

ORIGINAL ARTICLE

Health status in women with Turner syndrome: a questionnaire study on health status, education, work participation and aspects of sexual functioning

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Summary

Context Turner syndrome (TS) is a complex medical condition with specific cognitive and psychosocial characteristics and frequent medical morbidity. Few studies have investigated the influence this has on health status, education and ability to work.

Objective To explore health status, level of education, work participation, medical conditions, physical activity, satisfaction with life and aspects of sexual functioning in adult TS women and compare with a matched control group.

Design A questionnaire was sent to 168 adult women with TS >18 years registered in a database of Frambu Resource Centre for Rare Disorders and The TS Association in Norway. We assessed health status with Short Form 36, education with Norwegian Standard Classification of Education, and employment with The General Nordic Questionnaire. Life satisfaction was measured with LiSat-9, and questions on psychological strain during life phases were included.

Results Eighty women with TS (34.0 ± 11.7 years) and 214 controls (32.9 ± 10.6) responded. The TS group reported significantly more health problems and impaired health status in the two subscales “physical functioning” and “general health” ($P < 0.001$). Level of education and work participation was similar among TS and controls. TS moved away from their parents’ home later than controls (20.4 ± 4.0 vs. 18.7 ± 2.1 , $P = 0.001$). Age at sexual debut differed significantly (21.2 ± 4.3 vs. 17.3 ± 2.4 years, $P < 0.001$).

Conclusion TS attains the same level of education and level of employment as controls, they report more frequent occurrence of medical conditions, but scored lower on only two subscales in the SF-36. Despite considerable medical morbidity, TS seem to cope well with life.

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Introduction

Turner syndrome (TS) is a genetic disorder affecting only females, characterized by the absence of one or part of one X-chromosome. The incidence of TS is one in 2000 live female births.¹ Characteristics of TS are short stature, gonadal dysgenesis and cardiovascular malformations. Gonadal dysgenesis leads to oestrogen insufficiency, incomplete pubertal development, impaired fertility and increased risk of osteoporosis and fractures.² Increased morbidity with a number of medical conditions such as diabetes mellitus, hypothyroidism, hypertension and aortic dissection is documented.² Conductive and sensorineural hearing deficit are common and result in hearing impairment in early adult life.³ Growth hormone treatment (GHT) is recommended to enhance final height, and hormone replacement therapy (HRT) to induce puberty and maintain female sex characteristics.

Studies have documented deficits in four cognitive areas that interact, including visual-spatial organization, social cognition, nonverbal problem solving and psychomotor functioning.⁴ McCauley *et al.*⁵ described the psychosocial functioning in girls with TS and concluded increased risk of selective impairment in the social, cognitive and behavioural domains. A recent review describes the beneficial effects of hormonal treatment, whereas still arguing for screening by psychoeducational specialists before entering school and careful monitoring of social skill functioning.⁶ Young women with TS, in spite of the above documented medical and neuropsychological impairments, reported normal health-related quality of life when they had age-appropriate induced puberty and reached normal height.⁷

Thus, TS is a complex medical condition with specific cognitive characteristics and frequent medical morbidity. However, no studies have investigated the influence this has on health status using a population wide approach. It is not clear how the described medical and psychological impairments affect general well-being. We undertook this prospective cohort questionnaire study that explores self-reported health status, level of education, work participation, medical conditions and aspects of sexual functioning in adult women with TS compared with a control group matched for age and place of residence.

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Methods and materials

Participants were recruited from Frambu Resource Centre for Rare Disorders. Frambu is a national centre in Norway servicing patients with rare disorders, including TS, and those who seek services from Frambu are included in a database register. Participants were recruited from this register and from the Norwegian Turner Syndrome Association. We only had information on age and place of residence of the participants and the nonrespondents.

The questionnaires were sent to a total of 168 women with TS aged 18 years and above and, after receiving response from 80 TS respondents, to a control group approximately 10 times as large of 770 women established by The Norwegian National Registry, matched for age and place of residence of the TS women. A reminder was sent after 3 weeks to both groups.

