ORIGINAL ARTICLE ÖZGÜN ARAŞTIRMA

Psychometric and Psychosocial Evaluation of Adolescents with Turner Syndrome in a Multidisiplinary Approach: A Preliminary Study

Turner Sendromlu Ergenlerin Multidisipliner Yaklaşımla Psikometrik ve Psikososyal Açıdan Değerlendirilmesi: Ön Çalışma

Burak Karakök* (0000-0002-2191-072X), Devrim Akdemir* (0000-0003-2116-4461), Sıddıka Yalçın (0000-0001-9061-4281), Hacer Seniz Özusta* (0000-0002-3715-9213), Gülen Eda Utine*** (0000-0001-6577-5542), Özlem Doğan*** (0000-0002-8310-4053), Pelin Özlem Şimşek Kiper*** (0000-0001-7244-7766), Gizem Ürel Demir*** (0000-0002-9928-3236)**

*Hacettepe University Hospital, Clinic of Child and Adolencance Psychiatry, Ankara, Turkey

**Hacettepe University Hospital, Clinic of Social Pediatrics, Institute of Child Health, Ankara, Turkey

***Hacettepe University Hospital, Clinic of Pediatric Genetics, Ankara, Turkey

Abstract

Introduction: The aim of this study is to compare neurocognitive and psychosocial characteristics in adolescents with Turner Syndrome (TS) and age-matched adolescents with short stature (SS) and normal karyotypes.

Materials and Methods: Seven patients with TS and 7 patients with SS and normal karyotypes were included in the study. Their comorbid psychopathologies, cognitive functioning, quality of life, self-esteem, emphatic tendencies, mentalizing abilities and coping strategies were investigated.

Results: Although the adolescents with SS had higher levels of anxiety and conduct problems, there were no significant differences between the TS and SS groups in terms of comorbid psychopathologies, social cognition skills, quality of life, self-esteem and coping strategies. However, the cognitive functioning of adolescents with TS was found to be lower than both of the adolescents with SS and community samples.

Conclusions: According to this preliminary study, anxiety/conduct problems and cognitive functioning of patients with TS should be evaluated in order to prevent subsequent negative outcomes.

Öz

Giriş: Bu çalışmanın amacı, TS olan ergenler ile kısa boylu ve normal karyotipi olan benzer yaştaki ergenleri nörobilişsel ve psikososyal olarak karşılaştırmaktır.

Gereç ve Yöntem: Çalışmaya TS olan yedi hasta ile kısa boylu ve normal karyotipi olan yedi hasta dahil edilmiştir. Eşlik eden psikopatolojiler, bilişsel işlevsellik, yaşam kalitesi, benlik saygısı, empatik eğilimler, zihinselleştirme becerileri ve baş etme stratejileri araştırılmıştır.

Bulgular: Kısa boylu ergenlerde anksiyete ve davranım problemleri daha yüksek olmasına karşın, eşlik eden psikopatolojiler, sosyal biliş becerileri, yaşam kalitesi, benlik saygısı ve başetme stratejileri açısından TS olan ergenler ile kısa boylu ergenler arasında önemli bir fark bulunmamıştır. Bununla birlikte, TS olan

Keywords
Turner syndrome, psychopathology, psychosocial characteristics, cognitive functioning, short stature

Anahtar kelimeler
Turner sendromu, psikopatoloji, psikososyal özellikler, bilişsel işlevsellik, kısa boy

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Address for Correspondence/Yazışma Adresi: Burak Karakök MD, Hacettepe University Hospital, Clinic of Child and Adolencance Psychiatry, Ankara, Turkey
Phone: +90 542 649 30 09
E-mail: burak.karakok@hotmail.com

ergenlerin bilişsel işlevlerinin hem kısa boylu ergenlere hem de toplum örneklemine göre daha düşük olduğu bulunmuştur.
Sonuç: Bu ön çalışmaya göre, ileride gelişebilecek olumsuz sonuçları önlemek için TS'li hastalar anksiyete / davranış sorunları ve bilişsel işlevler açısından değerlendirilmelidir.

