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CONGENITAL UNILATERAL FACIAL NERVE AGENESIS : CASE REPORT

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ABSTRACT

Facial nerve agenesis should be considered in cases with irreversible congenital facial paralysis. We report two cases of facial nerve palsy in young children; facial nerve aplasia diagnosis was observed with magnetic resonance imaging (MRI) and computed tomography (CT).
Three-dimensional constructive interference in steady state magnetic resonance imaging (3D CISS-MRI) and high-resolution computed tomography scanning (HR-CT) are very useful for diagnosing congenital facial paralysis due to anatomic anomalies.

Keywords: case report; congenital facial paralysis; facial nerve aplasia; 3D-constructive interference in steady state magnetic resonance imaging; high-resolution computed tomography

INTRODUCTION

The incidence of congenital facial paralysis is around 0.8–2.3/1000 live births [1]. Congenital unilateral facial paralysis is generally considered to be the result of either developmental (including Möbius syndrome) or traumatic etiology [1].

Traumatic cases (mostly due to birth injury) recover completely within several weeks [2-5].
In contrast, congenital unilateral facial nerve paralysis not due to birth injury is irreversible [5-7], this group is considered to be the result of a developmental or genetic anomaly of the facial nerve or facial motor nucleus.

These case-reports detail two patients with congenital unilateral facial nerve aplasia diagnosed with 3D-constructive interference in steady state (3D-CISS) magnetic resonance imaging (MRI) and high-resolution (HR) computed tomography (CT) scanning.

Patients and observations

Case no 1:

A 2-year-old girl was referred for evaluation of right-sided congenital facial palsy. Otoscopy was normal.

On HRCT scanning, the mastoid and tympanic segments of the fallopian canal on the right side were absent (Figure 1(a), arrow), with no evidence of IAC stenosis (Figure 1(b), arrow). Bony facial canal of the left side is normal (Figure 1(c), white arrow). It showed features of bilateral chronic otitis media with bony erosion suggesting cholesteatoma.

MRI showed features of otomastoiditis but no brain abnormalities (Conventional T1 and T2-weighted and T1-gadolinium sequences). There were no significant lateral differences in the facial nucleus. The 3D-CISS MRI (heavily T2-weighted images) (Figure 2) demonstrated absence of the right facial nerve.

The left facial nerve was seen in the internal auditory canal (IAC) (Figure 2;d-e-f-g; red arrows), but the facial nerve was absent in the right IAC (Figure 2;d-e-f-h; black arrows). Both sides showed normal cochlear shapes and semicircular canals.

Case no 2:
A 1.5-year-old girl was referred for suspicion of facial nerve paralysis based on facial deformity since birth. Otoscopy was normal. No evidence of hear loss based on auditory evoked potential. General examination showed no associated deformities.

On HRCT scanning, the three segments of the fallopian canal on thc left side were absent -figure 3(a;b;c)- with no evidence of IAC stenosis.

DISCUSSION
The internal acoustic canal (IAC) is a bony canal within the petrous portion of the temporal bone that transmits nerves and vessels from within the posterior cranial fossa to the auditory and vestibular apparatus.

There are five nerves that run through the IAC :
- Nervus intermedius (sensory component of CN VII)
- Facial motor root (motor component of CN VII)
- Cochlear nerve (component of CN VIII)
- Inferior vestibular nerve (component of CN VIII)
- Superior vestibular nerve (component of CN VIII)

The falciform crescent, which is an horizontal ridge, divides the canal into superior and inferior portions [fig 4] :
- Superior: facial nerve and superior vestibular nerve (SVN); the facial nerve is anterior to the SVN and is separated from it laterally by Bill's bar, a vertical ridge of bone.
- Inferior: cochlear nerve and inferior vestibular nerve (IVN); the cochlear nerve is situated anteriorly.

Birth trauma is the most frequent cause (78–91%) of congenital unilateral facial paralysis [2-4]. Developmental disorders are divided into syndromes and isolated; syndromes include Goldenhar syndrome, Treacher–Collins syndrome, and hypoxic ischemic encephalopathy (such as Möbius syndrome) [1]; facial nerve deficiency/aplasia is one reason for isolated developmental congenital facial nerve paralysis [6].

Computed tomography (CT) and magnetic resonance imaging (MRI) are well established imaging modalities for examining the facial canal as well as the course of the facial nerve itself [8].

