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In-home Learning Support for Preschooler Suffering from Osteogenesis Imperfecta

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Abstract: For normal pre-school children, social and cultural learning is promoted through play. These experiences form the basis for learning as they move on in life after preschool. However, for children with severe motor and intellectual disabilities or intractable diseases that restrict opportunities for interpersonal inter-changes and being able to leave their homes or care facilities, the opportunities to be influenced by their peers to stimulate their development are very limited, which presents a serious problem. Therefore, finding ways to provide a substitute for this aspect of development and learning for such incapacitated preschoolers is an important problem, but little attention has been given to the need for this kind of early development support. This study reports on an experiment in learning support using home visits to a preschooler with Osteogenesis Imperfecta. We visited and provided in-home educational support by directing learning activities about Japanese cursive characters using a tablet-type device and a bowling game to interest the child. Through instructions, this child was able to improve reading abilities and recognition of quantities. This gives confirmation, to an extent, that in-home teaching activities can be effective in engaging home-bound children in eagerly learning information that is important in daily life. One conclusion is that such interventions may enhance the prognosis for learning for children with intractable diseases.

Key words: child with Osteogenesis Imperfecta, In-home learning support, preschooler

1. Introduction

Osteogenesis imperfecta (OI) is a congenital condition characterized by fragile bones and defective connective tissue. The symptoms and severity of OI widely vary across cases. The Sillence Classification is widely used to categorize OI based on its features. Treatments for increased susceptibility to fractures have advanced, and the underlying causes of the condition have been more fully understood in recent years.

Japanese guidelines on OI, drafted in 2006 (Tanaka, Tanaka, Kanzaki, Sugihara, Yokotani, Hasegawa, Harada, Fujieda, 2006), include survey data on the Sillence Classification of Japanese patients with OI according to a survey, in which, 31.7% of patients had type I, 1.7% had type II, 15.7% had type III, and 24.3% had type IV OI; the remaining 26.5% could not be classified. OI is reported to have an incidence of 1 in 20,000.

The symptoms exhibited by children with OI vary from mild to severe. These children have a wide range of intellectual capacities; therefore, some attend a regular school and others a school for students with special needs, depending on their condition. One important indicator of the severity of OI is whether the child is ambulatory at the age when he or she should normally enter elementary school. A previous study (Tanaka, 2015) contended that if a child with OI had difficulty walking, it was crucial

to maintain the maximum level of upper limb function. However, research has not yet arrived at a clear consensus on the best educational approach for children who have difficulty walking.

New treatments for OI have become available because of continuing advances in medicine and pharmacology, creating new options for both the treatment and upbringing of children with OI.

We have described a case of a preschooler with OI who received academic support from a home tutor and also regarding the child's learning process and tutoring content. Based on this information, we discuss the effectiveness of early academic support for children with OI who have limited opportunities to learn.

2. Case Presentation

The child, referred to as Case A, was a 6-year-old boy who had been diagnosed with OI congenita (type II). A college student (recruited through an advertisement for a "volunteer to visit a child with an illness or disability") and the current author provided academic support for Case A twice a month as home tutors.

Case A was born at 32 weeks of gestation and subsequently spent about 1 year in the neonatal intensive care unit. At 6 months of age, he underwent a tracheotomy. He has particularly weak lower limbs and ribs, which are highly susceptible to pressure from outside the body. Therefore, he often requires special adjustments to be able to sit in a seated position, although he can retain that position for short periods, such as during physical therapy exercises. In everyday life, he often adopts a supine or lateral position, can turn over in bed and manipulate objects (e.g., grasping a pen or toy), and move around indoors by crawling on his back. At the age of 3 years, physical therapy was initiated, and he began receiving tutoring at a local day care center. Case A suffers several fractures a year in the upper arms and hips. During treatment, his activities of daily living are limited.

Before beginning the research, all ethical regulations were explained to Case A and his guardian. They were assured that the support would continue to be provided and that they would not be penalized in any way even if they withdrew their consent to participate in this study. They were informed that the data collected in this study would be included in a research manuscript. They gave consent with the stipulation that their personal information would be safeguarded and privacy protected. These provisions were communicated in writing, and the study began only after receiving written consent from Case A's guardian.

Our assessment of Case A covered his developmental and academic levels. Restrictions related to Case A's disability precluded the administration of face-to-face development tests, so the Enjoji Scale of Infant Analytical Development (Enjoji, 1977) and an academic achievement checklist (Tokunaga, 2014) were used. His guardian was interviewed during testing. In addition, his behavior was observed through a video and was analyzed by a clinical psychologist specializing in developmental psychology.

