Original article

Survey of behaviour problems in children with neuromuscular diseases

Joanne Darkea, Kate Bushbyb, Ann Le Couteurc, Helen McConachie

ABSTRACT

Previous research on clinic samples has suggested that children with neuromuscular diseases may be affected by mental health problems. The aim of this study was to establish the carer-reported prevalence of social, communication, and behavioural problems in middle childhood in a total population with neuromuscular diseases.

From a target population of 111, 82 carers of children aged 5–13 years with a diagnosed neuromuscular disease living in the Northern Region of UK were interviewed about service utilisation and needs, and 66 completed the Strengths and Difficulties Questionnaire, Social Communication Questionnaire and Children’s Communication Checklist.

Two-fifths of children scored above the clinical cut-off on at least one questionnaire. These results were significantly higher than are reported for national and normally developing samples. Nine out of 82 had a diagnosis of autism spectrum disorder. Carers of children with problems reported significantly higher levels of unmet need.

Behaviour, social and communication problems are common in children with neuromuscular diseases and Regional Neuromuscular Clinics should consider mental health screening and assessment.

© 2006 European Paediatric Neurology Society. Published by Elsevier Ltd. All rights reserved.

Research suggests that 20–30% of children with chronic illness or physical disability experience behavioural or emotional problems at some point during childhood or adolescence.1 Of children with a muscle disease or weakness in the British national survey of the mental health of children and adolescents, 30% were found to have a mental disorder, around three times the rate of behavioural or emotional problems in children who are physically healthy.2

Previous studies suggest that children with neuromuscular diseases (NMDs) may be affected by a range of mental health problems, including personality disorders, social problems, attention deficits, affective disorders, anxiety and depression.3-6 In particular, there is evidence that children with Duchenne muscular dystrophy (DMD) are selectively impaired in the areas of social behaviour and communication.7,8 Problems with verbal learning, fluency, verbal IQ (VIQ), verbal expression, reading and verbally mediated attention memory are recurrent themes in the DMD literature.9-13 Furthermore, at the extreme end of language and social impairment are a number of case-reports and one clinic prevalence report describing autism in association with DMD.14-17
DMD, an X-linked disorder, which only normally affects males, is the most common NMD to affect children. Thus, the high rates of mental health problems observed in children with NMDs may be explained, in part, by the preponderance of males in this population, as male gender is a potent risk factor for mental health problems in children. A number of other reasons have also been proposed to account for the increased rates of mental health problems in this group. For example, the high prevalence of learning difficulties, high levels of familial stress, and increased risk of socio-economic disadvantage. Nereo et al., found increased stress in mothers of boys with DMD, and that problem behaviours, particularly in social interactions, were a significant predictor of stress.

The results of epidemiological studies suggest that high levels of unmet need are frequently reported amongst families of children with severe physical disabilities. Indeed, though the availability of resources is thought to markedly reduce the negative effects of stressful life events, studies involving families of disabled children have failed to demonstrate a consistent relationship between receipt of services and stress levels of the family. This may be because of parental dissatisfaction with the help received in managing children with physical disabilities. It is therefore important to obtain a clear picture of such needs from the parent’s point of view, particularly as evidence suggests parental perceptions of need differ considerably from those of professionals. The clinical picture of need amongst families of children with NMDs is unclear, but specialist neuromuscular clinics perceive very high levels of difficulty for some parents in managing their children’s behaviour.

Aims of the study:

- To carry out a population-based survey to establish the carer-reported prevalence of social, communication and behavioural problems in children with NMDs in middle childhood.
- To explore the carer-reported use of services to assist with children’s behaviour problems, satisfaction with current services, and perceptions of unmet need.

1. Methods

1.1. Participants

The study approached the main carers of 111 children aged 5–13 years, with a diagnosed NMD and living in the northern region of UK, to take part in the study. This was a population-based study as children were identified through the regional muscle clinics, which are thought to have full ascertainment of all children with a diagnosed NMD living in the northern region. The target population included 51 with DMD, 13 with Becker muscular dystrophy, 12 with myotonic dystrophy, 11 with spinal muscular atrophy (SMA), and 24 with other diagnoses. Seven further children were excluded, five where there was another child with NMD in the family, one child had recently been taken into care, and one was not approached because of family distress.

