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A group of Brazilian Turner syndrome patients: Better quality of life than the control group

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Abstract

Current literature presents no consensus regarding which aspects influence health-related quality of life (HRQoL) of women with Turner syndrome (TS). The objective of the present study was to compare HRQoL in the TS and control group, using components and domains of the Medical Outcomes Study 36-Item Short Form Health Survey (SF-36). Observational, descriptive, and cross-sectional study with 44 women with TS aged between 18 and 30 years (TS group) and 44 healthy women of the same age which were sisters, relatives, or friends of the TS group (control group). A registration form and the SF-36 questionnaire were used to analyze HRQoL in TS in relation to the control group. The TS group presented better scores in the mental component summary and in the role physical, bodily pain, general health, social function, and role emotional domains compared to the control group. This study presented some unexpected results, different from those found in the current literature, showing the possibility of TS patients presenting better coping strategies. It is necessary to develop a specific questionnaire to assess QoL in TS and analyze in great detail which factors may influence the HRQoL of these patients.

KEYWORDS
adaptation, psychological, health related quality of life, Turner syndrome

1 | INTRODUCTION

Turner syndrome (TS) is defined as the total or partial absence of a sex chromosome in female subjects (Bondy, 2006; Elsheikh, Dunger, Conway, & Wass, 2002; Gravholt et al., 2017; Polivka & Merideth, 2015). TS has an incidence of 25–50 per 100,000 women (Gravholt et al., 2017), being considered the most common sex chromosome abnormality in female individuals (Polivka & Merideth, 2015).

Women with TS may present several phenotypic characteristics, malformations, and dysmorphic features. Short stature and gonadal dysgenesis are the most frequent symptoms, being identified in 90–100% of cases. Other common alterations include cardiovascular anomalies, thyroiditis and hypothyroidism, lymphedema on the back of the hands and feet, decreased bone mineral content, elevated liver enzymes, webbed neck, among others (Elsheikh et al., 2002; Gravholt et al., 2017; Neto, Lemos-Marini, Faria, Guerra-Junior, & Maciel-Guerra, 2011).

Considered a complex medical condition with multiple dysfunctions and frequent morbidity, TS seems to influence patients' quality of life (QoL) (Nadeem & Roche, 2014; Naess, Bahr, & Gravholt, 2010). The term health-related quality of life (HRQoL) has been used frequently and covers several aspects, among them, psychological condition, functional abilities, financial factors, social interactions, vocation, personal well-being, and physical health (Berzon, Hays, & Shumaker, 1993; Seidl & Zannon, 2004).

According to the review developed by Reis, de Assumpção, Guerra-Junior, and de Lemos-Marini (2018), the literature has a lot of controversial data regarding QoL in TS; however, this issue is apparently impaired in these patients. Thus, the objective of this study was to evaluate and compare HRQoL in women with TS and a control group.

2 | METHOD

This study was approved by the Research Ethics Committee of State University of Campinas (UNICAMP), protocol 786.743, and the informed consent forms were signed by all the study participants.

It was an observational, analytical, and cross-sectional study, which was carried out at the Clinical Hospital of State University of Campinas (HC-UNICAMP) in the Pediatric Endocrinology Outpatient Clinic. This hospital is the reference hospital for 86 cities and 6.5 million inhabitants and considered of high complexibility.

All of the TS patients from the Outpatient Clinic of this public university hospital who were in the age range and fit the criteria for the study were invited to participate and none of them declined participation. Forty-four women with TS aged between 18 and 30 years who had reached the final height (growth rate less than 1 cm/year), and whose diagnosis was confirmed by the karyotype, were included. All the members of this group were monitored periodically in this service, being seen at least twice a year by the same team for a significant period (13.7 ± 6.4 years), with a minimum period of 3.7 years and a maximum of 29.9 years.

The control group consisted of 44 women aged between 18 and 30 year and had also reached the final height. Sisters, relatives, and friends of subjects in TS group were invited to participate in the control group. A registration form was filled in to confirm that this group consisted of healthy individuals.

The registration form for participants in TS group included data on income, level of education, and information obtained from the medical record, such as anthropometric data, pubertal aspects, karyotype, and global healthcare information (comorbidities, medical history, medications, surgeries, gestation, perinatal intercurrences, and birth data).

