LESS READY FOR ADULTHOOD?—TURNER SYNDROME HAS AN IMPACT ON TRANSITION READINESS

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Abstract

Objective: Young women with Turner syndrome (TS) are known to be at risk for loss to medical follow-up. Recent literature indicates that there are disparities regarding transition readiness between different chronic conditions. So far, studies in young women with TS investigating their transition readiness compared to youths with other chronic conditions with no or minor neurocognitive challenges have not been reported.

Methods: Patients (n = 52), 26 patients with Turner syndrome (mean age 17.24 ± 2.10) and 26 controls with type 1 diabetes or a rheumatic disease (mean age 17.41 ± 2.44), were recruited from specialized paediatric endocrine outpatient clinics. The Transition Readiness Assessment Questionnaire TRAQ-GV-15 was used to compare transition readiness scores between TS and controls. In addition, information on individual handling of the questionnaire was obtained. Descriptive statistics and nonparametric methods were used to analyse the data.

Results: Significant differences for transition readiness scores were found between the two study groups. The global TRAQ-GV-15 score was significantly lower for females with TS. In particular, subscale 1 ‘autonomy’ of the TRAQ-GV-15 showed lower scores in patients with TS. Patients with TS needed significantly more help and more time to complete the questionnaire.

Conclusion: Special attention should be given to young women with Turner syndrome in the preparation for the transitional phase. By incorporating the assessment of transition readiness specialists will find it easier to identify underdeveloped skills and knowledge gaps in their patients. Unless a multidisciplinary young adult clinic is established, an older age than 18 years at transfer to adult endocrine care might be beneficial.

KEYWORDS
adolescent health, assessment questionnaire, chronic disease, health autonomy, healthcare transition, TRAQ, Turner syndrome
Transition paths are still not clearly marked for patients with Turner syndrome (TS)\(^1\) and transition outcomes proved to be poor in the past.\(^3\)\(^,\)\(^4\) Appropriate medical care after transition is only found in 3.5% per cent of TS patients.\(^5\)\(^,\)\(^6\) As the process of transfer from comprehensive paediatric to adult medical care has been studied more closely within the last years, considerations and recommendations for successful transition are becoming theoretically underpinned.\(^7\)\(^,\)\(^8\) Assessment of readiness for transition in the preparatory process has been recognized as beneficial for the imminent transfer of adolescents to adult medical care.\(^9\)\(^,\)\(^10\) At present, assessment of readiness for transition with disease-specific and nonspecific questionnaires evolves to a more common practice internationally.\(^10\)\(^-\)\(^12\) However, recent literature points out that the type of disorder might account for differences in transition readiness levels.\(^13\) Neurocognitive involvement and delayed developmental maturation in complex chronic conditions affect successful transfer to adult care.\(^14\) TS can be associated with mild neurocognitive challenges and difficulties in social behaviour\(^15\)\(^-\)\(^18\) in addition, puberty is often delayed and personal maturity sets on later in life in young girls and women with TS. This is associated with decreased self-confidence and with a lower degree of autonomy than peers.\(^19\)

The main objective of the study was to examine whether individuals with TS show differences in transition readiness scoring compared to an age- and gender-matched population diagnosed with other chronic conditions, such as type 1 diabetes or childhood-onset rheumatic disease. We hypothesized that girls and young women with TS would have lower indicators for transition readiness than female adolescents with diabetes or arthritis, in particular when it comes to skills that include self-management, self-organization and self-advocacy.

2 | PATIENTS AND METHODS

Our study sample comprised female adolescents and young adults with TS and 26 female adolescents and young adult with type 1 diabetes or childhood-onset rheumatic disease, treated at specialized paediatric endocrine outpatient clinics at Vienna and Graz. The clinics provide comprehensive care for patients with Turner syndrome according to international guidelines\(^20\) and also for a broad range of childhood-onset chronic conditions.

Inclusion criteria were as follows: Adolescent girls and young women born between 1994 and 2003 and diagnosed with either TS, type 1 diabetes or with a rheumatic disease at least six months prior to the start of the study.

Primary exclusion criteria were lack of German language skills and/or severe cognitive impairment as documented in the medical record.

