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EDUCATIONAL NEEDS OF CHILDREN WITH RARE DISEASES AND LONG-TERM HOME VENTILATION (SMARD1) - CASE STUDY

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SUMMARY

Diagnosing a child with a rare disease that involves acute respiratory distress dramatically changes his/ her educational situation. Therefore, it is important to determine the child's educational needs which will help select an adequate teaching strategy. One of such diseases is Spinal Muscular Atrophy with Respiratory Distress Type 1 (SMARD1). In Poland, two children were diagnosed with it, whereas around the world there are over 60 cases. The study encompasses one boy with SMARD1. The purpose of the study was to describe the child's educational needs, as well as to indicate which of them are related to the rare disease and which to respiratory distress that requires mechanical ventilation. Based on the performed study, it was concluded that not all educational needs related to the rare disease were present. On the other hand, all needs related to home ventilation were present.

Key words: children with educational needs, Spinal Muscular Atrophy with Respiratory Distress Type 1, rare disease

INTRODUCTION

Researchers of various academic disciplines, including teachers, speech therapists, sociologists and psychologists started to tackle the issue of education of children with rare diseases more and more frequently. Most often, such studies refer to a specific disease. In spite of the increasing interest in this subject, academic publications describing educational needs of children with rare diseases and home ventilation are still missing. In the conducted study, the author assumed that "a rare disease is a health condition that affects a small number of people compared with other prevalent diseases in the general population. To date, between 5000 and 8000 distinct rare diseases have been documented" (Richter, Nestler-Parr, Babela et al., 2015: 2). Prevalence of a rare disease may vary, depending on the continent, country and region. In one state, it may be a rare disease, whereas in another one a disease with high prevalence. In Europe it is assumed that a rare disease affects 5 out of 1000 people. Thus, it may be concluded that in the European Union, there are 26-27 million people suffering from such diseases, whereas in the United States, 25 million people with such diagnosis (Richter, Nestler-Parr, Babela et al., 2015). The author also assumed, following Isobel Brooks, that long-term ventilation (LTV) is: "dependence on mechanical ventilation, whatever the interface, for all or part of a 24 hour period, for at least the last three months, in a medically stable child or young person." The term LTV is generally used to describe patients where the intention is to deliver ongoing ventilation at home. The interface used to deliver ventilation is either a tracheostomy - so called "invasive ventilation", or a mask (which may be full face, nasal, mouthpiece or face-shield) - "non invasive ventilation" (Brooks, 2019: 167).

Spinal Muscular Atrophy with Respiratory Distress Type 1 (SMARD1) is a rare disease and its exact prevalence remains unknown. So far, over 60 people have been diagnosed around the world (Eckart, Guenther, Idkowiak et al., 2012). At the present moment in Poland, there are two children with SMARD1 (four cases were diagnosed, two children died). It is inherited in an autosomal recessive pattern. SMARD1 is known to be caused by changes (mutations) in the IGHMBP2 gene (Grohmann, Schulke, Diers et al., 2001). Symptoms are already noticeable in the pre-natal period. Almost 1/3 of children are born prematurely, most often in the 37th week of pregnancy (some sources inform about 34th – 35th week of pregnancy). Other characteristics include low birth weight and weak sucking reflex (Porro, Rinchetti, Magri et al., 2014). SMARD1 symptoms may be classified into three groups: respiratory tract, nerve and muscle system and autonomous and sensory nervous system (figure 1). The first symptoms include respiratory distress along with progressing muscle weakness. Based on case studies, the researchers noted that frequent symptoms of respiratory distress include inspiratory stridor and/ or silent cry. As noted by Porro et colleagues it is also significant that “the diaphragmatic paralysis arises as dyspnea, with eventration of one or both hemidiaphragms and ultimately requires permanent respiratory support” (Porro, Rinchetti, Magri et al., 2014: 36). As far as muscle weakness is concerned, it initially affects distal muscles. It is usually more intense in lower extremities. Additionally, children may manifest elevated pain threshold, neurogenic bladder, elevated liver enzyme levels, problems with gastrointestinal system and cardiological problems (Porro, Rinchetti, Magri et al., 2014).