Introduction

Turner syndrome (TS) is a relatively common chromosomal disorder, affecting one in 2000-2500 live-born females (1). The condition is cytogenetically characterized by partial or total loss of one of the two X chromosomes. Main clinical features include short stature and puberty / fertility problems caused by premature ovarian insufficiency. Frequently, there are also dysmorphic features, and sometimes major malformations or various systemic disorders may affect the patients. Various inherent neurocognitive and psychosocial characteristics may be observed in this syndrom and additional psychosocial problems originating from perception of physical problems may further complicate the condition.

The typical neurocognitive profile in TS consists of deficits in visual-spatial skills, executive functions and social cognition (2). IQ may be low-normal and performance may be relatively weak in mathematics. Attention deficit and hyperactivity disorder (ADHD) may contribute to dysfunction in both academic and social adaptation; and psychosocial characteristics may be determined by both inherent and environmental factors. Particularly, impulsivity, slower processing speed, attention deficit, impaired use of nonverbal behaviors may contribute to psychosocial morbidity of adolescents. Consequently, lower self-esteem, social isolation, anxiety and depression symptoms may be common in adolescents with TS (3). Studies revealed that women with TS may be dissatisfied with life due to feelings of loneliness and also short stature and various handicaps (4). Many environmental factors may complicate the condition as indicated by a study showing that even the age at diagnosis may contribute to psychosocial health, peripubertal diagnosis more commonly leading to depression, substance use, and perceptions of insufficient competence (5).

Studies on psychosocial characteristics of adolescents with TS are limited in number. We aimed to compare various aspects of neurocognitive and psychosocial characteristics in adolescents with TS and age-matched adolescents with short stature and normal karyotypes in a multidisiplinary approach as a preliminary study. Although there are several previous studies performed using several tests, the present study is a comprehensive one including many tests. In particular, Schedule for Affective Disorders and Schizophrenia for School Age Children - Present and Lifetime Version (K-SADS-PL), a semi-structured clinical psychiatric interview for diagnosing the present and lifetime psychopathologies of children and adolescents has not been previously applied to adolescents with TS.

Materials and Methods

Participants

This case-control study was performed between January 1, 2019 and May 1, 2019 in collaboration with Department of Child and Adolescence Psychiatry, Units of Pediatric Genetics and Social Pediatrics in a university hospital in Turkey.

The study group consisted of patients with Turner Syndrome (TS) diagnosed and followed by the Pediatric Genetics Unit and who were 12-17 years of age at the time of the study. During study period 12 parents and their girls were invited for the study and seven couple were wanted to participate. And, seven girls between 12-17 years of age who admitted to the same clinic with short stature (SS) and had normal chromosome analysis were included in the control group.

Procedure

The study protocol was approved by the Institutional Review Board of Hacettepe University (project no: GO17/583). The patients and their parents were informed about the study by phone before they came to their routine visits.

Voluntary parents and their adolescents were referred to the Department of Child and Adolescent Psychiatry. First, a semi-structured clinical psychiatric interview was conducted by a child and adolescent psychiatrist with adolescents and their parents who volunteered to participate in the study and gave written informed consent. Then, RME and WISC-IV were applied to the adolescents by a child and adolescent psychiatrist

and an experienced clinical psychologist, respectively. Self-reported questionnaires were explained in details to the adolescents and their parents. PedsQL and SDQ were filled by both the adolescents and their parents, SRS was filled only by parents, and other scales were filled only by the adolescents. Body weights and height were measured, and z scores of height and body mass index for age (HAZ and BAZ) were calculated with WHO Anthro plus program (6).

Materials and Methods

Schedule for Affective Disorders and Schizophrenia for School Age Children - Present and Lifetime Version (K-SADS-PL). The K-SADS-PL is a semi-structured clinical interview for diagnosing the present and lifetime psychopathologies of children and adolescents (7).