2.1 | Recruitment

Female patients aged 14 to 23 were selected from the registry of the paediatric departments. Patients who seemed eligible to the study were contacted via email/ telephone or were asked personally about participation during routine clinical visits. After full explanation of the purpose and the course of the study, informed consent was obtained from patients and below the age of 18 years, also from parents. The study was approved by the local ethic committees (Medical University of Vienna EK Nr: 1465/2016 and Medical University of Graz EK Nr 30-297 ex 17/18).

32 patients with TS matching the inclusion criteria were identified at the two study sites. Contact was established in all cases, and 28 patients responded positively and participated in the study. One girl had to be excluded because parent’s consent was obtained too late, another girl was excluded due to inadequate compliance towards oestrogen substitution therapy, leading to a study group of 26 female patients with TS.

As for the control group, 38 female patients with type 1 diabetes and 43 female patients with a rheumatic disease were identified. Contact was established in all cases. All individuals responded positively and took part in the study.

We compared different diagnosis groups derived from our sample by using the transition readiness assessment questionnaire TRAQ-GV-15.\(^21\) First, female patients diagnosed with type 1 diabetes (n = 29) were compared to an age-matched group of female patients with a rheumatic condition (n = 29). Analysis did not reveal differences regarding transition readiness scores measured by the TRAQ-GV-15 or regarding the handling of the questionnaire between these two diagnosis groups (data not shown).

Therefore, this sample served as a control cohort for our group of patients with TS. After once more matching for age, twenty-six pairs of female participants (n = 52), 26 patients with TS, and 26 female patients diagnosed with either type 1 diabetes or a rheumatic condition were established.

Finally, data regarding transition readiness scores and application of the questionnaire were compared between TS participants (n = 26) and age-matched patients of the control group (n = 26).

One or two clinical appointments at the respective specialized outpatient clinic per year were stated from 71.4% of the TS patients and from 10.5% of the control group. Three or four doctor visits per year were reported from 28.6% of the participants in the TS group, whereas 89.5% of the controls reported three, four or even more routine clinical visits per year.

Mean age of patients with TS was 17.24 ± 2.10 years (range 14.49-22.56 years). Karyotype was 45, X in 12 patients and mosaicism involving a 46, XX or 46, X (iXq) line in 14 patients. Patients with ring chromosome X were not included in the study. 22/26 patients had been treated with growth hormone, with ongoing GH treatment in 8 patients. 4/26 had experienced spontaneous puberty. One participant was diagnosed in her origin country, and detailed information on history of oestrogen substitution was not obtained. The rest of the participants underwent pubertal induction and standardized oestrogen substitution following international consensus guidelines\(^20\)\(^,\)\(^22\) starting at a mean age of 12.1 years in doses of 0.05-0.07 mg/kg/d (3.7-mcg/d) per transdermal route\(^23\) or 0.2-0.3 mg/day micronized 17β estradiol orally.\(^24\) At the time of testing, all
patients were on cyclic sequence preparations with 1 to 2 mg oral 17β-estradiol or 50-100 mcg transdermal estradiol (depending on body weight and oestrogen levels) had Tanner stage 4 or 5 and regular menstrual bleeding.

All participants in the control group presented with a normal pubertal development for age as judged by pubertal staging (Tanner stage B4 to B5) and/or reporting of regular menstruations. Control subjects did not suffer from severe inflammatory or nutritional disorders and did not require hormonal substitution therapy. Mean age was 17.41 ± 2.44 years, range 14.75-23.39 years).

First language was German in 76.9% of patients with Turner syndrome and 46.2% of patients in the control group.

### 2.2 | Methods

#### 2.2.1 | Testing

Patients were either seen during routine medical visits or invited for new appointments at the study sites in Vienna or Graz between March 2017 and August 2018.

#### 2.2.2 | TRAQ-GV-15

The German version of the TRAQ 5.0⁹ comprises 15 items within three subscales: autonomy, health literacy and adherence (Table 1). TRAQ-GV-15 item scores range on a 5-point Likert scale between 1 to 5, total TRAQ scores fall between maximum 75 and minimum 15. Lower scores indicate less readiness for transition.

