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# Interdisciplinary care and support of child development with Williams syndrome

## Interdyscyplinarna opieka i wspomaganie rozwoju dziecka z zespołem Williamsa

**Streszczenie**

**Cel:** Zespół Williamsa to rzadkie, wieloukładowe, genetycznie uwarunkowane zaburzenie, występujące jednakowo u dziewczynek, jak i u chłopców na całym świecie, niezależnie od grupy etnicznej. Celem pracy jest przedstawienie znaczenia interdyscyplinarnej opieki i wspomagania rozwoju dziecka z zespołem Williamsa.

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**Metody:** Interdyscyplinarne podejście do opieki i wspomagania rozwoju dziecka z zespołem Williamsa.

**Wyniki:** Zespół Williamsa objawia się charakterystycznymi nieprawidłowościami fizycznymi (dysmorficzne cechy twarzy), medycznymi (schorzenia sercowo-naczyniowe), poznawczymi (umiarkowane upośledzenie umysłowe) oraz społeczno-emocjonalnymi. Dziecko powinno być pod opieką lekarzy różnych specjalności i innych specjalistów: logopedy, psychologa czy fizjoterapeuty.

**Wnioski:** Terapia musi obejmować usprawnianie motoryki, funkcji sensorycznych, poznawczych, ćwiczenia z zakresu komunikacji interpersonalnej i zachowań społecznych. Dobra współpraca między specjalistami jest niezbędna w prawidłowym zaplanowaniu i prowadzeniu terapii.

**Słowa kluczowe:** zespół Williamsa, opieka nad dzieckiem, interdyscyplinarna opieka, wspomaganie rozwoju.

**Abstract**

**Aim:** Williams syndrome is a rare, multi-system, genetically conditioned disorder, occurring equally at girls and boys around the world regardless of the ethnic group.

The aim of the work is the presentation of the meaning of interdisciplinary care and support in the development of a child with the Williams syndrome.

**Methods:** Interdisciplinary approach to care and support of development of a child with the Williams syndrome

**Results:** Williams syndrome manifests itself in characteristic physical (dysmorphic facial features), medical (cardiovascular), cognitive (moderate mental retardation) and social-emotional disorders. The child should be under the care of different types of doctors and medical specialists, a speech therapist, psychologist and physiotherapist.

**Conclusions:** Therapy must include improving motor activity, sensory and cognitive functions, exercises in interpersonal communication and social behavior. Good cooperation between specialists is essential in the proper planning and conducting of therapy.

**Keywords:** Williams syndrome, childcare, interdisciplinary care, suport of child development.

Williams syndrome (Williams-Beuren syndrome, hereinafter: WS) is a rare, multi-system, genetically conditioned disorder, affecting 1 in 7,500–20,000 children born alive. It occurs with the same frequency in boys and girls globally regardless of their ethnic group[1]. The syndrome is not inherited from parents.

[1] M. Buchnat, K. Pawelczak, *Nieznane? Poznane, Zaburzenia rozwojowe u dzieci z rzadkimi zespołami genetycznymi i wadami wrodzonymi*, Wydawnictwo Naukowe Uniwersytetu Adama Mickiewicza, Poznań 2011; J. Wierzba (ed.), *Medycyna Elfów, Kompendium wiedzy o zespole Williamsa*, Presscom, Wrocław 2017; T. Hutyra, K. Mowszet, A. Stawarski (ed.), *Dzieci chore, niepełnosprawne i z utrudnieniami w rozwoju*, Oficyna Wydawnicza "Impuls", Kraków 2008.

The disease is caused by a random loss of genes (deletion includes about 20 genes)[2]. It manifests itself in characteristic physical (dysmorphic facial features), medical (cardiovascular diseases), cognitive (moderate intellectual disability) and socioemotional disorders[3]. Persons with Williams syndrome are characterised by a different profile of cognitive and socioemotional functioning. They are mildly to moderately intellectually disabled, however, they are empathetic, sociable, and initiate interpersonal relations with ease. They have musical and language acquisition abilities[4]. Children affected by Williams syndrome have the following symptoms: metabolic problems, idiopathic hypercalcemia, delayed growth, general developmental delays, impaired eyesight, and cardiologic disorders (supravalvular aortic stenosis, heart murmur, hypertension)[5].

