Abstract

Purpose

To ascertain whether young people with dystonia are more likely than the general population to have mental health and/or behavioural difficulties, and to explore factors that may contribute to these difficulties.

Method

Using a quasi-experimental design, 50 young people with dystonia aged 7–17 and their carers were recruited from the Evelina London Children's Hospital. Young people completed the Beck Youth Inventories and the Strengths and Difficulties Questionnaire. Carers completed the Strengths and Difficulties Questionnaire-Parent version and the Paediatric Pain Profile. Important medical factors, such as age of onset, motor severity and manual function were obtained from medical records.

Results

One sample z tests showed young people with dystonia self-reported significantly higher levels of anxiety ($p < .001$) and prosocial difficulties ($p < .01$), with 48% experiencing clinically significant anxiety levels. They experienced significantly lower levels of anger, disruptive behaviour and conduct problems (all $p \leq .01$). Carers reported significantly higher rates of emotional problems, hyperactivity and peer problems, and significantly lower prosocial behaviours (all $p \leq .01$). Pearson's correlation coefficients showed lower levels of self-esteem were related to higher levels of anxiety ($p = .015$). High levels of pain were related to parent-rated conduct problems ($p = .004$). Age of dystonia onset and motor severity did not correlate with any of the psychological or behavioural measures.

Interpretation/conclusions

Our study suggests high rates of anxiety and behaviours that challenge in children with dystonia. Screening in movement clinics would be helpful in early identification and signposting for support.

Keywords

DystoniaMental healthAnxietyPainSelf-esteemDyskinetic cerebral palsy

1. INTRODUCTION

Dystonia is a movement disorder in which sustained or intermittent muscle contractions cause twisting, repetitive movements or abnormal postures.[1,2] Whilst most forms of dystonia do not affect a person's lifespan or cognition; muscle contractions can cause pain or affect everyday activities such as walking, talking, eating and sleeping.[3]

There is a high prevalence of mental health difficulties in adults with dystonia, with a systematic review suggesting that between 12-71% experience depression and anxiety over their lifetime.[4] While it is clear that there is a higher prevalence of mental health difficulties in those with dystonia, the cause of this is unclear. Some studies suggests that anxiety and depression are related to the neuropathology of dystonia[5–7] with the growing recognition that disruption to the cortical-limbic–striatal function is implicated in depression and other mood disorders.[8,9] Alternatively psychological distress may be caused by the functional impact of the disease itself, such as difficulties performing everyday tasks.[10,11]

Despite the known challenges that adults with dystonia face, there is a paucity of research into the mental health of children and young people (CYP) living with this condition. One recent study looked at the mental health and behavioural needs of CYP with acquired dystonia after basal ganglia stroke.[12] Parent rated questionnaires suggested significantly higher levels of anxiety and depression in CYP with dystonia after stroke as compared to those with stroke and no dystonia. Motor outcomes and cognition were not related to mental health outcomes, though it relied on parental report and did not ask CYP with dystonia about their own psychological experience.

The overreliance on parental report is also seen in studies of mood and behaviour in CYP with cerebral palsy (CP), a neurodisability that can experience similar bio-psycho-social challenges as those with dystonia. One large-scale study (*n*=818), based on parent report, found that a quarter of CYP with CP had significant psychological symptoms or behavioural difficulties.[13] Another study, that notably assessed the child themselves, found that over half of CYP with CP met criteria for a psychiatric disorder, most commonly behavioural difficulties, with half of the sample fulfilling criteria for ADHD.[14] Emotional disorders were much less prevalent, with only ten percent fulfilling a diagnosis of obsessive compulsive disorder (OCD). In summary, behavioural difficulties and to a lesser extent emotional disorders, are found to be more prevalent in both CYP with dystonia after stroke, and CYP with CP, in comparison to the general population.

