



# AUDITORY PHENOTYPES AND DYNAMICS OF HEARING THRESHOLDS IN 246 TURNER SYNDROME FEMALES



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## Background

The association of otitis media, hearing loss (HL) and Turner Syndrome (TS) was reported in the early 60's, being confirmed by later studies. It is recognized that individuals with TS have a higher incidence of middle ear disease and hearing problems than non-TS subjects. The associated hearing impairment has been described as both conductive and sensorineural, indicating both middle and inner ear involvement.

## Objective

To describe the auditory phenotype and dynamics of hearing thresholds in patients with TS at different ages.

## Materials and Methods

Cross-section study evaluating the hearing thresholds in 246 TS patients (age range 4-44 yrs). Patients were divided into three age groups: Group 1 (79 TS, age range 4.0-12.9 yrs); Group 2 (101 TS, age range 13.0-25.9 yrs,) and Group 3 (66 TS, age range 26.0-44.9 yrs,). Pure tone audiometry (PTA) with evaluation of frequencies ranging from 250 to 12,000 Hz was assessed to define 5 types of audiograms according to HEAR classification: 1. Increasing (hearing loss at low frequencies); 2. U-shaped or Dip (medium frequency hearing loss HL); 3. "Gentle" slope (hearing loss at high frequencies); 4. "steep" slope (hearing loss at high frequencies); 5. Plate.

## Results

- 108 (43.9%) TS females presented HL.
- Percentage of TS HL patients increased with age (Figure 1).
- Only in Group 3, ENT remote pathology was more frequently positive in TS with HL (80% vs 42.9%,  $p = 0.003$ ).
- The degree of HL was not different in the three groups (Figure 2).
- Sensorineural HL (SNHL) significantly increased with age (Figure 3).

Table 1. Distribution of chromosomal alterations among TS patients with or without hearing loss

Karyotype	Total population n. 246	Normoacusic pts n. 138	Hypoacusic pts n. 108	$\chi^2$	p
45,X0	43.9%	43.5%	44.4%	n.s	n.s
Mosaicism	42.3%	43.5%	40.7%	n.s	n.s
Structural alterations	13.8%	13.8%	14.8%	n.s	n.s

Figure 1. Percentage of Turner syndrome patients with hearing loss in different age groups

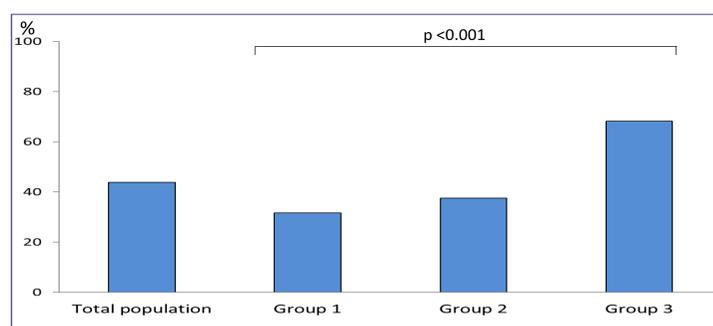


Figure 3. Type of hearing loss in the different age groups

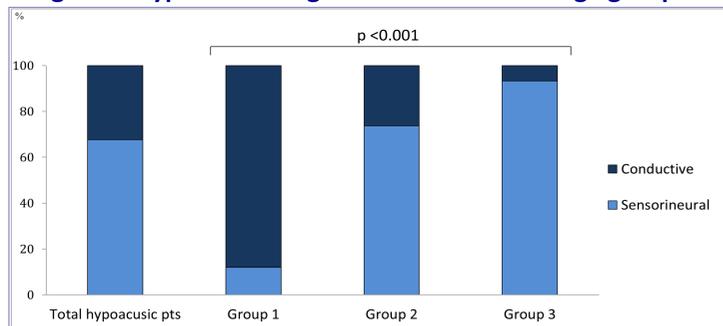
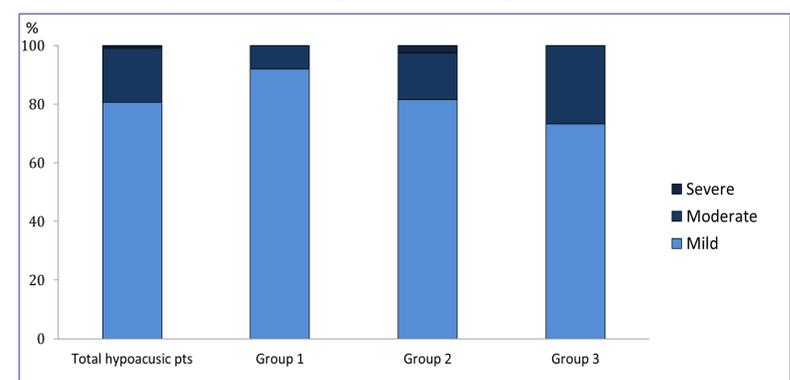


Figure 2. Degree of hearing loss according to HEAR classification



- Type 5 was the more frequent audiogram documented (34.2%), followed by types 2 (28.7%) and 3 (18,5%). Types 1 (6.5%) and 4 (12%) were rarer. Only the frequency of type 5 audiogram significantly decreased with age (Group 1 60.0%, Group 2 31.6%; Group 3 22.2%;  $p < 0.001$ ).
- The univariate and multivariate logistic regression analyses demonstrated that age (Odds 1.62) and a positive ENT remote pathology (Odds 2.007) were significant predictors of deterioration of auditory outcome in TS.

## Conclusions

- ENT remote pathology and age are predictive factors for HL in TS.
- SNHL in TS dramatically increased from the age of 13 yrs.
- Type 5 is the more frequent audiogram in TS under the age of 45.