#### 2.2.3 | Statistics

Differences between overall TRAQ scores, subscale scores and single item scores of the TRAQ-GV-15 were analysed by nonparametric Wilcoxon signed rank test, in case of non-normal distribution. We

### TABLE 1  TRAQ-GV-15²¹ Single Item Mean Score for TS group and Controls

| TRAQ itemsa | Domain | Controls | | TS | | P |
|-------------|--------|----------|---|---|---|---|
| 1. Do you fill a prescription if you need to? | 1 | 52 | 3.50 | 1.63 | 3.35 | 1.33 | .791 |
| 4. Do you reorder medications before they run out? | 1 | 50 | 3.80 | 1.47 | 3.56 | 1.53 | .463 |
| 5. Do you call the doctor’s office to make an appointment? | 1 | 52 | 3.92 | 1.32 | 2.96 | 1.46 | .003** |
| 7. Do you arrange for your ride to medical appointments? | 1 | 52 | 4.12 | 1.34 | 3.12 | 1.53 | .004** |
| 12. Do you fill out the medical history form? | 1 | 52 | 4.69 | 0.84 | 4.54 | 0.91 | .323 |
| 13. Do you keep a calendar or list of medical and other appointments? | 1 | 52 | 4.46 | 0.91 | 4.15 | 1.12 | .307 |
| 15. Do you answer questions that are asked by the clinic staff? | 1 | 52 | 4.85 | 0.37 | 4.81 | 0.40 | .705 |
| 2. Do you know what to do if you are having a bad reaction to your medications? | 2 | 50 | 3.72 | 1.51 | 3.32 | 1.63 | .365 |
| 9. Do you apply for health insurance if you lose your current coverage? | 2 | 52 | 2.50 | 1.68 | 2.19 | 1.23 | .436 |
| 10. Do you know what your health insurance covers? | 2 | 52 | 2.58 | 1.55 | 2.58 | 1.45 | .874 |
| 11. Do you manage your money and budget household expenses (eg, use checking/debit card)? | 2 | 52 | 2.62 | 1.65 | 1.85 | 1.22 | .052 |
| 3. Do you take medications correctly and on your own? | 3 | 50 | 4.76 | 0.72 | 4.80 | 0.65 | .832 |
| 6. Do you follow-up on any referral for tests, check-ups or laboratories? | 3 | 52 | 4.38 | 0.98 | 4.31 | 1.09 | .873 |
| 8. Do you call the doctor about unusual changes in your health (eg, allergic reactions)? | 3 | 52 | 4.27 | 1.19 | 4.38 | 1.06 | .886 |
| 14. Do you tell the doctor or nurse what you are feeling? | 3 | 52 | 4.65 | 0.56 | 4.54 | 0.99 | .742 |

Abbreviations: dom., domain; Domain 1, Autonomy; Domain 2, Health Literacy; Domain 3, Adherence; SD, standard deviation; TRAQ, Transition Readiness Assessment Questionnaire.

*Items: derived from the American TRAQ 5.0.⁹

*P-value of <.05

**P-value of <.01
used the Mann-Whitney U test to test for significant differences between the two patient samples in handling the TRAQ questionnaire regarding fill-in time, frequency of help needed and consultation time. We considered a P-value of <.05 on a two-sided level as statistically significant. Analysis was performed with IBM® SPSS® software version 22.

Power analyses were performed post hoc, using the software G*Power® 3.1.9.4.25 on a significance level of 5% (2-tailed). For the sensitivity power analysis, the power (1-ß) was set to >0.80. Sample effect sizes (Cohen's d) were calculated with the online tool 'Psycmetrika.'26 Generally, effect sizes > 0.2 are considered small effects, >0.5 medium effects and >0.80 large effects.27

3 | RESULTS

3.1 | Transition readiness indicators

Total mean item score of the TRAQ differed significantly between the two groups, with TS participants scoring lower than controls (3.63 ± 0.64 in TS vs 3.92 ± 0.70 in controls, P = .029, d = 0.44, Table 2).

TRAQ-GV-15 subscales: Among mean item scores, a significant difference was found in subscale 1 ‘Autonomy’ (TS group 3.77 ± 0.87 vs. control group 4.20 ± 0.77, P = .009, d = 0.52) whereas results in subscale 2 ‘Health Literacy’ and in subscale 3 ‘Adherence’ did not differ significantly between the two groups (Table 2).

