Self-perception, Behavior and Social functioning in Swedish Girls with Turner Syndrome: A population-based study

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Wide Boman, U. Albertsson Wikland, K. and Möller, A. Self-perception, Behavior and Social functioning in Swedish Girls with Turner Syndrome: A Population Based Study. Göteborg Psychological Reports, 2000, 30, No. 5. The objective of this study was to describe self-perception, behavior and social functioning in all known girls with Turner syndrome (TS), 7-16 years of age, in the region of Western Sweden of 1.5 million inhabitants, 400,000 children and adolescents. Thirty-seven girls with TS (mean age, 12.6 years; range, 7-16 years) participated in a cross-sectional study using the self-perception questionnaire I Think I Am, the Child Behavior Checklist, and a semi-structured interview. The girls with TS reported self-perception at the same level as the normative sample, while their parents rated them as having significantly more behavioral problems and lower social competence than did parents in a normative sample. Half of the girls with TS reported peer-related problems. Both negative and positive consequences of TS were described by those girls aware of their diagnosis. The results from this population-based sample support previous studies from other countries that girls with TS are at higher risk of having social and behavioral problems than the general population, although not at the level of clinically referred children. The use of multiple informants in assessing the psychological functioning in girls with TS is discussed.

Keywords: Behavior problems, self-perception, Turner syndrome.

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Turner syndrome (TS) is a sex-chromosome disorder that occurs in 1 in 2500 female births. TS is caused by the total or partial absence of one X chromosome in some or all cells. The principal features are short stature and dysfunctional gonads, resulting in a lack of sex hormones, incomplete pubertal development and impaired fertility. Other medical complications and minor abnormalities in appearance occur to varying degrees (Hall & Gilchrist, 1990). Medical treatment during childhood includes hormonal treatment with growth hormone (GH), oxandrolone and estrogen to stimulate growth and induce pubertal development (Rosenfeld et al., 1994). Deficits in non-verbal and spatial processing skills have been reported (Romans, Roeltgen, Kushner & Ross, 1997; Temple & Marriott, 1998).

An increased risk of behavioral and social problems in girls with TS, as measured by parental rating on the Child Behavior Checklist (Achenbach, 1991), are most commonly reported in the social, immaturity and attentional / hyperactivity dimensions (Swillen et al., 1993; Rovet & Ireland, 1994; McCauley, Ross, Kushner & Cutler, 1995; Lagrou et al., 1998; Skuse, Elgar & Morris, 1999). Thought problems (Rovet & Ireland, 1994; Skuse, Elgar & Morris, 1999), aggressive and delinquent behavior (McCauley et al., 1995; Skuse, Elgar & Morris, 1999) and withdrawal (Lagrou et al., 1998) have also been described. Girls with TS have rated themselves as having social problems (Swillen et al., 1993; Lagrou et al., 1998; Skuse, Elgar & Morris, 1999), and have reported being dissatisfied with their physical appearance (McCauley et al., 1995).

An important limitation of many studies of TS is the unknown representativeness of the samples. In many countries, participants may be recruited from potentially biased sources, such as parent support groups or through their participation in medical interventions. In order to obtain a population-based sample, the present study aims to include all known cases in a geographic area.

Knowledge of such rare clinical conditions as TS is based on studies from different countries. The importance of cross-cultural validation of results is clear. Recently, data on behavior and social functioning in girls with TS have been reported from Belgium (Swillen et al., 1993; Lagrou et al., 1998), Canada (Rovet & Ireland, 1994), the USA (McCauley et al., 1995), and the UK (Skuse, Percy & Stevenson, 1994; Skuse, Elgar & Morris, 1999).

The purpose of the present study was to describe self-perception, behavior and social functioning of all identified girls with TS, aged 7-16 years, in the Region of Western Sweden. The importance of obtaining the perspective of the child herself was considered in the design of the study (Rosenbaum & Saigal, 1996).

Method

Subjects

Participants were recruited from the Göteborg Pediatric Growth Research Centre at Queen Silvia Children’s Hospital Sahlgrenska University Hospital, Göteborg, between January 1995 and June 1997. All identified girls with TS between 7 and 16
years of age in Western Sweden (including the areas connected to the hospitals of Jönköping, Halmstad, Göteborg, Borås, Mölndal, Uddevalla, Vänersborg, Lidköping, Skövde and Karlstad, with 1.5 million inhabitants, 400,000 children and adolescents) were invited to participate, in order to obtain a representative sample. Forty-one girls were approached, 37 of them agreed to participate while three girls did not participate because their parents refused permission. One girl attending a special school for children with mental retardation was not included because it was considered that she would not be able to complete the self-rating scale.