Temporal bone CT is particularly useful for [1;7;8]
- Imaging the fallopian canal in the temporal bone, from the porus acusticus to the stylomastoid foramen.

- Evaluating the caliber and the course of the IAC and bony facial nerve canal in the temporal bone, which can provide key information regarding facial nerve pathology and prove critical in surgical planning for otologic surgery
- Erosion and destruction of the facial nerve canal.
- Diagnose cochlea-vestibular anomalies.
- Anatomical relationship of the facial nerve canal to the ossicles (which are not seen on MR) and cochlea-vestibular structures; these relationships are critical during surgical planning.
- Identifying temporal bone fractures that violate the facial nerve canal.

The protocol includes high-resolution temporal bone CT, with non-contrast 0.3mm axial slices through the bilateral temporal bones for raw data, with 0.6mm thick axial, coronal, and Pöschl reformats through the individual temporal bones [8]. The administration of intravenous contrast is not systematically performed but may be included if there is a clinical suspicion of a neoplasm or vascular abnormality [8]

From the perspective of radiation exposure, it is not the primary diagnostic tool for children's facial nerve aplasia (1).

3D-CISS MRI (**c**onstructive **i**nterference in **s**teady **s**tate**:** a heavily T2-weighted sequence) gives high contrast for cerebrospinal fluid and shows a filling defect, and it usually shows facial nerves in the IAC with clear distinction from the cochlear-vestibular nerves [1;11].

3D-CISS MRI sequence indicated the aplasia of the facial nerve in the IAC in the first case, it was difficult to prove facial nerve aplasia by conventional MRI or CT.

3D-CISS MRI could be, therefore, a valuable tool for the early differentiation of congenital facial nerve aplasia from birth injury and from other congenital causes in cases of delayed/irreversible recovery [1].
The key roles of MRI are [8]

- Image the facial nerve from the brainstem to the fundus of the internal auditory canal using high-resolution sequences.
- Evaluating soft tissue abnormalities around the facial nerve.
- Determine the presence of peri-neural spread from parotid malignancies.

In patients with Moebius syndrome, CISS imaging is particularly useful in confirming the absence or small caliber of the facial nerve within the CPA and IAC [8].

Standard MRI imaging for evaluation of the facial nerve and inner ear pathology should include an IAC protocol with the following sequences [8]:

- Non-contrast axial 5mm whole brain T2-weighted sequence; an axial 3 mm.
- T1-weighted sequence of the IAC angled perpendicular to dorsal aspect of the brainstem.
- Axial constructive interference in the steady-state (CISS) sequence (0.6 mm) from the occipital bone to superior petrous ridge.
- Axial 5mm whole brain post-contrast FLAIR and T1-weighted sequences.
- Coronal 4mm post-contrast T1-weighted sequence of the IAC.
- Axial 3mm T1-weighted fat-saturated sequence of the IAC.
- Reconstructed sagittal oblique CISS images through the internal auditory canals obtained perpendicular to the course of the IAC enable the evaluation of the caliber of the facial nerve.

3D-CISS MRI and HRCT confirmed that a developmental malformation of the unilateral facial nerve was the pathogenic condition in both patients.

From the perspective of radiation exposure, MRI including 3D-CISS should be the primary diagnostic tool for imaging children with congenital unilateral facial nerve paralysis [1].

CONCLUSION

Even though it is rare, facial nerve agenesis should be considered in patients with facial palsy. Imaging, especially 3D-CISS MRI of the brain and brainstem can provide critical informations for establishing the diagnosis and planning the treatment.

Competing interests: The authors declare no competing interest.

