LONGITUDINAL EVALUATION OF AUDIOLOGICAL PATTERN IN TURNER SYNDROME

Tommaso Aversa[1], Rocco Bruno[2], Simona Santucci[1], Maria Francesca Messina[1], Simona Borrello[1], Emanuela Scarano[3], Annamaria Perri[3], Federica Tamburrino[3], Mariella Valenzise[1], Angela Alibrandi[4], Laura Mazzanti[3], Malgorzata Wasniewska[1]

1) Pediatric Unit, Department of Human Pathology in Adulthood and Childhood, University of Messina, Messina, Italy; 2) Otorhinolaryngology Unit, Department of Human Pathology in Adulthood and Childhood, University of Messina, Messina Italy; 3) Rare Diseases and Auxological Unit, Department of Pediatrics, Sant'Orsola-Malpighi Hospital, University of Bologna, Bologna, Italy 4) Department of Economics, Unit of Statistics and Mathematical Sciences, University of Messina, Messina, Italy

P3-164

Disclosure statement: nothing to disclose.

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 investigate prognostic markers (age, initial hearing level, karyotype, chronic hormonal therapies, and presence/absence of a mid-frequency dip influence) for HL in TS.

Materials and Methods

DESIGN: Longitudinal cross-sectional and retrospective study.
STUDY POPULATION: 61 TS females (age range 4 - 45 yrs), diagnosed by cytogenetic analysis (49,2% monosomy X, 41% mosaicism and 9.8% structural aberration of X chromosome), 90,2% of them treated with growth hormone (GH), 88.5% underwent pubertal induction ,78.7% presented positive otolaryngological (ENT) remote anamnesis.
METHODS: ENT anamnesis and physical examination, pure tone audiometry (PTA) (frequencies ranging from 0.25 to 12 Hz) were performed. In each patient, at least 2 PTAs were evaluated through 10 years; 1st and 2nd PTAs were performed at median age of 11 yrs (range 4 – 29) and 26 yrs (range 15-45), respectively. The median follow-up period was 13 yrs (range 10-30).

Results

- HL frequency increased from 41 to 59%, sensorineural HL (SNHL) from 18 to 56%, whilst conductive HL (CHL) decreased from 23 to 3%, from 1st and 2nd PTAs (Figure 1).
- In young adult patients SNHL, mainly involved the high frequencies, from 8 to 12 kHz. The mid-frequency dip (2-4 kHz), considered as early and predictive sign of future SNHL, had been pointed out only in patients over 12 years (prevalence 16% for worse ear).
- Eight TS with CHL at 1st PTA, were normoacusis in the 2nd one (Figure 2).
- HL was significantly more common in patients with karyotype 45, X0 (52%) than these with mosaicism (28%) or chromosome X aberrations (20%) (p =0.044).
- The logistic regression detected 2 variables that significantly worsen auditory outcomes in TS: GH therapy (Odds 2.5) and a positive ENT remote pathology (Odds 3.0).

