



National Hospice and Palliative Care Organization
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**COMMUNICATION BETWEEN PARENTS
AND HEALTH CARE PROFESSIONALS
ENHANCES SATISFACTION AMONG
PARENTS OF CHILDREN WITH SEVERE
SPINAL MUSCULAR ATROPHY**

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FOCUS FOR THIS REPORT

In this report, based on findings derived from a nationwide survey about parents' perception of the care of their child with severe spinal muscular atrophy (SMA) (Lövgren, Sejersen et al. 2016a, Lövgren, Sejersen et al. 2016b) we specifically highlight the following areas: a parents' perception of disease information and treatment decisions; the bereaved parents' wishes about where and how the child would pass away and what bereaved parents recalled as being especially supportive or distressing in connection to their child's death; and bereaved parents' perception of support to siblings.

BACKGROUND

SMA is a rare, autosomal recessive disorder characterized by slowly progressive muscle weakness (Markowitz, Singh et al. 2012). It is classified into four grades of severity where the most severe form, type I, presents by definition during the first six months of life, and death usually occurs within the first two years of life without aggressive respiratory support. Type II presents slightly later, at 6-18 months, and shortens life expectancy (Markowitz, Singh et al. 2012, Bladen, Thompson et al. 2014). There are promising therapeutic drugs in the pipeline for SMA, including the most severe forms, but currently there is no cure for any form of SMA. Health providers are therefore guided to focus on preventing complications from muscle weakness and maintaining quality of life, primarily through respiratory and nutritional support (Iannaccone 2007, Wang, Finkel et al. 2007).

Research is very limited on how parents of children with severe SMA experience the child's care, despite the fact that they face difficult ethical decisions regarding life-prolonging treatment and often also have to endure their child's early death. However, the following studies are exceptions (Parker, Maddocks et al. 1999, Mah, Thannhauser et al. 2008, Rallison and Raffin-Bouchal 2013, Lawton, Hickerton et al. 2015, Qian, McGraw et al. 2015, Higgs, McClaren et al. 2016, Yang, Mu et al. 2016). Some of the few studies report that parents of children with SMA and other severe neuromuscular diseases note the following; perceive aspects of care as being of poor quality, e.g. lack of knowledge/competence among health care staff (Mah, Thannhauser et al. 2008, Rallison and Raffin-Bouchal 2013, Lawton, Hickerton et al. 2015, Qian, McGraw et al. 2015, Yang, Mu et al. 2016); lack of coordination of care (Parker, Maddocks et al. 1999) and health care staff behaving in an unhelpful manner (Qian, McGraw et al. 2015).

On the other hand, Higgs, McClaren et al. (2015) reported that parents perceived that their child's care was of good quality. Increased knowledge from the families' experiences, wishes and concerns may help health providers to develop strategies to improve the care for these children and their families.

MATERIAL AND METHODS

We conducted a nationwide survey in 2013, of the parents of children who were born in Sweden between the years 2000 and 2010 and later diagnosed with SMA type I or II, and for whom respiratory support was considered by the health care professionals during the first year of life. Seventy parents of 37 children were contacted by phone and asked about their willingness to participate. Both mothers and fathers were invited. Fourteen parents of seven children who were still living with SMA and 56 bereaved parents of 30 children were sent a letter of invitation in February 2013. In all, 61 parents of 36 children participated (participation rate: 87%).

The questionnaires to bereaved parents and non-bereaved parents consisted of similar questions with the addition of questions regarding the loss of their child to bereaved parents. The questionnaires for non-bereaved involved 59 items while the questionnaire to bereaved included 75. The first part focused on information, decision making, and support to the family from the time of diagnosis, and the second part included parents' perceptions of the quality of care at the hospital and at the rehabilitation clinic.

RESULTS

Characteristics of the parents and their children

- The participating parents consisted of an almost equal number of mothers and fathers.
- The parents' mean age was close to 40 years at time of follow-up.
- The children still alive at follow-up had a mean age of 9.7 years.
- The deceased children's mean age at the time of death was 9.5 months.
- Fifty-three children had SMA type I and seven had SMA type II (one child had type I or II).
- The most common type of respiratory support reported by the parents of the children who were alive at follow-up was non-invasive ventilation by mask (N=11).
- The deceased children had most commonly received supplementary oxygen by nostril or mask (N=16), followed by ventilation by mask (BIPAP) (N=15), and no respiratory support at all (N=11).
- None of the children had ventilator by tracheostomy.

Parents perception of information and treatment decisions

None of the parents reported that the health care staff made decisions concerning the child's treatment without informing them first, and 80% reported feeling confident about the decisions made. A vast majority reported that discussions regarding whether or not to treat the child had been carried out in a considerate manner. However, nearly a quarter (23%) of the bereaved parents, reported that they got no information about respiratory support, compared to 15% of non-bereaved. Bereaved parents were more likely to report being satisfied with and understanding the information given about the illness and its treatment than non-bereaved parents.

Bereaved parents' experiences and wishes near the child's death

Parents' wishes about where the child would pass away

The children's' death took place either at home, at a children's hospital or in a car on their way home. Almost equal numbers of children near death were seen at home as at a children's hospital, and one child was seen in a children's hospice. Two thirds of the parents had a preference for where their child would pass away. Of the parents who had a preference for location of death, a majority (66%) had their wishes fulfilled. All of those who wanted their child to die at a children's hospital had their wishes fulfilled. Not all children whose parents wanted the child to die at home died there, instead their death took place at a children's hospital or in a car on their way home.

