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Self-eteem evaluation of girls of Turner syndrome

Poziom samooceny u dziewcząt z zespołem Turnera

Turner syndrome (TS) is a condition caused by a complete or partial absence of genetic information in one of the two X chromosomes in women. It is estimated that there are 1.5 million women living with this condition in the world. The approximate incidence rate is 1/2500 female births. Currently, there are 9-10 thousand cases in Poland, with around 100 children with Turner syndrome being born each year [3,4,10]. The condition, described in 1938 by H. H. Turner, affects women and is associated with a characteristic phenotype and a number of somatic abnormalities. All individuals with Turner syndrome have growth disorders that begin in intrauterine life. Delayed growth and skeletal maturation, relatively mild in the first few years following birth, become more pronounced from 7 years of age onwards. Limbs tend to grow at an even slower rate than torso, which further contributes to disproportions in stature. The most apparent growth deficiency (compared to general population) of over 25 cm occurs between 11 and 15 years of age as a result of the so-called "puberty spurt" in girls with Turner syndrome [3]. A characteristic feature is the lack of sexual development in puberty and infertility (innate gonadal dysgenesia). The most significant morphological features are directly related to the chromosomal condition and include: facial dysmorphia, webbed neck, puffy hands and feet, arms turned out at elbows, long fingers, low-set and/or unusually rotated ears, broad chest, etc. [7]. There are also concurrent health problems and internal organ disorders, such as: impaired vision, pigmented nevi, kidney and cardiovascular anomalies, skeletal abnormalities, recurrent otitis media, hearing loss, arterial hypertension, insulin-independent diabetes, etc. [3,7]. Early diagnosis during the perinatal period allows for early monitoring of the patient's development and effective case management. Unfortunately, Turner syndrome tends to be diagnosed later in life, when delayed growth and lack of puberty become a cause for concern. Even though the situation of women with TS in Poland has recently been cleared up and the need for routine pharmacological treatment (hormonal treatment of growth deficiencies and lack of sexual development) is no longer questioned, there are still a number of unresolved issues with regard to psychological and educational support for the affected women. While focusing on the medical aspect of TS, we should not overlook psychological problems that these women face. According to a number of researchers, along with physical problems, individuals with Turner syndrome have characteristic profiles of psychosocial and neuropsychological functioning. There is a general consensus that women with TS suffer deficiencies in visuo-spatial abilities, spatial imagination, visuo-motor coordination, visual and visuo-spatial memory [6,9]. They have characteristic difficulties in sorting visual data, i.e. in spatial and visuo-motor organisation that requires integration skills. These problems contribute to educational problems and difficulties in learning mathematics [6,7], as well as causing lower efficiency in executive processes and motor planning [8]. Ongoing research studies focus on the search for neuroanatomical and neurophysiological basis of those deficiencies. Their results imply a presence of anomalies in the hemispherical lateralisation that cause the right hemisphere to be underdeveloped as compared with typical asymmetry found in healthy, age-adjusted controls [5]. It is suggested that damage in parietal and occipital lobes of individuals with TS causes deficiencies in visuo-spatial skills [6] and that there is some damage in subcortical

structures, such as cerebellum and pons, hippocampus, lentiform nucleus, and optic thalamus [6,7]. Abnormalities in those regions could constitute a neurological basis for the deficiencies in executive processes and motor planning. The most common social competence disorders include: difficulties in social interaction and problems in forming close, friendly relationships at school [9,4]. Mature women with TS reach independence from their parents at a later age than their healthy peers and often achieve lower professional status than expected for their level of education [4,7]. Studies on intellectual functioning demonstrate that girls with Turner syndrome typically have an unbalanced intelligence profile, scoring higher on the verbal scale than the executive scale. This trend continues into adulthood [6]. Descriptions of the emotionality of women with TS emphasize their immaturity and withdrawal, increased anxiety, loneliness, attention deficits and emotional over-sensitivity, and difficulties with emotional control [2]. Besides the severity of the aforementioned disorders, of equal importance are individual differences among women with TS, since the aforementioned problems occur in some of them, but not others.

