International Journal of Pediatric Otorhinolaryngology (2009) **73**, 57—65

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International Journal of Pediatric Otorhinolaryngology

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Surgical management of otologic disease in pediatric patients with Turner syndrome☆

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Received 2 July 2008; received in revised form 4 September 2008; accepted 9 September 2008
Available online 20 November 2008

KEYWORDS
Turner syndrome;
Otologic disease;
Surgery;
Surgical management;
Otitis media;
Turner

Summary

Objectives: Previous reports suggest that patients with Turner syndrome have a predisposition for acute and chronic otitis media. However, the role of early or aggressive surgical management of otologic disease has not been explored in the pediatric population. With respect to otitis media in pediatric Turner syndrome patients, we examined (1) the impact of timing and (2) the type of surgical intervention in the treatment of disease.
Methods: Retrospective 10-year review of patients with Turner syndrome and chronic otitis and its surgical management at a single pediatric tertiary institution.
Results: One hundred and seventy-eight patients with Turner syndrome were evaluated at our institution from 1997 to 2007. Thirty-two (18.0%) were diagnosed with middle ear disease. Eighteen (10.1%) were referred to otolaryngology for evaluation. Average age at presentation was 4.7 years (range: 1 month to 12 years). The 18 patients referred to otolaryngology required a mean of 16 clinic visits each for otologic symptoms. A mean of 6.7 pressure equalization tubes (PET) were required per patient (range: 0—25). Middle ear effusions ($n = 14$, 78%) along with tympanic membrane retractions and/or perforations ($n = 10$, 55.6%) were the most common otoscopic findings. Patients with tympanic membrane retractions (8/18) required a higher average number of PET (9.1) and cumulatively underwent a total of five tympanoplasty-type procedures. Six ears had evidence of cholesteatoma. Two patients underwent myringoplasty, 6 patients underwent tympanoplasty (33.3%, mean age 11.6 years), and 3 patients (16.7%, mean age 9.4 years) underwent tympanomastoidectomy. Revision procedures were common. Older age at first PET placement was significantly correlated with the need for later tympanoplasty and/or tympanomastoidectomy operations ($p < 0.036$). Tympanoplasty or tympanomastoidectomy patients had their first PET placed on average at

☆ Presented at the Society for Ear, Nose and Throat Advances in Children, Milwaukee, WI, December 2007.

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0165-5876/$ — see front matter
doi:10.1016/j.ijporl.2008.09.022

5.2 years as compared to 2.6 years in those not requiring tympanoplasty or tympanomastoidectomy operations.
Conclusions: Recurrent and chronic otitis media is common in patients with Turner syndrome. Once established, disease is recalcitrant and leads to multiple surgical procedures. Early PET insertion is advocated to offset the future necessity of more extensive tympanic procedures.

1. Introduction

Turner syndrome (TS) is a rare disorder arising from a monosomy 45, X karyotype. TS may also result from mosaicism of complete or partial X chromosome deletions. Phenotypic expression is variable but typical features include webbing of the neck, gonadal dysgenesis, coarctation of the aorta, short stature, high-arched palate, and renal anomalies [1]. Although less frequently reported, patients with TS have a high incidence of eustachian tube dysfunction and otitis media [2]. Ear disease in adult patients with TS is complicated by progressive sensorineural hearing loss, temporal bone abnormalities, and a need for multiple procedures [1—3]. The impact of otologic disease in patients with TS has been substantiated by several reports illustrating progressive and long-term otologic manifestations of the syndrome [1,2,4]. Nonetheless, specific examination of the surgical management of otologic disease in pediatric TS patients has not been performed. The role of early surgical intervention of otitis media has not been explored in this population. Understanding the purpose, incidence, and level of otologic surgery required in these patients is important so that interventions can be designed with respect to the natural history of ear disease in these patients and their current status. The objective of this study was to examine a single pediatric institution's experience with otologic disease in children with TS. Attention was placed on the impact of timing and the type of surgical intervention performed for middle ear disease.

