

ORIGINAL ARTICLE

A 6-year Follow-up survey of health status in middle-aged women with Turner syndrome

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Summary

Objective Studies suggest younger women with Turner syndrome (TS) have good quality of life. Less is known about everyday functioning in adults with TS. In a 6-year follow-up study, multiple areas of functioning were compared between TS women and controls. **Design:** Women with TS and controls were mailed a self-report survey 6 years after a baseline study.

Patients Fifty-seven women with TS (*M* age 40.6 ± 11.1 years) and 101 controls (*M* age 38.8 ± 10.6 years, *ns*) responded.

Measurements Measures of background information, experienced life strain and presence/impact of health conditions were developed for this study. The QPS Nordic measured perceived workload challenges. The LiSat-9 measured life satisfaction. The Rosenberg Self-Esteem Scale measured self-esteem.

Results More TS women lived alone, fewer had biological children, and more had adoptive children. TS women reported fewer sex partners and less sexual confidence. Controls had higher education. There was no difference in employment status. More TS women received disability pensions. TS women reported their work as more physically challenging, less positively challenging and requiring less knowledge skills. TS women experienced more life strain in school, adolescence and late working life. Controls reported higher overall life satisfaction, with no difference between samples on specific domains. TS women reported lower self-esteem. For TS women only, physical health at baseline predicted length of education and mental health at baseline predicted self-esteem.

Conclusions Women with TS face more challenges than controls on several domains of functioning. Early physical and mental health may influence later educational achievement and self-esteem for women with TS.

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Introduction

Turner syndrome (TS) is a genetic disorder involving total or partial absence of one X chromosome, affecting an estimated 1:2500 women.¹ Short stature, ovarian failure, oestrogen and thyroid hormone deficiency, and increased risk of somatic conditions (e.g. cardiac malformations, hearing impairment) have been documented for women with TS in several studies.^{2–4} Intellectual functioning is normal, with some reports of specific non-verbal learning problems.⁵ Women with TS seem to attain equal or higher level of education compared to controls, with no difference in work status,^{6–9} although increased occurrence of early retirement has been documented.⁹ More women with TS live without a partner and fewer become mothers.⁹ Earlier studies have shown increased shyness and lower self-esteem in girls with TS compared to controls, whereas more recent studies have not found such differences.⁸

In 2010, we published a cross-sectional survey of 80 women with TS aged 19–64 years (mean 34.4 years) compared to 214 healthy controls.⁷ The main findings were that women with TS reported (i) lower general health and poorer physical functioning compared to controls; (ii) higher satisfaction with leisure and private economy compared to controls, with no differences between groups in other areas of satisfaction; and (iii) later sexual debut, fewer sexual partners and less sexual confidence compared to controls.

A handful of studies have investigated similar areas of functioning in women with TS.^{9–13} In a large birth cohort registry study from Denmark, socio-economic status in 979 women with TS and healthy controls was compared, showing women with TS were less likely to have a partner and to be mothers, and more likely to retire early compared to controls.⁹ Education and employment rates, as well as income after the age of 30 years, were comparable between the groups. In an observational case-series study, only half of 26 women with TS (aged 20–50 years) were sexually active.¹² Furthermore, in this sample, sexually active women reported lower sexual arousal compared to healthy controls on a female sexuality self-report questionnaire. Another study reported only 30% of women with TS being sexually well functioning.¹⁰ Late sexual debut or no sexual activity among women with TS has also been described.¹¹ A recent Brazilian

study showed that social skills of young TS adults were similar to their biological sisters, despite the fact that their mothers regarded their relationship with their TS daughter as more problematic compared with their daughters without TS.¹⁴ The same study showed that more women with TS had been teased during their childhood and adolescence, and fewer had sexual experiences.¹⁴

In summary, a somewhat contradictory picture emerges concerning social and sexual functioning in women with TS. On one hand, there are documented difficulties in physical and social functioning. However, such problems do not seem to affect educational or vocational functioning, or quality of life. A substantial amount of this research evidence is based on cross-sectional studies, making it difficult to ascertain how factors affecting the lives of women with TS are associated. To address this gap, the present report presents follow-up data for the women with TS and the controls examined by us earlier⁷ (hereafter, 'baseline'), 6 years after the original study.