Emotional problems are related to and can be manifested by, for example, low self-esteem. Research conducted by E. McCauley et al. (1995) on girls with TS aged 7-14 demonstrated that self-esteem in terms of appearance, anxiety, popularity among peers, and happiness is lower in girls with Turner syndrome as compared to healthy girls of the same age. These tendencies become more pronounced with age (especially between 11 and 13 years of age). Socio-economic status of the family had no effect on the level of self-esteem in girls who participated in the study [4]. Adolescence is a particularly high-risk period for girls with TS: self-esteem tends to drop in puberty. Unfriendly interactions, negative peer attitudes are experienced with particular acuteness in adolescence and can have severe consequences for future functioning. Adolescence is marked, among other things, by noticeable physical, physiological, and psychological changes that are likely to result in low self-esteem. Therefore it seems worthwhile to analyze those changes in terms of the multiple dimensions of self-esteem.
The purpose of the present study was to compare the level of self-esteem in girls with Turner syndrome and their healthy peers, and to find out whether there were any differences in terms of: self-esteem with regard to health and physical abilities, assessment of their own appearance, intellectual abilities and attitudes towards schoolwork, attitudes towards parents, work, teachers, attitudes towards themselves, others, life, and the attitude towards boys among the girls in the study.

SUBJECTS

There were two groups of girls in the study. The experimental group consisted of 30 girls with TS; the control group of 30 healthy girls. Age distribution was similar in both groups. Subjects were aged 10-15 years. All girls came from two-parent families.

METHODS

The “What am I like?” Self-Esteem Scale devised by Paulina Sears and Janusz Kostrzewski was used in the study. The scale measures self-esteem and consists of 138 questions in 9 self-esteem categories, such as: health status and physical abilities, appearance, intellectual abilities and attitude towards schoolwork, attitude towards parents, work, teachers, attitudes towards self, others, and life, attitude towards boys and girls. The Educational Environment Questionnaire (1971) devised by Jan Konopnicki and Marian Ziemba was used to collect information about the environment in which the girls were being raised.

RESULTS

Subjects were asked to complete the Educational Environment Questionnaire (1971) devised by Jan Konopnicki and Marian Ziemba in order to determine whether there were any statistically significant differences with regard to educational environment. General results showed no statistically significant environmental differences between the two groups of girls ($p=0.01$). All subjects came from similar settings that encouraged personal development. Statistical analysis of data obtained from the study leads to the following conclusions:

1. There are statistically significant differences in the level of self-esteem with regard to self, others, life ($p+0.001$); attitude towards boys ($p=001$), self-esteem with regard to health status

and physical abilities (p=0.003), appearance (p=0.002) between girls with TS and their healthy peers. In those dimensions of self-esteem, girls with TS score lower than their peers.

2. There are no statistically significant differences between the level of self-esteem in girls with TS and their healthy peers with regard to intellectual abilities and attitude towards schoolwork (p=0.422); attitude towards parents (p=0.434); attitude towards teachers (p=705); attitude towards girls (p=0.172). No differences in terms of those dimensions of self-esteem were found between the two groups of girls in the study.
3. The results suggest the presence of very similar relationships between all dimensions of self-esteem with regard to "idealized self" of girls with TS and their healthy peers. All girls in the study wanted to improve themselves and their relationships with others in the near future.

DISCUSSION

Results obtained in the study demonstrate statistically significant differences in the level of self-esteem in subjects. Typically, girls with TS have lower self-esteem with regard to appearance (short stature), physical features (disproportionate body build) and assessment of health status and physical abilities (health problems, lower fitness), all of which are TS-specific disorders. These results are in line with the data obtained by McCauley et al. [4]. Presumably, years of treatment for various health problems significantly affect the subjects' level of self-acceptance in terms of the categories analyzed in the present study. In addition, research conducted by Gracka-Tomaszewska and Radoszewska (2001) shows that parents of girls with TS who describe their daughters tend to focus on their abnormal appearance, which is the core of Turner syndrome, and often overlook some positive aspects of their functioning [1]. The perception of girls with TS should not be influenced by their appearance, short stature or disproportionate body build. They need to be treated according to their age rather than their height, and special attention must be paid to the difficulties they experience. Those issues also affect their attitude towards boys and their interactions with the opposite sex. Girls in the present study often felt as being unattractive partners in play and conversation with their male peers and failed to establish close relationships with boys.