## Changes in the central nervous system in Williams syndrome

Thanks to their better developed auditory cortex patients with Williams syndrome have high musical abilities[6]. The chromosomal defect does not affect the network of connections between the frontal lobe, temporal lobe, and cerebellum, which is why patients with Williams syndrome have the opportunity to develop their language skills. Research has demonstrated that the general volume of cerebral cortex is lower than that of healthy persons, although the anatomic structure of the brain does not significantly deviate from normal[7]. The brain volume of persons with Williams syndrome is lower by about 13%. There is a significant loss in the mass of the brain stem (ca. 20%). The volume of white matter is visibly lower. A proportionally larger cerebellum is typical[8].

## Characteristic of patients with Williams syndrome

In Williams syndrome dysmorphia of facial skeleton occurs: large, wide mouth with prominent lips, wide jaw, small mandible and chin, full cheeks,

[2] M. Buchnat, K. Pawelczak, *Nieznane? Poznane, Zaburzenia rozwojowe*..., op. cit.
[3] A. Sampaio, J. Belsky, I. Soares et al., *Insights on Social Behavior From Studying Williams Syndrome*, „Child Development Perspective" 2017, pp. 1–6.
[4] A. Stefanowicz, M. Hajducka, M. Krajewska et all., *Problemy zdrowotne dziecka z Zespołem Williamsa-Beurena*, „Nowa Pediatria" 2016, pp. 183–190.
[5] Ibidem.
[6] Ibidem.
[7] M. Giers, *Wspomaganie rozwoju dzieci z zespołem Willamsa. Rozwój poprzez terapię. Interdyscyplinarne aspekty pedagogiki leczniczej*, [in:] M. Bidzan, W. Budziński (eds.), Wydawnictwo GWSH, Gdańsk 2011.
[8] Ibidem.

small snub nose with hollow bridge with a large round tip, nostrils leaning forward, flat malar region, long philtrum, broad forehead, relatively large eyes with an epicanthic fold, swollen orbital cavities, blue iris with a lace-like pattern, narrow and small teeth (particularly incisors) with noticeable interdental gaps, as well as frequently enlarged and protruding auricles[9]. Because of the characteristic appearance and friendly, joyful attitude children with Williams syndrome are described as "dwarf children" or "elf children"[10].

## Williams syndrome as a multiorgan disease

In Williams syndrome defects connected with the cardiovascular system frequently occur. Typically it is a supravalvular aortic stenosis, whichaffects 75% of patients. The pulmonary aorta or renal vessels are an example of anatomicaldefects in the structure of other vessels. Cardiovascular pathologies are resultant from hypertrophy of tissues, which leads to the narrowing of blood vessels. The level of the narrowing varies between patients. Heart murmurs and narrowing of the main blood vessels are anomalies which are found in 67% of new-borns and infants. They may lead to severe cardiac defects. The narrowing of vessels may also result in hypertension. This pertains to 40% adults and 46% of children with Williams syndrome. In connection with the changes in cardiovascular system the risk of cardiac infarction and stroke is heightened[11].

Yet another problem is the disorders of the urinary system[12] as well as endocrinological disorders. Idiopathic hypercalcemia, that is, increased levels of calcium[13] affects 15% of patients[14]. In older children the hypercalcemia tends to be mild, however, in infants it can be significant and lead to irritation, hypotonia, abdominal colic, and loss of appetite. 15% of children suffer from other metabolic dysfunctions. There are difficulties in feeding, colic, abdominal pains, constipation, and vomiting. These may result in irritation and delays in psychomotor development. Metabolic abnormalities lead to an increased risk of later problems such as nephrocalcinosis and sclerotization of long bones[15].

