

Unilateral facial nerve hypoplasia without evident facial palsy

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DESCRIPTION

A 2-year-old boy was referred to our neuropaediatrics unit by his otorhinolaryngologist (ORL) for the evaluation of cranial nerves abnormalities found in a brain MRI previously obtained. He was born at a gestational age of 36 weeks, by a normal delivery, with the unremarkable Apgar score. With 2 months of age, parents started to notice some hearing difficulties. He was evaluated by an ORL and some tests were performed (auditory evoked potentials, otoacoustic emissions and tympanogram), which revealed no response on the right side. Then, an MRI was obtained and it revealed an apparent absence of the right facial nerve, also with no identifiable ipsilateral cochlear nerve. Despite that, the internal auditory canal (IAC), the labyrinthine segment of the facial canal and the facial colliculus were symmetrically present (figure 1). There were no abnormal signal emissions in the exit root of the facial nerve at the pons. Physical examination revealed a slight drooping of the lower eyelid on the right side and subtle flattening of the right nasolabial sulcus, the last one only remarkable in some specific movements (eating, crying and smiling) (figure 2).

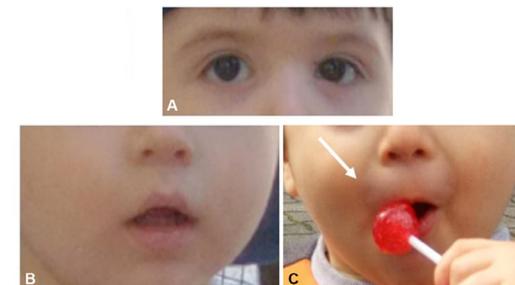


Figure 2 At rest, very slight drooping of the lower eyelid in the right side can be identified. No other significant asymmetries have been identified (A and B). Slight asymmetry of the nasolabial sulcus, only evident in eating movements (C, arrow).

Unilateral facial nerve hypoplasia is a rare condition.¹ It causes an ipsilateral facial palsy, as the healthy side is responsible for a contralateral attraction. Congenital unilateral facial palsy is also rare, with an incidence of around 2 in 1000 live births. A birth injury is the most frequent cause, with developmental causes being responsible for a minority of

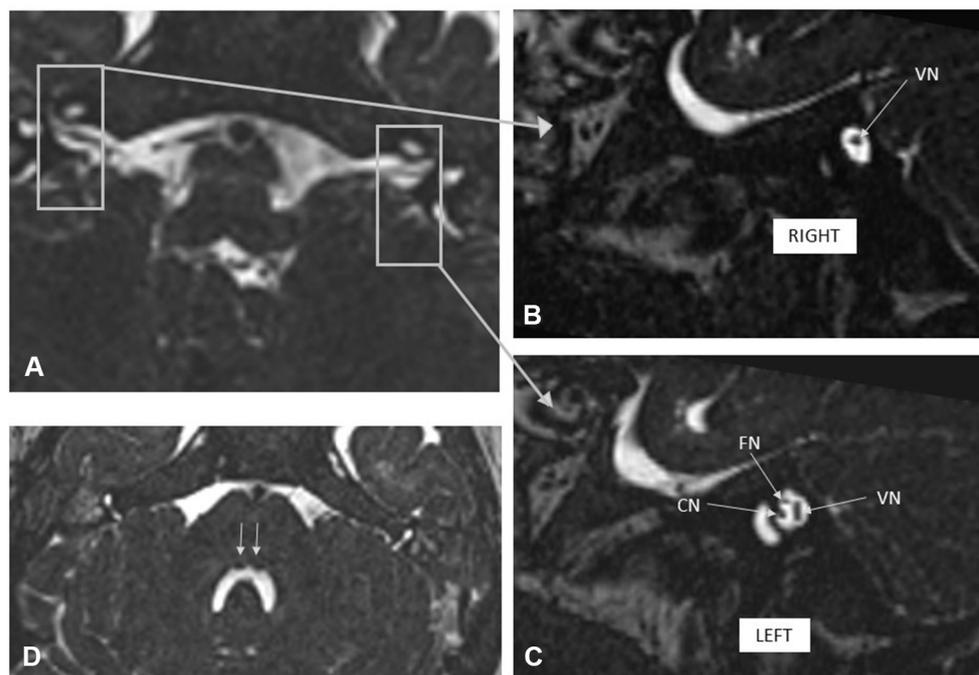


Figure 1 (A) Cranial MRI axial SPACE sequence showing the symmetric right and left IAC. (B) Right IAC, where only the vestibular nerve can be identified. (C) The cochlear, facial and vestibular nerves present in the left IAC. (D) Axial SPACE sequence at the level of the facial colliculus, showing the symmetry of this structure (white arrows). CN, cochlear nerve; FC, facial canal; FN, facial nerve; IAC, internal auditory canal; VN, vestibular nerve.



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cases.¹ Both facial and vestibulocochlear nerves exit from the pontomedullary junction. MRI provides excellent images of the cisternal and intracanal segments of the facial nerve and the three divisions of the vestibulocochlear nerve: the superior vestibular, inferior vestibular and cochlear nerves.² Facial nerve anomalies have been associated with hearing loss. Aplasia or stenosis of the IAC affects the intracanal portion of the facial nerve that accompanies the vestibulocochlear nerve.³

In our case, IAC has symmetrical dimensions and hearing loss is explained by cochlear nerve absence. We were not able to identify the right facial nerve in the MRI. However, the patient has only a discrete facial asymmetry, which makes unlikely facial

nerve agenesis. For that reason, we assume that this patient has severe hypoplasia of the right facial nerve, which cannot be seen in the obtained images. Despite that, this boy has a mostly preserved facial mimic, defining a significant dissociation between clinical findings and radiological measures and making it different from other cases reported in the literature.

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Learning points

- ▶ Facial nerve hypoplasia can result only in a discrete facial asymmetry.
- ▶ The association between facial nerve palsy and cochlear nerve anomalies is already established.
- ▶ Although facial nerve cannot be identified in the MRI, normal dimensions of the canal make unlikely facial nerve agenesis.

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