Participants in the control group filled in the registration form with information related to anthropometric data, global health information (diseases, medical history, medications, surgeries, gestation, perinatal intercurrences, and birth data), and sociodemographic aspects such as level of education and per capita income.

The Medical Outcomes Study 36-Item Short Form Health Survey (SF-36) is a multidimensional questionnaire for generic health assessment (Ciconelli, Ferraz, Santos, Meinão, & Quaresma, 1999). This instrument was used for QoL assessment with individuals from the TS and control groups, being explained to both groups by the same researcher and applied in a standardized and objective way, in order not to influence the answers of the participants.

The SF-36 was published by Ware and Sherbourne (1992), developed by McHorney, Ware, and Raczek (1993) and validated by McHorney, Ware, Lu, and Sherbourne (1994) with 3.445 patients and replicated across 24 subgroups. This instrument was translated and validated to Portuguese by Ciconelli et al. (1999) in a study with 50 patients who had rheumatoid arthritis, in which the author mentioned the importance of using the SF-36 due to the difficulty of finding a generic QoL questionnaire in Portuguese, besides its easy application and comprehension. After its translation and validation to Portuguese this instrument has been used in many studies to assess QoL of patients with chronic diseases.

The application of the SF-36 was authorized by QualityMetric Incorporated, protocol: QM033635. This questionnaire consists of two components (physical and mental), each divided into four domains. The physical component summary consists of the physical function, role physical, bodily pain, and general health domains and the mental component summary is composed by the vitality, social function, role emotional, and mental health domains (Ciconelli et al., 1999; QualityMetric Incorporated, 2016; Ware & Sherbourne, 1992; Ware, Snow, Kosinski, & Gandek, 1993).

Scoring of the questionnaire was performed by the software QualityMetric Health Outcomes™ Scoring services 4.5 (QualityMetric Incorporated, 2016). This software uses non-normative scores and norm-based scores. Non-normative scores only consider the participants of the study in question and norm-based scores compare the scores obtained by the participants of the group with those obtained in the evaluation of the U.S. population in 1998, stipulating a mean of 50 and a *SD* of 10 (QualityMetric Incorporated, 2016).

The software uses algorithms that consist of numeric codes to associate the responses of the items in the questionnaire with the scales and formulas that calculate the final scores for each scale. The physical and mental components are calculated by summarized measures from the scales of the questionnaire. In each SF-36 domain, the highest scores represent better QoL, with scores ranging from 0 to 100 (QualityMetric Incorporated, 2016).

Scores of TS and control groups were processed using the Statistical Package for the Social Sciences (SPSS, Inc., version 16.0, Chicago) for Windows® and analyzed statistically by comparing the physical and mental component summaries and considering the non-normative and norm-based data of the eight domains in the SF-36 questionnaire.

3 | RESULTS

Despite the attempt of reducing socioeconomic and cultural differences between the two groups, with the control group being formed by sisters, relatives, and friends of the TS group, the monthly per capita dollar income of the TS group (median: U$ 310.17) was significantly higher than of the control group (median: U$ 206.10) (p = .004). The education level was not significantly different among TS and controls (p = .418).

Data in Table 1 show that TS and control groups had no significant difference considering age and body mass index (BMI) Z-Score; statistical

TABLE 1 Distribution and comparison of data on Z-score height, Z-score weight, and Z-score BMI between TS and control group

Variable	TS (n = 44)	Control (n = 44)	p
Age (years)	21.4 (18.0–30.5)	22.5 (18.4–30.7)	.145*
Height (Z-score)	−2.72 ± 0.98	−0.04 ± 1.04	<.001**
Weight (Z-score)	−0.77 (−5.72–1.42)	−0.02 (−2.54–2.53)	<.001*
BMI (Z-score)	0.50 (−2.44–1.90)	0.18 (−3.43–2.12)	.155*

Abbreviations: BMI, body mass index; TS, Turner syndrome.
*Mann–Whitney *U* test (median – minimum and maximum); **Student's *t*-test (mean ± SD).