2. Materials and methods

This study was approved by the Institutional Review Board at Cincinnati Children's Hospital Medical Center (CCHMC). A retrospective database query was performed on patients with TS seen at CCHMC. Patients with TS visiting CCHMC between 1997 and 2007 were identified based upon the ICD-9-CM diagnostic code. These patients were screened for diagnostic codes (ICD-9-CM) for every type of middle ear disease. Specific diagnostic codes included chronic and acute otitis media with or without effusion (purulent, serous, secretory, and mucoid) and otitis media with cholesteatoma. These patients were thereby selected for retrospective chart review.

The charts of patients who were evaluated in the otolaryngology clinic were analyzed for age at presentation, number of clinic visits, audiometric results, disease symptoms, temporal bone computed tomography (CT) scan findings, type and number of operative procedures, follow-up findings, associated ear disease, complications from treatment, and related otologic diagnoses. The number of pressure equalization tubes required per patient was noted and the average number of tubes placed per patient was calculated. Pressure equalization tubes placed at our institution as well as outside institutions (when this data was available) were included in this analysis. Otoscopic findings that were noted in each chart (including middle ear effusions, tympanic membrane retractions, and perforations) were also noted. Otoscopic findings in each patient were correlated with the number of pressure equalization tubes (PETs) placed per patient and tympanoplasty and/or tympanomastoidectomy procedures. The number of patients with cholesteatoma was determined from chart review of physical examination findings and CT scanning. Operative reports were analyzed for procedure, surgical approach, and middle ear findings. Patient data at the time of each individual operation was utilized to calculate the mean patient age for each procedure, average time from presentation in clinic to surgical management, and the number of patients requiring each specific intervention. The surgical course and progression was outlined for each patient. Specific characteristics (including age at first PET placement) of patients requiring tympanoplasty or tympanomastoidectomy were examined. Surgical procedures in these patients were aimed to repair retractions and perforations or to remove cholesteatoma. Analysis of clinic appointments included review of audiometric results and otoscopic and microscopic examinations. Pure-tone averages (PTAs) were calculated at the thresholds of 500, 1000, 2000, and 4000 Hz according to AAO-HNS reporting criteria for 4-tone PTA. The nature of the hearing loss (sensorineural, mixed hearing, etc.) was explored.

3. Results

A total of 178 patients with TS were evaluated for management of their disease and associated abnormalities at our institution from 1997 to 2007. Exploration of all ICD-9 diagnostic codes for acute and chronic otitis media revealed that 32 patients (18.0%) had middle ear disease at any one clinic visit. Eighteen patients (10.1%) were referred to otolaryngology for evaluation. Each of these patients underwent some form of surgical intervention for their middle ear disease. The mean age at presentation was 4.7 years (range: 1 month to 12 years), with an overall average of 16 clinic visits per patient (range: 2—47). Generally, more severe middle ear disease (i.e. cholesteatoma) translated into a greater number of patient clinic visits. Specifically, children diagnosed with cholesteatoma were seen a mean of 25.3 times. Patients diagnosed with chronic otitis media alone required the fewest number of visits (average: 4.0). A 5 year old who underwent a tympanomastoidectomy for cholesteatoma and subsequent paper patch myringoplasty was the most frequently seen patient (47 clinic visits).

3.1. Exam

Table 1 illustrates the otologic findings in children with TS referred for evaluation to otolaryngology. Middle ear effusions ($n = 14$), tympanic membrane pathology (retractions [$n = 8$] and perforations [$n = 5$]), and otorrhea ($n = 9$) were the most common abnormalities identified on otoscopic examination. Sixteen patients (88.9%) presented with bilateral ear disease while 2 had unilateral otitis media. Eight of the 10 patients with reported chronic tympanic membrane retractions ($n = 3$) or perforations ($n = 5$) were diagnosed prior to presentation to otolaryngology. Three children had evidence of serous and bloody otorrhea. Six patients (33.3%) were found to have cholesteatoma. This was found on physical examination in 4 patients with the extent of disease clarified by CT scanning of the temporal bones. Two other patients were diagnosed with having cholesteatoma based upon erosive changes or discrete findings on CT scans.

