explored to determine if they are linked to mental health difficulties or are a result of their dystonia.

Despite a large body of research suggesting that adults with dystonia are vulnerable to experiencing mental health difficulties, it is difficult to establish whether these mental health difficulties are more than what would be expected in the general population. This is due to both groups not being compared fairly (i.e., with similar measures and with similar inclusion or exclusion criteria). Thus, it is important to review the evidence available where adults with dystonia are compared to adults without dystonia, to establish whether one group is more likely to have mental health difficulties than the other. In addition, it is important to explore a larger range of mental health difficulties, as conditions such as psychosis, somatization, and eating disorders, may have been overlooked.

Biopsychosocial Factors and Mental Health Problems Among Adults with Dystonia

Whilst it is difficult to establish causality for mental health problems in dystonia, research has suggested that mental health problems and dystonia share a common neurological basis (Barahona-Corrêa et al., 2011); such as genetic for those with primary dystonia (Heiman et al., 2004), or an impairment of dopaminergic and serotonergic neurotransmission (Brüggemann et al., 2014) in the basal ganglia (Smit, Bartels, et al., 2016). Whilst biological aspects are likely to contribute towards

mental health, other factors will also influence health outcomes. The biopsychosocial model of health (Figure 1) displays the interrelation of biological, psychological and social aspects that influence how a person might experience a health problem (Engel, 1977). This model explains how psychological factors, such as stress, have an adverse effect on one's health, that cannot be explained by biological mechanisms alone. Sociological factors, such as hierarchy in the social system, can influence health seeking behaviour or the quality of care someone can access (Engel, 1980). This model has been applied to movement disorders, where the exacerbation of movement symptoms was associated with psychological difficulties and historical traumatic experiences (Kranick et al., 2011). Psychological and social factors impact how a person responds to being diagnosed with a long-term health condition, and consequently how they approach treatment. Movement disorders have been found to be associated with emotional state (Tomic et al., 2016). This association is likely due to complex interplay between biological (e.g., neurobiological predisposition, pain and sleep disturbance), psychological (e.g., frustration and self-esteem) and social (e.g., perceived stigma) factors (Degirmenci et al., 2013; Hertenstein et al., 2016; Lewis et al., 2008; van den Dool, Tijssen, Koelman, Engelbert, & Visser, 2016).

The association between physical health problems and depression is well established (Barnett et al., 2012). Twenty percent of people with a chronic physical health problem (such as cancer, diabetes, heart disease or stroke) have depression (NICE, 2010), which can have a significant negative impact on health outcomes and functioning (Gili et al., 2013). Higher levels of disability in movement disorders are associated with lower activity levels (Tomic et al., 2016), which are a risk factor for

depression (Camacho, Roberts, Lazarus, Kaplan, & Cohen, 1991) according to behavioural models (Ferster, 1973). For example, a person with dystonia may stop working due to becoming depressed because of fatigue, uncontrollable movements and pain affecting their ability to perform at work (Molho et al., 2016; Yeo & Sawyer, 2005). In dystonia, disfigurement, negative body concept and low self-esteem are associated with self-reported depression (Lewis et al., 2008).

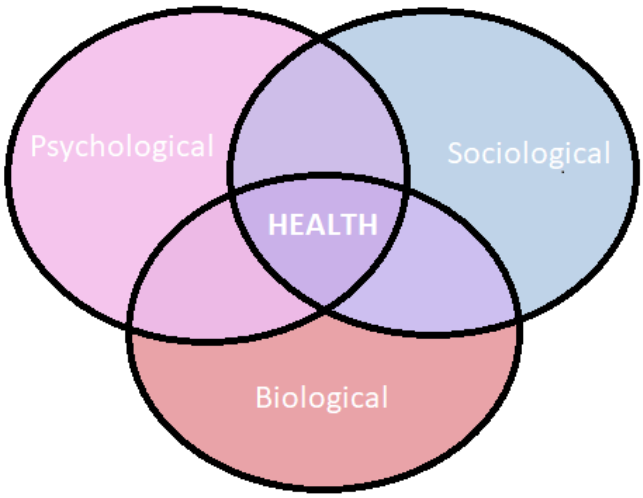

Figure 1: *The Biopsychosocial Model of Health (Engel, 1980)*

Many studies have identified factors explaining the link between anxiety and dystonia. Stigma towards people with physical differences and being stared at due to their movements, may cause an exacerbation of anxiety in social situations and impact how people with dystonia interact socially (Mordin, Masaquel, Abbott, & Copley-Merriman, 2014). One study on cervical dystonia found that participants

performed poorly on socio-cognitive tasks (Czekóová, Zemánková, Shaw, & Bareš, 2017), suggesting that people with dystonia may have poorer social skills. Although it is difficult to determine if these differences are due to movement disorder symptoms, or due to lack of experience in social situations, it supports research showing that dystonia is associated with social anxiety (Lauterbach, Freeman, & Vogel, 2004). Experiencing symptoms of dystonia (i.e., muscle spasms) can cause increased anxiety and panic if these symptoms are misinterpreted as something dangerous (Walker, 2002). Anxiety can predate dystonia onset, for example one study found that stressful life events often preceded the onset of blepharospasm (Johnson et al., 2007).

Whilst many studies have explored depression and anxiety in dystonia, fewer have explored other mental health difficulties. One study exploring movement disorders in antipsychotic naïve schizophrenic patients, found siblings of these patients were 17-23% more likely to have movement disorder symptoms than healthy controls (Koning, Kahn, Tenback, van Schelven, & van Harten, 2011). This supports the biological link between mental health problems and movement disorders. It is important to acknowledge that dystonia does not necessarily cause mental health difficulties. It can be difficult to determine the effects of dystonia on mental health when many adults had difficulties before their dystonia diagnosis (Tomić et al., 2017). One study found that mood disorders tend to develop after a diagnosis of dystonia, whilst anxiety conditions tend to develop before (Lauterbach et al., 2004). However, these results were based on participants retrospectively reporting their psychiatric and medical histories and so may not be accurate.

Aims of This Review

Mental health problems in dystonia have been widely reported, however most research has focussed on one specific mental health problem in one specific dystonia subtype, making it difficult to ascertain the association between dystonia and mental health problems generally. Whilst systematic reviews have been published exploring non-motor symptoms in genetic dystonia (Peall et al., 2013) and dystonia-plus syndromes (Sunga & Rosales, 2014), there has been no published review exploring the presence of mental health difficulties across a range of dystonia types, in comparison to the general population. This review aims to systematically review the research on mental health problems in adults with dystonia in comparison to adults without dystonia, to determine whether adults with dystonia are more likely to have mental health difficulties. The clinical implications of this literature and the key avenues for future research shall be identified and discussed.

Methods

Inclusion and Exclusion Criteria

The inclusion criteria for studies were: having one group of participants with a diagnosis of dystonia, and another group consisting of a healthy control or population-based sample; data from both groups specifying numbers of participants scoring above clinical cut-off on a valid and reliable measure of a mental health problem, and; participants over 18 years of age. The exclusion criteria for studies were: participants with conditions which were not explicitly dystonia (e.g., hemifacial

spasm, neurocirculatory dystonia); inclusion of participants with tardive dyskinesia; studies selecting or excluding participants based on whether they had mental health problems; and reviews and meta-analyses.

Search Strategy

Two electronic databases were used to generate the literature searches; PubMed and PsycINFO. These databases were selected to capture publications from both medical and psychological fields, deemed to be most relevant to the research question.

The search terms (Table 1) were used in the Abstract/Title field in PubMed. For PsycINFO, two separate searches were made, one for Abstract, and one for Title. For both databases, filters were applied to limit the searches to All Journals for Publication Type, and Human for Population Group.

Table 1: *The Search Terms Used in the Database Searches*

Dystonia	**Mental Health**		**Control**
dystonia	mental health	anorexic	case-control
blepharospasm	mental disorder*	anorexia	controlled
writer's cramp	mental illness	bulimic	healthy
Meige syndrome	mental disab*	bulimia	controls
	wellbeing	adjustment disorder	compared
	psychological	post traumatic stress disorder	comparison
	psychopathology	post-traumatic stress disorder	
	psychiatric	PTSD	
	mood	somatic	
	affective	somatoform	
	depressed	somatization	
	depression	psychosis	
	depressive	psychotic	
	dysthymic	schizophrenia	
	dysthymia	schizophrenic	
	anxiety	manic	
	panic	mania	
	agoraphobic	bipolar	
	agoraphobia	personality disorder	
	phobia	substance abuse	
	phobic	substance dependence	
	OCD	drug abuse	
	obsessive compulsive disorder	drug dependence	
	obsessive-compulsive disorder	alcohol abuse	
	body dysmorphic disorder	alcohol dependence	
	BDD		

The dystonia search terms were chosen to capture all forms of dystonia, not including conditions where dystonia is only one potential criteria for diagnosis (such

as Hallervorden-Spatz syndrome). This was to limit the effects of other neurological or medical comorbidities influencing results. The mental health terms were chosen according to the diagnostic categories on the Mini International Neuropsychiatric Interview- Plus (MINI+; Sheehan, 1998) and the Structured Clinical Interview for DSM-IV (SCID-IV; First, Spitzer, Gibbon, & Williams, 2002), and terms used in a previously conducted systematic review on mental health (De Silva et al., 2014). The aim was to capture the largest range of mental health problems possible. For the searches, title and abstract were chosen to ensure that mental health data were likely to be reported in the full text article. The search was restricted to journal articles to ensure that the full text article could be assessed for eligibility. The search was restricted to humans to eliminate animal laboratory studies. The searches were conducted in December 2017, and repeated in April 2018 to identify more recently published articles.

Process of Study Selection

All references were imported into Mendeley (Mendeley, 2015), a reference manager. Mendeley was used to identify duplicates and sort references into folders according to which papers were reviewed at each stage. Stage 1 involved removing duplicate articles. Stage 2 involved screening for eligibility based on articles' titles and abstracts, removing those which clearly did not meet the inclusion criteria, or met exclusion criteria. Stage 3 involved reading the full texts of articles and removing those which did not meet inclusion criteria, or met exclusion criteria. The author of

this review completed these screening stages and extracted the details of the final studies by collating them into a Microsoft Excel spreadsheet. No supplementary information was sought from the publishers of the chosen studies.

Process of Data Extraction

The characteristics of the studies chosen for reporting were Author, Date, Title, Country of Origin, Design, Groups (*n* per group), Inclusion/Exclusion criteria, Recruitment Procedure/Site, Age, Sex, Treatment Details and Measure of Mental Health. Details such as ethnicity and social economic status were not recorded as they were rarely reported in the studies.

Process of Quality Analysis

All studies were rated for quality by the author, using the Newcastle-Ottawa scale (Wells et al., 2013). This scale is recommended as a tool for evaluating quality of case-control studies (Zeng et al., 2015), and is suitable for use in systematic reviews (Deeks et al., 2003). The Newcastle-Ottawa scale (original version available in Appendix 1) is a star-based system, where studies are awarded stars according to specific criteria. A maximum of 9 stars can be awarded, higher numbers meaning the study is of higher quality. The measure has three main categories; for this review, the criteria for awarding stars were adapted (see Table 2).

Table 2: *Adapted Newcastle-Ottawa Scale Categories*

Category			Description
Selection	**1**	**a**	dystonia diagnosis confirmed (e.g., by a neurologist) *
		b	relied on self-report
	2	**a**	dystonia participants deemed representative *
		b	dystonia participants deemed not to be representative
	3	**a**	community controls *
		b	hospital controls
		c	no description or snowball sampling
	4	**a**	inclusion of controls with mental health problems *
		b	inclusion of mental health problems not clarified
Comparability	**1**	**a**	age *
		b	sex *
Exposure	**1**	**a**	medical record *
		b	structured interview blinded *
		c	structured interview not blinded
		d	self-report measure
	2	**a**	both groups used the same measure *
		b	groups used different measures
	3	**a**	both groups had the same non-response rate *
		b	non- respondents described for both groups
		c	response rate was unknown for one or both groups

Note: * 1 star is awarded for meeting this criterion

The Selection category refers to: 1) if the diagnosis of dystonia participants was clarified; 2) how representative dystonia participants were of the general population with the same diagnosis; 3) how control participants were recruited, and; 4) if the study specified whether controls with mental health problems were included. The Comparability category refers to variables of which the dystonia and

control participants were matched in the design or adjusted for in the analysis. The Exposure category refers to: 1) the main measure used; 2) whether both dystonia and control participants were assessed using the same measure; and 3) details on the response rate.

Process of Data Synthesis

Due to the lack of consistency between studies it was deemed inappropriate to compare them in a meta-analysis as the data would not meet the assumption of homogeneity required (i.e., similar measures and inclusion and exclusion criteria; Boland, Cherry, & Dickson, 2014).

To make comparisons between studies, odds ratios and confidence intervals were calculated using online software (MedCalc, 2017) where possible. Odds ratios were not calculated where authors had calculated them, or where one of the groups had no participants with that mental health problem. Odds ratios were calculated by comparing the numbers of people in the dystonia group who did and did not have a mental health problem, with the numbers of people in the control group who did and did not have a mental health problem. The odds of having a mental health difficulty is the probability that one group would have a mental health difficulty in comparison to another group (see Table 3 for interpretation using cut-offs identified by Monson, 1990).

Table 3: *Cut-Offs for Interpreting Odds Ratios (Monson, 1990)*

	Controls	**Dystonia**
No Association	0.9 - 1.0	1.0 - 1.2
Weak Association	0.7 - 0.9	1.2 - 1.5
Moderate Association	0.4 - 0.7	1.5 - 3.0
Strong Association	0.1 - 0.4	3.0 - 10.0
Infinite Association	<0.1	>10.0

Results

The process of study selection is displayed in Figure 2. The initial search resulted in 553 papers. After removing duplicates at Stage 1, 440 papers remained. After screening titles and abstracts at Stage 2, 44 papers remained. At Stage 3, papers were excluded for the following reasons: numbers scoring above cut-off on mental health measure not reported (10); participants excluded for having mental health problems (9); study not using a valid measure of mental health (4); dystonia sample including participants under 18 years old (2); dystonia sample not exclusively diagnosed with dystonia (2); no healthy control group (2), and; mental health measure not given to healthy controls. Fourteen papers were therefore included in the body of this review.

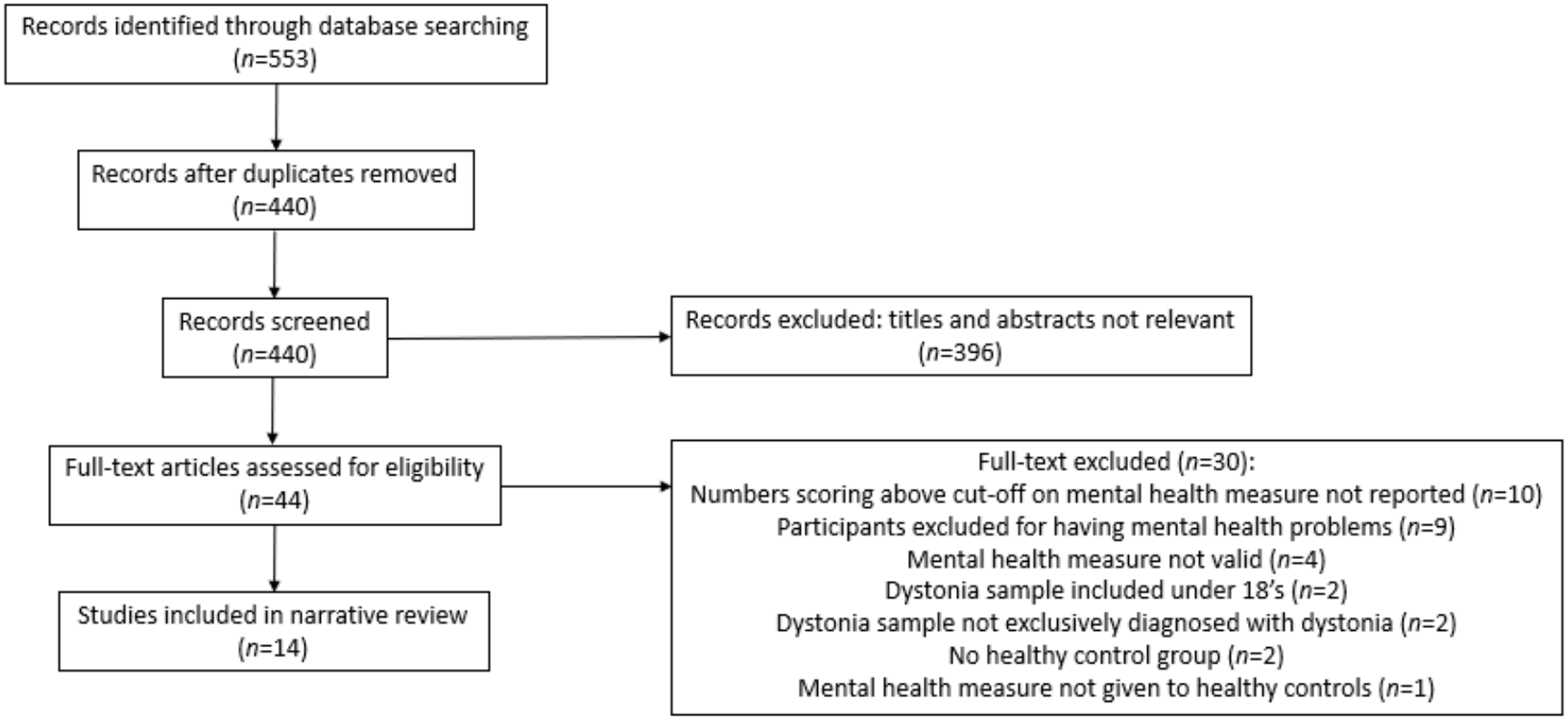

Figure 2: *PRISMA Diagram Showing Stages of Screening of Eligible Articles*

Methodology of Included Studies

The included studies vary widely in methodology and are shown in Table 4.

Design

All studies used a case-control design. Case-control studies are typically used in medical research to determine the odds of exposure to the risk factor of interest (i.e., mental health) for cases (i.e., adults with dystonia) and controls (i.e., healthy adults). Of the studies, 10 out of 14 matched participants in some way (one study only matched participants on some analyses (Lencer et al., 2009)).

Recruitment

The studies span across eight countries (four from the United States of America, two from Italy, Germany, and the Netherlands, and one from Portugal, Australia, Thailand and China).

None of the studies recruited from the same sites, it is therefore assumed that different participants took part in each study. The dystonia samples were recruited from a variety of site types (4 from Movement disorder clinics, 4 from neurology clinics, 3 from dystonia support networks, and 3 from Botox clinics), with several studies recruiting from multiple sites, and one study not reporting where they recruited from (Brashear et al., 2012).

The control samples were recruited from a variety of sources (5 from friends and family of patients or authors, 3 from a now-healthy hospitalised population, and 1 via advertisements); with two studies using pre-existing data from population studies (Lehn, Mellick, & Boyle, 2014; Lencer et al., 2009), and four studies not specifying the origin of their control group.

Inclusion and Exclusion Criteria

The dystonia population varied between studies. Eleven studies had participants with focal dystonia (11 blepharospasm, 8 cervical dystonia, 1 writer's cramp, 1 arm dystonia, 1 laryngeal dystonia, 1 Meige syndrome, and 1 cranio-cervical dystonia), one study had segmental and generalised dystonia (Lauterbach et al., 2004), one study had myoclonus-dystonia (van Tricht et al., 2012), and one study had rapid-onset dystonia-parkinsonism (Brashear et al., 2012).

Seven studies reported inclusion and/or exclusion criteria for participants with dystonia and controls, four studies reported this for participants with dystonia only (Fabbrini et al., 2010; Lauterbach et al., 2004; Lencer et al., 2009; Yang et al., 2016), one reported this for controls only (Lehn et al., 2014), and two studies did not provide this information (Brashear et al., 2012; Johnson et al., 2007).

Six studies required the participants with dystonia to have no secondary dystonia (Barahona-Corrêa et al., 2011; Eichenseer, Stebbins, & Comella, 2014; Fabbrini et al., 2010; Lencer et al., 2009; van Tricht et al., 2012; Yang et al., 2016), four required no other neurological abnormalities (Fabbrini et al., 2010; Paus et al.,

2011; Smit, Kuiper, et al., 2016; Yang et al., 2016), and one required no comorbid movement disorders (Setthawatcharawanich, Sathirapanya, Limapichat, & Phabphal, 2011). There was inconsistency in terms of control inclusion and exclusion criteria; for example, Smit, Kuiper, et al., (2016) required controls to have no family history of dystonia, whereas other studies specifically recruited from family members of dystonia patients (Paus et al., 2011; van Tricht et al., 2012). Three studies excluded participants who were taking medication (Barahona-Corrêa et al., 2011; Setthawatcharawanich et al., 2011; Smit, Kuiper, et al., 2016). Two studies included a cut-off relating to intellectual ability for both groups (e.g., MMSE score; Barahona-Corrêa et al., 2011; Mula et al., 2012).

Sample Size

There was variation in sample size across and within studies. Dystonia samples ranged from 16 to 221 participants whereas control samples varied from 23 to 3943 participants. As some studies broke down the dystonia group into types of dystonia, this meant that some statistical comparisons were done with as few as 10 participants per group (Fabbrini et al., 2010) which would have led to the studies not having enough statistical power to find differences between groups.

Participant Characteristics

Most studies reported the mean age of participants. Dystonia participant mean ages were 40 to 69 years, the range being 20 to 89 years. The mean ages of control participants were 36 to 69 years, the range being 18 to 84 years.

One study did not report the sex of participants (Smit, Kuiper, et al., 2016). The majority of studies (except Barahona-Corrêa et al., 2011 and Brashear et al., 2012, and one subtype in Fabbrini et al., 2010 and van Tricht et al., 2012) had more female participants than male. This may be due to more women being affected by many types of dystonia than men (Soland, Bhatia, & Marsden, 1996).

All studies (except Brashear et al., 2012) included participants who took medication. In seven out of 14 studies, all dystonia participants were either currently taking medication, or had recently had Botox injections (Eichenseer et al., 2014; Fabbrini et al., 2010; Johnson et al., 2007; Lehn et al., 2014; Lencer et al., 2009; Mula et al., 2012; Setthawatcharawanich et al., 2011). Five studies provided details on medication that controls were taking, such as antidepressants (Eichenseer et al., 2014; Fabbrini et al., 2010; Lehn et al., 2014; Paus et al., 2011; van Tricht et al., 2012).

Table 4: *Extracted Information From Included Studies*

Author, Date & Title	**Country of Origin**	**Groups (*n*)**	**Inclusion/ Exclusion Criteria**	**Recruitment (procedure/ site)**	**Age (mean ± SD)**	**Sex (m/f)**	**Treatment/ Medication**	**Measure(s) of Mental Health**
Barahona-Corrêa et al. (2011) Obsessive-Compulsive Symptoms in Primary Focal Dystonia: A Controlled Study	Portugal	Dystonia Primary focal (BL 15, CD 15, WC 15) Controls Similar age & gender (30)	Dystonia >18; primary pure focal, late-onset, normal MRI All MMSE below cut-off, no anticholinergics, neuroleptics or antidepressants	Dystonia Dystonia case register Controls Friends of dystonia participants	Dystonia 54.1 ± 13.9 Controls 49.1 ± 14.0	Dystonia 30/15 Controls 14/16	Dystonia 60% Botox Controls Not reported	All Y-BOCS MINI MINI+
Brashear et al. (2012) Psychiatric Disorders in Rapid-Onset Dystonia-Parkinsonism	United States of America	Dystonia RDP (26) Controls NMC (3) NC (27)	All Not reported	Dystonia Not reported Controls Family members of dystonia participants	Dystonia 39.9 ± 15.0 Controls NMC: 62.7 ± 23.7 NC: 48.2 ± 14.6	Dystonia 19/7 Controls NMC: 1/2 NC: 12/15	All Not reported	All SCID-I

Author, Date & Title	Country of Origin	Groups (*n*)	Inclusion/ Exclusion Criteria	Recruitment (procedure/ site)	Age (mean ± SD)	Sex (m/f)	Treatment/ Medication	Measure(s) of Mental Health
Eichenseer et al. (2014) Beyond a Motor Disorder: A Prospective Evaluation of Sleep Quality in Cervical Dystonia	United States of America	Dystonia CD (54) Controls Matched for age & gender (55)	Dystonia >18 years, diagnosis primary CD. No other dystonia, prior surgical treatment, or sleep disorder. Controls No neurological or primary sleep disorders	Dystonia Botox treatment clinic Controls Medical Centre	Dystonia 62 ± 10.1 Controls 62 ± 10.3	Dystonia 20/80% Controls 19/81%	Dystonia 35% benzodiazepines 4% anticholinergics 7% baclofen 20% anti-depressants 100% serial Botox treatment Controls 9% benzodiazepines 11% antidepressants	All BDI-II HAM-A
Fabbrini et al. (2010) Psychiatric Disorders in Adult-Onset Focal Dystonia: A Case-Control Study	Italy	Dystonia Focal (CD 34, BL 28, AD 11 & LD 16) Controls 4 groups (CD 32, BL 23, AD 10 & LD 12). Matched for age, sex, SES & education.	Dystonia Diagnosed focal dystonia, classified by site of onset, no neurological abnormalities or secondary dystonia Controls Not reported	Dystonia Neurology clinic Controls Not reported	Dystonia CD: 59.4 ± 14.4 BL: 69.1 ± 8.8 LD: 63.3 ± 11.0 AD: 52.4 ± 16.4 Controls CD: 58.9 ± 14.5 BL: 67.3 ± 10.5 LD: 64.2 ± 10.9 AD: 54.7 ± 17.9	Dystonia CD: 9/25 BL: 7/21 LD: 2/14 AD: 10/1 Controls CD: 9/23 BL: 7/16 LD: 3/9 AD: 9/1	Dystonia All Botox >4 months previously 7 CD took anxiolytics Controls 8 CD on antidepressants & anxiolytics	All SCID-I

Author, Date & Title	Country of Origin	Groups (*n*)	Inclusion/ Exclusion Criteria	Recruitment (procedure/ site)	Age (mean ± SD)	Sex (m/f)	Treatment/ Medication	Measure(s) of Mental Health
Johnson et al. (2007) Closely Spaced Stressful Life Events Precede the Onset of Benign Essential Blepharospasm and Hemifacial Spasm	United States of America	Dystonia BL (16) Controls (23)	All Not reported	Dystonia Neuro-Ophthalmology Clinic Controls Ophthalmology Clinic	Dystonia 69.3 ± 11.5 Controls 69.0 ± 9	Dystonia 4/12 Controls 10/13	Dystonia Previous Botox Controls Not reported	All BDI-II
Lauterbach et al. (2004) Differential DSM-III Psychiatric Disorder Prevalence Profiles in Dystonia and Parkinson's Disease	United States of America	Dystonia segmental (CD 18 & non-CD 3), generalised (7) Controls Sex, race & age-matched (3528)	Dystonia Neurological & movement disorder exam Controls Not reported	Dystonia Dystonia support group in movement disorder centre Controls National Institute of Health ECA study	Dystonia 51.32 ± 9.76 Controls: Not reported	Dystonia 8/20 Controls Matched	Dystonia 5 on anticholinergics 15 on GABA agonists Controls Not reported	All DIS

Author, Date & Title	Country of Origin	Groups (*n*)	Inclusion/ Exclusion Criteria	Recruitment (procedure/ site)	Age (mean ± SD)	Sex (m/f)	Treatment/ Medication	Measure(s) of Mental Health
Lehn et al. (2014) Psychiatric Disorders in Idiopathic-Isolated Focal Dystonia	Australia	Dystonia Idiopathic-isolated focal (CD 83, BL 12, MS 3, CCD 5) Controls Not reported (93)	Dystonia Not reported Controls No neurological abnormalities	Dystonia Neurology clinic Controls Community project	Dystonia Range 37-89 Controls Not significantly different	Dystonia 34/69 Controls Not significantly different	Dystonia All Botox 25% antidepressants or anxiolytic Controls 9% antidepressants or anxiolytic	All Y-BOCS
Lencer et al. (2009) Primary Focal Dystonia: Evidence for Distinct Neuropsychiatric and Personality Profiles	Germany	Dystonia Primary focal (CD 70, BL 16). 57 for some analyses. Controls Age & gender matched for some analyses (3943)	Dystonia No secondary dystonia Controls Not reported	Dystonia Dystonia clinics Controls Registration office files	Dystonia Men: 50.0 Women: 57.9 (participants <65 years removed for some analyses) Controls 18-64 years	Dystonia 23/63 (unreported for some analyses) Controls 2001/1942	Dystonia All Botox Controls Not reported	Dystonia SCID Controls M-CIDI

Author, Date & Title	Country of Origin	Groups (*n*)	Inclusion/ Exclusion Criteria	Recruitment (procedure/ site)	Age (mean ± SD)	Sex (m/f)	Treatment/ Medication	Measure(s) of Mental Health
Mula et al. (2012) Obsessive-Compulsive-Spectrum Symptoms in Patients with Focal Dystonia, Hemifacial Spasm, and Healthy Subjects	Italy	Dystonia Focal (CD 19, BL 8) Controls Age & gender matched (23)	All No <18 years, reading level <6th grade, learning disabilities, or MMSE score <24	Dystonia Neurology Botox Clinic Controls Hospital records	Dystonia 61.4 ± 13.7 Controls 59.9 ± 9.6	Dystonia 1/18 Controls 3/20	Dystonia All Botox Controls Not reported	All SCID
Paus et al. (2011) Impaired Sleep Quality and Restless Legs Syndrome in Idiopathic Focal Dystonia: A Controlled Study	Germany	Dystonia Mostly isolated focal dystonia (CD 111, BL 110) Controls Not reported (93)	Dystonia No onset before adulthood, generalised dystonia, or psychosis All No other neurological disorders	Dystonia Movement disorders & Botox clinics Controls Relatives of authors & patients	Dystonia CD: 59.8 ± 11.5 BL: 66.2 ± 11.2 Controls 57.4 ± 12.2	Dystonia CD: 32/68% BL: 32/68% Controls 49/51%	Dystonia 92% Botox 24% (CD) & 5% (BL) analgesics or muscle relaxants All 8% antidepressants	All BDI

Author, Date & Title	Country of Origin	Groups (*n*)	Inclusion/ Exclusion Criteria	Recruitment (procedure/ site)	Age (mean ± SD)	Sex (m/f)	Treatment/ Medication	Measure(s) of Mental Health
Setthawatcharawanich et al. (2011) Factors Associated With Quality of Life in Hemifacial Spasm and Blepharospasm During Long-Term Treatment With Botulinum Toxin	Thailand	Dystonia BL (32) Controls (32)	Dystonia No other chronic debilitating illnesses or movement disorders Controls No neurological disease, chronic illness or medication use	Dystonia Botox clinic Controls Not reported	Dystonia 63 ± 12.1 Controls 63 ± 11.6	Dystonia 8/24 Controls 8/24	Dystonia All Botox Controls No medications	All TDI
Smit, Kuiper, et al. (2016) Psychiatric Co-Morbidity is Highly Prevalent in Idiopathic CD and Significantly Influences Health-Related Quality of Life: Results of a Controlled Study	The Netherlands	Dystonia Idiopathic CD (50) Controls Age & sex matched (50)	Dystonia Adult onset Controls No family history of dystonia All No neurological co-morbidity or antidepressants	Dystonia Outpatient clinics & the dystonia patient association Controls Advertisement or friends of patients & investigators	Dystonia 54 (range 20-80) Controls 54 (range 24-83)	All NR	Dystonia Some Botox Controls Not reported	All MINI+ BDI BAI

Author, Date & Title	Country of Origin	Groups (*n*)	Inclusion/ Exclusion Criteria	Recruitment (procedure/ site)	Age (mean ± SD)	Sex (m/f)	Treatment/ Medication	Measure(s) of Mental Health
van Tricht et al. (2012) Cognition and Psychopathology in Myoclonus-Dystonia	The Netherlands	Dystonia Myoclonus (DYT11 31, non-DYT11 20) Controls Not reported (36)	Dystonia No secondary dystonia or myoclonus All Age >18 years, no stereotactic surgery or neurological comorbidity	Dystonia Movement disorder clinic Controls Family members & married spouses	Dystonia MD11: 45.9 ± 13.9 MD: 40.0 ± 15.8 Controls 36.7 ± 12.3	Dystonia MD11: 55/45% MD: 15/85% Controls 25/75%	Dystonia 4 Botox 21 no medication Others on trihexyphenidyl, clonazepam or anti-depressants Controls 2 on antidepressants 30 no medication	Dystonia SCID-I Controls MINI+
Yang et al. (2016) Nonmotor Symptoms in Primary Adult-Onset CD and BL	China	Dystonia Focal (CD 60 & BL 60) Controls Matched for age, sex, & education years (60)	Dystonia No secondary dystonia, other neurological abnormalities, or family history of movement disorders Controls Not reported	Dystonia Department of Neurology Controls Not reported	Dystonia CD: 40.23 ± 13.30 BL: 54.94 ± 11.51 Controls 47.87 ± 14.18	Dystonia CD: 25/35 BL: 22/38 Controls 30/30	Dystonia 38 receiving treatment (e.g., trihexyphenidyl or clonazepam) 28 had previous treatment (e.g., Botox) Controls Not reported	All HAM-D HAM-A

Note: AD arm dystonia. BL blepharospasm. CCD cranio-cervical dystonia. CD cervical dystonia. DYT11 myoclonic dystonia-11. LD laryngeal dystonia. MD dystonia myoclonus. MD11 myoclonic dystonia-11. MMSE Mini-Mental State Exam. MS Meige Syndrome. NC non-carrier. NMC non-manifesting mutation carrier. RDP rapid-onset dystonia-parkinsonism. WC writer's cramp.