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

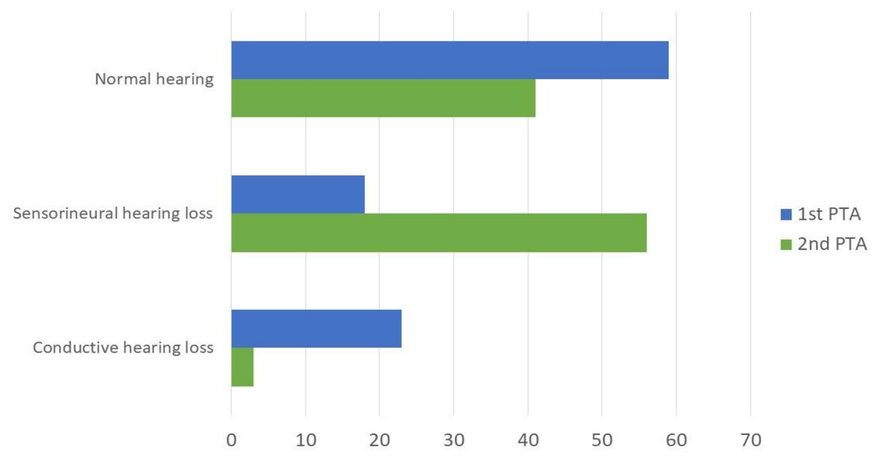
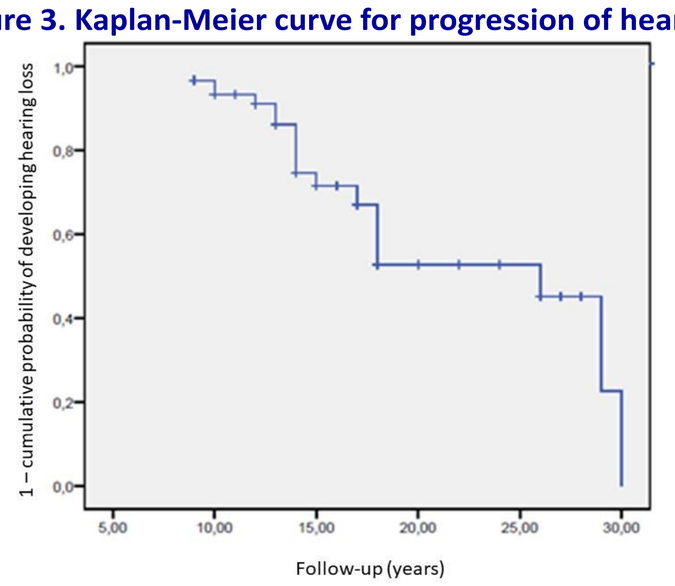
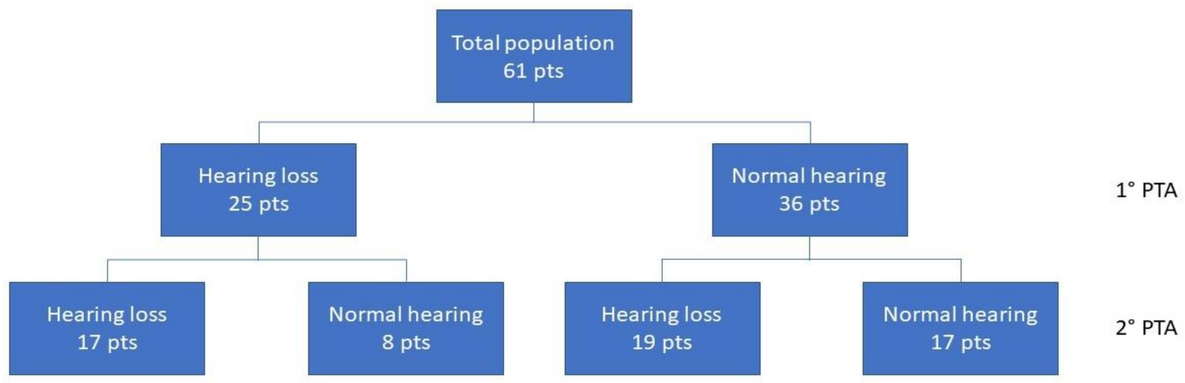


Figure 2. Evolution of hearing loss between first and second pure tone audiometry

Figure 3. Kaplan-Meier curve for progression of hearing loss





•The Kaplan-Meier curve confirmed that the risk of HL progressively increased with age (Figure 3); furthermore, a clear increased probability of HL was observed after 15 years of follow-up.

Conclusions

1) SNHL increased with age; 2) SNHL not always was preceded by a framework of CHL or mid-frequency dip, but it might be preceded by early high frequencies (8-12 kHz) HL; 3) GH therapy and ENT remote pathology resulted significant predictors of HL in TS.