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Experimental Research

Difficulties of Cochlear Implantation in Malformations of the Inner Ear

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Data from various authors indicate that 20% of children with sensorineural hearing loss or deafness have computed tomography results showing abnormalities in the structure of the temporal bones. Cochlear implantation in the presence of cochleovestibular anomalies is associated with a number of difficulties. These include details of how cochleostomy is performed and how electrodes are placed within the cochlea, the risk of damage to the facial nerve due to its abnormal location and its aberrant stimulation in the postoperative period, intraoperative liquorrhea, which can lead to the development of bacterial meningitis in the postoperative period, as well as the risk of penetration of the electrode into the internal auditory canal. The present report describes our method of performing cochlear implantation in the presence of a developmental malformation of the inner ear, i.e., common cavity. The method expands the possibilities of cochlear implantation to restore auditory perception in patients with this pathology.

Keywords: cochlear implantation, inner ear anomalies, common cavity, cochleostomy.

Introduction. Cochlear implantation (CI) is one of the most effective methods of rehabilitation of patients with sensorineural deafness. Data reported by a number of authors indicate that 20% of children with sensorineural hearing loss or deafness produce computed tomography results showing abnormalities in the structure of the temporal bones [Jackler et al., 1987; 1989; Papsin, 2005]. Such anomalies can be accompanied by different levels of impairment to auditory function and different levels of disease progression with and without developmental anomalies of other organs and systems [Park et al., 2000]. Auditory-verbal rehabilitation in children with developmental anomalies of the inner ear, in particular the cochlea, following cochlear implantation can also be associated with a number of difficulties and lower efficacy in terms of the quality of perception of speech and other sounds, duration of treatment, etc.

Cochlear abnormalities are believed to result from inhibition of embryogenesis, abnormal development during certain stages of the prenatal period, and genetic defects [Sennaroglu et al., 2002; Tucci et al., 1995]. Inactivation of one or more of the genes responsible for the normal development of the inner ear can occur [Morsli et al., 1999].

The most widely recognized classifications of cochleovestibular anomalies in otorhinolaryngology and otoneurosurgery are those proposed by Jackler (USA) and Sennaroglu (Turkey). The first classification (the Jackler classification) is based on data obtained from CT examination of the temporal bones [Jackler et al., 1987; 1989]. The second represents cochlear malformations, which Sennaroglu divided by severity into six categories depending on the time of disruption of the normal course of embryonic development [Sennaroglu et al., 2002]. This classification of cochlear malformations includes an incomplete separation of types I and II. The present work relies on the Jackler classification (Table 1).

In the light of cochlear implantation, category A – aplasia and malformations of the cochlea – is of great interest. Cochlear implantation is impossible in the first two types of aplasia. The only way to help a patient with such a pathology is brainstem implantation.

Three types of cochlear hypoplasia demonstrated on computed tomography are distinguished. The cochlea generally retains its internal architecture but is reduced in size: in

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TABLE 1. Jakler Classification of Inner Ear Malformations

Category A	Cochlear aplasia
	Labyrinth aplasia (Michel malformation)
	Cochlear aplasia, normal or malformed vestibule and semicircular canal system
	Cochlear aplasia, normal or malformed
	Incomplete cochlea, normal or deformed vestibule and semicircular canal system (Mondini malformation)
	Common cavity: cochlea and vestibule consist of a single space without internal architecture, normal of malformed semicircular canal system
	All malformations may be accompanied by dilated vestibular aqueduct
Category B	Normal cochlea
	Dysplasia of the vestibule and lateral semicircular canal, normal anterior and posterior semicircular canals
	Dilated vestibular aqueduct, normal or dilated vestibule, normal semicircular canal system

type I it is kidney-shaped, in Type II it is bag-shaped, and in Type III the cochlea has 1.5 turns. The third type of hypoplasia is very often confused with the Mondini malformation.

Incomplete division of the cochlea also occurs in three types: in Type I there is no modiolus or interscalene septa, in Type II only the basal part of the modiolus is present (the Mondini malformation), and in Type III the modiolus is absent and the interscalene septum is present (described in a case of X-linked deafness).

When there is a common cavity, the cochlea and vestibule generally form a single cavity, which has an oval or round shape.

Cochlear implantation in the presence of cochleovestibular anomalies is associated with a number of difficulties. These include features of performing cochleostomy and placing the electrode within the cochlea, the risk of damage to the facial nerve due to its abnormal location, intraoperative liquorrhea, which can lead to the development of bacterial meningitis in the postoperative period, as well as the danger of penetration of the electrode into the internal auditory canal.

