

Psychological well-being in women with Turner syndrome: Somatic and social correlates

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Our aim was to examine possible somatic and social correlates to psychological well-being in adult women with Turner Syndrome (TS), including hormone replacement treatment. Sixty-three women with a diagnosis of TS (mean age, 31.5 years) participated in a cross-sectional study, using interview data, ratings on the Psychological General Well-being (PGWB) Index, and data from medical examinations and medical records. Statistical analysis was performed by bivariate and multivariate analyses. Lack of sex hormones during adult life and the presence of hearing impairment were related to lower psychological well-being, as were higher age at diagnosis, higher age at menarche or induced bleeding, higher chronological age and retrospectively reported difficulties with school subjects. Age at diagnosis and difficulties with school subjects explained 25% of the variation in psychological well-being. This study has identified some correlates to psychological well-being in women with TS, which are important when considering the clinical management of adult women with TS.

Key words: Turner syndrome, psychological well-being, correlates, hormonal replacement treatment

INTRODUCTION

Turner syndrome (TS) is a sex chromosome disorder that occurs in about 1 in 2500 live female births¹. TS is caused by a total or partial absence or structurally abnormal second X chromosome, in all or a proportion of cells. The main features of TS, present in more than 90% of affected individuals, are short stature and dysgenesis of the gonads. Gonadal dysgenesis leads to sex hormone deficiency, incomplete pubertal development and impaired fertility. Changes in external appearance (stigmata), such as webbing of the neck, micrognathia (small jaw), shield thorax (broad chest) and cubitus valgus (wide angle of the elbows) occur to varying degrees. Malformations of organs (heart and urinary system) may occur, and some medical problems are more common than in the general population. A Swedish study comparing women with TS with an

age and gender matched control group, found hypertension to be more common in TS group (22 vs. 3%) as well as cardiovascular abnormalities (17 vs. 0.5%), while there was no difference in presence of hypothyreosis². A Danish study calculated risk ratio of medical diagnoses in TS compared to the national registry of patients, and for severe medical conditions found increased risk for hypothyreosis (RR 5.80), hypertension (RR 2.91) and congenital malformation of the heart (RR 13.35) in TS³. The majority of women with TS have a progressive sensory neural hearing impairment that may result in significant hearing impairment prominently in adult life⁴. Cognitive functioning in TS is often characterized by difficulties with spatial ability and arithmetic, while verbal intelligence is usually normal⁵. Non-verbal learning difficulties are common⁶.

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The diagnosis is usually made in childhood or early adolescence⁷, although some individuals are diagnosed in adult life. Modern medical treatment consists primarily of hormonal therapy, which aims to reduce the consequences of TS by allowing more normal development as well as an improved quality of life. Pubertal development is induced by estrogen treatment. Growth-promoting treatment with oxandrolone and growth hormone increases final height. Hormone replacement therapy (HRT) with estrogen and gestagen in adult life is recommended⁴.

Women with TS have been described as being at risk of moderate psychological and social difficulties, although there is a considerable variability and not all individuals have these difficulties⁸. In one study, women with TS reported a more negative self-concept than the normative group, and half of the sample had sought professional help because of mental health concerns⁹. Other studies have found impaired psychological well-being in women with TS, although general satisfaction with life was at the same level as in the comparison group¹⁰. Women with TS have also reported a health-related quality of life within the normal range¹¹. Difficulties in the area of social relationships have been repeatedly reported, and fewer women with TS have partner relationships than do women in the general population^{9,12}. In several recent studies, girls and adolescents with TS are described as being at risk of moderate behavioral problems and social difficulties^{13,14}. These findings on psychological and social functioning in TS raise questions about possible risk factors and how adaptation to TS can be facilitated. What are the psychological implications of different treatment regimens? Are the clinical manifestations of TS related to psychological well-being? Empirically based information on these issues could contribute to more appropriately focused healthcare. Indeed, the need for more specialized healthcare for adult women with TS has recently been acknowledged¹⁵.