Table 1 Ear findings by physical examination during clinic visits of pediatric patients with Turner syndrome referred for otologic surgical management.

Findings	No. of patients
Middle ear effusion	14 (78%)
Otorrhea	9 (50%)
Retraction	8 (44%)
Cholesteatoma	6 (33%)
Perforation	5 (28%)
Bloody drainage	4 (22%)
Myringosclerosis	2 (11%)
External ear abnormalities	2 (11%)
Palate abnormalities	2 (11%)
External otitis	1 (5.6%)

3.2. Testing

Preoperative audiometric results revealed mild to moderate conductive hearing loss in the affected ear of most patients. However, 4 patients (22.2%) had evidence of sensorineural deficits including 3 with mixed and 1 with isolated sensorineural hearing loss. Six patients (33.3%) had high-resolution computed tomography scans of the temporal bone and middle ear to evaluate for hearing loss, cholesteatoma, or tympanic membrane pathology. No abnormalities of the otic capsule or ossicular chain were discovered on these scans. However, findings consistent with cholesteatomas were confirmed in these 6 patients. Additional abnormalities in the tympanic membrane (inflammation, retraction, perforation) were noted by CT in 5 of these patients.

3.3. Surgery

In total, 17 children had placement of PET, 6 required tympanoplasties, 3 were managed with tympanomastoidectomies, and 2 underwent myringoplasties. All 18 children with TS had problematic middle ear disease (chronic or acute otitis media). One patient did not receive PET but underwent bilateral myringotomies. The average time from presentation to surgical management was 2.2 clinic visits. Overall, a mean of 6.7 PET procedures were performed per patient (range: 0—25 PET). This included the reported number of PET placed prior to presentation to our facility (5 patients, mean 7.2 PET) and those placed at our institution (range: 0—10 PET). One patient with 25 PET placed had 20 placed prior to being seen in our clinic. This patient eventually underwent a type I tympanoplasty. Patients with retractions (8/18) required a higher average number of PET (9.1) and a total of five tympanoplasty procedures in our cohort.

Patients that did not require tympanoplasty or tympanomastoidectomy had an average of 5.9 PET inserted. In this sub-population, patients received their first set of tubes at an average of 2.6 years of age. In contrast, children who underwent tympanoplasty had tubes first placed at 3.9 years of age while patients who underwent tympanomastoidectomy had tubes first placed at 6.8 years of age. Cumulatively, patients who underwent

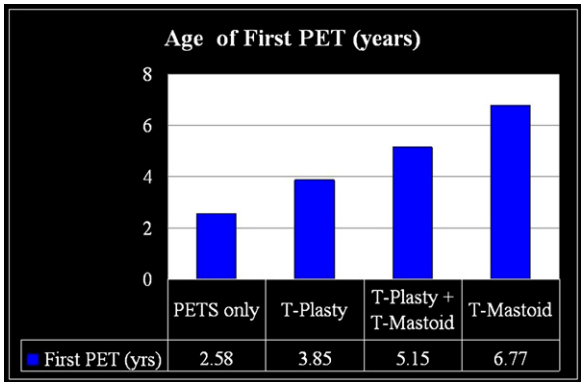

Fig. 1 Age of placement of first pressure equalization tube (PET) in patients with Turner syndrome for treatment of otitis media with PET alone or those undergoing more extensive tympanic procedures. A significant difference in age at first tube is noted in Turner syndrome patients who required tympanoplasty or tympanomastoidectomy (5.2 years) versus those who had only PET placed (2.6 years). This suggests an impact of the time of first surgical intervention on the disease course (T-plasty/mastoid: tympanoplasty or tympanomastoidectomy; %: percent; Dx: diagnosis; pts: patients).

tympanoplasty and/or tympanomastoidectomy had tubes first placed at 5.2 years of age (Fig. 1).