[9] A. Stefanowicz, M. Hajducka, M. Krajewska et all., *Problemy zdrowotne dziecka*..., op. cit.
[10] M. Buchnat, K. Pawelczak, *Nieznane? Poznane, Zaburzenia rozwojowe*..., op. cit.
[11] A. Stefanowicz, M. Hajducka, M. Krajewska et all., *Problemy zdrowotne dziecka*..., op. cit.
[12] Ibidem.
[13] J. Wierzba (ed.), *Medycyna Elfów*..., op. cit.
[14] N. Bishara, C.L. Clericuzio, *Zespoły dysmorficzne często spotykane w oddziałach intensywnej terapii noworodków*, „Pediatria po Dyplomie" 2009, pp. 70–82.
[15] T. Hutyra, K. Mowszet, A. Stawarski (ed.), *Dzieci chore*..., op. cit.

## Development of children with Williams syndrome

The duration and the course of the pregnancy in women whose children are born with Williams syndrome is normal. Children are born on time; however, their body mass and length are lower than those of healthy children. Infants with Williams syndrome reach the body length of ca. 48.3 cm. In the case of healthy children it is ca. 50 cm. Disorders of sucking and swallowing reflexes may occur in infants, as well as abdominal and oesophageal reflux, and vomiting. These disorders may lead to a lowered body mass in infancy. The problems with eating lead to anger and irritation of the infants[16].

Children with WS mature earlier, which even in spite of proper feeding may lead to decrease in fatty tissue. This, in turn, results in fatty degeneration[17]. Sexual maturity is reached 1–2 years earlier than in healthy persons and its course is normal. In girls it is on average 10 years of age and in boys 13 years of age[18]. The height of adults varies between 150 and 170 cm[19]. The average height of adults with Williams syndrome is 168 cm for men and 155 cm for women[20].

Problems pertaining to the muscular and skeletal system occur in ca. 50% of patients[21]. These are typically anomalies connected with body posture and the functioning of the joints. After the children are born they have problems with the joints of wide range of movement and muscle hypotonia. These disorders, in turn, weaken muscles, lower muscular elasticity, and influence body posture. The learning of sitting, standing up, and movement in infancy are thus delayed. During the process of maturity increased contractures occur, particularly in ankle and knee joints. The lowered muscle tension typically increases in adult life[22]. In adults pain around the spine occurs[23]. Adults may also be affected by scoliosis[24]. The gait of persons with Williams syndrome is stiff and clumsy. When standing or moving the posture of the body is leaning forward (in particular the upper part of the body). The muscle mass in thighs and lower legsis increased. In about 50% of affected children inguinal hernia and umbilical hernia occur. Persons with the above mentioned disorders require orthopaedic therapy and regular

[16] A. Stefanowicz, M. Hajducka, M. Krajewska et all., *Problemy zdrowotne dziecka*..., op. cit.
[17] Ibidem.
[18] J. Wierzba (ed.), *Medycyna Elfów*..., op. cit.
[19] A. Stefanowicz, M. Hajducka, M. Krajewska et all., *Problemy zdrowotne dziecka*..., op. cit.
[20] J. Wierzba (ed.), *Medycyna Elfów*..., op. cit.
[21] S.M. Reis, R. Schader, H. Milne, *Music and minds: using a talent, developmental approach for young adults with Williams Syndrome*, „Exceptional Children" 2003, pp. 293–313.
[22] M.A. Martens, S.J. Wilson, D.C. Reutens, *Research review: Williams syndrome: a critical review of the cognitive, behavioral and neuroanatomical phenotype*, „Journal of Child Psychology and Psychiatry" 2008, pp. 576–608.
[23] A. Stefanowicz, M. Hajducka, M. Krajewska et all., *Problemy zdrowotne dziecka*..., op. cit.
[24] B.R. Pober, *Medical Progress: Williams-Beuren Syndrome*, „The New England Journal of Medicine" 2010, pp. 239–252.

rehabilitation[25]. Patients with Williams syndrome are affected by dental problems. Both deciduous and adult teeth are smaller and set wider apart than in healthy individuals. Interdental gaps are present. In spite of reaching dental maturity a number of molars are missing. In majority of children bite anomalies occur – in over 85–90% of patients[26] – as well as caries. Caries is typically caused by vomiting which had occurred in infancy[27]. In patients with Williams syndrome various types of bite anomalies are found; usually of class II and III, open bite, deep bite, and scissor bite. Furthermore, micrognathism of the mandible and osteosclerotic changes in the bundle bone of the dental alveolus, in particular in the proximity of premolars and molars. Changes in soft tissues of the oral cavity are also noticed. They are thinned and wrinkled mucosa of the cheeks, as well as hypertrophied fraenum of the upper lip[28]. The tongue can be large and push unto the teeth[29].