Measure(s) of Mental Health

Studies either used a diagnostic interview (Barahona-Corrêa et al., 2011; Brashear et al., 2012; Fabbrini et al., 2010; Lauterbach et al., 2004; Lencer et al., 2009; Mula et al., 2012; van Tricht et al., 2012; Yang et al., 2016), a self-report measure (Johnson et al., 2007; Lehn et al., 2014; Paus et al., 2011; Setthawatcharawanich et al., 2011) or a combination of the two (Eichenseer et al., 2014; Smit, Kuiper, et al., 2016). See Table 5 for a description of each measure of mental health used in the included studies.

In total, eight out of 14 studies used diagnostic interviews such as the SCID and MINI+, which assess a large range of mental health difficulties (Barahona-Corrêa et al., 2011; Brashear et al., 2012; Fabbrini et al., 2010; Lauterbach et al., 2004; Lencer et al., 2009; Mula et al., 2012; Smit, Kuiper, et al., 2016; van Tricht et al., 2012). Three studies used diagnostic interviews for specific mental health problems; OCD (Y-BOCS, Barahona-Corrêa et al., 2011), depression (HAM-D, Yang et al., 2016) or anxiety (HAM-A, Eichenseer et al., 2014; Yang et al., 2016). Of the six studies using self-report measures, five used a measure of depression (BDI, BDI-II or TDI, Eichenseer et al., 2014; Johnson et al., 2007; Paus et al., 2011; Setthawatcharawanich et al., 2011; Smit, Kuiper, et al., 2016), one used a measure of anxiety (BAI, Smit, Kuiper, et al., 2016), and one used a measure of OCD (Y-BOCS-SR, Lehn et al., 2014).

Table 5: *Descriptions of Measures Used in Included Studies*

Measure of Mental Health	Abbreviation	Summary	Studies
Beck Anxiety Inventory	BAI	21-item self-report measure of anxiety Scores >10: above clinical cut-off for anxiety	Smit, Kuiper, et al. (2016)
Beck Depression Inventory	BDI	21-item self-report measure of depression Scores >10: above clinical cut-off for depression	Paus et al. (2011) Smit, Kuiper, et al. (2016)
Beck Depression Inventory-II	BDI-II	21-item self-report measure of depression Scoring: >14: above clinical cut-off for depression (Eichenseer et al., 2014) >10: above clinical cut-off for depression (Johnson et al., 2007)	Eichenseer et al. (2014) Johnson et al. (2007)
Diagnostic Interview Schedule	DIS	Structured psychiatric interview that non-clinicians can use to diagnose patients as accurately as trained clinicians using criteria from DSM-III (Diagnostic and Statistical Manual of Mental Disorders, third edition)	Lauterbach et al. (2004)
Hamilton Anxiety Scale	HAM-A	14 item clinician-rated interview measuring anxiety Scores >14: above clinical cut-off for anxiety	Eichenseer et al. (2014) Yang et al. (2016)
Hamilton Depression Scale	HAM-D	17 item clinician-rated interview measuring depression Scores >20: above clinical cut-off for depression	Yang et al. (2016)

Measure of Mental Health	Abbreviation	Summary	Studies
Munich Composite International Diagnostic Interview	M-CIDI	Interview using computerised scoring to assess DSM-IV symptoms, syndromes, and diagnoses	Lencer et al. (2009)
Mini-International Neuropsychiatric Interview (-PLUS)	MINI MINI+	Short structured diagnostic interview for DSM-IV and ICD-10 psychiatric disorders	Barahona-Corrêa et al. (2011) Smit, Kuiper, et al. (2016) van Tricht et al. (2012)
Structured Clinical Interview for DSM-IV Axis I Disorders	SCID-I	Semi-structured interview to make a diagnosis based on the DSM-IV (Diagnostic and Statistical Manual of Mental Disorders, fourth edition)	Brashear et al. (2012) Fabbrini et al. (2010) Lencer et al. (2009) Mula et al. (2012) van Tricht et al. (2012)
Thai Depression Inventory	TDI	20 item self-report measure of depression Scores >21: above clinical cut-off for depression	Setthawatcharawanich et al. (2011)
Yale-Brown Obsessive-Compulsive Symptom Checklist and Yale-Brown Obsessive-Compulsive Scale	Y-BOCS	Semi-structured interview to assess for obsessive–compulsive symptoms Scores >16: above clinical cut-off for OCD	Barahona-Corrêa et al. (2011)
Yale-Brown Obsessive-Compulsive Scale (self-report version)	Y-BOCS-SR	10 item self-report measure of obsessive-compulsive symptoms Scores >16: above clinical cut-off for OCD	Lehn et al. (2014)

Measures and Results

Study measures, outcomes (the numbers and percentages of participants who scored above clinical cut-off on mental health measures), statistics and odds ratios are shown in Table 6. For ease of interpretation, this review categorised data into mood disorders, anxiety disorders, other mental health problems, and overall levels of psychopathology (presence of any mental health problem).

Several studies used a mental health measure to explore the influence of a mental health difficulty on another factor (e.g., quality of life; Smit, Kuiper, et al., 2016); for these studies, numbers of participants with mental health problems in each group were not compared statistically (e.g., Lencer et al., 2009). For several studies, it was unclear which statistical test was used for comparisons (Mula et al., 2012; Setthawatcharawanich et al., 2011; Smit, Kuiper, et al., 2016; van Tricht et al., 2012). Some studies reported the number of participants with each diagnosis, but did not specify overall how many participants had mental health difficulties (Brashear et al., 2012; Eichenseer et al., 2014; Lauterbach et al., 2004; Yang et al., 2016), making it unclear if any participants had more than one diagnosis, thus potentially overestimating the number of participants in the sample with mental health problems.

Results of Included Studies

Mood Disorders

Six out of seven studies found that adults with dystonia were significantly more likely to have depression than controls (Lauterbach et al., 2004; Paus et al., 2011; Setthawatcharawanich et al., 2011; Smit, Kuiper, et al., 2016; van Tricht et al., 2012; Yang et al., 2016; all $p < .05$). Barahona-Corrêa et al. (2011) found no significant difference between the two groups in terms of numbers of adults with depression. For odds ratios, one study reported no association between depression and the presence of dystonia (Johnson et al., 2007) potentially due to modest numbers of participants, five studies reported a moderate association (Barahona-Corrêa et al., 2011; Brashear et al., 2012; Lencer et al., 2009; Paus et al., 2011; Smit, Kuiper, et al., 2016, MINI+), three studies reported a strong association (Lauterbach et al., 2004; Setthawatcharawanich et al., 2011; Smit, Kuiper, et al., 2016, BDI) and two studies reported an infinite association (Eichenseer et al., 2014; van Tricht et al., 2012). Overall, the results suggest that adults with dystonia are significantly more likely to have depression than adults without dystonia.

Two studies compared the incidence of dysthymic disorder between participants with dystonia and controls, and found no significant difference (Barahona-Corrêa et al., 2011; Smit, Kuiper, et al., 2016). Lencer et al. (2009) found an odds ratio of 3.08 (95% CI: 1.31-7.23), suggesting the odds of having dysthymic disorder for people with dystonia was three times that of controls. Lauterbach et al. (2004) found a significant difference between adults with dystonia and controls for

both bipolar affective disorder (p< .001) and atypical bipolar disorder (p< .001), with infinite associations. Brashear et al. (2012) found a strong association between bipolar affective disorder and the presence of dystonia.

Three studies statistically compared the dystonia and control groups on presence of a mood disorder (combining the results from separate mood disorder diagnostic interviews). Two studies found that adults with dystonia were significantly more likely to have a mood disorder than controls, with strong associations (Brashear et al., 2012; Smit, Kuiper, et al., 2016; both p< .03). Fabbrini et al. (2010) compared four different dystonia groups with four control groups, and found that participants with cervical dystonia and blepharospasm were significantly more likely to have a mood disorder than controls (strong and infinite associations respectively; both p< .01), whereas the results for arm dystonia and laryngeal dystonia were not significant. Overall, the results suggest that adults with dystonia are significantly more likely to have a mood disorder than controls.

Anxiety Disorders

Both studies comparing the groups on anxiety severity measures found that significantly more dystonia participants scored above the cut-off for anxiety than controls (Smit, Kuiper, et al., 2016; Yang et al., 2016; both p< .001). Odds ratios indicated a strong association between anxiety severity and dystonia (Eichenseer et al., 2014; Lencer et al., 2009; Smit, Kuiper, et al., 2016), with one indicating an infinite association (Yang et al., 2016, OR 27.13 [95% CI: 6.07-121.31]).

Two out of four studies found that adults with dystonia were significantly more likely to have generalised anxiety disorder (GAD) than controls (Lauterbach et al., 2004 and Smit, Kuiper, et al., 2016, both $p < .05$; Barahona-Corrêa et al., 2011 and van Tricht et al., 2012, not significant). Odds ratios indicated no (Barahona-Corrêa et al., 2011), and strong (Lauterbach et al., 2004, OR 4.25 [95% CI: 1.77-10.22]; Lencer et al., 2009, OR 4.89 [95% CI: 1.46-16.36]) associations between adults with dystonia and a diagnosis of GAD. One study found that controls had more GAD diagnoses than adults with dystonia (Brashear et al., 2012, OR 0.65 [95% CI: 0.14-3.04]), however statistical significance was not calculated.

One out of five studies found that adults with dystonia were significantly more likely to have OCD than controls (Lehn et al., 2014, $p < .01$). A further study found 26% of dystonia participants to meet criteria for OCD, compared to no controls (Mula et al., 2012). Moderate and strong associations between OCD and the presence of dystonia were found (Brashear et al., 2012, OR 2.42 [95% CI: 0.21-28.31]; Lencer et al., 2009, OR 8.4 [95% CI: 1.7-38.9]). Conversely, one study found a weak association between OCD and the absence of dystonia (van Tricht et al., 2012, OR 0.7 [95% CI: 0.04-11.57]).

For post-traumatic stress disorder (PTSD), there were no significant differences between groups, with more controls meeting criteria for PTSD in two (Barahona-Corrêa et al., 2011; Lencer et al., 2009) out of three studies. For adults with dystonia, the odds of having PTSD was 2.55 [95% CI: 0.43-15.2] that of controls in one study (Brashear et al., 2012).

Lauterbach et al. (2004) reported that adults with dystonia were 3.76 times [95% CI: 1.75-18.07] more likely to have any phobic disorder (combining the results of separate phobia diagnostic interviews) than controls. Two studies on simple phobia found conflicting results; one found dystonia and control groups to be significantly different (Smit, Kuiper, et al., 2016, p=.05); and one found no significant difference (van Tricht et al., 2012). Moderate (Lauterbach et al., 2004; Lencer et al., 2009; Smit, Kuiper, et al., 2016) and strong (van Tricht et al., 2012, OR 3.8 [95% CI: 0.43-34.05]) associations were found between simple/specific phobia and presence of dystonia.

Three out of four studies found that adults with dystonia were significantly more likely to have social anxiety than controls (Lauterbach et al., 2004, Smit, Kuiper, et al., 2016 and van Tricht et al., 2012, all p< .05; Barahona-Corrêa et al., 2011, not significant). Two studies had strong associations between social anxiety and presence of dystonia (Lauterbach et al., 2004; van Tricht et al., 2012) and one had an infinite association (Lencer et al., 2009, OR 21.6 [95% CI: 11.1-41.9]). One study had a weak association between social anxiety and the absence of dystonia (Brashear et al., 2012, OR 0.85 [95% CI: 0.17-4.19]).

One out of three studies found that adults with dystonia were significantly more likely to have agoraphobia than controls (Smit, Kuiper, et al., 2016, p< .01; Barahona-Corrêa et al., 2011 and van Tricht et al., 2012, not significant). Two studies found a moderate association (Barahona-Corrêa et al., 2011; Lauterbach et al., 2004), and two found an infinite association (Lencer et al., 2009; Smit, Kuiper, et al., 2016) between agoraphobia and presence of dystonia. One out of three studies

found a significant difference between dystonia and control groups for panic disorder (van Tricht et al., 2012, $p < .05$; Barahona-Corrêa et al., 2011 and Smit, Kuiper, et al., 2016, not significant). Moderate (Lauterbach et al., 2004), strong (Smit, Kuiper, et al., 2016), infinite (Lencer et al., 2009), and no (Brashear et al., 2012) associations between panic disorder and presence of dystonia were found.

Four studies statistically compared the dystonia and control groups on presence of any anxiety disorder (combining the results from separate anxiety diagnostic interviews). Two studies found that adults with dystonia were significantly more likely to have an anxiety disorder than controls, with strong associations (Smit, Kuiper, et al., 2016; van Tricht et al., 2012; both $p < .01$), and two studies found no significant difference between groups, despite weak and moderate associations (Brashear et al., 2012; Fabbrini et al., 2010). An additional study found a strong association between any anxiety disorder and presence of dystonia (Lencer et al., 2009, OR 3.4 [95% CI: 2.18-5.31]).

Other Mental Health Problems

Various mental health problems that are not specific mood or anxiety disorders were reported by the studies. Two studies reported that alcohol abuse was more common among adults with dystonia than controls (Lauterbach et al., 2004, OR 4.11 [95% CI: 1.22-13.85]; Lencer et al., 2009, OR 9.6 [95% CI: 4.5-20.3], however another study found this relationship to not be significant (Barahona-Corrêa et al., 2011). Alcohol dependence was found to not be significantly increased in adults with

dystonia in comparison to controls (van Tricht et al., 2012), with weak associations found between alcohol dependence and presence (Lencer et al., 2009) and absence (Lauterbach et al., 2004) of dystonia. Barahona-Corrêa et al. (2011) found that no participants met criteria for heroin abuse. One study found that adults with dystonia were significantly more likely to score above clinical cut-off for substance abuse or dependence (p=.04), with a moderate association (Brashear et al., 2012).

Two studies exploring the relationship between psychosis and dystonia had conflicting results. One study found that adults with dystonia were significantly more likely than controls to have any psychotic disorder (Brashear et al., 2012, p=.02), whilst another study found no significant difference between the groups for psychotic syndrome (Barahona-Corrêa et al., 2011).

No significant difference was found between groups for combined anxiety and depression (Smit, Kuiper, et al., 2016), adjustment disorder (Fabbrini et al., 2010), or somatization disorder (Barahona-Corrêa et al., 2011). Lencer et al. (2009) found an infinite association between bulimia nervosa and presence of dystonia (OR 23.22 [95% CI: 2.09-258.52]).

Total Psychopathology

Three out of four studies found that adults with dystonia were significantly more likely to have any mental health problem than controls, with strong associations (Fabbrini et al., 2010, Smit, Kuiper, et al., 2016 and van Tricht et al., 2012, all p< .05; Barahona-Corrêa et al., 2011, not significant). In addition, Lencer et

al. (2009) found adults with dystonia were 4.5 times more likely to have an Axis 1 mental health problem (i.e., a psychological diagnosis that is not a personality disorder or intellectual disability) than controls.

Table 6: *Results of Included Studies*

Author & Date	**Measure**	**Statistical Test**	**Summary Outcome Data (*n* (%))**	**Reported Significance Level**	**Odds Ratios & Confidence Intervals**
Barahona-Corrêa et al. (2011)	Y-BOCS	Fisher's exact test	Y-BOCS		
			>16: Dystonia 11 (24.4%); Controls 0	NR	NA
	MINI & some MINI+		MINI+		
			No psychopathology: Dystonia 18 (40%); Controls 17 (56.6%)	**NS**	**0.51 (0.2-1.3)**
			Major depression: Dystonia 2 (4.4%); Controls 2 (6.7%)	NS	0.65 (0.09-4.98)
			Dysthymia: Dystonia 0; Controls 1 (3.35%)	NS	NA
			GAD: Dystonia 7 (15.6%); Controls 5 (16.7%)	NS	0.92 (0.26-3.23)
			Agoraphobia (current/lifetime): Dystonia 4 (8.9%); Controls 1 (3.3%)	*p*=.09	2.83 (0.3-26.64)
			Panic disorder (± agoraphobia): Dystonia 1 (2.2%); Controls 0	NS	NA
			Social phobia: Dystonia 1 (2.2%); Controls 0	NS	NA
			PTSD (current/lifetime): Dystonia 1 (2.2%); Controls 1 (3.3%)	NS	0.66 (0.04-10.96)
			OCD: Dystonia 3 (6.7%); Controls 0	*p*=.06	NA
			Psychotic syndrome (current/lifetime): Dystonia 2 (4.4%); Controls 0	NS	NA
			Somatization disorder: Dystonia 0; Controls 1 (3.3%)	NS	NA
			Heroin abuse (in remission): Dystonia 0; Controls 0	NS	NA
			Alcohol abuse (in remission): Dystonia 2 (4.4%); Controls 0	NS	NA

Author & Date	Measure	Statistical Test	Summary Outcome Data (*n* (%))	Reported Significance Level	Odds Ratios & Confidence Intervals
Brashear et al. (2012)	SCID-I	Fisher's exact test	Any anxiety disorder: MMC 12 (48%); NMC 0; NC 11 (41%)	*p*=.16	1.48 (0.51-4.32)
			GAD: MMC 3 (12%); NMC 0; NC 5 (19%)	NR	0.65 (0.14-3.04)
			PTSD: MMC 4 (17%); NMC 0; NC 2 (7%)	NR	2.55 (0.43-15.2)
			OCD: MMC 2 (8%); NMC 0; NC 1 (4%)	NR	2.42 (0.21-28.31)
			Panic disorder: MMC 5 (19%); NMC 0; NC 5 (19%)	NR	1.19 (.30-4.68)
			Social phobia: MMC 3 (12%); NMC 0; NC 4 (15%)	NR	.85 (.17-4.19)
			Any mood disorder: MMC 13 (50%); NMC 0; NC 6 (22%)	*p*=.03	4 (1.23-13.01)
			Depression: MMC 9 (35%); NMC 0; NC 5 (19%)	NR	2.65 (.75-9.28)
			Bipolar disorder: MMC 4 (15%); NMC 0; NC 1 (4%)	NR	5.27 (.55-50.55)
			Any psychotic disorder: MMC 5 (19%); NMC 0; NC 0	*p*=.02	NA
			Psychosis NOS: MMC 3 (12%); NMC 0; NC 0	NR	NA
			Brief psychotic disorder: MMC 2 (8%); NMC 0; NC 0	NR	NA
			Any substance abuse/dependence: MMC 10 (38%); NMC 0; NC 7 (27%)	*p*=.04	2.05 (.65-6.54)
			Alcohol: MMC 9 (35%); NMC 0; NC 7 (27%)	NR	1.74 (.54-5.6)
Eichenseer et al. (2014)	BDI-II	Not statistically compared	BDI-II		
			>14: CD 14 (26%); Controls 1 (2%)	NR	18.9 (2.39-149.72)
	HAM-A		HAM-A		
			>18: CD 5 (9%); Controls 1 (2%)	NR	5.51 (0.62-48.82)

Author & Date	Measure	Statistical Test	Summary Outcome Data (*n* (%))	Reported Significance Level	Odds Ratios & Confidence Intervals
Fabbrini et al. (2010)	SCID-I	χ^2 test	Mood		
			CD 9 (26.4%); Controls 2 (6%)	*p*< .01	5.4 (1.07-27.33)
			BL 11 (39.3%); Controls 1 (4%)	*p*< .01	14.24 (1.67-121.32)
			LD 2 (12.5%); Controls 2 (16.6%)	NS	0.71 (0.09-5.96)
			AD 0; Controls 0	NS	NA
			Anxiety		
			CD 9 (26.4%); Controls 5 (15.6%)	NS	1.94 (0.57-6.59)
			BL 7 (25.0%); Controls 3 (11.5%)	NS	2.22 (0.5-9.81)
			LD 3 (18.7%); Controls 2 (16.6%)	NS	1.15 (0.16-8.27)
			AD 0; Controls 0	NS	NA
			OCD		
			CD 1 (3%); Controls 0	NS	NA
			BL 1; Controls 0	NS	NA
			LD 0; Controls 0	NS	NA
			AD 1 (9%); Controls 0	NS	NA
			Adjustment		
			CD 4 (11.7%); Controls 1 (3%)	NS	4.13 (0.44-39.14)
			BL 0; Controls 2 (7.6%)	NS	NA
			LD 3 (18.7%); Controls 1 (10%)	NS	2.54 (0.23-28.02)
			AD 1 (9%); Controls 0	NS	NA
			Total diagnosis		
			CD 23 (67.6%); Controls 8 (25%)	*p*< .01	6.27 (2.14-18.39)
			BL 19 (67.8%); Controls 9 (34.6%)	*p*< .01	3.28 (1.04-10.41)
			LD 8 (50%); Controls 5 (41.6%)	NS	1.4 (0.31-6.33)
			AD 2 (18%); Controls 0	NS	NA
			All dystonia 51 (57.3%); All controls 15 (24.1%)	***p*< .01**	**5.55 (2.75-11.21)**

Author & Date	Measure	Statistical Test	Summary Outcome Data (*n* (%))	Reported Significance Level	Odds Ratios & Confidence Intervals
Johnson et al. (2007)	BDI-II	Not statistically compared	>10: BL 8 (50%); Controls 11 (48%)	NR	1.09 (.30-3.91)
Lauterbach et al. (2004)	DIS	Odds ratios & Fisher's exact test	Major depression: Dystonia 7 (25%); Controls 195 (5.53%)	*p*< .001	5.697
			Dysthymic disorder: Dystonia 3 (10.71%); Controls 138 (3.91%)	NR	2.95
			Bipolar: Dystonia 2 (7.14%); Controls 19 (0.54%)	*p*< .001	14.21
			Atypical bipolar disorder: Dystonia 2 (7.14%); Controls 6 (0.18%)	*p*< .001	27.06
			Any phobic disorder: Dystonia 11 (39.29%); Controls 518 (14.67%)	NR	3.76
			Agoraphobia: Dystonia 3 (10.71%); Controls 237 (6.72%)	NR	1.67
			Social Phobia: Dystonia 5 (17.86%); Controls 99 (2.81%)	*p*< .001	7.53
			Simple Phobia: Dystonia 7 (25%); Controls 442 (12.53%)	NR	3.33
			Panic disorder: Dystonia 1 (3.57%); Controls 71 (1.98%)	NR	1.81
			GAD: Dystonia 7 (25%); Controls 108 (7.28%)	*p*< .001	4.25
			OCD: Dystonia 0; Controls 92 (2.60%)	NA	NA
			Alcohol abuse: Dystonia 3 (10.71%); Controls 100 (5.68%)	*p*< .05 NS	4.11
			Alcohol dependence: Dystonia 2 (7.14%); Controls 293 (8.31%)	NR	0.849
Lehn et al. (2014)	Y-BOCS	Fisher's exact test	>16: Dystonia: 8 (7.8%); Controls 0	*p*< .001	NA

Author & Date	Measure	Statistical Test	Summary Outcome Data (*n* (%))	Reported Significance Level	Odds Ratios & Confidence Intervals
Lencer et al. (2009)	SCID (Dystonia) M-CIDI (Controls)	Odds Ratios & Breslow Day Test		**OR- matched sample**	**OR- whole sample**
			Axis I: Dystonia 53 (61.6%); Controls 1383 (35.1%)	**4.5**	**2.98 (1.92-4.62)**
			Anxiety: Dystonia 32 (37.2%); Controls 586 (14.9%)	NR	3.4 (2.18-5.31)
			Social phobia: Dystonia 20 (23.3%); Controls 72 (1.8%)	21.6 (11.1-41.9)	16.09 (9.27-27.93)
			Specific phobia: Dystonia 14 (16.3%); Controls 411 (10.4%)	NR	1.67 (0.94-2.99)
			Agoraphobia disorder: Dystonia 9 (10.5%); Controls 41 (1.0%)	16.7 (7.4-37.3)	11.14 (5.23-23.73)
			Panic disorder: Dystonia 7 (8.1%); Controls 36 (0.9%)	11.5 (4.5-29.2)	9.63 (4.16-22.3)
			GAD: Dystonia 3 (3.5%); Controls 29 (0.7%)	NR	4.89 (1.46-16.36)
			OCD: Dystonia 2 (2.3%); Controls 20 (0.5%)	8.4 (1.7-38.9)	4.68 (1.08-20.33)
			Major depression: Dystonia 23 (26.7%); Controls 446 (11.3%)	3.0 (1.7-5.5)	2.87 (1.76-4.67)
			Dysthymic Disorder: Dystonia 6 (7.0%); Controls 94 (2.4%)	NR	3.08 (1.31-7.23)
			Alcohol abuse: Dystonia 13 (15.1%); Controls 178 (4.5%)	9.6 (4.5-20.3)	3.77 (2.05-6.94)
			Alcohol dependence: Dystonia 4 (4.7%); Controls 149 (3.8%)	NR	1.24 (0.45-3.44)
			Drug abuse: Dystonia 2 (2.3%); Controls 30 (0.8%)	NR	3.11 (0.73-13.23)
			Drug dependence: Dystonia 2 (2.3%); Controls 14 (0.4%)	14.9 (3.1-70.7)	6.69 (1.5-29.91)
			Bulimia nervosa: Dystonia 1 (1.2%); Controls 2 (0.1%)	NR	23.22 (2.09-258.52)
			PTSD: Dystonia 1 (1.2%); Controls 54 (1.4%)	NR	0.85 (0.12-6.2)
Mula et al. (2012)	SCID	χ^2 or Fisher's exact test	OCD: Dystonia 5 (26%); Control 0	NR	NR
Paus et al. (2011)	BDI	χ^2 test	>10: CD 25%; BL 27%; Control 10%	$p < .01$	2.55 (1.24-5.28)

Author & Date	Measure	Statistical Test	Summary Outcome Data (*n* (%))	Reported Significance Level	Odds Ratios & Confidence Intervals
Setthawatcharawanich et al. (2011)	TDI	Mann-Whitney U or Student's *t* test	>21: BL 10 (31.3%); Control 3 (9.4%)	*p*=.03	4.39 (1.08-17.89)
Smit, Kuiper, et al. (2016)	MINI+	χ^2 test or Fisher's Exact Test	MINI+		
			Depressive episode: Dystonia 16 (32%); Control 7 (14%)	*p*=.03	2.89 (1.07-7.82)
			Dysthymia: Dystonia 4 (8%); Control 0	*p*=.11	NA
			Combined anxiety/depression: Dystonia 1 (2%); Control 0	*p*=1	NA
			Mood total: Dystonia 19 (38%); Control 7 (14%)	*p*< .01	3.76 (1.41-10.05)
			Panic disorder: Dystonia 5 (10%); Control 1 (2%)	*p*=.20	5.44 (0.61-48.4)
			Agoraphobia: Dystonia 12 (24%); Control 1 (2%)	*p*< .01	15.47 (1.93-124.3)
			Social phobia: Dystonia 9 (18%); Control 0	*p*< .01	NA
			Simple phobia: Dystonia 8 (16%); Control 2 (4%)	*p*=.05	4.57 (0.92-22.73)
			GAD: Dystonia 6 (12%); Control 0	*p*=.03	NA
			Anxiety total: Dystonia 21 (42%); Control 4 (8%)	*p*< .001	8.33 (2.6-26.72)
			Total with diagnosis: Dystonia 32 (64%); Control 14 (28%)	***p*< .01**	**4.57 (1.96-10.65)**
	BDI		BDI		
			>10: Dystonia 23 (46%); Control 6 (12%)	*p*< .001	6.25 (2.26-17.69)
	BAI		BAI		
			> 10: Dystonia 20 (40%); Control 4 (8%)	*p*< .001	7.67 (2.38-24.65)

Author & Date	Measure	Statistical Test	Summary Outcome Data (*n* (%))	Reported Significance Level	Odds Ratios & Confidence Intervals
van Tricht et al. (2012)	SCID-I (Dystonia) MINI+ (Controls)	χ^2 test or Fisher's exact test	MDD: MD-11 10 (32%); MD 9 (45%); Control 2 (6%)	*p*< .001	10.09 (2.17-46.84)
			Any anxiety disorder: MD-11 16 (52%); MD 3 (15%); Control 4 (11%)	*p*< .01	4.75 (1.45-15.53)
			Social phobia: MD-11 8 (26%); MD 1 (5%); Control 2 (6%)	*p*=.02	3.64 (0.74-18)
			Panic disorder: MD-11 7 (23%); MD 3 (15%); Control 0	*p*< .05	NA
			Agoraphobia: MD-11 3 (10%); MD 2 (10%); Control 0	NS	NA
			Specific phobia: MD-11 5 (16%); MD 0; Control 1 (3%)	NS	3.8 (0.43-34.05)
			GAD: MD-11 3 (10%); MD 1 (5%); Control 0	NS	NA
			OCD: MD-11 1 (3%); MD 0; Control 1 (3%)	NS	0.7 (0.04-11.57)
			Alcohol dependence: MD-11 4 (13%); MD 2 (10%); Control 0	NS	NA
			Total MD-11 17 (55%); MD 10 (50%); Control 5 (14%)	***p*< .05**	**6.98 (2.34-20.81)**
Yang et al. (2016)	HAM-D	χ^2 test	HAM-D		
			CD 12 (20%); BL 8 (13.3%); Controls 0	*p*< .001	NA
	HAM-A		HAM-A		
			CD 17 (28.3%); BL 12 (20%); Controls 2 (3.3%)	*p*< .001	27.13 (6.07-121.31)

Note: AD arm dystonia. BL blepharospasm. CD cervical dystonia. LD laryngeal dystonia. MD-11 myoclonic dystonia DYT11. MD myoclonic dystonia. MDD major depressive disorder. MMC manifesting mutation carrier. NA not applicable. NC non-carrier. NMC non-manifesting mutation carrier. NR not reported. NS not significant. OR odds ratio.