The questionnaires included demographic data, socio-educational status, the Short Form 36 version 2 (SF-36v2),⁸ Norwegian Standard Classification of Education,⁹ The General Nordic Questionnaire for Psychological and Social factors at work (QPSNordic),¹⁰ Labour Force Survey LFS 1996,¹¹ a Life Satisfaction instrument¹² and a questionnaire measuring psychological strain¹³ during different life phases. All forms but the last are validated.

SF 36 consists of eight subscales: mental health (mh), role functioning emotional (re), social functioning (sf), vitality (vt) and general health (gh), bodily pain (bp), physical functioning (pf) and role functioning physical (rp). All subscales of the SF 36 have 0–100 scores (except the vitality scale which has scores from 0 to 90). Best health status score is 100.

Norwegian Standard Classification of Education (NUS2000) functions as a classification norm for all educational activities in Norway. This norm classifies years of education into nine levels starting with primary education and ending with postgraduate education and unspecified.

QPSNordic is a general questionnaire for psychological and social factors at work. The instrument renders information on how to improve working conditions, how to prevent health problems, increase efficiency and job satisfaction. Our survey has used questions from Chapter 2 in the survey regarding demands in the work situation. QPSNordic is tested for reliability and validity in two studies with data from four Nordic countries.¹⁴

The Labour Force Surveys (LFS) in Norway allows a classification of the population into categories as employed, unemployed or as persons not in the labour force. Characteristics of patterns of working time, permanency of the job and field of study are covered in our questionnaire.

Life Satisfaction instrument (LiSat-9) measures satisfaction with “life as a whole” and eight domains of life. The eight domains are “vocational situation”, “financial situation”, “leisure”, “contact with friends”, “sexual life”, “activities of daily living”, “family life” and “partnership relationship”. The nine variables are rated on an ordinal scale where very dissatisfying represent “one” and very satisfying is represented by “six” and has been epidemiologically validated in a representative Swedish sample.¹²

To examine possible differences in experienced psychological strain during different life phases between the women with TS and

the controls, we used a questionnaire designed by a Norwegian psychologist.¹³ The variables are rated “no strain”, “some strain”, “and heavy strain” and “not applicable”. The first questions concern experienced strain during primary school, life as a teenager, as a young adult, as a parent, when children leave home and ends with aging and possible disability.

The questions on self-reported health complaints were compiled with the current knowledge of medical conditions thought to affect females with TS and after consultations with leading members from the Norwegian Turner Syndrome Association in order to examine if suspected conditions were more frequent among TS. Other areas of concern were self physical activity, use of GHT and HRT and sexual functioning (age of sexual debut, number of sex partners, self confidence as a woman and sex partner).

The National Committee for Research Ethics (Ref: 401-60163 1:2006:840) and Norwegian Social Science Data Services (Project number 14879) approved the study in June 2006.

The study has been performed in compliance with the recommendations of the Declaration of Helsinki.

Statistical analysis

SF-36 was scored according to the Health survey manual & interpretation guide¹⁵ (QualityMetric Incorporated, Lincoln, RI, USA) and data entered into SPSS 16.0 for Windows. Descriptive analyses showed that the data in seven of the eight subscales (except in the vitality subscale) were not normally distributed in both groups. We applied the Mann–Whitney *U*-test to compare the two groups. All categorical data were analysed using Pearson’s χ^2 test and in case of <6 in a cell, a protected *P* value was used. Correlations were analysed using Spearman rank order correlation. Level of education was categorized as follows: 1 = up to 12 years, 2 = up to 15 years, 3 = >15 years. Self-reported medical conditions data were reported in four categories. Because of too few answers in each stratum, categories were combined into two categories: “have this ailment/problem”, “is assessed for it now” and “is treated for” were combined into “yes, has this problem” and “no symptoms” remained the same. To be able to statistically analyse the data because of small numbers in LiSat-9, we dichotomized the scale into 1–4 (not satisfied) and 5–6 (satisfied). This dichotomy has been proved statistically valid in a previous study in 2002.¹⁶