There are several issues of clinical importance where few data are available. These include the long-term psychological consequences of the management of pubertal induction and of HRT in adults, which is necessary for almost all females with TS. Beneficial effects of estrogen treatment in girls and adolescents with TS on self-concept and memory have been reported^{16,17}. How-

ever, there has been some debate concerning the timing of pubertal induction. Puberty is usually induced at a relatively late age compared with normal puberty, as early estrogen treatment reduces final height. One study reported that girls with TS suffered from considerable psychological distress prior to pubertal induction that was considerably delayed, while their status improved following pubertal induction¹⁸. In adult life, up to 25% of women with TS may not get the HRT that is recommended to continue following pubertal induction¹⁹. In a survey of women with TS, those without HRT had a more negative body image than those receiving HRT⁹. Another question concerns the consequences of short stature in adulthood, as one of the reasons for growth-promoting treatment in TS is to reduce the negative effects of short stature on quality of life. In a few recent studies, height was found not to be related to psychological functioning in women with TS^{9,10}.

Methods for measuring the possible importance of the visible clinical manifestations of TS differ between studies. One study of women with gonadal dysgenesis (90% with a diagnosis of TS) found that the degree of stigmata (cubitus valgus and webbed neck) was not related to social functioning²⁰. Studies on girls with TS have found that behavior and social problems were not related to height¹⁴, but also that height was related to social functioning but not to behavior problems¹³. A study using an extensive stigmata index found no relationship between the degree of stigmata and most measures of psychological and social functioning¹⁴. In conclusion, therefore, the few studies performed on the effects of the various manifestations of TS on aspects of psychological functioning have produced discrepant results.

The present study is part of an ongoing project (Göteborg Turner project) with coordinated medical and psychological examinations of women with TS. We have previously reported that the majority of women with TS in this project have been given standard medical treatment²¹, and have not differed with regard to psychological well-being and self-rated health compared with normative data, with the exception that they reported more social difficulties¹². Furthermore, it was found that both higher age and lower self-rated health were related to lower psychological well-

being¹². A relationship between self-rated health and psychological well-being has been reported previously⁹, thus encouraging further study into these issues. The present study set out to examine correlates to psychological well-being in women with TS, and their possible clinical implications. Detailed somatic and psychological data were collected in co-ordinated examinations, specifically examining somatic and social status, including age at diagnosis, pubertal development, and adult HRT, as were retrospective data on social and educational functioning.

METHOD

Subjects

This cross-sectional study included women above 18 years of age with a diagnosis of TS. The study is part of an ongoing multidisciplinary Swedish nationwide project, which is aiming to provide recommendations for optimal care of individuals with TS. Recruitment of participants was made with the intention to invite all known adult women with TS living in Western Sweden. The two genetic laboratories in the area identified 175 women aged > 16 years with TS in the total population of 604,500 in Western Sweden (one TS per 3500). It is known that some of the missing women had moved out of the area, but some not diagnosed at the genetic laboratories in the area had moved in. For ethical reasons it was not possible to recruit participants from via the genetic laboratories. Participants were recruited via referral from their doctors, and by an advertisement in the National Turner patient organization newsletter. Eighty-four women were invited to participate. Of these, 66 women agreed to participate in the multidisciplinary project, while 18 declined due to lack of time or interest. The final sample consisted of 63 women with a mean age of 31.5 years (range, 18–59 years). Two women were too physically ill to participate, and one woman was excluded due to mental retardation.

The diagnosis of TS was confirmed by leucocyte karyotyping. Forty women had a 45,X karyotype, 11 had a 45,X/46,XX karyotype and 12 had a karyotype including structural abnormalities. Medical data compared with normative data are presented elsewhere²¹. The study was approved by the Ethic's committee of Göteborg University. All participants gave their informed consent.

Procedures

The psychological and medical examinations were integral parts of the multidisciplinary study, in which the participants underwent 2 days of psychological and medical examinations. The results presented here comprise one part of a larger investigation. Each participant took part in a semi-structured interview with a trained psychologist and completed self-rating scales, of which the Psychological General Well-being (PGWB) index²² is reported in this study. Each participant also underwent blood sampling and medical examinations.

Measurements

Psychological well-being: The PGWB index was constructed to measure personal affective states reflecting well-being or distress²². The scale consists of 22 items formed as questions, where responses are given on a six-grade scale. The items are summarized into an overall psychological general well-being score (total PGWB score) and may also be divided into subscales covering the states of anxiety, depressed mood, positive well-being, self-control, general health and vitality. A low score represents lower well-being. In this study, the total PGWB score was used as a measure of psychological well-being. For the total PGWB score, the internal consistency (Crombach's Alpha Coefficient) was 0.94 for the original sample²² and 0.95 for the Swedish version²³. Internal consistency for the present TS sample was 0.95. The PGWB has been tested for validity, and been found to be highly correlated with self-reported needs and utilization of mental health services, with scales of mental health distress, and with interviewers ratings of depression. The PGWB index also discriminated between mental health clients and community subjects²². The PGWB index has been used in studies of several clinical groups; for example, patients with upper gastrointestinal disease²⁴ and with postmenopausal symptoms²⁵.