Results of the sensitivity power analysis demonstrate that with the given sample size of n = 52 the minimal detectable effect was 0.41. This implies that small to moderate effects reach statistical significance. Results for the post hoc power analyses (observed power 1-ß) for the total score and the subscale scores are presented in Table 2.

Comparison of the two groups on a single item level displayed significant differences for the scores of two items both belonging to subscale 1 ‘Autonomy’: Item 05 (‘Do you call the doctor’s office to make an appointment?’) and item 07 (‘Do you arrange for your ride to medical appointments?’) with lower scores in patients with TS (Table 1).

3.2 | Handling of the TRAQ-GV-15

Statistically significant differences could be found concerning ‘fill-in time’ and ‘time of consultation’. 56.5% of the TS group finished the questionnaire within 5 minutes. 26.1% of the TS patients answered the survey within ten minutes, but 17.4% needed up to twenty minutes for the task. In contrast, 84.6% of the controls completed the TRAQ-GV-15 within five minutes and 15.4% within ten minutes.

With regard to the consultation time, for 8.3% of the TS patients less than fifteen minutes of consultation time were documented, whereas 62.5% of the TS group needed thirty minutes of counselling and for 29.2% of the TS participants sixty minutes of counselling was documented. For controls, 84.6% of the participants consumed around fifteen minutes, four youth up to 30 minutes. No significant difference between the two groups was found with respect to ‘help needed’ in filling out the TRAQ (Table 3).

4 | DISCUSSION

We assessed healthcare transition readiness (HCT) in girls and young women with Turner syndrome applying a standardized method in comparison to an age-matched control group of youths with type 1 diabetes or childhood-onset rheumatic disease. Our study was powered to detect small to medium group differences.

We found statistically significant lower overall TRAQ scores as well as lower scores in subscale 1 ‘Autonomy’ for girls and young women with TS. For subscale 2 ‘Health Literacy,’ the difference in means between the TS group and the control group did not reach

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**TABLE 2** TRAQ-GV-15 Subscale Mean Item Score and Overall Mean Score for TS group and controls

| Item level | TRAQ-GV-15 total score | Domain 1 Autonomy | Domain 2 Health Literacy | Domain 3 Adherence |
|------------|------------------------|-------------------|-------------------------|-------------------|
|            | Mean ± SD (Md)         | Mean ± SD (Md)    | Mean ± SD (Md)          | Mean ± SD (Md)    |
| **TS**     | 3.63 ± 0.64 (3.50)     | 3.77 ± 0.87 (3.79)| 2.47 ± 1.03 (2.50)      | 4.51 ± 0.60 (4.75)|
| Controls   | 3.92 ± 0.68 (3.70)     | 4.20 ± 0.77 (4.50)| 2.86 ± 1.28 (2.75)      | 4.52 ± 0.65 (4.75)|

| Total score level | Mean ± SD (Md) | Mean ± SD (Md) | Mean ± SD (Md) | Mean ± SD (Md) |
|-------------------|---------------|---------------|---------------|---------------|
| **TS**            | 54.38 ± 9.61 (55.5) | 26.38 ± 6.08 (26.5) | 9.90 ± 4.11 (9.2) | 18.04 ± 2.39 (19.0) |
| Controls          | 58.78 ± 10.20 (56.0) | 29.37 ± 5.37 (31.5) | 11.42 ± 5.12 (10.0) | 18.08 ± 2.59 (19.0) |

Note: Abbreviations: 1-ß, observed power; d, observed effect size (Cohen’s d,27 standardized difference between two means); GV, German version; Md, median; SD, standard deviation; TRAQ, Transition Readiness Assessment Questionnaire.

*P-value of <.05.
**P-value of <.01.
statistical significance. Effect sizes for this scale were small, so the sample size may not have been sufficient to detect the actual effect.