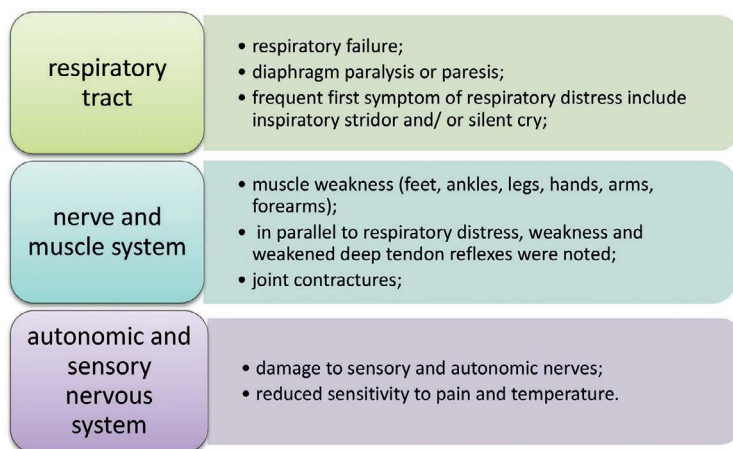


Figure 1. Spinal Muscular Atrophy with Respiratory Distress Type 1 clinical signs and symptoms Source: M. Jędrzejowska (2010). *Przeponowa postać rdzeniowego zaniku mięśni (SMARD1)*, *Neurologia Dziecięca*, vol. 19 (38), pp. 51-54, K. Grohmann, R. Varon, P. Stolz et al. (2003). *Infantile Spinal Muscular Atrophy with Respiratory Distress Type 1 (SMARD1)*. American Neurological Association, Vol. 54, No. 6, pp. 719-724, A. M. Kaind, U. P. Guenther, S. Rudnik-Schöneborn et al. (2008). *Spinal Muscular Atrophy with Respiratory Distress Type 1 (SMARD1)*. *Journal of Child Neurology*, Vol. 23, No. 2, pp. 199-204.

Child With a Rare Disease and Home Ventilation Vs. Education

The priority, as far as care for a child with mechanical ventilation is concerned, is stabilisation of the child's health condition. This allows for the child's discharge from a medical facility where he/she was subject to intense paediatric care to home environment. The stage of health condition stabilisation is a time deprived of experiences that allow for the child's development. Return home is difficult, because the time of hospitalisation could have exposed the child to improper types and levels of stimulation, which is related to the absence of positive and adequate experiences based on interactions with other persons. Thus, intensive medical care in hospital conditions cannot provide support in the realm of developmental, social and educational needs (Hewitt-Taylor, 2004).

Anna Ludvigsen and Jan Morrison, based on a study performed in the area of Great Britain focused on support for children with home ventilation, indicated problems that such children may experience. Among them, the following ones were listed:

- "Barriers to discharge resulting in prolonged hospitalisation, in particular delayed transfer from intensive care, and increased risk of infection,
- Negative impact on quality of life, in particular privacy and dignity and effect on independence,
- Lack of consistency in short break respite provision" (Lawrence 2011: 20; Ludvigsen & Morrison, 2003: 3).

The situation is even more complicated when the child has been diagnosed with a rare disease and participates in clinical trials or undergoes experimental treatment. Frequency of hospital stays related to it, as well as the fact that the child is primarily surrounded by adults may lower the level of the child's psycho-social functioning. Return home is also often related to the commencement or continuation of the interrupted educational process, both on the pre-school and school level. The majority of children with mechanical ventilation are provided with individual teaching or they attend integration kindergartens or schools.

Educational Needs of a Child With Rare Disease and Home Ventilation

Thinking about educational needs of a child with difficulties in development and/or a disability is changing. This results, among others, from the fact that the modern studies provide us with medical data and information pertaining to the development of cognitive functions which supplement the hitherto level of knowledge about the disease or overthrow existing theories about it. This also translates to the education of children with disabilities, as well as the process of their therapy. In literature, educational needs of children with difficulties in development are called special educational needs (SEN) and are defined as: "... a restriction in the capacity of the person to participate in and benefit from education on account of an enduring physical, sensory, mental health or learning disability, or any other condition which results in a person learning differently from a person without that condition" (Griffin, 2014: 10).

In individual countries, definitions of SEN may vary, yet their common element is the fact that they refer to students who experience difficulties - in contrast to their peers - that make learning difficult for them, at every stage of education, also pre-school education (Malbogot, 2017). The aforementioned difficulties may refer to, among others: hearing and sight defects, neurological disorders, movement disorders, etc. (figure 2).