Results

Eighty women with TS and 214 controls responded (response rate: 47.6% and 27.8%, respectively), because all nonresponders received one reminder. The characteristics for both groups are presented in Table 1. The two groups differed in height and weight as expected, and women with TS had a higher BMI. There was a negative correlation between height and age in the TS group showing that the oldest women were the shortest ($r = -0.423$, $P < 0.001$). More women with TS lived alone or with parents than with a partner. The level of education and employment status was similar. The TS group consisted of 45 × and mosaics (45 × : 29 women, mosaics: nine, karyotype not established: 42).

	Turner syndrome (N = 80)		Controls (N = 214)		P-value
	n (%)	Mean ± SD (range)	n (%)	Mean ± sd (range)	
Age (years)		34.4 ± 11.7 19–64		32.9 ± 10.6 18–63	0.342
Weight (kg)		59.3 ± 12.7 34–97		67.01 ± 12.1 44–130	0.001
Height (cm)		152.8 ± 6.2 138–165		168.2 ± 6.2 143–187	0.001
BMI (kg/m ²)		25.5 ± 5.0 16.9–39.5		23.7 ± 3.9 17.2–42.5	0.005
Smoking					
Daily	4 (5)		39 (18)		0.002
Sometimes	6 (8)		32 (15)		
Never	69 (86)		143 (67)		
Daily medication use	69 (85)		39 (18)*		<0.001
Marital status					
Lives alone	37 (46)		42 (20)		<0.001
With parents	12 (15)		22 (10)		
With partner	31 (39)		149 (70)		
Age when leaving parents	67	20.4 ± 4.4 9–42	196	18.7 ± 2.1 13–25	0.001
Education					
Up to 12 years	44 (55)		94 (44)		0.2
Up to 15 years	23 (29)		77 (36)		
>15 years	13 (16)		43 (20)		
Employment status					
Employed or under education	71 (89)		186 (91)		0.096
Disability pension	7 (8)		10 (5)		
Out of work and domestic work	2 (2)		18 (5)		
Parenthood					
Yes	11 (14)		110 (52)		<0.001

Data are presented as mean ± SD and range or as categorical data. The Mann–Whitney *U*-test was used to test continuous data and the Pearson's χ^2 statistic for testing categorical data.

*Mainly medication for asthma, hypothyreosis, depression and hypertension.

SF 36 questionnaire

Women with TS reported impaired health status compared with the control group in the two subscales “physical functioning” and “general health”. Here a highly significant difference was present, whereas all other subscales showed comparative results (Table 2). In the TS group, we found that significant correlations between age and all subscales of SF 36 ($r = -0.231$ to -0.495 , $P < 0.05$) except in “social functioning”, whereas among controls there was only significant correlations between age and the subscales “mental health” and “role emotional” ($r = 0.172$ – 0.227 , $P < 0.05$). There was a significant negative correlation between “age when the women with TS were diagnosed” and four subscales: “bodily pain”, “physical functioning”, “role physical” and “role emotional” ($r = -0.199$ to -0.298 , $P < 0.05$). Height correlated positively to several subscales like “vitality”, “bodily pain”, “physical functioning” and “role physical” in the TS group, ($r = 0.312$ – 0.446 , $P < 0.01$), showing that the taller a given person was, the better the score on these subscales. There were no such correlations in the control group. Level of education in the TS group

showed a positive correlation in the three subscales “bodily pain”, “social functioning” and “physical functioning” ($r = 0.256$ – 0.314 , $P < 0.05$), while in controls level of education was positively

Table 2. Data from the questionnaire Short Form 36 (SF 36)

