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SOCIAL FUNCTIONING OF A STUDENT WITH PRADER-WILLI SYNDROME IN EDUCATIONAL SYSTEM

INTRODUCTORY COMMENTS

Prader-Willi syndrome (PWS) is a very rare genetic disorder. It is estimated that for about 20,000 newborns only 1 is affected by this disease (for comparison, at the same time, approximately 623 children with Down's syndrome will be born). The disorder owes its name to the Swiss endocrinologists Andrei Prader and Heinrich Willi, who in 1956, together with Alexis Labhart, described the syndrome for the first time. At that time, its cause was unknown, however, the dominant symptoms were so characteristic and unique that Prader and Willi were able to distinguish a new disease entity, initially called the H3O syndrome (*hypotonia* = reduced muscle tone, *hypomentia* = intellectual disorder, *hypogonadism* = malformation of the genital organs, *obesitas* = obesity)[1].

CAUSES OF PWS

The genetic origin of this disorder is complex and concerns a unique phenomenon in human biology, which is parental genomic stigma (all body cells have the same set of genes, but not all genes are equally active). Prader-Willi syndrome arises as a result of major changes in the genome arrangement found in chromosome 15. This means that if genes

[1] See: Leaflet of the Polish Association of Prader-Willi Syndrome, M. Libura, *Moje dziecko ma Zespół Pradera-Williego. Jak mogę mu pomóc?*, Polish Association of Prader-Willi Syndrome, Warsaw 2007, p. 12; T. Kaczan, R. Śmigiel, M. Michalska-Idaszek, *Wspomaganie rozwoju dzieci z zespołem Pradera-Williego*, [in:] T. Kaczan, R. Śmigiel (ed.), Wczesna interwencja i wspomaganie rozwoju u dzieci z chorobami genetycznymi, Publishing House „Impuls", Cracow 2012, p. 63.

from a particular fragment of chromosome 15 originating from one of the parents are altered, then the backup of the same genes from the second parent will not replace their function. There are three types of genetic errors that determine PWS. The most common (75% of cases) is the so-called *deletion* of the chromosome 15 originating from the father. It means that there is a lack of genes from the critical region, because this fragment of the chromosome has been lost and the maternal counterparts of these genes are not active and cannot compensate for the resulting deficiency. The second most frequent occurrence (20% of cases), *is maternal uniparental disomy*. This means that due to the incorrect division of the cell the child receives two copies of the chromosome 15 from mother, while the paternal copy is lost. Due to the aforementioned genomic stigma, genes on the maternal copy are not active and do not work. The third mechanism responsible for the creation of PWS is called the *mutation of the imprinting center* – IC. The imprinting center is a fragment of the chromosome that directs the operation of the region critical for PWS. As a result of the IC mutations, the genes, although physically present, do not show activity on which the proper development depends. In most people with Prader-Willi syndrome, genetic changes are *de novo* (the defect is accidental and sporadic) and for the vast majority of cases they are not inherited. In rare events (mutation in the IC) the risk of family re-syndrome can reach up to 50%[2].

PHENOTYPIC TRAITS AND SYMPTOMS IN PEOPLE WITH PRADER-WILLI SYNDROME

The *craniofacial dysmorphic* characteristic is already visible in infants with PWS. It becomes more pronounced with the child's age and is characterized by: a disproportionately long face and skull, a narrow forehead, almond-shaped eyes, narrow palpebral fissures, downturned corners of the mouth, a narrow upper lip, thick, sticky saliva drying at the corners of the

[2] Cf. M. Libura, *Moje dziecko ma Zespół Pradera-Williego. Jak mogę mu pomóc?*, Polish Association of Prader-Willi Syndrome, Warsaw 2007, p. 14; T. Kaczan, R. Śmigiel, M. Michalska-Idaszek, *Wspomaganie rozwoju dzieci z zespołem Pradera-Williego*, [in:] T. Kaczan, R. Śmigiel (ed.), Wczesna interwencja i wspomaganie rozwoju u dzieci z chorobami genetycznymi, Publishing House „Impuls", Cracow 2012, p. 63–64.

mouth. In patients with PWS, there is also hypopigmentation of the skin and hair, manifested in the light complexion of the skin and light hair. In the external appearance small hands and feet in relation to the rest of the body can be also observed[3].

