Internal Auditory Canal Stenosis and Congenital Facial Nerve Palsy

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INTRODUCTION

Internal Auditory Canal (IAC) Stenosis

- 12% of congenital sensorineural hearing loss (SNHL) is associated with a narrow internal auditory canal (<3mm)[1]
- Typically facial nerve function is preserved in IAC stenosis[2]
- 2-8 mm is the normal range for the IAC[3]

Facial Nerve Paralysis in the Newborn

- Primarily traumatic in etiology and complete recovery expected in 89-91% of cases
- 16-22% of congenital facial nerve palsy is developmental in etiology[4,5]
- Developmental facial nerve paresis typically accompanies other craniofacial abnormalities
  - Möbius syndrome
  - Goldenhar syndrome
  - asymmetric crying facies

METHODS

- **Subjects**
  - Children seen in the otolaryngology clinic at the Children's Hospital of Wisconsin from June 1996 to June 2009 with each of the following findings
    - Congenital facial nerve palsy
    - Congenital sensorineural hearing loss
    - Internal auditory canal stenosis
  - Exclusion criteria
    - Diagnosis of a syndrome or other major congenital abnormalities
- **Retrospective chart review of the two identified subjects**

Case one

History

- 9 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 audiogram
- Subject did not recall a time when he could hear from his left ear
- Unable to close left eye and has had facial asymmetry since birth
- Birth history
  - No perinatal, ototoxic medications
  - No perinatal infections
  - No hx of jaundice or hyperbilirubinemia
  - Full-term cesarian section
  - Apgar scores unremarkable
- Family history- mother has Diabetes Mellitus Type I

Physical Exam

- House—Brackmann facial-nerve grading system: IV/VI
  - Asymmetric face at rest with decreased expression of left nasolabial fold
  - Incomplete closure of his left eye
  - No movement of his left forehead
  - Mouth is symmetric at rest and with movement
- Left ear has lop ear deformity with absence of antihelical fold
- Cranial nerves intact except left CN 7 and 8

Diagnostics

- Audiogram
  - Profound left SNHL throughout all frequencies
  - Normal right hearing
- High-resolution computed tomography (Figure 1)
  - Narrow left internal auditory canal (1.0 mm)
  - Normal right internal auditory canal (4.0 mm)
  - Bony anatomy of bilateral external, middle, inner ears were otherwise normal

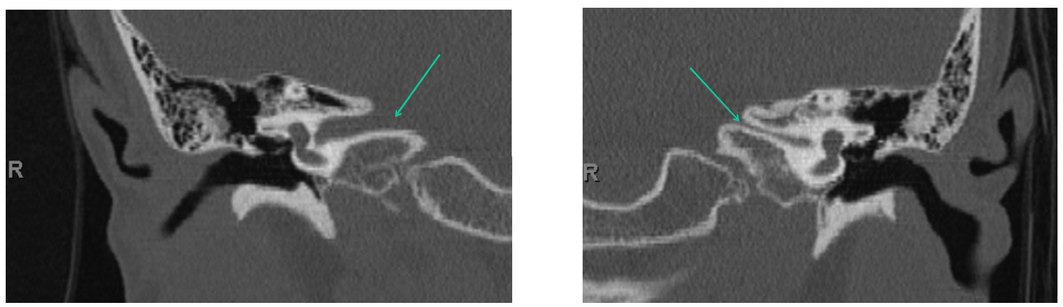

Figure 1. Coronal CT scan demonstrating a normal right internal auditory canal in Subject 1 (image on the left) and a narrow left internal auditory canal (image on the right).

Case two

History

- 5 month-old healthy female referred to the otolaryngology clinic for hearing loss, facial weakness, and bilateral pre-auricular skin tags
- Had failed her newborn hearing screening
- Left facial weakness since birth with inability to close her eye; had had no improvement in function
- Birth history
  - No perinatal, ototoxic medications
  - No perinatal infections
  - No hx of jaundice or hyperbilirubinemia
  - Full-term spontaneous vaginal delivery
  - Apgar scores unremarkable
- Family history- mother has Diabetes Mellitus Type II, multiple paternal relatives with pre-auricular skin tags

Physical Exam

- House—Brackmann facial-nerve grading system: V/VI
  - Asymmetric face at rest
  - Incomplete closure of his left eye, mild exposure keratitis evident
  - No movement of his left forehead
  - Mouth is asymmetric at rest
- Left preauricular skin tag and two right pre-auricular skin tags
- Cranial nerves intact except left CN 7 and 8

Diagnostics

- Audiology
  - Abnormal left auditory brainstem response (ABR) and absent left otoacoustic emissions (OAE)
- Electroneuronography
  - No response of left orbicularis oculi or nasalis
- Electromyography
  - Decreased muscle unit action potentials (MUAPs) in orbicularis oris and frontalis on left and absent spontaneous or voluntary activity in orbicularis oculi
- High-resolution computed tomography (Figure 2)
  - Narrow left internal auditory canal
  - Normal right internal auditory canal
  - Bony anatomy of bilateral external, middle, inner ears were otherwise normal
- Magnetic resonance imaging
  - Hypoplastic left 7th and 8th cranial nerves.

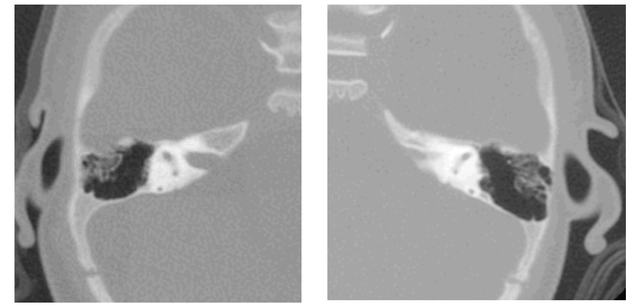

Figure 2. Axial CT scan demonstrating a normal right internal auditory canal in Subject 2 (image on the left) and a narrow left internal auditory canal (image on the right).

DISCUSSION

- Congenital internal auditory canal stenosis is rarely associated with both facial nerve palsy and ipsilateral sensorineural hearing loss.
  - There are less than five reported cases[7,8,9]
- Theories explaining the association of Hearing Loss with IAC Stenosis[6]
  - **1st theory:** The 8th cranial nerve is hypoplastic secondary to an abnormal chemotactic environment or a lack of end organs. The bony canal is narrow as a result of the thin or absent 8th nerve.
  - **2nd theory:** Stenosis is a primary defect that then inhibits the growth of the 8th 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

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