Difficulties may also arise in the postoperative period during rehabilitation. Some researchers have found that inducible stimulation of the facial nerve occurred in 80% (8 out of 10) of patients with cochlear malformations [Papsin, 2005; Rah et al., 2016]. The presence of cochlear anomalies is a risk factor for aberrant facial nerve stimulation, preventing programming of optimal stimulation levels as compared with children with a normal cochlea [Aljazeeri et al., 2021]. An abnormal course of the facial nerve, passing near the oval window or along the promontorium, often accompanies certain inner ear defects and affects emplacement of the electrode array. In addition, cases of stimulation of the facial nerve resulting from use of a cochlear implant have been described in children with inner ear malformations [Kuzovkov et al., 2022]. These anomalies complicate the conditions of cochlear implantation for restoration of auditory perception in patients. To overcome this problem, we proposed an original method for cochleostomy and introduction of the electrode array, which was successfully used in clinical practice.

Methods. ***General clinical practice.*** Of the 845 cochlear implantations we performed, almost 800 were performed on children. A variety of inner ear malformations were identified in 27 children. In 21 of these, the inner ear malformation involved the cochlea: 14 children had type II incomplete separation of the cochlea (the Mondini malformation), one child had type III incomplete separation of the cochlea, four children had cochlear hypoplasia, and two had a common cavity. In two cases, transient paresis of the facial nerve was observed in the postoperative period, with recovery after appropriate therapy. Intraoperative liquorrhea occurred in four cases and was eliminated after direct insertion of the entire active part of the electrode into the cochlea by plugging the cochlear window or by cochleostomy with fragments of myofascial flaps.

The most interesting case in the context of this work was a clinical case with an inner ear defect, i.e., a common cavity, where the original method we proposed for cochleostomy and introduction of the electrode array was first used.

Clinical case. The parents of patient T. (female) attended the clinic with complaints regarding the lack of responses to sounds and speech in a three-year-old child. The child (a fourth pregnancy without complications, no heredity) was born at term with birth weight 2980 g.

Examination established a diagnosis of chronic bilateral sensorineural hearing loss grade IV, of congenital etiology; secondary receptive and expressive language disorder. In addition, the following were observed: sequelae of perinatal CNS damage; movement disorder syndrome; atonic-astatic syndrome; delayed psychomotor development.

The child's mother reported that the girl was late in starting to hold her head up (at 6 months), sit (at 9–10 months), and walk (at about two years). From age six months the child was registered with an ENT specialist, a neurologist, an audiologist, and a pediatrician. At the age of 1.4 years, she received binaural high-power hearing aids, which had no posi-

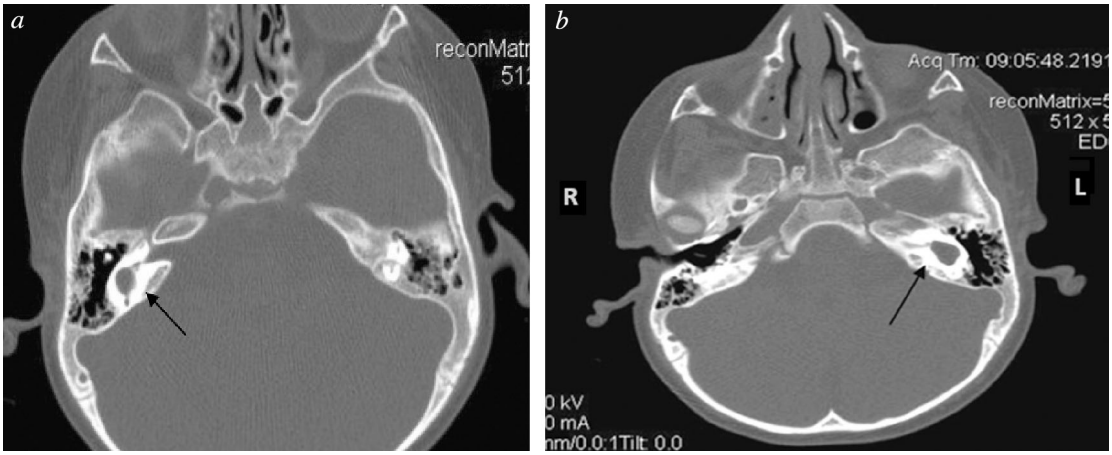

Fig. 1. Computed tomography child T. Inner ear malformation: common cavity *a*) right; *b*) left.

tive effect on auditory perception. Audiological examination showed that short-latency auditory evoked potentials were identified on the right side at a stimulation level of 90 dB; on the left side, the V peak was not recorded at maximum stimulation of 100 dB. No otoacoustic emissions were detected on either side.