Eyesight problems also occur more frequently than in healthy individuals[30]. These may include heterotropia, particularly esotropia (27–78%), hyperopia (68%), amblyopia (24%), and impaired dual eyesight[31]. In adults with Williams syndrome chronic diseases may be present, such as cataract, impaired eyesight, or senile hypermetropia[32].

Sleep disorders accompany Williams syndrome. These include: difficulties in falling asleep, anxiety while asleep, recurring waking during sleep, and involuntary leg movement[33]. The majority of patients with WS present behavioural problems. These are, among others, fear, lack of concentration, lowered tolerance of dissatisfaction, low adaptability to new conditions, variant behaviours (obsessions, stereotypes)[34]. Children may be recalcitrant, hyperactive, emotionally unstable, and aggressive. The majority of patients are affected by intellectual disability. Patients have problems with learning, reading, and writing. Children have difficulties with doing and understanding basic mathematical tasks, such as arithmetic operations or using numbers[35].

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[25] B.R. Pober, *Fulfilling Dreams. A handbook for parents of people with Williams syndrome*, Williams Syndrome Association, Oak 2002.

[26] J. Wierzba (ed.), *Medycyna Elfów*..., op. cit.

[27] A. Stefanowicz, M. Hajducka, M. Krajewska et all., *Problemy zdrowotne dziecka*..., op. cit.

[28] K. Gerreth, T. Pawlaczyk-Kamieńska, *Szczególny charakter postępowania stomatologicznego u pacjentów z zespołem Williamsa*, „Przegląd Lekarski" 2016, pp. 671–674.

[29] J. Szczepańska, E. Pawłowska, *Cechy kliniczne i morfologia części twarzowej czaszki u dzieci z zespołem Williamsa – opis dwóch przypadków*, „Czasopismo Stomatologiczne" 2008, pp. 401–408.

[30] J. Wierzba (ed.), *Medycyna Elfów*..., op. cit.

[31] P. Kaplan, *Williams-Beuren Syndrome. Research, evaluation and treatment*, "The Johns Hopkins University Press", Baltimore 2006.

[32] A. Stefanowicz, M. Hajducka, M. Krajewska et all., *Problemy zdrowotne dziecka*..., op. cit.

[33] Ibidem.

[34] Ibidem.

[35] A. Stefanowicz, M. Hajducka, M. Krajewska et all., *Problemy zdrowotne dziecka*..., op. cit.; B.R. Pober, *Medical Progress*..., op. cit.; K. O'Hearn, B. Luna, *Mathematical skills in Williams*

The results of intelligence tests in persons with Williams syndrome vary significantly between 40 and 90 IQ points. About 55% of studied children achieve results below 50 points, which is interpreted as moderate intellectual disability. Scores of over 70 points are achieved by 4% of the patients, which means that they are within the intellectual development norm[36].

Nany Grejtak, having analysed a typical cognitive profile of students with Williams syndrome, outlined their strengths and weaknesses. The strengths include:

— good memory; good verbal skills, musical talent, the ability to learn with the use of the visual, audial, and kinaestheticmethods, enthusiasm, high level of motivation, dedication[37].

Difficulties of persons with Williams syndrome pertain to visual and motor coordination, abstract and cause-and-effect thinking, planning the organisation of complex tasks, spatial thinking and imagination, difficulty in comprehension of nonverbal messages, stereotypical interests and monopolizing conversation subjects, tendency to focus on details, low concentration, sporadic problems with finding the correct word, problems in modulating emotions, impulsivity, and hypersensitivity to certain sounds[38]. Motor agility of patients with Williams syndrome is impaired. Problems arise with gross motor skills, that is, motor agility of the entire body, as well as fine motor skills – fine, precise hand movements. Motor development may be delayed in comparison with healthy children[39].