Wechsler Intelligence Scale for Children, Fourth Edition (WISC-IV). WISC-IV is the latest revision of a widely used general intelligence test (8).

The Pediatric Quality-of-Life Inventory (PedsQL). PedsQL is a health-related quality-of-life scale consisting of 23 items on physical health as well as psycho-social functionality (9).

Child Depression Inventory (CDI). CDI is a self-report scale for children aged 6-17 years developed by Kovacs to measure the level of depressive symptoms (10)

Screen for Child Anxiety and Related Disorders (SCARED). SCARED was developed in order to screen childhood anxiety disorders between 8-18 years-of-age (11).

Strenghts and Difficulties Questionarie (SDQ). SDQis a brief emotional and behavioural screening questionnaire for children and adolescents aged 3-16 years (12).

Rosenberg Self-esteem Scale (RSES). RSES was developed to measure self-esteem particularly in adolescents (13).

Ways of Coping Questionnaire (WCQ). WCQ is a self-report, four-point Likert type scale developed to evaluate the coping styles with stress (14).

KA-SI Empathic Tendency Scale - Adolescent Form (KA-SI). KA-SI was developed by Kaya and Siyez (15) in order to measure empathic tendency of adolescents with a tool specific to Turkish culture.

Social Responsiveness Scale (SRS). SRS was developed by Constantino et al. (16) and used to assess the autism-like symptom cluster.

Reading The Mind in the Eyes Test (RME). The 'Eyes Test' was designed by Baron-Cohen et al. (17) to investigate mentalizing abilities according to the Theory of Mind.

Statistical Analysis

Data were studied by the SPSS version 22.0 (SPSS Inc., Chicago, IL, USA). Descriptive data were

Table 1. Socio-Demographic Characteristics of the Study and Control Groups

	Turner syndrome	Short stature	p
Age, years*	14.7 14 (12, 17)	13.6 14 (12, 15)	0.209
Maternal age, years*	42 43 (34, 47)	40 38.5 (37, 48)	0.520
Paternal age, years*	46 45 (36, 54)	42.7 41(40, 51)	0.399
Maternal education≥12 years**	2 (28.6)	3 (42.9)	0.577
Parental education≥12 years**	3 (42.9)	3 (42.9)	0.593
Maternal occupation, working**	2 (28.6)	1 (14.3)	0.515
Birth order, first child**	2 (28.6)	3 (42.9)	0.577
HAZ*	-3.2 -3.2 (-4.2, 2.1)	-2.7 -2.9 (-3.2, -1.8)	0.097
BAZ*	0.6 0.8 (-1.0, 1.9)	-0.4 -0.4 (-2.0, 1.7)	0.165

*Mean, median (range), **n (%)

included mean, median, range (minimum-maximum), frequency, and percentages. Continuous variables of groups were evaluated by Mann–Whitney U test due to limited number of available cases. Categorical variables between the groups were compared with Chi square test. Relationship between mother and child reports of SDQ and PedsQL were analysed with Spearman correlation. Statistical significance was taken as P value of < 0.05.

Results

As shown in Table 1, sociodemographic characteristics of the two groups showed no statistically significant differences. The mean age of the adolescents with TS was 14.7 (Median:14) and the mean age of the adolescents with SS was 13.6 (Median:14), without significant difference between the two groups. Parental age and education levels of both groups were similar. There were no significant differences between the two groups in terms of HAZ and BAZ scores.

The mentalization skills and the autism-like symptoms of the participants in this study were evaluated with the RME, KA-SI and SRS, and no significant differences were found between the two groups (Table 2). Even though two adolescents in the SS group had low self-esteem according to the RSES scores, there was no statistically significant difference in the mean scores of the RSES between the study and the control groups (Table 2).