On the other hand, the symptoms of PWS in various age ranges (fetus, newborn, child up to 3 years, a child over 3 years old, teenager, adult) are different. Newborns and infants with PWS display *muscular hypotonia*, which manifests itself during fetal life with poor fetal movements, its abnormal position during delivery and lack of labour progression. The child is born limp, motionless and with reduced muscle tension which leads to delay in psychomotor development. The child is not able to hold the head, sit down, crawl nor walk. In children with PWS there is weak sucking reflex, poor mimic expression, and they have characteristic wailing, silent crying. Due to the dysfunction of the hypothalamus, the child's body, from birth has also *problems with thermoregulation*. Another characteristic feature of PWS is *the growth hormone deficiency*, and as a consequence *short stature and lack of muscle mass.* Although after birth the body length of children with PWS usually falls within the normal range, later on they develop more slowly than peers and without applying growth hormone usually remain very short. The human growth hormone also plays a very important role in muscle formation. Muscle deficiency can be observed in infants with PWS. This is another factor that hinders normal development of children right after hypotension. They have skimpy muscle tissue, they have difficulty in moving and are not very active, consequently they do not have much opportunity to explore the surroundings, and as a result they learn and develop more slowly than their peers. It is not only for this reason that *delay in intellectual development and speech delay* is visible in PWS patients. Children with PWS exhibit learning difficulties. This applies in particular to areas involving abstract thinking. Most often they read well but do worse in writing. They have big problems remembering current information, which requires frequent repetitions. As mentioned above, speech development is also disturbed. Children with PWS understand much more than they can say. Articulation problems coexist with linguistic system disorders. There are many factors involved in impaired speech development in PWS,

[3] Ibid.

including muscle weakness of the entire speech apparatus, incorrect oral cavity structure (gothic palate), sequential processing disorders and the low level of cognitive development. The next symptom that affects all people with PWS is *hypogonadism*, or hormonal malfunction of the genital organs. It is visible from the moment of birth, and is manifested in underdeveloped genitals and cryptorchidism in boys, and small labia and clitoris in girls. Sexual hormone deficiency affects the course of puberty, which in PWS patients is incomplete: boys usually do not undergo mutation, there is no facial hair, girls do not have menstruation, or they are irregular. Thus, the vast majority of people with PWS are infertile. Also typical of the Prader-Willi syndrome is the occurrence of osteoporosis, scoliosis and / or kyphosis and the lack of gag reflex. However, the feature that distinguishes the Prader-Willi syndrome from other disorders is abnormally increased appetite – *hyperphagia*. This symptom appears between 2 and 4 years of age, and is rising gradually. Lack of feeling of satiety (uncontrollable, unrestrained hunger), which is the result of congenital disturbances in the functioning of mechanisms responsible for maintaining the energy balance and reduced metabolism, can become a cause of significant obesity in a short time. Considerable obesity and subsequent complications such as diabetes, heart failure, circulatory system diseases, breathing disorders including sleep apnea, postural defects, degeneration of the skeletal system and joints are widely regarded as the most serious clinical problems in the Prader-Willi syndrome. These disorders not only hamper the functioning of the patient and deepen his disability, but also constitute the most common cause of premature deaths of people affected by this syndrome[4].

BEHAVIOURAL ASPECT OF PRADER-WILLI SYNDROME

In the hypothalamus, which functioning in people with PWS is disturbed, there are many centres responsible for individual features of the organism that have more or less indirect impact on human behaviour. Here, in addition to the satiety centre, there are also areas responsible

[4] Cf. M. Libura, *Moje dziecko ma...,* op. cit., p. 22–28; T. Kaczan, R. Śmigiel, M. Michalska-Idaszek, *Wspomaganie rozwoju dzieci...*, op. cit., p. 64–66.

for regulating the day-night cycle, pain sensation and thermoregulation. Moreover, hypothalamus is also responsible for the control of emotions and memory. Therefore, patients with PWS usually have problems in these areas.[5]