1.2. Measures

The child’s age and sex, home address postcode, and diagnosis were accessed from the medical notes. The postcode was used to generate a Townsend Index, referenced to the northern region, to measure the level of deprivation of the child’s family based on census returns concerning unemployment, overcrowding, house ownership and car ownership (positive scores indicate greater deprivation).

An information questionnaire collected data on the child’s family characteristics, the type of school attended by the child, and presence of a diagnosed autism spectrum disorder (ASD). Parents were asked if their child had learning difficulties (i.e. whether moderate/severe or mild/none). In addition, data about the families’ use of services to deal with behaviour problems and their satisfaction with these were elicited. A list of 13 areas of need which families of children with a physical disability sometimes experience was presented, and a score calculated for unmet needs, i.e. those areas rated as ‘already getting help but need more’ or ‘not getting help but need it’.

The Strengths and Difficulties Questionnaire (SDQ) is a 25-item questionnaire, which enquires about children’s behaviour in five areas (emotional symptoms, conduct, hyperactivity, peer problems and prosocial behaviour) rated on a 0–2 scale. The first four areas are summed to generate a total difficulties score for each child. A score of 17 or above is the ‘abnormal’ cut-off point. The Social Communication Questionnaire (SCQ) comprises 40 questions that screen for ASDs in children aged four and over. The questions focus on the three main areas of functioning that are affected in children with ASDs: social interaction, communication and stereotyped behaviour. A score of 15 and above is taken as the cut-off point to suggest the child may have ASD.

The Children’s Communication Checklist (CCC) is a 70-item questionnaire that assesses pragmatic aspects of children’s communication, for example, the use of stereotyped language, restriction of language to specific topics, over-literal interpretation of language, and failure to take the perspective of conversational partners into account. The questionnaire comprises nine scales, five of which assess pragmatic aspects of communication and are therefore used to calculate the child’s ‘pragmatic composite’. Parents are asked to complete the checklist if their child’s speech is intelligible, and s/he speaks in sentences. A composite score of 132 or below may suggest the presence of a pragmatic language impairment in the child.

1.3. Procedures

Ethical approval for the study was granted by the Newcastle and North Tyneside Joint Ethics Committee.

Carers were sent an information sheet and consent form for the study, before attending their regular twice-yearly appointment in the Regional Neuromuscular Clinic. Carers who consented were interviewed (by JD) using the information questionnaire in an allocated room, while waiting to see a member of the muscle clinic team. Following the
interview, carers were given copies of the SDQ, SCQ and CCC with a stamped addressed envelope and instructions for self-completion of the questionnaires. Families who did not attend an appointment within the time of the study were sent the questionnaires. If questionnaires were not returned after 3 weeks, a reminder letter was sent, and a second set after a further 3 weeks.

1.4. Analysis

The SDQ has a validated level of 10.5% abnormal ratings in the UK child population. On the basis of previous studies of children with physical disability, 20–30% are to be expected in this study to score above the clinically abnormal cut-off point. The current study therefore had 80% power at the 5% level to detect a difference with a response rate of 45%.

Analysis used chi-squared tests to compare children with NMDs to national norms and data from studies on relevant samples for the three behaviour questionnaires. The proportion of children with NMDs scoring above the cut-off on the SDQ was compared to the proportion of males in the National Child and Adolescent Mental Health Survey who scored above cut-off (13.4%), and to results for males in lone-parent families and males from the lowest income quintile in the Health Survey for England. The proportions of children with NMDs to score above the suggested clinical cut-off on the SCQ and CCC were compared to results from previous studies by Rutter et al., and Bishop and Baird, respectively. Service utilisation and unmet need data were tabulated. T-tests, chi-squared and Fisher’s exact tests were used in comparing respondents with non-respondents, and children with DMD or not, as appropriate.

2. Results

2.1. Representativeness of sample

Information questionnaire data were available for 82 children (73.9%) (see Fig. 1). These children were compared to the 29 whose carers did not participate in the study, on information obtained from the medical notes (see Table 1). Analysis suggests that data from the information questionnaire may be representative of the whole sample.