Six children with TS (33.3%, average age 11.6 years) underwent tympanoplasty at our institution over the past 10 years (Table 2). Those who underwent tympanoplasty had an average of 6.8 PET prior to tympanoplasty. Of the 7 tympanoplasty procedures performed on these 6 children (Table 3), 6 incorporated temporalis fascia grafts and 1 used a tragal cartilage graft. Two myringoplasties were performed, one on a patient who had undergone prior tympanoplasty and another on a patient who had also undergone a prior tympanomastoidectomy. Of those patients who underwent tympanoplasty, 3 were discovered to have cholesteatoma during the procedure. A mean 4.9 dB (decibel) closure of their air-bone gap was observed.

Three patients (16.7%, average age 9.4 years) and a total of 4 ears required tympanomastoidectomy for cholesteatoma (Table 2). Children with TS who underwent tympanomastoidectomy received an average 3.3 pressure PET prior to their procedure. Of the 7 tympanomastoidectomies performed on these 3 patients, 1 included a canal wall down approach. The same patient had a bone anchored hearing aid (BAHA) inserted at the time of canal wall down tympanomastoidectomy. Ossicular chain reconstructions were not performed in any patient. One patient required tympanomastoidectomy in both ears along with three revision tympanomastoidectomies.

Middle ear abnormalities noted during the tympanomastoidectomies included cholesteatoma in the mastoid antrum, the middle ear cleft, and in the mastoid bowl, granulation tissue in the hypotympanum and the antrum, and two eroded incus bones. Canaloplasty was performed in 2 patients. Also note that patients numbered 3 and 6 are listed in both Tables 2 and 3 as they each required a tympanoplasty and tympanomastoidectomy.

An average 3.8 dB increase in their air-bone gap was observed. Postoperative PTAs showed a marked decline in conductive hearing of ear number 6 and number 7 (opposite ears of same patient). Following an initial left tympanomastoidectomy, this patient underwent a contralateral tympanomastoidectomy and 3 revision tympanomastoidectomies that showed large cholesteatomas or granulomas in the middle ear upon each operation. One patient (patient 9 in Table 3) awaits tympanomastoidectomy for cholesteatoma at the time of this study. Her history is significant for tympanic membrane retraction and placement of 8 pressure equalizing tubes.

The surgical treatments and outcomes of all 9 patients who required tympanoplasty or tympanomastoidectomy are outlined in Table 3. Two patients required tympanomastoidectomy after tympanoplasty for evidence of cholesteatoma. The patient listed as number 6 (left ear) and 7 (right ear) required left tympanomastoidectomy, left tympanoplasty, right tympanomastoidectomy, and 3 revision tympanomastoidectomies due to recurrent or recidivistic disease.

4. Discussion

Turner syndrome is a rare disease with known genotypic and phenotypic variability. Along with a constellation of other features, otologic abnormalities have been described in this patient population. These factors also appear to follow a pattern of variable expression with an unpredictable rate of progressive sensorineural hearing loss, temporal bone and soft tissue aberrancies, and otitis media. Regardless, a predisposition for acute and chronic otitis media and sensorineural hearing loss has been clearly established [2,4—7]. In the literature, incidences range from 48 to 100% for recurrent middle ear infections and 11—67% for sensorineural hearing loss for patient populations up 42 years of age [11].

In this study we report a 10-year otologic experience with pediatric patients with TS at a single institution. A population of 178 patients with TS was identified with 18% (n = 32) evaluated for middle ear infections by pediatricians. Ten percent were ultimately referred for otologic evaluation by pediatric otolaryngologists. The most common manifestations of surgical disease were middle ear

Table 2 Pediatric patients with Turner syndrome over a 10-year period at a single institution requiring extensive tympanic procedures. Preoperative and postoperative audiometric findings are shown. Six patients required tympanoplasty (mean age: 11.6 Yr). Three patients underwent tympanomastoidectomy (mean age: 9.4 Yr). Patients 6 and 7 are the same patient with opposite ear procedures (Yr: year; Mos: months; A: air conduction; B: bone conduction; N/A: not available; CHL: conductive hearing loss; PTA: pure tone average; Preop: preoperative; Postop: postoperative; PETs: pressure equalization tubes).