Psychiatric symptoms of the participants were evaluated with CDI, SCARED and SDQ. There was no significant difference in depressive symptoms (Table 2), but the anxiety symptoms (Table 2) and conduct problems (Table 3) were significantly higher in patients with SS. There were five adolescents in the SS group who received a score of 25 or higher from SCARED, which is a warning sign for the diagnosis of an anxiety disorder. However, there were no adolescents with these scores in the TS group. Adolescents with SS were found to have significantly higher scores than those of adolescents with TS only in the subscale of conduct problems of the SDQ (Table 3). Correlations between the child and parent reports for SDQ subscales in the two groups were low, except for those of emotional symptoms in the group of adolescents with TS. Parents generally reported higher symptoms than adolescents with TS, lower symptoms than adolescents with SS (Table 3).

In this study, there were no significant differences between the two groups in terms of physical health, psychosocial health and total scores of PedsQL (Table 4). Correlations between the child and parent reports for PedsQL subscales were higher in TS group than those in SS group (Table 4). In addition, there were no significant differences in WCQ subscale scores in the both groups (Table 5).

Table 2. Scores of the scales in the study and control groups

		Turner syndrome	Short stature	
Reading The Mind in the Eyes Test		18.1 19 (10-25)	19.6 19 (15-23)	
KA-SI Empathic Tendency Scale		47.7 50 (35-57)	53.6 53 (44-64)	
Social Responsiveness Scale		50 47 (34-65)	46.9 42 (21-64)	
Rosenberg Self-esteem Scale (RSES)		1.9 1 (1-4)	2.9 2 (1-6)	
RSES	High 0-1 Moderate 2-4 Low 5-6	5 (71.4)% 2 (28.6)% 0 (0.0)%	4 (57.1)% 1 (14.3)% 2 (28.6)%	
Child Depression Inventory (CDI)		5 4 (0-13)	9.3 11 (3-13)	
Screen for Child Anxiety and Related Disorders (SCARED)		17.6 19 (11-23)	32.4 38 (8-50)	
SCARED ≥25	0 (0)	5 (71.4)%	**0.021**	

Mean,median (range)

Table 3. SDQ scores in the study and control groups and correlations between the child and parent scores

	SDQ child*			SDQ parent*			Correlations between the child and parent reports**
SDQ subscales	Turner syndrome	Short stature	p	Turner syndrome	Short stature	p	Turner syndrome
Emotional Symptoms	2 1 (0-5)	3.4 3 (0-4)	0.220	5 3 (1-9)	3.7 3 (1-9)	0.516	**0.82 (0.024)**
Conduct Problems	0.4 0 (0-1)	1.7 2 (0-4)	**0.027**	1.4 1 (0-5)	1.1 1 (1-2)	0.765	0.08 (0.872)
Hyperactivity-Inattention	3.4 3 (0-7)	5.7 5 (5-7)	0.052	3.1 4 (1-6)	3.1 3 (1-5)	0.896	0.58 (0.171)
Peer Problems	2.3 3 (0-5)	2.4 3 (0-4)	0.896	3.7 3 (2-6)	2.6 3 (1-4)	0.209	-0.25 (0.585)
Pro-social Behaviour	8.7 9 (7-10)	8.6 8 (7-10)	0.740	8.3 8 (7-10)	3.3 8 (6-10)	0.789	-0.10 (0.824)
Total Difficulties Score	8.1 9 (0-16)	13.3 14 (8-17)	0.072	13.3 11 (6-22)	10.6 11 (5-16)	0.367	0.72 (0.068)

Mean, median (range)

Table 4. PedsQL scores in the study and control groups and correlations between the child and parent scores