The aims of the present study were to examine change in demographic, educational and vocational data, incidence of health conditions and their impact, perceived workload and challenges, experienced life satisfaction and strain, and sexual functioning 6 years after baseline.⁷ We hypothesize that main findings will be similar to baseline, and we argue this will validate the results from the original study.

Methods

Samples and procedures

The sample comprised 56 women with TS (mean age = 40.6 ± 11.1 years) and 101 controls (mean age = 38.8 ± 10.6 years; *ns*). We attempted to contact all responders from baseline, that is 80 women with TS and 214 controls. At baseline, women with TS were recruited from the database register of Frambu Resource Center for Rare Disorders in Norway. The control group was established by the Norwegian National Registry and matched on age and place of residence with the TS women. The response rates at baseline were 47.6% for women with TS and 27.8% for controls. Nonresponders at baseline were considered to not have consented to participation and were not contacted at follow-up.

For the current follow-up study, the response rate in the TS sample was 70.0% (i.e. 56 of 80). Since the 2010 study, two participants were deceased, four questionnaires were returned with 'address unknown', one questionnaire was returned with active nonconsent and 17 questionnaires were not returned for unknown reasons. In the control sample, the response rate was 47.2% (i.e. 101 of 214). We were not able to identify current addresses for 47 of the 214 original controls; thus, questionnaires were sent to 167 controls. Of these, five questionnaires were returned with 'address unknown', nine were returned with active nonconsent and 52 were not returned for unknown reasons. A reminder letter with new questionnaires was mailed to nonresponders in both samples 2 months after the first request.

For both samples, there were no significant differences between responders and nonresponders to follow-up in terms of education level, age or health scores at baseline.

Measures

Background information. The following information was collected from both samples: marital and family status, education level, work and employment status, alcohol and tobacco use, and height/weight. The TS sample received additional questions regarding oestrogen treatment and health service use.

Health conditions and their impact. Both samples were asked to rate past or current diagnosis, or current assessment (*yes/no*) of 22 health conditions/diseases (e.g. hearing loss, osteoporosis). The list was compiled based on current knowledge of medical conditions among women with TS and on consultations with board members from the Norwegian Turner Syndrome Association. In addition, participants were asked to rate current impact of confirmed health conditions on a 9-point Likert scale from (0) lowest impact to (8) highest impact. Participants were also asked to rate extent of joint/muscle pain on a 5-point scale from (0) never to (5) daily, to indicate pain site (e.g. back, head, shoulders) and to report any experience of surgery.

The QPS Nordic¹⁵ was used to measure perceived workload and challenges. The QPS Nordic is a self-report measure of 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. We used questions from Chapter 2 in the survey regarding demands in the work situation. QPS Nordic has been tested for reliability and validity in two studies with data from four Nordic countries.¹⁵ In the present study, interitem reliability of the QPS Nordic was good for the TS sample ($\alpha = 0.88$) and acceptable for controls ($\alpha = 0.69$).

The Life Satisfaction instrument (LiSat-9)¹⁶ developed at the University of Umeå, Sweden, was used to measure satisfaction with *life as a whole* and eight specific life domains of life (i.e. *vocational, financial, leisure, contact with friends, sexuality, activities of daily living, family and partnership/relationship*). The nine variables are rated on a 7-point scale from very dissatisfying (1) to very satisfying (6). The LiSat-9 has been epidemiologically validated in a representative Swedish sample.¹⁶ In the present study, interitem reliability of the LiSat-9 was good in both samples (TS sample $\alpha = 0.85$; controls $\alpha = 0.84$).

Experienced life strain was examined with a questionnaire designed by a Norwegian psychologist.¹⁷ Sixteen different life phases (e.g. *early primary school, adolescence, old age*) were rated either *no strain, some strain, heavy strain* or *not applicable*. Interitem reliability was good in both samples (TS sample $\alpha = 0.85$; controls $\alpha = 0.83$).

