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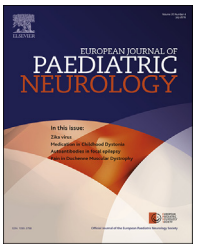

Original article

Parents' advice to healthcare professionals working with children who have spinal muscular atrophy

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ABSTRACT

Aim: To explore parents' advice to healthcare professionals working with children with spinal muscular atrophy (SMA).

Materials and methods: This study derives from a Swedish nationwide survey and uses content analysis to make inferences from answers to an open-ended question concerning parent's advice to healthcare professionals. Of eligible parents who had a child born in Sweden between 2000 and 2010, diagnosed with SMA type 1 or 2, and for whom respiratory support was considered in the first year of life, 61 participated in the study (response rate: 87%). Of these, 51 parents answered the question about advice to healthcare professionals working with children with SMA.

Results: More than half of the advice from parents was related to professional–family relations. The second most frequent type of advice related to two aspects of knowledge about SMA: desire that healthcare professionals possess knowledge, and desire that they provide knowledge. The parents also had advice concerning support in daily life, both to the parents and to the affected child. Other pieces of advice were related to organization of care and the parents' desire to be involved in the child's care.

Conclusions: Parents advised healthcare professionals to increase their disease-specific knowledge, to treat the parents as experts on their child, and to treat the family with respect, particularly in situations where the child's case is used as an opportunity to improve healthcare professionals' competence. Increased practical support in daily life and a case coordinator is also among parents' advice to healthcare professionals.

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1. Introduction

Little is known about how parents of children with spinal muscular atrophy (SMA) perceive care and what they want from healthcare professionals (HCPs). SMA is a severe genetic neuromuscular disease with autosomal recessive inheritance; it results in progressive proximal muscle weakness and paralysis. With an estimated incidence of 1 in 10,000 live births and a carrier frequency of 1/40–1/60, SMA is the second most common fatal autosomal recessive disorder among children.[1] SMA is typically classified into three grades of severity (type 1, 2, 3) based on age at presentation and disease severity, although an alternative five-grade classification has also been suggested.[2] The most severe type, SMA type 1, causes symptoms in the first 6 months of life and the child usually dies within two years unless respiratory and nutritional support is initiated. SMA type 2 is an intermediate form with similar symptoms but later presentation of first symptoms (before 18 months of age) and slower progression. This is the most common type of SMA in older children and adolescents. SMA type 2 usually leads to a shortened life span.[3] Despite promising potential for new medication,[4] there is currently no cure for SMA. Treatment therefore focuses on preventing complications caused by weakness and maintaining quality of life, and should include management of respiratory failure, nutritional support, orthopedic care, rehabilitative interventions, and palliative care.[5]

Palliative care research on children with neurological disease is sparse as compared to research on children with cancer.[6] A few studies of how parents of children with SMA experience their child's care have been published; however, the focus is not on palliative care in all of them, despite the fact that these parents face difficult ethical decisions regarding life-prolonging treatment and often have to endure their child's early death.[7–13] Some of these studies report that parents of children with SMA and other severe neuromuscular diseases perceive aspects of care as having poor quality, e.g. lack of knowledge/competence among HCPs,[7,8,10,12–15] lack of coordination of care[9] and HCPs behaving in a unhelpful manner.[8] On the other hand Higgs et al.[11] reported that parents perceived that their child's care was of good quality. This is also in line with the findings by Lövgren et al., that all parents were informed regarding treatment decisions and that a majority of the parents who had expressed a preference concerning their child's location of death had their wish fulfilled.[14,15] However, the results[14] also indicate that care guidelines were not always followed in full. For example, a fourth of the parents did not receive information about available respiratory support options.[16]

Since the number of studies in pediatric palliative care of children with neurological diseases is small, and knowledge about parents' wishes about care is limited, this study aims to explore parents' advice to HCPs working with children who have SMA and their families.

2. Materials and methods

2.1. Study design and participants

This study derives from a Swedish nationwide survey focusing on parents' experiences of the care of their severely ill child with SMA.[14,15] The survey included parents of children who were born in Sweden between 2000 and 2010, diagnosed with SMA type 1 or type 2, and for whom respiratory support was considered as treatment by HCPs during the first year of life. The children and their parents were identified through searches in the comprehensive registers maintained by the National Board of Health and Welfare and the Swedish Tax Agency, respectively. Seventy-eight parents of 40 children were identified. Of these, seventy parents of 39 children had an identifiable phone number and were therefore contacted and asked if they wanted to participate in our survey. In total, 61 parents of 39 children (48 bereaved parents of 32 children and 13 non-bereaved parents of seven living children) participated (response rate: 87%). The present study is based on the 51 parents who responded to the question about advice to HCP (Fig. 1).

The survey was approved by the Ethics Committee in Stockholm/Karolinska Institutet, reference number: 2009/1702-31/2.

2.2. Data collection

Eligible parents were sent an information letter describing the study, one week to 10 days after the parents were contacted by the study interviewer and asked if they wanted to participate. If so, questionnaires were sent to the child's mother and father separately. About ten days later, a combined thank-you and reminder card was mailed to the parents who had agreed to participate. Parents who did not return the questionnaire were contacted by telephone and asked if they had any particular concerns or needed assistance with responding to the questionnaire.

2.3. The questionnaires

The questionnaires, one for bereaved and one for non-bereaved parents, were constructed according to a method developed by Charlton[17] and others.[18–20] The questionnaires consist of 75/59 questions (bereaved/non-bereaved parents), including both closed and open-ended questions. The present study is based on the parents' written responses to one open-ended question: "What advice would you give to staff working with children who have spinal muscular atrophy and their families?".

2.4. Data analysis

In order to explore parents' advice to HCPs, the responses to the open-ended question about advice were analyzed with

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