	Turner syndrome		Controls		P-value
	n	Median (range)	n	Median (Range)	
Mental health	75	80.0 (10–100)	214	80.0 (0–100)	0.5
Vitality	75	55.0 (10–90)	214	60.0 (10–90)	0.4
Bodily pain	76	84.0 (22–100)	214	84.0 (0–100)	0.4
General health	76	68.5 (10–100)	214	87.0 (5–100)	<0.001
Social functioning	78	100.0 (0–100)	214	100.0 (0–100)	1.0
Physical functioning	76	95.0 (15–100)	214	100.0 (20–100)	<0.001
Role physical	76	100.0 (0–100)	214	100.0 (0–100)	0.3
Role emotional	74	100.0 (0–100)	214	100.0 (0–100)	0.3

Data are presented as median and range for both groups. Best health status score is 100. The Mann–Whitney *U*-test was used to test data.

correlated to the “mental health”, “general health”, “physical functioning”, “role physical” and “role emotional” subscales ($r = 0.140-0.203$, $P < 0.05$). Hearing loss influenced how both groups reported their health status, with correlations between reported hearing loss and all subscales except “bodily pain” in TS women ($r > 0.361$, $P < 0.05$) and all subscales except “bodily pain” and “general health” in the control group ($r > 0.226$, $P < 0.05$).

Life satisfaction

The control group reported that they were significantly more dissatisfied with their financial and leisure situation than the TS group ($P = 0.009$ and 0.03 , respectively). In the other domains, there were no significant differences (Fig. 1).

Experienced psychological strain

The two groups experienced important life phases differently. Transition periods such as commencing school, the last years of primary school and life as a teenager showed significant differences between the groups, revealing that the women with TS reported more psychological strain during these years than their peers without TS (all $P < 0.05$). The controls reported more strain during education ($P = 0.001$).

Self-reported medical conditions

TS reported suffering from significantly more medical conditions in most areas covered by this questionnaire than the control group (Table 3).

Fractures

There were no significant differences between the groups concerning sustained fractures (data not shown).

Sexual functioning

The TS group ($n = 48$) had their sexual debut significantly later than controls ($n = 206$) (21.2 ± 4.3 vs. 17.3 ± 2.4 years, $P < 0.0001$).

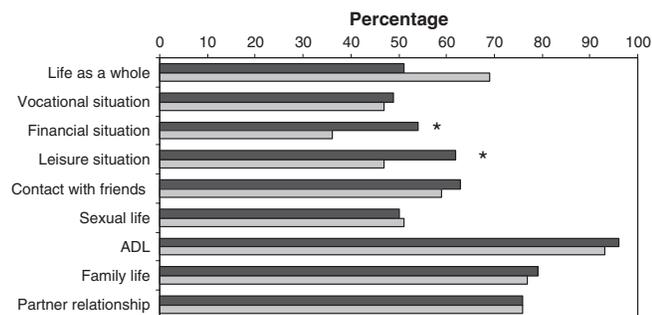


Fig. 1 Data from LiSat-9 showing satisfaction with life in different areas P -value calculated using the χ^2 statistic. Dark bars depict Turner syndrome and grey bars depict control. * $P < 0.05$.

Table 3. Self-reported health complaints

Health complaints	TS n (%)	Controls n (%)	P-value
Hearing loss	52 (64)	14 (7)	<0.0001
Strabismus	27 (34)	8 (4)	<0.0001
Shortsightedness	44 (55)	74 (35)	<0.0001
Longsightedness	22 (28)	35 (16)	0.01
Kidney problems	3 (4)	2 (1)	0.1*
Hypothyreosis	9 (11)	4 (2)	0.0008*
Osteoporosis	6 (8)	1 (1)	0.001*
Scoliosis	12 (15)	6 (3)	<0.0001
Kyphosis	3 (4)	2 (1)	0.1*
Oedema of the legs	15 (19)	12 (6)	0.0005
Type 1 diabetes	1 (1)	1 (1)	0.4*
Type 2 diabetes	6 (8)	0 (0)	0.0002*
Bowel disease	12 (16)	9 (4)	0.0004
High blood pressure	15 (19)	4 (2)	<0.0001*
Myocardial infarction	3 (4)	1 (1)	0.05*
Coarction of the aorta	5 (6)	1 (1)	0.004*
Dissection of aorta	0 (0)	0 (0)	†
Dental malalignment	20 (25)	8 (4)	<0.0001
Disease of the gingiva	8 (10)	8 (4)	0.04*
Underbite, undershot jaw	3 (4)	3 (1)	0.2*
Overbite, receding jaw	19 (24)	23 (11)	0.002
Overlapping molars	6 (8)	2 (1)	0.004*