Sexuality was assessed with two questions developed for baseline in co-operation with board members from the Norwegian Turner Syndrome Association. Participants were asked to rate number of sexual partners and confidence as a sexual partner (*yes/no*). In the present study, rating of confidence as a sexual

partner on an 8-point scale from (0) very uncomfortable to (8) very confident was added.

Self-esteem was measured with the Rosenberg Self-Esteem Scale.¹⁸ This is a ten-item scale measuring different aspects of self-esteem (e.g. *I take a positive attitude on myself, I feel that I have a number of good qualities*) rated on a four-item scale from 1 (*strongly agree*) to 4 (*strongly disagree*). Interitem reliability was good in both samples (TS sample $\alpha = 0.91$; controls $\alpha = 0.90$).

Statistics

Data were analysed with IBM Statistics SPSS version 22.0, New York, NY, USA. Descriptive statistics (Fisher's exact tests, chi-square analyses and Pearson's r correlations) were applied to examine differences in frequencies between samples and relations between variables. Independent sample t -tests were applied to examine differences between samples when variables were scaled. Linear regression models were applied to examine predictors from the original study of education level, overall life satisfaction and self-esteem at follow-up.

Ethics

The National Committee for Research Ethics (Ref: 401-601631Æ2006Æ840) and Norwegian Social Science Data Services (Project number 14879) approved the study in June 2006 and the follow-up study in 2012. The study has been performed in compliance with the recommendations of the Declaration of Helsinki.

Results

Background information

Controls more frequently reported living with a partner and having children ($P < 0.05$). More women with TS reported having adopted children ($n = 7$) compared to controls ($n = 2$; $P < 0.05$). In the TS sample, seven participants confirmed at least one successful pregnancy, two without medical intervention and five through egg donation. Six participants had been assessed for infertility. Since baseline, two additional participants in the TS sample reported pregnancy, three more participants reported assessment for infertility and two more reported treatment for infertility in the form of egg donation (Table 1). Controls had a higher level of education ($P < 0.001$).

In terms of work status, more women with TS reported to be on disability pensions, while there was no difference between the samples in frequency of working, working part time or holding a permanent position (Table 1). Women with TS were shorter in length, but with similar weight and consequently a higher BMI compared to controls.

Among women with TS, 89% had received long-term sex hormone replacement therapy, with 77% currently receiving treatment. The mean age of onset for oestrogen treatment was 17.1 ± 4.6 (10–32) years, and for subjects that had ended oestrogen treatment the age was 41.7 ± 17.6 (17–62) years.

Table 1. Background variables for women with Turner syndrome and controls at 6-year follow-up compared to baseline

	TS (N = 56)	Controls (N = 101)
Mean age \pm SD	40.6 \pm 11.1	38.8 \pm 10.6
Marital and family status (%)		
Live alone	50	7**
Live with partner	34	60**
Have biological children	20	74***†
Have adopted children	13	2*
Highest education (%)		
Primary + Secondary school	8	2
High school	36	17
<3-year University/college degree	27	30
>3-year University/college degree	23	44***†
Work and employment (%)		
Currently working	68	77
Holds permanent position	73	69
Works part time	34	22
On disability pension	14	1**
Alcohol and cigarette use		
Daily smoker	2	7‡
Drinks >6 alcohol units per week	4§	12§
Mean height (cm) \pm SD	152 \pm 6	168 \pm 6**
Mean weight (kg) \pm SD	64 \pm 19	68 \pm 13
Mean body mass index \pm SD	27.6 \pm 9.2	23.8 \pm 4.3**

TS, Turner syndrome; SD, standard deviation;

*Difference between samples is significant at the $P < 0.05$ level.

**Difference between samples is significant at the $P < 0.001$ level.

†Significant within-sample increase from baseline.⁷

‡Significant within-sample decrease from baseline.⁷

§Data not available from baseline.⁷

Since baseline, six participants had ended oestrogen treatment at a mean age of 42.3 (25–63) years.