His chronological age was 72 months at the time of study, but his posture, movement, and development were greatly limited because of his disability. His locomotion and hand movement was equivalent to that of a 7-month-old and 36-month-old, respectively.

His language comprehension and speech was equivalent to that of a 4-year-old and 36-month-old, respectively. Due to his tracheotomy, he had difficulty articulating some sounds and part of his speech was unclear. However, he could converse by speaking slowly and repeating his words when necessary. His formal language comprehension and expression had developed through watching and listening to television programs; furthermore, if he failed to comprehend a topic in conversation, he would instead begin talking about his topic of interest.

His basic social skills were equivalent to those of a 24-month old. His developmental level was deemed to be limited as items assessed at 24 months and beyond are related to postural adjustments

and movement. In terms of interpersonal relationship skills, his development age was 27 months. Because of fewer opportunities to leave the house or interact with children of his age, he had limited learning opportunities.

On the academic achievement checklist, Case A attained 60, 48, 36, 24, and 6 points for "talking;" "listening," "reading," and "writing;" "amounts and measurements" and "shapes;" "routine activities;" and "movement and activity," respectively. The scores on this checklist indicate a child's approximate developmental age in each area. Therefore, Case A exhibited basic academic skills equivalent to a 4- or 5-year-old, but he had relatively low scores in areas requiring repeated hands-on learning. This was presumably because of the very high risk of fractures restricting his ability to leave the house or engage in certain activities. It also resulted in frequent interruptions in learning.

3. The tutoring plan

Case A's guardian was interviewed to formulate a tutoring plan. The guardian indicated a desire for Case A to interact with various people since he had interacted with only family and staff at the hospital and day care center. Case A also had limited experience in socializing because of his illness. Furthermore, the guardian wanted Case A to engage in developmental tasks such as following social etiquette (e.g., waiting one's turn) and understanding when an activity or event was over.

The guardian indicated that Case A was unable to read some hiragana (basic Japanese characters akin to letters in the English alphabet) and he often misunderstood hiragana characters with a similar phonology and gradually began to understand the concept of numbers and counting. Considering this, the guardian wanted Case A to receive basic tutoring in words and counting.

Tutoring took place once every 2 weeks at Case A's home; sessions were not held if he was in poor health. The tutoring lasted 5 months, with a total of 10 sessions. The guardian and current author were present at all sessions. As he had undergone a tracheotomy, sessions were interrupted when sputum needed to be aspirated, which was managed by the guardian. Case A's condition was checked before starting each session, and he was taught for 45 min, after which the tutor discussed the session with the guardian. Each visit lasted for approximately 60 min.

With the consent of Case A and his guardian, each session was recorded. After each session, the effectiveness of tutoring was reviewed based on Case A's behavior and speech during the session. In addition, Case A's actions were observed to determine whether his posture would increase the potential for a fracture. Video review assisted in planning the next session.

The tutoring covered the four areas described below.

(1) Reading picture books

In this, Case A learned through looking at pictures and words (improving his visual cognition) and conversations about materials. Specifically, the tutor read a picture book aloud and questioned Case A questions about it. To encourage Case A to elaborate, the tutor would ask him questions like "What is happening?" "In what way?," and "Why?"

(2) Crafts

Case A acquired fine finger movements through hands-on learning, which provided him a sense of accomplishment. He completed craft projects such as origami or making a rocket from paper cups by following the tutor's instructions or looking at an example.

(3) Learning hiragana

Case A learned to differentiate hiragana characters and to improve his pronunciation and speech. He learned by looking at a sample character on a tablet personal computer (PC) and responded with the name of that character.

(4) Learning to count

Case A learned to identify and recite numbers and to decide whether a given amount was a lot or little. The tutor and Case A played a bowling game in which A counted how many pins had fallen and recorded that number on a board.

4. Progress of tutoring

In the following descriptions of the actual tutoring content by session, the sessions are designated as S1–S10 and words spoken by the tutor are in quotation marks and those by A are italicized and in quotation marks.

Given Case A's development level, the picture books used had relatively few Japanese characters, simple language, color illustrations or a theme appropriate to the season. Initially (S1 and S2), the tutor read aloud to Case A. In S3–S6, the tutor questioned Case A and prompted him to explain his understanding of the story in conversation. Finally, in S7–S10, the activity involved shared roles, with Case A and the tutor reading aloud.

At the beginning of tutoring (S1 and S2), the tutor would point to a passage in the picture book and ask, "What does it say here?" If Case A was able to read what was written, he would immediately answer, if not, he would say "I don't understand" or "Mom, what does this say?"