The factors influencing mood and behaviour continue to be explored in adults and CYP with dystonia and the evidence is mixed. This is unsurprising given that it is likely that a dystonic individual's genetic and medical characteristics interact with their environment to shape their psychological development.[15] Evidence shows a high incidence of mood disorders in adults with acquired or idiopathic dystonias;[4,16] for example, one study found that both adult onset cervical dystonia and blepharospasm were associated with higher rates of anxiety, depression and apathy.[17] Genetic factors influencing mood have also been found to be important especially for those with monogenetic inherited dystonia. Those with Torsin-A DYT-1 genetically inherited dystonia are found to have a higher incidence of mood disorder regardless of whether the dystonia is manifested,[5] suggesting depression is an independent expression of DYT-1 mutation and is a non-motor symptom. Likewise, those with the Epsilon-Sarcoglycan DYT-11 mutation leading to myoclonus-dystonia are found to have a much higher prevalence of psychiatric conditions with two thirds experiencing social anxiety, alcoholism or OCD.[18]

Pain is another non-motor manifestation and is one of the most disabling and frequent symptoms reported in dystonia.[19] Unrecognised or difficult to manage pain can lead to psychological distress, severely reduced quality of life, poor sleep and difficulties with socialising.[13,20,21] Interestingly, studies that use both self-report and parental measures have shown that the link between mental health and skeletal pain is only shown through self-report; carers will often overestimate a child's emotional distress and do not recognise the factors,

such as pain, which contribute to this distress.[20] The management of pain is crucial and often one of the main aims of medical interventions such as Deep Brian Stimulation (DBS) surgery and Intrathecal Baclofen Pump (ITB) implantation in those with generalised dystonia.[22] Other medical factors have been implicated in psychological development, but the link to emotional and behavioural outcomes is less robust. Earlier age of onset has been related to worse mental health outcomes in adults[18,23] and could affect CYP if they are prevented from accessing education or peer relationships.[24] Some studies, however have found no such clear relationships.[25,26] Higher levels of motor severity can lead to stigma, pain, emotional, and social difficulties.[27] Some research has found poorer mental health outcomes for those with milder levels of motor severity,[13] possibly due to increased awareness of perceived differences between affected CYP and unaffected peers, although this has not been fully explored.

Societal factors can also play an important role in the development of mood difficulties but are challenging for quantitative studies to measure. Self-esteem is a psychological construct that pertains to someone's confidence in their own worth or abilities; for a person with a disability, this is influenced by how one evaluates their capacity to perform in society.[28] In adults with dystonia, self-esteem has been related to higher incidence of mental health difficulties due to factors such as disfigurement and negative body concept.[26] CYP with movement disorders and disability are more likely to face discrimination due to the structures of society and the attitudes of others.[29] They may experience exclusion or lack of opportunity and participation in activities,[30] leading to self-esteem issues.[31,32] CYP who exhibit visible movement difficulties may become particularly self-conscious as adolescents,[33] especially if they have experienced bullying as a result.

The current study investigated the mental health and behavioural experiences of CYP with dystonia of varying aetiologies. Due to the high incidence of mental health difficulties in adults with dystonia and CYP with other movement disorders; we investigated whether there is also high incidence of mental health difficulties in CYP with dystonia. We used self-report and parent ratings of emotional and behavioural difficulties, in addition to exploring the roles of potentially important factors that may influence psychological development: aetiology of dystonia, gross motor severity, manual ability, self-concept, pain and age of onset, in addition to other demographic variables.

2. METHOD

2.1 Design

This research used a cross-sectional design in which every participant completed a battery of standardised psychological questionnaires. A quasi-experimental design was used to compare data from CYP with dystonia to normed samples.[34,35] A within-subjects design was used to explore factors associated with mental health/behavioural measures. Ethical approval for the study was granted by the London-Dulwich Research Ethics Committee (17/LO/1160) and by the Research Ethics Committee at Royal Holloway, University of London.