REFERENCES

1. Mari Shimada Dias, Kenichi Nakamura, Toru Sasaki, Hisashi Sugimoto & Makoto Ito (2016) A case of congenital unilateral facial nerve aplasia, Acta Oto-Laryngologica Case Reports, 1:1, 102-105, DOI: 10.1080/23772484.2016.1247648
2. Smith JD, Crumley RL, Harker LA. Facial paralysis in the newborn. Otolaryngol Head Neck Surg. 1981 Nov-Dec;89(6):1021-4. doi: 10.1177/019459988108900628. PMID: 6801582..
3. Toelle SP, Boltshauser E. Long-term outcome in children with congenital unilateral facial nerve palsy. Neuropediatrics. 2001 Jun;32(3):130-5. doi: 10.1055/s-2001-16610. PMID: 11521208.
4. Falco NA, Eriksson E. Facial nerve palsy in the newborn: incidence and outcome. Plast Reconstr Surg. 1990 Jan;85(1):1-4. doi: 10.1097/00006534-199001000-00001. PMID: 2293714.
5. Shapiro NL, Cunningham MJ, Parikh SR, Eavey RD, Cheney ML. Congenital unilateral facial paralysis. Pediatrics. 1996 Feb;97(2):261-4. PMID: 8584391.
6. Pavlou E, Gkampeta A, Arampatzi M. Facial nerve palsy in childhood. Brain Dev. 2011 Sep;33(8):644-50. doi: 10.1016/j. Brain Dev.2010.11.001. Epub 2010 Dec 8. PMID: 21144684.
7. Held P, Fellner C, Fellner F, Seitz J, Graf S, Hilbert M, Strutz J. MRI of inner ear and facial nerve pathology using 3D MP-RAGE and 3D CISS sequences. Br J Radiol. 1997 Jun;70(834):558-66. doi: 10.1259/bjr.70.834.9227246. PMID: 9227246.
8. Gupta S, Mends F, Hagiwara M, Fatterpekar G, Roehm PC. Imaging the facial nerve: a contemporary review. Radiol Res Pract. 2013;2013:248039. doi: 10.1155/2013/248039. Epub 2013 May 23. PMID: 23766904; PMCID: PMC3676972.
9. Lo YL, Fook-Chong S, Leoh TH, Dan YF, Lee MP, Gan HY, Chan LL. High-resolution ultrasound in the evaluation and prognosis of Bell's palsy. Eur J Neurol. 2010 Jun 1;17(6):885-9. doi: 10.1111/j.1468-1331.2010.02950.x. Epub 2010 Feb 11. PMID: 20158516.
10. Terzis JK, Anesti K. Experience with developmental facial paralysis: Part I. Diagnosis and associated stigmata. Plast Reconstr Surg. 2011 Nov;128(5):488e-497e. doi: 10.1097/PRS.0b013e31822b6791. PMID: 22030508.
11. Hwang JY, Yoon HK, Lee JH, Yoon HM, Jung AY, Cho YA, Lee JS, Yoon CH. Cranial Nerve Disorders in Children: MR Imaging Findings. Radiographics. 2016 Jul-Aug;36(4):1178-94. doi: 10.1148/rg.2016150163. PMID: 27399242.

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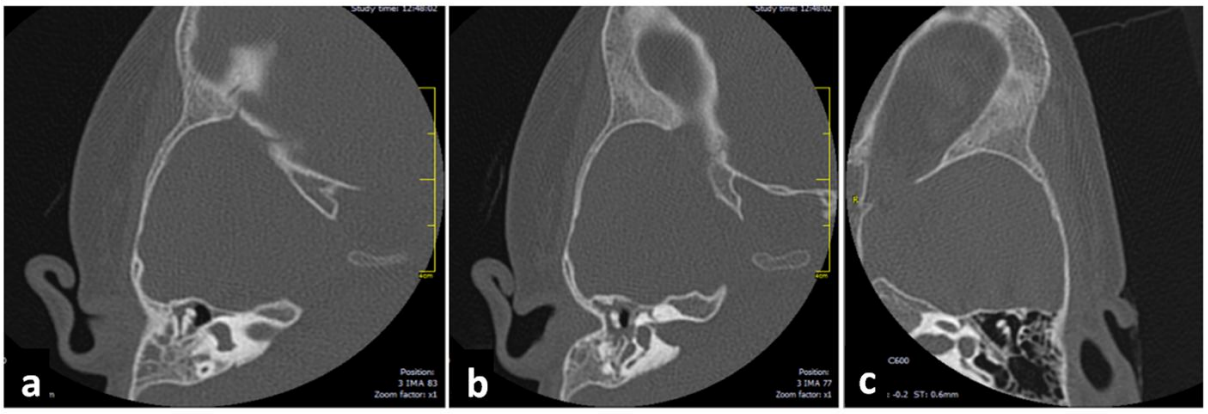

Figure 1: **Right erosive chronic otomastoiditis.**
(a;b) Axial CT images show opacification of the right middle ear and right mastoid air cells with erosion of the bony facial canal in its tympanic segment cavity suggesting cholesteatoma.
(c) Left ear: normally aerated mastoid cells and middle ear with normal tympanic appearance of the facial canal.