Quality Analysis

The quality of studies was assessed using the Newcastle-Ottawa scale (Wells et al., 2013). For these 14 studies (Table 7), five achieved a 5-star rating (Barahona-Corrêa et al., 2011; Lencer et al., 2009; Mula et al., 2012; Setthawatcharawanich et al., 2011; Yang et al., 2016), four achieved a 4-star rating (Eichenseer et al., 2014; Fabbrini et al., 2010; Johnson et al., 2007; Paus et al., 2011), four achieved a 3-star rating (Brashear et al., 2012; Lauterbach et al., 2004; Lehn et al., 2014; Smit, Kuiper, et al., 2016), and one achieved a 1-star rating (van Tricht et al., 2012).

Studies achieved 0-3 stars in the Selection scale (out of 4). The most common score was 2 stars (8 studies, Brashear et al., 2012; Johnson et al., 2007; Lauterbach et al., 2004; Lehn et al., 2014; Mula et al., 2012; Paus et al., 2011; Setthawatcharawanich et al., 2011; Yang et al., 2016). Eleven studies used neurological examinations or hospital records to confirm dystonia diagnoses, whilst three studies relied on self-report (Fabbrini et al., 2010; Lehn et al., 2014; Smit, Kuiper, et al., 2016). Five studies had a dystonia sample believed to be representative of the general population with that type of dystonia (Barahona-Corrêa et al., 2011; Lauterbach et al., 2004; Paus et al., 2011; Setthawatcharawanich et al., 2011; Yang et al., 2016), the remaining nine dystonia samples were not generalisable (i.e., recruited from Botox treatment clinics or had strict exclusion criteria). Five studies recruited controls from the community, (Barahona-Corrêa et al., 2011; Brashear et al., 2012; Fabbrini et al., 2010; Lehn et al., 2014; Lencer et al., 2009), three from hospitals (Eichenseer et al., 2014; Johnson et al., 2007; Mula et al., 2012), and six did not specify recruitment procedures or used family and friends

(Lauterbach et al., 2004; Paus et al., 2011; Setthawatcharawanich et al., 2011; Smit, Kuiper, et al., 2016; van Tricht et al., 2012; Yang et al., 2016). Four studies included control participants with mental health problems (Johnson et al., 2007; Lehn et al., 2014; Lencer et al., 2009; Mula et al., 2012), the remaining ten did not specify whether they included control participants with mental health problems.

Studies achieved 0-2 stars in the Comparability scale. Seven studies matched participants based on age and sex, two studies matched with age only (Johnson et al., 2007; Paus et al., 2011), one matched with sex only (Lauterbach et al., 2004), and four studies did not use matching.

Studies achieved 0-2 stars in the Exposure scale (out of an available 3). Ten studies achieved 1-star, one study achieved 2 stars (Barahona-Corrêa et al., 2011), and two studies achieved no stars (Lauterbach et al., 2004; Lencer et al., 2009). Only one study (Barahona-Corrêa et al., 2011) used a diagnostic interview where the interviewer was blind to if the participant had dystonia, likely as this would be difficult to hide from an interviewer. The remaining studies used diagnostic interviews (Brashear et al., 2012; Eichenseer et al., 2014; Fabbrini et al., 2010; Lauterbach et al., 2004; Lencer et al., 2009; Mula et al., 2012; Smit, Kuiper, et al., 2016; van Tricht et al., 2012; Yang et al., 2016) or self-report measures (Johnson et al., 2007; Lehn et al., 2014; Paus et al., 2011; Setthawatcharawanich et al., 2011). Most studies used the same measure for all participants; two used population-based control samples which used different measures to the dystonia group (Lauterbach et al., 2004; Lencer et al., 2009). Most studies did not report the response rates for one or both groups.

Table 7: *Study Quality Ratings (Newcastle-Ottawa Scale; Wells et al., 2013)*

	Selection				Comparability		Exposure			Rating
	1	2	3	4	Age	Sex	1	2	3	
Barahona-Corrêa et al. (2011)	a*	a*	a*	b	n	n	b*	a*	c	5*
Brashear et al. (2012)	a*	b	a*	b	n	n	c	a*	c	3*
Eichenseer et al. (2014)	a*	b	b	b	y*	y*	c	a*	c	4*
Fabbrini et al. (2010)	b	b	a*	b	y*	y*	c	a*	c	4*
Johnson et al. (2007)	a*	b	b	a*	y*	n	d	a*	b	4*
Lauterbach et al. (2004)	a*	a*	c	b	n	y*	c	b	c	3*
Lehn et al. (2014)	b	b	a*	a*	n	n	d	a*	c	3*
Lencer et al. (2009)	a*	b	a*	a*	y*	y*	c	b	c	5*
Mula et al. (2012)	a*	b	b	a*	y*	y*	c	a*	c	5*
Paus et al. (2011)	a*	a*	c	b	y*	n	d	a*	c	4*
Setthawatcharawanich et al. (2011)	a*	a*	c	b	y*	y*	d	a*	c	5*
Smit, Kuiper, et al. (2016)	b	b	c	b	y*	y*	c	a*	c	3*
van Tricht et al. (2012)	a*	b	c	b	n	n	c	b	c	1*
Yang et al. (2016)	a*	a*	c	b	y*	y*	c	a*	c	5*

Discussion

The purpose of this review was to determine whether adults with dystonia are more likely to have mental health difficulties than adults without dystonia.

Systematically searching two online databases (PubMed and PsycINFO) according to inclusion and exclusion criteria resulted in 14 studies being reviewed. Information from these studies was extracted, data were presented on numbers of participants who had mental health problems, and studies were quality assessed.

Summary of Results

Mood Disorders

Most studies found that adults with dystonia were significantly more likely to have depression, with 4-50% meeting criteria, as compared to 0-48% controls. Only one study out of eleven did not find any significant differences (Barahona-Corrêa et al., 2011). Johnson et al. (2007) found similar rates of depression in both groups (50% dystonia, 48% controls), however this study had several limitations including a small sample size (*n*=16), the inclusion and exclusion criteria were not reported, and a self-report measure was used. Therefore, the levels of depression reported in this study may not be accurate. No other control group had more than 19% of participants meeting criteria for depression. Only two studies found rates of depression in adults with dystonia to be less than 25% (Barahona-Corrêa et al., 2011; Yang et al., 2016). Barahona-Corrêa et al. (2011) found controls to have higher rates of depression than dystonia participants (4.4% dystonia, 6.7% controls), however these results may have been influenced by the exclusion of antidepressants. A significant difference was found for one study looking at bipolar affective disorder (Lauterbach et al., 2004), whereas differences in dysthymia were not found to be

significantly different in two studies (Barahona-Corrêa et al., 2011; Smit, Kuiper, et al., 2016). This may be due to an insufficient sample size to detect a condition which is not common (Sansone & Sansone, 2009), or the exclusion of antidepressants. Overall, the results indicate that adults with dystonia are more likely to have mood difficulties (particularly depression) than adults without dystonia.

Anxiety Disorders

The results of anxiety disorders were mixed. Only two studies explored anxiety in general (using severity measures), both finding a significant difference between adults with dystonia and controls. Studies found that participants scored above clinical cut-off for anxiety in 9-40% of participants with dystonia as compared to 2-15% in controls. Only one study found the rate of anxiety in adults with dystonia to be less than 26% (Eichenseer et al., 2014). Three out of four studies found adults with dystonia to be significantly more likely to have social phobia; finding social phobia in 2-26% of participants as compared to 0-13% of controls. However, for GAD, OCD, PTSD, specific phobias, and agoraphobia, half or fewer studies found a significant difference between adults with dystonia and controls, with some studies finding more controls having mental health difficulties than dystonia participants. Most of these conditions were statistically compared by four or fewer studies; and considering the varied methodology between studies in this review, it is difficult to ascertain whether adults with dystonia are more likely to have these difficulties than adults without dystonia. As adults with dystonia were found to have an 8.4 increased

risk of having OCD than controls in one study with a large sample size (Lencer et al., 2009), some studies may have been underpowered to detect a difference.

Other Mental Health Problems

In terms of mental health problems not categorised as a mood or anxiety disorder, few studies explored these and reported statistical comparisons. Despite several studies showing associations between mental health difficulties and presence of dystonia, only two studies showed a significant difference between adults with dystonia and controls. Brashear et al. (2012) found that adults with dystonia were significantly more likely to have any psychotic disorder, or a substance abuse or dependence problem. Lencer et al. (2009) found that adults with dystonia were significantly more likely to have bulimia nervosa than controls; in this study only one person with dystonia was found to have bulimia (1.2%) in comparison to two in the population-based control group (0.1%). This highlights the difficulty in exploring specific mental health problems in studies with small sample sizes.

Total Psychopathology

All studies but one found that adults with dystonia were significantly more likely to have a mental health difficulty than controls. The rates of total mental health difficulties were 55-64% for adults with dystonia, and 14-37% in controls. The one study that did not show a significant difference had methodological qualities that may have influenced results (Barahona-Corrêa et al., 2011), such as strict

inclusion and exclusion criteria. Despite this, the results from this study showed mental health difficulties to be present in 60% of adults with dystonia.

Summary

Overall, results suggest that adults with dystonia are more likely than adults without dystonia to have mental health difficulties, particularly for depression, overall anxiety, and social anxiety. Calculated odds ratios indicated associations between some additional mental health difficulties (i.e., simple/specific phobia, agoraphobia and alcohol abuse) and presence of dystonia, however statistical significance was not explored.

It is important to note that, in comparison to other studies included in this review which found more non-significant results (Barahona-Corrêa et al., 2011; Fabbrini et al., 2010), studies which consistently provided significant results (Brashear et al., 2012; Lauterbach et al., 2004; Smit, Kuiper, et al., 2016; van Tricht et al., 2012) were found to be of poor methodological quality on the Newcastle-Ottawa scale. These studies tended not to report the selection of controls or match on both age and sex. In addition, both Lauterbach et al. (2004) and van Tricht et al. (2012) used different measures for dystonia cases and controls. Factors such as these calls the comparability of the groups into question and may have impacted the results. Despite this, studies of high methodological quality did report some significant results (Setthawatcharawanich et al., 2011; Yang et al., 2016).

Strengths and Limitations of Included Studies

For the studies included in this review, some consistently showed that adults with dystonia were significantly more likely to have mental health problems than adults without dystonia (e.g., Smit, Kuiper, et al., 2016), whereas other studies consistently found no significant difference (Barahona-Corrêa et al., 2011; Fabbrini et al., 2010). These differences may indicate that results were influenced by the studies methodologies, for example, Smit, Kuiper, et al. (2016) recruited from outpatient clinics whilst Barahona-Corrêa et al. (2011) recruited from the Dystonia Association case register. Recruiting from a hospital setting means that participants are more likely to be suffering from issues which impact mental health (such as pain), in comparison to those not seeking treatment for their dystonia. In addition, across all studies the measures differed in terms of those which are diagnostic (e.g., SCID-I) and those which are indicative (e.g., BDI-II) of a mental health problem.

One factor likely influencing results is sample size. A large sample size is often necessary to detect effects and calculate odds ratios which are reflective of the true population. This is especially the case for mental health difficulties that have a low prevalence; for example, bipolar affective disorder was found in 0.54% of a control group of 3528 participants (Lauterbach et al., 2004), meaning that a sample size of nearly 200 is needed to detect just one case. Fabbrini et al. (2010) separated their analyses of specific mental health problems into four different types of dystonia; this resulted in two of the groups having fewer than 20 participants. Significant differences were found only between the dystonia and control groups for those with larger sample sizes. This is reflected in how one control group had one person with a

mood disorder, which was 4% of that group, and another had two people which amounted to 16.6% of that group; such small sample sizes make it difficult to ascertain if differences found reflect true prevalence rates in the population.

Other methodological features require some consideration. Further sources of bias in these studies could be lack of blinding of which group each participant was in when completing diagnostic interviews, however as many adults with dystonia have visible movements, or need aids to walk or communicate, this would not have been possible without eliminating many potential participants and affecting the generalisability of results. It is possible that interviewers' questions and interpretations of answers were influenced by their knowledge of a person having or not having dystonia. Not all studies specified their recruitment strategy or response rate, making it unclear whether participants were selected randomly to take part. Not doing so may have introduced a source of bias into results. One further impact may be the matching of controls to cases. Whilst matching to age, sex, and other characteristics ensures that the control sample is not too different from the dystonia sample; it is argued that matching can introduce a source of bias, as it limits the variability in the control sample and makes this data less generalisable to the population in general (Rose & van der Laan, 2009). The studies in this review used a variety of matching techniques which makes it difficult to compare the control samples. Additionally, it is difficult to know what the ideal inclusion and exclusion criteria are; for example, excluding use of antidepressants could impact the numbers of people with mental health difficulties for either group and makes both samples

less representative, however including participants taking antidepressants may also introduce bias into results.

Strengths and Limitations of the Review

One strength of this review is that all journals meeting criteria were accessed in full to determine eligibility and extract the relevant data. In addition, there was no language bias influencing results, as studies in all languages were included. Conversely, only journals were accessed for this purpose of this review which may have introduced a source of bias due to significant results being more likely to be published. This may have resulted in the differences between the dystonia and control groups being overestimated (Dickersin, 1990).

The search criteria are considered sufficient to have found all relevant published studies, including as wide a range of relevant dystonia subtypes and mental health problems as possible. It was deemed important to be able to ascertain whether adults with dystonia are more likely to have mental health problems than adults without dystonia, however, there were relatively few studies which included a control group. This was surprising considering the large amount of literature on mental health in adult dystonia. The searching of reference lists of included studies, an additional database, or non-journal resources may have found additional material. Reporting additional data, such as the mean scores on measures of mental health, would also have yielded more information from a greater range of studies,

rather than limiting the extraction of data to numbers and percentages of people reaching above clinical threshold.

The lack of studies means that some results which are widely reported in the literature, such as rates of OCD being high in adults with dystonia (e.g., Bihari, Pigott, Hill, & Murphy, 1992; Cavallaro et al., 2002; Saunders-Pullman et al., 2002), were not found in this review. However, the evidence in this review was clearer for broader terms such as depression and anxiety, and more in line with the general literature (e.g., Heiman et al., 2004; Peall et al., 2013; Tomic et al., 2016; Wenzel et al., 1998). The included studies used a range of different measures and reported the data in different ways (e.g., reporting mental health problems in combination), in addition to some not doing statistical comparisons with the data or reporting effect sizes. This was particularly problematic for making comparisons considering the small number of studies. It was not possible to discuss subtypes of dystonia separately due to the lack of consistency in the types of dystonia, methods and data reported. Whilst effect sizes were calculated where possible, the confidence intervals were very large which suggests these measures of association are not very reliable. The use of the Newcastle-Ottawa scale was a useful tool in terms of assessing methodological quality, and a strength of this review is that the factors of importance in addressing the quality of the included studies were considered using this scale.

Implications for Professional Practice

The results from this review are fairly generalisable considering the wide range of methodologies, types of dystonia, and countries of origin. However, due to the small number of studies, there is limited consistency in reported results which makes it difficult to make concrete conclusions. The results which show depression, anxiety (in a broad sense) and social anxiety to be higher in adults with dystonia were the most consistent, however most of the studies included were of poor methodological quality so it is difficult to ascertain whether these results are reflective of the true prevalence of these mental health problems in adults with dystonia.

Considering this, clinicians working in services attended by adults with dystonia should be aware that these patients may be more likely to have depression, anxiety, or social anxiety than adults without dystonia. It is advisable that people working within these settings know about the signs for these difficulties and monitor people whom they are concerned about. Alternatively, a general depression or anxiety screen could be routinely offered in these settings. Based on these results, a face-to-face screen may be more accurate than a self-report measure.

Implications for Future Research

As the studies in this review were low in numbers, especially considering that 'dystonia' is a broad term, more research on mental health is needed with control groups. This would provide a clearer picture of which types of dystonia are more

likely to be associated with mental health difficulties than the general population. This would make it easier for consideration of these difficulties to be incorporated into care plans of dystonic patients. Future research would need larger sample sizes, a wider variety of recruitment sites, and less restrictive inclusion and exclusion criteria; for small effects to be detectable, and for results to be more generalisable. It is important to note that these results cannot be generalised to people with dystonia who are under 18 years of age, so research with younger participants is also needed.

Conclusions

Overall, most studies in this review found that adults with dystonia are more likely to have depression, anxiety, or social phobia than controls. Other specific anxiety disorders (e.g., OCD, GAD and agoraphobia) and other mental health problems (e.g., psychosis and alcohol dependence) provided varied results. This may have been attributable to small sample sizes, varied recruitment procedures and lack of studies exploring specific mental health problems. The overall likelihood of having a mental health problem was significantly greater in adults with dystonia than controls. However, as the methodological quality of these studies overall was quite poor and varied greatly in terms of dystonia population, measures and interpretation, they should be treated with caution. Services working with adults with dystonia would likely find that patients benefit from general mental health screening.

Empirical Study: Mental Health and Behaviour in Childhood Dystonia

Abstract

Several studies have published data on mental health difficulties in adults with dystonia and children with other movement disorders; however, few studies have explored mental health or behavioural difficulties in young people with dystonia. Fifty young people with dystonia (aged 8-17 years old) and their parents or guardians were recruited from the Evelina London Children's Hospital. Young people completed the Beck Youth Inventories (BYI) and the Strengths and Difficulties Questionnaire (SDQ). Parents completed the parent version of the SDQ (SDQ-P), a demographic and clinical questionnaire and the Paediatric Pain Profile (PPP). Gross Motor Function Classification System Expanded and Revised (GMFCS-E&R) scores were obtained from medical records. Young people with dystonia had significantly higher levels of anxiety and peer problems, and significantly lower levels of disruptive behaviour than population norms. Parents reported significantly higher total difficulties, emotional problems, hyperactivity and peer problems, and significantly lower prosocial behaviours than population norms. Young people with dystonia did not have high levels of depression or anger. Self-concept and pain were related to anxiety and parent-rated total difficulties, whereas age of onset of dystonia and severity of movement disorder did not correlate with any of the psychological or behavioural measures. Interventions targeting self-esteem and pain may therefore be beneficial for young people with dystonia. Due to methodological

problems such as a small sample size and missing data, conclusions drawn from this study are offered tentatively and require replication in a larger sample. Future research could also explore specific anxiety disorders and identity issues in young people with dystonia.

Introduction

There have been several studies which have reported mental health difficulties in adults with dystonia and children with other movement disorders; however, few studies have explored mental health or behavioural difficulties in children and young people with dystonia. The present study aimed to address this by examining relationships between dystonia and mental health and behavioural problems among young people with dystonia.

This introduction will begin by defining dystonia and describing the challenges associated with living with it. Due to the lack of literature on childhood dystonia; the adult literature will be reviewed, followed by literature on childhood movement disorders. Contributory factors to mental health and behavioural problems among children and adults with movement disorders will then be reviewed. Finally, an outline of the research questions of this study, and the hypotheses will be provided.

Definitions of Dystonia and Challenges Associated with Living with It

Dystonia is a movement disorder in which "sustained or intermittent involuntary muscle contractions cause twisting or repetitive movements or abnormal postures'' (Sanger, 2003, p. 1509). Typically, Bressman's (2003) distinction between primary (genetic cause) or secondary (associated with other neurological or metabolic diseases) types of dystonia is used in clinical settings. Dystonia diagnoses are classified in terms of location on the body which is affected by abnormal movements or postures (see page 18 for details of dystonia definition, classifications, prevalence and treatment). Dystonia is considered to be a basal ganglia disorder; however, the neurological mechanisms remain unclear (Degirmenci et al., 2013).

Dystonia can cause problems with physical functioning due to muscle contractions impacting everyday activities such as sitting, walking, talking and eating (Hertenstein et al., 2016; Pavone et al., 2013). Constant movement causes fatigue which can impact a person's ability to concentrate, often causing them to underachieve at school or work (Molho et al., 2016). Patients may have difficulties sleeping due to pain caused by muscle spasms and stiffness, and they may face stigma which impacts their self-esteem and social interactions (Mordin et al., 2014). Many people with dystonia have comorbid health-related difficulties (such as scoliosis, cerebral palsy or stomach problems) which have an additional impact on wellbeing (Schneider & Bhatia, 2010).

Mental Health Problems Among Adults with Dystonia

Many studies have reported that over half of people with dystonia have mental health problems (Moraru et al., 2002; Wenzel et al., 1998). These levels are higher than what would be expected in the general population (Fabbrini et al., 2011; Smit, Kuiper, et al., 2016). Mental health difficulties are associated with poorer quality of life and worse health outcomes (Page et al., 2007; Simon, 2001). It is therefore important that mental health problems are identified to enable these individuals to access psychological support, which may also reduce need for further medical interventions.

As many as 50% of adults with dystonia have been reported to have clinical levels of depression (Demartini et al., 2017). The prevalence of anxiety disorders is also reported to be high among people with dystonia (van Tricht et al., 2012), with as many as 60% of participants scoring within the clinical range (Demartini et al., 2017). The prevalence of specific anxiety disorders such as obsessive-compulsive disorder (OCD) and social anxiety have been found to be high in people with dystonia (Lauterbach, Freeman, & Vogel, 2004; Lehn, Mellick, & Boyle, 2014; see page 21 for further statistics on depression and anxiety in dystonia). In addition to depression and anxiety, people with dystonia can have high levels of alcohol abuse, potentially due to alcohol's depressive effect on dystonia movements (Biary & Koller, 1985; Lauterbach et al., 2004), substance abuse (Wenzel et al., 1998) and bulimia nervosa (Lencer et al., 2009). In a study conducted over five years, mental health difficulties were shown to be persistent in people with dystonia (I. Berardelli et al., 2015),

suggesting they may not be accessing adequate support for mental health difficulties.

Despite many studies finding that people with dystonia are more likely than the general population to have mental health problems, some studies did not replicate these results (Barahona-Corrêa et al., 2011; Johnson et al., 2007). This inconsistency may be due to methodological flaws, such as small sample sizes and insufficient measures to distinguish between symptoms of dystonia and symptoms of mental health problems (such as fatigue or feeling shaky). Overall, studies exploring mental health problems among adults with dystonia have provided evidence that they can be varied in aetiology, are highly prevalent and persistent, and have a negative impact on quality of life (I. Berardelli et al., 2015; Lauterbach et al., 2004; Page et al., 2007).

It is important that mental health problems are identified and addressed as early as possible to minimise the longer-term impact they have on wellbeing and health outcomes. For instance, a person may become depressed as a result of fatigue, uncontrollable movements and pain affecting their ability to perform at work (Molho et al., 2016; Yeo & Sawyer, 2005). Being able to access psychological support to help the person adapt to their impaired physical abilities, and manage their fatigue, pain, and mood may help them to keep their job and therefore maintain independence. Alternatively, some people with dystonia may feel embarrassed about their uncontrollable movements so avoid social events; accessing psychological support to develop their social skills may help their ability to make meaningful relationships in later life. As many forms of dystonia typically begin in

childhood (Hertenstein et al., 2016; Kinugawa et al., 2009), an important direction for research into mental health in dystonia is to focus on children and adolescents and to identify areas where they may be in more need of psychological support.

Mental Health and Behavioural Problems Among Children with Movement Disorders

There have been few investigations of mental health problems among young people with dystonia beyond published case studies (e.g., Blackburn & Cirillo, 2012). A systematic review of 88 studies reporting non-motor symptoms in paediatric movement disorders revealed that many children with such disorders have cognitive and behavioural problems, attention deficit hyperactivity disorder (ADHD), depression, and/or anxiety (Ben-Pazi, Jaworowski, & Shalev, 2011). This review however, contained just three studies of dystonia; one study relied on historical reports (Koukouni, Martino, Arabia, Quinn, & Bhatia, 2007), and two had small sample sizes (*n*= 4 Hahn et al., 2001; *n*= 16 Saunders-Pullman et al., 2002). Small sample sizes make it difficult to ascertain the extent of mental health or behavioural difficulties in dystonia due to them not having sufficient power to establish such relationships.

Studies on children with other movement disorders (such as cerebral palsy) are helpful in identifying the difficulties young people with dystonia may have. Cerebral palsy is characterised by spasticity, ataxia, and at times, dystonia; therefore, studies on cerebral palsy may include children where dystonia is part of their

presentation. One large-scale study of 8-12 year olds with cerebral palsy (*N*=818) found that a quarter had significant psychological symptoms, as measured by the Strengths and Difficulties Questionnaire (Parkes et al., 2008). They scored particularly highly on the emotional problems, hyperactivity and peer problems subscales. Another study of 47 children aged six years old found that over half of children met criteria for a psychiatric disorder, most commonly ADHD or OCD (Bjørgaas, 2015). In this study, most children who did not meet criteria for diagnosis of an emotional problem reached sub-threshold. Conversely, another study found that children with cerebral palsy did not differ from their siblings in terms of anxiety (Cohen, Biran, Aran, & Gross-Tsur, 2008), however this may be due to there being high levels of anxiety in the household overall.

Considering that mental health and behavioural problems appear to be highly prevalent in children with cerebral palsy (Bjørgaas, 2015), it is probable that young people with dystonia are more likely to have mental health and behavioural difficulties than the general child population. Understanding the mental health needs of children with cerebral palsy has led to interventions being developed to manage their specific needs, such as helping them cope when undergoing painful procedures (Yu, Liu, Li, & Ma, 2009). As different movement disorders have varying motor and non-motor profiles (Dias et al., 2011), studies of other childhood movement disorders cannot be used to make decisions about interventions which may help young people with dystonia. The present study was designed to address this issue by exploring the relationships between dystonia and mental health and behavioural problems among young people.