Regarding health service use, women with TS reported that contact with the healthcare system most frequently involved a general practitioner (63%), gynaecologist (30%) and ENT doctors (20%). Eight participants in the TS sample (14%) reported that no healthcare professional followed them up for TS. On a 1–5 scale, the mean overall satisfaction with TS-related health services was 3.1 ± 0.8 .

Health conditions and their impact

Using a questionnaire developed for this study, women with TS confirmed nine health conditions more frequently than controls: hearing loss, strabismus, scoliosis, kidney problems, type 2 diabetes, bowel disease, oedema of the legs, high blood pressure and dental misalignment. Other diseases were reported with insignificantly higher frequency, for example thyroid disease and aortic dissection. There was no difference in reported perceived impact of health conditions between the samples (Table 2).

Five women in the TS sample had been diagnosed with diabetes type 2 since baseline, which was a significant increase ($P < 0.05$), while there were no other significant changes from baseline in the occurrence of any other health conditions in both samples.

Table 2. Frequency and impact of health concerns for women with Turner syndrome and controls

	TS (N = 56)		Controls (N = 101)	
	% confirming concern	Impact‡ M ± SD	% confirming concern	Impact‡ M ± SD
Hearing loss	68	3.3 ± 2.0	12**	2.2 ± 1.6
Strabismus	38	1.4 ± 1.5	6**	1.2 ± 1.5
Short-sightedness	43	1.7 ± 1.7	39	1.6 ± 1.8
Long-sightedness	29	1.8 ± 2.2	18	2.0 ± 2.1
Kidney problems	7	0.3 ± 0.6	0*	–
Hypothyreosis	11	4.2 ± 2.2	4	2.0 ± 2.2
Osteoporosis	9	2.7 ± 3.1	3	0.7 ± 0.6
Scoliosis	23	3.1 ± 1.6	3**	1.0 ± 1.0
Kyphosis	5	–	1	1.0 ± –
Oedema of the legs	32	2.5 ± 2.0	9**	2.3 ± 2.3
Diabetes type 1	2	6.0 ± –	0	–
Diabetes type 2	9†	7.0 ± 1.4	1*	6.0 ± –
Bowel disease	20	5.0 ± 2.3	4*	6.3 ± 1.3
High blood pressure	18	1.5 ± 1.5	4*	2.0 ± 1.0
Myocardial infarction	4	1.5 ± 2.1	0	–
Coarctation of the aorta	0	–	0	–
Dissection of aorta	4	–	0	–
Dental misalignment	23	1.2 ± 1.0	6*	0.5 ± 0.6
Disease of the gingiva	11	3.8 ± 1.5	4	2.8 ± 2.1
Underbite, undershot jaw	5	0	2	1.5 ± 2.1
Overbite, receding jaw	14	2.0 ± 3.4	5	0.3 ± 0.5
Overlapping molars	7	0.5 ± 0.7	3	0 ± –

TS, Turner syndrome.

*Difference in frequency between samples is significant at the $P < 0.05$ level.

**difference in frequency between samples is significant at the $P < 0.001$ level.

†Significant within-sample increase from original study ($P < 0.05$).

‡Respondents were asked to rate the impact of a given condition on a 0–8 scale, with higher numbers indicating more impact. There was no difference between samples in impact on any of these variables.

Muscle/joint pain frequency was identical in the TS sample and in controls ($M = 3.3$ on a 1 (*never*) to 5 (*daily*) scale; *ns*). In both samples, neck, back and shoulders were the most frequently reported pain sites. Pain frequency had not significantly changed from baseline for either sample (data not presented).

More women with TS confirmed ear, eye and heart surgery compared to controls (all $P < 0.05$). There was no significant increase in any type of surgery between baseline and follow-up for either sample (data not presented).

Perceived workload and challenges (QPS Nordic)

There were no differences between the samples on 8 of 12 items on perceived workload and challenges. Women with TS rated their work required more physical endurance compared to controls ($P < 0.05$). Controls scored higher on conducting work

tasks they needed more training to perform, work requiring new knowledge and skills, and work being challenging in a positive way (all $P < 0.05$). For women with TS, there was a significant decrease in scores on work being challenging in a positive way from baseline (Table 3).

