Spectrum of developmental abnormalities of cranial nerve VIII (statoacoustic)

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Learning objectives

- To review the main causes of congenital hearing loss causes
- To remember the most common findings in developmental anomalies of VII cranial nerve
- To demonstrate the most appropriate imaging techniques and protocols for their evaluation

Background

Sensorineural hearing loss (NSHL) is a major cause of disability, with an estimated prevalence of 1 per 2000 live births. Early diagnosis is of great importance, not only because of the hearing impairment and associated alterations in the daily and language development, but also affects social development, emotional and academic. Screening programs for neonatal HNS have reduced significantly the average age of diagnosis from 24 months years ago until to 2-3 months today.

Although most neurosensory hearing loss are secondary to degenerative processes of the inner ear that increase progressively with age, there are different types of birth defects that affect the inner ear causing hypoplasia and hearing loss.

During embryological development of the inner ear of the progressive growing of petrous bone structures will condition a series of pathologies according to the moment when it is interrupted. According to the classification of Jackler, the more severe is Michel aplasia or labyrinth aplasia, in which there is an absolute absence of the labyrinthine structures, and for which there are no therapeutic options nowadays. In a more advanced step of embryological development appears unique cochlear aplasia with present SCC and vestibulum, and successively it will develope with less severity cochlear hypoplasia, incomplete cochlea and common cavity. In a second phase, with the cochlea adequately differentiated, development anomalies can be divided between lateral-SCC dysplasia, vestibular dysplasia, and enlarged vestibular aqueduct.

Fig. 8 on page 3

Agenesis and hypoplasia of the eighth cranial nerve (statoacoustic) are rare development disorders, although within the range of causes of neuropathies with disorder auditory (ENDA) are an important cause to be considered. Around 10% of hearing loss in children are due to ENDA, and of these, about 10% are caused by agenesis or hypoplasia of VIII cranial nerve. The internal auditory canal is created by the inhibition of cartilage on the lateral aspect of the otic vesicle in the 9th gestation week of. The existence of the VIII cranial nerve is required so that this process can take place. The cochlear nerve size
depends directly on the number of ganglion cells in the basal turn. If VIII cranial nerve is aplastic, IAC will have a diameter proportional to the nerve size. The main symptom is sensorineural deafness from birth, that can be treated with a cochlear implant if the nerve is present, and with a brainstem implant if aplastic.

In the study of young patients with NSHL many techniques are required, but in each center different protocols are considered according to their sanitary infrastructure, which often include audiometry, evoked potentials, complete neurological examination, and advanced neuroimaging studies within a complete multidisciplinary approach.

Images for this section:

Fig. 8
High resolution studies of CT and MRI of posterior fossa allow to detect the presence of the VIIIth cranial nerve and depict the morphology of IAC and inner ear, in order to plan potential therapeutic targets, since their identification is key when considering the use of cochlear implants.

MRI is the technique of choice to show integrity of cranial nerves, especially in its cisternal paths. Advances in MRI have been extremely important, especially in the higher homogeneity of the magnetic field developing stronger gradients, and antennas in Phase - array, allowing a greater signal to noise ratio. Using T2 high spatial resolution in posterior fossa (CISS. ..) or mixed sequences True- FISpP, allow to recognize and define the eighth cranial nerve in cisternal and canalicular paths, and to define cochlear and vestibular branches. In this MPR-MRI in posterior fossa CISS sequence, we can see how, under normal conditions, inside IAC we can delineate the seventh pair at anterior and upper location (red arrow), the two branches of the vestibular located downstream (green arrow), and in the anterior and basal location, the cochlear branch of the eighth cranial nerve (yellow arrow).
Furthermore, it is necessary to evaluate the morphology of the membranous labyrinth structures, and eventually the brain stem, as in the case of aplasia of the VIII cranial nerve, patients may be candidates for central implant. MRI also allows Volume Rendering 3D reconstructions for better delineation of the membranous labyrinth.

More recently it has been developed cranial nerve neurography, based on Diffusion tensor sequences and tractography adapted to the cranial nerves, using cutt thicknesses below 2 mm, maximum gradients using powerful and isotropic structural sequences with overlap in posterior fossa, allowing to analyze the fractional anisotropy in auditory brainstem nuclei, as well as the integrity of the nerve fibers. Specific configuration is required in the acquisition parameters and image reconstruction in DTI. For example, an acquisition can be carried out by a DWISE sequence with TR 11395 and TE 70ms, flip angle 90 °, voxel size 2x2x1.20mm, with 16 directions and 2 values B (0 to 1000). In the reconstruction and post-processing the values must be adapted with respect to the conventional tractography, using a minimum value of fractional anisotropy 0.06 (versus...
0.15 conventional) with a maximum angle of direction change of the fibers of 36.5 ° (versus 27 °), and a minimum length of 7mm fibers.

Fig. 2

References: Neurorradiología, Clínica las Nieves - Jaén/ES

In addition, high-resolution CT can accurately define bone structure of middle and inner ear, allowing to study accurately with curved and multiplanar reconstructions both diameter and morphology of IAC´s, cochlea and other structures for surgical planning. The acquisition with isotropic studies, with voxels of approximately 1.25 mm shaft, allows to delineate accurately all the petrous structures, make the necessary measurements in cochlear canal and the bony labyrinth, and perform necessary curves and multiplanar reconstructions so as to study the viability of the implant and schedule it.
Fig. 3

References: Neurorradiología, Clínica las Nieves - Jaén/ES
In the case described below, a patient 10 months old, was sent to study due to negative potentials, and absence response to auditory stimuli. The study of high-resolution CT of posterior fossa showed absence of the formation of CAIS, with a single cavity in irregular inner ears. MRI failed to locate the VIII cranial nerve, with an VII cranial nerve in an ectopic location. This is a case and complete bilateral labyrinthine agenesis with VIII cranial nerve aplasia.
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**Fig. 9**

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In the second case, the patient underwent cochlear implant. However, after the operation the patient did not improve his hearing loss as expected, and CT study showed severe hypoplasia of both CAI's, with accessory bone channel for the seventh cranial nerve.
It is important to remember that even when there are abnormalities in the conformation of the inner ear structures, as Mondini dysplasia, common cavity deformity or cystic cochlear malformation, if the cochlear nerve is present, conventional devices for cochlear implant may be useful, so that in these cases, the accurate identification of the nerve becomes even more. This nine years old patient affected of left NSHL, shows in with the CT stenosis of the internal auditory cannal, while the MRI showed nerve structures properly developed.
We must also bear in mind that there are other anomalies of cranial nerves that may hinder the diagnosis of agenesis of the cochlear nerve or of the complete VII cranial nerve, as in this case of Moebius syndrome, wherein the patient suffers from agenesis of cranial nerves VII and IV, but the VIII is present in all its branches. In left IAC we can appreciate the left facial and cochlear branches, but in the right side only the eighth cranial nerve can be identified.
Fig. 5

References: Neurorradiología, Clínica las Nieves - Jaén/ES

Conclusion

- Developmental abnormalities of the VIII cranial nerve are disorders that require specific diagnosis, and correlation between CT and MRI can detect the degree of disease in order plan the most appropriate treatment.

- The proper preoperative study may allow better patient selection, avoiding unnecessary surgeries.

References


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