Lower scores in subscale 1 'Autonomy' and items referring to practical issues in medical follow-up go in line with reports from the literature emphasizing less autonomy, higher dependence on parents and decreased self-organization in girls with TS compared to the reference population.28,29 Moreover, these results are in line with another recent study investigating the impact of physical health restraints versus neurocognitive impairments on transition readiness: higher indicators for transition readiness were found in patient groups diagnosed with a chronic physical condition without neurocognitive distinctiveness whereas lower indicators for HCT readiness were found in cohorts with cognitive and behavioural shortcomings, coming along with the primary chronic condition.13

The characteristic neuropsychological, social and emotional profile of girls and women with TS has now been well recognized and extensively studied.16-18,30,31 The effect of X-chromosomal loss on neurocognition and behaviour per se and mediated by oestrogen deficiency is complex and has been discussed in detail before.32

As all the participants in the TS study group were adequately substituted with oestrogens and the pubertal stage was comparable to the female individuals in the control group, we conclude that the discrepancies in TRAQ scoring in the TS group cannot be explained by differences in pubertal development. Thus further studies are needed to define if very early and ultra-low-dose oestrogen treatment could moderate cognitive profile and improve behavioural aspect related to autonomy.33

Participants of the TS group needed significantly more time to fill-in the questionnaire and consumed more time for discussion and for personal support before and after filling in the questionnaire. It has been reported that reduced working speed is related to slower processing of information and impaired working memory in patients with TS.15 Thus, executive functions (EF) play an important role in the process of transition. The term executive function is a collective term from brain research and neuropsychology. It refers to those mental functions used to control own behaviour, taking into account the conditions of a person’s environment. EFs such as planning, problem-solving and reasoning34 have been found to be impaired in TS.35-37

In this study, we did not perform neuropsychological testing in our cohort and thus, we cannot relate individual strengths and weaknesses in executive functions—especially abilities to plan and organize—with the results of the TRAQ. However, our data provide evidence for impaired specific abilities necessary for health autonomy in the group of girls and young women with TS when compared to the control group.

Recent consensus guidelines and reviews on the care of patients with TS univocally include summaries on the specific neurocognitive profile in TS17,20,38 and recommend age-appropriate psychological testing, training and support in order to optimize school performance and satisfaction with educational and occupational achievements.17,18 Likewise, transition and transfer of care for girls and young women with TS is given considerable space in recently published work cited above—notably, authors stress, that the age of transfer to adult medical care might have to be individualized. On the basis of our results, we recommend to use questionnaires like the TRAQ—which can easily be implemented in routine visits—in order to determine the individual readiness for transition as determined by items which refer to everyday-life situations important in a successful transfer to adult medical care. Clinical value of the TRAQ emerges from detecting gaps in health literacy of juvenile patients. Assessment of transition readiness will facilitate standardized discussions on the topic of transition between patient, caregivers and HCP, including the valuable opportunity of addressing uncertainties and outstanding issues. In addition, we recommend that healthcare professionals prepare for longer and intensified individual consultancy for adolescent girls with TS during the transition period. This additional time should also be dedicated to early introduction of general HCT-related topics (12, 25).

Still, in a considerable number of young women with TS transfer to the adult services might take place at an older age than 18. Along with longer life-expectation in the general population, the term ‘childhood and adolescence’ is mirrored in the term ‘emerging adulthood’.39,40 Healthcare autonomy is more likely to be achieved once education is finished and young adults move out.

In conclusion, our results indicate that patients with TS are less ready for transition than age-matched patients with less impairing chronic conditions. Assessing transition readiness in the context of a structured transition process will help in identifying underdeveloped skills or gaps in knowledge concerning disease management. We are convinced that the use of transition readiness
assessment questionnaires in routine standard care will be useful in the preparatory phase of adolescents with special health care needs for transfer.

**Strengths:** Our results concerning health autonomy in patients with TS have a direct impact on optimizing strategies during transition. This study has been performed in a homogeneous sample of patients treated at two tertiary care services according to international consensus recommendations and during personal appointments.

**Limitations:** This study was cross-sectional, and the study population was a convenience sample; therefore, results should be interpreted with caution. Furthermore, the sample size in this study was small. This fact applies to most studies including patients with rare diseases; however, a verification of our results on the impact of TS on the items and subscales of the TRAQ would need a larger population. Future research should systematically differentiate between the influence of structural, individual and conditional differences.

**CONFLICT OF INTEREST**

The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of this study.

**DATA AVAILABILITY STATEMENT**

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

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**REFERENCES**

1. Downing J, Gleeson HK, Clayton PE, Davis JRE, Wales JK, Callery P. Transition in endocrinology: the challenge of maintaining continuity. *Clin Endocrinol (Oxf)*. 2013;78(1):29-35.