Life satisfaction (LiSat-9) and experienced life strain

Women with TS reported significantly lower overall life satisfaction compared to controls ($P < 0.05$), but there was no difference in satisfaction with specific areas of life (Table 4). To compare with baseline, satisfaction scores were dichotomized so that scores 1–4 represented *low satisfaction* and scores 5–6 represented *high satisfaction*. In both samples, there was a significant increase from baseline in overall life satisfaction, as well as satisfaction with work, economy, leisure, friends and sexuality (all $P < 0.03$), whereas frequency of satisfaction with family, independence and partner/spouse remained stable from baseline.

Women with TS reported more life strain than controls in four of 16 life phases (i.e. *starting school, last years of primary school, teenage years, after several years working*; all $P < 0.05$; Table 5). Compared to baseline, women with TS reported significantly more strain during education and work-life ($P < 0.05$). Controls reported significantly more strain as parents of teenagers compared to baseline ($P < 0.05$).

Sexuality

Women with TS reported having had fewer sex partners than controls ($M_{TS} = 3.1 \pm 5.6$; $M_C = 7.3 \pm 6.3$; $P < 0.001$). In both samples, there was no overall increase in number of sex partners since baseline. Fewer women with TS confirmed feeling confident as a sex partner compared to controls (39%_{TS} vs. 70%_C; $P < 0.05$), with no change from baseline within each sample. With sexual confidence measured on a scale from 1 to 8, controls scored higher ($M_{TS} = 5.3 \pm 1.9$; $M_C = 6.4 \pm 1.6$; $P < 0.001$). This item was not included at baseline.

Rosenberg Self-Esteem Scale

Self-esteem was significantly lower among women with TS ($M = 2.8 \pm 0.6$) compared to controls ($M = 3. \pm 0.5$; $P < 0.001$). Self-esteem was not measured at baseline.

Relations between health at baseline and follow-up data

For women with TS, physical and mental health at baseline was positively correlated with education, overall life satisfaction and self-esteem reported at follow-up (all $P < 0.05$). Thus, three linear regression models are presented with physical and mental health at baseline as predictors of education, overall life satisfaction and self-esteem at follow-up. Physical health (not mental health) at baseline significantly predicted length of education reported at follow-up ($P < 0.05$; explained variance 11.1%). Mental health (not physical health) at baseline significantly pre-

Table 3. Perceived workload and challenges for women with Turner syndrome and controls

Item	<i>M</i> ± <i>SD</i> on a scale from 1 (<i>never</i>) to 5 (<i>always</i>)	
	TS (<i>N</i> = 56)	Controls (<i>N</i> = 101)
Do you have to work overtime?	2.4 ± 1.1‡	2.3 ± 1.2†
Does your work require physical endurance?	3.1 ± 1.3	2.5 ± 1.1*
Does your work require quick decisions?	3.6 ± 1.2‡	3.8 ± 0.9¶
Are your work tasks too challenging for you?	1.8 ± 0.8	1.7 ± 0.6
Does your work require maximal attention?	4.0 ± 1.1	4.2 ± 0.9
Does your work require complex decisions?	2.9 ± 1.3	3.2 ± 1.0¶†
Do you conduct work tasks that you require more training to perform?	2.0 ± 1.0†	2.4 ± 0.9*
Do you feel that your educational skills are being used in your work?	3.7 ± 1.3†	4.0 ± 1.1‡
Is your work challenging in a positive way?	3.6 ± 1.0§	4.1 ± 0.8*†
Do you regard your work as meaningful?	4.2 ± 0.9	4.4 ± 0.8
Does your work require you to acquire new knowledge and skills?	3.4 ± 1.0‡	3.8 ± 1.0*
Do you have social relations with work colleagues?	4.4 ± 0.7	4.6 ± 0.7

TS, Turner syndrome.

*Difference between samples is significant at the $P < 0.05$ level.