Data are presented as the number (percentage) of patients reporting a specific health complaint. P value calculated using the χ^2 statistic unless otherwise stated.

*Fishers exact test.

†Not possible to compute a P value.

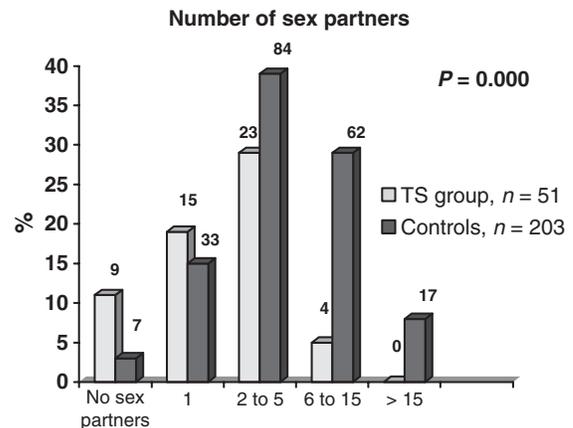


Fig. 2 Number of sex partners among women with Turner syndrome ($N = 80$) and controls ($N = 214$). Values are shown in number and percentage. P value calculated using the χ^2 statistic. Please note that not all recipients answered questions regarding sexual functioning, and actual respondents are shown in the figure.

More TS reported reduced self-confidence as a woman and sex partner [TS: 44 (51%) felt confident, 10 (13%) felt less confident, 15 (19%) felt uncertain; controls: 180 (84%), 11 (5%), 19 (9%), $P < 0.001$]. Women with TS also reported having significantly fewer sex partners than controls (Fig. 2). Furthermore, we found that age at onset of GHT and HRT both were positively related to

age at sexual debut (GHT: $r = 0.563$, $P < 0.001$; HRT: $r = 0.520$, $P < 0.001$).

Physical activity, physical therapy and chiropractor use

There were no differences between the groups concerning present or past exercise habits ($P = 0.6$ and $P = 0.2$, respectively). Significantly more TS women were swimming ($P < 0.001$), whereas the control group reported more activity in different sports ($P < 0.012$). Among those who were reporting inactivity, more TS felt clumsy ($P = 0.006$). The control group reported that during childhood and adolescence they liked physical activities better than the women with TS both during school time ($P < 0.003$) and during leisure time ($P < 0.001$). No differences were reported concerning how often the two groups used physiotherapy ($P = 0.5$) or a chiropractor ($P = 0.6$).

Operations

Eight women in the TS group had had a heart operation (two in the control group, $P < 0.001$), and the TS group had a significantly higher frequency of ear operations ($n = 27$) compared with the controls ($n = 4$) ($P < 0.001$). Sixteen women with TS (six in the control group) reported that they had had an eye operation ($P < 0.001$). Two TS women had bladder operations, none in the control group (too few data for statistical analysis). Operations on the back, in the ovaries and other unspecified operations showed no significant differences.

Growth hormone therapy, hormone replacement therapy and age at diagnosis

The younger women with TS were diagnosed on average 11 years earlier than the oldest group. The youngest group also started GHT and HRT on average 5 years earlier than the oldest group (Fig. 3).