Computed tomography of the temporal bones (see Fig. 1, *a, b*) revealed cochleas with rudimentary alterations on both sides – the semicircular canals were not clearly differentiated and merged with the expanded vestibule; the inner ear on both sides was represented by a single irregularly shaped cavity measuring 10.3 × 7.2 mm on the right and 12.6 × 6.9 mm on the left.

The auditory ossicle chains were not altered. Internal auditory canal diameter was 1.4–2.7 mm on the right and 3.3 mm on the left.

The changes in the temporal bones identified on CT led to the decision to obtain additional MRI imaging of the brain and inner ear (Fig. 2).

The results revealed MRI signs of rudimentary cochleas with partially formed semicircular canals and asymmetry of the internal auditory canals on both sides. The presence of vestibulo-cochlear nerves extending from the brain stem was also noted on both sides. However, at the level of the pyramid of the temporal bones, the tissue structure was visible more clearly on the right than the left. These MRI results led to the decision to proceed with right cochlear implantation (right ear).

Approach to Cochlear Implantation and Its Results. Under general endotracheal anesthesia, infiltration of soft tissues with saline solution plus adrenaline was followed by making an S-shaped incision in the postauricular area. Skin and musculoperiosteal flaps were formed and separated. Bores were used to perform mastoidectomy. A short process of the incus was found in the typical location of the aditus ad antrum. At the same level, on the medial wall of the tympanic cavity, the tubercle of the common cavity of the inner ear was identified (Fig. 3, *a*).

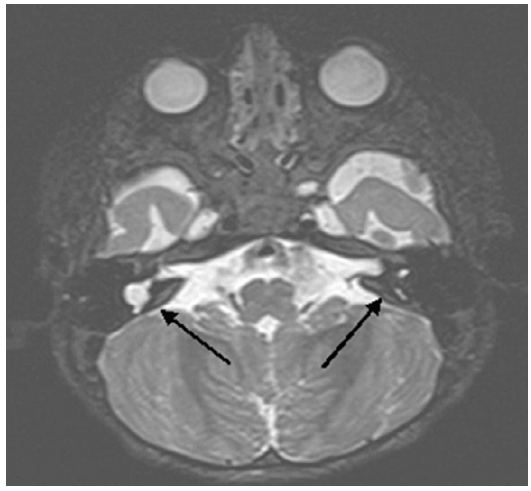

Fig. 2. MRI scan, child T.

The decision was made to open the cavity at that point without proceeding to posterior tympanotomy, with a aim of avoiding the risk of injury to the facial nerve, as we were unable to exclude the possibility that it occupied an abnormal location.

Burrs were used to prepare a bed for the implant and holes for its attachment. The common cavity was opened with a diamond burr (1.2 mm), and a cochleostomy was performed. The shape of the cochleostomy was oval rather than the conventional round. We termed this type of cochleostomy the "post box" method. No liquorrhea was seen on opening the common cavity or at subsequent stages of the procedure. A model CI24RE straight electrode array (ST) implant (Cochlear, Australia) was then emplaced. The electrode was inserted into the cavity using a loop method: holding the free end, the loop was gradually pushed into the cavity (Fig. 3, *b, c*). Once maximum insertion was reached, the free end was released into the cochlear cavity (*d*).

This procedure allowed emplacement of 19 electrodes out of 22. Intraoperative testing of the implanted device