If a passage had words that Case A did not understand, he would remain silent. The tutor would then point to each character, read it aloud, and prompt him to repeat the character name. Eventually, Case A was able to repeat the character name. When a picture book had a seasonal theme (e.g., Christmas), Case A would think about his own life and describe his experiences.

Starting in S6, Case A made remarks like "I don't understand" less often and voluntarily read characters on some occasions. However, he still mistook some characters for similar-looking ones (e.g., reading "Ru" as "Ro"). The tutor would get Case A's attention and point to the character saying, "Try again. Look here." When Case A realized his error and read aloud the character correctly, the tutor commended him by saying something like "That's right. That's the right answer." If Case A failed to realize his mistake, the tutor would read aloud the character correctly and instruct him to repeat the character name. After S7, Case A made fewer errors while reading and was able to read long passages.

Crafts involved seasonal projects, with the tutor showing Case A an example and encouraging him to make the object in question. Initially (S2), Case A responded, "I can't make that." The tutor encouraged Case A, saying "I'll teach you how I made this and we can make one together." Upon completing a paper cup rocket, Case A beamed as he blasted the rocket around the room. When crafts were taught at S4 and thereafter, Case A repeatedly remarked, "How do you make that?" or "Tell me [how you did it]."

To encourage Case A's active involvement in learning hiragana, he was taught on a tablet PC. During S1 and S2, Case A paid little attention to the tutor's words or directions and would repeatedly touch the screen. Series of instructional steps "touch the screen … the screen changes … a sound comes out" riveted his attention. After S3, Case A had become accustomed to the tablet PC and educational software. In response to a question by the tutor, he would touch the tablet's screen and take an extended period of time to compare the sample and the answer choices. After S7, Case A answered ≥17 questions correctly out of the 20 in a task that required matching a particular character. However, if the matching task involved an easily misread character and Case A answered incorrectly, he would touch various places on the tablet PC in confusion.

Case A was also taught to draw lines and recognize character shapes. To teach Case A to place his finger on the screen of the tablet PC, the tutor would hold his finger and show him how to move it. Case A would then be asked to answer. Ultimately, by S9, A was able to correctly match all of the hiragana in a character matching task.

The bowling task described above was used to teach numbers and counting. It was designed to teach Case A to point out and recite numbers and comprehend the concept of amounts. During S2 and S3, the tutor and Case A competed to see who could knock down the most pins of the 10 standing. During S4 and S5, the tutor asked Case A to count the number of pins that had fallen. Case A would point and count, but would often count incorrectly. When the tutor asked Case A, "How many pins are left?" he responded with the correct number. When all the pins fell, he was unable to understand all 10 pins had fallen; when Case A and the tutor knocked over the same number of pins, he did not understand that the score was a tie. Given Case A's demonstrated limitations, only five bowling pins were used after S6, and the pins that fell were placed in a holder so that A could more easily visualize the number.

When counting small numbers of pins, Case A practiced by successively counting numbers while pointing at each pin from a distance. He was able to accurately determine amounts up to three. Eventually, when the tutor and Case A knocked over the same number of pins, he would remark, "We knocked over the same number, so it's a tie." During S10, Case A was still answering incorrectly regarding amounts up to five, but in some cases he was able to answer correctly when prompted to count again.

5. Effectiveness of tutoring

The Enjoji Scale of Infant Analytical Development was administered at the conclusion of tutoring, and there were no changes from the results prior to tutoring. However, on the academic achievement checklist, Case A's scores for "reading" and "numbers and counting" improved by 12 points each. Five months had passed since the start of tutoring. These improved scores were presumably the result of tutoring.

During the conversations after the tutoring sessions, Case A's guardian indicated that he was making progress in hiragana since he was able to read characters on signs when he left the house. Case A also said "I want to write a letter to my tutor" in anticipation of the tutor's visit, and he wrote as he spoke aloud what he wanted to write.

Case A suffered two fractures during everyday activities while tutoring was taking place. At the behest of Case A and his guardian and in the light of instructions from a physician, the learning setting was modified and learning activities continued.

6. Discussion

This report describes a case of a preschooler with OI who received academic support from a home tutor, documenting the effectiveness of early academic support for a child with limited learning opportunities due to OI.