Behaviour problems become most evident during adolescence, because child with PWS after birth is rather apathetic, sleepy and little interested in the outside world. At the age of 1–2 years, child becomes cheerful, kind, friendly and obedient. Easily and willingly makes contact with the surrounding. Nevertheless, young children already show attachment to controlled situations and have difficulties in interrupting the activity. In the 2 to 3 years of life, along with the growing hyperphagia, behaviours related to obsessive search for food appear. The child inquires about the time of meals and their type, which also translates into other forms of compulsive behaviour. Problems arise especially when there is an unexpected change in earlier plans or a typical order of daily activities and routines. With age, unwanted behaviours intensify and older children are characterized by growing stubbornness and emotional lability with characteristic anger attacks. People with PWS are described as stubborn, impulsive, irritable, susceptible to fluctuating changes in mood, self-cantered, demanding towards others, prone to tantrums in moments of frustration. They try to manipulate others and tend to be less flexible with age. The PWS children have also a tendency to depression, anxiety and sporadically they can fall into psychosis. Aggression and auto-aggression are also frequent problems in their conduct. About 1/3 of people with PWS tend to scratch the wounds and pick the skin. This is especially onerous and dangerous behaviour as they show less sensitivity to pain, therefore picking the skin can take the form of obsession and can lead to the formation of difficult to heal wounds and numerous scars.[6]

On the basis of the theoretical considerations presented in the first part of the paper, the methodology of my own research will be presented below, along with a description of social functioning of the student with Prader-Willi syndrome in the education system.

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[5] T. Kaczan, R. Śmigiel, M. Michalska-Idaszek, *Wspomaganie rozwoju dzieci...*, op.cit., p. 75

[6] Ibid., p. 75–76, M. Libura, *Moje dziecko ma Zespół...*, op. cit., p. 28–32.

METHODOLOGICAL ASSUMPTIONS OF THE RESEARCH

Aim: to learn about the social functioning of the student with Prader-Willi syndrome in education system.

Subject of research: social functioning of a student with Prader-Willi syndrome in education system.

Research problems:

- main:
  - How does a student with Prader-Willi syndrome function in the education system?
- specific:
  - What was the didactic success of the student with Prader-Willi syndrome at particular educational stages?
  - What was the relation of the student with Prader-Willi syndrome with other students?
  - What was the relation of the student with Prader-Willi syndrome with teachers?
  - What abnormal behaviours resulting from PWS manifested the student with Prader-Willi syndrome in the education environment?

Table 1. Conceptual framework of research problems

Conceptual framework of research problems	
didactic successes	School certificates confirming completion: 1st educational stage (3rd year of Primary School): descriptive assessment. 2nd educational stage (6th year of Primary School): descriptive assessment. Junior high school (3rd year): descriptive assessment. Apprenticeship school: descriptive assessment.
relations with other students	Positive: working in a group, spending time together during breaks, talking, making friends, sharing interests, respecting basic social norms in relationships with other students. Negative: inability to work in a group, lack of willingness to make contact with other students, lack of common topics for conversations, lack of common interests, disrespect of basic norms in relations with other students.
relations with teachers	Positive: fulfilling teacher's orders, keeping a proper distance, correctly fulfilling the role of the student, Negative: failure to follow the teacher's instructions, lack of a proper distance to the teacher, incorrect fulfilment of the student's role.

cd. Table 1.

Conceptual framework of research problems	
abnormal behaviour associated with Prader-Willi syndrome	- hyperphagia – taking food and drink from other people, obsessive food-seeking behaviour, asking for the time and composition of meals, – temper tantrums, – emotional lability, – stubbornness, – quarrelsomeness, – obsessive-compulsive behaviour, – self-injury – skin picking, – high tolerance to pain, – lack of mental flexibility and reluctant acceptance of changes, – difficulties with interrupting initiated activity, – aggression.

Source: own study.

Method: case study.

Technique: interview, document analysis.

Tool: an interview questionnaire with teacher and student's parent, and a document analysis sheet.

The research was carried out and developed in accordance with the procedure used in the quality strategy[7].

SOCIAL FUNCTIONING OF THE STUDENT WITH PRADER-WILLI SYNDROME – CASE STUDY – MARCIN K.

Due to the size of the study, the presentation of the collected empirical data in the concept-ordered matrix was abandoned and the case description and analysis were immediately made.