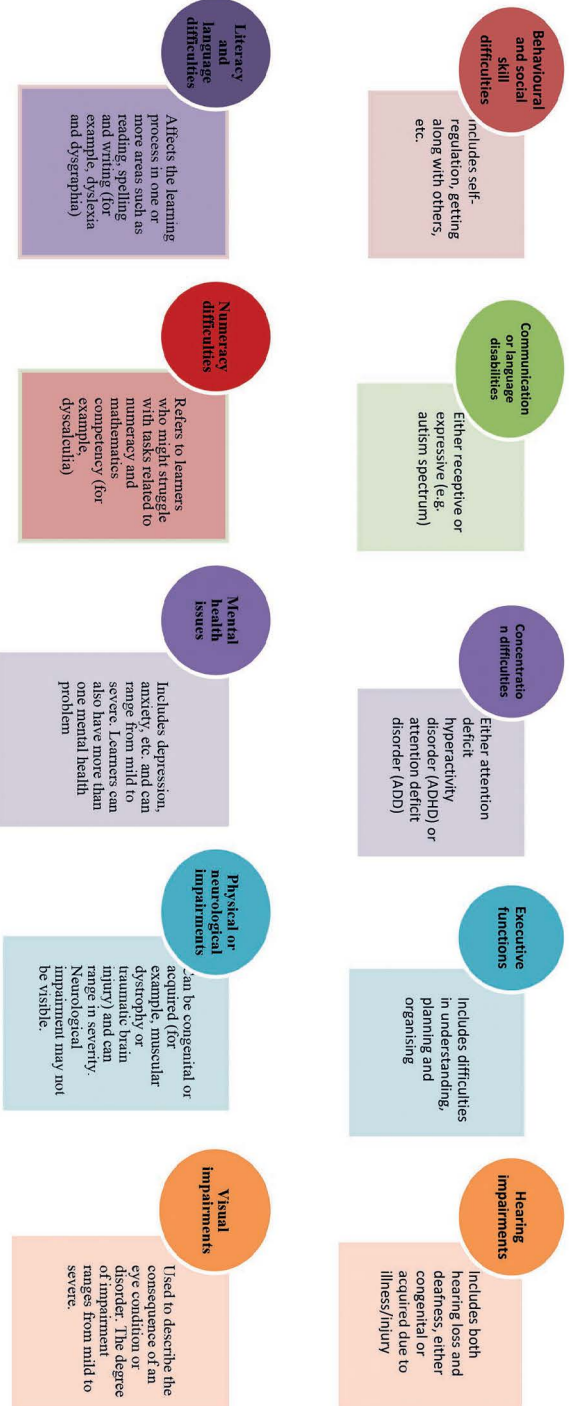


Figure 2. Categories of educational needs

Source: Malbogat, L. (2014). Special educational needs. Cambridge Assessment International Education, IGCSE

Diagnosis of disruptions in development or a disability may not affect education and in such case, no additional support is necessary. On the other side, the experienced development or health difficulties may hinder the process of education when there is no such additional support. Staying in a classroom together with peers, learning in teams may sometimes offer sufficient support, in spite of the fact that it takes place without the teacher’s extra support or additional assistance in the form of adjusted teaching strategies. Other students with SEN may need additional didactic support and/ or care and assisting technologies. Irrespective of the child’s needs, it is possible to guarantee diversified educational programme and qualified teaching personnel (Griffin, 2014: 21).

In the conducted study, the author assumed that a child with a rare disease and home ventilation experiences limitations in participation in the educational process. This results from current health condition of the child which requires changes in the strategy of teaching and designates educational needs different than the needs of healthy peers. Simultaneously, the author acknowledges the conclusions from studies carried out by Gernsbachear, Raimond Balinghasay, & Boston (2016) which showed that the term “special educational needs” is perceived more negatively by adults (a sample of 530 persons) than the concept of disability. Thus, in the study the author assumed that a child with a rare disease and home ventilation has different educational needs than his/ her healthy peers, yet their character is individual and personalised, but not special. Based on an analysis of literature devoted to the functioning of a child with Spinal Muscular Atrophy with Respiratory Distress Type 1, the author singled out the child’s educational needs referring both to pre-school and early school age. They refer to the main symptoms of SMARD1 taking into account, among others, muscle weakness and respiratory distress that require home ventilation and limitations in participation in education outside of home related to it.