†Within-sample positive correlation with length of education is significant at the $P < 0.05$ level.

‡Within-sample correlation with length of education is positive and significant at the $P < 0.001$ level.

§Significant within-sample decrease from baseline⁷ ($P < 0.05$).

¶Significant within-sample increase from baseline⁷ ($P < 0.05$).

Table 4. Life satisfaction for women with Turner syndrome and controls

Satisfaction area	<i>M</i> ± <i>SD</i> from 1 (<i>very dissatisfied</i>) to 6 (<i>very satisfied</i>)	
	TS (<i>N</i> = 56)	Controls (<i>N</i> = 101)
Overall	4.6 ± 1.0†	5.0 ± 0.9*†
Work	4.1 ± 1.1†	4.4 ± 1.1†
Economy	4.6 ± 1.1†	4.5 ± 1.2†
Leisure	4.5 ± 1.0†	4.5 ± 1.0†
Friends	4.7 ± 1.1†	4.6 ± 1.1†
Sexual	4.1 ± 1.3†	4.1 ± 1.4†
Independence	5.7 ± 0.7	5.9 ± 0.5
Family	5.0 ± 1.0	5.2 ± 0.8
Partner/spouse	5.2 ± 1.1	5.2 ± 1.1

TS, Turner syndrome.

*Difference is significant at the $P < 0.05$ level.

†Significant within-sample increase from baseline ($P < 0.05$).

Table 5. Perceived life strain for women with Turner syndrome and controls

Life phase	Mean strain ± <i>SD</i>	
	TS (<i>N</i> = 56)	Controls (<i>N</i> = 101)
Early primary school	1.6 ± 0.6	1.2 ± 0.6*
Late primary school	1.8 ± 0.7	1.5 ± 0.7*
Adolescence	2.2 ± 0.7	1.7 ± 0.7*
Leaving parental home	1.4 ± 0.6	1.4 ± 0.6
Young adult	1.5 ± 0.6	1.6 ± 0.7
During education	1.7 ± 0.7†	1.6 ± 0.6
Early work years	1.7 ± 0.8†	1.5 ± 0.6†
Late work years	1.8 ± 0.7†	1.5 ± 0.6*
Newly-wed/cohabiting	1.8 ± 0.6	1.9 ± 0.6
As parent of toddlers	1.8 ± 0.6	1.9 ± 0.6
As parent of teenagers	1.4 ± 0.5	1.6 ± 0.6†
When children left home	1.8 ± 0.6	1.8 ± 0.6
After divorce	1.3 ± 0.5	1.9 ± 0.6
As widow	1.5 ± 1.0	1.8 ± 1.0
Old age	1.5 ± 0.7	1.4 ± 0.5
Starting disability benefits	1.8 ± 0.8	1.6 ± 0.9

TS, Turner syndrome. *SD*, standard deviation.

*Difference is significant at the $P < 0.05$ level.

†Significant within-sample increase from baseline ($P < 0.05$).

dicted self-esteem at follow-up ($P < 0.05$; explained variance 20.0%). Finally, mental health (not physical health) at baseline significantly predicted overall life satisfaction at follow-up ($P < 0.05$; explained variance 22.0%).

For controls, neither physical nor mental health at baseline was significantly correlated with education, overall life satisfaction and self-esteem reported at follow-up, and linear regression models predicting education, overall life satisfaction and self-esteem at follow-up from physical and mental health at baseline were also nonsignificant (data not shown).

Discussion

This 6-year follow-up of women with TS compared to controls provides new insights into multiple areas of life and functioning for women with TS. Follow-up data confirmed findings from baseline⁷ that women with TS more frequently lived alone and fewer had biological children, which has also been previously shown.¹⁹ However, more women with TS had adopted children compared to controls. Furthermore, those with a partner were no less satisfied with their family or intimate relationships compared to controls. This is in line with results from a Swedish survey of sexuality among 57 women with TS.¹⁹ Similar to baseline, women with TS reported fewer sex partners than controls and felt less confident as sexual partners. Sexual confidence may be an issue to consider for counsellors of women with TS.