2.2 Participants

The convenience sample consisted of 50 CYP with dystonia and their carer (Table 1). Participants were recruited from the Evelina London Children's Hospital between October 2017 and March 2018. Inclusion criteria included if they had a diagnosis of dystonia and were aged between 7 and 17 years old. Exclusion criteria were CYP who had a diagnosis of a degenerative condition, lacked capacity to consent to taking part, or were unable to understand the questions or communicate their answers. Eligible participants were invited to take part during their routine clinical appointments; the participation rate was 57%.

2.3 Measures

1) Demographic and clinical information was collected from carers using a questionnaire, and from medical records.
2) The Beck Youth Inventories (BYI);[34] a self-report questionnaire consisting of 20-item subscales for depression, anxiety, anger, disruptive behaviour and self-concept. Details of scoring, interpretation and reliability have been reported elsewhere.[34,36]
3) The Strengths and Difficulties Questionnaire;[37] a behavioural screening questionnaire with 5-item subscales for emotional problems, conduct problems, hyperactivity, peer (friendship) problems, and prosocial (kind and helpful)

behaviour. Parental data (SDQ-P) was collected for all participants and self-report data (SDQ) was collected for those aged 11-17 years. Details of scoring, interpretation, reliability and validity have been reported elsewhere.[37–41]

4) The Paediatric Pain Profile (PPP);[42] a 20-item behaviour rating scale completed by carers for those CYP who experienced pain because of their dystonia or comorbid health problem. Details of scoring, interpretation and reliability have been reported elsewhere.[42,43]
5) The Gross Motor Function Classification System Expanded and Revised (GMFCS-E&R)[44] and The Manual Ability Classification System (MACS)[45] classify CYP into five levels according to their level of motor severity and manual ability function respectively. An estimated "equivalence" grading score was applied to those cases with a diagnosis other than CP.[46]

Table 1: *Demographic and Clinical Characteristics of the sample of CYP with dystonia*

Characteristic		***n* (%)**	**Mean (SD)**	**Median (range)**
Age		50 (100%)	13.5 (2.8)	14.0 (8.5-17.8)
Sex	**Male**	22 (44%)		
	Female	28 (56%)		
Age of Onset		39 (78%)	5.8 (4.7)	4.8 (0-14)
Paediatric Pain Profile		33 (66%)	19.03 (12.3)	13.0 (4-57)
GMFCS		49 (98%)		
	I	18 (36%)		
	II	4 (8%)		
	III	4 (8%)		
	IV	14 (28%)		
	V	9 (18%)		
MACS		48 (96%)		
	I	14 (28%)		
	II	7 (14%)		

	III	10 (20%)
	IV	11 (22%)
	V	6 (12%)
Dystonia Aetiology	**Monogenetic Inherited**	15 (30%)
	Acquired	27 (54%)
	of which CP	19 (38%)
	Idiopathic	5 (10%)
	Metabolic	2 (4%)
Medication (Y)		33 (66%)
DBS-Surgery (Y)		19 (38%)
Intrathecal Baclofen Pump (Y)		5 (10%)
Comorbid Health Problem (Y)		31 (62%)
Learning Needs (Y)		22 (44%)

n = number of participants, SD = Standard Deviation, GMFCS = Gross Motor Function Classification System Expanded and Revised, MACS = Manual Ability Classification System for children with cerebral palsy, CP = cerebral palsy, Y = Yes, DBS-Surgery = Deep Brain Stimulation Surgery, Learning Needs = diagnosed learning disability or specific learning difficulties

2.4 Statistical Analyses

IBM SPSS Statistics version 21 was used for all analyses. Data was checked for normality; outliers were removed, and skewed subscales underwent square root transformations which resulted in them being normally distributed. One-sample z tests were used to compare the dystonia group and normed control group on the BYI and SDQ. For the BYI comparisons, the normed data from two separate age groups (7-14 years and 15-17 years) were combined[47] so as not to split our sample. For the SDQ, normed data was only available for those aged 11-15 years. For demographic characteristics, chi-square tests (sex), Kruskal-Wallis tests (aetiological group and intervention type), and *t*-tests (learning needs, pain and medication) were used to investigate their relationships with psychological and behavioural measures.