Self-report screening tools may overestimate mental health difficulties in people with movement disorders as they may score highly due to their movement disorder symptoms. One study addressed this difficulty using a sample of 56 children with cerebral palsy (Bjørgaas, Elgen, Boe, & Hysing, 2013). Comparisons with clinical interviews revealed that a screening tool (the Strengths and Difficulties Questionnaire) could capture the complexity of difficulties that children with movement disorders face. One further criticism of measurement tools in research is that children often do not complete them themselves. This is problematic as differences have been found between young people and their parents in terms of how they perceive their wellbeing (Janssen, Voorman, Becher, Dallmeijer, & Schuengel, 2010). It is important that young people report on their own difficulties to get a more accurate picture of the issues they may need support with.

Factors Contributing to Mental Health and Behavioural Problems Among Young People with Dystonia

Dystonia does not necessarily cause mental health difficulties. The biopsychosocial model of health shows the interrelation of biological, psychological and social aspects that may affect a young person with dystonia (Engel, 1977; see page 23 for further discussion). For example, physical symptoms and medication effects may impact a young person's ability to take part in activities other young people take part in, and as a result impact their social skills and self-esteem (Bøttcher & Dammeyer, 2013). Young people who exhibit visible movement

symptoms may become particularly self-conscious as adolescents (Lees, 2002). Young people may also lack a support network or the specific coping skills required to deal with difficult emotions associated with living with dystonia, which may result in maladaptive coping strategies or behavioural difficulties. In comparison to adults, children may have additional problems relating to dystonia; for example, children who miss school due to attending frequent medical appointments may struggle to complete work to their full potential, or may have parents who are very anxious about their unwell child (Raina et al., 2005). These factors may contribute to a mental health or behavioural difficulty; therefore, it is important that social and psychological issues are monitored and addressed in conjunction with physical health care.

Several factors relating to dystonia may impact on wellbeing. High pain levels in children with cerebral palsy have been linked to higher rates of mental health difficulties (Parkes et al., 2008). In dystonia, one study found that mental health problems and pain make the largest contributions to disability (van den Dool et al., 2016), whilst physical functioning made no significant contribution. These studies suggest that pain plays a role in wellbeing, sometimes more so than the physical limitations of the disability. Improvements in pain management can also have an effect on symptoms of dystonia and depression, for example they may improve sleep which would therefore reduce fatigue (Ramstad et al., 2012).

Higher levels of disability in movement disorders have been associated with stigma, emotional state, pain, social life, and activity levels (Tomic et al., 2016). In dystonia, motor severity has been associated with depression and quality of life

(Ben-Shlomo, Camfield, & Warner, 2002; Queiroz, Chien, & Barbosa, 2011), suggesting that physical limitations have an impact on wellbeing. In children with cerebral palsy, higher levels of disability have been associated with lower levels of social functioning (van Schie et al., 2013). Young people with poorer social skills are at increased risk of social exclusion, and therefore mental health difficulties.

The age of onset of dystonia is likely to have an impact on mental health. One study on Parkinson's disease found that young-onset patients had depression more frequently than older-onset patients (Schrag, Hovris, Morley, Quinn, & Jahanshahi, 2003), due to loss of employment, perceived stigmatisation and poorer quality of life. Duration of motor symptoms has also been associated with higher incidence of psychiatric disorders in dystonia (Peall et al., 2013). The experience of having a movement disorder as a child can differ from that of an adult, by impacting on their performance at school and potentially preventing them from building relationships with peers (van Schie et al., 2013). Being diagnosed as a teenager may contribute to increased self-consciousness as they become 'different' from peers (Bjørgaas, 2015).

In movement disorders, depression has been associated with negative body perception and low self-esteem (Lewis et al., 2008). Low self-esteem can have a significant effect on quality of life (Basurovic, Svetel, Pekmezovic, & Kostic, 2012). One study found that low self-esteem was associated with anxiety in young people with cerebral palsy, despite levels of self-esteem not differing from controls (Borkowska, 2015). Children with cerebral palsy who have better self-esteem have higher levels of social participation and positive perceptions of disability (Manuel, Balkrishnan, Camacho, Smith, & Koman, 2003), and therefore may be protected from

mental health problems arising due to social exclusion. A study of adults with multiple sclerosis found that having a positive view of one's own disability and incorporating this into their identity (such as by attending support groups) has a good impact on mental wellbeing (Bogart, 2015). This fits with social identity theory, which states that a sense of belonging, social identity and pride can result from belonging to a social group (Tajfel & Turner, 1979). Having a more positive sense of self could be protective for young people with dystonia, therefore interventions targeting self-esteem or social inclusion could be beneficial.

Factors such as pain, severity, age of onset and self-esteem have been associated with mental health problems in adults and children with movement disorders; therefore, it is important to consider the influence of these factors when investigating mental health problems in young people with dystonia.

The Present Study

In total, 50 young people with dystonia and a parent or guardian completed self-report questionnaires assessing for mental health and behavioural difficulties. This study aimed to answer the following research questions: 1) Are young people with dystonia more likely to have mental health difficulties (particularly with depression and anxiety) and behavioural problems than the general child and adolescent population; and 2) To what extent are self-concept, severity of movement disorder, pain, and age of onset of dystonia associated with mental health and behavioural problems in young people with dystonia?

The first question which the study aimed to address is whether young people with dystonia are more likely to have mental health difficulties and behavioural problems than the general child population. Research with adults has shown that adults with dystonia and children with movement disorders are more likely to have depression, anxiety and behavioural problems than the general population. It was hypothesised that young people with dystonia would score significantly higher on measures of mental health and behavioural difficulties than normed controls. Assessing young people with dystonia would help determine whether they are more likely to have mental health or behavioural difficulties, and whether it is necessary to routinely screen or provide additional support to them.

Secondly, the study aimed to explore the extent to which pain, severity of movement disorder, age of onset of dystonia, and self-concept are associated with mental health and behavioural problems in young people with dystonia. These factors have previously been linked to mental health difficulties in adults with dystonia and children with movement disorders. It was hypothesised that higher levels of severity and pain, younger age of onset and lower self-concept would be significantly associated with higher levels of mental health and behavioural problems in young people with dystonia. Exploring these factors would indicate which young people with dystonia may be more at risk of developing mental health or behavioural difficulties, and where intervention may be beneficial.

Method

Design

This research used a cross-sectional design in which every person participated once by completing a battery of standardised psychological questionnaires. For the first research question; a quasi-experimental design was used where young people (*n*=12-31) with dystonia were compared to normed samples on the Beck Youth Inventories (BYI) and the Strengths and Difficulties Questionnaire (SDQ), measures of mental health and behaviour. The independent variable was the presence of a diagnosis of dystonia, and the dependent variables were psychological and behavioural difficulties as measured by the BYI and SDQ. For the second research question, a within-subjects design was used (*n*=23-50). The independent variables were the age of onset of dystonia, pain, severity of movement disorder and self-concept. The dependent variables were psychological and behavioural difficulties as measured by the BYI and SDQ.

Participants

<u>The Dystonia Sample</u>

The final sample consisted of 50 young people with dystonia (Table 8) and a parent or guardian for each young person. Young people aged 8.5-17.8 years participated. The range for age of onset of dystonia was 0-14 years.

Of the total sample, 38% had primary dystonia (caused by genetic mutation) and 62% had secondary dystonia (associated with another health problem). Thirty-eight percent of the sample had undergone deep brain stimulation surgery (DBS). Twenty-two percent of parents reported that their child had mental health or behavioural needs, such as problems with low mood (*n*=3), anxiety (*n*=2), obsessive-compulsive traits (*n*=2), anger (*n*=2), and self-esteem (*n*=1). Seventy-four percent of parents said their child had a statement of special education needs; 44% due to learning needs (such as a learning disability or specific problem with attention or memory).

Table 8: *Demographic and Clinical Characteristics*

Characteristic		*n* (%)	Mean (SD)	Median (range)
Age		50 (100%)	13.5 (2.8)	14.0 (8.5-17.8)
Gender	**Male**	22 (44%)		
	Female	28 (56%)		
Ethnicity	**Asian**	3 (6%)		
	Black	5 (10%)		
	Mixed	4 (8%)		
	White	38 (76%)		
Age of Onset		39 (78%)	5.8 (4.7)	4.8 (0-14)
Dystonia Type	**Primary**	19 (38%)		
	Secondary	31 (62%)		
Medication (Y)		33 (66%)		
Deep Brain Stimulation Surgery (Y)		19 (38%)		
Comorbid Health (Y)		31 (62%)		
Mental Health or Behavioural Problem (Y)		11 (22%)		
Special Educational Needs (Y)		37 (74%)		
Paediatric Pain Profile		33 (66%)	19.03 (12.3)	13 (4-57)
GMFCS-E&R		43 (86%)	2.95 (1.5)	3 (1-5)

Note: GMFCS-E&R gross motor function classification system expanded and revised. *n*'s vary due to missing data.

Comparison of Dystonia Sample with Population Sample

The dystonia sample was compared with population norms of the BYI and SDQ (Beck, Beck, & Jolly, 2005; Meltzer, Gatward, Goodman, Ford, & Melzer, 2000). For the BYI, the dystonia sample was split into children (aged 7-14 years, *n*=31) and adolescents (aged 15-17 years, *n*=17) to make direct comparisons with the published norms according to age group. The standardisation sample was recruited in the

United States of America and consisted of 107 children and 148 adolescents. For the SDQ and SDQ-P, only data from those aged 11-15 years (*n*=21) in the dystonia sample was compared, due to the norms available. The standardisation sample was recruited from the United Kingdom and consisted of 4228 children and 4443 parents.

Recruitment

All dystonia participants were recruited from the Evelina London Children's Hospital between October 2017 and March 2018. Twenty-six children were recruited from the Complex Movement Disorders Service, 20 children were recruited from the Neurology clinics, and four children were recruited from the Botulinum toxin (Botox) clinics. Several clinics were selected to access greater numbers of eligible participants, and to facilitate a greater variety of participants in the sample (i.e., in terms of severity, type, and treatment of dystonia).

In total, 88 children and their parents were approached about the study. The participation rate was 57%. Reasons for not taking part included; parents feeling that the child would not understand the questions (*n*=11), not being able to stay after their hospital appointment (*n*=5), and parents feeling that their child was too stressed to take part (*n*=4). Two participants declined without giving a reason. In some cases, a potential participant agreed to take part, but it was not possible to meet them at any of their future appointments (*n*=16).

Inclusion and Exclusion Criteria

Participants were invited to take part in the study if they had a diagnosis of dystonia and were aged between 7 and 17 years old. Those children who had a diagnosis of a life-limiting condition (e.g., PANK-2 disorder) were excluded from the study as their needs are likely to be different from children who are not on the palliative pathway. Additionally, those who lacked capacity to consent to taking part, those who were unable to understand the questions in the questionnaire, or could not communicate their answers were excluded from the study to ensure informed consent was taken and that answers were valid.

Service-User Involvement

Before applying for NHS ethical approval, two patients from the Complex Movement Disorders Service were asked for feedback on the project materials. Both completed the BYI and SDQ and could complete these questionnaires without difficulty. The parent of one child with attention difficulties advised reading out the information sheet to ensure the child understood what was being asked of them. This was incorporated into the research protocol.

Ethical Approval

The Doctorate in Clinical Psychology Research Committee at Royal Holloway, University of London, approved the initial study proposal and design. NHS ethical

approval for the study was subsequently granted by the London-Dulwich Research Ethics Committee, the Health Research Authority and Research and Development at Guys and St Thomas's NHS Trust. Finally, ethical approval was granted by Royal Holloway, University of London's Research Ethics Committee. Appendix 2 contains copies of the approval documents.

Measures

The following measures were completed. Appendix 3 contains copies of these measures.

Beck Youth Inventories

The Beck Youth Inventories (BYI; Beck et al., 2005) are designed to measure a range of mental health and behavioural difficulties, in addition to self-concept among children and adolescents. The BYI is a 100-item self-report questionnaire, which includes 20 items for each subscale; depression, anxiety, anger, disruptive behaviour and self-concept. Items are rated on a four-point scale (never, sometimes, often, always) and children choose which word best describes them. All scores are positively weighted and range from 0-60 for each subscale. Raw scores are converted to *t* scores. For the self-concept scale the clinical ranges are as follows: >55 above average; 45-55 average; 40-44 lower than average, and; <40 much lower than average. For all other scales the clinical ranges are: <55 average; 55-59 mildly elevated; 60-69 moderately elevated, and; 70+ extremely elevated. Participants

could complete this measure without assistance, or with assistance (i.e., verbally, pointing or using eye gaze).

The BYI has been tested for internal consistency reliability across the age range of participants for this study (Cronbach's α range= .86-.96). Test-retest reliability after 7-8 days indicates acceptable reliability of scores (*r*= .74-.93, Beck et al., 2005). A large correlation has been reported between the Children's Depression Inventory and the BYI depression subscale (*r*= .81, Steer, Kumar, Beck, & Beck, 2001), and moderate correlations have been reported between the Revised Children's Manifest Anxiety Scale and child (*r*= .70) and adolescent (*r*= .64) scores on the BYI anxiety subscale (Beck et al., 2005).

Strengths and Difficulties Questionnaire

The Strengths and Difficulties Questionnaire (SDQ and SDQ-P; Goodman, Meltzer, & Bailey, 1998) is a 25-item behavioural screening questionnaire which contains five items for each subscale; emotional problems, conduct problems, hyperactivity, peer problems, and prosocial behaviour. All items are rated on a three-point scale (not true, somewhat true, certainly true). Most questions are positively rated; however, five items are reverse scored. Scores for each subscale range from 0-10, and all subscales except prosocial behaviour are combined to give a total difficulties score between 0 and 40. Different cut-offs are applied for each subscale to classify the child into the following categories: close to average; slightly raised/lowered; high/low; or very high/low. Parental data were collected for all

participants and self-report data were collected for those aged 11-17 years, due to the age-range the questionnaire is validated for. Participants could complete this with or without assistance.

This measure has substantial concurrent validity with similar measures, such as the Child Behaviour Checklist (*r*= .70 for total score; Muris, Meesters, & van den Berg, 2003). It has satisfactory internal consistency reliability (mean Cronbach's α= .73; Goodman, 2001) and test-retest reliability (*r*= .85 for total difficulties; Goodman, 1999).

Demographic and Clinical Questionnaire

Demographic and clinical information about young people was collected from parents using a demographic and clinical questionnaire. The information included date of birth, age of diagnosis of dystonia, details of any comorbid physical health diagnoses, medication use, ethnicity, gender, details of any current or previous known mental health difficulty, and whether their child has had any cognitive assessments or has a statement of special educational needs. Further details were collected as to what type and level of learning disability (LD) they have.

The Paediatric Pain Profile

The Paediatric Pain Profile (PPP; Hunt, Mastroyannopoulou, Goldman, & Seers, 2003) is a 20-item behaviour rating scale which was completed by parents.

Parents rate their child on a range of behaviours that they may exhibit when in pain. All items are rated on a four-point scale (not at all, a little, quite a lot, a great deal). Two items are reverse scored. The range of scores is from 0-60. Scores of 14 or more are considered as indicative of moderate or severe pain. Parents also rate their child's pain as none, mild, moderate, severe, or very severe. Parental data were collected for all participants who experienced pain because of their dystonia or comorbid health problem.

A study on 140 children whose parents and clinicians rated their pain before and after surgery, concluded that this scale has good interrater reliability (ICC= .87) and internal consistency reliability (Cronbach's α range= .75-.89) for assessing child pain (Hunt et al., 2004).

The Gross Motor Function Classification System Expanded and Revised

The Gross Motor Function Classification System Expanded and Revised (GMFCS-E&R; Palisano, Rosenbaum, Bartlett, & Livingston, 2007) is typically used in cerebral palsy, although is used clinically with young people with dystonia. It is a clinician-rated scale which classifies children over the age of two years into five levels according to their level of motor function and performance in everyday environments (i.e., sitting, walking and use of mobility devices). This information was collected from clinical records. The classification was made after a full physiotherapy assessment by a Physiotherapist.

The GMFCS-E&R is moderately correlated with other established tests of gross motor function, and therefore has criterion-rated validity (Bodkin, Robinson, & Perales, 2003). This measure has excellent inter- and intra-rater reliability (ICC= .97 and .98 respectively; Piscitelli, Vercelli, Meroni, Zagnoni, & Pellicciari, 2017). The measure is appropriate to use in children with dystonia (Elze et al., 2016) as it has a strong relationship with the Burke-Fahn-Marsden Dystonia Rating Scale-movement (a broader measure of dystonia severity, *r*= .84).

Medical Records

Medical records were checked in cases where parents were unable to provide information on the demographic and clinical questionnaire, such as type of dystonia, comorbid health difficulties, and medication.

Procedure

The appointment lists for the recruiting clinics were screened for eligible young people by the medical professionals running the clinics. Eligible participants were told about the study initially via a telephone call or face-to face during a visit to the hospital. The researcher then arranged to meet them during their next visit to the hospital. This was either in a private room within the clinic, in the clinic waiting room or at their bedside, depending on room availability, level of assistance required and if they were at hospital for a medical procedure. Upon meeting, the information sheets and consent forms (Appendix 4) were discussed, and the young person and

parent or guardian were given the opportunity to ask questions. If happy to take part, the young person and parent or guardian were given the questionnaires to complete. The researcher was available whilst the measures were being completed, to answer any further questions and to offer assistance.

When the young person could complete the measures themselves, it took an average of 20 minutes for the questionnaires to be completed. Twenty-six young people needed assistance with questionnaires, whereby the researcher read out the questions, and the young person answered verbally, by pointing, or using eye gaze. For these young people, it took between half an hour and one hour to complete the measures. A small number of young people did not complete all the BYI subscales (*n*=13) or the SDQ (*n*=3) due to them being unable to concentrate or becoming disengaged due to the length of time it took to complete the questionnaires. Completing all the subscales in these cases may have led to invalid answers or the young person becoming distressed.

Once completed, participants were thanked and told that they would receive a summary of the results once the study ended. Twelve children selected 'sometimes', 'often' or 'always' for the question on the BYI "I wish I was dead". For these children, advice was sought from the Clinical Psychologist supervising the project and departmental risk protocols were followed.

Data Analyses

Statistical power was calculated using online software (Soper, 2018). For the first research question using one sample z tests; a sample of 20 participants was required for each analysis for a medium effect size and power of .80. Sufficient power was achieved for the BYI child, SDQ and SDQ-P comparisons. For the BYI adolescent sample, all comparisons were underpowered. For the second research question using hierarchical multiple regression; a medium effect size and power of .80 (with three predictor variables in set A) required a sample of 57 for the analyses using one predictor variable in set B, and a sample of 70 for analyses using two predictor variables in set B. These analyses were therefore underpowered.

Data were cleaned prior to analysis. Participants were categorised into Asian, Black, Mixed or White. Dystonia type was coded into primary or secondary. Medical records were checked where this information was not provided or was unclear. Age of onset was categorised into the age the child was when diagnosed. For children under 1 year, a 0 was used. Statement of special educational needs were categorised into: physical disability; learning disability; learning needs; visual impairment; hearing impairment; and autism.

For missing items on the BYI and SDQ, the guidelines as determined by the measures' authors were followed, where the average score for the other items on that subscale was used. There were no incidences of BYI or SDQ data being excluded due to missing data (i.e., no more than two items were missing from each subscale). As per the measure's guidelines, missing scores from the PPP were entered as 0. The medical records of 19 participants did not contain a GMFCS-E&R score. For 12 of

these participants, a Physiotherapist allocated an estimated score based on their medical records. Data remained missing for seven participants where it was not possible to estimate a score.

IBM SPSS Statistics version 21 was used for all analyses. Results of the BYI, SDQ, SDQ-P and PPP were checked for outliers. Scores which were three standard deviations from the mean were removed (one from each of the BYI anger and disruptive behaviour subscales, SDQ conduct problems subscale and the PPP). The distribution of subscales on the BYI, SDQ, SDQ-P and PPP were checked for normality (z scores of >2.58 on measures of skew or kurtosis). For the subscales which were skewed (BYI anger, SDQ conduct problems, SDQ-P total difficulties, emotional problems and conduct problems, and the PPP), square root transformations were carried out, resulting in them being normally distributed. Agreement ratings between child and parent respondents on the SDQ were calculated using intraclass correlation coefficients.

Descriptive statistics were generated for participant demographic and clinical characteristics, and percentages of young people who scored above clinical cut-off on the BYI and SDQ. Chi-square tests, *t*-tests and one-way ANOVAs were used to investigate relationships between psychological and behavioural measures and demographic characteristics. For Chi-square tests, Fisher's exact test was used in instances where the expected frequency in any cell was less than five. For *t*-tests, separate variance estimates were used when homogeneity of variance assumptions were not met. Fisher's protected *t*-tests were used to make comparisons between individual groups when *F* values from one-way ANOVAs were significant.

For the BYI and SDQ measures, one sample z tests were used to compare the mean scores from the dystonia group with those from the normed control group. One sample z tests were chosen over *t* tests as the standard deviations of the normed population sample were known and this variance could therefore be taken into account in the analysis. Pearsons's product moment correlation coefficients were used to examine which variables (self-concept, age of onset of dystonia, pain, and severity of movement disorder) were significantly associated with the psychological and behavioural variables as measured by the BYI and SDQ. Hierarchical multiple regression analyses were conducted to identify significant independent predictors of anxiety and total difficulties among children and adolescents with dystonia. Bonferroni corrections were applied to correlations to control for multiple comparisons within the dystonia group, however were deemed too strict to apply to the one sample z tests comparing the dystonia and normed control groups.

Results

Intraclass Correlations (ICC) Between Young People and their Parents on the SDQ

For those participants for whom the SDQ and SDQ-P were completed, the inter-rater agreements for each subscale were explored. The inter-rater agreements between young people and their parents were moderate for the total difficulties (ICC(32,32) = .55 [95% CI: .08-.78]), emotional problems (ICC(32,32) = .54 [95% CI: .07-.77]), conduct problems (ICC(32,32) = .65 [95% CI: .31-.83]), hyperactivity

(ICC(32,32) = .62 [95% CI: .25-.81]) and peer problems (ICC(32,32) = .61 [95% CI: .21-.81]) subscales. The inter-rater agreement was good for the prosocial subscale (ICC(32,32) = .80 [95% CI: .60-.90]).

Relationships Between Demographic and Clinical Characteristics, and the BYI and SDQ

The relationships between demographic and clinical characteristics, and psychological and behavioural factors measured using the BYI and the SDQ, were examined. Female participants were significantly more likely to score above clinical threshold than participants who were male, on the BYI anxiety subscale (females 60.7%; males 30.0%; $\chi^2(1) = 4.41$, p=.04), and the SDQ total difficulties (females 47.1%; males 6.3%; χ^2, df= 1; p=.01), and emotional problems (females 58.8%; males 6.3%; $\chi^2(1) = 10.25$, $p< .01$) subscales.

Participants who had DBS scored significantly higher than participants who had not had DBS surgery on the BYI anxiety subscale (mean=56.84, SD=9.0 vs. mean=50.59, SD=11.2 respectively; $t(46) = 2.04$, $p< .05$). Additionally, participants with learning needs scored significantly higher than participants without learning needs on the BYI anxiety subscale (mean=56.36, SD=10.9 vs. mean=50.27, SD=10.0 respectively; $t(46) = -2.03$, $p< .05$).

Young people whose parents did not report that their child has pain as a result of dystonia scored significantly higher on the BYI disruptive behaviour subscale than young people whose parents did report pain (mean=46.27, SD=5.3 vs.

mean=41.05, SD=6.5 respectively; $t(35) = -2.60$, $p=.01$). Young people whose parents reported that their child had a moderate or severe level of pain scored significantly higher on the BYI depression (mean=50.7, SD=10.1 vs. mean=42.3, SD=8.6 respectively; $t(28) = -2.4$, $p=.02$) and SDQ-P total difficulties (mean=15.0, SD=7.9 vs. mean=9.59, SD=4.9 respectively; $t(24.7) = -2.4$, $p=.03$) and conduct problems (mean=1.81, SD=1.5 vs. mean=0.71, SD=1.3 respectively; $t(29.7) = -2.3$, $p=.03$) subscales, than young people whose parents reported mild pain.

Percentages of Young People Scoring Above Clinical Cut-Off on the BYI and SDQ

The percentages of young people who scored above clinical cut-off for the subscales on the BYI, SDQ and SDQ-P are shown in Tables 9 and 10.

On the BYI, 47.9% of young people with dystonia scored above clinical cut-off for anxiety (51.6% of children and 41.2% of adolescents). For depression, 23.4% scored above clinical cut-off (19.4% of children and 31.3% of adolescents). On the anger subscale, 21.4% of adolescents scored above clinical cut-off compared to 7.7% of children. On the disruptive behaviour subscale, 12.0% of children scored above clinical cut-off compared to 7.7% of adolescents. For the SDQ, 27.3% of young people with dystonia self-reported slightly raised, high, or very high total difficulties, and parents reported 32.0% of their children as being above cut-off for total difficulties.

Table 9: *Participants Scoring Above Clinical Cut-Off on BYI Subscales*

Dystonia Sample	**BYI Subscale**	**Total *n***	***n* above cut-off**	**% above cut-off**
Whole	**Anxiety**	48	23	47.9
	Depression	47	11	23.4
	Anger	40	5	12.5
	Disruptive Behaviour	38	4	10.5
Child	**Anxiety**	31	16	51.6
	Depression	31	6	19.4
	Anger	26	2	7.7
	Disruptive Behaviour	25	3	12.0
Adolescent	**Anxiety**	17	7	41.2
	Depression	16	5	31.3
	Anger	14	3	21.4
	Disruptive Behaviour	13	1	7.7

Table 10: *Participants Scoring Above Clinical Cut-Off on SDQ Subscales*

Respondent	SDQ Subscale	Total *n*	*n* above cut-off	% above cut-off
Child	**Total Difficulties**	33	9	27.3
	Emotional Problems	33	11	33.3
	Conduct Problems	33	5	15.1
	Hyperactivity	33	10	30.3
	Peer Problems	33	13	39.4
	Prosocial	33	8	24.2
Parent	**Total Difficulties**	50	16	32.0
	Emotional Problems	50	16	32.0
	Conduct Problems	50	8	16.0
	Hyperactivity	50	19	38.0
	Peer Problems	50	28	56.0
	Prosocial	50	16	32.0

Comparison of Dystonia Sample and Population Norms on the BYI and SDQ

Scores on all subscales of the BYI and SDQ (child and parent versions) were compared against the population norms (Beck, Beck, & Jolly, 2005; Meltzer, Gatward, Goodman, & Ford, 1999; see Table 11). On the BYI anxiety subscale, children (z= 3.05, $p< .01$, $d= 0.55$) and adolescents (z= 2.34, $p=.02$, $d= 0.57$) with dystonia scored significantly higher than the population norms. Children with dystonia scored significantly lower on the BYI depression (z= -2.06, $p=.04$, $d= -0.37$),

and anger (z= -3.37, *p*< .001, *d*= -0.66) subscales than population norms, although adolescents did not. On the BYI disruptive behaviour subscale, children (z= -3.07, *p*< .01, *d*= -0.61) and adolescents (z= -1.99, *p*< .05, *d*= -0.58) with dystonia scored significantly lower than the population norms.

On the SDQ, young people with dystonia self-reported significantly higher levels of peer problems (z= 3.21, *p*< .01, *d*= 0.70) and significantly lower conduct problems (z= -2.45, *p*=.01, *d*= -0.54) than the population norms. They did not report significantly higher or lower levels of total difficulties, emotional problems or hyperactivity in comparison to the general population. In contrast, parents reported that young people with dystonia experience significantly higher levels of total difficulties (z= 5.21, *p*< .001, *d*= 1.04), emotional problems (z= 5.05, *p*< .001, *d*= 1.01), hyperactivity (z= 3.31, *p*< .001, *d*= 0.66) and peer problems (z= 7.47, *p*< .001, *d*= 1.49) and significantly lower prosocial behaviours (z= -2.50, *p*=.01, *d*= -0.50) than the population.

Table 11: *Population Norms vs. Dystonia Sample on the BYI and SDQ*

Measure & Respondent	Subscale	Norms			Sample			z score	*p* value	Cohen's *d*
		n	Mean	SD	*n*	Mean	SD			
BYI	**Self-Concept**	107	51.6	9.1	31	51.7	7.7	0.08	.94	0.01
(child sample)	**Anxiety**	107	48.4	9.4	31	53.6	10.7	3.05	**< .01**	0.55
	Depression	107	48.7	8.4	31	45.6	7.4	-2.06	**.04**	-0.37
	Anger	107	48.6	8.2	26	43.2	8.0	-3.37	**< .001**	-0.66
	Disruptive Behaviour	107	48.4	8.5	25	43.2	7.7	-3.07	**< .01**	-0.61
BYI	**Self-Concept**	148	51.6	8.8	17	52.8	9.9	0.57	.57	0.14
(adolescent sample)	**Anxiety**	148	48.2	7.0	17	52.2	11.0	2.34	**.02**	0.57
	Depression	148	48.1	7.2	16	50.9	11.0	1.54	.12	0.39
	Anger	148	47.4	7.6	13	43.9	8.9	-1.68	.09	-0.47
	Disruptive Behaviour	148	47.8	8.2	12	43.1	2.8	-1.99	**< .05**	-0.58

Measure & Respondent	Subscale	Norms			Sample			z score	*p* value	Cohen's *d*
		n	Mean	SD	*n*	Mean	SD			
SDQ	**Total Difficulties**	4228	10.3	5.2	21	11.8	4.1	1.33	.18	0.29
11-15 years	**Emotional Problems**	4228	2.8	2.1	21	3.4	2.2	1.38	.17	0.30
	Conduct Problems	4228	2.2	1.7	21	1.3	1.3	-2.45	**.01**	-0.54
	Hyperactivity	4228	3.8	2.2	21	4.6	2.8	1.71	.09	0.37
	Peer Problems	4228	1.5	1.4	21	2.5	1.4	3.21	**< .01**	0.70
	Prosocial	4228	8.0	1.7	21	7.3	2.5	-1.91	.06	-0.42
SDQ-P	**Total Difficulties**	4443	8.2	5.8	25	14.2	7.6	5.21	**< .001**	1.04
11-15 years	**Emotional Problems**	4443	1.9	2.0	25	3.9	3.0	5.05	**< .001**	1.01
	Conduct Problems	4443	1.5	1.7	25	1.4	1.5	-0.41	.68	-0.08
	Hyperactivity	4443	3.2	2.6	25	4.9	3.1	3.31	**< .001**	0.66
	Peer Problems	4443	1.5	1.7	25	4.0	2.1	7.47	**< .001**	1.49
	Prosocial	4443	8.6	1.6	25	7.8	2.1	-2.50	**.01**	-0.50

Note: Significant *p* values in bold.