	PedsQL child*			PedsQL parent*			Correlations between the child and parent reports**
PedsQL subscales	Turner syndrome	Short stature	p	Turner syndrome	Short stature	p	Turner syndrome
Physical health total score	557 525 (400-800)	650 675 (450-800)	0.223	492.9 450 (200-800)	596.4 675 (375-800)	0.518	**0.87 (0.010)**
Psychosocial health total score	1107 1250 (500-1375)	1125 1150 (900-1275)	0.404	964.3 950 (175-1450)	1185.7 1175 (875-1425)	0.249	**0.76 (0.050)**
The scale total score	1664 1750 (1025-2175)	1775 180 (1350-2075)	0.654	1457.1 1400 (550-2250)	1782.1 1775 (1250-2225)	0.277	**0.93 (0.003)**

*Mean, median (range), **Spearman correlation coefficient, r (p), PedsQL: Pediatric Quality-of-Life Inventory

The scores of verbal comprehension and perceptual reasoning domains of WISC-IV did not differ significantly in the TS and in the SS groups. However, TS group was found to have lower scores in the working memory, processing speed and total IQ domains. WISC-IV results were summarized in Table 6.

Table 5. WCQ scores in the study and control groups

WCQ subscales	Turner syndrome	Short stature	p
Self-confident	14 14 (8-21)	13 14 (9-16)	0.845
Optimistic	11.9 13 (7-15)	8.9 9 (7-11)	0.081
Seeking of social support	8 8 (2-11)	8 8 (6-10)	0.558
Submissive	5.4 5 (1-9)	6.1 7 (2-8)	0.606
Helpless	5.7 5 (4-8)	10.6 12 (5-18)	0.143

Mean, median (range), WCQ: Ways of Coping Questionnaire

Table 6. WISC-IV scores in the study and control groups

WISC-IV domains	Turner syndrome	Short stature	p
Verbal comprehension	93.4 94 (72-110)	102.3 104 (86-120)	0.404
Perceptual reasoning	74.6 79 (62-89)	88.0 89 (77-102)	0.063
Working memory	68.4 68 (59-77)	89.1 85 (62-115)	**0.021**
Processing speed	77 74 (53-103)	100.9 100 (88-121)	**0.029**
WISC-IV total	71.9 70 (52-86)	93.6 93 (77-107)	**0.013**

Mean, median (range)

Relationship between the SCARED and the WISC-IV total scores in the study and control groups was also demonstrated in Figure 1.

According to K-SADS-PL, only one adolescent with TS was diagnosed with both obsessive-compulsive disorder (OCD) and social anxiety disorder, and the remaining six adolescents in TS group did not meet the diagnostic criteria of any other psychiatric disorder. Past history revealed presence of enuresis nocturna in two of the adolescents with TS. In the SS group, one adolescent was diagnosed with enuresis nocturna and the other one was diagnosed with both specific phobia and enuresis nocturna. Remaining five adolescents in SS group did not meet the diagnostic criteria of any other psychiatric disorder for the present time. One of the adolescents was diagnosed with both separation anxiety and enuresis nocturna, and the other one was diagnosed with both depression and tic disorder in the past in the SS group. There were no significant differences between the two groups in terms of the total number of past or present psychiatric diagnoses.

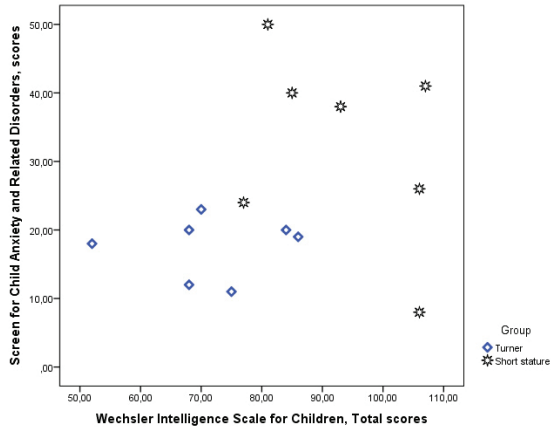

Figure 1. Relationship between the SCARED and the WISC-IV total scores in the study and control groups

Discussion

In this preliminary short-term study, with semi-structured clinical psychiatric interview, K-SADS-PL, only one adolescent with TS was diagnosed with both OCD and social anxiety disorder, and the other six adolescents with TS did not meet the diagnostic criteria of any psychiatric disorder. In control group, one adolescent was diagnosed with enuresis nocturna, and another was diagnosed with both specific phobia and enuresis nocturna. Remaining five adolescents in the control group did not meet the diagnostic criteria of any psychiatric disorder. There was no significant difference between the two groups in terms of the total number of present psychiatric diagnoses, indeed the total psychopathology rates were not high in either of the groups.