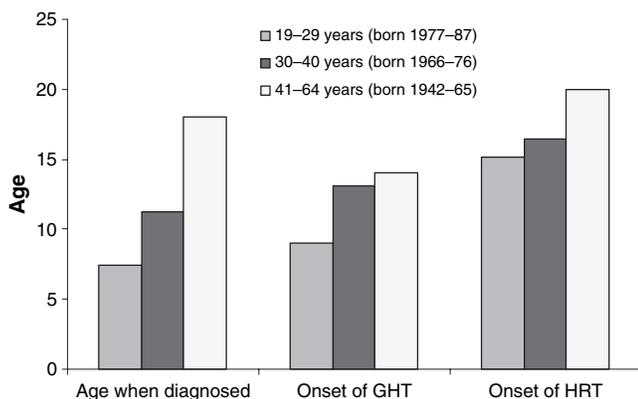


Fig. 3 Age when diagnosed, self-reported onset of treatment with GHT and HRT shown in age groups. Note that older women in the two older cohorts born before 1976 reporting receiving GH treatment, have probably been given other growth promoting therapy in the form of oxandrolone or other steroid therapy, as neither recombinant nor pituitary GH were available at the time they report starting GHT.

Self-reported medical follow-up in relation to the TS diagnosis

Twelve women (15%) reported that they were very dissatisfied or dissatisfied with the medical follow-up that they received. Twenty-one percent reported satisfaction and 13% were very satisfied with their follow-up. The women were followed by physicians from various medical disciplines, but only six women were followed up by a cardiologist. Eight women reported no medical follow-up.

Education and employment

There were no significant differences concerning level of education, but the two groups reported significant differences in relation to field of study ($P = 0.042$). More TS chose to qualify as kindergarten teachers and, compared with the controls, more rarely entered economic or administrative professions. There were no significant differences in working hours per week and working shifts (day/night). Employment status showed significant differences between the two groups ($P = 0.009$). The TS group (68%) more often held a permanent job position than the control group (54%). None of the women with TS were self-employed (4% in controls), and 8% were employed in a temporary job position (19% in controls).

Demands in the work situation

The TS group reported that they had to make quick and complicated decisions more rarely or never compared with the controls. Twenty-four percent of the women with TS never or very rarely experienced that they needed more education or training to cope with their tasks at work (16% among controls, $P < 0.05$).

Discussion

The main result from the present study dealing with aspects of health status in TS is apparently disparate. On the one hand, TS women report a much more frequent occurrence of medical conditions, a later sexual debut and diminished sexual confidence, live less frequently with a partner and score lower on two subscales (general health and physical functioning) from the SF-36 questionnaire. On the contrary, they attain the same level of education and employment, have a close attachment to working life, and reported a higher degree of satisfaction with their financial and leisure situation. To our knowledge, TS is the only rare and chronic disorder where afflicted individuals cope in such a manner with the demands of an increased burden of medical conditions in addition to the described sociopsychological challenges. The present study corroborates and extends previous reports and as such provides an opportunity to evaluate the impact of increased medical morbidity and medicinal treatment on health status.

We demonstrate that young TS women move significantly later from their parents' home than controls and fewer lived with a partner, thus corroborating earlier reports.^{17,18} Civil status was reported in a US study from 1995 where only 19.6% cohabited,¹⁸ and 18% cohabited in a Polish study,¹⁹ in contrast to 31% living with a partner in the present study.

Carel *et al.*²⁰ used the SF 36 questionnaire in their French cohort study of 568 young women (22.6 ± 2.6 years) with TS. They found no significant differences between these women and a control group of the same age from the general population. Our present study of older women (34 ± 11.7 years) shows significant differences in two subscales: general health and physical functioning, which may indicate that the physical impairments and conditions become more prominent as TS women grow older. Boman *et al.*²¹ reported similar results from their sample of 63 women with a mean age of 31.5 years. There was also a significant correlation between higher age at diagnosis and lower score in four subscales (bodily pain, physical functioning, role physical and role emotional) in our study suggesting that these areas are vulnerable to late induction of puberty. Boman *et al.*²² studied well-being in Swedish TS using The Psychological General Well-being Index and found that higher age at diagnosis was related to lower well-being. All these findings might also indicate that not only ageing affects health status but also the fact that the eldest women did not receive age-appropriate treatment.