Correlations Between Age of Onset, Severity, Pain and Self-Concept, and the BYI and SDQ Subscales

Pearson's product moment correlation coefficients were examined to explore the relationships between the hypothesised predictor variables (age of onset of dystonia, severity of movement disorder, pain and self-concept) and the mental health and behavioural outcome measures (BYI and SDQ child and parent versions). Results are shown in Tables 12-14 and includes bivariate and partial correlations (controlling for age group, gender and type of dystonia). After applying Bonferroni corrections to control for multiple comparisons, all the significant correlations (except for pain and parent-rated conduct problems) became non-significant, indicating that the results should be interpreted with caution as it is not possible to rule out the possibility of type 1 errors.

There was a significant negative correlation between self-concept and anxiety (measured by the BYI), that is, lower levels of self-concept were associated with higher levels of anxiety ($r(46) = -.35$, $p=.02$) among young people with dystonia. There were significant negative correlations between self-concept and total difficulties and emotional problems (measured by the SDQ child version), indicating that lower levels of self-concept were associated with higher levels of total difficulties ($r(31) = -.41$, $p=.02$) and emotional problems ($r(31) = -.41$, $p=.02$). In addition, on the SDQ-P, there were significant negative correlations between self-concept and total difficulties ($r(46) = -.29$, $p< .05$) and emotional problems ($r(46) = -.31$, $p=.03$) among young people with dystonia.

Higher levels of pain among young people with dystonia were associated with significantly higher levels of anger ($r(22) = .44$, $p=.03$) and lower levels of prosocial behaviour ($r(21) = -.41$, $p< .05$). Higher levels of pain were also associated with higher levels of parent-rated total difficulties ($r(30) = .36$, $p=.04$), conduct problems ($r(30) = .49$, $p< .01$), and hyperactivity ($r(30) = .41$, $p=.02$).

Table 12: *BYI Subscales and Hypothesised Predictor Variables Correlations*

Variables	**BYI Subscale**			
	Anxiety	**Depression**	**Anger**	**Disruptive Behaviour**
	(*n*=48)	(*n*=47)	(*n*=39)	(*n*=37)
Self-Concept	-.35*	-.27	.00	.15
(*n*=48)	(-.30*)	(-.23)	(.05)	(.14)
Age of Onset	.00	-.05	-.15	-.02
(*n*=37)	(.11)	(-.04)	(-.02)	(.03)
Pain	.15	.37	.44*	-.03
(*n*=30)	(.05)	(.31)	(.38)	(-.04)
Severity	.03	.08	.17	.02
(*n*=43)	(-.15)	(.00)	(-.02)	(-.04)

Note: *$p < .05$. **$p < .01$. *p* values in bold indicate significance after Bonferroni corrections $p = .0125$. Values in brackets indicate partial correlations controlled for age group, gender and type of dystonia.

Table 13: *SDQ Subscales and Hypothesised Predictor Variables Correlations*

Variables	**SDQ Subscale (child completed)**					
	Total	**Emotional**	**Conduct**	**Hyper.**	**Peer**	**Prosocial**
	(*n*=33)	(*n*=33)	(*n*=32)	(*n*=33)	(*n*=33)	(*n*=33)
Self-Concept	-.41*	-.41*	-.25	-.34	-.09	.29
(*n*=33)	(-.34)	(-.32)	(-.25)	(-.34)	(-.05)	(.33)
Age of Onset	-.04	-.07	-.23	.15	-.10	-.02
(*n*=25)	(-.05)	(-.19)	(-.24)	(.21)	(-.03)	(-.05)
Pain	.31	.31	-.21	.26	.05	-.41*
(*n*=23)	(.22)	(.26)	(-.27)	(.23)	(-.02)	(-.50*)
Severity	.11	-.05	-.04	.03	.25	.03
(*n*=28)	(.06)	(-.03)	(-.09)	(-.04)	(.17)	(.05)

Note: *$p < .05$. **$p < .01$. *p* values in bold indicate significance after Bonferroni corrections *p*=.0125. Values in brackets indicate partial correlations controlled for age group, gender and type of dystonia.

Table 14: *SDQ-P Subscales and Hypothesised Predictor Variables Correlations*

Variables	**SDQ Subscale (parent completed)**					
	Total	**Emotional**	**Conduct**	**Hyper.**	**Peer**	**Prosocial**
	(*n*=50)	(*n*=50)	(*n*=50)	(*n*=50)	(*n*=50)	(*n*=50)
Self-Concept	-.29*	-.31*	-.14	-.17	-.16	.16
(*n*=48)	(-.26)	(-.28)	(-.10)	(-.14)	(-.22)	(.16)
Age of Onset	.14	.22	.06	-.03	.23	-.02
(*n*=39)	(.25)	(.18)	(.09)	(.18)	(.28)	(-.05)
Pain	.36*	.14	**.49****	.41*	.05	-.29
(*n*=32)	(.33)	(.16)	(**.48****)	(.35)	(.09)	(-.30)
Severity	-.02	-.12	-.08	.06	-.14	.09
(*n*=43)	(-.16)	(-.06)	(-.15)	(-.23)	(-.18)	(.15)

Note: *$p < .05$. **$p < .01$. *p* values in bold indicate significance after Bonferroni corrections *p*=.0125. Values in brackets indicate partial correlations controlled for age group, gender and type of dystonia.

Hierarchical Multiple Regression Analyses of Anxiety and Total Difficulties Subscales

Hierarchical multiple regression analyses (Table 15) were conducted to identify significant independent predictors of anxiety (on the BYI) and total difficulties (on the SDQ child and parent versions) among young people with dystonia, once age group, gender and type of dystonia had been accounted for.

Age group, gender and type of dystonia did not explain a significant amount of variance in anxiety ($F(3,44) = 1.85$, $p=.15$; $R^2 = .11$, adjusted $R^2 = .05$). More importantly, self-concept contributed an increase in variance explained from 11% to 19%, adjusted $R^2 = .12$, a change that was significant ($F(1,43) = 4.33$, $p=.04$). In the final equation, only self-concept ($t(43) = -2.08$, $p=.04$) made a significant unique contribution to explaining anxiety, whereas age group ($t(43) = -0.45$, $p=.66$), gender ($t(43) = 1.09$, $p=.28$) and type of dystonia ($t(43) = 1.89$, $p=.07$) did not. This suggests that young people with dystonia who report negative self-concept are more likely to experience greater levels of anxiety than young people with dystonia who report a more positive self-concept.

Age group, gender and type of dystonia did not explain a significant amount of variance in self-reported total difficulties ($F(3,29) = 2.73$, $p=.06$; $R^2 = .22$, adjusted $R^2 = .14$). Self-concept contributed an increase in variance explained from 22% to 32%, adjusted $R^2 = .22$, a change that was a non-significant trend ($F(1,28) = 3.92$, $p=.058$). In the final equation, gender ($t(28) = 2.18$, $p=.04$) made a significant unique contribution to explaining total difficulties and there was a non-significant trend for

self-concept (t(28) = -1.98, p=.058), whereas age group (t(28) = 0.67, p=.51) and type of dystonia (t(28) = 1.62, p=.12) did not provide a significant unique contribution. This suggests that young people with dystonia who are female and/or report negative self-concept may be more likely to experience greater levels of emotional and behavioural difficulties than young people with dystonia who are male and/or report a more positive self-concept.

Age group, gender and type of dystonia did not explain a significant amount of variance in parent-reported total difficulties (F(3,26) = 0.93, p=.44; R^2 = .10, adjusted R^2 = -.01). Self-concept and pain contributed an increase in variance explained from 10% to 46%, adjusted R^2 = .35, a change that was significant (F(2,24) = 8.13, p< .01). In the final equation, type of dystonia (t(24) = -2.69, p=.01) and pain (t(24) = 3.72, p< .01) made significant unique contributions to explaining total difficulties, whereas gender (t(24) = -1.21, p=.24), self-concept (t(24) = -0.76, p=.45), and age group (t(24) = -1.11, p=.28) did not. This suggests that young people with secondary dystonia who experience high levels of pain are more likely to experience greater levels of emotional and behavioural difficulties than young people with primary dystonia who do not have high levels of pain.

Table 15: *Hierarchical Multiple Regression Analyses*

Subscale	Set A	Set B	R^2	Adjusted R^2	F Change	*p* value (F Change)	*t*	*p* value (Coefficients)
Anxiety (*n*=48)								
	Age Group		.11	.05	1.85	.15	-0.45	.66
	Gender						1.09	.28
	Type of Dystonia						1.89	.07
		Self-Concept	.19	.19	4.33	**.04**	-2.08	**.04**
SDQ Total Difficulties (*n*=33)								
	Age Group		.22	.14	2.73	.62	0.67	.51
	Gender						2.18	**.04**
	Type of Dystonia						1.62	.12
		Self-Concept	.32	.22	3.92	.06	-1.98	.06
SDQ-P Total Difficulties (*n*=31)								
	Age Group		.10	-.01	.93	.44	-1.11	.28
	Gender						-1.21	.24
	Type of Dystonia						-2.69	**.01**
		Self-Concept	.46	.35	8.13	**< .01**	-0.76	.45
		Pain					3.72	**< .01**

Note: Significant *p* values in bold.

Discussion

This section shall discuss the main findings in relation to the aims of the study and previous literature. It shall then outline the strengths and limitations of this study and the impact on results. Finally, clinical implications and areas for future research will be discussed.

Summary of Main Findings

Young people with dystonia self-reported higher levels of anxiety and peer problems, and lower levels of conduct problems and disruptive behaviour than population norms. Children with dystonia reported significantly lower levels of depression and anger than population norms, whereas for adolescents, there was no significant difference in depression and anger between those with dystonia and population norms. Young people with dystonia did not report significantly different levels of total difficulties, emotional problems, hyperactivity, or prosocial behaviour to population norms. Parents reported higher levels of total difficulties, emotional problems, peer problems and hyperactivity, and lower levels of prosocial behaviour, than population norms. Self-concept was a significant independent predictor of anxiety, and pain was a significant independent predictor of total difficulties (parent-reported). Age of onset and severity of movement disorder did not correlate with any of the emotional or behavioural problems measured in the present study.

Comparison of Dystonia Sample With Population Norms

Both children and adolescents reported higher levels of anxiety than population norms which supports the hypotheses of this research, and is consistent with previous findings in adults with dystonia and children with movement disorders (Demartini et al., 2017). Previous literature has associated anxiety with having complex medical needs which require regular hospital appointments, traumatic experiences, changing of employed assistants to care for the young person and concerns about how they are perceived by others (Bjørgaas, 2015; Mordin et al., 2014). An additional factor associated with greater anxiety in the present study was experience of DBS surgery. This could be due to stress caused by the surgery and follow-up appointments, unexpected complications, or worry about the future if symptoms have not resolved as anticipated (Austin, 2015).

Both self- and parent-rated levels of peer problems were higher than population norms, which supports the hypotheses of this research and previous literature on children with movement disorders (Yude, Goodman, & McConachie, 1998). These children experience more negative social experiences such as exclusion and bullying, in addition to reduced social opportunities due to their physical limitations (Parkes, McCullough, & Madden, 2010).

Both children and adolescents reported lower levels of disruptive behaviour than population norms. This is contradictory to the study's hypotheses and is inconsistent with previous literature (Ben-Pazi et al., 2011; Bjørgaas, 2015). One potential explanation is the use of the BYI to measure behaviour. Children who

cannot move independently were less likely to score highly on this measure due to the nature of the questions (for example “I skip school”). However, young people also self-reported lower levels of conduct problems, and parents reported no significant difference from population norms. High numbers of children with cerebral palsy have been found to have conduct problems, with one in seven reported to have oppositional defiance disorder (Bjørgaas, Hysing, & Elgen, 2012; Parkes et al., 2008), suggesting that the results for conduct problems are not in line with previous studies. Parents rated their child as being significantly higher on the hyperactivity scale than population norms. This finding supports previous literature showing that children with movement disorders are more likely to have ADHD (Bjørgaas et al., 2012).

Contradictory to the study’s hypotheses, children reported significantly lower levels of depression and anger, and adolescents did not differ significantly from the population norms. Whilst there is little in the adult literature regarding anger, these results (taken alongside the conduct problems and disruptive behaviour subscales) conflict with literature on childhood movement disorders. One possible explanation could be social desirability, as many participants required assistance to complete the questionnaires and may have wanted to appease the researcher (Van de Mortel, 2008). The majority of the adult literature suggests that people with dystonia have higher rates of depression than the general population (Fabbrini et al., 2011; Wenzel et al., 1998). For children with movement disorders, whilst there are several studies showing high rates of depression (Ben-Pazi et al., 2011; Koukouni et al., 2007), at least one study has found low rates (Bjørgaas, 2015). The results of this study

suggest that young people with dystonia are not more likely to have depression; however, it is difficult to determine whether other factors influenced results. It has been suggested that people with dystonia become increasingly aware of their difficulties and being 'different', leading to higher rates of depression as they become older (Bjørgaas, 2015). This may explain why adolescents had a higher mean score on the BYI depression subscale than children, and why higher rates of depression have been found in adult studies. Additional factors such as having dystonia from birth, attending a school to meet their specific needs, having an intellectual impairment and not having a sibling, may limit a child's awareness of how different they are from children without dystonia.

Children did not self-report significantly higher difficulties than population norms on any of the SDQ subscales, other than peer problems. It is unclear to what extent these results are in line with previous literature, because most studies on childhood movement disorders relied on parent data only (Brossard-Racine et al., 2013; Parkes et al., 2008). In the present study, parent-reported SDQ scores were significantly higher than population norms on all subscales, except conduct problems. The agreement ratings (intraclass correlation coefficients) reported in this study support these conflicting results as agreement was moderate at best for all subscales, except for the prosocial scale. Differences in perception of wellbeing between children and their parents has been found in previous studies (Janssen et al., 2010; White-Koning et al., 2007). There is evidence that children with cerebral palsy are more positive about their health-related quality of life than their parents

think they are (Janssen et al., 2010), this resilience may account for why young people with dystonia report fewer difficulties than parents report.

Relationships Between Biopsychosocial Factors and Mental Health and Behavioural Outcome Measures

The biopsychosocial variables explored in this study were self-concept, age of onset of dystonia, pain and severity of movement disorder. Each of these is discussed in turn.

Self-Concept

Results of the present study supported the hypothesis that young people with dystonia who have lower self-concept have higher levels of anxiety, as well as both self- and parent-rated total difficulties and emotional problems. However, self-concept was not significantly associated with depression, anger, or any of the behavioural subscales. Self-esteem has been associated with self-consciousness about visible movement symptoms, lack of participation in activities with other young people, and negative perception of the impact of their disability (Bøttcher & Dammeyer, 2013; Lees, 2002; Manuel et al., 2003). Self-esteem's association with anxiety supports previous literature on cerebral palsy, where scores on the State-Trait Anxiety Inventory were found to be highly associated with global self-esteem and self-efficacy (Borkowska, 2015). The lack of association between self-esteem and depression in this study contradicts the adult literature, where a study of 329

community based dystonia patients found that 56% of the variance in scores on the Beck Depression Inventory (BDI) was explained by scores on self-esteem measures (Lewis et al., 2008). This difference may be partly due to the participants in this study having higher rates of depression (30% moderately to severely depressed) than found in the present study.

Age of Onset of Dystonia

Contradictory to the hypothesis of the present study, age of onset of dystonia was not associated with any of the psychological or behavioural measures reported. This conflicts with previous studies associating lower age of onset of a movement disorder with greater severity of mental health problems (Peall et al., 2013; Schrag et al., 2003). These studies used self-reported (BDI) and clinician-rated measures of mental health difficulties, however did not report how age of onset was measured. It is therefore difficult to ascertain whether there are methodological differences that may account for conflicting findings. Alternatively, age of onset may not be associated with mental health problems in dystonia; some of the adult literature has not found an association (Lewis et al., 2008; Page et al., 2007), and therefore there may not be an association between childhood dystonia and age of onset.

One possible reason for not observing the associations between age of onset and mental health difficulties seen in adult literature, is that the possible range for age of onset is much smaller in this study. In adult literature, the range of age of onset can be 18-66 years (Balint & Bhatia, 2014). It may be that age of onset is an

important factor when comparing young- verses adult-onset dystonia, where there is wider variability in age range in comparison to just children and adolescents. Another possible reason is the difficulty in establishing an accurate age of onset of dystonia in a population of young people. Many parents reported difficulty in specifying an age, as dystonia can often begin with subtle symptoms that may not be spotted initially or be attributed to dystonia specifically. This was especially the case for young people with secondary dystonia and/or comorbid health problems. Many parents reported that it took several years to get a diagnosis and therefore it was difficult to remember when clear symptoms of dystonia began, this was particularly the case for children who were not born with dystonia.

Pain

Pain was found to be moderately correlated with anger, child-rated prosocial behaviour, and parent-rated total difficulties, conduct problems and hyperactivity. In addition, levels of depression and disruptive behaviour were higher for young people who experienced moderate or severe pain in comparison to those who experienced no pain or mild levels of pain. This supports previous research on children with movement disorders, where difficulties with sleep and difficulties participating in activities with peers have been suggested as possible explanations for the influence that pain has on mental health and behaviour (Parkes et al., 2008; Ramstad et al., 2012; Russo, Miller, Haan, Cameron, & Crotty, 2008).

It is possible that pain was associated more frequently with subscales of the SDQ-P because both the PPP and SDQ-P are rated based on parents' observations of their child's behaviour. Child pain has been found to influence parental reports of quality of life in a study of children with cerebral palsy (White-Koning et al., 2007); this may explain why there was a significant association between pain and parent-rated total emotional and behavioural difficulties, but not between pain and child-rated total emotional and behavioural difficulties.

Severity of Movement Disorder

The severity of movement disorder was not significantly associated with any of the psychological or behavioural measures. This conflicts with previous studies conducted with adults with dystonia and children with cerebral palsy (Ben-Shlomo et al., 2002; Tomic et al., 2016; van Schie et al., 2013) which found that higher levels of disability were associated with emotional state and social functioning. One potential reason for this discrepancy is the measure used to assess severity. Whilst the GMFCS-E&R is used clinically for assessing gross motor function in dystonia (Elze et al., 2016), the measure was designed for use in cerebral palsy and does not capture other areas of disability such as fine motor movements or communication. Difficulties in these areas can have a large impact on a person's ability to carry out everyday activities and therefore may influence mental health and behaviour.

An alternative explanation is that dystonia severity does not influence mental health and behavioural problems. Several studies have not found severity to be an

influencing factor on mental health (Fabbrini et al., 2010; Heiman et al., 2004). Another study found that children who needed total assistance with moving about had lower parent-rated SDQ total difficulties scores than children who were able to move without assistance (Parkes et al., 2008). Contradictory findings may be, in part, due to conflicting interpretation of severity; for example, one study found that severity of disability had an impact on quality of life, but not severity of motor symptoms themselves (Smit, Kuiper, et al., 2016). This is an area which warrants future research.

Strengths and Limitations

The present study had some limitations that may have affected results. The sample size was smaller than intended due to the availability of eligible participants being lower than expected, and practical issues around researcher availability. As the study was underpowered for the regression analyses and correlations, there is the possibility that type 2 errors occurred where non-significant results were found. Additionally, most results which were significant did not remain so after Bonferroni correction; therefore, the conclusions drawn from this study are offered tentatively due to the possibility of type 1 errors. Nonetheless, the present study was small and exploratory in nature, and provides useful and novel information regarding possible relationships between biopsychosocial factors and mental health and behavioural outcomes among young people with dystonia.

In addition to the small overall sample size, there was also a substantial amount of missing data. The PPP could not be completed in cases where the child did not have pain. This reduced the number of participants available for statistical comparisons using this variable and affected the size of the correlation coefficient needed to detect a significant effect. In addition, the GMFCS-E&R was estimated retrospectively for 12 of the participants due to the medical records not containing details of a full assessment. It is possible that these issues influenced the validity of the results. The age ranges for the population norms caused some restrictions in the data analysis. Splitting up the sample into children and adolescents did enable some observations about differences between ages to be made but meant that the sample size was small for some comparisons which led to a reduction in power, particularly for the adolescent group.

Measurement tools in research with children have been criticised as children often do not fill them out themselves (Janssen et al., 2010), a strength of this study is that every child completed the BYI and therefore was able to self-report psychological and behavioural difficulties. Both the child and parent versions of the SDQ were used in this study; however, self-reported data were only collected from those aged over 11 years. As there are differences between the children and adolescents on the BYI, this suggests that valuable information may have been missed by not being able to give the SDQ to younger participants.

This study had broad inclusion criteria for results to be as generalisable to young people with dystonia as possible; however, recruiting from a hospital, and the high numbers of young people who had DBS surgery or could not take part due to

capacity or communication issues, limits the generalisability of results. Whilst the high number of comorbidities is more reflective of the actual dystonia population than only including participants with primary dystonia, the presence of comorbidities makes it difficult to exclude possible confounding factors from conclusions.

Implications of Findings

Based on the findings of the present study it is possible to make tentative suggestions regarding clinical implications, whilst bearing in mind that these findings require replication in a larger sample before any firm conclusions can be drawn.

Clinical Implications

These results suggest that mental health screening (particularly for anxiety) could be useful in neurology, movement disorder and Botox clinics. The BYI depression and anxiety subscales were highly correlated, suggesting that children with higher levels of anxiety may be at risk of depression if it is not addressed. It is also important to consider parents viewpoints in terms of difficulties their child is having but may not be aware of. As 12 out of 47 young people answered 'sometimes', 'often' or 'always' for the question "I wish I was dead", this highlights a need for clinicians to be aware of difficulties with mood and suicidal ideation in this population. It is important to consider that these children have a long-term health condition and may have to withstand painful and risky procedures; they therefore may think about death more frequently than a typical child. Even so, it is important

for clinicians to be aware of the significant distress that children can be under when being treated for a physical health problem, and ensure the appropriate support is offered particularly during times of painful or invasive treatment.

The lack of relationship between age of onset of dystonia and psychological and behavioural variables in this study suggests that both having a diagnosis from birth and being diagnosed as an older child or adolescent have their own challenges, and no age is more likely to have a psychological or behavioural problem. Similarly, the results suggest that young people who are not able to walk independently, and young people whose dystonia is more difficult to detect, are equally likely to have psychological or behavioural difficulties. The results indicate that interventions targeting self-esteem and pain could be useful for young people with dystonia. Targeting self-esteem may help to reduce anxiety. Play therapy and compassion-focused therapy (CFT) have been used with young people to help self-esteem (Carona, Rijo, Salvador, Castilho, & Gilbert, 2017; Post, 1999). Targeting pain may help reduce depression, anger and behavioural problems. This could be via a psychological intervention such as cognitive-behavioural therapy (CBT) or relaxation, which have been found to be useful in treating chronic pain in children and adolescents (Eccleston, Morley, Williams, Yorke, & Mastroyannopoulou, 2002).

Implications for Future Research

This study found that people with dystonia are more likely to have higher levels of anxiety than people without dystonia. As clinically there are many different

anxiety diagnoses, future research could explore the nature of anxiety further. Considering the high levels of difficulties with peers which were reported, social anxiety could be of interest. This would be in line with previous research showing that adults with dystonia have high levels of social anxiety (Dias et al., 2011). Further exploration into the variables associated with anxiety, and how they relate to each other could provide information that may be helpful for interventions. As in adult studies, only a small range of mental health difficulties have been explored in the present study. It is worth considering that areas that are not widely researched in adults (for example psychosis, somatization, eating disorders, agoraphobia and panic disorder), are worth exploring in adults and young people.

The theoretical basis for self-esteem being a protective factor for mental health problems was that identifying with others who have similar difficulties can give a sense of belonging which is related to lower levels of mental health problems (Bogart, 2015). In the present study, anxiety (and total difficulties and emotional problems) was related to self-concept; however, factors that might be related to identify (such as peer problems and prosocial behaviour) were not. Future research could explore identity further, by seeing if identifying with other people with dystonia and/or other physical disabilities is associated with fewer mental health or behavioural difficulties (particularly with anxiety and peers). This would be in line with research on neurodevelopmental disorders (Crabtree, Haslam, Postmes, & Haslam, 2010).

As several results were not as hypothesised or in line with previous literature and may have been impacted by measures or small sample size, it would be

important to explore these further. The low incidence of depression is contradictory to many studies of adult dystonia and childhood movement disorders, and therefore warrants further research. Additional factors could be explored, such as whether the child goes to a school which meets their specific needs. Despite conflicting literature on the relationship between mental health and severity of movement disorder, it would be important to explore this relationship further in childhood dystonia. Future research could use a greater range of measures of severity; for example, measures assessing for communication and fine motor skills in addition to gross motor skills, particularly as communication problems are significantly related to psychiatric disorders (Bjørgaas et al., 2012).

It was considered important to access the viewpoint of the child in this study; therefore, valid and reliable child measures were used that would give as much information about mental health and behaviour as possible. However, one limitation of this study, was the inaccessibility of the questionnaires for many children who had learning difficulties. This was particularly noticeable for younger children, as older children and adolescents more often had a level of understanding that enabled them to access the questionnaires despite problems with learning. Forty-four percent of the final sample had learning needs, this reflects the high number of children with dystonia who have problems with learning. This has been problematic in previous studies, where children with intellectual disabilities could not be diagnosed with a psychiatric disorder due to the severity of their physical or mental condition (Bjørgaas et al., 2012). Eleven children could not complete the study due to difficulty understanding the questionnaires, and future research could use alternative means

to access the emotional and behavioural states of these children, to have more of an understanding of the needs of all young people with dystonia.

Conclusions

Overall, young people with dystonia were found to have greater difficulties with anxiety and peers, and fewer difficulties with disruptive behaviour than population norms. Parents reported a greater range of emotional and behavioural difficulties than children. Self-concept was found to be a significant independent predictor of anxiety, and pain was found to be a significant independent predictor of total difficulties (parent-reported). Psychological interventions targeting self-esteem and pain may therefore be helpful for this population. These results should be interpreted with caution due to methodological issues such as the small sample size and missing data leading to insufficient power, and age-restrictions of published norms. Future research should investigate anxiety disorders and depression further in a larger sample, in addition to factors influencing these such as identity.

Integration, Impact and Dissemination Plan

Integration

This section aims to provide an overview of the whole research process. This includes critical evaluation and reflection on the process of undertaking the Systematic Review and the Empirical project, including any challenges and decisions made along the way.

Comparisons Between the Systematic Review and the Empirical Paper

Design

The aim of the Systematic Review was to determine if adults with dystonia were more likely to have mental health problems than adults without dystonia. It was therefore important to only include studies which used a control group. Similarly, it was considered important that a control group was included in the Empirical study to help services designed to support people with dystonia gain a better understanding of their mental health. For the Empirical study, there was limited time and resources available for recruitment. As two of the adult studies had included pre-existing data from population studies, it was considered appropriate for the Empirical study to use population norms as a control group.

One difficulty with using norms is the lack of control over who is included in the study. The choosing of inclusion and exclusion criteria and matching participants

in the adult studies, helps the dystonia and control groups be more comparable. In the Empirical study however, there may be differences between the dystonia and population norms that cannot be controlled for. Despite this, using population norms ensured the dystonia sample was compared to a large, varied sample. This was more appropriate for the Strengths and Difficulties Questionnaire (SDQ) as it used UK norms, whereas the Beck Youth Inventories (BYI) used an American sample where there may be cultural variations.

Inclusion and Exclusion Criteria

The inclusion and exclusion criteria for the Systematic Review did not pose restrictions on type of dystonia (except for tardive dyskinesia), however some of the individual studies did pose more strict criteria (such as only including primary dystonia, or one specific subtype). The Empirical study had broad criteria as it was the first to explore mental health and behaviour in childhood dystonia in general, therefore it was important for criteria to be as inclusive as possible.

Measures

The studies included in the Systematic Review incorporated a large range of mental health problems, however most of the significant comparisons were for depression and anxiety. As this was the first study exploring mental health in young people with dystonia, depression and anxiety were also the focus for this study.

Most of the included studies in the Systematic Review used clinical interviews. Whilst clinical interviews such as the Kiddie Schedule for Affective Disorders and Schizophrenia (K-SADS) are widely used in research, it was not possible to use these in the Empirical study due to time and resources available. Self-report measures were found to be a good alternative in a similar study of childhood cerebral palsy (Bjørgaas, 2015) and were therefore used in the Empirical study.

What I Gained from the Systematic Review Process

One advantage of carrying out the Systematic Review was that it helped me understand the vast number of health conditions that use the term 'dystonia'. This gave me insight into which participants were suitable for the Empirical study. For those with a comorbid condition, it helped me understand what difficulties may be due to dystonia and what may be due to other conditions. The Systematic Review highlighted that many mental health difficulties aside from depression and anxiety have not been explored extensively; this helped me reflect on what avenues would be worthwhile for future research to help meet the mental health needs of this population.