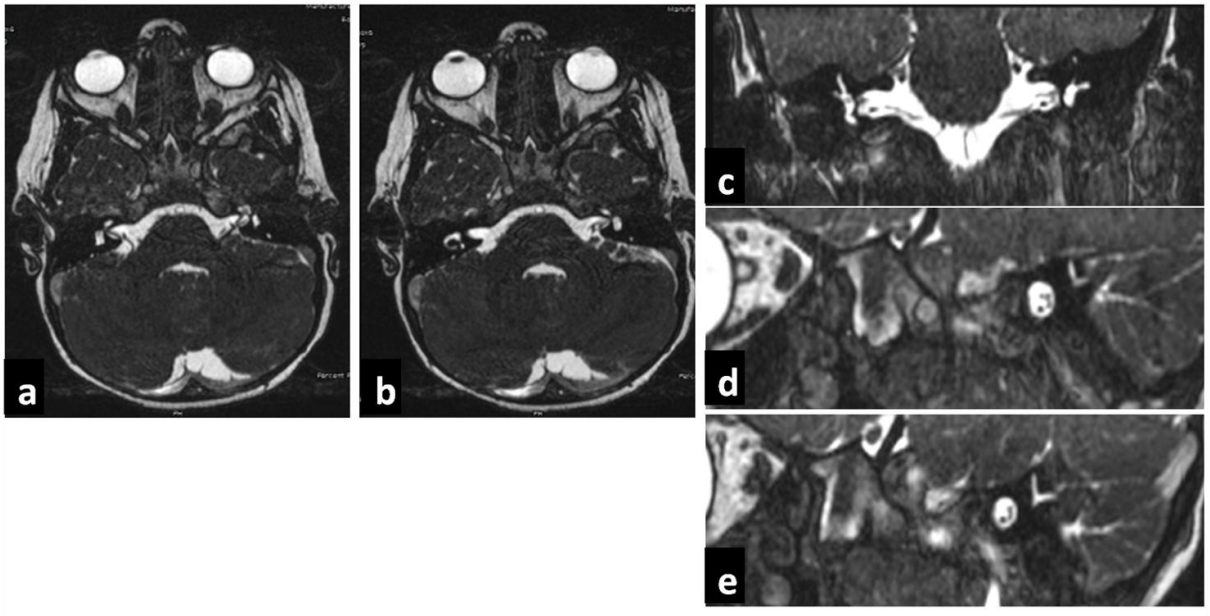

Figure 2: **MRI of the same patient**
Axial CISS **(a;b)** coronal **(c)** and sagittal oblique left(d) and right **(e)**.
Lack of normal facial nerve on the right side with absence of the cisternal segment (arrowhead). Note normal position of facial nerve on the left side. Opacification of the mastoid cells enhanced after gadolinium administration with no focal lesion in diffusion.