Tympanoplasty

Patient/ear	Age	Preop PTA	Postop PTA	Otic findings
1/Right	11 Yr	A: 8.8 B: 2.5	A: 8.8 B: N/A	Cholesteatoma Perforation
2/Right	11.7 Yr	A: 32.5 B: 5	A: 31.7 B: 8.8	Cholesteatoma Otorrhea Perforation Retraction
3/Left	5.8 Yr	A: 42.5 B: 8.8	A: 40 B: 10	Cholesteatoma Retraction
4/Right	15.6 Yr	A: 51.3 B: 17.5	A: 30 B: 6.3	Perforation Cholesteatoma Retraction
5/Right	15 Yr	A: 8.8 B: N/A	A: 13.8 B: 5	Bloody otorrhea Myringosclerosis Perforation Retraction
6/Left	10.5 Yr	A: 38.8 B: 6.3	A: 40 B: 8.8	Bloody otorrhea Cholesteatoma Perforation Retraction

Tympanomastoidectomy

Patient/ear	Age	Preop PTA	Postop PTA	Otic findings
3/Left	9.3 Yr	A: 48.8 B: 7.5	A: 23.8 B: 2.5	Cholesteatoma Retraction
6/ Left	10.3 Yr	A: 53.8 B: 13.8	A: 70 B: 13.8	Bloody otorrhea Cholesteatoma Perforation Retraction
7/Right	11.9 Yr	A: 32.5 B: 3.8	A: 53.8 B: 10	Cholesteatoma Chronic otitis media Retraction
8/Left	6.2 Yr	A: 7.5 B: N/A	A: 18.8 B: N/A	Bloody otorrhea Cholesteatoma Perforation Retraction

effusions, tympanic membrane abnormalities (perforations and retractions), and otorrhea. These findings support the previously suggested middle ear and eustachian tube problems reported in TS children. More importantly, this study illustrates the increased need for otologic operations (10.1%) in the treatment of chronic otitis in pediatric patients with TS. Previous reports approximate that 4% of children in the general population will require pressure equalization tubes [8,9]. A well-designed prospective study to explore the incidence of all patients undergoing otologic intervention at our institution would better delineate this difference but was beyond the scope of this study. Nonetheless it was apparent that once chronic disease was established in a Turner syndrome child, the likelihood of otolaryngologic interventions was high. For example, over 50% of the patients with TS with a diagnosis of middle ear disease at our institution ultimately required PET placement (18/32). In

Table 3 Surgical otologic management beyond tympanostomy and tube insertion in 9 pediatric Turner syndrome patients identified at our institution over a 10-year period. Patients 6 and 7 reflect the same patient with opposite ear procedures. Multiple interventions were required as reflected by two or more procedures required in 5 of these patients.

Patient	Ear	1st operation/age	2nd operation/age	3rd operation/age	4th operation/age
1	Right	Tympanoplasty with temporalis fascia graft/11 Yr	—	—	—
2	Right	Transcanal type I tympanoplasty/11.7 Yr	Fat myringoplasty/13.3 Yr	—	—
3	Left	Tympanoplasty with ossicular reconstruction/5.8 Yr	Tympanomastoidectomy/9.3 Yr	Tympanoplasty with ossicular reconstruction/9.8 Yr	—
4	Right	Tympanoplasty/15.6 Yr	—	—	—
5	Right	Type I tympanoplasty with temporalis fascia graft/15 Yr	—	—	—
6	Left	Tympanomastoidectomy with canaloplasty/10.3 Yr	Tympanoplasty with cartilage graft/10.5 Yr	Revision tympanomastoidectomy with canaloplasty/10.8 Yr	Revision modified radical tympanomastoidectomy/ 12.8 Yr
7	Right	Tympanomastoidectomy/ BAHA/11.9 Yr	Revision canal wall down tympanomastoidectomy/14.6 Yr	—	—
8	Left	Tympanomastoidectomy/6.2 Yr	Paper patch myringoplasty (bilateral)/6.7 Yr	—	—
9	Left	Tympanomastoidectomy pending	—	—	—