Carrying out the Systematic Review highlighted that few studies have a control group, report specific data on mental health (as opposed to how it impacts other factors, such as quality of life), or are of high quality (with a large, generalisable dystonia sample). This made the use of a control group, detailed reporting of mental health and behavioural data, and the Empirical study being of a

high quality, more important. One final benefit was the process of synthesising a lot of data in the Systematic Review helped me organise and process my own data. This was particularly helpful when data conflicted and helped me make meaningful conclusions from it.

Impact

Immediate Impact of the Study

The largest immediate impact of the study was that some of the young people who took part were not currently receiving psychological support yet scored highly on some of the measures or said 'sometimes', 'often' or 'always' to the "I wish I was dead" question. This resulted in conversations between myself, the Clinical Psychologist supervising the project, and the Consultant in charge of the young person's care to decide how best to support the child. In some cases, this resulted in the child being offered support from the Clinical Psychologist or being referred to the local Child and Adolescent Mental Health Service (CAMHS). In addition, the Consultant received a letter for every young person that took part in the study, with a summary of their scores and any risk information. Consultants were encouraged to discuss these letters with participants, thus considering their psychological wellbeing as part of their routine clinical care. It is hoped that this will have a knock-on effect for other young people the Consultants see.

Clinical Impact of the Study

Increasing Awareness About Mental Health Needs

Results from the study suggest that mental health screening in clinics which support young people with dystonia may be a vital way of gaining information that might otherwise be missed. Whilst it is important for clinicians to have face-to-face conversations about wellbeing, the NHS is continually stretched and often there is not time in routine appointments (which can be as short at 15 minutes long) to ask everyone about their wellbeing beyond their motor symptoms and side effects of any medications. Using a questionnaire, such as the SDQ, is a quick way to gain a lot of valuable information that might be missed, particularly as parents or young people may not know where to discuss psychological, social, or behavioural difficulties. In addition, clinicians may not prioritise asking about wellbeing when a family appears to be coping well.

Anxiety was seen in nearly half of children. As most adult anxiety disorders are preceded by anxiety disorders experienced in childhood (Martin, 1998), it is important to address anxiety in childhood in order to protect against anxiety disorders in later life. Similarly, many participants identified difficulties with peers, which can impact mental health in adulthood (Lereya, Copeland, Costello, & Wolke, 2015).

Whilst young people reported fewer or no differences to population norms on depression, anger, and disruptive behaviour; parents highlighted difficulties in most psychological and behavioural areas. A quarter of participants answered

'sometimes', 'often' or 'always' to "I wish was dead". Whilst not all respondents were actively suicidal or depressed, many young people with dystonia have had this thought, and it is important that this is known by those close to them. Clinicians asking about mental health and suicidality sets a precedent that it is acceptable to talk about difficult feelings, and that medical appointments are an appropriate place to have these discussions.

For young people, an indicator of mental health difficulties is often their behaviour. Children who are not disruptive may be missed in a family or school environment where more subtle signs of a mental health difficulty may not be noticed. This research highlights the high prevalence of anxiety, and the need for discussions around mental health to be had with all children, before difficulties become a longer-term problem.

This study has highlighted who might be more likely to have mental health or behavioural difficulties, such as those who have difficulties with peers, have had deep brain stimulation surgery (DBS), have learning difficulties, are female, have a comorbid health problem, have lower self-esteem, or experience high levels of pain. In addition, it highlights that the age of onset and severity of movement disorder do not make young people more likely to have a mental health problem. However, as this was an exploratory study which lacked power due to small sample size, the possibility of type 1 and type 2 errors cannot be ruled out. Replication in a larger sample is therefore warranted to support these conclusions.

Increasing Interventions for Mental Health Difficulties

This research has highlighted what avenues for intervention may be useful. Poorer self-concept was associated with greater anxiety. This suggests that interventions for improving self-esteem may be useful to reduce anxiety in people with dystonia. One way to target this is through play therapy, where children use toys to express their feelings, needs, and experiences. Play therapy has been found to prevent at-risk children from developing low self-esteem (Post, 1999), and to improve self-esteem, anxiety, and depression (Baggerly, 2004). This intervention could be particularly useful for children who have learning needs or difficulty communicating (Landreth, 1987). For adolescents, compassion-focused therapy (CFT) may be useful to help decrease shame and self-criticism, and increase the young person's ability to access and tolerate their emotions (Gilbert, 2014; Leaviss & Uttley, 2015). Targeting self-esteem may also have an impact on peer relationships and prosocial behaviour.

Higher levels of pain were found to be associated with higher levels of anger and behavioural problems. Psychological interventions aimed at reducing pain include cognitive behavioural therapy (CBT), which incorporates biofeedback (the use of monitoring devices to help gain control over physiological processes) and relaxation training (Eccleston et al., 2014). A meta-analytic review of psychological therapies for young people with chronic pain found evidence that CBT can lead to a reduction in disability and improve emotional functioning (Palermo, Eccleston, Lewandowski, Williams, & Morley, 2010). Whilst not all children will be able to

access CBT, some of the techniques could be adapted for younger children, such as guided imagery or relaxation (Ball, Shapiro, Monheim, & Weydert, 2003).

Psychological interventions could be offered via Clinical Psychologists or other mental health professionals attached to the teams working with children within hospital settings, or can be offered by the young person's local CAMHS. For a child to receive an intervention, staff working closely with these children (i.e., Consultants, Nurses, Physiotherapists, Speech Therapists) need to be able to assess whether a child would benefit from such support and know how to access appropriate support. Research such as this further reinforces the need for mental health professionals to work in parity with physical health services, and policies and funding need to account for this (Parkin & Powell, 2017).

Research Impact of the Study

This research has supported findings of previous studies which have shown that children with movement disorders are more likely to have higher levels of anxiety (Bjørgaas, 2015), peer problems (Parkes et al., 2008) and hyperactivity (Tate, Allison, Pranzatelli, & Verhulst, 2005). However, the present study has not replicated results from previous studies showing relationships between dystonia and higher levels of depression (Ben-Pazi et al., 2011), disruptive behaviour (Yude et al., 1998) and conduct problems (Parkes et al., 2008). As this was the first study exploring these variables in childhood dystonia, further investigation is needed in a larger sample, to establish the nature of these relationships.

The present study differed from previous studies because both child and parent data were collected. Children did not self-report significantly higher difficulties than population norms on any of the SDQ subscales other than peer problems, whereas parents reported them to have higher levels of total difficulties, emotional problems and hyperactivity, and lower levels of prosocial behaviour. Whilst the parent data were similar to previous studies of young people with cerebral palsy (Brossard-Racine et al., 2013; Parkes et al., 2008; Tate et al., 2005), ascertaining the viewpoint of the child is important. Future research should use child as well as adult reported data. One reason for this is because learning difficulties were found to be related to anxiety. This really highlights the need to consider the viewpoint and wellbeing of children with severe communication difficulties and learning needs, who may not be able to communicate their emotional difficulties to parents. Further development of suitable measures to access these children is necessary.

The present study has reported findings that indicate future avenues for research. Anxiety was reported for nearly half of young people. Future research could investigate if the presence of anxiety translates to clinically diagnosable mental health problems, such as OCD, separation anxiety, or specific phobias. Social anxiety may be a particularly common difficulty for these young people considering the high levels of difficulties with peers, and low levels of prosocial behaviour reported.

Personal Impact of the Study

As a person who has not previously worked with young people who have a health condition or a disability, I was completely in awe of the strength of these young people and their families. I thoroughly enjoyed working with the young people who enthusiastically answered so many questions, despite them being of a sensitive nature. I met young people who were in wheelchairs, found it difficult to use their hands and communicate, yet they could demonstrate an amazing sense of humour during the process of completing the questionnaires.

There were several instances where working with these young people was quite distressing, for example, when seeing a child cry due to severe pain. One child told me he thought about dying, and looked ashamed to say so. It was incredibly moving when his father told him it was "OK" to feel that way, and that he had also had those thoughts in the past. This highlighted to me how important it is that the difficult questions are asked, as the door had been opened for this father and son to talk about suicidal thoughts (between themselves and with their clinical team as this information was shared according to the hospital's safeguarding policy), and to support each other in the future.

Parents were very willing to tell their stories, which included experiences such as the often difficult and lengthy process of receiving a diagnosis for their child and the everyday difficulties that they face, but more so praise about the wonderful staff they had worked with in hospital and school settings, and the achievements and progress that their child had made. Many parents reflected on the resilience of

their child and spoke about the joy of parenting them. I felt that many of the children and their parents had an incredibly close relationship, which I noticed when parents would translate their child's answer if it was unclear due to communication issues. I felt that it was a shame that these stories were lost due to the study being of a quantitative nature. Future research could aim to better understand the narratives around coping with and adjusting to living with dystonia.

This experience taught me that, whilst these children are facing difficult circumstances, the support children receive was often extensive, with a parent becoming a full-time carer, or the child going to a school which met their needs. This is likely a large protective factor for these children and may be why many younger children scored significantly lower on measures of mental health and behavioural difficulties than the population norms. I also learned that, despite such extensive difficulties, many of these young people could access activities that other young people access, such as sports, youth clubs, and social media. It was lovely to have children teach me about their interests. For a subject area which was expected to reveal many difficulties, it was nice to learn that many young people and their parents are coping very well. The process of doing this thesis has encouraged me to work more with children who have health problems or neurodevelopmental difficulties. I have seen how much a child can thrive in the right context despite difficult circumstances.

Dissemination

Participants

Participants have access to their individual scores via their Consultant. All participants will receive a summary of the study's main results. This will include the percentages of young people who scored above clinical cut-off on the various scales, and information on how young people with dystonia differ from typical young people their age. The factors that appear to be linked to higher levels of mental health difficulties will be highlighted (i.e., being female, having DBS surgery, learning difficulties, low self-esteem, high pain levels and peer problems) in addition to how other factors do not appear to be linked (i.e., age of onset of dystonia and severity of movement disorder).

It will be highlighted that this was the first study to look at mental health difficulties in young people with dystonia, and that further research is needed to support these findings, provide more information on factors associated with them, and evaluate potential interventions. In addition, it will be stressed that having one or multiple factors associated with these difficulties does not mean a person will have or develop a mental health or behavioural problem, and conversely that not having these factors means that a person will not have any difficulties.

Parents will be encouraged to share this research with the healthcare professionals they work with and the schools their children attend. To help parents feel as though they can benefit from this information, it will be suggested that developing self-esteem, peer relationships, and managing pain may be useful steps

to help support their child. They will be encouraged to speak to a member of their medical team if they wish to have support with this. Hopefully, young people reading this information may feel that their needs are being heard, and parents may be more aware of difficulties their children may face and how to support them. It is hoped that this will increase conversations about mental health to address difficulties earlier on. This may reduce the incidence of difficulties that occur in adulthood.

The Evelina London Children's Hospital

This research will be presented to the Evelina London Children's Hospital Complex Movement Disorder Service, who supported much of this research. They are keen to have information on the mental health needs of this population and to be aware of any additional screening or support they could be offering routinely if there is a need for it. The high levels of anxiety and the factors associated with it (i.e., having DBS surgery, learning difficulties and low self-esteem) will be shared.

It will also be highlighted that a quarter of young people have thoughts about death, and therefore it is important to normalise these thoughts by asking questions about difficult thoughts and emotions in medical appointments and to monitor risk. As parents report more difficulties than young people, it might be appropriate to direct questions regarding behavioural difficulties to parents. Factors such as pain and peer problems will also be suggested as avenues clinicians may wish to explore with patients. It is hoped that overall, clinicians will feel more confident in knowing what risk signs to look out for and discussing mental health with patients.

It will be suggested that screening measures could be handed out in waiting rooms before routine clinical appointments, to capture child and parent viewpoints quickly and to encourage the perception that medical appointments are an appropriate place to discuss mental health. The development of a new measure or use of an existing one may help in the detection of young people who are struggling but may not have spoken to anyone or be showing obvious signs. Scores could also be monitored over time, particularly for children who may be more likely to have a mental health or behavioural difficulty.

Wider Dissemination

It is intended that this research is published in the Journal of Neurology, Neurosurgery & Psychiatry (impact factor=7.35), this would hopefully reach other teams who work with young people with dystonia for them to be aware of what mental health problems to look out for and address. Also, other researchers conducting research into movement disorders and mental health may be able to build on the findings reported here. Other journals which may publish the data include: The European Journal of Paediatric Neurology; The Journal of Clinical Movement Disorders; Parkinsonism & Related Disorders, and; Journal of Neurology; Movement Disorders.

Attending conferences would help disseminate the research to researchers and clinicians interested in movement disorders and mental health. The data could be presented at events such as: The International Dystonia Symposium; The British

Paediatric Neurology Association Annual Conference; The Parkinson's Disease and Other Movement Disorders Conference, or; the Cognitive Neuroscience Society Annual Meeting.

The Dystonia Society is a national charity which provides support, advocacy and information for anyone affected by dystonia. Their website has a research page and a children page, which information from this study could be added to. This would enable more widespread dissemination of this research, for example to schools, medical teams, and young people with dystonia and their parents who may not receive as much intensive support from schools or hospitals. This information could offer guidance and support for those who are concerned about a child's mental health or behaviour. Conversely, due to young people with dystonia having lower levels of difficulty in some areas, this information may provide some comfort for people to know that a diagnosis of dystonia does not necessarily result in a mental health problem, and that children may be more resilient than their parents think. Being aware of such information could help schools make efforts to help young people with dystonia integrate with peers.

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Appendices

Appendix 1: The Newcastle-Ottawa Scale (Wells et al., 2013)

CODING MANUAL FOR CASE-CONTROL STUDIES

SELECTION

1) Is the Case Definition Adequate?

a) Requires some independent validation (e.g. >1 person/record/time/process to extract information, or reference to primary record source such as x-rays or medical/hospital records) ☆
b) Record linkage (e.g. ICD codes in database) or self-report with no reference to primary record
c) No description

2) Representativeness of the Cases

a) All eligible cases with outcome of interest over a defined period of time, all cases in a defined catchment area, all cases in a defined hospital or clinic, group of hospitals, health maintenance organisation, or an appropriate sample of those cases (e.g. random sample) ☆
b) Not satisfying requirements in part (a), or not stated.

3) Selection of Controls

This item assesses whether the control series used in the study is derived from the same population as the cases and essentially would have been cases had the outcome been present.

a) Community controls (i.e. same community as cases and would be cases if had outcome) ☆
b) Hospital controls, within same community as cases (i.e. not another city) but derived from a hospitalised population
c) No description

4) Definition of Controls

a) If cases are first occurrence of outcome, then it must explicitly state that controls have no history of this outcome. If cases have new (not necessarily first) occurrence of outcome, then controls with previous occurrences of outcome of interest should not be excluded. ☆
b) No mention of history of outcome

COMPARABILITY

1) Comparability of Cases and Controls on the Basis of the Design or Analysis

A maximum of 2 stars can be allotted in this category

Either cases and controls must be matched in the design and/or confounders must be adjusted for in the analysis. Statements of no differences between groups or that differences were not statistically significant are not sufficient for establishing comparability. Note: If the odds ratio for the exposure of interest is adjusted for the confounders listed, then the groups will be considered to be comparable on each variable used in the adjustment.

There may be multiple ratings for this item for different categories of exposure (e.g. ever vs. never, current vs. previous or never)

Age =☆ , Other controlled factors = ☆

EXPOSURE

1) Ascertainment of Exposure

Allocation of stars as per rating sheet

2) Non-Response Rate

Allocation of stars as per rating sheet

Appendix 2: Ethical Approval Documents

a. Doctorate in Clinical Psychology Research Committee Project Approval

Memorandum

To:	Lauren Bates
From:	Andy MacLeod (on behalf of the Research Sub-Committee and Course Executive)
Date:	3rd February 2017
Copy To:	Jess Kingston
Re:	Main Research Project Proposal

The Research Sub-Committee has considered your Main Research Project Proposal response and has decided to give you **Approval**. Your research costs have also been approved. Please note that if these costs change and you do not re-submit an amended form for approval prior to incurring any additional costs, these additional costs will not be reimbursed.

- Rules of thumb should not guide calculations on the number of participants needed. A proper power analysis with 5 predictor variables indicates 91 participants to detect a medium effect size. Sixty may be more practically feasible for you but you should be clear about what power that sample gives you.

- You acknowledge your study does not inform causality but you need to be vigilant about causal language: in responding to Reviewer 1 Point 5 you talk about your regression testing whether mental health difficulties are "impacted" by …

Systematic review – approved.

The title should probably not be about "prevalence" given that prevalence studies will only be one source of information and it has a strict meaning.

Now that you have received approval it is time to apply for ethics. Please keep Annette informed and where possible provide copies of all applications, letters and approvals. Also, please ensure that if RHUL is your sponsor, Annette is sent all participant signed consent forms.

b. London-Dulwich Research Ethics Committee Project Approval

NHS
Health Research Authority
London - Dulwich Research Ethics Committee
Health Research Authority
Skipton House
80 London Road
London
SE1 6LH

Telephone: 020 7972 2561

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

07 August 2017

Miss Lauren Bates
Trainee Clinical Psychologist
Camden and Islington NHS Foundation Trust
Doctorate in Clinical Psychology, Department of Psychology
Royal Holloway, University of London
Egham
TW20 0EX

Dear Miss Bates

Study title:	**Mental Health and Behaviour in Childhood Dystonia**
REC reference:	**17/LO/1160**
IRAS project ID:	**224326**

Thank you for your letter responding to the Committee's request for further information on the above research and submitting revised documentation.

The further information has been considered on behalf of the Committee by the Chair.

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 opinion letter. Should you wish to provide a substitute contact point, require further information, or wish to make a request to postpone publication, please contact hra.studyregistration@nhs.net outlining the reasons for your request.

Confirmation of ethical opinion

On behalf of the Committee, I am pleased to confirm a favourable ethical opinion for the above research on the basis described in the application form, protocol and supporting documentation as revised, 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 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 within 6 weeks of recruitment of the first participant (for medical device studies, within the timeline determined by the current registration and publication trees).

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

NHS 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" below).

Non-NHS sites

Approved documents

The final list of documents reviewed and approved by the Committee is as follows:

Document	*Version*	*Date*
Confirmation of any other Regulatory Approvals (e.g. CAG) and all correspondence [Initial Project Proposal]	1	
Covering letter on headed paper [REC response letter]	1	03 August 2017
Evidence of Sponsor Insurance or Indemnity (non NHS Sponsors only) [Indemnity Insurance]	1	15 June 2017
GP/consultant information sheets or letters [Letter to Neurologist]	1	15 June 2017
IRAS Application Form [IRAS_Form_23062017]		23 June 2017
Letter from funder [Cost Approval from RHUL REC]	1	03 February 2017
Non-validated questionnaire [Demographic Questionnaire (parents)]	1	15 June 2017
Participant consent form [Assent form (young people) V2]	2	27 July 2017
Participant consent form [Assent form (young people) V2 track changes]	2	27 July 2017
Participant consent form [Consent form (young people) V1]	1	27 July 2017
Participant consent form [Consent form (parent) V2]	2	27 July 2017
Participant consent form [Consent form (parent) V2 track changes]	2	27 July 2017
Participant information sheet (PIS) [PIS young person (assent) V2]	2	27 July 2017
Participant information sheet (PIS) [PIS young person (assent) V2 track changes]	2	27 July 2017
Participant information sheet (PIS) [PIS young person (consent) V1]	1	27 July 2017
Participant information sheet (PIS) [PIS parent V2]	2	27 July 2017
Participant information sheet (PIS) [PIS parent V2 track changes]	2	27 July 2017
Referee's report or other scientific critique report [Initial Project Proposal]	1	17 November 2016
Referee's report or other scientific critique report [Proposal Response from RHUL REC]	1	16 December 2016
Referee's report or other scientific critique report [Feedback to RHUL REC]	1	27 January 2017
Referee's report or other scientific critique report [Project Approval from RHUL REC]	1	03 February 2017
Referee's report or other scientific critique report [Correspondence with RHUL REC re. project changes]	1	19 June 2017
Referee's report or other scientific critique report [Summary of Proposal Changes for RHUL REC]	1	15 June 2017
Research protocol or project proposal [Research Protocol]	2	27 July 2017

Summary CV for Chief Investigator (CI) [Chief Investigator CV (Lauren Bates)]	1	22 June 2017
Summary CV for student [Chief Investigator CV (Lauren Bates)]	1	22 June 2017
Summary CV for supervisor (student research) [Supervisor CV (Sarah Rudebeck)]	1	22 June 2017
Summary CV for supervisor (student research) [Supervisor CV (Jessica Kingston)]	1	22 June 2017
Validated questionnaire [Strengths and Difficulties Questionnaire (young person)]	1	15 June 2017
Validated questionnaire [Strengths and Difficulties Questionnaire (parent)]	1	15 June 2017
Validated questionnaire [Paediatric Pain Profile (parents)]	1	15 June 2017
Validated questionnaire [Gross Motor Function Classification System Expanded and Revised (medical records)]	1	15 June 2017

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

We are pleased to welcome researchers and R&D staff at our training days – see details at http://www.hra.nhs.uk/hra-training/

17/LO/1160	Please quote this number on all correspondence

With the Committee's best wishes for the success of this project.

Yours sincerely

pp
Dr Michael Philpot
Chair

Email:nrescommittee.london-dulwich@nhs.net

Enclosures: "After ethical review – guidance for researchers"

Copy to: *Ms Annette Lock*
Mariusz Marcinkowski, Guys and St Thomas's NHS Foundation Trust

c. Health Research Authority Project Approval

NHS
Health Research Authority

Miss Lauren Bates
Trainee Clinical Psychologist
Camden and Islington NHS Foundation Trust
Doctorate in Clinical Psychology, Department of Psychology
Royal Holloway, University of London
Egham
TW20 0EX

Email: hra.approval@nhs.net

07 August 2017

Dear Miss Bates

Letter of HRA Approval

Study title:	**Mental Health and Behaviour in Childhood Dystonia**
IRAS project ID:	**224326**
REC reference:	**17/LO/1160**
Sponsor	**Royal Holloway University of London**

I am pleased to confirm that HRA Approval has been given for the above referenced study, on the basis described in the application form, protocol, supporting documentation and any clarifications noted in this letter.

Participation of NHS Organisations in England

The sponsor should now provide a copy of this letter to all participating NHS organisations in England.

Appendix B provides important information for sponsors and participating NHS organisations in England for arranging and confirming capacity and capability. **Please read *Appendix B* carefully**, in particular the following sections:

- *Participating NHS organisations in England* – this clarifies the types of participating organisations in the study and whether or not all organisations will be undertaking the same activities
- *Confirmation of capacity and capability* - this confirms whether or not each type of participating NHS organisation in England is expected to give formal confirmation of capacity and capability. Where formal confirmation is not expected, the section also provides details on the time limit given to participating organisations to opt out of the study, or request additional time, before their participation is assumed.
- *Allocation of responsibilities and rights are agreed and documented (4.1 of HRA assessment criteria)* - this provides detail on the form of agreement to be used in the study to confirm capacity and capability, where applicable.

Further information on funding, HR processes, and compliance with HRA criteria and standards is also provided.

It is critical that you involve both the research management function (e.g. R&D office) supporting each organisation and the local research team (where there is one) in setting up your study. Contact details and further information about working with the research management function for each organisation can be accessed from www.hra.nhs.uk/hra-approval.

Appendices

The HRA Approval letter contains the following appendices:

- A – List of documents reviewed during HRA assessment
- B – Summary of HRA assessment

After HRA Approval

The document "*After Ethical Review – guidance for sponsors and investigators*", issued with your REC favourable opinion, gives detailed guidance on reporting expectations for studies, including:

- Registration of research
- Notifying amendments
- Notifying the end of the study

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

In addition to the guidance in the above, please note the following:

- HRA Approval applies for the duration of your REC favourable opinion, unless otherwise notified in writing by the HRA.
- Substantial amendments should be submitted directly to the Research Ethics Committee, as detailed in the *After Ethical Review* document. Non-substantial amendments should be submitted for review by the HRA using the form provided on the HRA website, and emailed to hra.amendments@nhs.net.
- The HRA will categorise amendments (substantial and non-substantial) and issue confirmation of continued HRA Approval. Further details can be found on the HRA website.

Scope

HRA Approval provides an approval for research involving patients or staff in NHS organisations in England.

If your study involves NHS organisations in other countries in the UK, please contact the relevant national coordinating functions for support and advice. Further information can be found at http://www.hra.nhs.uk/resources/applying-for-reviews/nhs-hsc-rd-review/.

If there are participating non-NHS organisations, local agreement should be obtained in accordance with the procedures of the local participating non-NHS organisation.

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

We are pleased to welcome researchers and research management staff at our training days – see details at http://www.hra.nhs.uk/hra-training/

Your IRAS project ID is **224326**. Please quote this on all correspondence.

Yours sincerely

Beverley Mashegede
Assessor

Email: hra.approval@nhs.net

Copy to: Ms Annette Lock, Sponsor Contact

Mariusz Marcinkowski, Lead NHS R&D Contact

IRAS project ID	224326

Appendix A - List of Documents

The final document set assessed and approved by HRA Approval is listed below.

Document	*Version*	*Date*
Confirmation of any other Regulatory Approvals (e.g. NIGB) and all correspondence [Initial Project Proposal]		
Contract/Study Agreement template [Statement of Activities]	1	04 August 2017
Covering letter on headed paper [REC response letter]		03 August 2017
Evidence of Sponsor Insurance or Indemnity (non NHS Sponsors only) [Indemnity Insurance]		27 July 2016
GP/consultant information sheets or letters [Letter to Neurologist]	1	15 June 2017
IRAS Application Form [IRAS_Form_23062017]		23 June 2017
IRAS Application Form XML file [IRAS_Form_23062017]		23 June 2017
IRAS Checklist XML [Checklist_03082017]		03 August 2017
Letter from funder [Cost Approval from RHUL REC]		03 February 2017
Non-validated questionnaire [Demographic Questionnaire (parents)]	1	15 June 2017
Other [Schedule of Events]	1	04 August 2017
Participant consent form [Assent form (young people) V2]	2	27 July 2017
Participant consent form [Consent form (young people) V1]	1	27 July 2017
Participant consent form [Consent form (parent) V2]	2	27 July 2017
Participant information sheet (PIS) [PIS young person (assent) V2]	2	27 July 2017
Participant information sheet (PIS) [PIS young person (consent) V1]	1	27 July 2017
Participant information sheet (PIS) [PIS parent V2]	2	27 July 2017
Referee's report or other scientific critique report [Proposal Response from RHUL REC]		16 December 2016
Referee's report or other scientific critique report [Feedback to RHUL REC]		27 January 2017
Referee's report or other scientific critique report [Project Approval from RHUL REC]		03 February 2017
Referee's report or other scientific critique report [Correspondence with RHUL REC re. project changes]		19 June 2017
Referee's report or other scientific critique report [Summary of Proposal Changes for RHUL REC]		15 June 2017
Research protocol or project proposal [Research Protocol]	2	27 July 2017
Summary CV for Chief Investigator (CI) [Chief Investigator CV (Lauren Bates)]		
Summary CV for student [Chief Investigator CV (Lauren Bates)]		
Summary CV for supervisor (student research) [Supervisor CV (Sarah Rudebeck)]		
Summary CV for supervisor (student research) [Supervisor CV (Jessica Kingston)]		
Validated questionnaire [Strengths and Difficulties Questionnaire (young person)]		
Validated questionnaire [Strengths and Difficulties Questionnaire (parent)]		
Validated questionnaire [Paediatric Pain Profile (parents)]		
Validated questionnaire [Gross Motor Function Classification System Expanded and Revised (medical records)]		

IRAS project ID	224326

Appendix B - Summary of HRA Assessment

This appendix provides assurance to you, the sponsor and the NHS in England that the study, as reviewed for HRA Approval, is compliant with relevant standards. It also provides information and clarification, where appropriate, to participating NHS organisations in England to assist in assessing and arranging capacity and capability.

For information on how the sponsor should be working with participating NHS organisations in England, please refer to the *participating NHS organisations, capacity and capability* and *Allocation of responsibilities and rights are agreed and documented (4.1 of HRA assessment criteria)* sections in this appendix.

The following person is the sponsor contact for the purpose of addressing participating organisation questions relating to the study:

Name: Mariusz Marcinkowski

Tel: 020 7188 7188

Email: mariusz.marcinkowski@gstt.nhs.uk

HRA assessment criteria

Section	HRA Assessment Criteria	Compliant with Standards	Comments
1.1	IRAS application completed correctly	Yes	No comments
2.1	Participant information/consent documents and consent process	Yes	No comments
3.1	Protocol assessment	Yes	No comments
4.1	Allocation of responsibilities and rights are agreed and documented	Yes	The Sponsor intends to use the Statement of Activities as the form of agreement with the participating organisation.
4.2	Insurance/indemnity arrangements assessed	Yes	Where applicable, independent contractors (e.g. General Practitioners) should ensure that the professional indemnity provided by their medical defence organisation covers the activities expected of them for this

Section	HRA Assessment Criteria	Compliant with Standards	Comments
			research study.
4.3	Financial arrangements assessed	Yes	No application for external funding made. No funds will be provided to the participating organisation to support this study.
5.1	Compliance with the Data Protection Act and data security issues assessed	Yes	No comments
5.2	CTIMPS – Arrangements for compliance with the Clinical Trials Regulations assessed	Not Applicable	No comments
5.3	Compliance with any applicable laws or regulations	Yes	No comments
6.1	NHS Research Ethics Committee favourable opinion received for applicable studies	Yes	Provisional Opinion issued 24 July 2017. Further Information Favourable Opinion issued 07 August 2017.
6.2	CTIMPS – Clinical Trials Authorisation (CTA) letter received	Not Applicable	No comments
6.3	Devices – MHRA notice of no objection received	Not Applicable	No comments
6.4	Other regulatory approvals and authorisations received	Not Applicable	No comments

IRAS project ID	224328

Participating NHS Organisations in England

This provides detail on the types of participating NHS organisations in the study and a statement as to whether the activities at all organisations are the same or different.
This is a non-commercial student (PhD) study and there is one site type. The Chief Investigator or sponsor should share relevant study documents with participating NHS organisations in England in order to put arrangements in place to deliver the study. The documents should be sent to both the local study team, where applicable, and the office providing the research management function at the participating organisation. For NIHR CRN Portfolio studies, the Local LCRN contact should also be copied into this correspondence. For further guidance on working with participating NHS organisations please see the HRA website. If chief investigators, sponsors or principal investigators are asked to complete site level forms for participating NHS organisations in England which are not provided in IRAS or on the HRA website, the chief investigator, sponsor or principal investigator should notify the HRA immediately at hra.approval@nhs.net. The HRA will work with these organisations to achieve a consistent approach to information provision.