DESCRIPTION AND CASE STUDY

At Marcin's, the Prader-Willi syndrome was diagnosed when he was about one year old. His mother was troubled by her son's problems with sucking and his extreme flexibility: *"immediately after birth, my son's muscles were floppy, my breastfeeding was difficult because he did not have sucking reflex. I contacted the doctor and he paid attention to his appearance. That's how*

[7] Cf. H. Rudomska, *Analiza jakościowa zachowań prawidłowych i nieprawidłowych uczniów głuchoniewidomych na tle edukacji rodzinnej i instytucjonalnej – rozwiązania metodologiczne*, [in:] M. Chomczyńska-Rubacha (ed.), Educational Research Review no. 18 (1/2014), The Nicolaus Copernicus University Scientific Publishing House, Toruń 2014, p. 171–194.

we got to the genetic counselling centre." The mother was informed about the course of the disease, the child's rehabilitation was also started: *"the geneticist doctor talked to me about the disorder, gave the referral for Vojta rehabilitation, then we went to the sanatorium".*

Until the second year of life, the boy had problems with physical development: *"his muscles were very floppy, he could not sit down or roll over for a long time"*. From the second year of his life, he started to attend specialized nursery where he made progress in gross motor skills: *"Rehabilitation process and the stay in the nursery got strengthen his muscles. He started to roll over and sit down, and he slowly began to stand up to walk by the hand."* From the records kept in the nursery, it appears that in a 5-year-old boy; *"Systematic progress in development is observed. Marcin moves independently, but not very efficiently. Despite the physical progress, intellectual functioning is still reduced. He understands speech, he makes verbal contact with the environment very easily. He uses a pencil quite well, though the legibility of drawing is rather poor. The level of play activities is average. Coordination is getting better. A good emotional contact with researcher, cognitive contact somewhat difficult (distracts attention, repeats stereotypical questions). The child differentiates basic shapes, recognizes objects of everyday life on pictures. Marcin has rather limited range of vocabulary and multiple articulation disorders.. The child requires to stay in the group with peers to better adapt to life (periodically shows aggression in behaviour)."*

At the age of 6 Marcin went to non-public kindergarten where he had various adaptation problems. It was mainly due to the fact that he was qualified for a large group of children, with only one educator. During the course he had difficulty concentrating, he exhibited aggressive behaviours directed at his peers*: "he did not cope in the nursery school, it was probably because of noise and high temperature, he could not concentrate, he supposedly kicked a girl".* Lack of understanding by teaching staff of the PWS children's specific functioning and lack of willingness to support and help the child caused that Marcin was removed from the preschool list: *"after 2 weeks my son was expelled from the kindergarten, I do not even remember on what pretext, but it was something trivial."* Next, the boy began education in special care nursery school. According to the Psychological and Pedagogical Counselling due to*: "delayed mental development with behavioural disorders",* he was qualified for individual teaching with the request for:

"*enabling participation in social activities organized in the kindergarten, parties, trips, etc.*" There, in cooperation with his mother, the consistent implementation of the rules of dealing with the child began, especially in case of difficult behaviours: *"in the second kindergarten he also happened to behave badly, he teased children who were sitting close to him, sometimes he could strike a child. He also resisted teachers' orders, and when they did not give way, he screamed terribly. We agreed with the teachers that we would reward good behaviour and get small pieces of jelly or raisin for a certain stage of the material being processed. Besides, we praised him loudly, we applauded him, he got a kiss from me".* Consistent treatment of Marcin, above all proceeded by his mother but also of all persons taking care of him in the nursery school, resulted in longer periods of concentration and accomplishment of tasks. In spite of this, the Psychological and Pedagogical Counselling's research continued: *"increased fatigability, difficulty in controlling affects, functioning at the level of a light degree of mental retardation, reduced range of knowledge and skills necessary to take up the school duty."* Hence, his school obligation was postponed twice and the recommendation was to repeat the class "0".

At the age of 9, in accordance with the opinion of Psychological and Pedagogical Counselling about special education needs, the boy started: *"special education for mentally retarded children"*. Unfortunately, up to the age of 12, Marcin was unable to meet the role of the student. His mother mentions this period as very difficult for both of them: *"he could not sit on the lesson for too long, he was provoking others, he was teasing, he was destroying his glasses, he was leaving the classroom. Under unfavourable conditions, he was beating the one who stood closest, kicking, spitting, he scratched his skin and scabs. I always had to be at school to calm him down. I led him to the toilet to let him scream and cry, and after all, he did not want to go back to the classroom."* Such situations indicate the incompetence of teachers who ceded responsibility for Marcin's behaviour only to his mother. They did not take the effort to continue the principles developed at kindergarten and did not cooperate with the parent to prevent and to eliminate the effects of problematic behaviours of the child, which did not result from parental education mistakes and were caused by PWS. Subsequently behaviour problems have affected school performance. The certificate ending the third grade of primary school stated*: "Marcin is a social and obliging boy. He always comes to school carefully prepared.*