Table 1. *Symptoms of Spinal Muscular Atrophy with Respiratory Distress Type 1. Clinical signs and symptoms vs. the child’s educational needs*

Spinal Muscular Atrophy with Respiratory Distress Type 1 clinical signs and symptoms	Educational needs
weak neck and trunk muscles, difficulties with holding head up	due to greater fatigue, more frequent breaks during work, division of tasks into stages and materials into smaller batches that allow for keeping the level of knowledge taught or the course or the train of thought;
no possibility to change body position, lift arms, turn sideways	preparation of the work place, adjustment of teaching materials and form of tasks and exercises to limited mobility (e.g. a possibility of eye-pointing, preparation of materials that are aimed at expanding experience in the realm of touch, including Newtonian fluid in small containers that can be moved and put close to the child); adaptation of work sheets, school textbooks admitted for school use (e.g. proper positioning of drawings and photographs, specific size of text on a page with larger side margins); taking the individual communication system into account, supporting messages with graphics and gestures during communication in turns;

no head movement which limits the field of vision	<p>presentation of teaching aids, instructions for tasks to be performed within the child's field of vision, most frequently in front of the child and at a proper height;</p> <p>adjustment of the work place: proper positioning of the wheelchair or the child's seat to allow him/ her observe what is going on around him/ her, who is entering and exiting the room, etc.;</p> <p>adaptation of work sheets, school textbooks admitted for school use (e.g. proper positioning of drawings and photographs, specific size of text on a page with larger side margins);</p> <p>taking the individual communication system into account, supporting messages with graphics and gestures during communication in turns;</p>
respiratory distress resulting in mechanical home ventilation	<p>application of the demonstrative method, enabling multi-sensory cognition;</p> <p>combining the content taught with the closest environment of the child's life and education, as well as individual experience which is limited on account of no movement; whilst teaching, taking the individual system of communication into account, e.g. instructions for tasks supported with gestures and graphics;</p> <p>necessity of functional teaching, based on the child's functioning, his/ her activities, engagement, experiences, discoveries and cognition;</p> <p>boosting self-confidence during activities in groups and individual tasks;</p> <p>extension of work time;</p> <p>adjustment of the work place: proper positioning of the child's wheelchair or seat so that he/ she can fully participate in group activities;</p>
no milestones accomplished, i.e. sitting, crawling, walking	<p>combining the teaching content with the closest environment of the child's life and education, as well as individual experiences that are limited on account of lack of mobility;</p> <p>necessity of functional teaching, based on the child's functioning, his/ her activities, engagement, experiences, discoveries and cognition;</p>
elevated pain threshold	<p>the necessity to work out principles pertaining to classes based on tactile experiences, i.e. learning the temperature of water by putting a foot in a container with water;</p>
no verbal speech ^a , facial mimicry, possibility of alternative communication	<p>use of devices and techniques designed for communication, preparation of instructions for tasks supported with graphics, pictograms and/ or gestures;</p> <p>application of a broad array of inclusive teaching methods, adjusted to the objectives of the class;</p>

^a Some children diagnosed with SMARD1 have verbal speech; however, it is characterised by articulatory disruptions, as well as disrupted (short) pattern of breathing.

auditory hypersensitivity	adjustment of the place of classes (e.g. lining that absorbs noise, classroom located at a distance from places generating noise); limitation of sudden, unexpected sounds from the environment, e.g. bursts of laughter, elevated voice of the teacher, possibility of using muffling headphones, resignation from aids and toys generating high-pitched sounds;
visual hypersensitivity (sunlight, bright lights in the classroom)	adjustment of the place of work by use of lights with a possibility of intensity regulation; limitation of sudden and diverse visual stimuli by, e.g. a cap with a visor, or sunglasses.

Source: Author's own study based on Eckart M., Guenther UP., Idkowiak J., Varon R., Grolle B., Boffi P., Van Maldergem L., Hübner C., Schuelke M., von Au K. (2012). The natural course of infantile spinal muscular atrophy with respiratory distress type 1 (SMARD1). *Pediatrics*, Vol. 129 (1), pp. 148-56, Kaind A. M., Guenther U. P., Rudnik-Schöneborn S. *et al.* (2008). Spinal Muscular Atrophy with Respiratory Distress Type 1 (SMARD1). *Journal of Child Neurology*, Vol. 23, No. 2, pp. 199-204, Kamyk-Wawryszak A. (2019). *Educational needs of preschool children with recognized rare disease and intellectual disability - teachers' perspective*, *Journal Plus Education*, Vol. XXIV, Special issue, pp. 8-19.