Social functioning: An interview was conducted following a standardized protocol constructed for the study. For the purpose of this study, questions were included that covered issues of the subject's present life situation: whether the participant had a stable partner relationship (married or cohabiting) (yes/no), was employed or studying vs. unemployed or on sickness pension, and

had at least one close friend (yes/no). Retrospective data about childhood and adolescence were also requested: whether the participant had experienced difficulties with subjects in school (yes/no), had experienced teasing (yes/no), and had at least one close friend (yes/no). A self-rating scale was used to measure if the individual had experienced negative life events during the past year. That is, if any of the following events had happened and were rated by the participant to have a strong negative effect (yes/no): sickness, sickness of close relative, death in the family, separation, moving house, change of occupation, unemployment, insecurity in working life, economic problems or legal problems.

Somatic status and medical treatment: Medical examination included blood sampling, and analysis of thyroid-stimulating hormone (TSH) and serum free thyroxin (T_4). If earlier karyotyping was based on less than 30 metaphases, blood sampling for a repeat karyotype was also performed. Body mass index (BMI) was calculated as body weight divided by height squared (kg/m^2). According to the World Health Organization, a BMI of 19–24 kg/m^2 represents a normal weight, a BMI of 25–29 kg/m^2 represents slightly overweight and a BMI above 30 kg/m^2 represents overweight. Systolic and diastolic blood pressures were measured. An electrocardiogram and echocardiography were performed. For details concerning medical examinations and measurement see²¹. The physician rated the presence (as 0 or 1) of four common stigmata of TS: webbing of the neck, micrognathia, shield thorax and cubitus valgus. The scores were then combined into a stigmata index (range, 0–4). The presence of cardiovascular abnormalities, hypothyreosis and hypertension was recorded from the participant's history and the medical examinations. The presence of impaired hearing was recorded from the medical interview. Data on age at diagnosis (age at karyotyping) were obtained from medical records, as were details on childhood and adolescent hormonal therapy and HRT throughout adult life. Those women who had not received adult HRT because they had sufficient endogenous sex-hormone levels to attain spontaneous pubertal development and following bleedings were included in the same category as those with HRT. The physician rated pubertal breast development according to Tanner²⁶. The participants were divided into two groups: those with incomplete pubertal de-

velopment (Tanner stage I–III) and those with complete pubertal development (Tanner stage IV–V). Data on whether the participant had had spontaneous bleeding or bleeding induced by hormonal treatment, and age at first vaginal bleeding (spontaneous or induced by estrogen) was obtained from medical records.

Statistical methods

Distributions of continuous variables are given as means, standard deviations (SD) and ranges. Spearman's rank correlation coefficient was used for correlation analysis between continuous variables and the dependent variable. The Mann–Whitney non-parametric *U*-test was used for comparison between the two levels of each dichotomous variable regarding the dependent variable. All significance tests were two-tailed and conducted at the 5% significance level. Those independent variables attaining a *p*-value of < 0.10 in the preceding bivariate analysis were included in a forward stepwise regression analysis. Verification of the independent predictors from the stepwise regression model with non-parametric methods was performed using Spearman's partial correlation analysis and Mantel's technique of pooling²⁷ applied to the Mann–Whitney *U*-test.

RESULTS

Somatic variables in relation to psychological well-being

Thirteen participants (21%) had not received HRT continuously in adult life. Five of these, however, did not meet the indications for HRT due to sufficient gonadal functioning, as indicated by spontaneous pubertal development and following bleedings. In order to investigate the importance of sex hormone (especially estrogen) deficiency in adult women with TS, these five participants were combined with the remaining participants who had received continuous HRT in adult life, into a 'sufficient sex hormone' group. The remaining eight participants without HRT, who were considered to have a lack of sex hormones, had a lower psychological well-being ($p=0.006$) than those participants who were considered to have had sufficient levels of sex hormones in adult life. (The two subgroups with sufficient sex hormones in adult life had similar ratings of psychological well-being (data not shown)). The reasons

for not having received HRT in adult life were diagnosis of TS later in adult life ($n=2$), diagnosis of TS later in adult life in combination with insufficient medical follow-up ($n=4$), insufficient medical follow-up ($n=1$) and lack of compliance ($n=1$) (Tables 1 and 2).