In spite of confirming several health conditions more frequently than controls, thus possibly experiencing a larger burden of disease, there was no difference in perceived impact of such health conditions between the samples. Having multiple health conditions tends to cause larger overall impact.²⁰ The fact that

women with TS report the same level of impact as controls, in spite of more diseases, may indicate that women with TS harbour good coping strategies. We did not examine coping strategies; however, numerous factors influencing coping in women with TS have been identified in previous trials, including quality of family relationships, height, cognitive functioning and physical appearance.²¹

More women in the control group had higher education compared to women with TS. This is in contrast to baseline⁷ and large studies from Belgium, Denmark and the United States.^{6,8,9} There was a significant within-sample increase in higher education for controls, indicating that over time, more controls achieve higher education compared to women with TS. Importantly, for women with TS, physical health at baseline was a significant predictor of education at follow-up. This indicates the physical impact of TS may prevent them from achieving higher education. Women with TS also reported increased strain during education compared to baseline, while for controls there was no increase.

There was no difference in employment status or work hours between samples. However, more women with TS reported early retirement. This is in line with a previous study.⁹ Women with TS reported their work required more physical endurance, was less challenging in a positive way and required less training, knowledge and skills compared to controls. Together, these findings indicate that women with TS hold less intellectually challenging and more physically challenging jobs, compared to controls. Several work tasks may be more physically demanding for a person of short stature.²² Length of education was significantly correlated with perceived work training, skills and knowledge needs within the TS sample, indicating that the lower education of women with TS may help explain these findings. Significantly, fewer women with TS regarded their work challenging in a positive way compared to baseline, with no difference for controls, indicating that positive work challenges may decline for women with TS over time.

While women with TS reported lower overall life satisfaction compared to controls, there were no differences in satisfaction within specific areas of life. This is in contrast to baseline, where women with TS were more satisfied with their leisure and financial situation. The difference may be due to change in the satisfaction or due to the fact that we did not reach 100% response in the follow-up part of this study among TS. Whereas the satisfaction scale was dichotomized at baseline, it was represented as a 6-point scale at follow-up. Both samples more frequently scored in the high end of the satisfaction scale since baseline, both in terms of overall life satisfaction and all other areas of life except satisfaction with family, independence and partner.

Women with TS reported lower self-esteem compared to controls, which is in line with some previous trials,²³ but in contrast to others.⁸ A global measure of self-esteem may not be ideal for women with TS. This is because previous studies have shown that in some domains of self-esteem, women with TS score lower than controls (e.g. popularity, physical appearance^{23,24}). However, in other domains of self-esteem, women with TS have

scored similar to controls (e.g. intelligence, behaviour, performance^{23,24}).

Our study revealed some important relations between variables. Whereas mental health at baseline predicted self-esteem at follow-up for women with TS, this was not the case for controls. Mental health at baseline also predicted overall life satisfaction for women with TS at follow-up, but not for controls. This indicates that early mental health interventions may be particularly important for women with TS to influence later areas of psychological functioning. Furthermore, self-reported physical health at baseline influenced education level at follow-up for women with TS, but not for controls. Potentially, interventions to reduce the physical impact of having TS may facilitate educational achievement for women with TS.

Limitations of the present study include that less than half of controls responded, challenging generalizability of sample comparisons. Although genetic confirmation is required to be registered in the database wherefrom we recruited women with TS, we did not have access to test details due to anonymity of the data and were not able to examine differences between different karyotypic TS groups. We do also not know much about the women with TS that chose not to participate. In addition, not all women with TS in Norway are known to us and they could thus not be approached and there is always the question of TS that are nondiagnosed and evidently not known to anybody.⁹

In conclusion, this follow-up study shows that women with TS face more challenges than controls on several domains of functioning. Domains that were comparable between the samples at baseline, that is education, overall life satisfaction, were significantly lower for the TS sample at follow-up. Early physical and mental health intervention for women with TS may facilitate later educational achievement and self-esteem for this group of women.

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