Any analysis of self-esteem should go beyond external appearance and take into account social interactions. Findings suggest that social relations with girls of the same age are not affected by the fact that girls with TS tend to have low self-esteem with regard to their appearance, health status and physical abilities. Subjects with TS evaluate them as satisfactory, they experience the sense of attachment, being understood and belonging to a peer group. They enjoy spending time with their girlfriends and feel accepted in those interactions. The need for a close extrafamilial attachment is prominent and typical for that developmental period: girls want to make friends among their peers. The fact that there were no differences between the two groups in terms of self-esteem with regard to intellectual abilities, attitude towards schoolwork and teachers might suggest that girls with TS try to compensate for their low assessment of their appearance and lack of physical attractiveness with educational achievements and good cognitive functioning. In addition, both groups in the study gave positive assessments of their relationships with parents: they like to play and talk to them, they are willing to help them with household chores, and enjoy mutual understanding and trust. Findings show that it is necessary to promote the formation of appropriate self-esteem in girls with TS in all of its aspects. Low self-esteem is detrimental to psychological stability and may encourage girls to adopt ineffective patterns of responding to adverse situations.

CONCLUSION

1. Girls with TS are in the high-risk group for physical disorders and psychosocial difficulties.
2. They require integrated, interdisciplinary medical assistance for TS-specific health problems, as well as psychotherapeutic and educational support at various stages of life. The period of adolescence, which is particularly important for building self-esteem, is particularly testing for girls with Turner syndrome. It is during that time that they require a special kind of support, motivating them to make use of the psychological resources that will help them face everyday problems. The aim is to augment their sense of self-worth, encourage confidence in their competence and

intellectual abilities, show them their strengths and weaknesses. That potential is necessary if they are to be capable of facing challenges and taking roles that come with adulthood.

3. Turner syndrome should not prevent women from leading full and productive lives. All help and support schemes should be aimed at providing individuals suffering from TS with a quality of life comparable to that of their peers.

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ABSTRACT

The purpose of the study was to determine the level of self-esteem in girls with Turner syndrome as compared with their healthy peers. Thirty girls participated in the study. Subjects were aged 10-15. The "What am I like?" Self-Esteem Scale by Paulina Sears and Janusz Kostrzewski was used. We found that girls with TS have lower self esteem with regard to themselves, others, life, boys, health status and physical abilities, appearance, and attitude towards work as compared with healthy girls of the same age. There were no statistically significant differences between the level of self-esteem in girls with Turner syndrome and their healthy peers with regard to intellectual abilities and attitude towards schoolwork, parents, teachers, and other girls.

STRESZCZENIE

Celem badań było określenie poziomu samooceny dziewcząt z zespołem Turnera w porównaniu do ich zdrowych rówieśniczek. W badaniu uczestniczyło 30 dziewcząt. Wiek badanych wynosił 10-15 lat. Zastosowano Skalę samooceny Pauliny Sears i Janusza Kostrzewskiego „Jaki jestem?". Stwierdzono, że dziewczęta z ZT mają niższy poziom samooceny w stosunku do siebie, do ludzi, do życia, do chłopców, samooceny stanu zdrowia i możliwości fizycznych, oceny swojego wyglądu zewnętrznego oraz stosunku do pracy w porównaniu ze zdrowymi rówieśnicami. Nie ma statystycznie istotnych różnic w poziomie samooceny dziewcząt z ZT i ich zdrowych rówieśnic w zakresie oceny swoich możliwości intelektualnych oraz stosunku do nauki szkolnej, do rodziców, nauczycieli oraz do dziewcząt.