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［原著］

Milestones for Communication Development in Japanese Children with Spinal Muscular Atrophy Type I

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Abstract

The objective of this study was to create communication milestones, including the use of communication devices, that enable supporters of children with spinal muscular atrophy (SMA) type I to more easily set appropriate developmental goals. We conducted a questionnaire survey regarding communication skills, in cooperation with the Network for Spinal Muscular Atrophy in Japan, for 58 parents who had children aged 15 years or younger with SMA type I. Responses were received from a total of 36 parents (response rate: 62%). From these responses, developmental milestones consisting of 3 items (communication using devices, communication without using devices, and communication methods) and 19 sub-items were constructed. Although the sample size was small, a developmental course of communication specific to children with SMA type I was evident, and the milestones described in this study are therefore expected to contribute to better communication support.

Keywords: spinal muscular atrophy, SMA type I, milestone, communication device

Introduction

Spinal muscular atrophy (SMA) is an autosomal recessive disorder characterized by muscle atrophy and weakness due to degeneration of the anterior horn cells in the spinal cord[1)]. SMA is classified into three clinical types based on age of onset and control of motor function. Type I, the most severe subtype of SMA, is defined by the International Consortium on SMA as onset between birth and 6 months of age, unable to sit up without support, and death typically before the age of 2 years if artificial ventilation is not applied[2)].

Most children with SMA type I have respiratory muscle weakness and require a tracheostomy, and therefore find it difficult to speak. Furthermore, their severe muscle weakness prevents them from using non-verbal communication such as gestures. Because both

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their verbal and non-verbal communication is severely restricted, health care professionals such as occupational or speech-language-hearing therapists have provided communication support using special devices that exploit well preserved cognitive and physical functionality such as a normal IQ[3] and simple finger movements[4]. Sakai *et al*. (2012) reported that 60.4% of parents of children with SMA type I expected that their children would be able to actively communicate using methods such as special communication devices[5]. This suggests that these children have not received sufficient support from health care professionals such as therapists. One possible reason for this may be that, due to the rarity of SMA type I, health care professionals have little information and experience in relation to support for children with the disorder[6]. In fact, to the best of our knowledge, no studies regarding communication support for children with SMA type I have been reported.

Although developmental milestones are common for healthy children, those for children with disabilities have rarely been reported[7] [8]. However, milestones seem to be valuable because children with the same disease are presumed to follow the same developmental course. By applying milestones, the developmental level of individual children can be compared to others with the same disability, allowing a better understanding of that child's current developmental level and an expectation of the skills that they are likely to acquire in the future. Because children with SMA type I likely follow a similar developmental course specific to the disability[5], and because SMA type I is a rare disease for which information tends to be poor and no treatment has been developed, the creation of milestones is expected to be useful.

The objective of this study was to create communication milestones, including the use of communication devices, that enable supporters of children with SMA type I to more easily set appropriate developmental goals.

Methods

We conducted an anonymous postal questionnaire survey from September to October 2012 in cooperation with the Network for Spinal Muscular Atrophy (NESMA) in Japan among 58 parents who were NESMA members and had children aged 15 years or younger with SMA type I. The study protocol was approved by the Ethics Committee of the Faculty of Health Sciences, Hokkaido University (approval number 12-36). Informed consent was implied by the voluntary return of the questionnaire.

2.1. Questionnaire

To clarify the communication skills of children with SMA type I, the questionnaires asked parents to provide information on the following: (1) all communication methods (from among 10 choices) that were currently being used by their children; (2) the types of electronic communication devices being used, such as voice output communication aids; (3) what their children were able to express using these devices (from among 9 choices, e.g., "Can your child differentiate the use of 'yes' and 'no'?") ; (4) the types of non-electronic communication devices being used, such as communication boards; (5) what their children were able to express using these devices (from among 9 choices; questions identical to those in (3) above) ; and (6) parents' free responses as to what their children were able to express through other methods of communication.

2.2. Communication Milestones

Communication milestones for children with SMA type I were developed based on their age and their parents' responses to questions (1) through (6) above. The milestones were classified into the following three major items:

1. Communication using devices

This item was created based on responses regarding the children's communicative skills using electronic or non-electronic devices

(questions (3) and (5)). Achievement norms for each sub-item corresponded to age, in ascending order from youngest to oldest, of the first, the second to fourth, and the fifth child. Although, for example, the DENVER II Developmental Screening Test uses age-based norms set to 25%, 50%, 75%, and 90% of the children successfully passing check-list items[9], we adopted similar achievement norms based on the number of children so that we could provide valuable information even in a small sample. In addition, the youngest children who had not yet achieved the sub-items, but were using communication devices, were also noted.