Confirmation of Capacity and Capability

This describes whether formal confirmation of capacity and capability is expected from participating NHS organisations in England.
Participating NHS organisations in England **will be expected to formally confirm their capacity and capability to host this research**. • Following issue of this letter, participating NHS organisations in England may now confirm to the sponsor their capacity and capability to host this research, when ready to do so. How capacity and capacity will be confirmed is detailed in the *Allocation of responsibilities and rights are agreed and documented (4.1 of HRA assessment criteria)* section of this appendix. • The Assessing, Arranging, and Confirming document on the HRA website provides further information for the sponsor and NHS organisations on assessing, arranging and confirming capacity and capability.

Principal Investigator Suitability

This confirms whether the sponsor position on whether a PI, LC or neither should be in place is correct for each type of participating NHS organisation in England and the minimum expectations for education, training and experience that PIs should meet (where applicable).
A Local Collaborator is expected at the participating organisation. GCP training is not a generic training expectation, in line with the HRA statement on training expectations.

IRAS project ID	224328

HR Good Practice Resource Pack Expectations

This confirms the HR Good Practice Resource Pack expectations for the study and the pre-engagement checks that should and should not be undertaken
Where arrangements are not already in place, an NHS to NHS confirmation of pre-engagement checks letter is expected (if NHS employed). These should confirm enhanced DBS checks, including appropriate barred list checks, and occupational health clearance.

Other Information to Aid Study Set-up

This details any other information that may be helpful to sponsors and participating NHS organisations in England to aid study set-up.
The applicant has indicated that they do not intend to apply for inclusion on the NIHR CRN Portfolio.

d. Research and Development (Guys and St Thomas's NHS Trust) Project Approval

From: Shearman Kirstie <Kirstie.Shearman@gstt.nhs.uk>
Sent: 14 September 2017 14:47
To: Bates, Lauren (2015)
Cc: Lock, Annette; Rudebeck Sarah
Subject: IRAS ID: 224326 Confirmation of GSTFT participation

Dear Lauren,

Study Title: Mental Health and Behaviour in Childhood Dystonia
Sponsor: RHUL
Chief Investigator: Lauren Bates

Guy's and St Thomas' NHS FT has agreed to host your research study (please see attached the confirmation of capacity and capability email to the Sponsor). Your study can therefore now start at GSTFT.

You have committed to recruit 80 patients between 14/09/2017 and 31/07/2018. Please refer to the attached document for capturing recruitment activity onto the EDGE database.

Please note your responsibilities in the attached 'Conditions of GSTFT participation'. These conditions must be adhered to for the Trust to continue to host and support your research.

Documents approved:

If you wish to make any changes to the approved documents, please contact the R&D department or refer to our guidance on submitting amendments (delete if GSTT are not the sponsor).

Document	*Version*	*Date*
GP/consultant information sheets or letters [Letter to Neurologist]	1	15 June 2017
IRAS Application Form [IRAS_Form_23062017]		23 June 2017
Non-validated questionnaire [Demographic Questionnaire (parents)]	1	15 June 2017
Participant consent form [Assent form (young people) V3]	3	21 August 2017
Participant consent form [Consent form (young people) V2]	2	21 August 2017
Participant consent form [Consent form (parent) V3]	3	21 August 2017
Participant information sheet (PIS) [PIS young person (assent) V2]	2	27 July 2017
Participant information sheet (PIS) [PIS young person (consent) V1]	1	27 July 2017
Participant information sheet (PIS) [PIS parent V2]	2	27 July 2017

Research protocol or project proposal [Research Protocol]	2	27 July 2017
Validated questionnaire [Strengths and Difficulties Questionnaire (young person)]		
Validated questionnaire [Strengths and Difficulties Questionnaire (parent)]		
Validated questionnaire [Paediatric Pain Profile (parents)]		
Validated questionnaire [Gross Motor Function Classification System Expanded and Revised (medical records)]		

Kind regards,
Kirstie

Kirstie Shearman
Research & Development Facilitator
NIHR GSTFT/KCL Biomedical Research Centre
T: +44 (0)20 7188 7188 | Ext: 51071 | F: 0207 188 8330 |
E: kirstie.shearman@gstt.nhs.uk | W: www.guysandstthomas.nhs.uk/

e. Royal Holloway, University of London's Research Ethics Committee Project Approval

Ethics Review Details

You have chosen to self certify your project.	
Name:	Bates, Lauren (2015)
Email:	NCJT001@live.rhul.ac.uk
Title of research project or grant:	Mental Health and Behaviour in Childhood Dystonia
Project type:	Royal Holloway postgraduate research project/grant
Department:	Psychology
Academic supervisor:	Jessica Kingston
Email address of Academic Supervisor:	jessica.kingston@rhul.ac.uk
Funding Body Category:	No external funder
Funding Body:	
Start date:	04/09/2017
End date:	31/08/2018

Research question summary:

Dystonia is a movement disorder. This study, 'Mental Health and Behaviour in Childhood Dystonia' aims to answer the research questions: 1) Are children with dystonia more likely to have mental health difficulties (particularly with mood and anxiety) and behavioural problems than the general child population? and; 2) To what extent are self-concept, severity of movement disorder, pain and age of onset of dystonia associated with mental health problems in children with dystonia?

Research method summary:

This research will use a mixed, cross-sectional design- where every person shall participate once only. For the first research question; a quasi-experimental design shall be used where 80 children with dystonia will be compared to a normed sample. For the second research question; a within-subjects design on the children with dystonia shall be used.

The participants will be approximately 80 children with a formal diagnosis of dystonia made by a Neurologist. The age range will be between 7 years and 17 years. The study will include all levels of disability and causes of dystonia to capture an overall picture of what mental health difficulties children and adolescents with dystonia may have. Children on the palliative pathway or nonverbal children who cannot consent for themselves will not be asked to participate.

All testing will take place at the Evelina Children's Hospital.

Children aged 7-10 will complete the Beck Youth Inventories, a 100-item self-report questionnaire, which includes the subscales: depression, anxiety, anger, disruptive behaviour and self-concept.

Children aged 11-17 will complete the Beck Youth Inventories and the Strengths and Difficulties Questionnaire, a brief behavioural screening questionnaire consisting of 25 items. This measure contains subscales measuring emotional, conduct, attention and peer relationship problems, in addition to pro-social behaviour.

Parents will complete a demographic questionnaire, parent version of the Strengths and Difficulties Questionnaire, and the Paediatric Pain Profile, a 20-item behaviour rating scale which discriminates between mild, moderate and severe pain.

Medical records shall be checked to confirm type of dystonia and age of onset; in addition to obtaining Gross Motor Function Classification System Expanded and Revised scores which classifies children into five levels according to their level of motor function and performance in everyday environments.

Risks to participants

Does your research involve any of the below?

Children (under the age of 16).

Yes

Participants with cognitive or physical impairment that may render them unable to give informed consent,
No

Participants who may be vulnerable for personal, emotional, psychological or other reasons,
Yes

Participants who may become vulnerable as a result of the conduct of the study (e.g. because it raises sensitive issues) or as a result of what is revealed in the study (e.g. criminal behaviour, or behaviour which is culturally or socially questionable),
No

Participants in unequal power relations (e.g. groups that you teach or work with, in which participants may feel coerced or unable to withdraw),
No

Participants who are likely to suffer negative consequences if identified (e.g. professional censure, exposure to stigma or abuse, damage to professional or social standing),
No

Details,
Participants will be between 7 and 17 years old.

Participants will have a diagnosis of dystonia and may be suffering from mental health difficulties.

This research has been approved by the London-Dulwich Research Ethics Committee.

Design and Data

Does your study include any of the following?

Will it be necessary for participants to take part in the study without their knowledge and/or informed consent at the time?,
No

Is there a risk that participants may be or become identifiable?,
No

Is pain or discomfort likely to result from the study?,
No

Could the study induce psychological stress or anxiety, or cause harm or negative consequences beyond the risks encountered in normal life?,
Yes

Does this research require approval from the NHS?,
Yes

If so what is the NHS Approval number,
17/LO/1160

Are drugs, placebos or other substances to be administered to the study participants, or will the study involve invasive, intrusive or

potentially harmful procedures of any kind?,
No

Will human tissue including blood, saliva, urine, faeces, sperm or eggs be collected or used in the project?,
No

Will the research involve the use of administrative or secure data that requires permission from the appropriate authorities before use?,
No

Will financial inducements (other than reasonable expenses and compensation for time) be offered to participants?,
No

Is there a risk that any of the material, data, or outcomes to be used in this study has been derived from ethically-unsound procedures?,
No

Details,
Participants may become distressed completing questionnaires about their mental health. They will be told before consenting the nature of the questions. A Trainee Clinical Psychologist is administering the questionnaires and can offer support if needed. Participants can also take a break or stop if they want to.

This project has been given ethical approval by the London-Dulwich Research Ethics Committee.

Risks to the Environment / Society

Will the conduct of the research pose risks to the environment, site, society, or artifacts?,
No

Will the research be undertaken on private or government property without permission?,
No

Will geological or sedimentological samples be removed without permission?,
No

Will cultural or archaeological artifacts be removed without permission?,
No

Details,

Risks to Researchers/Institution

Does your research present any of the following risks to researchers or to the institution?

Is there a possibility that the researcher could be placed in a vulnerable situation either emotionally or physically (e.g. by being alone with vulnerable, or potentially aggressive participants, by entering an unsafe environment, or by working in countries in which there is unrest)?,
Yes

Is the topic of the research sensitive or controversial such that the researcher could be ethically or legally compromised (e.g. as a result of disclosures made during the research)?,

Yes

Will the research involve the investigation or observation of illegal practices, or the participation in illegal practices?,
No

Could any aspects of the research mean that the University has failed in its duty to care for researchers, participants, or the environment / society?,
No

Is there any reputational risk concerning the source of your funding?,
No

Is there any other ethical issue that may arise during the conduct of this study that could bring the institution into disrepute?,
No

Details,
Working with children who are physically impaired or distressed may be emotionally demanding for the researcher. The researcher shall ensure that they meet regularly with the supervisors for this project to discuss such issues.

Working with children there is the risk of safeguarding issues being made aware to the researcher; this may be anxiety provoking for the researcher when deciding a course of action to take. All decisions of this nature will be made with one or several of the following people; the child's named clinician, the duty Clinical Psychologist, and the two Clinical Psychologists supervising the researcher (Dr. Rudebeck and Dr. Kingston).

Declaration
By submitting this form, I declare that the questions above have been answered truthfully and to the best of my knowledge and belief, and that I take full responsibility for these responses. I undertake to observe ethical principles throughout the research project and to report any changes that affect the ethics of the project to the University Research Ethics Committee for review.

Working with children there is the risk of safeguarding issues being made aware to the researcher; this may be anxiety provoking for the researcher when deciding a course of action to take. All decisions of this nature will be made with one or several of the following people; the child's named clinician, the duty Clinical Psychologist, and the two Clinical Psychologists supervising the researcher (Dr. Rudebeck and Dr. Kingston).

Declaration
By submitting this form, I declare that the questions above have been answered truthfully and to the best of my knowledge and belief, and that I take full responsibility for these responses. I undertake to observe ethical principles throughout the research project and to report any changes that affect the ethics of the project to the University Research Ethics Committee for review.

Certificate produced for user ID, NCJT001

Date:	21/08/2017 16:08
Signed by:	Bates, Lauren (2015)
Digital Signature:	Lauren Bates
Certificate dated:	8/21/2017 4:50:49 PM
Files uploaded:	Assent form (young people) V3.pdf Consent (young person) V2.pdf Consent form (parent) V3.pdf PIS parent V2.pdf PIS young person (assent) V2.pdf PIS young person (consent) V1.pdf

Appendix 3: Measures

a. Beck Youth Inventories

Here is a list of things that happen to people and that people think or feel. Read each sentence carefully, and circle the one word (Never, Sometimes, Often, or Always) that tells about you best. THERE ARE NO RIGHT OR WRONG ANSWERS.

		0	1	2	3
1.	I work hard.	Never	Sometimes	Often	Always
2.	I feel strong.	Never	Sometimes	Often	Always
3.	I like myself.	Never	Sometimes	Often	Always
4.	People want to be with me.	Never	Sometimes	Often	Always
5.	I am just as good as the other kids.	Never	Sometimes	Often	Always
6.	I feel normal.	Never	Sometimes	Often	Always
7.	I am a good person.	Never	Sometimes	Often	Always
8.	I do things well.	Never	Sometimes	Often	Always
9.	I can do things without help.	Never	Sometimes	Often	Always
10.	I feel smart.	Never	Sometimes	Often	Always
11.	People think I'm good at things.	Never	Sometimes	Often	Always
12.	I am kind to others.	Never	Sometimes	Often	Always
13.	I feel like a nice person.	Never	Sometimes	Often	Always
14.	I am good at telling jokes.	Never	Sometimes	Often	Always
15.	I am good at remembering things.	Never	Sometimes	Often	Always
16.	I tell the truth.	Never	Sometimes	Often	Always
17.	I feel proud of the things I do.	Never	Sometimes	Often	Always
18.	I am a good thinker.	Never	Sometimes	Often	Always
19.	I like my body.	Never	Sometimes	Often	Always
20.	I am happy to be me.	Never	Sometimes	Often	Always
				BSCI–Y Total RS	

Here is a list of things that happen to people and that people think or feel. Read each sentence carefully, and circle the one word (Never, Sometimes, Often, or Always) that tells about you best, especially in the last two weeks. THERE ARE NO RIGHT OR WRONG ANSWERS.

		0	1	2	3
21.	I worry someone might hurt me at school.	Never	Sometimes	Often	Always
22.	My dreams scare me.	Never	Sometimes	Often	Always
23.	I worry when I am at school.	Never	Sometimes	Often	Always
24.	I think about scary things.	Never	Sometimes	Often	Always
25.	I worry people might tease me.	Never	Sometimes	Often	Always
26.	I am afraid that I will make mistakes.	Never	Sometimes	Often	Always
27.	I get nervous.	Never	Sometimes	Often	Always
28.	I am afraid I might get hurt.	Never	Sometimes	Often	Always
29.	I worry I might get bad grades.	Never	Sometimes	Often	Always
30.	I worry about the future.	Never	Sometimes	Often	Always
31.	My hands shake.	Never	Sometimes	Often	Always
32.	I worry I might go crazy.	Never	Sometimes	Often	Always
33.	I worry people might get mad at me.	Never	Sometimes	Often	Always
34.	I worry I might lose control.	Never	Sometimes	Often	Always
35.	I worry.	Never	Sometimes	Often	Always
36.	I have problems sleeping.	Never	Sometimes	Often	Always
37.	My heart pounds.	Never	Sometimes	Often	Always
38.	I get shaky.	Never	Sometimes	Often	Always
39.	I am afraid that something bad might happen to me.	Never	Sometimes	Often	Always
40.	I am afraid that I might get sick.	Never	Sometimes	Often	Always

BAI–Y Total RS

Here is a list of things that happen to people and that people think or feel. Read each sentence carefully, and circle the one word (Never, Sometimes, Often, or Always) that tells about you best, especially in the last two weeks. THERE ARE NO RIGHT OR WRONG ANSWERS.

		0	1	2	3
41.	I think that my life is bad.	Never	Sometimes	Often	Always
42.	I have trouble doing things.	Never	Sometimes	Often	Always
43.	I feel that I am a bad person.	Never	Sometimes	Often	Always
44.	I wish I were dead.	Never	Sometimes	Often	Always
45.	I have trouble sleeping.	Never	Sometimes	Often	Always
46.	I feel no one loves me.	Never	Sometimes	Often	Always
47.	I think bad things happen because of me.	Never	Sometimes	Often	Always
48.	I feel lonely.	Never	Sometimes	Often	Always
49.	My stomach hurts.	Never	Sometimes	Often	Always
50.	I feel like bad things happen to me.	Never	Sometimes	Often	Always
51.	I feel like I am stupid.	Never	Sometimes	Often	Always
52.	I feel sorry for myself.	Never	Sometimes	Often	Always
53.	I think I do things badly.	Never	Sometimes	Often	Always
54.	I feel bad about what I do.	Never	Sometimes	Often	Always
55.	I hate myself.	Never	Sometimes	Often	Always
56.	I want to be alone.	Never	Sometimes	Often	Always
57.	I feel like crying.	Never	Sometimes	Often	Always
58.	I feel sad.	Never	Sometimes	Often	Always
59.	I feel empty inside.	Never	Sometimes	Often	Always
60.	I think my life will be bad.	Never	Sometimes	Often	Always

BDI–Y Total RS

Here is a list of things that happen to people and that people think or feel. Read each sentence carefully, and circle the <u>one</u> word (Never, Sometimes, Often, or Always) that tells about you best. THERE ARE NO RIGHT OR WRONG ANSWERS.

		0	1	2	3
61.	I think people try to cheat me.	Never	Sometimes	Often	Always
62.	I feel like screaming.	Never	Sometimes	Often	Always
63.	I think people are unfair to me.	Never	Sometimes	Often	Always
64.	I think people try to hurt me.	Never	Sometimes	Often	Always
65.	I think my life is unfair.	Never	Sometimes	Often	Always
66.	People bully me.	Never	Sometimes	Often	Always
67.	People make me mad.	Never	Sometimes	Often	Always
68.	I think people bother me.	Never	Sometimes	Often	Always
69.	I get mad at other people.	Never	Sometimes	Often	Always
70.	When I get mad, I stay mad.	Never	Sometimes	Often	Always
71.	When I get mad, I have trouble getting over it.	Never	Sometimes	Often	Always
72.	I think people try to control me.	Never	Sometimes	Often	Always
73.	I feel people try to put me down.	Never	Sometimes	Often	Always
74.	I feel mean.	Never	Sometimes	Often	Always
75.	I feel like exploding.	Never	Sometimes	Often	Always
76.	I think people are against me.	Never	Sometimes	Often	Always
77.	I get angry.	Never	Sometimes	Often	Always
78.	When I get mad, I feel mad inside my body.	Never	Sometimes	Often	Always
79.	I hate people.	Never	Sometimes	Often	Always
80.	I get mad.	Never	Sometimes	Often	Always
				BANI–Y Total RS	

Here is a list of things that happen to people and that people think or feel. Read each sentence carefully, and circle the one word (Never, Sometimes, Often, or Always) that tells about you best. THERE ARE NO RIGHT OR WRONG ANSWERS.

		0	1	2	3
81.	I steal.	Never	Sometimes	Often	Always
82.	Other people get me into trouble.	Never	Sometimes	Often	Always
83.	I think about running away from home.	Never	Sometimes	Often	Always
84.	I do mean things.	Never	Sometimes	Often	Always
85.	I break into cars, houses, or other places.	Never	Sometimes	Often	Always
86.	I fight with others.	Never	Sometimes	Often	Always
87.	I like getting people mad.	Never	Sometimes	Often	Always
88.	I skip school.	Never	Sometimes	Often	Always
89.	I hate listening to other people.	Never	Sometimes	Often	Always
90.	I argue with adults.	Never	Sometimes	Often	Always
91.	I hurt people.	Never	Sometimes	Often	Always
92.	I like being mean to others.	Never	Sometimes	Often	Always
93.	I break the rules.	Never	Sometimes	Often	Always
94.	I like it when people are scared of me.	Never	Sometimes	Often	Always
95.	I like to hurt animals.	Never	Sometimes	Often	Always
96.	I like to bully others.	Never	Sometimes	Often	Always
97.	I tell lies.	Never	Sometimes	Often	Always
98.	I like to trick people.	Never	Sometimes	Often	Always
99.	I break things when I am mad.	Never	Sometimes	Often	Always
100.	I swear at adults.	Never	Sometimes	Often	Always
				BDBI–Y Total RS	

b. Strengths and Difficulties Questionnaire (Child Self-Report Version)

Strengths and Difficulties Questionnaire S 11-17

For each item, please mark the box for Not True, Somewhat True or Certainly True. It would help us if you answered all items as best you can even if you are not absolutely certain or the item seems daft! Please give your answers on the basis of how things have been for you over the last six months.

Your Name ..... Male/Female

Date of Birth.....

	Not True	Somewhat True	Certainly True
I try to be nice to other people. I care about their feelings	☐	☐	☐
I am restless, I cannot stay still for long	☐	☐	☐
I get a lot of headaches, stomach-aches or sickness	☐	☐	☐
I usually share with others (food, games, pens etc.)	☐	☐	☐
I get very angry and often lose my temper	☐	☐	☐
I am usually on my own. I generally play alone or keep to myself	☐	☐	☐
I usually do as I am told	☐	☐	☐
I worry a lot	☐	☐	☐
I am helpful if someone is hurt, upset or feeling ill	☐	☐	☐
I am constantly fidgeting or squirming	☐	☐	☐
I have one good friend or more	☐	☐	☐
I fight a lot. I can make other people do what I want	☐	☐	☐
I am often unhappy, down-hearted or tearful	☐	☐	☐
Other people my age generally like me	☐	☐	☐
I am easily distracted, I find it difficult to concentrate	☐	☐	☐
I am kind to younger children	☐	☐	☐
I am often accused of lying or cheating	☐	☐	☐
Other children or young people pick on me or bully me	☐	☐	☐
I often volunteer to help others (parents, teachers, children)	☐	☐	☐
I think before I do things	☐	☐	☐
I take things that are not mine from home, school or elsewhere	☐	☐	☐
I get on better with adults than with people my own age	☐	☐	☐
I have many fears, I am easily scared	☐	☐	☐
I finish the work I'm doing. My attention is good	☐	☐	☐

Do you have any other comments or concerns?

c. Strengths and Difficulties Questionnaire (Parent Version)

Strengths and Difficulties Questionnaire P 4-17

For each item, please mark the box for Not True, Somewhat True or Certainly True. It would help us if you answered all items as best you can even if you are not absolutely certain or the item seems daft! Please give your answers on the basis of the child's behaviour over the last six months.

Child's Name ..... Male/Female

Date of Birth.....

	Not True	Somewhat True	Certainly True
Considerate of other people's feelings	☐	☐	☐
Restless, overactive, cannot stay still for long	☐	☐	☐
Often complains of headaches, stomach-aches or sickness	☐	☐	☐
Shares readily with other children (treats, toys, pencils etc.)	☐	☐	☐
Often has temper tantrums or hot tempers	☐	☐	☐
Rather solitary, tends to play alone	☐	☐	☐
Generally obedient, usually does what adults request	☐	☐	☐
Many worries, often seems worried	☐	☐	☐
Helpful if someone is hurt, upset or feeling ill	☐	☐	☐
Constantly fidgeting or squirming	☐	☐	☐
Has at least one good friend	☐	☐	☐
Often fights with other children or bullies them	☐	☐	☐
Often unhappy, down-hearted or tearful	☐	☐	☐
Generally liked by other children	☐	☐	☐
Easily distracted, concentration wanders	☐	☐	☐
Nervous or clingy in new situations, easily loses confidence	☐	☐	☐
Kind to younger children	☐	☐	☐
Often lies or cheats	☐	☐	☐
Picked on or bullied by other children	☐	☐	☐
Often volunteers to help others (parents, teachers, other children)	☐	☐	☐
Thinks things out before acting	☐	☐	☐
Steals from home, school or elsewhere	☐	☐	☐
Gets on better with adults than with other children	☐	☐	☐
Many fears, easily scared	☐	☐	☐
Sees tasks through to the end, good attention span	☐	☐	☐

Do you have any other comments or concerns?

d. Demographic and Clinical Questionnaire

Demographic Questionnaire

Mental Health and Behaviour in Childhood Dystonia

Please answer the following questions about your child. If you are unsure about how to answer, please ask the researcher. If you do not wish to answer any of the questions, then you do not have to do so.

Date of birth ____ /____ /_____ Ethnicity _____ Gender _____

Type of dystonia _____ Age of diagnosis _____

Details of any current medications _____

Has your child had Deep Brain Stimulation Surgery? Yes ☐ (Date _____) No ☐

Details of any other current/past physical health diagnoses

_____

_____

Has your child ever received a diagnosis of or received treatment for a mental health or behavioural difficulty? Yes ☐ No ☐

If so, please provide details (nature of difficulty, past/current, any treatment received)

_____

_____

Does your child have a statement of special educational needs? Yes ☐ No ☐

If so, please provide details (e.g. reason for statement, type of learning disability)

_____

_____

If known, what level of learning disability do they have?

Mild ☐ Moderate ☐ Severe ☐ Profound ☐

Thank you for answering these questions.

e. The Paediatric Pain Profile

Pain Profile

Most troublesome pain (Pain A)

1 For each item please circle the number that best describes your child's behaviour when they have this pain.
2 Enter the number you have circled in to the "score" column.
3 Add up the numbers in the "score" column to give the total score.
4 Record the score on the Summary Graph

When my child has this pain, he or she...	Not at all	A little	Quite a lot	A great deal	Score
Is cheerful	3	2	1	0	
Is sociable or responsive	3	2	1	0	
Appears withdrawn or depressed	0	1	2	3	
Cries / moans/groans / screams or whimpers	0	1	2	3	
Is hard to console or comfort	0	1	2	3	
Self-harms e.g. biting self or banging head	0	1	2	3	
Is reluctant to eat / difficult to feed	0	1	2	3	
Has disturbed sleep	0	1	2	3	
Grimaces / screws up face / screws up eyes	0	1	2	3	
Frowns / has furrowed brow / looks worried	0	1	2	3	
Looks frightened (with eyes wide open)	0	1	2	3	
Grinds teeth or makes mouthing movements	0	1	2	3	
Is restless / agitated or distressed	0	1	2	3	
Tenses / stiffens or spasms	0	1	2	3	
Flexes inwards or draws legs up towards chest	0	1	2	3	
Tends to touch or rub particular areas	0	1	2	3	
Resists being moved	0	1	2	3	
Pulls away or flinches when touched	0	1	2	3	
Twists and turns / tosses head / writhes or arches back	0	1	2	3	
Has involuntary or stereotypical movements / is jumpy / startles or has seizures	0	1	2	3	
				TOTAL	

Please **tick the box** next to the word that best describes the severity of this pain

☐ **None** ☐ **Mild** ☐ **Moderate** ☐ **Severe** ☐ **Very severe**

f. The Gross Motor Function Classification System Expanded and Revised

CanChild Centre for Childhood Disability Research
Institute for Applied Health Sciences, McMaster University,
1400 Main Street West, Room 408, Hamilton, ON, Canada L8S 1C7
Tel: 905-525-9140 ext. 27850 Fax: 905-522-6095
E-mail: canchild@mcmaster.ca Website: www.canchild.ca

GMFCS – E & R
Gross Motor Function Classification System
Expanded and Revised

(Reference: Dev Med Child Neurol 1997;39:214-223)

INTRODUCTION & USER INSTRUCTIONS

The Gross Motor Function Classification System (GMFCS) for cerebral palsy is based on self-initiated movement, with emphasis on sitting, transfers, and mobility. When defining a five-level classification system, our primary criterion has been that the distinctions between levels must be meaningful in daily life. Distinctions are based on functional limitations, the need for hand-held mobility devices (such as walkers, crutches, or canes) or wheeled mobility, and to a much lesser extent, quality of movement. The distinctions between Levels I and II are not as pronounced as the distinctions between the other levels, particularly for infants less than 2 years of age.

The expanded GMFCS (2007) includes an age band for youth 12 to 18 years of age and emphasizes the concepts inherent in the World Health Organization's International Classification of Functioning, Disability and Health (ICF). We encourage users to be aware of the impact that **environmental** and **personal** factors may have on what children and youth are observed or reported to do. The focus of the GMFCS is on determining which level best represents the **child's or youth's present abilities and limitations in gross motor function**. Emphasis is on usual **performance** in home, school, and community settings (i.e., what they do), rather than what they are known to be able to do at their best (capability). It is therefore important to classify current performance in gross motor function and not to include judgments about the quality of movement or prognosis for improvement.

The title for each level is the method of mobility that is most characteristic of performance after 6 years of age. The descriptions of functional abilities and limitations for each age band are broad and are not intended to describe all aspects of the function of individual children/youth. For example, an infant with hemiplegia who is unable to crawl on his or her hands and knees, but otherwise fits the description of Level I (i.e., can pull to stand and walk), would be classified in Level I. The scale is ordinal, with no intent that the distances between levels be considered equal or that children and youth with cerebral palsy are equally distributed across the five levels. A summary of the distinctions between each pair of levels is provided to assist in determining the level that most closely resembles a child's/youth's current gross motor function.