2. Bernard V, Donadille B, Le Poulennec T, Neddou M, Martinerie L, Christin-Maitre S. MANAGEMENT OF ENDOCRINE DISEASE: Transition of care for young adult patients with Turner syndrome. *Eur J Endocrinol*. 2019;180:R1-R7.

3. Gawlik A, Kaczor B, Kaminska H, Zachurzok-Buczynska A, Gawlik T, Malecka-Tendera E. Quality of medical follow-up of young women with Turner syndrome treated in one clinical center. *Horm Res Paediatr*. 2012;77(4):222-228.

4. Ertl D-A, Gleiss A, Schubert K, et al. Health status, quality of life and medical care in adult women with Turner syndrome. *Endocr Connect*. 2018;7(4):534-543.

5. Devernay M, Ecosse E, Coste J, Carel J-C. Determinants of medical care for young women with Turner syndrome. *J Clin Endocrinol Metab*. 2009;94(9):3408-3413.

6. Gawlik A, Malecka-Tendera E. Transition in endocrinology: treatment of Turner’s syndrome during transition. *Eur J Endocrinol*. 2014;170(2):R57-R74.

7. Rollo A, Salardi S, Ciavarella A, et al. Transition from pediatric to adult care. Eight years after the transition from pediatric to adult diabetes care: metabolic control, complications and associated diseases. *J Endocrinol Invest*. 2014;37(7):653-659.

8. Gray WN, Schaefer MR, Resmini-Rawlinson A, Wagoner ST. Barriers to transition from pediatric to adult care: a systematic review. *J Pediatr Psychol*. 2018;43(5):488-502.

9. Wood DL, Sawicki GS, Miller MD, et al. The Transition Readiness Assessment Questionnaire (TRAQ): its factor structure, reliability, and validity. *Acad Pediatr*. 2014;14(4):415-422.

10. Moynihan M, Saewyc E, Whitehouse S, Paone M, McPherson G. Assessing readiness for transition from paediatric to adult health care: Revision and psychometric evaluation of the Am I ON TRAC for Adult Care questionnaire. *J Adv Nurs*. 2015;71(6):1324-1335.

11. Kaugars AS, Kichler JC, Alemzadeh R. Assessing readiness to change the balance of responsibility for managing type 1 diabetes mellitus: adolescent, mother, and father perspectives. *Pediatric Diabetes*. 2011:12:547-555.

12. Jensen PT, Paul GV, LaCount S, et al. Assessment of transition readiness in adolescents and young adults with chronic health conditions. *Pediatr Rheumatol*. 2017;15(1):1-7.

13. Beal SJ, Riddle IK, Kichler JC, et al. The associations of chronic condition type and individual characteristics with transition readiness. *Acad Pediatr*. 2016;16(7):660-667.

14. Godbout A, Tejedor I, Malivoir S, Polak M, Touraine P. Transition from pediatric to adult healthcare: assessment of specific needs of patients with chronic endocrine conditions. *Horm Res Paediatr*. 2012;78(4):247-255.

15. Ross J, Roeltgen D, Zinn A. Cognition and the sex chromosomes: studies in turner syndrome. *Horm Res*. 2006;65(1):47-56.

16. Hong D, Scalia Kent J, Kesler S. Cognitive profile of Turner syndrome. *Dev Disabil Res Rev*. 2009;15(4):270-278.

17. Culen C, Ertl D-A, Schubert K, Bartha-Doering L, Haeusler G. Care of girls and women with Turner syndrome: beyond growth and hormones. *Endocr Connect*. 2017;6(4):R39-R51.

18. Hutaff-Lee C, Bennett E, Howell S, Tartaglia N. Clinical developmental, neuropsychological, and social–emotional features of Turner syndrome. *Am J Med Genet C Semin Med Genet*. 2019;181(1):42-50.

19. Carel J-C, Elie C, Ecosse E, et al. Self-esteem and social adjustment in young women with Turner syndrome—impact of pubertal management and sexuality: population-based cohort study. *J Clin Endocrinol Metab*. 2006;91(8):2972-2979.