Pearson's product-moment correlation coefficients were used to examine whether self-concept, age of onset or pain were significantly associated with the psychological and behavioural variables. Bonferroni corrections were applied to correlations to control for multiple comparisons within the dystonia group. Sufficient power of 0.80 was achieved for all comparisons. In all cases a *p* value of < .05 was considered statistically significant.

3 RESULTS

50 young people participated; 13 of whom did not complete all the BYI subscales and/or the SDQ due to them becoming fatigued or disengaging due to the length of time it took to complete the questionnaires, especially in those who were non-verbal or who could not write for themselves. The PPP was completed by 33 parents, as 17 of them reported their child experienced no pain.

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

One sample z-tests (see Table 2) revealed that for the BYI, CYP with dystonia scored significantly higher than population norms on the anxiety subscale (z= 4.13, *p*< .001, *d*= 0.60), and significantly lower on the anger (z= -3.55, *p*< .001, *d*= -0.57) and disruptive behaviour (z= -3.56, *p*< .001, *d*= -0.59) subscales. On the depression subscale, CYP with dystonia did not differ significantly to population norms (z= -0.84, *p*=.40, *d*= -0.12); in fact, the younger age group (7-14 years, as specified by BYI norms) scored significantly lower than population norms (z= -2.06, *p*=.04, *d*= -0.37).

On the SDQ, CYP 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. There were no significant differences in levels of total difficulties, emotional problems or hyperactivity between the CYP with dystonia and population norms. In contrast, carers reported that CYP with dystonia experienced 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 norms.

Table 2: *Population Norms vs. Dystonia Sample Scores on the BYI and the SDQ*

Measure	Subscale	Norms			Sample			z score	*p* value	Cohen's *d*
		n	Mean	SD	*n*	Mean	SD			
BYI	**Self-Concept**	255	51.6	8.9	48	52.1	8.5	0.38	.70	0.06
	Anxiety	255	48.3	8.1	48	53.1	10.8	4.13	< .001*	0.60
	Depression	255	48.4	7.7	47	47.4	8.8	-0.84	.40	-0.12
	Anger	255	47.9	7.9	39	43.4	8.3	-3.55	< .001*	-0.57
	Disruptive Behaviour	255	48.1	8.3	37	43.2	6.5	-3.56	< .001*	-0.59
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

* significant *p* value

n = number of participants, SD = Standard Deviation, BYI = Beck Youth Inventories, SDQ = Strengths and Difficulties Questionnaire, SDQ-P = Strengths and Difficulties Questionnaire- Parent version. *n* for BYI subscales vary as some CYP were unable to complete all subscales, despite assistance offered.

3.2 Percentages of CYP Scoring Above Clinical Cut-Off on the BYI and SDQ

The percentages of CYP who scored above clinical cut-off for the subscales on the BYI and SDQ are shown in Figures 1-2. On the BYI, 47.9% of CYP with dystonia scored above clinical cut-off for anxiety and 23.4% for depression. For the SDQ, 27.3% of CYP with dystonia and 32.0% of carers reported above the clinical cut-off for total difficulties, and 39.4% of CYP and 56.0% of carers reported above the clinical cut-off for peer problems.

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

The relationships between demographic and clinical characteristics, and psychological and behavioural factors were examined. A Kruskal-Wallis test showed there was no significant difference in emotional or behavioural difficulties according to aetiological group (monogenetic inherited, acquired non-CP, acquired CP, idiopathic and metabolic; all $p>0.05$). The same analysis showed that surgical intervention (DBS, ITB, and no intervention) was not associated with differences in psychological experience (all $p>$ .05).

Female participants were significantly more likely to score above clinical threshold than 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 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; $t(46) = -2.03$, $p< .05$).