In the literature, some studies indicate that the rates of psychopathologies are not high in patients with TS, whereas some others report that the rates of ADHD, autism spectrum disorder, social anxiety disorder and depression are increased (18-24). It was shown that adolescents with TS, when compared to the control groups (short stature or healthy adolescents), were more isolated socially and incompetent in social relationships, had fewer friends and lower self-esteem (25-28). Downey et al. (19) stated that disorders inducing physical impairment, those with chronic courses, those that require hospitalization or multidrug

regimens may be the risk factors for development of psychopathologies in TS. The factors such as not having any additional medical disorders requiring hospitalization, being sufficiently informed about and adapted the disease due to long follow-up, and not being an adult might have prevented the patients in this TS sample from having a psychopathology.

Studies on the psychological health of the patients with TS have mostly been conducted with adult patients and have focused on the effects of physical stigmata on the psychological development of female patients, such as short stature, regression in sexual development and infertility. However, social life and functionality of adults are different from adolescents. Compliance and passivity -'weakness in emotional stimulation' observed in girls with TS by Money and Mittenthal (29) are more common behaviours in childhood and adolescence and these adaptive behaviours may obscure the recognition of other developmental deficiencies. In addition, infertility causes anxiety in adulthood because of fear of rejection by their partners due to being incomplete as a female; and difficulties in marriage, sexual experience and establishing friendships are reported to increase especially with age (28).

There may be more than one reason of obtaining variable results on comorbid psychiatric disorders in TS patients in these studies. Evaluation of comorbid psychiatric disorders with different tools may be an important factor. In the literature, comorbid psychiatric symptoms and disorders in adolescents with TS were assessed with self-report measures, while no study exists using the K-SADS-PL as a clinical psychiatric interview tool. In addition, number of inadequate samples, usage of retrospective data, different geographical and cultural aspects of the studies may have caused variability of results.

In this study, through the self-report scales, participants' empathy, perception of social cues, mentalization skills such as reading the mind through eyes, and coping strategies with stress were evaluated and no significant differences were found between the two groups. In addition, when the groups were compared in terms of psychiatric symptoms, there was no significant difference in depressive symptoms, but the anxiety symptoms and conduct problems were significantly higher in patients with SS. These results might be associated with the fact that adolescents with TS and their families are informed about and adapted to the possible developmental problems from the early period of life when compared to those with SS. However, delay in the physical development of children with SS may predispose them to be more fragile and vulnerable to comments from their peers, and this may possibly canalize families and teachers to behave in a more protective way towards these children. In fact, overprotective attitudes of parents are shown to strongly predict peer victimization in school settings (30). This vicious cycle may result in inadequate social maturity, undemanding attitudes, shyness, and lacking functional independence. On the other end, people might attempt to overcompensate their short stature by adopting overly assertive and bold manner (31). Nevertheless, our sample is too small to generalize these results to all adolescent patients with SS.