Social, communication and behaviour questionnaire data were returned for 66 children, an overall response rate of 59.5%. These children were compared with the 16 non-respondents with respect to data from the information questionnaire and from the children’s medical notes. Respondents were significantly less deprived on the Townsend Index than carers who had not returned the behaviour questionnaires (t = 2.19, p = 0.032). In addition, the children of respondents were significantly younger than the children of non-respondents (t = 2.26, p = 0.027). The proportion of children from lone-parent families, or with behavioural diagnoses or moderate/severe learning difficulties was not significantly different for respondents and non-respondents.

Bushby et al. found a significantly raised level of deprivation in families of children with DMD seen at the Northern Region Muscle Clinic over a 30-year period (median Townsend score 3.44, compared with random samples from the same enumeration districts, median 1.46, p < 0.0001). In this population of current patients in middle childhood, the mean Townsend Index score of the 50 children with DMD was 0.65 (SD 3.17), indexed to the northern region, thus not significantly different from the northern region population.

---

**Table 1** - Child and family characteristics of the whole sample, comparing children whose carers completed the information questionnaire, with children whose carers did not take part

<table>
<thead>
<tr>
<th>Variable</th>
<th>Respondents (n=82)</th>
<th>Non-respondents (n=29)</th>
<th>Test</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age in years (SD)</td>
<td>8.70 (2.50)</td>
<td>8.72 (2.40)</td>
<td>t = 0.054</td>
<td>0.957</td>
</tr>
<tr>
<td>Diagnosis: DMD* [no. (%)]</td>
<td>37 (45.1%)</td>
<td>14 (48.3%)</td>
<td>$\chi^2$ = 0.086</td>
<td>0.770</td>
</tr>
<tr>
<td>No. (%) still walking</td>
<td>55 (67.1%)</td>
<td>21 (72.4%)</td>
<td>$\chi^2$ = 0.283</td>
<td>0.595</td>
</tr>
<tr>
<td>No. (%) taking steroids</td>
<td>7 (8.5%)</td>
<td>2 (6.9%)</td>
<td>Fisher’s test</td>
<td>1.000</td>
</tr>
<tr>
<td>Gender: no. (%) male</td>
<td>70 (85.4%)</td>
<td>26 (89.7%)</td>
<td>Fisher’s test</td>
<td>0.755</td>
</tr>
<tr>
<td>Mean Townsend index (Standard Deviation)</td>
<td>0.47 (2.94)</td>
<td>0.47 (3.20)</td>
<td>t = 0.013</td>
<td>0.990</td>
</tr>
</tbody>
</table>

A subsidiary analysis was carried out to determine whether or not there was a significant difference between those children whose carers had received the information questionnaire through the post, and those who had completed it in an interview. Since, the analysis revealed there were no significant differences between the groups, the data are reported together.

* Duchenne muscular dystrophy.
2.2. Behaviour questionnaires

Seventeen of the children with NMDs scored above the cut-off on the total difficulties score of the SDQ (26.2%; nb. total of 65 as one SDQ had missing data). This is a significantly higher proportion than for males in the National Child and Adolescent Mental Health Survey ($\chi^2 = 6.02$, $p < 0.02$). Furthermore, this proportion remains more than that obtained for males in the lowest income quintile (20%) and males from lone-parent families in The Health Survey for England (17%) (see Fig. 2).

30.6% (19 out of 63) of children with NMDs scored above the cut-off on the SCQ and 30.2% (16 out of 53) on the CCC. Children with NMDs were significantly more likely to score above the cut-off on the SCQ and CCC than children from normally developing samples (in which no children were found to score above the cut-off) ($p < 0.001$ and $< 0.01$, respectively) $^{29,33}$

Children with DMD have been thought particularly likely to have behaviour problems. In this study, they were not found significantly more likely to score above cut-off than the rest of the children with NMDs on any of the three questionnaires. However, when children with DMD (28) were combined with children with Becker muscular dystrophy (BMD) (8), they were significantly more likely to score above cut-off on the SDQ ($\chi^2 = 4.14$, $p < 0.05$) than children with other NMDs. Of the 82 children in the overall study, three out of eight children with BMD had a pre-existing diagnosis of ASD and 2 out of 37 children with DMD. Children with myotonic dystrophy were particularly likely to score over cut-off on the SCQ; this is partly explained by level of learning difficulty, but also by physical difficulties in social characteristics such as appropriate facial expression. Nevertheless, 2 out of 10 individuals had a pre-existing diagnosis of ASD. No child with SMA was reported by their parents as having behavioural or communication problems.