TABLE 2 Aspects related to puberty, menarche, and estrogen of the TS group

Puberty	*N*	Menarche		Estrogen	
		Induced	Spontaneous	Yes	No
No spontaneous puberty	31	22	-	31	0
Incomplete spontaneous puberty	8	5	-	7	1
Complete spontaneous puberty	5	-	5	-	5
Total	44	27	5	38	6

Abbreviation: TS, Turner syndrome.

TABLE 3 Scores of the physical and mental component summaries of both groups

	TS (*n* = 44)	Control (*n* = 44)	*p**
PCS	56.4 (41.8–65.7)	55.9 (42.7–64.3)	.523
MCS	54.7 (16.8–62.0)	47.9 (11.6–62.1)	.004

Abbreviations: MCS, mental component summary; PCS, physical component summary; TS, Turner syndrome.
*Mann–Whitney U test. Data were expressed as median, minimum, and maximum.

TABLE 4 Non-normative scores of SF-36 domains between groups

	TS (*n* = 44)	Control (*n* = 44)	*p**
Physical function	95.0 (55.0–100.0)	95.0 (70.0–100.0)	.154
Role physical	100.0 (50.0–100.0)	87.5 (25.0–100.0)	<.001
Bodily pain	84.0 (51.0–100.0)	74.0 (22.0–100.0)	.009
General health	82.0 (17.0–100.0)	77.0 (20.0–100.0)	.024
Vitality	72.5 (40.0–100.0)	70.0 (10.0–100.0)	.142
Social function	100.0 (12.5–100.0)	87.5 (12.5–100.0)	.040
Role emotional	100.0 (0.0–100.0)	66.7 (0.0–100.0)	<.001
Mental health	80.0 (32.0–100.0)	78.0 (16.0–100.0)	.259

Abbreviation: TS, Turner syndrome.
*Mann–Whitney U test. Data were expressed as median, minimum, and maximum.

differences were only observed for weight and height, in which TS group had lower values, as already expected, since TS presents a smaller height.

The frequency of the karyotype in the TS group was: 45,X (*n* = 17); structural aberration of chromosome X or Y with or without mosaicism (*n* = 16) and mosaic without structural aberration of the chromosome X or Y (*n* = 11).

Regarding the aspects related to puberty, 31 participants in the TS group did not present spontaneous puberty; 13 presented spontaneous puberty (complete or incomplete); 32 participants presented menarche (spontaneous or induced); and the 12 participants who had not presented menarche were in the process of pubertal induction or had inadequate adherence to treatment. Progesterone was prescribed for patients who had induced menarche after the date of the first bleeding (Table 2). In addition, five patients used recombinant human growth hormone (rhGH).

TABLE 5 Norm-based scores of SF-36 domains between groups

	TS (*n* = 44)	Control (*n* = 44)	*p**
Physical function	55.1 (38.3–57.1)	55.1 (44.6–57.1)	.154
Role physical	56.2 (42.1–56.2)	52.7 (35.0–56.2)	<.001
Bodily pain	55.9 (41.8–62.8)	51.6 (29.4–67.5)	.026
General health	55.6 (25.1–64.0)	53.2 (26.5–64.0)	.024
Vitality	57.3 (42.0–70.4)	56.2 (27.8–70.4)	.136
Social function	57.1 (19.1–57.1)	51.7 (19.1–57.1)	.037
Role emotional	55.3 (23.7–55.3)	44.8 (23.7–55.3)	<.001
Mental health	52.7 (25.5–64.1)	50.4 (16.4–64.1)	.165

Abbreviation: TS, Turner syndrome.
*Mann–Whitney U test. Data were expressed as median, minimum, and maximum.

Considering comorbidities, the control group was formed by healthy individuals who presented no significant comorbidities and the TS Group presented the following issues: audiology (*n* = 12), cardiovascular (*n* = 15), bone metabolism (*n* = 25), hepatic (*n* = 4), thyroid (*n* = 28), insulin resistance (*n* = 1), Kabuki syndrome (*n* = 1), and agenesis of the corpus callosum (*n* = 1).

Scores of TS patients and controls were compared in relation to the physical and mental component summaries and the non-normative and norm-based data of the eight domains in the SF-36 questionnaire, to evaluate the global aspects of the QoL.

Regarding the physical and mental component summaries, the results showed no statistically significant difference between TS and control groups in relation to the physical component and showed that TS participants had a significantly better score in the mental component than the control group (Table 3).