2. Communication without using devices

This item was created based on responses to question (6) regarding current communication skills such as vocalizing, finger pointing, and sign usage. Each sub-item was classified according to what they were able to express regardless of the method. The expressions that more than 5 children had accomplished were used as sub-items, and achievement norms were adopted in the same manner as described above in "Communication using devices".

3. Communication methods

This item was created to assess the current communication methods that children were using at the time their parents received and answered the questionnaires. Sub-items were constructed based on whether the children were currently using particular methods. The communication methods that more than 5 children had used were adopted as sub-items.

Results

Completed questionnaires were collected from a total of 36 children (17 boys, 19 girls; median age, 4 years 5 months; age range, 11 months to 15 years) (response rate: 62%).

Regarding demographic data, the five communication methods most frequently used by the children were as follows: signs (e.g., eye movements indicating 'yes' or 'no'; n=18, 50%) ; eye fixation (n=17, 47.2%) ; electronic communication devices (n=17, 47.2%) ; vocalizing (n=11, 30.6%) ; and non-electronic communication devices (n=8, 22.2%). Meanwhile, no communication methods were used by 19.4% (n=7) of the children to convey their intentions. The three most frequently used electronic communication devices were as follows: Let's Chat (Panasonic Corp., Osaka, Japan) (n=11) ; Tobii™ Communicator (Tobii Technology, Ltd., Tokyo, Japan) (n=4) ; and SuperTalker™ Progressive Communicator (Ablenet, Inc., Roseville, United States) (n=2). The three most frequently used non-electronic communication devices were picture cards (n=3), communication boards (n=2), and whiteboards (n=2). Milestones were created using these data (Figure 1).

Discussion

Some characteristic tendencies were observed in relation to the milestones. First, the ability to communicate without using devices, such as through eye fixation, vocalization, or use of signs, tended to be acquired earlier than the ability to communicate using electronic or non-electronic devices. The developmental order from eye fixation to using signs was the same, even though it was later, than that of healthy children, which suggests that use of communication devices is important for language development. Second, regarding communication devices, use of an electronic device was reported by a 3-year 4-month-old child, as was use of a device to express intention by a 4-year 1-month-old child. Although the ages of achievement among the children were different, these milestones suggest that some children were able to use devices by the age of 4 years. Finally, it was difficult to "call people" without using a device, because even if children could vocalize, they were unable to use a loud voice. However, the ability of children with SMA type I to use a communication device is thought