Development of perception also constitutes a problem. Frequently difficulties in visual and spatial functioning occur. They are present in the form of problems in differentiating between directions, assessment of distance, and locating objects in space. Differentiating between objects and geometrical figures is problematic, and so are their remembering, sequencing, and classifying[40]. Patients often cannot differentiate between left and right, forwards and backwards, above and below, and they are lost in their surroundings. They also have trouble with putting things together, including jigsaw puzzles and constructions from blocks[41]. Thanks to their good audial memory and ability to perceive and process audial stimuli the patients present high levels of the development of verbal skills[42]. Initially language development may be delayed compared with

*syndrome: Insight into the importance of underlying representations*, „Developmental Disabilities Research Reviews" 2009, pp. 11–20.

[36] M. Buchnat, K. Pawelczak, *Nieznane? Poznane, Zaburzenia rozwojowe*..., op. cit.

[37] M. Giers, *Wspomaganie rozwoju dzieci*..., op. cit.; N. Grejtak, *The gift of learning*, „Williams Syndrome Association", Oak 2002.

[38] Ibidem.

[39] M. Buchnat, K. Pawelczak, *Nieznane? Poznane, Zaburzenia rozwojowe*..., op. cit.

[40] A. Maurer, I. Bołtuć, *Dzieci z zespołem Williamsa, Diagnoza i terapia*, Oficyna Wydawnicza "Impuls", Kraków 2002.

[41] M. Buchnat, K. Pawelczak, *Nieznane? Poznane, Zaburzenia rozwojowe*..., op. cit.

[42] Ibidem.

healthy children[43]. A year's delay is typically observed in children in comparison with children with no developmental disorders; occasionally the delay may be prolonged. What is interesting is that children with WS first utter a word and then point fingers, while in healthy children the sequence is reversed[44]. The differences decrease in school age. Then the speech of most patients with Williams syndrome is easy to understand, clear, fluent, and highly expressive. Children with WS at this age happen to use a broader vocabulary than their peers; they construct complex expressions[45]. That is why they appear significantly more intelligent than their IQ test results would suggest.

Persons with Williams syndrome are characterised by high memorising skills. This is expressed in their precise, nearly flawless recollection of song lyrics, poems, faces, or tone of voice. At the same time they present deficits connected with remembering instructions, facts, or the location of objects in space. They are better at memorising words than visual and spatial information[46].

## Use of physiotherapy in patients with Williams syndrome

Fromthe first days of their lives the development of children with Williams syndrome flows slower than in their healthy peers. Delays in the appearance of milestones are observed. In the first months of life one can observe impairments in central nervous coordination, posture asymmetry, and pathological movement models. Because of the generally lowered muscle tension and elastopathy the learning of sitting, crawling, and verticality is impaired, and after the children have learnt to walk they experience problems with maintaining balance and coordination as well as clumsy gait[47].

In adolescence disproportions in muscle tension appear: it is axially lowered, and distally heightened, muscle contractures occur, especially in the lower limbs. The weak abdominal and back muscles lead to asymmetry of the trunk, resulting in scoliosis – a deformity of the spine in three planes. Spinal-disc-related and degenerative changes in the spine may occur earlier than in the healthy population, leading to pain. Deformations of bones and joints also occur frequently, typically in joints of the hips, knees, and feet. That is why every child with WS needs to be closely monitored for faulty posture duringthe time of

[43] M.A. Martens, S.J. Wilson, D.C. Reutens, *Research review...*, op. cit.
[44] J. Cieszyńska-Rożek, P. Sobolewski, D. Grzesiak-Witek (eds.), *Zaburzenia mowy w wybranych zespołach uwarunkowanych genetycznie*, Wydawnictwo Czelej, Lublin 2018.
[45] M. Buchnat, K. Pawelczak, *Nieznane? Poznane, Zaburzenia rozwojowe...*, op. cit.
[46] M. Giers, *Wspomaganie rozwoju dzieci...*, op. cit.
[47] E. Semel, S. Rosner, *Understanding Williams Syndrome: Behavioral Patterns and Interventions*, Taylor & Francis, London 2011.