CYP whose carers did not report that their child has pain as a result of dystonia scored significantly higher on the BYI disruptive behaviour subscale than CYP whose carers did report that their child has pain (mean=46.27, SD=5.3 vs. mean=41.05, SD=6.5 respectively; $t(35) = -2.60$, $p=.01$). CYP whose carers reported that their child had a moderate or severe level of pain scored significantly higher on the BYI depression subscale (mean=50.7, SD=10.1 vs. mean=42.3, SD=8.6 respectively; $t(28) = -2.4$, $p=.02$) and the 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 CYP whose carers reported that their child experiences mild pain. Whether or not the child took medication did not significantly affect measures of emotional or behavioural difficulties (all $p> .05$).

3.4 Relationships Between Predictor Variables, and the BYI and SDQ

Correlations between the BYI and SDQ and the predictor variables (self-concept, age of onset and pain) are shown in Table 3. There was a significant negative correlation between self-concept and anxiety ($r(46) = -.35$, $p=.015$; see Figure 3(A)), and trends toward significant negative correlations between anxiety and self-reported total difficulties ($r(31) = -.41$, $p=.018$) and emotional problems ($r(31) = -.41$, $p=.018$). This suggests that low self-esteem is related to higher levels of anxiety and overall emotional and behavioural difficulties. In addition, there were trends toward significant negative correlations between self-concept and parent-rated total difficulties ($r(46) = -.29$, $p=.048$) and emotional problems ($r(46) = -.31$, $p=.034$).

There was a significant positive correlation between pain and parent-rated conduct problems ($r(30) = .49$, $p=.004$); that is, higher levels of pain were associated with higher levels of behavioural difficulties. In addition, there were trends towards significant positive correlations between pain and anger ($r(22) = .44$, $p=.030$; see Figure 3(B)), and parent-rated total difficulties ($r(30) = .36$, $p=.041$) and hyperactivity ($r(30) = .41$, $p=.020$). There was a trend towards a significant negative correlation between pain and self-reported prosocial behaviour ($r(21) = -.41$, $p=.049$).

Table 3: *BYI and SDQ, and Predictor Variables Correlations*

		Self-Concept	**Age of Onset**	**Pain**
		(*n*=32-48)	(*n*=25-39)	(*n*=23-32)
BYI	**Anxiety** (*n*=48)	-.35**	.00	.15
	Depression (*n*=47)	-.27	-.05	.37
	Anger (*n*=39)	.00	-.15	.44*
	Disruptive Behaviour (*n*=37)	.15	-.02	-.03
SDQ	**Total** (*n*=33)	-.41*	-.04	.31
	Emotional (*n*=33)	-.41*	-.07	.31
	Conduct (*n*=32)	-.25	-.23	-.21

	Hyperactivity (*n*=33)	-.34	.15	.26
	Peer (*n*=33)	-.09	-.10	.05
	Prosocial (*n*=33)	.29	-.02	-.41*
SDQ-P	**Total** (*n*=50)	-.29*	.14	.36*
	Emotional (*n*=50)	-.31*	.22	.14
	Conduct (*n*=50)	-.14	.06	.49**
	Hyperactivity (*n*=50)	-.17	-.03	.41*
	Peer (*n*=50)	-.16	.23	.05
	Prosocial (*n*=50)	.16	-.02	-.29

*p< .05. **correlations remained significant after Bonferroni corrections p< .017.

n = number of participants, BYI = Beck Youth Inventories, SDQ = Strengths and Difficulties Questionnaire, SDQ-P = Strengths and Difficulties Questionnaire- Parent version. *n* for BYI subscales vary as some CYP were unable to complete all subscales, despite assistance offered.

A Kruskal-Wallis test investigated if there was a difference in BYI and SDQ scores in regard to a young person's MACS and GMFCS level. No significant differences were found (both p<0.05) suggesting that neither manual ability or motor severity were associated with emotional or behavioural difficulties in CYP with dystonia.