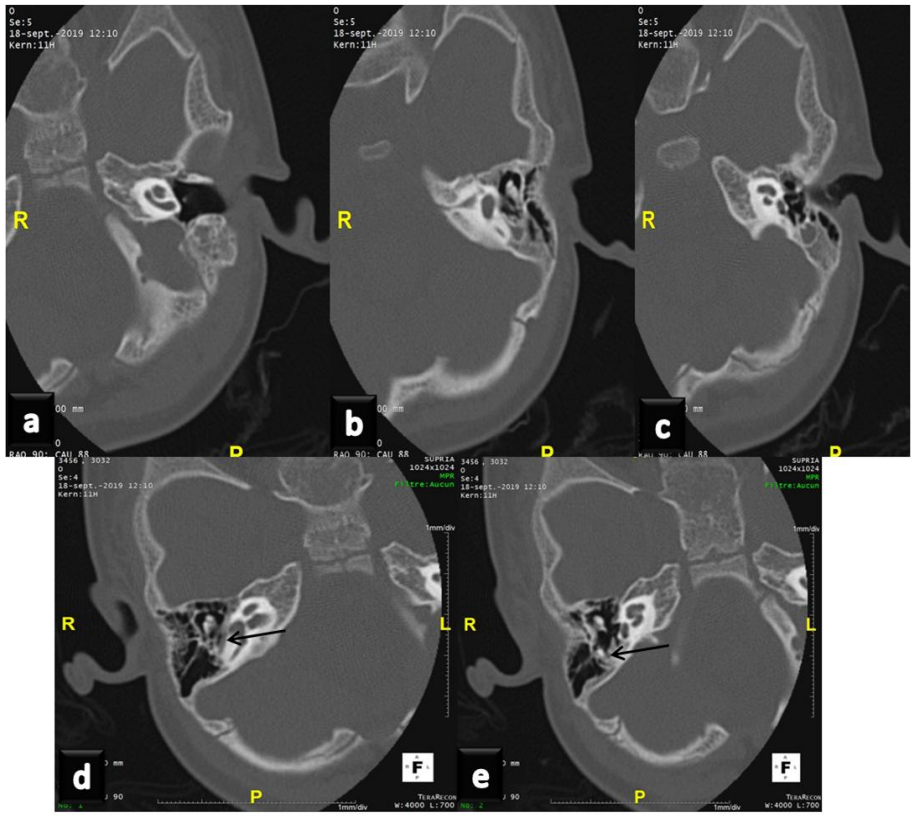

Figure 3: 5-year-old girl patient with left facial deformity noticed since birth. **a,b,c** : absence of left facial nerve canal absent (labyrinthine, tympanic and mastoid segments are absent); **d.e**: normal position of facial nerve on the right side (arrow)

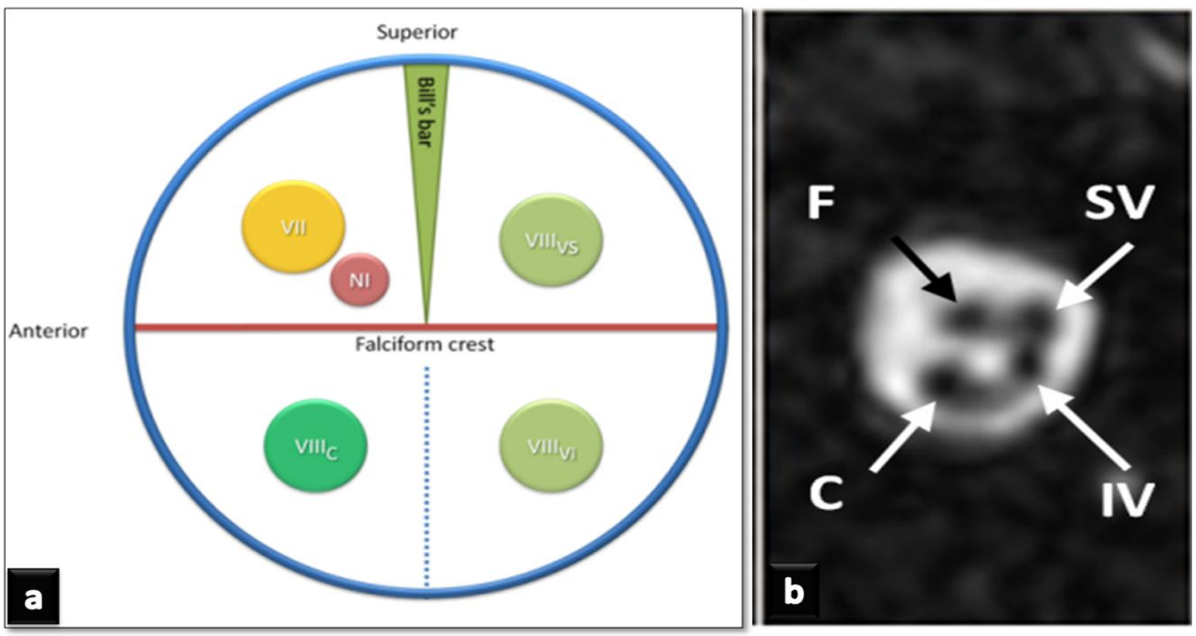

Figure 4 :
[a] Diagram of the contents of the internal auditory canal (*Case courtesy of Assoc Prof Frank Gaillard, Radiopaedia.org, rID: 36049*);
[b] Sagittal T2 CISS MRI of the IAC (*Case courtesy of Eldda Banciu and Arnaud Attyé, clinique universitaire de radiologie-CHU de Grenoble*).
NI : nervus intermedius; SV :superior vestibular nerve; IV inferior vestibular nerve; F : facial nerve; C : cochlear nerve