Hearing impairment, reported by 26 of the participants, was related to a lower psychological well-being ($p=0.029$). Higher age at diagnosis was related to a lower psychological well-being ($p=0.001$), as was higher age in itself ($p=0.04$) (Tables 1 and 2).

Higher age at menarche/induced bleeding was related to a lower psychological well-being ($p=0.040$). This relationship did not remain, however, when two women with very late induced bleeding (above age 50) were excluded. Psychological well-being did not differ between the participants who had received induced bleeding compared with those with spontaneous bleeding (menarche), or between those with complete pubertal development compared with those with incomplete pubertal development. The remaining somatic variables included in this study (BMI, height, presence of stigmata, cardiovascular abnormalities, hypertension and hypothyreosis) were not related to psychological well-being (Tables 1 and 2).

Social variables in relation to psychological well-being

The participants who had experienced difficulties with school subjects (retrospective reports) ($n=48$) now had a lower psychological well-being ($p=0.004$). The majority ($n=39$) had experienced difficulties with mathematics, while the remaining nine participants most commonly mentioned difficulties with language and sports. The participants' present social situation (having a stable partner relationship or not, working or studying vs. unemployed or on sickness pension) was not related to their psychological well-being. Whether participants had experienced teasing and whether they had had close friends during childhood was not related to their present psychological well-being (Table 3).

Multivariate analyses

Using the results of the bivariate analyses, multivariate analyses were conducted to investigate to what degree these variables could explain the variation in psychological

Table 1 Somatic variables (measured as continuous data) in relation to psychological well-being (total PGWB score) in women with TS. A high PGWB score reflects more positive well-being

Variable	Mean (SD) (range)	Correlation coefficient	p-value
Age, years	31.5 (11.5) (18–59)	–0.26	0.040
Age at diagnosis, years	11.6 (11.1) (0–58)	–0.41	0.001
Age at menarche/induced bleeding ^a	17.2 (7.0) (12–54)	–0.26	0.04
Age at menarche/induced bleeding (exclusive of 2 outliers) ^b	16.0 (2.2) (12–23)	–0.21	n.s., 0.11
Body mass index	25.6 (4.5) (18.7–35.9)	–0.02	n.s., 0.86
Height, cm	151 (6.1) (135–164)	0.01	n.s., 0.95
Stigmata index ^c	2.05 (0.80) (0–3)	0.03	n.s., 0.82

^a $n=62$, ^b $n=60$, ^c $n=61$

Table 2 Somatic variables (measured as dichotomous data) in relation to psychological well-being (total PGWB score) in women with TS. A high PGWB score reflects more positive well-being

Variable	n (%)	Total PGWB score Mean (SD)	p-value
Cardiovascular abnormalities			
yes	5 (8%)	102.0 (14.5)	n.s., 0.63
no	58 (92%)	102.8 (17.4)	
HRT/endogenous estrogen			
yes	55 (87%)	104.9 (16.3)	0.006
no	8 (13%)	87.9 (15.2)	
Hypertension			
yes	14 (22%)	97.0 (15.3)	n.s., 0.06
no	49 (78%)	104.3 (17.3)	
Hypothyreosis			
yes	12 (19%)	96.2 (22.3)	n.s., 0.30
no	51 (81%)	104.2 (15.5)	
Impaired hearing			
yes	26 (41%)	97.8 (16.8)	0.029
no	37 (59%)	106.1 (16.6)	
Incomplete pubertal breast development ^a			
yes	20 (32%)	108.0 (13.8)	n.s., 0.13
no	42 (68%)	100.0 (18.1)	
Induced bleeding			
yes	57 (90%)	103.5 (16.2)	n.s., 0.57
no	6 (10%)	94.8 (24.5)	

^a $n=62$

well-being, that is the total PGWB score. Included in the multivariate analyses as possible predictors were the variables related to the total PGWB score with a p -value of < 0.10 : age, age at diagnosis, age at spontaneous or induced bleeding, hypertension, impaired hearing, lack of estrogen during adulthood and school subject difficulties. Independent predictors of the total PGWB score were age at diagnosis and school

