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Bifurcation of the intratemporal facial nerve: A rare anatomical anomaly

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ABSTRACT
The anatomical position of the facial nerve is a critical factor in determining surgical candidacy in patients with congenital aural atresia (CAA). All patients with CAA must preoperatively be evaluated using a grading score based on information gained from a high resolution CT scan. In patients not suitable for surgical reconstruction, implantation of novel hearing implants is increasingly used for hearing rehabilitation. We, here, describe a bifurcation of the intratemporal part of the facial nerve in a 5-year old boy with CAA undergoing implantation with a bone conductive hearing device.

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Introduction
Congenital aural atresia (CAA) is caused by the abnormal development of the first and second branchial arches [1]. As the absence of an external ear canal precludes fitting of conventional hearing aids, the hearing rehabilitation must be provided through surgical reconstruction of the ear canal and middle ear or implantation of a hearing implant. Since the atresiaplasty is associated with an abundance of complications such as restenosis and recurrent granulations of the reconstructed ear canal, different hearing implants such as bone conduction or active middle ear devices are used for hearing rehabilitation. One of the most important anatomical prerequisites for surgical repair is the position of the facial nerve, which may also influence on the application of hearing implants [2].

Case report
A 3-year-old boy with bilateral CAA and microtia was referred to the ENT clinic. Since birth, he was fitted with bilateral BAHA on soft band. CT examination (Figure 1) revealed on the right side a narrowed middle ear space, a sclerotic oval window and an atretic round window. The tympanic segment of the facial nerve was positioned lateral to the posterior semicircular canal and a bony canal, interpreted as chorda tympani, was identified inferior of the lateral semicircular canal. This canal appeared to communicate with the tympanic segment of the facial nerve. The pneumatization of the mastoid was poor and the middle ear was opaque. The right external ear canal was narrowed and filled with soft tissue. The external ear was valued as Microtia grade II.

On the left side, the external ear was normal. On the CT scan, the left middle ear space was evaluated as narrow and the oval window was partially sclerotic. Although the tympanic portion of the facial nerve was covering the oval window and the mastoid portion was slightly anterior positioned, the course of the facial nerve on this side was considered as less aberrant than on the right side. Moreover, the size of the round window was normal and the external auditory ear canal was very narrow. Consequently, the patient was not eligible for surgical reconstruction. At the age of four he was implanted with a fixture applied BAHA. The fitting was limited through severe skin problems and pain and he was therefore planned for an implantation with a semi-implantable BonebridgeR device on the right side. A renewed CT scan at the age of five years showed unchanged anatomical conditions.

At the mastoidectomy, a thin string of soft tissue in a bony canal was detected in the middle part of the mastoid. The canal was positioned posterior of the remnant of the external bony canal wall running...
superficially to the posterior part of the microtic external ear. The nerve was monitored during the whole operation with a Nerve monitor (Neurosign). The structure was directly stimulated with 0.8 mV without response. To avoid contact with the unknown structure, the implant was therefore placed further posteriorly than initially planned.

After discharge on the day of the operation he complained about a numb feeling on the right side of the face. A facial nerve dysfunction graded House Brackman (HB) IV was observable at the first postoperative day, more evident in the ramus zygomaticus and ramus marginalis mandibulae. The patient was immediately explanted and the structure was identified as a portion of the facial nerve.

The electrophysiological examination one month after explantation revealed an axonal blockage in 50% and a conductive blockage in 50%. Several active motoric units were measured in the frontal muscle at the EMG, less activity documented in the orbicularis and oculi muscles. The function of the facial nerve was completely restored after 4 months.

A renewed evaluation of the CT scan including 3D oblique reformats showed the duplication of the facial nerve at the transition between the tympanic and mastoid portion with one lateral component displaying an aberrant course laterally in the mastoid cavity (Figure 2). The second component of the duplicated nerve had a slight abnormal ventral position in the mastoid and covered the oval window. The duplication was located directly below the lateral semi-circular canal (Figures 3 and 4).

One year after explantation, the boy was aided with a bone-aided hearing aid, ADHEAR (Med-El, Innsbruck, Austria)

Discussion
The development of the facial nerve is closely related with the development of the middle ear and the mastoid process as these structures derive from the first and second branchial arches and the facial nerve derives from the second branchial arch [2]. The orientation of the facial nerve is established in the 8th week of gestation but the ultimate position and covering of the facial nerve are determined by further development of structures like the stapes, labyrinthine capsule, mastoid bone and tympanic bone. The final position of the nerve in the temporal bone is established during the second year of life.

The most common abnormalities of the facial nerve in CAA are displacement of the nerve and bony dehiscence in the tympanic segment [3]. The bony dehiscence can also be seen in normal temporal bones up to 53% and it is considered as a normal anatomical variation [4]. An aberrant course of the nerve appears in 0.3% of normal ears. Goldsztein and Roberson presented the anatomical findings in 209 atresia cases and found that in 39% of the cases, the facial nerve had an abnormal course, located more
lateral and anterior, not identified in 1% and dehiscent in 57% [5].

Bifurcation of the facial nerve was described in the literature but is a very rare anomaly. Marquet reported bifurcation of the intratemporal part of the facial nerve in 0.7% from anatomical specimens [6]. It was also described in a series of 972 patients undergoing cochlear implantation were seven patients had an abnormal facial course and two of those had a duplication of the facial nerve [7].

We found only two previous case reports of congenital duplication of the facial nerve. In one of these reports, a duplicated mastoid segment of the facial nerve in a young female with middle ear anomalies was described [8] and another author reported three cases [9].

Facial nerve injury is a well-recognized risk of atresiaplasty. Its incidence has been quoted in the literature to range from 0% to 11% [10]. Since the introduction of perioperative facial nerve monitoring, the risk probably has decreased.

We conclude that although the course of the facial nerve in atretic ears is almost normal in most cases the meticulous evaluation of the preoperative CT scan is mandatory in the surgical planning. In present case study, the bifurcation of the nerve was overseen despite repeated imaging. The mechanical pressure of the skin retractor used at the surgery was considered as the cause of the postoperative facial nerve dysfunction.

**Summary**

Surgery of CAA remains one of the most challenging procedures in otology. As facial nerve anomalies,
often, are associated with middle- and inner ear anomalies the careful evaluation of the preoperative CT scan in different projections is essential before a surgical intervention and a prerequisite for a successful surgery.

Disclosure statement
A written consent for publication was obtained by the patient’s parents.

The authors report no conflict of interests.

References