The mean age of the 37 participants was 12.6 years (SD 2.9 years). Diagnosis of TS was confirmed by leukocyte karyotyping. Fifteen girls (40%) were diagnosed during their first year of age, 8 girls (22%) between 1 and 7 years of age, and 14 girls (38%) between 8 and 13 years of age. At the time of the examination, 3 girls (8%) had not received hormone treatment, 11 (30%) had received GH treatment, 9 (24%) had received GH and oxandrolone, 13 (35%) had received GH, oxandrolone and estrogen, and 1 girl (3%) had received GH and oxandrolone. Pubertal status, breast stage according to Tanner (Tanner, 1982) was prepubertal (stage 1) in 21 girls (57%). Twelve girls (32%) had started pubertal development (stage 2-3), and 4 girls (11%) had almost completed or completed pubertal development (stage 4-5). The educational level of the families did not differ from that of the general Swedish population.

**Procedure**

Each girl was interviewed for about 15 minutes by the same psychologist (UWB), and then completed the I Think I Am scale (Ouvinen-Birgerstam, 1985). At the same time, but in another room, the primary caregiver completed the Child Behavior Checklist (CBCL) (Achenbach, 1991), and a questionnaire about educational background. A medial examination was made at the same visit to the clinic.

*The semi-structured interview.* The semi-structured interview was conducted following a standardized protocol, including questions about social activities, peer relationships, repeated teasing and school performance. There were also questions about their reaction to receiving the TS diagnosis, contact with other girls with TS and with the patient support organization. Questions related to TS were asked only to those girls who were already aware of their diagnosis (n=29). Questions required either a choice of alternatives or were open-ended, in which case the response was written down verbatim.

*The I think I am scale.* The Swedish self-perception scale I Think I Am has one longer version for older children (grade 4-9, age 10-16 years) and one shorter version for younger children (grade 1-3, age 7-9 years). Reliability and validity of the scale are well-documented (Ouvinen-Birgerstam, 1985). The long version consists of 72 statements. The girl reads each statement and gives her written answer whether it is true for her, choosing between the four alternatives. The short version consists of 32 statements (read by the psychologist to the child) to which the girl is asked to reply yes or no. The scores are summarized into a total self-perception score, and may be divided into five subscales of physical appearance, skills and talents, psychological well-being, relationships with family and relationships with others. A high score
reflects favourable self-perception. The Swedish normative raw data is transformed into a stanine scale with normal distribution and a range of 1 to 9 (mean=5, SD=2), the stanine transformation made specific for sex and school grade. Stanine scores of subjects of different ages are thereby directly comparable, and differences between ages in raw-score distribution are held constant. Normative stanine scores are available for the total self-perception score and the subscales for the long version of the scale, while for the short version they are available only for the total score (Ouvinen-Birgerstam, 1985). In this study, each girl in the sample was assigned a stanine score according to the normative sample (n=778) representing her sex and grade.

The Child Behavior Checklist (CBCL). The CBCL questionnaire gives a global picture of the competencies and emotional and behavioral problems of the girl, as assessed by the parent (Achenbach, 1991). Reliability and validity of the scale are well-documented (Achenbach, 1991). The competence scale provides a total score divided in the dimensions activities, social relationships and school performance. A high competence score reflects good competence. The problem scale provides a total score and groups the problems into two broad categories: internalizing (covering somatic complaints, withdrawn and anxious/depressed behavior), and externalizing (covering aggressive and delinquent behavior). The presence of attention, social and thought problems is also investigated. Higher problem scores reflect a greater degree of problematic behavior. The Swedish version of the CBCL was used in the normative Swedish study (Larsson & Frisk, 1999) and has been used in studies of Swedish children with other chronic illnesses (Engström, 1992). The TS group was compared with a proportional female sample (n=165) from the Swedish normative study, with similar distribution regarding age (mean=11.9 years; range, 7-16 years) and socio-economic status.

Statistical methods. Students t-test was used to compare I Think I Am scores with normative data, and the Mann-Whitney test was used to compare CBCL scores with normative data. Spearman's rank correlation coefficient ($r_s$) was used to examine the relationship between age, I Think I Am scores and CBCL scores. Descriptive statistics are given as means and standard deviations (SD), in transformed stanine scores for the I Think I Am, and as raw scores for the CBCL.

Ethics. The study was approved by the Local Ethical at Göteborg University. Informed consent was obtained from all participants and/or parents.