He works slowly, uses the help and suggestions of the teacher. He tries to establish positive relationships with peers, but is not always able to control his emotions. During classes, he often deals with matters not related to the topic of the lesson, refuses to do the task. The student eagerly participates in conversations and talks about his experiences and reflections. He does not always listen carefully to other people's statements. He reads syllables and words well. While rewriting, he occasionally makes spelling mistakes. He has trouble writing words from memory and from hearing. He can add and subtract on the specific examples within 20. He can recognize coins and banknotes. He can read the full hours on the clock, he correctly lists days of the week, little worse names of months. The student solves simple text problems with the help of a teacher. He knows the rules of getting around the roads. He takes care of personal hygiene. He enjoys playing percussion instruments and is able to use simple tools."

The school situation improved, when Marcin, at the age of 12, came under the care of a new teacher-therapist. Then the appropriate procedure for reacting to abnormal behaviours, based on the assumptions of behavioural therapy, was implemented: *"he still used to shout, hit, bite or kick someone, nevertheless the new therapist was able to deal with these behaviours as he knew that they did not result from Marcin's bad will. I did not have to be at school anymore which was a great relief for me. We introduced special tables for school and home conduct. Marcin collected pluses and minuses for his behaviour. For the right amount of pluses collected during the week, he received a significant prize, e.g. horse riding or other interesting activity. The prize was cancelled for the minus points and he could not, for example, sit next to the driver on his way to and from school. The rules were clear to my son, he knew exactly what he was working on, how many pluses he must have to get a reward. This system made his behaviour better."* At the end of primary school the following descriptive assessment was made: *"Marcin's behaviour has significantly improved. The student knows and applies adequate politeness phrases. He is always well prepared for classes. He honestly fulfils the tasks entrusted to him. Marcin understands the information and follows the instructions very well. He has a rich vocabulary. He readily and correctly rewrites the text, and also writes by ear. The student copes well with adding and subtracting in writing. Multiplication and division requires further exercises. He knows the rules of using a calculator. He can solve tasks with content with help of a teacher,*

can substitute data for a formula and calculate fields of basic figures. He identifies units of measurement, time, weight, length and gravity. He knows the value of money, independently does the shopping. Music classes make Marcin happy, he can remember the content and melody of songs, readily sings, has a sense of rhythm, can determine the tempo of the song. Besides he likes dancing and playing instruments. During the art classes, Marcin always finishes his work, likes to draw and paint, properly arranges the elements on a piece of paper. The works are aesthetic and precise. Marcin's efficiency in technical operations requires improvement, however the student deals with manual and construction activities quite well. He enjoys sticking and cutting. The student is able to operate basic audiovisual devices, he enjoys working on a computer. Marcin shows moderate interest in physical exercises. He is able to independently overcome short distances by running, he eagerly swims and participates in recreational and motor activities."

At the age of 15, Marcin began junior high school for special needs education. The teacher changed again and he did not continue the worked out procedures for dealing with Marcin. The new lecturer was not able to establish a positive relationship with the student. The attitude of the teacher caused Marcin's reluctance to school and escalation of incorrect behaviours. The boy's mother again had to deal with problematic situations with her son alone, without support and help from the school environment: *"I had to be in school again because he began to disregard the teacher's instructions, he left the classroom and slammed the door when the problem was overgrowing, he started shouting, waving arms and legs. During the attack, apart from shouting, he called names, spited and even pissed himself. After calming down, he had to clean up, sort out what he had dumped, and at home he had to wash his clothes. Relatively quickly, he stopped polluting himself, because he stated that washing is quite boring. I asked to reintroduce the notebook with pluses and minuses. I was consistent. It was the table with the pluses and minuses that gave him control over the behaviour. The transparency of this form is perfect. Exceeding 3 minuses per week cancelled the possibility of getting a prize."* Years spent in junior high school were not a time for raising the level of school achievements for Marcin. In the leaving certificate there was no new skill mentioned in relation to the primary school.