The results of non-normative (Table 4) and norm-based (Table 5) scores of SF-36 domains were analyzed. In both types of score, there was no significant difference between TS and control group for physical function, vitality, and mental health domains. However, there was statistically significant difference for the role physical, bodily pain, general health, social function, and role emotional domains, in which the TS group presented higher scores.

4 | DISCUSSION

SF-36 was the instrument chosen in this study to assess QoL, because despite being developed for chronic diseases in general, considering

the systematic review conducted by Reis et al. (2018), it was possible to determine that the SF-36 was the most frequent internationally used instrument to assess QoL in TS and that this instrument had still not been used with Brazilian TS patients. Moreover, the authors considered that using the SF-36 in this study would make it possible to compare the results with other international studies.

Although such review considered that the QoL in TS is apparently impaired, in the present study, the TS group obtained a significantly better performance in the mental component summary compared to the control group. It also obtained better performance in the role physical, bodily pain, general health, social function, and role emotional domains.

When analyzing the physical and mental component summaries of the SF-36, Taback and Van Vliet (2011) evaluated the interference of rhGH use in the HRQoL and compared these results with data on the general population of Canadian women. TS groups that used and did not use rhGH scored similarly to the general population of Canadian women, considering data from the physical and mental component summaries in the SF-36. This result is not similar to that found in the present study, in which the TS group presented similar scores in the physical component summary, but significantly better scores in the mental component summary in relation to the control group.

Considering the SF-36 domains, studies regarding the use of SF-36 to assess QoL for TS patients have found different results; therefore, it is important to separate the analysis by specific domains.

Regarding the physical function domain, two studies found a similar score for the TS group in relation to the data of the comparison group, as well as in the present study. Bannink, Raat, Mulder, and de Muinck Keizer-Schrama (2006) compared data from the TS group in relation to the general Dutch population and Carel et al. (2005) compared the TS group data in relation to the French female population.

However, two studies reported that women with TS had a significantly worse score in the physical function domain in relation to the data used for comparison. Naess et al. (2010) compared TS patients in relation to a control group and Nadeem and Roche (2014) compared data from TS patients in relation to the general female Canadian population.

Four studies reported that patients with TS had similar scores in the role physical domain in relation to the group used for comparison. Naess et al. (2010) performed this comparison in relation to the control group and Nadeem and Roche (2014), Bannink et al. (2006), and Carel et al. (2005) in relation to, respectively, the general Canadian, Dutch, and French female population. These results were different from those found in the present study where the patients had better scores than the control group.

Considering the bodily pain domain, three studies found a similar score between the TS group and the control group (Naess et al., 2010) or data from the general Canadian (Nadeem & Roche, 2014) and French (Carel et al., 2005) female population. Bannink et al. (2006) reported better scores of women with TS compared to data from the general Dutch female population, a result similar to the present study.

In three studies, the general health domain was considered with a similar score between the TS group and the general Canadian (Nadeem & Roche, 2014), Dutch (Bannink et al., 2006), and French (Carel et al., 2005) female population. The study by Naess et al. (2010) reported worse scores of women with TS compared to the control group. None of the articles reported similar scores to the present study, in which the TS group had better scores in the general health domain compared to the control group.

Regarding the vitality domain, Naess et al. (2010), Nadeem and Roche (2014), Bannink et al. (2006), and Carel et al. (2005) considered the score between the TS group similar to the control group and the general Canadian, Dutch, and French female population, respectively. These results are in accordance with the present research, which also found similarity between TS and control groups in the vitality domain. Three studies considered the scores in the social function domain of the TS group similar to the control group (Naess et al., 2010) or to the general Canadian (Nadeem & Roche, 2014) and French (Carel et al., 2005) female population. Only Bannink et al. (2006) found similar results to the present study, reporting that patients found scores significantly better in the social function domain than the general Dutch female population.

Regarding the role emotional domain, three studies considered the scores of the TS group similar to the control group (Naess et al., 2010) or general Canadian (Nadeem & Roche, 2014) and French (Carel et al., 2005) female population. However, Bannink et al. (2006) found similar results to the present study, reporting that patients found scores significantly better in the role emotional domain than the general Dutch female population.