Results

Self-perception and CBCL data: comparison with normative data

The girls with TS did not differ in the I Think I Am total self-perception score (mean=4.92, SD 2.1, n=36; data missing for one participant due to an administrative error) compared to the normative sample (mean=5.00, SD 2.0, n=778). In this analysis, the power to detect a difference of 1 stanine score on the total self-perception score was 0.86 at the p<0.05 level. There were no significant differences between the normative sample and the TS group on the subscales covering physical
appearance, skills and talents, psychological well-being, relationships with family and relationships with others, as analysed in the 28 older girls (age 10-16 years) who completed the long version of the scale (see Table 1).

Table 1
I think I am self-perception dimension scores for girls with TS and the normative group, age 10-16 years. Scores are given as transformed stanine mean score and standard deviation (SD). High scores represent a positive self-perception. No significant differences were found

<table>
<thead>
<tr>
<th></th>
<th>TS group (n=28) Mean (SD)</th>
<th>Normative group (n=597) Mean (SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Physical appearance</td>
<td>4.6 (2.1)</td>
<td>5.0 (2.0)</td>
</tr>
<tr>
<td>Skills and talents</td>
<td>5.2 (2.1)</td>
<td>5.0 (2.0)</td>
</tr>
<tr>
<td>Psychological well-being</td>
<td>4.9 (1.8)</td>
<td>5.0 (2.0)</td>
</tr>
<tr>
<td>Relationships with family</td>
<td>5.4 (2.2)</td>
<td>5.0 (2.0)</td>
</tr>
<tr>
<td>Relationships with others</td>
<td>5.0 (2.2)</td>
<td>5.0 (2.0)</td>
</tr>
</tbody>
</table>

On the CBCL, the girls with TS received higher (p=0.006) behavioral problem scores (mean=22.97, SD 16.63, n=34; data missing for three girls due to an administrative error) than did the girls in the normative sample (mean=15.02, SD 13.81, n=165) (see Table 2). The girls with TS were rated as having more problems with aggressive behavior (p=0.007) but not more delinquent behavior. The TS group scored higher on the combined externalizing dimension (p=0.030). On the subscales included in the internalizing dimension, the TS group was rated to be more withdrawn (p=0.035), while there was no difference between the groups concerning anxious/depressed behavior or somatic complaints. On the combined internalizing dimension, there was no significant difference between the TS group and the normative group. On the remaining subscales, the TS group was rated as having significantly more problems than the normative group: attention problems (p<0.001), social problems (p<0.001), and thought problems (p=0.011).
Table 2

*CBCL behavior problem scores for girls with TS and the normative group. Scores given as raw mean score and standard deviation (SD). High scores represents more problems.*

<table>
<thead>
<tr>
<th></th>
<th>TS group (n=34)</th>
<th>Normative group (n=165)</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Problem score</td>
<td>23.0 (17.6)</td>
<td>15.0 (13.8)</td>
<td>0.006</td>
</tr>
<tr>
<td>Aggressive Behavior</td>
<td>6.20 (4.79)</td>
<td>4.31 (4.44)</td>
<td>0.007</td>
</tr>
<tr>
<td>Anxious/Depressed</td>
<td>3.41 (4.21)</td>
<td>2.30 (2.91)</td>
<td>n.s.</td>
</tr>
<tr>
<td>Attention Problems</td>
<td>3.56 (3.64)</td>
<td>1.45 (2.05)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Delinquent Behavior</td>
<td>0.76 (1.35)</td>
<td>1.03 (1.59)</td>
<td>n.s.</td>
</tr>
<tr>
<td>Social Problems</td>
<td>2.38 (2.32)</td>
<td>0.84 (1.60)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Somatic Complaints</td>
<td>1.36 (1.80)</td>
<td>1.10 (1.48)</td>
<td>n.s.</td>
</tr>
<tr>
<td>Thought Problems</td>
<td>0.50 (0.96)</td>
<td>0.16 (0.61)</td>
<td>0.011</td>
</tr>
<tr>
<td>Withdrawn</td>
<td>2.32 (2.60)</td>
<td>1.32 (1.48)</td>
<td>0.035</td>
</tr>
<tr>
<td>Externalizing (aggressive, delinquent)</td>
<td>6.97 (5.68)</td>
<td>5.34 (5.65)</td>
<td>0.030</td>
</tr>
<tr>
<td>Internalizing (somatic, withdrawn, anxious/depressed)</td>
<td>6.91 (7.25)</td>
<td>4.66 (4.66)</td>
<td>n.s.</td>
</tr>
</tbody>
</table>