20. Gravholt CH, Andersen NH, Conway GS, et al. Clinical practice guidelines for the care of girls and women with Turner syndrome: proceedings from the 2016 Cincinnati International Turner Syndrome Meeting. *Eur J Endocrinol*. 2017;177(3):G1-G70.

21. Culen C, Herle M, König M, et al. Be on TRAQ — Cross-cultural adaptation of the Transition Readiness Assessment Questionnaire (TRAQ 5.0) and pilot-testing of the German Version (TRAQ-GV-15). *J Transit Med*. 2019;11(1):1–8.

22. Klein KO, Rosenfield RL, Santen RJ, et al. Estrogen replacement in Turner Syndrome: literature review and practical considerations. *J Clin Endocrinol Metab*. 2018;103(5):1790-1803.

23. Ankarberg-Lindgren C, Kriström B, Norjavaara E. Physiological estrogen replacement therapy for puberty induction in girls: a clinical observational study. *Horm Res Paediatr*. 2014;81(4):239-244.

24. Davenport ML. Approach to the patient with Turner Syndrome. *J Clin Endocrinol Metab*. 2010;95(4):1487-1495.

25. Faul F, Erdfelder E, Lang A-G, Buchner A. G*Power 3: a flexible statistical power analysis program for the social, behavioral, and biological sciences. *Behav Res Methods*. 2007;39(2):175-191.

26. Lenhart W, Lenhart A. *Calculation of Effect Sizes*. Dettelbach, Germany: Psychometrica; 2016. https://doi.org/10.13140/RG.2.2.17823.92329

27. Cohen J. *Statistical Power Analysis for the Behavioral Sciences*, 2nd ed. Hillsdale, NJ: Lawrence Erlbaum Associates, Publishers. 1988.

28. Ross JL, Roeltgen D, Kushner H, Wei F, Zinn AR. The Turner syndrome–associated neurocognitive phenotype maps to Distal Xp. *Am J Hum Genet*. 2000;67(3):672-681.

29. Lepage J-F, Dunkin B, Hong DS, Reiss AL. Impact of cognitive profile on social functioning in prepubescent females with Turner syndrome. *Child Neuropsychol*. 2013;19(2):161-172.
30. Kesler SR. Turner syndrome. *Child Adolesc Psychiatr Clin N Am*. 2007;16(3):709-722.
31. Knickmeyer RC, Davenport M. Turner syndrome and sexual differentiation of the brain: implications for understanding male-biased neurodevelopmental disorders. *J Neurodev Disord*. 2011;3(4):293-306.
32. Bakker J. The sexual differentiation of the human brain: role of sex hormones versus sex chromosomes. *Curr Top Behav Neurosci*. 2019;43:45-67.
33. Ross JL, Quigley CA, Cao D, et al. Growth hormone plus childhood low-dose estrogen in turner’s syndrome. *N Engl J Med*. 2011;364(13):1230-1242.
34. Diamond A. Executive functions. *Annu Rev Psychol*. 2013;64(1):135-168.
35. Romans SM, Stefanatos G, Roeltgen DP, Kushner H, Ross JL. Transition to young adulthood in Ullrich-Turner syndrome: neurodevelopmental changes. *Am J Med Genet*. 1998;79(2):140-147.
36. Kirk JW, Mazzocco MMM, Kover ST. Assessing executive dysfunction in girls with fragile X or turner syndrome using the Contingency Naming Test (CNT). *Dev Neuropsychol*. 2005;28(3):755-777.
37. Mauger C, Lancelot C, Roy A, Coutant R, Cantisano N, Le Gall D. Executive functions in children and adolescents with turner syndrome: a systematic review and meta-analysis. *Neuropsychol Rev*. 2018;28(2):188-215.
38. Lin AE, Prakash SK, Andersen NH, et al. Recognition and management of adults with Turner syndrome: from the transition of adolescence through the senior years. *Am J Med Genet A*. 2019;179(10):1987-2033.
39. Arnett JJ. Emerging adulthood: a theory of development from the late teens through the twenties. *Am Psychol*. 2000;55(5):469-480.
40. Hochberg Z, Konner M. Emerging adulthood, a pre-adult life-history stage. *Front Endocrinol*. 2020;10:918.

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