4 DISCUSSION

Consistent with previous research, in this study CYP with dystonia reported higher levels of anxiety than population norms, with 48% of the dystonia group reporting clinically significant levels of anxiety. It is concerning that almost half our sample reported significant anxiety, and reflects the adult literature which has found a high prevalence of anxiety disorders in those with dystonia.[48] Although 24% of our sample did report clinically significant levels of depression, this did not reach significance in comparison to the population norms. Interestingly, the younger age group scored significantly lower on the depression measure than population norms. While this is lower than has been found in adults with dystonia, the fact that this significant difference disappears once combined with the older group data, suggests that the risk of developing depression with dystonia increases as people become older. It has

been suggested that an awareness of being 'different' increases with age, which may be associated with higher incidence of depression.[14]

Our sample reported significantly lower levels of anger, conduct problems and disruptive behaviour than their peers. While contradictory to the research in CYP with CP, it may be due to the CP literature often focusing on spastic CP and not dyskinetic CP, as in our sample. Both self- and parent-rated levels of peer problems were higher than population norms. CYP did not self-report significantly higher difficulties than population norms on any other SDQ subscales; carers however, reported significantly higher levels of total difficulties, emotional problems and hyperactivity, and significantly lower levels of prosocial behaviour. Differences in perception of wellbeing between CYP and their carers has been found in previous studies[49,50] with CYP commonly reporting better outcomes than parental report.[20] There is also evidence that CYP with CP are more positive about their health-related quality of life than their carers think they are.[49]

In the present study, carers reported significantly higher levels of behavioural difficulties, whereas the CYP reported more emotional distress. The reasons behind this may be multifactorial. Diagnostic overshadowing is a well-known concept in learning disabilities and a large body of research has suggested that emotional distress can present as challenging behaviour (i.e., screaming, aggression and self-injurious behaviour) instead of symptoms usually associated with mood disorder.[51] It may be in our sample, half of which had learning needs, CYP exhibit externalising behaviours of distress as a way of communicating their emotional distress. Alternatively, it may be that the CYP in our study were less likely to have insight into their behavioural needs, potentially due to their learning ability or age. Lastly, they could have been influenced by social desirability biases in cases where assistance was required to complete questionnaires.[52]

In line with previous findings, pain was found to be significantly positively correlated with parent-rated conduct problems. There were also trends towards significant correlations between pain and anger, child-rated prosocial behaviour, and parent-rated total difficulties and hyperactivity. In addition, levels of depression and disruptive behaviour were higher for CYP who experienced higher levels of pain. The link between pain and emotional distress is well known in those with other neurodisabilities,[13] however, this is the first study to hint at the potential relationship in childhood dystonia. This may be as specific clinical psychometric measures, like the BYI, which enable insight into the range of emotional experiences, have not been utilised in previous studies. The adult literature on chronic pain reveals how

the inability to express anger is seen to lead to higher intensity of pain, disability and behavioural expressions of anger.[53] In our sample, carers related higher levels of pain to behavioural difficulties, but not emotional distress. The lack of recognition of the relationship between mental health and pain by carers has been found in previous studies of CYP with CP.[54] As learning needs were related to higher levels of self-reported anxiety, our study adds to the evidence that it is essential to include self-report measures in mental health studies, particularly in populations where learning is impaired.

In this study, CYP with dystonia who had lower self-concept experienced significantly higher levels of anxiety, as well as there being trends towards significantly higher self- and parent-rated total difficulties and emotional problems. This supports previous movement disorder literature; Borkowsha[55] found that CYP did not have lower self-esteem than their peers, however poorer self-esteem was found in those with higher anxiety levels. The link between self-esteem and mental health difficulties is thought to be due to factors such as negative body image, self-consciousness, discrimination, bullying and social exclusion.[26,30] Additionally, in CYP with disabilities, it is thought that anxiety is exacerbated by circumstances that threaten their self-worth, such as academic performance.[56] This may be more of a factor for those with more subtle motor or cognitive symptoms, who are in mainstream schools and therefore more likely to compare themselves to able-bodied peers. It is important to acknowledge that excessive worrying as part of an anxiety disorder, can negatively impact on self-esteem due to impacting how one views themselves in the context of their comorbid anxiety and movement disorders.[55]