Table 3 Social variables in relation to psychological well-being (total PGWB score) in women with TS. A high PGWB score reflects more positive well-being

Variable	n (%)	Total PGWB score Mean (SD)	p-value
Close friend			
yes	55 (87%)	102.2 (17.5)	n.s., 0.70
no	8 (13%)	106.0 (13.9)	
Negative life events ^a			
yes	20 (34%)	97.8 (18.9)	n.s.
no	39 (66%)	105.1 (16.4)	
Partner relationship			
yes	18 (29%)	100.8 (17.0)	n.s., 0.53
no	45 (71%)	103.5 (17.2)	
Unemployed/sickness pension			
yes	8 (13%)	94.1 (16.5)	n.s., 0.10
no	55 (87%)	103.9 (3.3)	
Childhood teasing			
yes	30 (48%)	102.4 (18.6)	n.s., 0.90
no	32 (52%)	102.6 (15.9)	
Close friend in childhood ^b			
yes	44 (73%)	102.2 (17.3)	n.s., 0.73
no	16 (27%)	102.7 (12.2)	
School subject difficulties ^c			
yes	48 (77%)	99.3 (17.7)	0.004
no	14 (23%)	113.4 (8.5)	

^an = 59, ^bn = 60, ^cn = 62

Table 4 Model predicting psychological well-being (total PGWB) in women with TS

Model	Beta coefficients (SE)	Adjusted p-value
Age at diagnosis	-0.59 (0.18)	0.001
School subject difficulties Yes = 1, No = 0	-10.62 (4.66)	0.026

Model R² = 0.26, Residual SD = 14.7

subject difficulties. These variables explained 26% of the variation in psychological well-being. Multivariate non-parametric analyses verified that age at diagnosis and school subject difficulties were independent predictors with adjusted *p*-values of 0.003 and 0.032 (Table 4).

DISCUSSION

The aim of this study was to investigate correlates of psychological well-being in adult women with TS, including both somatic and social variables. Current and retrospective data were used in order to obtain a life-span perspective of the disorder, and psychological, social and medical data were collected in co-ordinated examinations.

The large majority of females with TS have gonadal dysgenesis, leading to sex

hormone deficiency and primarily estrogen deficiency, and therefore meet the indications for HRT in adult life. It has been reported that up to 25% of adult women with TS do not receive HRT¹⁹. In the present study, about 20% had not received HRT continuously in adult life. After excluding those not meeting the indications for HRT, 13% still did not receive HRT as needed, and these women with sex hormone deficiency reported a lower psychological well-being. This result indicates a positive psychological effect of HRT in TS. HRT in women with TS contributes to physical health. For example, Landin-Wilhelmsen *et al.*²¹ found osteoporosis only among those women without continuous HRT in adult life. In fact, Gravholt *et al.*³ have argued that the profile of medical problems in females with TS, including increased risk of fractures, heart disease, hypertension, vascular insults of the brain, overweight, diabetes and other endocrine diseases, is similar to that of postmenopausal women, although the problems in TS occur at an earlier age. Similar positive effects of HRT on physical health and quality of life have been reported in postmenopausal women²⁵, when the same psychological measurement was used as in the present study. Normal levels of sex hormone may contribute to a better well-being secondary to the effects on physical health. There may also be a direct effect of gonadal hormones on the CNS, contributing to a better psychological well-being. It is not possible to separate these mechanisms in the present clinical study.

Related to the importance of HRT is the induction of pubertal development that is required in the majority of females with TS. There is some discussion in the literature about the timing of pubertal development. On the one hand, pubertal development at a time consistent with their peers is thought to be preferable not least for psychological reasons. The median age for menarche in the US as well as in Sweden is about 13 years. On the other hand, however, early estrogen treatment reduces final height in these individuals who are already of short stature. In this study, there was a slight relationship between having menarche or induced bleeding at an earlier age and a better well-being. This relation did not remain, however, when those women with induced bleeding very late in life were excluded. Our data therefore support the importance of pubertal induction during adolescence, while the age

variation within this age range was not related to subsequent well-being. It remains to be investigated whether differences in pubertal development (timing, completion) is related to other aspects of gender development such as gender identity, and to sexuality, areas where conflicting results have been reported^{9,28}.

