

## Middle Ear Cholesteatoma Prevalence in Children with Turner Syndrome (TS)

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### DESCRIPTION

An anomalous group of keratinized squamous epithelial cells in the middle ear known as a cholesteatoma can be locally invasive, cause facial paralysis, hearing loss, and bacterial infections of the central nervous system. With a small male predominance of 1.4:1, the estimated prevalence of middle ear cholesteatoma in youngsters is 0.03-0.15%. The risk factors for middle ear cholesteatoma should be known by doctors since clinical monitoring can identify them early and enable definitive surgical removal before bone erosion or local invasion takes place. Cleft palate has a 200 times higher risk than normal and Down syndrome has a 7 times higher risk than normal for middle ear Cholesteatoma [1].

The most prevalent sex chromosomal abnormality in females, Turner Syndrome (TS) is defined by a missing or altered second X chromosome and affects 1 in 2500 female infants. Numerous developmental anomalies, including as congenital heart defects, short stature, a wide neck, and scoliosis, are linked to TS. Additionally, people with TS are more likely to experience hearing loss, external ear abnormalities, and certain otologic problems such recurring middle ear infections. Patients with TS frequently experience chronic otitis media, which greatly contributes to the high rate of tympanostomy tube placement procedures in this population, which ranges from 9.6% to 32% [2,3].

Seven variations of cholesteatoma were found in a 2014 study that looked at 179 individuals with TS. This resulted in a prevalence of 3.9%, which is much higher than the reported prevalence in the general population of 0.03%-0.15% but was severely constrained by sample size. Although even more constrained by sample size, a second widely regarded study that detected 3 occurrences of cholesteatoma among 60 children with TS gave a prevalence of 5%. To our knowledge, there hasn't been a sizable population-level examination evaluating the occurrence and relative risk of middle ear cholesteatoma in kids with Turner Syndrome [4].

In order to compare risks between the TS group and a large control sample of kids without TS, the prevalence of middle ear cholesteatoma will also be determined. The presence of cleft

palate, one of the most significant risk factors for cholesteatoma and a potential contributory condition, will be checked in the patient group that includes children with TS as a secondary outcome. The obvious therapeutic relevance as it will inform otologic care for children with TS by indicating the importance of vigilant and frequent monitoring for cholesteatoma in this cohort [5]. Based on the known elevated risk for middle ear infection and smaller-scale studies looking at cholesteatoma in TS patients, a significantly higher risk for cholesteatoma among children with TS compared to children without TS.

### CONCLUSION

Middle ear cholesteatoma is an abnormal collection of keratinized squamous epithelial cells within the middle ear. It can be locally invasive and potentially lead to hearing loss, facial paralysis, and bacterial infection of the central nervous system. Children with TS found to have a 16 times higher rate of middle ear cholesteatoma than children without TS. Middle ear cholesteatoma in children with TS, the largest sample size thus providing reliable evidence for prevalence in this population. Clinical monitoring for cholesteatoma should be especially rigorous and more frequent in this population so definitive surgical intervention can prevent progression to hearing loss.

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