Learning is defined as "a given experience in a certain setting that causes or that has the potential to cause a change in an individual's behavior in the same or a similar setting." (Vandenbos, 2007). Learning can be divided into two types: intentional learning, based on a motivation to learn and incidental or spontaneous learning, with no expressed motivation to learn (Tatsuno, 2009). In the present case, tutoring was intended to foster the basic ability to learn before a child started school. However, Case A probably perceived working with the tutor as an extension of play. Given the limitations posed by Case A's OI and development level, a heavy emphasis on intentional learning would not have been appropriate. Instead, Case A needed to be given opportunities for structured incidental learning through play.

Case A was given the opportunity to notice seasonal changes in his surroundings through picture

books and crafts, to experience words and counting, and to discover that symbols present in his world actually carried meaning. These opportunities helped him to learn the material that was taught.

Home visits by tutors presumably had a positive effect by allowing Case A to encounter new sources of information (i.e., new people). Over the last few years, developmental studies (e.g., Harris, 2007; Corriveau & Harris, 2009) have examined the mechanisms of social and cultural learning in early childhood and have revealed that preschoolers learn from information disseminated by others with whom they are familiar and whom they trust (particularly adults). While interacting with an adult who knows and disseminates information, a child spontaneously takes in knowledge by observing and imitating that person's actions. However, children with a disability or illness often have limited opportunities for social and cultural learning because of the extent of their functional disability and the commensurate limitations on their activities. Because he interacted with a limited number of people in a limited number of settings, Case A's opportunities to learn were severely restricted.

Diligent support from a caregiver and the involvement of the staff of a medical facility and a day care center provided Case A with a safe, stable foundation for daily living. However, for greater learning to occur, this foundation needed to be complemented by further social interaction. The use of a tablet PC as a teaching tool facilitated Case A's learning.

Based on passages in the Manual on Educational Use of ICT (Ministry of education, culture, sports, science and technology-Japan, 2010), schools for students with special needs have introduced ICT devices that facilitate learning for children with certain disabilities. "Use of various assistive technologies and interaction with other people" allow young students with multiple disabilities "to better express themselves by making what they want to say clearer or by helping them to find other ways to express themselves." In the present case, Case A enjoyed touching the tablet PC for some time after he was shown it, but he did not listen to or pay attention to what the tutor was saying. Looking solely at this stage, one might mistakenly believe that the introduction of ICT was distracting him and thereby hampering his education. A tutor should use ICT with an eye toward transitioning from its initial introduction (becoming familiar with an ICT device, which may induce fascination) to its practical use (increasing the effectiveness of instruction).

The inability to perform an action because of a disability or illness should not be automatically viewed as a lack of ability. Instead, suitable customized learning opportunities should be provided, and educational materials and teaching tools should be used in accordance with how a child learns. A tutor and a child should have shared interest in what is being taught and should communicate in a synergistic and complementary manner (Tomasello, 1999). A child needs to experience "understanding—the ability to apply what is learned—the joy of learning."

Efforts such as the tutoring of Case A described in this article are designed to improve the academic outlook for children with severe motor and physical disabilities or intractable diseases are of great significance. In Japan, a day care program for children and individuals with severe motor and intellectual disabilities has been established by law, but numerous implementation problems have been reported (Mito, Takashima, Suemitu, 2014), such as the limited number of day care facilities, problems with transportation to and from those facilities, and deficiencies in medical care systems. Medical advances offer a longer life and alleviation of symptoms for such children, but facilities and support systems must be in place to improve their quality of life. In the present case, the academic support provided by a home tutor was not part of a system of public support, but rather, was delivered by a university department. Nonetheless, it demonstrates the type of practice that can improve the academic outlook for children with disabilities.

Once successful evidence regarding such academic support is available, it could serve as the basis for system and facility improvements. A neurologically typical child spontaneously learns concepts as part of life before starting school even if he or she is not intentionally taught hiragana or

how to recognize and count numbers. This is because such a child is given a host of opportunities to learn through mistakes. In contrast, things that should be learned spontaneously through daily life are difficult for children (and other individuals) with severe motor and intellectual disabilities or an intractable disease to learn because of their more limited experience and exposure. If these children are structurally given a setting for intentional learning before starting school, they can be expected to develop a greater interest in learning once they reach school age.

One limitation of the present report is that it examined only one case. Another limitation is that the material taught and the changes in the child's learning could not be objectively assessed. Multiple case studies should be conducted and compared so that we can gain greater understanding of how to best provide early learning support for children with severe disabilities or an intractable disease. In addition, differences between caring for and teaching preschoolers in the community and home education by a tutor should be examined. I hope to assemble evidence on the impact of the home experience on children with developmental limitations due to severe disabilities or an intractable disease and to compile the research findings so as to enhance coordination between medical personnel, teachers, and welfare service providers.

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