The visible deviations in appearance that may affect females with TS may be regarded as risk factors for adaptation. The visible physical deviations included in this study (height, overweight, common TS stigmata and incomplete pubertal development) were not related to psychological well-being. This result is similar to the few previous studies where variation in height and weight were not related to psychological well-being^{9,10}. These deviations in appearance, however, may have been stressors earlier in life, which was not investigated in this study. Whether empirical data support the contention that short stature *per se* should be considered a risk factor for adaptation has been discussed in the literature^{29,30}. Short people may be treated according to the age that they appear, rather than the age they actually are, following a mechanism called juvenilization³¹. This may have adverse effects on psychological and social development, for example by contributing to immaturity in behavior and self-image.

By definition, TS is present from birth, although the clinical manifestations may differ in expression throughout life. The diagnosis may therefore be made in girls and women of very different ages. In this study, earlier diagnosis was related to better well-being. Correct diagnosis is, of course, necessary in order to provide appropriate treatment. The length of time from diagnosis to study may also be important in the adaptation process; in a study of individuals with different chronic illnesses, those diagnosed most recently had a poorer mental health³². The women in the present study who were diagnosed with TS later in life may still be suffering from lack of treatment and also from the crisis of receiving the diagnosis. It should also be considered that, in a disorder such as TS, age at diagnosis is not always identical to age of disclosure.

Individuals with TS are at increased risk of learning disabilities, primarily in nonverbal areas, and may not receive the necessary extra educational support⁶. The participants in the present study who retrospectively reported difficulties with school subjects

now had a lower psychological well-being. Although retrospective data should be interpreted with caution, this result supports previous recommendations that the school situation for girls and adolescence with TS should be evaluated throughout their education. Interestingly, in the study by Rovet⁶ using current data, achievement and self-concept were not related.

The presence of hearing impairment might be one aspect of importance for women with TS, as the participants in this study who had impaired hearing also reported a lower psychological well-being. This result in itself is not surprising. Negative psychological and social consequences in non TS women with hearing loss has been described³³. However, although it has long been recognized that TS is associated with hearing loss, primarily in adult life, the psychological significance of hearing impairment in this group has not previously been investigated to our knowledge. Further research should include more detailed data on hearing function, and also evaluate clinical management.

We have previously reported that higher chronological age was slightly related to lower psychological well-being in women with TS¹², a correlation not found in the reference group for these age ranges²⁴. It follows from the cross-sectional design of this study that a difference between age groups could be due to a cohort effect, because the knowledge and treatment of the disorder that is standard today was not available earlier. In a previous Swedish study of middle-aged and older women with TS, most women were satisfied with their lives³⁴. The results are not quite comparable, however, as the study by Sylvén *et al.* was based on interviews and did not use a standardized measure of psychological functioning. In the present study, relatively few older women participated. In a study of all individuals diagnosed with TS in Denmark between 1913 and 1993, few individuals with TS were found in the birth cohorts from 1910–1940s³. The authors argue that before 1950 the knowledge of the disorder was low, and many females with TS probably died due to TS complications before a diagnosis was made.

Some limitations of the present study should be considered. The correlation design where most measures were performed at the same time, limits the possibilities of drawing conclusions about causal relationships. The

use of retrospective data on social functioning and school performance may insert a bias, as the level of problems reported may be influenced by later events and by the present status of the participant. The sensitivity of the scale used to measure psychological well-being could also be questioned following the many negative findings. On the other hand the scale has previously been reported to discriminate between individuals in other clinical groups on relevant variables. A common shortcoming in clinical studies of rare disorders is that relatively small sample sizes leads to low power for the statistical analyses. In this study, there were several absolute differences between subgroups indicating possible relationships of the variable and psychological well-being, that were not statistically significant. Further studies with larger sample sizes would be needed to further test the importance of these correlates.

In summary, this study of a broad sample of women with TS has identified the following possible important correlates or risk factors for psychological well-being which are important to consider in clinical practice: sex hormone deficiency, hearing ability, age at diagnosis, and presence of school difficulties. The message is encouraging and challenging for the healthcare management of females with TS throughout their lives. The results suggest that it is important to make an early diagnosis, to follow the situation in school and provide support if educational and other school subject difficulties occur, to provide HRT not only for pubertal development but also in adult life if indicated, and to evaluate hearing and provide hearing aid when needed. Other aspects of TS, such as visible deviations, appear to be of less importance. This study also revealed that the group of women with TS is heterogeneous regarding most of these variables, and that an individual approach is necessary. More specialized healthcare provided by multidisciplinary teams could facilitate the life situation for females with TS.

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