Parents' wishes about how the child would pass away

A majority of parents (65%) had specific preferences about how their child would pass away and many of them (84%) talked to a physician about it. Of those, only two parents (8%) did not get what they asked for. Of those with preferences about how the child would pass away, a vast majority (87%) had their wishes fulfilled. Parents wrote free comments about their desires regarding how their child would die:

- with no pain or anxiety or with as mild symptoms as possible
- together with the parents
- peacefully
- with their parents singing
- with no equipment
- while sleeping
- with their parents praying
- without attempts of resuscitation

Supportive or considerate situations in connection to their child's death

Many parents (83%) reported that health care staff said or did something in connection with the death of the child that the parents remembered as being especially supportive or considerate. Almost all parents who did not experience any such support had children who died in a hospital. Thirty-three of the parents (69%) wrote free comments about what had been especially supportive or considerate:

- Health care staff showed respect, empathy or did their utmost
- Staff supported both the parents and their child
- Staff were present in the moment
- The staff made it possible for relatives to say goodbye
- Staff supported them by giving information
- Especially supportive after death was when the health care staff dressed the child, held him/her and made the room beautiful, collected mementos, gave the parents time with the dead child, when staff showed feelings, opened the window to release the soul, and gave gifts.

Distressful situations in connection to the child's death

Close to a quarter of the parents (27%) reported that health care staff said or did something in connection with the child's death that they remembered as being distressing. Parents' reported the following difficult situations:

- Staff or the funeral agency treated the child and/or the parents insensitively
- Their child received inadequate pain relief
- The parents were given incorrect or no information
- There was a change in shift on the ward at the time of death
- The staff were stressed
- The staff did not listen to the parents about how to treat their child at the end of life

Support to siblings

Thirty-six parents reported that their child had siblings and that four of the siblings (11%) received professional psychological support after the death of their brother or sister (12 were described as too young). Parents' free comments about support for siblings often were associated with talking a lot within the family about illness and bereavement. Some parents wrote that they had been encouraged by health care staff to do that as a way of supporting siblings.

DISCUSSION

Treatment Decisions

All parents in this study were informed before treatment decisions were made but a quarter reported not having received information about respiratory support. The reasons for not having this information, even if it is recommended in care guidelines (Wang, Finkel et al. 2007), are unknown. However, one potential reason might be that the physician/health care staff thought that respiratory support might only contribute to more suffering for the child and his/her family. Regardless of this, parents need to be informed to be able to make clear decisions.

Receiving and Understanding Information

This study also showed that bereaved parents were more likely to report being satisfied with and understanding the information given about the illness and its treatment than non-bereaved parents. On the other hand, it was more bereaved parents who reported that they did not get the information about respiratory support. One reason for this might be that non-bereaved parents have been exposed to health care for longer periods than bereaved parents and the probability to have a negative experience increase with time. However, this result indicates that care provision needs to be improved for families of children living with severe SMA, especially now when there are promising therapeutic drugs for SMA.

Honoring End-of-Life Preferences

The results from this study showed that it was easier for parents to have their preferences honored if they wanted their child to die at a hospital compared with those who wanted their child to die at home. This is in line with previous findings by Dussel and colleagues (2009). Only in Stockholm, the capital city in Sweden, can homecare for children be provided around the clock. This means it is not an option for many families living elsewhere in the country. This might also be a reason why parents' preferences for their child to die at home was difficult to fulfill. It might also be one reason why "only" half of the parents wanted their child to die at home.

On the other hand, there is no clear evidence about home being the preferred location for their child's death, and more studies are therefore needed to explore this issue (Bluebond-Langner, Beecham et al. 2013). Not surprisingly, it was the families who had already used home care during the child's illness that wanted their child to die at home, and it is probably more likely that it is the children with less symptom burden that die at home. It also seems more likely to want to have your child cared for at home if you have additional children in family.

The findings from this study also showed that it was fruitful to talk to the physician about what they hoped their child's death would be like as almost all parents who had this discussion had their wish fulfilled in the end. This suggests that it is important to develop a good communication milieu among physicians, health care staff and families in order to make it comfortable for parents to express their wishes and concerns. All expressed wishes in this study were concrete and most were feasible enough to be manageable for health care providers.

Siblings

Siblings of children with cancer have been called "the forgotten grievers" for a long time. This study confirms that this is also the case for siblings of children with SMA. Even if these siblings' situation has gotten much more attention over the last 10 years, it still needs to be improved.

Conclusions

Although all parents reported having been informed before treatment decisions were made and a vast majority reported feeling confident about the decisions, it is troubling that a quarter of the parents reported that they received no information about respiratory support. This indicates that the parents did not sufficiently understand the available respiratory treatment options, and that their children may not have received the kind of care that is recommended in guidelines (Wang, Finkel et al. 2007).

Our study found that parents' communication with the physician about their wishes and concerns regarding their child's end-of-life care and their preferred location of death contributed to their desires being honored and fulfilled. Arranging for a child with severe SMA to die at home, if the parents so desire, must be facilitated, and the psychological and emotional support to healthy siblings needs to be an improved consideration. Moreover, non-bereaved parents, who were less satisfied with information given by health care staff, should consistently be provided with the information that they need to make decisions about the care and treatment of their child.

Implications for health care

- To create prerequisites in the clinic to follow care guidelines for SMA
- To provide tailored health care services for families living with SMA
- To create a climate conducive to good communication between physicians/health care staff and parents and to encourage parents to express their wishes and concerns for the end-of-life of their child.
- To develop a care system that allows providers the time and resources they need to be fully present with the family during illness and death, and to provide high quality care that is based on established guidelines.
- To avoid neglecting the healthy siblings' situation, inform and encourage their involvement with their severely ill sibling as well as with extended family members, friends, etc. who can be an important source of support for the healthy siblings throughout the illness trajectory and following the loss.

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