Procedure	% of all Turner Pts Seen	% Turner Pts with Otitis Dx	% Turner pts referred to Otology
PET	9.6%	53.1%	94.4%
T-plasty/mastoid	5.1%	28.1%	50%

Fig. 2 Incidence of patients with Turner syndrome receiving pressure equalization tubes (PETs) or undergoing extensive tympanic procedures as a reflection of the total number of patients with Turner syndrome seen at our institution, those with otitis diagnosis, and those referred for surgical evaluation (T-plasty/mastoid: tympanoplasty or tympanomastoidectomy; %: percent; Dx: diagnosis; pts: patients).

essence, once TS patients present with recurrent and chronic ear problems, these patients should be selected into a higher maintenance group. It remains important, however, that medical options are exhausted prior to surgical intervention.

Expressed as the total number of patients with TS seen at our institution, 9.6% (17/178) required tube placement while 5.1% (9/178) required either a tympanoplasty or tympanomastoidectomy (Fig. 2). Sculerati et al. documented that 50% of TS patients required myringotomy and pressure equalization tubes while 16% underwent tympanoplasty or tympanomastoidectomy [5]. The difference in our study compared to others may reflect a difference in patient populations examined. Wherein otolaryngology patients with TS were the base populations in previous studies, we extracted information from a central population of children with TS. This may also be indicative of the phenotypic variability in middle ear disease in the TS population. Finally, a direct comparison of our population with other studies is difficult since our average patient age (4.7 years) is much younger than in previous reports and examines an age group that has not been specifically discussed in the past. As the TS population ages, it is likely that the prevalence of middle ear disease increases as well.

When expressed as the number of patients with TS seen for ear infections in our group, 53.1% underwent surgical intervention while 28.1% (9/32) underwent a tympanic or mastoid procedure (Fig. 2). If we examine the patients referred to otolaryngology alone, 100% required some form of middle ear intervention with 50% requiring tympanoplasty or tympanomastoidectomy. This result underscores the propensity for chronic ear disease in TS patients presenting with otitis to a surgical clinic. Patients with TS have a tendency to require multiple procedures when surgical otologic disease is present [5]. This is further illustrated by an average 6.7 PET per patient, seven tympanoplasties, and seven tympanomastoidectomies (including three revision procedures) performed in this study. As seen in Table 3, revision procedures were common and reflect the recalcitrant nature of disease in these patients. Of note, patients with retractions (8/18) required a higher average number of PET (9.1) and a total of five tympanoplasty procedures in our cohort.

Perhaps the most critical finding in this study was the large difference in the age of first PET placement in pediatric TS patients who underwent tympanoplasty or tympanomastoidectomy (5.2 years) versus those who had only PET placed (2.6 years). The p value for a t-test performed on these data was determined to be 0.2162. However, closer examination of the data set for the PET only group revealed an outlier who had tubes placed at 141 months while the other patients had their PET initially placed at 6, 8, 8, 12, 13, 26, and 34 months. A Q-test (used to identify outliers in a data set) performed on this data set allows one to exclude the 141 months outlier at a 99% confidence level ($0.793 > 0.634$, type I error <0.01). If one excludes this outlier, the p value then becomes 0.0364. With a small data set, rejecting even one value can be problematic and warrants consideration. When these two data sets are viewed comparatively, however, the trend remains that tympanoplasty and tympanomastoidectomy patients had PET placed at a much later age than those who had PET and did not require tympanoplasty or tympanomastoidectomy.