We recognize that the manifestations of gross motor function are dependent on age, especially during infancy and early childhood. For each level, separate descriptions are provided in several age bands. Children below age 2 should be considered at their corrected age if they were premature. The descriptions for the 6 to 12 year and 12 to18 year age bands reflect the potential impact of environment factors (e.g., distances in school and community) and personal factors (e.g., energy demands and social preferences) on methods of mobility.

An effort has been made to emphasize abilities rather than limitations. Thus, as a general principle, the gross motor function of children and youth who are able to perform the functions described in any particular level will probably be classified at or above that level of function; in contrast, the gross motor function of children and youth who cannot perform the functions of a particular level should be classified below that level of function.

OPERATIONAL DEFINITIONS

Body support walker – A mobility device that supports the pelvis and trunk. The child/youth is physically positioned in the walker by another person.

Hand-held mobility device – Canes, crutches, and anterior and posterior walkers that do not support the trunk during walking.

Physical assistance – Another person manually assists the child/youth to move.

Powered mobility – The child/youth actively controls the joystick or electrical switch that enables independent mobility. The mobility base may be a wheelchair, scooter or other type of powered mobility device.

Self-propels manual wheelchair – The child/youth actively uses arms and hands or feet to propel the wheels and move.

Transported – A person manually pushes a mobility device (e.g., wheelchair, stroller, or pram) to move the child/youth from one place to another.

Walks – Unless otherwise specified indicates no physical assistance from another person or any use of a hand-held mobility device. An orthosis (i.e., brace or splint) may be worn.

Wheeled mobility – Refers to any type of device with wheels that enables movement (e.g., stroller, manual wheelchair, or powered wheelchair).

GENERAL HEADINGS FOR EACH LEVEL

LEVEL I	-	Walks without Limitations
LEVEL II	-	Walks with Limitations
LEVEL III	-	Walks Using a Hand-Held Mobility Device
LEVEL IV	-	Self-Mobility with Limitations; May Use Powered Mobility
LEVEL V	-	Transported in a Manual Wheelchair

DISTINCTIONS BETWEEN LEVELS

Distinctions Between Levels I and II - Compared with children and youth in Level I, children and youth in Level II have limitations walking long distances and balancing; may need a hand-held mobility device when first learning to walk; may use wheeled mobility when traveling long distances outdoors and in the community; require the use of a railing to walk up and down stairs; and are not as capable of running and jumping.

Distinctions Between Levels II and III - Children and youth in Level II are capable of walking without a hand-held mobility device after age 4 (although they may choose to use one at times). Children and youth in Level III need a hand-held mobility device to walk indoors and use wheeled mobility outdoors and in the community.

Distinctions Between Levels III and IV - Children and youth in Level III sit on their own or require at most limited external support to sit, are more independent in standing transfers, and walk with a hand-held mobility device. Children and youth in Level IV function in sitting (usually supported) but self-mobility is limited. Children and youth in Level IV are more likely to be transported in a manual wheelchair or use powered mobility.

Distinctions Between Levels IV and V - Children and youth in Level V have severe limitations in head and trunk control and require extensive assisted technology and physical assistance. Self-mobility is achieved only if the child/youth can learn how to operate a powered wheelchair.

Gross Motor Function Classification System – Expanded and Revised (GMFCS – E & R)

BEFORE 2ND BIRTHDAY

LEVEL I: Infants move in and out of sitting and floor sit with both hands free to manipulate objects. Infants crawl on hands and knees, pull to stand and take steps holding on to furniture. Infants walk between 18 months and 2 years of age without the need for any assistive mobility device.

LEVEL II: Infants maintain floor sitting but may need to use their hands for support to maintain balance. Infants creep on their stomach or crawl on hands and knees. Infants may pull to stand and take steps holding on to furniture.

LEVEL III: Infants maintain floor sitting when the low back is supported. Infants roll and creep forward on their stomachs.

LEVEL IV: Infants have head control but trunk support is required for floor sitting. Infants can roll to supine and may roll to prone.

LEVEL V: Physical impairments limit voluntary control of movement. Infants are unable to maintain antigravity head and trunk postures in prone and sitting. Infants require adult assistance to roll.

BETWEEN 2ND AND 4TH BIRTHDAY

LEVEL I: Children floor sit with both hands free to manipulate objects. Movements in and out of floor sitting and standing are performed without adult assistance. Children walk as the preferred method of mobility without the need for any assistive mobility device.

LEVEL II: Children floor sit but may have difficulty with balance when both hands are free to manipulate objects. Movements in and out of sitting are performed without adult assistance. Children pull to stand on a stable surface. Children crawl on hands and knees with a reciprocal pattern, cruise holding onto furniture and walk using an assistive mobility device as preferred methods of mobility.

LEVEL III: Children maintain floor sitting often by "W-sitting" (sitting between flexed and internally rotated hips and knees) and may require adult assistance to assume sitting. Children creep on their stomach or crawl on hands and knees (often without reciprocal leg movements) as their primary methods of self-mobility. Children may pull to stand on a stable surface and cruise short distances. Children may walk short distances indoors using a hand-held mobility device (walker) and adult assistance for steering and turning.

LEVEL IV: Children floor sit when placed, but are unable to maintain alignment and balance without use of their hands for support. Children frequently require adaptive equipment for sitting and standing. Self-mobility for short distances (within a room) is achieved through rolling, creeping on stomach, or crawling on hands and knees without reciprocal leg movement.

LEVEL V: Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Functional limitations in sitting and standing are not fully compensated for through the use of adaptive equipment and assistive technology. At Level V, children have no means of independent movement and are transported. Some children achieve self-mobility using a powered wheelchair with extensive adaptations.

BETWEEN 4TH AND 6TH BIRTHDAY

LEVEL I: Children get into and out of, and sit in, a chair without the need for hand support. Children move from the floor and from chair sitting to standing without the need for objects for support. Children walk indoors and outdoors, and climb stairs. Emerging ability to run and jump.

LEVEL II: Children sit in a chair with both hands free to manipulate objects. Children move from the floor to standing and from chair sitting to standing but often require a stable surface to push or pull up on with their arms. Children walk without the need for a hand-held mobility device indoors and for short distances on level surfaces outdoors. Children climb stairs holding onto a railing but are unable to run or jump.

LEVEL III: Children sit on a regular chair but may require pelvic or trunk support to maximize hand function. Children move in and out of chair sitting using a stable surface to push on or pull up with their arms. Children walk with a hand-held mobility device on level surfaces and climb stairs with assistance from an adult. Children frequently are transported when traveling for long distances or outdoors on uneven terrain.

LEVEL IV: Children sit on a chair but need adaptive seating for trunk control and to maximize hand function. Children move in and out of chair sitting with assistance from an adult or a stable surface to push or pull up on with their arms. Children may at best walk short distances with a walker and adult supervision but have difficulty turning and maintaining balance on uneven surfaces. Children are transported in the community. Children may achieve self-mobility using a powered wheelchair.

LEVEL V: Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Functional limitations in sitting and standing are not fully compensated for through the use of adaptive equipment and assistive technology. At Level V, children have no means of independent movement and are transported. Some children achieve self-mobility using a powered wheelchair with extensive adaptations.

BETWEEN 6TH AND 12TH BIRTHDAY

Level I: Children walk at home, school, outdoors, and in the community. Children are able to walk up and down curbs without physical assistance and stairs without the use of a railing. Children perform gross motor skills such as running and jumping but speed, balance, and coordination are limited. Children may participate in physical activities and sports depending on personal choices and environmental factors.

Level II: Children walk in most settings. Children may experience difficulty walking long distances and balancing on uneven terrain, inclines, in crowded areas, confined spaces or when carrying objects. Children walk up and down stairs holding onto a railing or with physical assistance if there is no railing. Outdoors and in the community, children may walk with physical assistance, a hand-held mobility device, or use wheeled mobility when traveling long distances. Children have at best only minimal ability to perform gross motor skills such as running and jumping. Limitations in performance of gross motor skills may necessitate adaptations to enable participation in physical activities and sports.

Level III: Children walk using a hand-held mobility device in most indoor settings. When seated, children may require a seat belt for pelvic alignment and balance. Sit-to-stand and floor-to-stand transfers require physical assistance of a person or support surface. When traveling long distances, children use some form of wheeled mobility. Children may walk up and down stairs holding onto a railing with supervision or physical assistance. Limitations in walking may necessitate adaptations to enable participation in physical activities and sports including self-propelling a manual wheelchair or powered mobility.

Level IV: Children use methods of mobility that require physical assistance or powered mobility in most settings. Children require adaptive seating for trunk and pelvic control and physical assistance for most transfers. At home, children use floor mobility (roll, creep, or crawl), walk short distances with physical assistance, or use powered mobility. When positioned, children may use a body support walker at home or school. At school, outdoors, and in the community, children are transported in a manual wheelchair or use powered mobility. Limitations in mobility necessitate adaptations to enable participation in physical activities and sports, including physical assistance and/or powered mobility.

Level V: Children are transported in a manual wheelchair in all settings. Children are limited in their ability to maintain antigravity head and trunk postures and control arm and leg movements. Assistive technology is used to improve head alignment, seating, standing, and and/or mobility but limitations are not fully compensated by equipment. Transfers require complete physical assistance of an adult. At home, children may move short distances on the floor or may be carried by an adult. Children may achieve self-mobility using powered mobility with extensive adaptations for seating and control access. Limitations in mobility necessitate adaptations to enable participation in physical activities and sports including physical assistance and using powered mobility.

BETWEEN 12TH AND 18TH BIRTHDAY

Level I: Youth walk at home, school, outdoors, and in the community. Youth are able to walk up and down curbs without physical assistance and stairs without the use of a railing. Youth perform gross motor skills such as running and jumping but speed, balance, and coordination are limited. Youth may participate in physical activities and sports depending on personal choices and environmental factors.

Level II: Youth walk in most settings. Environmental factors (such as uneven terrain, inclines, long distances, time demands, weather, and peer acceptability) and personal preference influence mobility choices. At school or work, youth may walk using a hand-held mobility device for safety. Outdoors and in the community, youth may use wheeled mobility when traveling long distances. Youth walk up and down stairs holding a railing or with physical assistance if there is no railing. Limitations in performance of gross motor skills may necessitate adaptations to enable participation in physical activities and sports.

Level III: Youth are capable of walking using a hand-held mobility device. Compared to individuals in other levels, youth in Level III demonstrate more variability in methods of mobility depending on physical ability and environmental and personal factors. When seated, youth may require a seat belt for pelvic alignment and balance. Sit-to-stand and floor-to-stand transfers require physical assistance from a person or support surface. At school, youth may self-propel a manual wheelchair or use powered mobility. Outdoors and in the community, youth are transported in a wheelchair or use powered mobility. Youth may walk up and down stairs holding onto a railing with supervision or physical assistance. Limitations in walking may necessitate adaptations to enable participation in physical activities and sports including self-propelling a manual wheelchair or powered mobility.

Level IV: Youth use wheeled mobility in most settings. Youth require adaptive seating for pelvic and trunk control. Physical assistance from 1 or 2 persons is required for transfers. Youth may support weight with their legs to assist with standing transfers. Indoors, youth may walk short distances with physical assistance, use wheeled mobility, or, when positioned, use a body support walker. Youth are physically capable of operating a powered wheelchair. When a powered wheelchair is not feasible or available, youth are transported in a manual wheelchair. Limitations in mobility necessitate adaptations to enable participation in physical activities and sports, including physical assistance and/or powered mobility.

Level V: Youth are transported in a manual wheelchair in all settings. Youth are limited in their ability to maintain antigravity head and trunk postures and control arm and leg movements. Assistive technology is used to improve head alignment, seating, standing, and mobility but limitations are not fully compensated by equipment. Physical assistance from 1 or 2 persons or a mechanical lift is required for transfers. Youth may achieve self-mobility using powered mobility with extensive adaptations for seating and control access. Limitations in mobility necessitate adaptations to enable participation in physical activities and sports including physical assistance and using powered mobility.

Appendix 4: Information Sheets and Consent Forms

a. Young Person (Assent) Participant Information Sheet

Study Title: Mental Health and Behaviour in Childhood Dystonia

Royal Holloway, University of London
Department of Clinical Psychology
Egham, Surrey, TW20 0EX
Tel: 01784 414012

Information Sheet for Young People

Mental Health and Behaviour in Childhood Dystonia

We are asking whether you would be interested in taking part in a research study.

Before you decide if you would like to take part, it is really important that you understand what the study is about, why the study is being done and what it would involve for you. So please read and think about this sheet carefully. You can also talk to your family and friends about this if you want.

If something isn't clear or you have more questions, you can ask your parents to call us, and we can discuss it with you and your parents. Thank you for reading this.

Why are we doing this research?
This research is being done to try and understand what it's like for young people with dystonia.

Why have I been invited to take part?
You have been invited because you are between 7 and 17 years old, and have been given a diagnosis of dystonia.

We are hoping to talk to 80 young people with dystonia.

Do I have to take part?
No, you don't. It is your choice whether you want to take part and you can always change your mind.

What will happen to me if I take part?

Someone from our team will come to see you and your parents when you next visit the hospital. They will be with you for about 1 hour.

At the hospital, you and your parents will have the opportunity to ask any more questions you might have.

Then you and at least one of your parents sign a form to say that you are both happy for you to take part in the study.

Then we will ask you to complete some questionnaires about your feelings and how often you do certain things. If you like we can help fill these out for you.

When we have finished speaking to the other young people in the study, we will let you and your parents know what we have found out.

Is there anything to be worried about when taking part?

The questionnaires will ask how you feel and behave. We do not think that taking part in the study will upset you. But if you do become upset while we are talking, we can take a break and you can decide to stop if you want. We can talk to you and your parents about what help you need.

What do I do if I don't want to take part in the research anymore?
Just tell your parents and the person who gives you the questionnaires that you don't want to take part anymore. You can stop taking part at any time. You don't have to give a reason. **It is YOUR choice.**

Will taking part in the study help me?
The study will not help you right now, but it will help us to understand more about what it's like to have dystonia. This may help you and other young people in the future. You may also enjoy taking part.

What happens when the research project stops?
The results of the study will be written up so that people can read about it, but they won't know that you were in the study. We will also let you and your parents know the results of the study.

What if something goes wrong?
If there is a problem you should talk to your parents first, or the person giving you the questionnaires.

Will my information be kept private? Will anyone else know that I am taking part?
All your information will be kept private. We may however have to speak to other people if we worry about you or someone else's safety.

Who is organising and funding the research?
This research is being organised by Royal Holloway, University of London, with the support of the Evelina Children's Hospital.

Who has reviewed this study?
Before any research is allowed to go ahead, it has to be checked by a group of people called the Research Ethics Committee. They make sure that the research is being carried out in a safe way. This study has been reviewed by the London-Dulwich Research Ethics Committee.

Thank you for your time and thinking about taking part in the study.

WHO SHOULD I ASK IF I HAVE FURTHER QUESTIONS?

If you have any questions, talk to your parents first. You can also contact the research team at Royal Holloway: Lauren Bates by telephone on 01784 414012

b. Young Person Assent Form

Royal Holloway, University of London
Department of Clinical Psychology
Egham, Surrey, TW20 0EX
Tel: 01784 414012

Guy's and St Thomas' NHS Foundation Trust

IRAS ID: 224326 Participant Identification Number:

ASSENT FORM for Young People – V2 27/07/2017

Mental Health and Behaviour in Childhood Dystonia

Name of Researcher: Lauren Bates

Thank you for thinking about taking part in this project. The project must be explained to you before you agree to take part. If you have any questions please ask before you decide whether to join in. You will be given a copy of this form to keep.

Please tick the boxes, if you agree and the answer is 'yes':

1. I have read the information sheet for Young People dated 27.07.2017 (version 2) and someone has explained it to me and answered my questions. ☐
2. I know that I can change my mind about joining in anytime, and I don't have to say why. ☐
3. I know what I say is private unless it is about somebody being hurt. ☐
4. I want to join in with the project. ☐

If any answers are 'no' or you don't want to join in, don't write your name.
If you do want to join in, write your name on the line.

Young person's name: _____ Date: _____

5. I have explained the study and answered any questions. ☐

_____ _____ _____

Name of researcher Date Signature

When completed: 1 copy for the family, 1 copy for medical notes, 1 (original) to be kept at Royal Holloway, University of London.

c. Young Person (Consent) Participant Information Sheet

Study Title: Mental Health and Behaviour in Childhood Dystonia

Royal Holloway, University of London
Department of Clinical Psychology
Egham, Surrey, TW20 0EX
Tel: 01784 414012

Information Sheet for Young People

Mental Health and Behaviour in Childhood Dystonia

You are receiving this information sheet as you are currently under the care of the Evelina Children's Hospital and have a diagnosis of dystonia. The clinic is participating in a research project about the wellbeing of children with dystonia, and would like to invite you and your parent to take part.

Before you decide if you would like to take part, it is really important that you understand what the study is about, why the study is being done and what it would involve for you. So please read and think about this sheet carefully. You can also talk to your family and friends about this if you want. The lead researcher will go through this information sheet with you and answer any questions you may have.

Why are we doing this research?
There are at least 8,000 children and young people in the UK with dystonia. This research is being done to try and understand what it's like for young people with dystonia. This research may find out about areas of extra support that people with dystonia and their families may need.

Why have I been invited to take part?
You have been invited because you are between 7 and 17 years old, and have been given a diagnosis of dystonia. We are hoping to recruit 80 young people with dystonia.

What would taking part involve?
A member of your medical team will ask if a member of the research team can call you. They will tell you about the research and answer any questions that you may have. If you are happy for us to, then someone from our team will come to see you and your parent when you next visit the hospital. They will be with you for about 1 hour. You will be given the chance again to ask any more questions you might have before you and your parent sign a form to say that you are both happy to take part in the study.

Then we will ask you to complete some questionnaires about your feelings and how often you do certain things. If you like we can help fill these out for you.

Taking part in this study will in no way impact the care that you may be currently receiving, or receive in the future, from the NHS.

Do I have to take part?
No, you don't. It is your choice whether you want to take part and you can always change your mind.

Will taking part in the study help me?
The study will not help you right now, but it will help us to understand more about what it's like to have dystonia. This may help professionals in future support children with dystonia and their families better. You may also enjoy taking part.

If the answers you put on your questionnaires suggest that you are struggling with how you are feeling; your doctor will be told to make sure that you are getting the support your need.

Is there anything to be worried about when taking part?
The questionnaires will ask how you feel and behave. We do not think that taking part in the study will upset you. But if you do become upset while taking part, we can take a break and you can decide to stop if you want. We can talk to you and your parent about what help you need.

What if something goes wrong?
If there is a problem you should talk to your parents first, or the person giving you the questionnaires. You can also call the researcher [Lauren Bates, 01784 414012] or one of the study supervisors [Dr. Sarah Rudebeck, 02071888533; or Dr. Jessica Kingston, 01784 414105], who will do their best to answer your questions.

Will my information be kept private? Will anyone else know that I am taking part?
All your information will be kept private within the research team. An ID number will be used on your questionnaires instead of your name.

Your doctor at the Evelina Children's Hospital will be told you are taking part, but they won't know what you put on the questionnaires. They will be told if the answers you put on your questionnaires suggest that you are struggling with how you are feeling.

The researcher may need to speak to other people if they are worried about you or someone else's safety. They would always aim to speak to you about this first.

What will happen to the results of this study?
We will let you know what we have found once the study is finished. This will be through posting a sheet summarising the findings for you.

The findings will be written up and may be published in research journals or presented at a professional conference. It will not be possible for anyone hearing about the research to know that you are someone who took part.

What do I do if I don't want to take part in the research anymore?
Just tell your parent and the person who gives you the questionnaires that you don't want to take part anymore. You can stop taking part at any time without a reason. If you want us to, we can remove your questionnaire responses from the results. This won't affect the care that you do or will receive from NHS services.

Who is organising this research study?
This research is being organised by Royal Holloway, University of London, with the support of the Evelina Children's Hospital.

How have the public been involved in this study?
Other young people with dystonia helped us with the study by giving us feedback on

Further information and contact details

Lauren Bates (Trainee Clinical Psychologist, Royal Holloway University of London)
Lauren.Bates.2015@live.rhul.ac.uk 01784 414012

Dr. Sarah Rudebeck (Study Supervisor, Clinical Psychologist, Evelina Children's Hospital)
Sarah.Rudebeck@gstt.nhs.uk 02071888533

Dr. Jessica Kingston (Study Supervisor, Clinical Psychologist, Royal Holloway University of London)
Jessica.Kingston@rhul.ac.uk 01784 414105

d. Young Person Consent Form

Royal Holloway, University of London
Department of Clinical Psychology
Egham, Surrey, TW20 0EX Tel: 01784 414012

Guy's and St Thomas' NHS
NHS Foundation Trust

IRAS ID: 224326 Participant Identification Number:

CONSENT FORM (young person) – V1 27/07/2017

Title of Project: **Mental Health and Behaviour in Childhood Dystonia**

Name of Researcher: Lauren Bates

Please initial boxes

1. I have read the information sheet dated 27.07.2017 (version 1) for the above project., and one of the researchers has talked to me about it. I have had enough time to think about it, and ask questions. ☐
2. I understand that taking part is my choice, and that my parent and I are free to stop at any time without giving any reason, and without my medical care or legal rights being affected. ☐
3. I am willing for the researcher to let my clinical team know that I am taking part in the study. ☐
4. I am willing for the researcher to contact my clinical team with any information relevant to my care (for example, if my questionnaire responses suggest I am struggling with how I am feeling, or the researcher is concerned about mine or someone else's safety). ☐
5. The clinical team can give the researchers relevant information from my clinical notes, if required (for example; an address, age, or confirm clinical information). ☐
6. I understand that some of the questions I will be asked may cause me to become upset. The researcher may stop the research if they decide it is in my best interests. ☐
7. I agree to my data being included in the write-up and publication of this study's findings. Any information that may identify me will be removed from these. ☐
8. I understand that information relating to my taking part in this study will be stored securely for up to five years. ☐
9. I agree to take part in the above study. ☐

Name of young person	Date	Signature

10. I have explained the study to this young person, and answered their questions honestly and fully.

Name of researcher taking consent	Date	Signature

When completed: 1 copy for the family, 1 copy for medical notes, 1 (original) to be kept at Royal Holloway, University of London.

e. Parent Participant Information Sheet

Guy's and St Thomas' NHS
NHS Foundation Trust

Royal Holloway, University of London
Department of Clinical Psychology
Egham, Surrey, TW20 0EX
Tel: 01784 414012

Information Sheet for Parents

Mental Health and Behaviour in Childhood Dystonia

You are receiving this information sheet as your child is currently under the care of the Evelina Children's Hospital and has a diagnosis of dystonia. The clinic is participating in a research project about the wellbeing of children with dystonia, and would like to invite you and your child to take part.

Joining the study is entirely up to you and your child and before you decide whether you would like to take part, we would like you to understand why the research is being done and what it would involve for you both. The lead researcher will go through this information sheet with you, to help you decide whether you would like to take part, and answer any questions you may have.

Explanation: Purpose of this research

There are at least 8,000 children and young people in the UK with dystonia; whilst there is research on the physical care of children with dystonia; little is known about their psychological wellbeing. Research on adults has suggested that dystonia can impact wellbeing. This research aims to explore the emotions and behaviour of children with dystonia; and determine which factors may be associated with a child being more or less likely to have problems with wellbeing or behaviour.

This research may highlight important areas of additional need and support required for families with dystonia. The findings may also help to develop recommendations for parents on how to support their child.

We are hoping to recruit 80 participants with dystonia, to gain an overview of the types of difficulties these children may be facing that impacts on their wellbeing.

What would taking part involve?

Initially, a member of your medical team will ask for your consent for a member of the research team to call you. They will tell you about the research and answer any questions that you may have. This phone call would be before your next appointment. Following this call, if you have given us your consent, a member of the

research team would meet with you at your next appointment and talk you through this Information Sheet.

At this stage you will be asked whether you would like to take part in the study. If you both agree you will both be asked to complete a Consent Form. Your child will be asked to complete some questionnaires about their wellbeing and you will be asked to complete some questions about your child's experiences. You can do this in the same room together and we think that it will last no longer than one hour.

Taking part in this study will in no way impact on the care that you and your child may be currently receiving, or receive in the future, from NHS services.

What are the possible benefits of taking part?
Although taking part may not necessarily benefit you or your child directly, we hope that the findings will increase our understanding of what difficulties young people with dystonia have. In the future, this may help professionals support families where a child has dystonia and enable increased access to psychological services through additional commissioning. Your child may also enjoy taking part in the study.

If your child indicates on their questionnaires that they are struggling with issues such as mood or anxiety; their clinician will be informed to ensure appropriate support is put in place.

What are the possible disadvantages and risks of taking part?
Although we think it is unlikely, there is a chance that your child may find taking part stressful or distressing. If this was to happen then your child would be invited to take a break, and reminded that they do not have to take part. We may also speak with their clinician in case your child may need further support.

What if something goes wrong?
If you have a concern about any aspect of this study, you should ask to speak to the researcher [Lauren Bates, 01784 414012] or one of the study supervisors [Dr. Sarah Rudebeck, 02071888533; or Dr. Jessica Kingston, 01784 414105], who will do their best to answer your questions. If you remain unhappy and wish to complain formally, you can do this by contacting the Guys and St Thomas' complaints line: 020 7188 3514.

What will happen if I don't want my child to carry on with the study?
Your decision to take part in this research is entirely voluntary, and you or your child can change your minds at any stage. If you decline the invitation for your child to take part, or you withdraw their participation later on, this will not affect the care that you do or will receive from NHS services.

If you withdraw from the study after your child has taken part, we will not include their questionnaire responses in our analyses, or in the write-up of the findings.

Will my information be kept confidential?
Your child will be given a unique code, which will be attached to their answers for the questionnaires. Any identifiable details, such as your child's name, will be kept separate from their answers. Only the research team will have access to your child's information and the questionnaire responses.

Your Paediatric Neurologist at the Evelina Children's Hospital will be informed that your child is taking part in the research. They will not be informed of your child's questionnaire responses; however, in the case that your child scores above threshold for any of the clinical disorders on the measures, the clinician responsible for your child's care will be informed so they can decide as to whether your child needs additional support.

As we have a duty of care to keep you and your family safe; if we become concerned your child or someone else is at risk of harm, we will need to share this information with other professionals to ensure we are providing the best care. Where possible we will always try and let you know about this before speaking to other people. This is in line with the Guy's and St Thomas' NHS safeguarding policy.

What will happen to the results of this study?
We will let you know what we have found once the study is finished. This will be through posting a sheet summarising the findings for you.

The findings will be written up as part of a doctoral piece of research, however they will also be submitted to scientific journals that publish research. The findings may also be presented at a professional conference.

Your child and your family will not be identifiable from any report or publication placed in the public domain.

Who is organising this research study?
This research is being organised by Royal Holloway, University of London, with the support of the Evelina Children's Hospital.

How have the public been involved in this study?
Other young people with dystonia who are seen in the Complex Movement Disorders Service at the Evelina Children's Hospital were involved in reviewing materials such as the young person Participant Information Sheet and Consent Form, and the questionnaires.

Who has reviewed this study?
All research in the NHS is looked at by an independent group of people, called a Research Ethics Committee, to protect your interests. This study has been reviewed and given a favourable opinion by the London-Dulwich Research Ethics Committee.

Further information and contact details

Lauren Bates (Trainee Clinical Psychologist, Royal Holloway University of London)
Lauren.Bates.2015@live.rhul.ac.uk 01784 414012

Dr. Sarah Rudebeck (Study Supervisor, Clinical Psychologist, Evelina Children's Hospital)
Sarah.Rudebeck@gstt.nhs.uk 02071888533

Dr. Jessica Kingston (Study Supervisor, Clinical Psychologist, Royal Holloway University of London)
Jessica.Kingston@rhul.ac.uk 01784 414105

f. Parent Consent Form

Royal Holloway, University of London
Department of Clinical Psychology
Egham, Surrey, TW20 0EX Tel: 01784 414012

Guy's and St Thomas' NHS
NHS Foundation Trust

IRAS ID: 224326 Participant Identification Number:

CONSENT FORM (parent) – V2 27/07/2017

Title of Project: **Mental Health and Behaviour in Childhood Dystonia**

Name of Researcher: Lauren Bates

Please initial boxes

1. I confirm that I have read the information sheet dated 27.07.2017 (version 2) for the above project, and one of the researchers has talked to me about it. I have had enough time to think about it, and ask questions. ☐
2. I understand that taking part is voluntary, and that my child and I are free to withdraw at any time without giving any reason, and without my child's medical care or legal rights being affected. ☐
3. I am willing for the researcher to let my child's clinical team know that my child is taking part in the study. ☐
4. I am willing for the researcher to contact my child's clinical team with any information relevant to my child's care, should this become apparent while they are taking part in the study (for example, if my child scores above threshold for any clinical disorder, or the researcher is concerned about my child's or someone else's safety). ☐
5. I give permission for the clinical team to provide the researchers with relevant information from my child's clinical notes, if required (for example; address, age, or confirm clinical information). ☐
6. I understand that the nature of some of the questions my child will be asked are sensitive and may cause my child to become upset. The researcher may withdraw my child from the study if it is deemed appropriate. ☐
7. I agree to my child's data being included in the write-up and publication of this study's findings. Any information that may identify my child will be removed from these. ☐
8. I understand that information relating to my child taking part in this study will be stored securely for up to five years. ☐
9. I agree for my child to take part in the above study. ☐

_____ _____ _____

Name of parent/carer Date Signature

10. I have explained the study to this parent/carer, and answered their questions honestly and fully.

_____ _____ _____

Name of researcher taking consent Date Signature

When completed: 1 copy for the family, 1 copy for medical notes, 1 (original) to be kept at Royal Holloway, University of London.