Overall, the data demonstrated that 41.5% of children (27/65) had been identified as having behavioural, social and communication problems by at least one of the questionnaires. These children were found to be significantly more likely to have a moderate or severe learning disability (17 out of 27) ($\chi^2 = 11.71$, $p = 0.001$) and significantly more likely to have had a pre-existing diagnosis of ASD (7 out of 27) than children without carer-reported problems (0 out of 38) (Fisher’s exact test, $p < 0.001$). The children with problems were not significantly more likely to be still walking or taking steroids; nor were they more likely to be from lone-parent families, or more deprived.

2.3. Service use

Results from the information questionnaire demonstrated predominantly low service use amongst children with significant carer-reported problems, 22.2% of whom had not seen any professionals in relation to their child’s behaviour problems in the 6 months preceding their muscle clinic appointment. The professionals most commonly consulted were: teacher (15), educational psychologist (8), physiotherapist (8), special support assistant (7) or the family care worker from the regional clinic (6). Carers were generally highly satisfied with the services they had encountered.

Unmet need scores were significantly higher amongst the 27 children with significant carer-reported problems ($Z = -3.716$, $p < 0.001$). The most frequently reported areas of unmet need were: child’s behaviour problems, teaching positive new skills, communication skills, information about child’s condition, information about services available, and classes to learn how to help their child.

3. Discussion

In this study, children with NMDs were reported by their carers as showing serious levels of behavioural, social, and communication problems that are likely to impact on their and their families’ daily lives. Those most likely to present with problems were children with a moderate or severe learning disability, children with Becker muscular dystrophy, or children who already had a diagnosis of an autism spectrum disorder. Furthermore, it was the families of these children with behavioural, social and communication problems who had the greatest levels of unmet need for services, despite reporting high satisfaction with the professionals they were consulting about their child’s behaviour.

Children were identified for this study through the Northern Region Muscle Clinic. This population sample may not have absolutely complete ascertainment of all children living in the northern region with an NMD, as children with currently undiagnosed conditions will not be registered with the clinic. However, this effect is likely to be small. The reason for the difference from the Bushby et al. study in levels of deprivation for children with DMD is not known; it is unlikely to be accounted for in full by the different method of comparison, but may arise from the previous study having covered a 30 year period.

The response rate (73.9% for the information questionnaire, 59.5% for the social, communication and behaviour questionnaires) might have introduced bias in to the study if respondents differed significantly from non-respondents in ways that affected answers to questionnaires. The fact that the children of respondents to behaviour questionnaires were significantly younger may have increased the number of children identified as having behaviour problems, as evidence indicates that younger children are more likely to score above the cut-off on the SDQ. However, the higher
levels of deprivation amongst non-respondents may have led to an underestimate of the prevalence of behaviour problems in children with NMDs, on account of the generally increased prevalence of behaviour problems found in children who are more socially deprived.2 Thus, the response rate is not thought to have biased the results to any significant extent (Table 2).

The findings show that children with some types of NMDs are at risk for developing significant behaviour, social and communication problems. The risk for children with chronic illness or physical disability is already known, but this population study suggests that there may be particular additional factors affecting children with NMD. Firstly, there is an increased risk where children have learning disability. However, the fact that 40% of the children with carer-reported problems were not classified as having a moderate/severe learning disability means other factors are also important. Secondly, different conditions had different levels of risk for behavioural, social or communication problems. While recognising that numbers are small, the survey suggests that children with Becker muscular dystrophy were particularly likely to have problems, including a diagnosis of ASD, more so than was the case for children with DMD. Nevertheless, the findings agree with a recent US clinic study that the rate of ASD is significantly raised in DMD.17 Children with SMA, often reported as having above average intelligence, did not present behavioural problems, in line with previous studies.34 Finally, it was notable that the study showed that parents are more likely to seek advice about their children's behavioural, social and communication difficulties from the educational sector or at the Regional Muscle Clinic, than from mental health professionals (such as local community child psychiatrists, clinical psychologists, or mental health nurses). Of the unmet needs surveyed, help with behaviour problems, and help with developing the child's communication skills were amongst the most commonly reported. Clinically, the study suggests that Regional Neuromuscular Clinics should consider screening in order to identify children who are at high risk of mental health problems. Children so identified would then require more detailed assessment in order to characterise their particular difficulties, and to identify the child and family's needs for additional services or support. The clinics may therefore consider forming regular links with Child and Adolescent Mental Health services and Learning Disability services. Interventions, which address children's needs in terms of social interaction skills, pragmatic communication skills and self-esteem, as well as advice to parents on