The studies that analyzed the mental health domain found statistical similarity between TS and control groups (Naess et al., 2010) or general Canadian (Nadeem & Roche, 2014), Dutch (Bannink et al., 2006), and French (Carel et al., 2005) female population, similarly to the present study.

In general, there are major controversies in the literature regarding QoL in TS: it is believed that QoL appears to be impaired in these patients in at least one aspect (Reis et al., 2018).

The results of the TS group in relation to HRQoL were higher in relation to the control group. Some data of this study were not expected, such as the fact that TS patients presented better scores in the role physical, bodily pain, general health, social function, and role emotional domains and in the mental component summary of SF-36.

Naess et al. (2010) also reported that patients with TS seemed to have a good adaptation to medical, social, and emotional difficulties, showing satisfaction with their financial and leisure conditions, besides a good relationship with their professional life.

According to Bannink et al. (2006), the physical health and emotional condition of patients with TS did not lead to social difficulties, possibly due to the fact that women with TS do not feel different from other women as adults.

Mccauley, Feuillan, Kushner, and Ross (2001) compared the psychosocial development of 122 adolescents with TS in relation to a control group with 108 adolescents without chronic diseases, through the application of questionnaires with participants of each group and with their mothers. The authors used the term “coping” to refer to a possible pattern of adaptation developed by patients to deal with their physical, cognitive, and emotional difficulties, because in the

questionnaires, the self-report of patients in relation to their clinical symptoms was better than the ones considered by their mothers.

Folkman and Lazarus (1980) defined the term "coping" as cognitive and behavioral efforts made by the individual, resulting from their interaction with the environment, so that it is possible to live and deal with stressful situations and reduce internal and external conflicts and demands. Yasmeen, Khan, Jamshaid, and Salman (2015) reported that through coping strategies people with chronic diseases are able to deal with difficulties and eliminate problems, learning new skills and making adverse situations more satisfying.

It is important to mention that the demonstration of coping strategies by this group of TS patients is a possibility considered by the authors, which cannot be confirmed by this study and that the results of this study were unexpected, especially due to the well-known high incidence of morbidity in TS patients. Therefore, the authors consider that it is extremely important to continue analyzing these issues in other populations of Brazilian TS patients in future studies with a bigger group of patients for external validity.

Another reason to analyze QoL aspects of TS patients in more detail and using a bigger population, is that sample size should be mentioned as a limitation of this study, because the number of TS patients in this study is smaller than expected considering the estimated incidence of these patients and the number of inhabitants of the cities this Clinical Hospital is a reference for. However, the authors believe that this is due to the fact that not all female TS patients were included in this study because of the selected age range and the fact that the study took place in Public University Clinical Hospital, and there is a number of TS patients in this area that use private practice.

Moreover, the fact that it was not possible to analyze a broader range of outcomes to assess all the aspects that might involve the QoL of TS patients is also a limitation of this study. The authors also consider that for future studies it would be important to analyze information from people who are close to these patients and achieve objective results regarding a broader range of QoL aspects in Brazilian TS patients.

5 | CONCLUSION

This group of Brazilian TS patients had better scores in some domains of the SF-36 compared to the control group. There are many aspects that may have contributed to this factor and one of the possibilities is that these patients have good coping strategies. This possibility needs to be better understood by health professionals involved with women with TS and should be better analyzed and evaluated in future studies.

It is also important to mention that the HRQoL of the TS group may have been positively influenced by the fact that this group consists of patients who have a periodical follow-up with the same service and by the same team, since all the patients attended the service since their diagnosis.

Besides, it is necessary to carry out more studies that evaluate in detail the influence of the aspects related to height in the HRQoL in TS and also to verify which other factors really influence the HRQoL of these patients.

In this study, besides the need to analyze a bigger number of patients with broader assessments, an important gap was found in the current literature, in which the instruments used to assess QoL in TS are the same ones developed for chronic diseases in general, therefore, there is not yet a questionnaire that specifically assesses QoL in TS. Therefore, it is crucial to create an instrument specifically for these patients and their main characteristics, and thus enable a better understanding of what factors influence their QoL.

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CONFLICT OF INTEREST

All authors declare that they have no conflict of interest.

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