Internal Auditory Canal Stenosis and Congenital Facial Nerve Palsy

Case one

History:
- 12-year-old healthy male referred to the otolaryngology clinic for hearing loss.
- Failed routine school hearing screening and was found to have a profound left SNHL on subsequent audiological evaluation.
- Subject did not recall a time when he could hear from his left ear.
- Unable to close left eye and had facial asymmetry.

Birth history:
- Primarily traumatic in etiology and complete recovery expected in 89-91% of cases.
- No perinatal infections.
- No family history or diabetes mellitus.
- Full-term cesarian section.
- Normal right hearing.

Diagnostics:
- High-resolution computed tomography.
- Magnetic resonance imaging.

Case two

History:
- 5-month-old healthy female referred to the otolaryngology clinic for hearing loss.
- Failed routine school hearing screening and was found to have a profound left SNHL on subsequent audiogram.
- Subject did not recall a time when she could hear from her left ear.
- Unable to close left eye and had facial asymmetry.

Birth history:
- Primarily traumatic in etiology and complete recovery expected in 89-91% of cases.
- No perinatal infections.
- No family history of diabetes mellitus.

Diagnostics:
- High-resolution computed tomography.
- Magnetic resonance imaging.

DISCUSSION

Congenital facial nerve palsy is rarely associated with both facial nerve palsy and bilateral sensorineural hearing loss.

Theories explaining the association of hearing loss with IAC stenosis:
- 1st theory: The IAC cranial nerve is hypoplastic secondary to an abnormal cholesteatotic environment or lack of end organs. The fonic canal is narrow as a result of the thin or absent IAC wall.
- 2nd theory: Facial nerve palsy is a primary defect that inhibits the growth of the IAC cranial nerve.

Major argument against the 2nd theory is that facial nerve function is typically preserved; these two cases counter that argument against the 2nd theory.

IAC stenosis should be considered in the differential diagnosis of developmental facial nerve palsy.

Facial nerve function may be less likely to recover in setting of IAC stenosis.

REFERENCES