After graduating from junior high school, Marcin moved to an education centre and continued learning at apprenticeship school. There, once

again, the mother introduced the specificity of the PWS person to the teachers dealing with Marcin, rose awareness to her son's behaviour and pointed out methods of preventing and mitigating difficult situations. Unfortunately not all pedagogues met the requirements of working with a student with special educational needs. A particular escalation of adverse behaviours was caused by a teacher who was in conflict with Marcin, trying to prove his superiority. Finally, at mother's request, during the school year the teacher was changed*: "I started working with Marcin in the middle of the second semester of the school year. I often witnessed Marcin standing at the door of the class and refusing to go inside, shouting, crying, calling names at the teacher. He was accompanied by his mother who was making an effort to ease the situation, unfortunately the teacher did not show any willingness to establish a proper relationship with Marcin. It happened that I approached the boy and tried, calmly or jokingly distract him. Therefore, as I was offered to work with Marcin, I willingly agreed. "* Due to previous conflicts between Marcin and the teacher, his mother was also present in a new school during school hours or somewhere near, to react immediately to her son's aggressive conduct. The situation improved when the teacher changed. The occurrence of Marcin's difficult comportment significantly reduced their frequency and strength. The leading teacher tried to comprehend the specificity of the student's functioning and understood that his behaviour is resulting from the disorder: *"Marcin was generally very cheerful and eager to work, but he required teacher's involvement. The prepared work cards had to be very attractive and varied for him. Besides he was given pluses for his behaviour during classes and on breaks, additionally there was always a crossword at the end of the class. From the beginning of the day, he inquired whether there would be a crossword, although he knew it was always there. At each lesson he was also asking what I would have for dinner. He liked talking about food."* During the lesson there were problematic behaviours, but the teacher, being attentive to the student's needs, was able to recognize them and ease the situation before the outbreak of uncontrolled anger and aggression*:" There were such days, especially related to the change of weather, I knew from Marcin's entering the classroom that it will be a hard day in school. He walked in such a characteristic way, he had changed facial expression, it was clear that he was irritated. In those situations, I asked what he would like to do, I suggested, for example, a walk or classes in the World Experience*

Hall. I think that he was angry at the same time with himself but he always appreciated my suggestions. At that time he could choose the form of activity he liked and it never happened that the irritation turned into an uncontrolled anger. During those difficult days, he was also picking the skin uncontrollably, sometimes I managed to divert his attention from it and took something else, but occasionally I had to cover the wounds with plaster, so that, as Marcin said, he was not tempted. I talked to him a lot at that time, and he easily made contact with people he liked." The teacher also experienced unusual situations requiring unconventional attitude*:"We were never bored, I loved taking with him because I never knew what will happen. Once he approached me and changed his voice and talked as some celebrity, actor or film hero. Sometimes I had to guess who he is. When I succeeded, he was very pleased then. I never discussed with him that after all he is not a famous actor or a hero of the film, on the contrary, I had a double dialogue with Marcin – telling him that during the lesson we would have a visitor known from television and I invited him to classes with my best, unique student. Sometimes, Marcin played a double role all day but it did not bother me as he did all the tasks with enthusiasm and flawlessly, because "the visitor" looks at him."* It can be said that in the relationship with the teacher Marcin was able to read moods, he was empathetic and had a sense of humour adequate to the situation*: "Once I felt really bad and I was not joking with Marcin as always. He looked at me and asked: 'Do you know how did I come to school today?' I answered: 'Maybe by tram?', he replied that it's trivial, 'Maybe by car?'I continue, 'Everyone can come by car', he responded. I said that I give up, because I have no idea how he could come, and he stated: 'Didn't you see a horse at the porter's lodge?; because that's how I came'... He made me smile and I started laughing. He asked me if I was feeling better because he just wanted to make me laugh. It was really amazing. Then he said that I always save him and understand how bad he might feel, now he took care of me."* Such good relations between Marcin and his teacher influenced the correct course of the lessons, and thus the school achievements. Unfortunately, these accomplishments were also determined by his psychophysical state: *"Marcin generally did not have problems with learning. I prepared for him working cards at the level of grades IV – VI with mathematical, science and humanistic content. Generally, he had no problems with filling them. His mother mentioned that he made the most progress in class IV – VI, when*