Simultaneously, the above-listed educational needs may be divided into two categories. The first one includes needs related to the child's home ventilation, whereas the second to the diagnosis of a rare disease.

Educational needs related to mechanical ventilation include, among others:

- more frequent breaks during work, division of tasks into stages and materials into smaller batches that allow for keeping the level of knowledge taught or the course or the train of thought due to greater fatigue;
- application of the demonstrative method, enabling multi-sensory cognition;
- combining the teaching content with the closest environment of the child's life and education, as well as individual experiences that are limited on account of lack of movement;
- the need for functional teaching, based on the child's functioning, his/ her activities, engagement, experiences, discoveries and cognition;
- boosting self-confidence of the pre-schooler during activities in groups and individual tasks;
- extension of work time;
- adjustment of the work place: proper positioning of the child's wheelchair or seat so that he/ she can fully participate in group activities.

Educational needs resulting from the diagnosis of a rare disease include:

- individual needs of a child, resulting from high physical fatigue or current treatment process;
- opportunity to participate in activities outside of the classroom (competitions, professions) adapted to the child's physical limitations;
- taking into account the slower pace of work resulting from specific physical limitations;
- planned breaks during didactic classes resulting from the specifics of a child's treatment and medical-hygiene procedures;

- very close cooperation with specialists during the creation of support programmes;
- supporting active contacts with a child during periods of hospitalisation;
- possibility of interchanging group and individual classes in the periods when the disease worsens;
- possibility of remote teaching during hospitalisation in medical facilities in a foreign country (Kamyk-Wawryszuk 2019).

Study Methods

The study relies on a quality strategy with the use of a case study method, observation techniques, interviews with parents and analysis of the child's medical documentation. The following tools were used during the study:

- sensory sensitivity assessment authored by Bill Nason (Nason 2014);
- author's interview questionnaire with the child's parents pertaining to the level of functioning of the child with home ventilation;
- author's interview questionnaire with the child's parents pertaining to the education of the child in a pre-school age with a rare disease and home ventilation.

The following main research problem was formulated:

- What are the educational needs of a child in a pre-school age with a rare disease and home ventilation?

Along with detailed questions:

- Which educational needs of a child in a pre-school age are related to the diagnosis of a rare disease?
- Which educational needs of a child in a pre-school age are related to home ventilation?
- Which teaching strategies can be applied in pre-school education of a child with a rare disease and home ventilation?

Examined Group

In Poland, there are currently two children diagnosed with SMARD1. The study refers to one of them, due to the fact that the level of functioning of the boys varies and therefore there is no possibility of comparing their educational needs. The differences refer to such areas of development as, for example, speech. The studied boy does not speak and is at the stage when alternative communication is being introduced. On the other hand, the other child uses verbal speech. Therefore, the study procedure encompasses only one child.

Piotr

Pregnancy and Birth

Piotr was born in 2014 with low birth weight (2260 g). Pregnancy progressed correctly and ended with natural birth at full term.

First Disease Symptoms and Diagnosis

At the time of the study, the boy was 6 years' old. After the 7th month of life, he was diagnosed with Spinal Muscular Atrophy with Respiratory Distress Type 1. When he was 2.5 months' old, difficulties with breathing started and the physicians diagnosed symptoms of respiratory distress. He was put on a respirator. Due to this, the boy was hospitalised for 99 days. At the age of 5 months, he underwent a tracheostomy procedure and had a tracheostomy tube inserted permanently (photo No. 1). Three months later, the boy was included in the home ventilation programme. He was initially fed by a probe. At the age of two he underwent the percutaneous endoscopic gastrostomy (PEG). At the age of five, he had an ear drainage procedure.



Photo No. 1 Piotr between 2014 and 2020. Source: private archives.