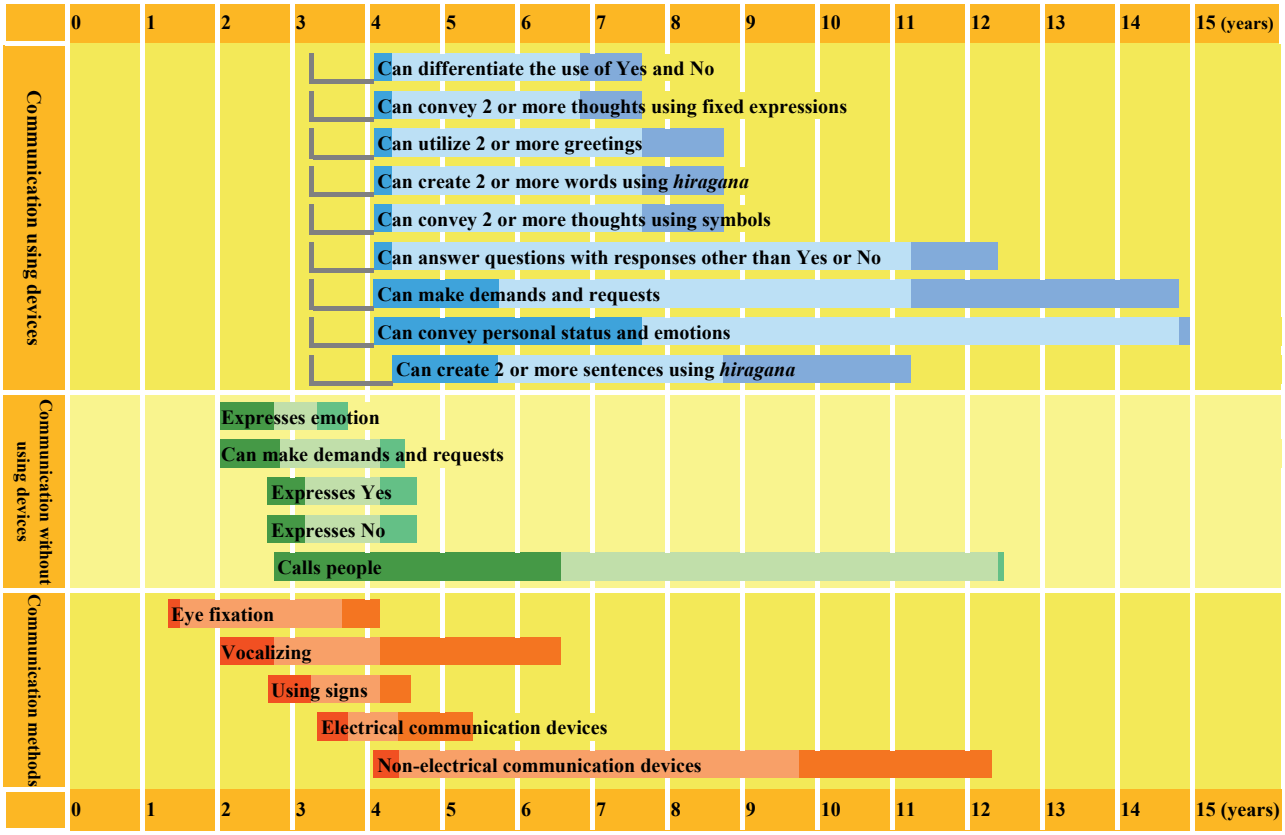

Figure 1. Milestones developed in this study for communication development in children with spinal muscular atrophy type I

Items in "Communication using devices" represent the development of communication methods using both electronic communication devices (e.g., voice output communication aids) and non-electronic communication devices (e.g., communication boards) . Items in "Communication without using devices" represent the development of communication methods including signs, eye fixation, and vocalizing.

[Dark colors] represent milestone achievement by 1 child,

[light colors] represent milestone achievement by 2 to 4 children, and

[dull colors] represent milestone achievement by 5 children. Milestone achievement by the sixth and subsequent children has been omitted. The └ [symbol] represents the youngest among the children with SMA type I who had been using communication devices but had not yet achieved that particular milestone.

Method of use

Please check the age of the subject child against these "Development milestones". Please confirm approximately how many children with spinal muscular atrophy type I of the same age have achieved each item, and judge whether the child also appears capable of achieving the item. For example, for items that have been achieved by even one person, the child may be able to achieve the item with practice. For items that have been achieved by five people, the child may be able to achieve the item with practice relatively easily. For specific practice methods, please consult specialists such as an occupational therapist or a speech-language-hearing therapist.

to be essential, particularly in emergency situations such as when having difficulty breathing.

The milestones developed in this study offer supporters of children with SMA type I information on a general developmental course in relation to communication skills, such as typical age of achievement of various communication skills. By applying these milestones, supporters will be able to compare the developmental level of a particular child with SMA type I to others with the same disorder, and to have more accurate expectations of the methods or expressions that the child is likely to acquire in the future; therefore, we believe that these milestones would contribute to more appropriate goal setting by supporters for individual children.

A major limitation of this study was the small sample size. If we had requested that parents answered questions regarding the past, this problem may have been somewhat alleviated. However, we requested that parents answer questions concerning what the children were able to do at the present time because we did not want the parents to provide inaccurate responses in retrospect. Nevertheless, because SMA type I is a rare disease for which information tends to be poor and no treatment has been developed, and the purpose of the milestones is not to screen, but rather to provide support based on understanding the course of communication development in children with SMA type I, this limitation is not thought to have affected the research findings.

In order to provide the best support possible, these milestones will continue to require updates and revisions as new communication devices are developed and utilized in the future.

Conclusions

Although the sample size was small, the communication milestones described in this study suggested a developmental course of communication specific to children with SMA type I. These milestones are expected to contribute to communication support for such children, especially those at a younger age.

Acknowledgements

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Conflicts of interest

The authors declare that they have no conflicts.

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