One must be cognizant of the fact that when a patient undergoes a tympanomastoidectomy, it is not always a direct result of progressive disease which failed earlier surgical procedures. For example, if a patient presents with an obvious cholesteatoma, then a tympanomastoidectomy would be performed without tympanoplasty. Thus, the surgical course for TS patients is not always step-wise. However, the aforementioned trend suggests that patients with TS should undergo early tympanostomy and PET insertion to offset the risk of progressive disease that could necessitate future tympanoplasty or tympanomastoidectomy. This is similar to cleft palate patients with known midfacial abnormalities contributing to chronic eustachian tube dysfunction. Although the mechanism of chronic middle ear disease is less clear in TS, our results illustrate a functional problem in clearing persistent middle ear disease once it has been established. Otologic management of patients with TS with known disease may thereby need to reflect the practice in patients with cleft palates [10]. Similarly, intervention with long lasting T-tubes may be appropriate with recurrent disease.

Standard PET can be placed with a low threshold for subsequent T-tube insertion in cases of recurrent or recalcitrant otitis media.

The etiology of otologic disease in TS patients is currently under investigation. Several authors have asserted that middle ear and temporal bone abnormalities facilitate the development of ear problems in patients with TS [1,11]. Specifically, Windle-Taylor et al. described a Mondini malformation with apical dilatation in the basal turn of the cochlea leading to middle ear obstruction [11]. Interestingly, we discovered no structural abnormalities within the temporal bone in 6 patient undergoing CT scans. Moreover, middle ear malformations were not present among those undergoing either tympanoplasty or tympanomastoidectomy. Although this is a small population of TS children, this suggests another etiology for middle ear disease beyond middle ear abnormalities. Several authors have noted abnormalities in the orientation of the external auditory canal and the development of the first branchial arch [4,12]. Two patients in the current series underwent canaloplasty but this does not necessarily reflect rare canal deformities. Phenotypic variation in midfacial abnormalities such as cleft or high-arched palates seen in patients with TS could contribute to abnormal anatomy of the eustachian tube and lead to the higher incidence of middle ear disease in these patients. Nonetheless, a final proposed etiology includes a X-linked hormonal difference in TS patients impacting the function of the eustachian tube compared to individuals without the disorder [3,13—16].

It is not entirely clear from a retrospective analysis whether pediatric TS patients who require tympanoplasty or tympanomastoidectomy are the result of progressive underlying disease that may have been circumvented by surgical intervention earlier in the disease course or the result of more aggressive disease when presenting later in childhood. Regardless of the etiology, this study suggests that pediatric TS patients with otitis media have a greater predisposition for the development of chronic and recalcitrant otologic disease. These patients are best managed with early surgical intervention (i.e. earlier PET placement) relative to the general population in order to decrease their risk of more severe otologic disease. Future studies with a prospective design, larger patient sample size, and multicenter collaboration would help to answer the aforementioned question and would ultimately elucidate the full benefit of early PET placement in pediatric TS patients. Unfortunately, such analysis may be difficult to perform when one considers the rarity of Turner syndrome in the population.

5. Conclusions

- Recurrent and chronic otitis media are more frequently encountered in TS patients than in the general population.
- Recalcitrant and chronic otitis media requiring multiple procedures is often present once disease is established in this patient population.
- 10.1% of all TS patients in our analysis required surgical intervention for recurrent or chronic otitis media.
- 5.1% of all TS patients in our analysis required either a tympanoplasty or tympanomastoidectomy.
- 53.1% of TS patients diagnosed with otitis media in our analysis required surgical management involving PET placement or more extensive procedures.
- Once surgical otologic disease is established in pediatric TS patients, parents and practitioners need to be aware that multiple procedures will likely be required to achieve clinically silent disease.
- Early surgical intervention with rapid progression to T-tube insertion, as seen in patient populations with recalcitrant middle ear disease and eustachian tube dysfunction, is advocated in patients with TS with chronic otitis media.
- The high incidence of SNHL with late progression advocates early and frequent hearing tests in TS patients into adult life.

Conflict of interest

None declared.

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