On the CBCL total competence scale, the girls with TS received lower scores than the normative sample (p<0.001), having poorer social relationships (p=0.019) and poorer school performance (p<0.001) (see Table 3).
Table 3

*Competence as measured on the CBCL for girls with TS and the normative group, given as raw mean score and standard deviation (SD). High scores represent better competence*

<table>
<thead>
<tr>
<th></th>
<th>TS group (n=34)</th>
<th>Normative group (n=165)</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Competence</td>
<td>15.0 (3.11)</td>
<td>17.2 (3.13)</td>
<td>0.001</td>
</tr>
<tr>
<td>Social relationships</td>
<td>5.84 (1.98)</td>
<td>6.75 (1.84)</td>
<td>0.019</td>
</tr>
<tr>
<td>Activities</td>
<td>4.78 (1.41)</td>
<td>5.28 (1.95)</td>
<td>n.s.</td>
</tr>
<tr>
<td>School performance</td>
<td>4.29 (0.94)</td>
<td>5.09 (0.76)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

*Self-perception and CBCL data in the TS group: relationship with age*

The total self-perception score was not related to age ($r_s$=-0.11, $p=0.52$), while a more negative self-perception on the physical appearance subscale, as analysed in the 28 older girls (age 10-16 years), was related to increasing age ($r_s$ = -0.54, $p=0.003$). The CBCL competence and behavioral problem scores were not related to age ($r_s$=0.17, $p=0.33$; $r_s$=-0.16, $p=0.38$).

*Relationship between self-perception and CBCL in the TS group*

A more negative total self-perception was related to a poorer school performance, as rated on the CBCL ($r_s$=0.62, $p<0.001$), and to more behavioral problems on the CBCL total problem score ($r_s$=-0.54, $p<0.01$).

*Social situation: results from the semi-structured interview*

In the semi-structured interview, 23 of the 37 girls (61%) reported that they participated in one or more scheduled activities each week, such as sports (e.g. horse riding, jujitsu, handball and golf) and playing musical instruments. Other common leisure activities were writing stories, reading and being with friends. About school performance, six girls (16%) said that mathematics was their favourite subject and 15 (41%) thought it was their worst subject. When asked about their friends, 19 (51%) girls had a best friend, 13 (35%) had no best friend but some close friends, and seven
girls (19%) had no close friends. Seventeen (49%) of the girls always socialized with classmates during school breaks, 16 (46%) did so sometimes, and two (6%) were always alone (data missing for two girls). Twenty girls (54%) reported that they had experienced repeated and distressing teasing by peers, and nine of these (24%) were currently teased. Some girls associated teasing with the features of TS, such as being short and chubby, while other girls could not tell why they were being teased.

**Experiences of diagnosis of TS: results from the semi-structured interview**

Answers are reported below from interviews with those 29 (78%) girls from the total sample who were aware of their diagnosis. Sixteen girls remembered their reactions when told about the diagnosis. Examples of reactions to being told about the diagnosis were: "It felt strange, like something very serious"; "I was afraid that I would become seriously ill"; and "It was hard to understand, but also good to know why I did not grow and develop like my peers". Nineteen girls reported that some classmates knew about their diagnosis. Twenty-five girls had met another girl with TS, and 20 of these had regular contact with another girl with TS. Twenty-two girls knew about the national TS patient support organization; 17 of these were members and 14 had taken part in its activities.

Most of the girls reported both negative and positive consequences of TS. Negative consequences included being short, future infertility, and the numerous medical interventions and measurements. Positive consequences mentioned by younger girls were "It is good to be more lithe"; "Being short makes it easier to play"; "I meet nice people". Older girls said for example: "I have learned to better understand and help other people with different problems"; "I don't judge people by their looks"; "I have a better knowledge of my body".

**Discussion**

In this population-based study, including the large majority of the subjects approached, the girls with TS reported a similar self-perception as a normative sample, while their parents rated them as having more behavioral problems, a lower social competence and a poorer school performance than a normative sample.

The finding that the girls with TS did not report a more negative self-perception, is, to some degree in contrast to the results of previous study in which the girls with TS reported a more negative self-perception than did the controls, in the areas of physical appearance and popularity (McCauley et al., 1995). Similar to this previous study, however, the older girls with TS in the present study rated their physical appearance more negatively than younger girls (McCauley et al., 1995). Similar age-dependent changes in self-perception have been observed in normative samples. However, the relation between age and negative physical appearance in the TS group in this study was found using transformed stanine scores, a transformation that holds
the effect of age constant in relation to the distribution of raw-scores in the reference group (see method section). This indicates that the general relationship between age and negative physical appearance may be emphasized in girls with TS. This may be due to the fact that the features of TS become more evident in adolescence, following the absence of a growth spurt and delayed pubertal development. Psychological developmental factors may also contribute by making older girls more capable of understanding the negative consequences of the condition, such as the future implications of impaired fertility.