Psychomotor and Cognitive Development

In his development, Piotr did not accomplish such milestones as sitting, crawling or walking. At the age of three, he was diagnosed with coupled disability of movement and sight. At that time, he could no longer control his body on account of the progressing muscle weakness. However, he is able to move his eyes and make slight movements with the pectoral girdle. Facial mimicry has been preserved. He can sit up with his torso and head propped up. He is not able to change his body position, lift arms or turn his head. At that time, a progressing eye condition was diagnosed. The boy has a limited field of vision which is consequence of the lack of head movement. Limitations resulting from no possibility to move restricted Piotr's capacity to examine the environment and acquire new experiences and skills. The level of cognitive functions was difficult to diagnose at that stage. However, based on conversations with the parents and analysis of opinions issued on the basis of psychological and pedagogical studies conducted at that time, Piotr was not diagnosed with difficulties characteristic for children with intellectual disability. The boy understands instructions and reacts to them with facial expressions. He shows interest in new people. If he wishes to attract attention, he performs an activity that produces sounds or noise, for example he grinds his teeth. With proper tooling that enables positioning, he points to an item or an image that is named and is able to put puzzle pieces together. He recognizes animals, vehicles and toys that he knows and likes on pictures. He makes use of eye-pointing. He gets bored quickly. He chooses persons with whom he wants to work. Given the vision defect (astigmatism, squint), Piotr experiences problems with eyeball coordination.

Sensory Processing

The boy manifests behaviour related to auditory hypersensitivity. He is sensitive to loud and sudden sounds, for example when someone laughs out loud. Noisy activities or classes make him isolate from the environment. It can be noticed that he is pressing his eyelids shut. He does not like songs for children. Sometimes, he ignores sounds from the environment that annoy him. He manifests tactile hypersensitivity in the orofacial sphere. He does not tolerate logopaedic massage or its elements. Piotr does not like to be touched on his face, but he tolerates and likes hygiene activities. He feels uncomfortable when his clothes are too tight or when he is massaged with sensory balls with spikes. He cools down quickly. Piotr has an elevated pain threshold. Within the realm of proprioception, he has a weak muscle tone, which results from the specific nature of the disease. In the realm of sight, he is hypersensitive to sunlight and bright light. He often squints. He is not overwhelmed with significant changes in the appearance of persons or people. He maintains eye contact.

Self-service

The boy does not perform self-service activities. On account of swallowing difficulties, he is fed via percutaneous endoscopic gastrostomy (PEG). He does not signal physiological needs and wears diaper. From time to time, he signals discomfort related to the experiencing of physiological needs. At the age of three, Piotr was diagnosed with

un-integrated bite reflex, excessive salivation resulting from reduced muscle tone in the area of the oral cavity. Before turning one, he tried new flavours, but he frequently spat the food out. He pressed his lips on a spoon. This was the stage when he was fed by a tube (Photo No. 2).



Photo No. 2 Feeding Piotr, 2014. Source: private archives.

Speech and Communication

Piotr babbled after turning one. However, no verbal speech has developed. Lack of independent breathing contributed to the lack of vocalisation. In spite of it, the boy is able to whisper the word “mama”. Nevertheless, mimicry that is used for communication is limited. Piotr cries silently, only his tears are visible. Reduced efficiency of the articulation apparatus is noticeable on account of muscle paresis manifested with reduced tension in the area of the oral cavity. At the age of three, Piotr could slightly press his lips and make them vibrate. No purposeful movements of the tongue, high vaulted palate, malocclusion. Additionally, there was increased mobility of the mandible and a tendency for mesio-occlusion.

In the area of pre-verbal communication, on the level of primary communication (contact with the environment on the level of own body perception), he is able to express impressions of own body in a specific mode. When he is uncomfortable at night and wants to change his body position or when he wants to sit up, he shows it with facial mimicry or makes “prrrr” sounds. Piotr reacts in a specific manner to individual selected stimuli from the environment, for example he follows a person with his eyes. On the level of sensory communication (contact with the environment via gestures and specific types of behaviour, e.g. screaming, hitting objects, laughter), Piotr reacts to voice: he is actively searching for the source of voice by smiling and trying to look in that direction; he initiates and maintains eye contact. When he is excited, for example, after the grandfather’s visit, he moves his shoulders. Within the realm of communication on the level of behaviour, Piotr reacts to his name and to verbal communications from the

environment (e.g. referring to daily, repetitive rituals, for example the story read at bedtime by his dad, watching cartoons in the morning). He initiates contact with other people in a manner enabled by his health condition, e.g. by looking at a specific person. He enjoys company. When he does not want to do something or when a given activity makes him uncomfortable, he shuts his eyes (Photo No. 3).



Photo No. 3 "No" communication Source: private archives.