Level of education is described as a strong predictor of health status in TS,²⁰ in other groups like short stature patients,²³ and in normative data.²⁴ Here, women with TS and higher education report better health status only in three subscales (bodily pain, social functioning and physical functioning) in contrast to both the control group and the normative data where six and eight subscales, respectively, were reported with better health status.²⁴ Thus, we argue that education is not an equally strong predictor of health status in TS women as in other women.

Hearing loss is associated with perceived health-related quality of life in adult women with TS,^{20,22} as also shown here, with positive correlations between the SF-36 subscales and hearing loss in both TS and controls. As 64% in this study sample reported hearing loss (7% in the controls) this pinpoints a major problem for TS women and implies that management of otological problems should be rigorously addressed as recommended in recent guidelines.³

The TS group reported a higher psychological strain than the controls during life phases that might be vulnerable for many children and young adults. This confirms that young TS girls and adults might be even more exposed due to their often late onset of puberty and short stature compared with their peers.

Our results show that TS women report a significantly higher frequency of medical conditions than controls, as would be expected. Frequent otological and ophthalmological findings, hypothyroidism,³ osteoporosis and scoliosis,^{3,25} type 2 diabetes, inflammatory bowel disease and hypertension^{1,3} are all well described. In this Norwegian cohort, we also find increased frequencies of dental malalignment, disease of the gingiva, receding jaw and overlapping molars, not commonly described in the literature. A study in 1996 described occlusal morphology²⁶ and the dental involvement are also emphasized in the guidelines from the Turner Syndrome Study Group.³

Maximum oxygen uptake has been described as decreased in adults with TS² and a Swedish study reported in 1999 that women with TS were significantly less physically active than the controls.²⁵ In contrast, we found that 33% of TS reported to be physically active on a regular basis in comparison with 22% among controls.

Our data corroborate previous Belgian and Swedish studies in showing that TS women are at least as well educated as the general

population. The Belgian study reported that 12.7% of TS women attended university (7.7% among controls).²⁷ Likewise, a Swedish study reported that 88% of the women with TS were students or employed²¹ compared with 89% in our study. We have found no other studies reporting experienced work demands by TS women. The results may indicate that women with TS choose jobs that demand less decision making and that they feel well educated for the work they do since they report less need for further education.

Epidemiological studies in Denmark^{1,2} show median age of 15 years at diagnosis. Our results correspond to previous results by Cunniff *et al.*¹⁸ who reported a diagnostic age 7.8 years (<20 years) and 17.4 years in the age category >35 years.

Sexual debut differed significantly in the two groups, being delayed in the TS group, similar to the Polish data (22.7 years).¹⁹ Furthermore, we show that the age at GHT and HRT start is related to age at sexual debut similar to French data concluding that it is of utmost importance that puberty is induced at a physiologically appropriate age to enhance self-esteem, social adjustment and sexual activity.²⁸

The present findings must be interpreted within the limitations of this study. Only a part of the Norwegian population of women with Turner Syndrome is registered in the database at Frambu Resource Centre for Rare Disorders or as member of the Turner Syndrome Association. The reason for the low response rate is not clear. Our clinical experience is that several TS women prefer to live their lives without acknowledging their diagnosis on a daily basis and therefore rejected our postal questionnaire. The low response rate may lead to selection bias, but it is not clear which direction this bias might have. However, the rate of participation is always a challenge in postal questionnaires.²⁹

In spite of the documented medical morbidity and psychosocial challenges several of these girls and women meet, they are well educated, are equally or more often fully employed and they report a health status that correspond to the control group in all but two (important) subscales. Further studies are needed to explore the changes that might come with ageing. A need for more systematic follow-up from medical specialists is recommended.

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Competing interests/financial disclosure

Nothing to declare.

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