Elevated scores for behavioral problem in girls with TS compared with controls, as found in this study, have been described previously. Also similar to previous studies, the problem scores were not related to age (Rovet & Ireland, 1994; McCauley et al., 1995). The average increase in the problem score in the present sample is similar to that reported for Swedish children with other chronic diseases (Engström, 1992), but not as high as the level found in children referred to a psychiatric clinic in Sweden (mean=39.1) (Larsson & Frisk, 1999). This moderate level of increased behavioral problem scores has been reported previously (McCauley et al., 1995), although one study reported that a considerable proportion of the study sample had behavior problem on a level of clinical referral (Skuse, Percy & Stevenson, 1994).

It has recently been argued that it is more accurate to interpret high CBCL problem scores as related to general psychopathology, rather than using the specific dimensions as more detailed indications of a specific psychiatric diagnosis (Hartman et al., 1999). On the other hand, similar findings on the dimension level could indicate the presence of a consistent TS phenotype, even if generic screening measures, such as the CBCL, may not accurately cover specific problems in clinical groups, such as girls with TS (Rovet & Ireland, 1994). In the present study, attention problems and social problems, including immaturity, were found, as have been reported commonly in other studies (Swillen et al., 1993; Rovet & Ireland, 1994; McCauley et al., 1995; Lagrou et al., 1998; Skuse, Elgar & Morris, 1999). The finding of increased thought problems in the present study has also been reported previously (Rovet & Ireland, 1994; Skuse, Elgar & Morris, 1999), as has aggressive behavior (McCauley et al., 1995; Skuse, Elgar & Morris, 1999) and withdrawal (Lagrou et al., 1998). The variation that still exists between studies could partly be due to sampling differences and differences regarding specific medical treatments. Hormonal treatment may have primary and secondary influence on psychological functioning. Differences in cultural values of behavior in children may also be of importance, although the CBCL has been shown to be robust between different cultures (Crijnen, Achenbach & Verhulst, 1997).

Both parental reporting and the girls’ answers in the interviews showed that several of the girls had social problems, such as experiencing teasing by peers and not having a close friend. Social difficulties in girls with TS have previously been reported on self-rating scales (Lagrou et al., 1998; Skuse, Elgar & Morris, 1999) as well as on parental reporting (Rovet & Ireland, 1994; McCauley et al., 1995).

The girls in this study had a poorer school performance, according to parental rating, compared with normative data, and the poorer school performance was related to a less favourable self-perception. The presence of non-verbal learning disabilities is increased among girls with TS, and could have a negative effect on self-perception. It
might also be that girls with a less favourable self-perception generally find it more
difficult to perform well in school and to handle possible learning disabilities.

It could be of interest to consider the differences between self- and parental
ratings in the present study. While the girls with TS reported on average normal self-
perception, their parents reported a higher frequency of behavioral problems. Evidently, the two measures of psychological functioning are not exactly comparable,
and they may differ in sensitivity. On the other hand, in a previous study of children
of short stature using the same methods for parental and self-ratings, it was also found
that parents rated their children’s well-being as lower than did the children
themselves (Erling, Wiklund & Albertsson-Wikland, 1994). The phenomenon of
different information being obtained from different informants could be interpreted as
a lack of validity. It might be more accurate, however, to assume that different
informants contribute different, but still valid, information (Achenbach, McConaughy
& Howell, 1987). The present study supports the interpretation of a limited self-
awareness in girls with TS, which may be seen as an adaptational mechanism. Also,
adults with chronic illnesses report less emotional distress compared with ratings
made by other informants (Faller, Lang & Schilling, 1995). The higher problem
scores reported by parents could, beside representing the problems of their daughters,
bé coloured by reactions to having a child with a chronic disorder. While discussing
these results, it should also be noted that those girls with TS who were rated as having
more problems by their parents also tended to report a less favourable self-perception.

To summarize, in this population-based study of girls with TS in Sweden, more
behavioral and social problems were reported compared with controls, although these
problems did not reach the level of problems found in children referred to a
psychiatric clinic. The girls themselves did not report impaired self-perception,
although older girls rated their physical appearance more negatively. These problems
should be considered when providing clinical care. The present study provides some
validation of the increased risk of behavior problems reported from studies of
potentially more biased samples.

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