The first attempts at introducing alternative communication were made in 2015; they relied on introduction of the switch tool and exercising the ability of eye-pointing to items.

Educational Needs of a Child with Rare Disease and Home Ventilation: Results of Own Studies

Based on the analysis of literature devoted to the functioning of a child with SMARD1, the author has prepared a list of educational needs. It includes two groups of needs. The first is related to the diagnosis of a rare disease, the second to the respiratory distress resulting in mechanical home ventilation. Based on study results, it may be concluded that in the first group, four needs were not present in the case of the child in question (Table 2 and 3). This is related to, among others, Piotr's health condition and no approved treatment procedure for SMARD1. At the present moment, clinical trials are under way (Centro Clinico Nemo in Milan), yet in Poland there is no procedure to apply and no principles and guidelines to be followed with respect to the treatment process. This situation resulted in the child's exclusion from hospitalisation in specialist medical centres located, among others, in Italy. Therefore, the needs listed in the catalogue that result from diagnosis of a rare disease are primarily related to the limitations that are a consequence of the weak muscle tone, characteristic for SMARD1, and not experimental treatment.

Table 2. *Piotr's educational needs related to diagnosis of a rare disease*

Educational needs	
related to the rare disease diagnosis	presence of a need
individual needs of a child, resulting from high physical fatigue or current treatment process,	YES
opportunity to participate in activities outside of the classroom (competitions, professions) adapted to the child's physical limitations,	YES
taking into account the slower pace of work resulting from specific physical limitations,	YES
planned breaks during didactic classes resulting from the specifics of a child's treatment and medical-hygiene procedures,	NO
supporting active contacts with a child during periods of hospitalisation,	NO*
very close cooperation with specialists during the creation of support programmes,	YES
possibility of remote teaching during hospitalisation in medical facilities in a foreign country,	NO
possibility of changing the form of classes: interchanging individual and group classes in the periods when the disease worsens.	NO

*Piotr's current health condition does not require extended periods of hospitalisation. Source: author's own study.

Within the scope of educational needs related to the respiratory distress and the necessity of using a respirator, based on the performed study it is possible to indicate that all of the needs listed in the catalogue were present.

Table 3. *Piotr's educational needs related to mechanical home ventilation*

Educational needs	
related to mechanical home ventilation	presence of a need
due to greater fatigue, more frequent breaks during work, division of tasks into stages and materials into smaller batches that allow for keeping the level of knowledge taught or the course or the train of thought,	YES
application of the demonstrative method, enabling multi-sensory cognition,	YES
combining the teaching content with the closest environment of the child's life and education, as well as individual experiences that are limited on account of lack of movement,	YES
the need for functional teaching, based on the child's functioning, his/her activities, engagement, experiences, discoveries and cognition,	YES
boosting self-confidence of the pre-schooler during activities in groups and individual tasks,	YES
extension of work time,	YES
adjustment of the work place: proper positioning of the child's wheelchair or seat so that he/ she can fully participate in group activities.	YES

Source: author's own study.

CONCLUSION

Determination of educational needs allows for personalisation of the teaching strategy. This is indispensable for making sure that both a pre-schooler and a school student master the curriculum and acquire new information and skills. In the modern times, limitations resulting from dependence on medical equipment, such as a respirator, hinder, but do not deprive of the possibility of self-fulfilment in the role of a pre-schooler or a student or, in the future, a qualified employee (e.g. remote work from home with adjusted assisting technology). Resources of persons permanently using medical equipment are still unmanaged, thus it is important to tackle the issues of education of children using such equipment in studies.

Based on the performed study it may be concluded that in the case of a pre-schooler with a rare disease and home ventilation, four out of eight educational needs resulting from a rare disease and all needs related to the respiratory distress and home ventilation were present. Thus, the catalogue of educational needs will comprise the following needs:

- individual needs of a child, resulting from high physical fatigue or current treatment process,
- opportunity to participate in activities outside of the classroom (competitions, professions) adapted to the child's physical limitations,
- taking into account the slower pace of work resulting from specific physical limitations,
- very close cooperation with specialists during the creation of support programs,
- more frequent breaks during work, division of tasks into stages and materials into smaller batches that allow for keeping the level of knowledge taught or the course or the train of thought,
- application of the demonstrative method, enabling multi-sensory cognition,
- combining the teaching content with the closest environment of the child's life and education, as well as individual experiences that are limited on account of lack of movement,
- necessity of functional teaching, based on the child's functioning, his/ her activities, engagement, experiences, discoveries and cognition,
- boosting self-confidence of the pre-schooler during activities in groups and individual tasks,
- extension of work time,
- adjustment of the work place: proper positioning of the child's wheelchair or seat so that he/ she can fully participate in group activities.