their intensive growth[48]. Adults with this syndrome experience the symptoms of aging sooner; this is caused by a deficit of elastin, which is responsible for elasticity of tissues and internal organs, including the heart, lungs, and intestines, which is why they wear out sooner and function worse. Adults with WS report pains of the joints and the spine[49]. All the problems result in the necessity to provide patients with WS of all ages with rehabilitation. Physiotherapeutic proceedings are initiated inthe first days of the life of the child. Because of the frequently occurring cardiac disorders the programme of the therapy ought to be developed in cooperation with a cardiologist to strictly take into account the fitness of a child. That is why the NDT-Bobath method is the therapy of choice – it makes it possible to adjust the type of exercises to the ability of the patient: from low-intensity care actions to highly advanced exercises. The conception of NDT (Neuro-Developmental Treatment)-Bobath, the basis of which isformed by a precise knowledge of the physiology of the movement sequence of a new-born, an infant, and a child, as well as the ability to interpret even the minutest of faults and more serious pathologies, is directed not only at problem solving, but also at discovering the origins of the disorder[50]. The assumptions of NDT-Bobath therapy encompass normalisation of muscle tension, halting improper reflexes, freeing up movement in the form approximating the correct one as closely as possible as well as using and strengthening the acquired motor abilities in everyday activities. Each movement is properly planned and combined with relocating the weight of the body and the centre of gravity, thanks to which a child achieves stabilisation of the limbs in supports with more ease and maintains a proper position of the entire body. The therapist facilitates a maximum personal input of the child into the therapy, while at the same time making sure not to trigger incorrect responses resulting from stress or too strenuous an effort[51].

With the growth of the child one can include other neurophysiological methods which require the cooperation of the patient. A child with WS, depending on their needs, ought to perform active free exercises, as well as active supported, corrective, breathing, manual, balance, motor coordination, and strengthening exercises, along with sensory gymnastics, and formative therapeutic games. The aims of the activities mentioned above are: improving gross and fine motor skills, practicing the contraposing of the thumb in grabbing, improving functions of the hand, forming an upright posture, strengthening the shoulder girdle, practising alternation of movements and coordination.

Proprioceptive neuromuscular facilitation (PNF) is the most commonly employed method. It is based on neurophysiological rules of conducting movement

[48] Ibidem.
[49] Ibidem.
[50] E. Wysoczańska, A. Skrzek, M. Pyzio-Kowalik, *Możliwości zastosowania metody NDT-Bobath w rehabilitacji pediatrycznej*, „Fizjoterapia" 2013, no. 21.
[51] A. Nawrat-Szołtysik, J. Pasek, *Fizjoterapeutyczne KNOW-HOW*, Elamed Media Group, Katowice 2017.

actions and their development in human life. Proprioceptive facilitation also influences the state of movement centres thanks to the stimulation of deep receptors found in such structures as muscles, ligaments, and joint capsules. Its main aims include the use of diverse movement models which all people perform automatically. In the method the models for shoulder blades, trunk, head, lower and upper limbs are developed in detail, along with models for unison movements of the limbs. The conceptions includes the technique of proprioceptive stimuli, which provides the opportunity of conducting coordinated movements within a proper range, thanks to the use of increasing resistance of the hand of the therapist. Muscle synergisms are employed to stimulate weaker muscle groups. Exercises are selected individually, taking into account the needs of the patient[52]. If a patient is affected by scoliosis, methods typically used with scoliosis can be employed, e.g., FITS, FED, Spiral Muscle Stabilisation System. In the case of spinal disc hernia the McKenzie therapy is highly effective. The FDM Neuro (Fascial Distortion Model) method is also worth using, because it is effective in children and adults with muscle tension pathology in the course of neurological diseases, as well as patients with other disorders. Neuro FDM deals with functional faults of the neuro-fascial system. It is recommended that the method be used as a supplement of the traditional functional techniques (International FDM Organisation (IFDMO)). All the methods can be successfully used in adult patients. Depending on the needs of the patient physicotherapy massages, and water therapy can be employed, however, the cardiac fitness of the patient needs to be taken into account every time.