he had good contact with the teacher and in fact I only revised that material. Unfortunately, due to intellectual disability, it was difficult to expand this knowledge and skills. There were days when the simplest mathematical activities caused him problems. Sometimes he wondered how much is 4 plus 4, he counted on his fingers and still gave wrong answer... Then, so that he could succeed and gain pluses, I had prepared tasks from class I – III of elementary school and of course an easier crossword puzzle. However, regardless of the psychophysical state, as he started to work on the task, he always had to bring it to the end, even if the break started or ended." Very difficult situations appeared when there was a change in the order of classes or the teacher was absent: *"when I had a day off or I was on exams, the other teacher took over Marcin's class. Marcin did not like it, he shouted that I should be with him in the class. It happened once that I was gone for a week, then he made a terrible row: he cried, he did not want to enter the class, he shouted that if I did not show up immediately, he would never come back to this school. He used vulgarisms, three people had to keep him so that he would not hurt himself or others. From that time on, when I was not present at school Marcin was staying at home."* The school events were also problematic, especially those with food, such as Christmas Eve or Children's Day*: "In situations when food appeared at school celebrations, in which Marcin participated, there always had to be present his mother. She was telling him that he could try every dish, but just a little, and all the cakes were divided in half. As Marcin knew the rules, he only sometimes happened to be slightly irritated telling that he would like to eat more, but it was never a reason to cause attack of anger or aggression."* It turns out that the attitude of his mother, the person who loves her son, was very important. She was exceptionally consistent with him and very kind and supportive towards the actions of Marcin's teachers: *"definitely we should emphasize here the merits of Marcin's mother. She is an extraordinary woman. Marcin is a slim boy thanks to her. People with PWS of his age often weigh over 120 kilos. She skilfully prepares food for him, so that he can eat at every break, but this is, for example, a quarter of an apple cut into thin slices which gives the impression that there is a lot of food. I am also impressed by her consistency in raising her son. She strictly adheres to established rules and applies prizes and penalties in accordance with them. As a result, currently recorded abnormal behaviours are sporadic and moderate in severity. Marcin knows that he must control himself, be-*

cause his mother cannot be manipulated. Thanks to his mother, he is also always prepared for classes, this is she who developed in him hygienic and orderly habits. Really, how well Marcin works is her great merit. "

At the age of 24 Marcin finished the apprenticeship school. He currently stays at home with his mother who is trying to assign specialist care services to her son.

CONCLUSION

1. The didactic achievements of the PWS student at individual educational stages were evaluated in a descriptive way. The assessments show that the student fulfilled the program contained in the individual didactic curriculum, adapted to his psychophysical abilities. His school progress at each stage was mainly determined by the attitude of the teachers.
2. The student with PWS did not establish closer relations with peers in the educational environment. In kindergarten, he showed unjustified aggression towards other children manifested in their beating, kicking, spitting. At school, despite individual teaching, he also physically abuse other children. In the educational centre, where he continued individual teaching, he also did not establish positive relations with other students, but he did not show any aggressive behaviours towards them.
3. The relations between teachers and the student affected by Prader-Willi syndrome were strongly dependent on the attitude of the former. The student preferred educators who were consistent, but accepting him and understanding his needs. During the lessons with the teacher he liked, as a rule, he behaved properly. He was eager to learn new material, follow teacher's instructions, and use courtesy phrases. He also talked about personal topics. Possible problems resulted, for example, from changes in weather or changes in the set rhythm of the day, however it was possible to alleviate the anger and frustration quickly. In turn, in the presence of teachers he did not sympathize with, the student behaved in a disrespectful and offensive manner. He often argued with them, he used vulgar vocabulary, aggression or he mutilated himself. It was difficult to calm him down.
4. In the initial stages of education a number of incorrect behaviours resulting from PWS were registered in a child. They included: asking for the time and composition of meals, uncontrollable tantrums, stub-

bornness, quarrelsomeness, self-harming – skin picking, lack of mental flexibility, reluctant acceptance of changes, difficulties with interrupting the initiated activity, various forms of aggression. Together with age, these behaviours lost intensity and frequency. This was caused mainly by the introduction of elements of behavioural therapy and rewarding normal conduct. It was not without significance here, the close coordination with the student's mother as well as consistent respecting the principles adopted earlier.

CONCLUSIONS

The presence of Prader-Willi syndrome carries a number of health and behavioural consequences. Quick diagnosis of a child allows an early rehabilitation and introduction of rules for dealing with the patient. The cooperation and working out a common front of interactions, between people taking care of a child with PWS, e.g. parents, physiotherapists, teachers, can significantly limit the negative effects of the disorder and improve the quality of life of the person affected.

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SUMMARY

The study analyzes the educational path of a student with Prader-Willi syndrome from the beginning of his education in the kindergarten, up to the completion

of vocational school. Particular emphasis was placed on the description of student's didactic achievements, building relationships with schoolmates and teachers as well as on the abnormal behaviours in the school environment as a consequence of this genetic disorder.

Key words: Prader-Willi syndrome, educational system, relations with students, relations with teachers, undesirable behaviours.