<table>
<thead>
<tr>
<th>Diagnostic grouping</th>
<th>Pre-existing ASD diagnosis n (%)</th>
<th>Above cut-off on SDQa n (%)</th>
<th>Above cut-off on SCQa n (%)</th>
<th>Above cut-off on CCCa n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Duchenne muscular dystrophy n=37</td>
<td>2 (5.4)</td>
<td>9 (32.1)</td>
<td>5 (18.5)</td>
<td>7 (30.4)</td>
</tr>
<tr>
<td>Becker muscular dystrophy n=8</td>
<td>3 (37.5)</td>
<td>4 (50.0)</td>
<td>4 (50.0)</td>
<td>3 (42.9)</td>
</tr>
<tr>
<td>Myotonic dystrophy type 1 n=10</td>
<td>2 (20.0)</td>
<td>2 (22.2)</td>
<td>6 (75.0)</td>
<td>2 (40.0)</td>
</tr>
<tr>
<td>Spinal muscular atrophy n=9</td>
<td>0 (0.0)</td>
<td>0 (0.0)</td>
<td>0 (0.0)</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td>Congenital muscular dystrophy n=7</td>
<td>1 (14.3)</td>
<td>0 (0.0)</td>
<td>1 (20.0)</td>
<td>1 (20.0)</td>
</tr>
<tr>
<td>Other neuromuscular diseases n=10</td>
<td>1 (9.1)</td>
<td>2 (18.2)</td>
<td>3 (27.3)</td>
<td>3 (27.3)</td>
</tr>
<tr>
<td>Total n</td>
<td>9/82</td>
<td>17/65</td>
<td>19/63</td>
<td>16/53</td>
</tr>
</tbody>
</table>

a SDQ, strengths and difficulties questionnaire; SCQ, social communication questionnaire. CCC, children's communication checklist.

phenotype, which includes problems with social interaction and communication. This is similar to the suggestions made by Yude et al.37 Based on their work with hemiplegic children, the authors proposed that the high levels of peer rejection and victimisation observed amongst children with hemiplegia may have occurred on account of neurologically driven deficits in social awareness and social skills. A further study conducted using children with hemiplegia suggested that social and emotional immaturity were associated with delayed maturation of 'theory of mind'.38 ‘To have a theory of mind is to be able to attribute independent mental states to self and others in order to understand and predict behaviour’ (Happe,39 p. 38). It is deficits in this ability to ‘mind read’ that are thought to contribute to the social impairments present in children with autism spectrum disorders (ASDs). If it were the case that some children with NMDs also have constitutional problems in ‘mind reading’ abilities, then this may help to explain their limited ability to initiate and maintain peer relationships.37 Further investigation of a specific genetic or other link with DMD, BMD and myotonic dystrophy is also indicated.17 In the long run, it will be important to understand the characteristic difficulties of children with NMDs, and the development of their strengths and difficulties over time, in order to ensure that the most effective interventions are available.

The study showed that parents are more likely to seek advice about their children’s behavioural, social and communication difficulties from the educational sector or at the Regional Muscle Clinic, than from mental health professionals (such as local community child psychiatrists, clinical psychologists, or mental health nurses). Of the unmet needs surveyed, help with behaviour problems, and help with developing the child’s communication skills were amongst the most commonly reported. Clinically, the study suggests that Regional Neuromuscular Clinics should consider screening in order to identify children who are at high risk of mental health problems. Children so identified would then require more detailed assessment in order to characterise their particular difficulties, and to identify the child and family’s needs for additional services or support. The clinics may therefore consider forming regular links with Child and Adolescent Mental Health services and Learning Disability services. Interventions, which address children’s needs in terms of social interaction skills, pragmatic communication skills and self-esteem, as well as advice to parents on

children with NMDs may show a characteristic behavioural
management of children’s attention and cooperation, will be more likely to be successful if introduced early on after diagnosis of the NMD.

REFERENCES