## Speech therapy

Because of the generally lowered muscle tension in infants with WS it is frequent that problems with sucking occur, leading to the necessity to introduce clinical speech therapy in the first days of their lives. Elements of the Castillo-Morales method are typically employed. It is crucial that the cooperation of the mother with a lactation consultant be maintained. In the following months of life one can observe progress in the development of active and passive speech, and, if needed, stimulation of prattling, imitating gestures and sounds, understanding and communication, can be introduced. Speech therapy ought to focus on visual analysis and synthesis in working with spatial materials, on tasks including cause-and-effect relationships, tasks employing visual memory, abstract thinking, comprehension of rules and imitation, understanding jokes and differentiating between truth and fiction, as well as expanding active and passive vocabulary. In spite of rich active vocabulary, children do not always comprehend the

[52] A. Stępień, K. Graff, A. Koloze et al., *Metoda PNF w odniesieniu do wytycznych Society on Scoliosis Orthopaedic and Rehabilitation Treatment* (*SOSORT*) *dotyczących leczenia zachowawczego osób ze skoliozami*, „Postępy Rehabilitacji" 2014, no. 4.

meaning of the word which they use. What also constitutes a problem is comprehension of simple words and relations of place (in, on, under, between), direction (left, right), time (earlier, later), size (bigger, smaller), equality (the same, different), exclusion (none, except for, all)[53]. Grammar exercises are required because of frequent generalisation of grammatical rules[54], as well as graphomotoric exercises.

## Pedagogical and psychological therapy

In relation to the socioemotional development of persons with WS one can notice problems in establishing relationships with peers: such persons, however, are exceptional in the fact that they are eager to enter into relations with adults and older children. Children with WS are very friendly and open in relationships. On the other hand, they are often excessively expressive, their need to be accepted is great, and they tend to be too trusting of strangers[55].

Diversity among children with Williams syndrome is high. First of all, the specific profile of the syndrome makes it stand apart from other disorders, because the language and social levels are high in relation to the perception and motor skills development and the ability to concentrate[56]. Furthermore, there are individual differences between persons with Williams syndrome. That is why when creating the programme supporting their development, or a therapeutic programme, one ought to take into account the possibility of major differences in cognitive and socioemotional development even between children whose intellectual development levels are comparable[57].

The Williams syndrome cognitive profile (WSCP) is measured with the use of Differential Ability Scales (DAS). What is assessed in the diagnosis are: "Constructive and visual and spatial skills on a level lower than the level of general intellectual abilities; Constructive and visual and spatial skills on a level lower than the level of short-term memory; Constructive and visual and spatial skills on a level lower than the level adequate for the age; Good audial memory and short-term memory and language abilities exceeding the level of general intellectual abilities" (Mervis 1999)[58].

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[53] J. Cieszyńska-Rożek, P. Sobolewski, D. Grzesiak-Witek (eds.), *Zaburzenia mowy*..., op. cit.

[54] M. Maratsos, L. Matheny, *Language specificity and elasticity: Brain and clinical syndrome studies*, „Annual Review of Psychology" 1994, no. 45.

[55] M. Giers, *Wspomaganie rozwoju dzieci z zespołem Willamsa. Rozwój poprzez terapię. Interdyscyplinarne aspekty pedagogiki leczniczej*, [in:] M. Bidzan, W. Budziński (eds.), Wydawnictwo GWSH, Gdańsk 2011b.

[56] A. Maurer & I. Bołtuć, *Zaburzenia rozwoju u dzieci z zespołem Willamsa*, [in:] J. Pilecki i M.A. Winzer (eds.) *Problemy edukacji i rewalidacji dzieci niepełnosprawnych*, WSP, Kraków 1996.

[57] M. Giers, *Wspomaganie rozwoju dzieci*..., op. cit.

[58] Cf.: M. Giers, *Wspomaganie rozwoju dzieci...,* 2011b; cf.: C.B. Mervis, B.F. Robinson, J. Bertrand, C.A. Morris, B.P. Klein-Tasman, S.C. Armstrong, *The Williams syndrome cognitive profile*, „Brain And Cognition" 2000, no. 44(3).