Interestingly, aetiology of dystonia appeared to have no influence on the reported emotional or behavioural experiences of our sample, despite the robust adult literature suggesting high prevalence of psychiatric conditions in monogenetic dystonia in adults.[57] Our findings may have been influenced by small sample sizes in the individual aetiology groups. Motor severity and manual function were also not associated with difference in psychological experience in our sample. The literature in those with CP is mixed and it is clear that the relationship between physical disability and emotional problems and behavioural difficulties is complex. Age of onset of dystonia was not associated with any of the psychological or behavioural measures reported. Factors contributing to results may be: that the range for age of onset is much smaller in this study in comparison to adult literature; many CYP's age of onset is recorded as their birth, and; difficulty in establishing an accurate age of onset in cases where it begins with symptoms that are subtle or alongside another movement disorder. Reassuringly, surgical

intervention for dystonia (i.e. DBS-surgery or ITB) was not related to more significant mood or behavioural needs.

The main limitation of this study was the sample size being smaller than intended, which meant that when the sample was subdivided further, groups became small. Nevertheless, this is the largest study of mental health, behaviour, and the relationship to pain and self-esteem in childhood dystonia, and the only one to include both self- and parent- report measures. The study has built on previous literature by giving a clinically-utilised questionnaire of mental health to CYP with dystonia and has enabled a richer understanding of their internal experiences. This study had broad inclusion criteria to increase generalisability and every effort was made to make this study accessible to CYP with learning needs and communication difficulties. Some CYP had the questions read out to them and they indicated answers through point or other non-verbal communication.

Future research could explore specific anxiety disorders and reasons for these elevated levels; considering the high levels of peer difficulties reported, and links in the literature with self-esteem,[58] social anxiety could be of interest. Indeed, identifying with other people with physical disabilities can be associated with reduced anxiety and/or peer difficulties.[59] This study did not have the scope to fully determine the complex and interacting factors that may contribute to the development of psychological difficulties in CYP with dystonia. Further understanding of the effects of under-explored environmental factors will be crucial, particularly in regard to the caregivers of those with dystonia. As with any child's development, parental mental health, parenting style and social supports may all be important factors to explore. The effects of other systems that directly influence CYP with dystonia, like schools, social support, access to friendships and societal attitudes have never been explored. This would be interesting especially in teenagers and young adults where peer relationships become highly important.[60] Finally, the dynamic relationship between pain, anger and externalising behaviours and the perception of CYP and caregivers would be a rich and interesting field to explore in those with dystonia and other neurodisabilities.

Considering research has recently shown that DBS-surgery leads to reductions in pain in those with dystonia,[22] an interesting avenue of research could determine if there is a related change in emotional distress after DBS-surgery, particularly anger and perceived challenging behaviour, which may be mediated by reduction in pain post DBS-surgery. This could be added to meaningful symptom-reduction, evidence currently lacking in CYP with dystonic CP.[61]

5 CONCLUSION

In conclusion, our findings suggest that mental health screening, particularly for anxiety and anger should be part of routine care, particularly for those experiencing pain and/or learning needs. The BYI depression and anxiety subscales were highly correlated, suggesting that CYP with higher levels of anxiety may be at risk of depression if it is not addressed. The importance of timely and accessible treatment including medication and psychological therapy will be vital for this group who undoubtably experience medical and environmental factors that make them at greater risk to mental health difficulties. Our findings also suggest that proactive pain management is vital to support wellbeing in CYP with dystonia and that those exhibiting reported "challenging behaviours" should also be assessed for pain and emotional distress that cannot be expressed. The differences between CYP and parent report highlight that both children and parents should be asked about emotional and behavioural difficulties during routine clinical appointments, and every effort should be made to engage CYP with communication needs. As anxiety and self-esteem are linked; it is important for anxiety treatment to consider factors which may influence both factors, such as lack of social opportunities, trauma from bullying or exclusion, and negative body perception or view of disability. For those CYP without an anxiety disorder, it will be helpful to consider these factors, in order to reduce the likelihood of developing anxiety or depression when older.[26]

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