When the quality of life was evaluated in the TS and SS groups, no significant differences were found between them in terms of physical and psycho-social health and total scores based on adolescents' and parents' reports in this study. The quality of life is defined by WHO as "the perception of its own situation in the society and value system in which one lives" (32). It is known that psycho-social adjustment and quality of life are low in children with chronic physical illness (33). Perceptions of patients with chronic illnesses that are associated with the course and treatment process of disease can affect their quality of life. In the literature, education level, physical appearance, general health status and psycho-social/emotional problems such as low self-esteem, social anxiety, shyness and sexual issues were determined as predictors of quality of life in patients with TS (34-38). Compatible with the result of this study, previous studies have shown that patients with TS have similar levels of quality of life as compared to the healthy female population (39-41). In this study, almost none of the adolescents with TS had any psychiatric disorders affecting quality of life. In addition, their high self-esteem and coping skills with stress, good general state of health, and similarity with control group in terms of physical appearance affecting their social functionality may have contributed to this result. While the parents of adolescents with SS group were found to sense only physical health challenge of their children, the parents of adolescents with TS group have recognized both physical and psycho-

social health challenges of their children in this study. This result might be related to the fact that the parents of children with chronic health problems have high levels of awareness and sensitivity of their children's health concerns (42).

In our study, while there were no significant differences between the two groups in terms of verbal comprehension and perceptual reasoning domains of WISC-IV, working memory, processing speed and total IQ scores were significantly lower in TS group. In the literature, studies that focused on the cognitive functioning of patients with TS, total IQ scores were found to be lower than the control groups and these results were stated to emanate from lower performance IQ scores. A subtest of performance IQ, namely visual-spatial processing, was significantly lower in patients with TS (43-45). In conclusion, specific IQ profiles of patients with TS are reported to be close to normal in verbal and low in performance dimensions (46). This particular cognitive pattern of patients with TS shows itself as weakness in visual-spatial function, visual memory, visuo-constructive abilities, visual attention, arithmetical abilities, executive functioning (particularly inhibitory control, auditory attention and planning), social interaction, recognition of facial expressions and specific aspects of language (47,48). While the "working memory" domain of WISC-IV evaluates short-term auditory memory, attention, concentration, and working memory; the "processing speed" domain evaluates mental and visual-motor processing speed. Both tasks require efficient production of multiple motor responses such as visual perception and organization, visual screening, effortful control of attention and speed. Since there was not any previous study using WISC-IV in adolescents with TS, we were unable to compare our findings head-to-head with the data in the literature. However, our findings on working memory, processing speed and total IQ in adolescents with TS support the results of existing literature on the IQ tests other than WISC-IV. In addition, a positive correlation was detected between total IQ and anxiety symptoms in our study. Even though there are some researches stating a negative correlation between the intelligence and anxiety, higher cognitive capacity may lead to the development of anxiety symptoms by increasing the awareness of the person to the probable challenges (49).

Study Limitations

This study has several strengths and limitations. In addition to self-report scales, application of of K-SADS-PL and WISC-IV to all participants in order to evaluate accompanying psychopathologies and cognitive functioning, respectively, is a major strength of this study. The most important limitation of this study is the small sample size. Low number of participants in the groups reduces the generalization of the study results.

Conclusion

As a conclusion, although adolescents with SS had higher level of anxiety and conduct problems, there were no significant differences in adolescents with TS compared to adolescents with SS in terms of comorbid psychopathologies, social cognition skills, quality of life, self-esteem and coping strategies. However, the cognitive functioning of adolescents with TS was found to be lower than both of the adolescents with SS and community samples. The well-known neurocognitive status of these patients enables to support both the patients and their families in many areas such as quality of life, need of education, career planning, treatment compliance, prevention of risky behaviours and family relationships. Conducting this evaluation at the time of diagnosis is vital in terms of monitoring the neurocognitive effects of disease and providing the required reinforcement. It is of great importance to illuminate the psychosocial and neurodevelopmental effects of TS, which is a chronic disease that requires lifelong follow-up and treatment. Other prospective studies including large numbers of patients are needed to take necessary measures and to provide support for children and young people with TS.

Ethics

Ethics Committee Approval: The study were approved by the Hacettepe University of Local Ethics Committee (2018/08-GO 17/583-10).

Conflict of Interest: No conflict of interest was declared by the authors.

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