In psychological and pedagogical therapy it is crucial to generate a description of the functioning of the child at home and at preschool/school in the didactic, emotional and motivational, and social (contacts with peer group, teachers, adaptation to the school environment, self-service, skillsand learning difficulties) areas. To achieve this it is worth to juxtapose the observations made by teachers and parents. Scales of behaviour assessment encompassing the level of adaptation of a child, self-service, and skills, have been developed by Iwona Bołtuć[59] and they are presented in the course book "Dzieci z zespołemWilliamsa – diagnozaiterapia" [Children with Williams syndrome – diagnosis and therapy]. The authors claim that the assessment of the behaviour of children marked on a scale by parents and teachers often vary significantly[60], which ought to be analysed when creating an individual programme of supporting development. Cooperation between parents, teachers, and other persons from the immediate environment of a child with Williams syndrome is indispensable at the stage of the diagnosis as well as in the course of the support programme. As has been previously mentioned, children with Williams syndrome like to find themselves in relations with persons older than them and they are too trusting; they can also be perceived as individuals who give too much attention to those who they wish to be friends with. That is why it is essential to create a safe environment for persons with WS and educate the people directly engaged in relations with them (but not exclusively). Apart from professional help parents can also take part in support groups. In the case of rare diseases and disorders this form of help is particularly important, because it counteracts the sense of loneliness in problems that parents need to face on a daily basis. Contemporarily, traditional support groups can be substituted by internet forums and internet websites for those interested in the problems of WS. The exceptional musical skills[61] and the willingness to cooperate can also be used in the work with children with WS. These are the traits which differentiate children with Williams syndrome from other persons with developmental disorders[62]. According to the assumptions of corrective-compensation therapy the weaknesses of the patient ought to be worked on by reinforcing the strengths. That is why music should be included in various areas of the life of children with WS, e.g. while studying (by learning songs), learning to cooperate with peers (singing in a choir), therapy (relaxation), or movement exercises (dance, which contributes to improved coordination and awareness of one's body). One ought to bear in mind, however, that apart from often absolute musical hearing children with WS are hypersensitive to loud sounds and noise[63].

[59] A. Maurer & I. Bołtuć, *Zaburzenia rozwoju u dzieci*..., op. cit., A. Maurer, I. Bołtuć, *Dzieci z zespołem Williamsa*..., op. cit.
[60] A. Maurer & I. Bołtuć, *Zaburzenia rozwoju u dzieci*..., op. cit.
[61] A. Stefanowicz, M. Hajducka, M. Krajewska et all., *Problemy zdrowotne dziecka*..., op. cit.
[62] M. Giers, *Wspomaganie rozwoju dzieci*..., op. cit.
[63] B. Gilbert-Dussardier, *La Revue Du Praticien*, „Williams-Beuren syndrome" 2006, no. 56(19).

That is why the music which they listen to should notbe too loud. Playing a musical instrument should be considered, because this contributes to the work on one's diligence and positively influences self-esteem and perception by others[64]. To enhance concentration and the particular areas of children's behaviour one can employ the techniques which were developed for children with psychomotor hyperactivity and attention deficit[65]. What can be helpful is to take care of the child's environment (by removing objects which may distract the child) and to divide tasks into shorter ones, with the child rewarded at the end of each part.

## Conclusions

Rehabilitation of children with WS ought to be an interdisciplinary process, to be conducted in close cooperation between specialists of various methods, and to be individually selected to meet the needs of the patient. Therapeutic proceedings need to be initiated as soon as the condition of the child allows and be carried out continuously, with an ongoing adaptation of the programme of therapy to the changing needs of the patient. An important method in the work with a child with WS is sensory integration, corrective and compensation methods using strengths, and the techniques developed for children with psychomotor hyperactivity and attention deficits. Because of the generally impaired development of the child ongoing motor and intellectual skills therapy is necessary. Children require diverse motor skills therapy, working on the development of orientation in the outline of the body and the sense of the body in space, and motor planning skills. Only thanks to systematic work on physical agility in connection with pedagogical and psychological work, as well as speech therapy, can children with WS be provided an opportunity to function well in the future.

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[64] M. Giers, *Wspomaganie rozwoju dzieci*..., op. cit., 2011b.
[65] Ibidem.

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