This shows that whilst designing education for such child, it is necessary to take into account needs related to the use of medical equipment, as well as to select such an educational path that incorporates the possibilities of professional functioning based on remote work and assisting technology. Technological progress, both in medicine and in tooling for people with motor disabilities is going to change in the next 20 years, allowing these students and in the future adult people to have greater participation in both social and professional life.

The conducted study allows for one more conclusion. It is not directly related to the study objective, but equally important. It is necessary to set up a research network that is going to tackle the issues of pre-school and school education in its projects, including support and adjustment of teaching strategies for persons dependant on medical equipment, e.g. children with mechanical ventilation and forging their potential into self-fulfilment/ independence both in the professional and social aspect.

REFERENCES

1. Brookes, I. (2019). Long-term ventilation in children. *Paediatrics and Child Health*, 29(4), 167-171.
2. Eckart, M., Guenther, U.P., Idkowiak, J., Varon, R., Grolle, B., Boffi, P., & von Au, K. (2012). The natural course of infantile spinal muscular atrophy with respiratory distress type 1 (SMARD1). *Pediatrics*, 129(1), 148-56.
3. Gernsbacher, M. A., Raimond, A. R., Balinghasay, M. T., Boston J. S. (2016). "Special needs" is an ineffective euphemism. *Cognitive Research: Principles and Implications*, 1(1), 1-13.
4. Griffin, T. (2014). *Children with Special Educational Needs. Information Booklet for Parents*, National Council for Special Education.
5. Grohmann, K., Schuelke, M., Diers, A., Hoffmann, K., Lucke, B., Adams, C., & Hübner, C. (2001). Mutations in the gene encoding immunoglobulin mu-binding protein 2 cause spinal muscular atrophy with respiratory distress type 1. *Nature Genetics*, 29(1), 29 - 75.
6. Grohmann, K., Varon, R., Stolz, P., et al. (2003). Infantile Spinal Muscular Atrophy with Respiratory Distress Type 1 (SMARD1). *American Neurological Association*, 54(6), 719-724.
7. Hewitt-Taylor, J. (2004). Children who require long-term ventilation: staff education and training. *Intensive and Critical Care Nursing*, 20, 93—102.
8. Jędrzejowska, M. (2010). Przeponowa postać rdzeniowego zaniku mięśni (SMARD1). *Neurologia Dziecięca*, 19(38), 51-54.
9. Kaind, A. M., Guenther, U. P., Rudnik-Schöneborn, S., et al. (2008). Spinal Muscular Atrophy With Respiratory Distress Type 1 (SMARD1). *Journal of Child Neurology*, 23(2), 199-204.
10. Kamyk-Wawryszuk, A. (2019). Educational needs of preschool children with recognized rare disease and intellectual disability – teachers` perspective. *Journal Plus Education*, Vol. XXIV, Special Issue, 8-19.
11. Lawrence, S. (2011). Promoting independence for children on long-term ventilation. *Nursing Children and Young People*, 23(10), 20-22.
12. Ludvigsen, A., & Morrison, J. (2003). *Breathing Space: Community Support of Children on Long Term Ventilation – Summary*. Barnardo's. Giving Children Back Their Future.
13. Malbogat, L. (2014). *Special educational needs*. Cambridge Assessment International Education, IGCSE.
14. Nason, B. (2014). *The Autism Discussion Page on the Core Challenge of Autism*. London: Jessica Kingsley Publishers Ltd.
15. Porro, F., Rinchetti, P., Magri, F., Riboldi, G., Nizzardo, M., Simone, Ch., ... & Corti, S. (2014). The wide spectrum of clinical phenotypes of spinal muscular atrophy with respiratory distress type 1: A systematic review. *Journal of the Neurological Sciences*, 346, 35–42.
16. Richter, T., Nestler-Parr, S., Babela, R., Khan, Z.M., Tesoro, T., Molsen, E., Hughes, D.A. (2015). Rare Disease Terminology and Definitions—A Systematic Global Review: Report of the ISPOR Rare Disease Special Interest Group. *Value in Health*, 18(6), 906-914.