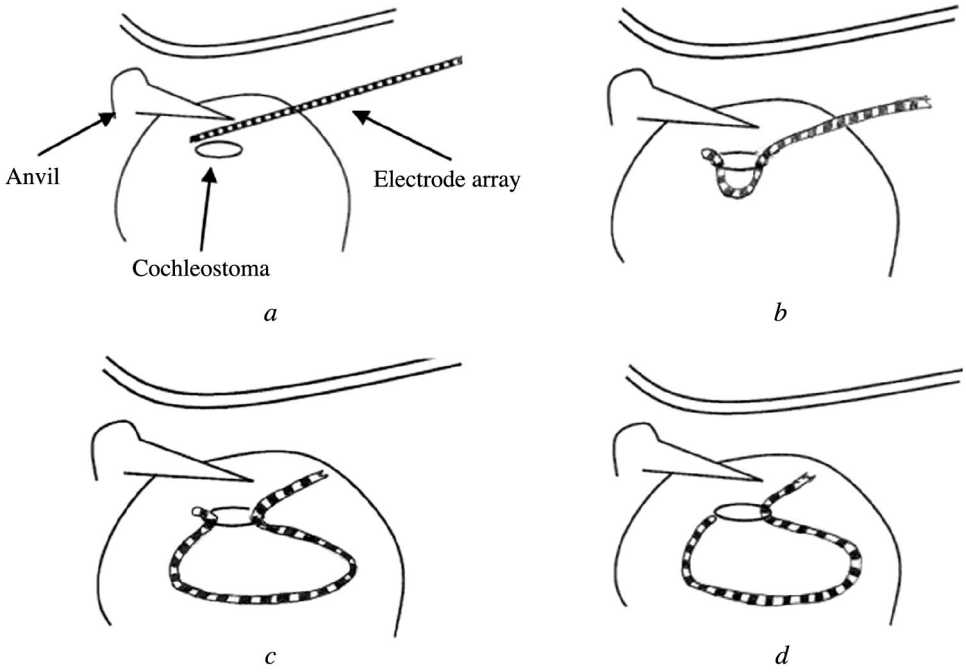

Fig. 3. Diagram showing steps in the emplacement of electrode array after cochleostomy. *a*) Formation of cochleostoma; *b, c, d*) stages in gradual insertion of electrode array).

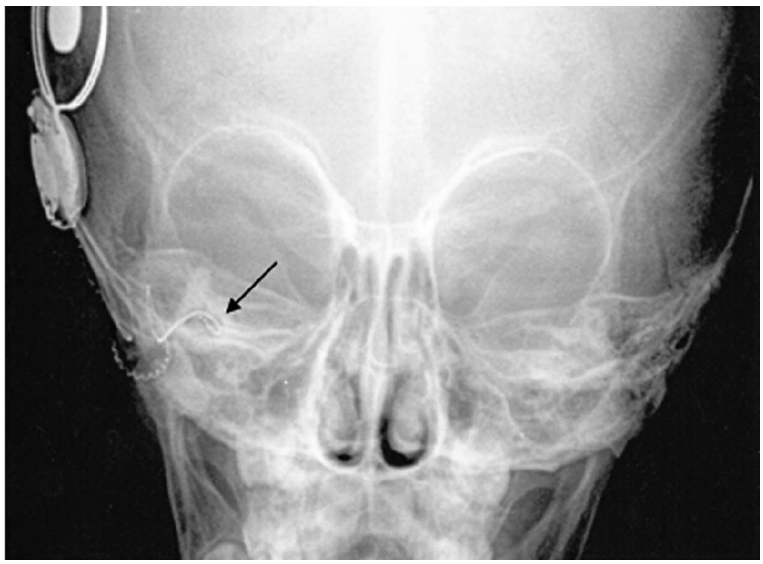

Fig. 4. Intraoperative x-ray. Electrode in common cavity.

by nerve response telemetry (NRT) showed that positive responses were obtained in 16 electrode channels out of 22. Postoperative transorbital radiography of the cochlea showed that the active electrode of the implant was located in the common cavity (Fig. 4). Thus, the cochlear implantation procedure was performed successfully.

After the operation, the child, together with an audiologist and a teacher of the deaf, underwent a course of auditory-speech rehabilitation. Long-term follow-up results showed that the patient's hearing aid was effective. During correctional pedagogical work, the girl developed appropriate auditory-speech behavior, allowing her to make active use of developing auditory sensations to perceive, understand, and reproduce oral speech. She is now studying successfully at secondary school.

Conclusions. Considering the anatomical features of the child's inner ear, the interesting aspect of this case lies in the proposed method of performing cochlear implantation, namely the method of performing cochleostomy ("post box") and the method of introducing the electrode array. It is im-

portant to note that cochlear implants with direct electrode arrays should be used for inner ear malformations of this type. In our view, these conditions led to the above intraoperative results, with avoidance of possible complications such as liquorrhea, damage to the facial nerve, and its aberrant stimulation in the postoperative period. The main goal was also achieved – successful auditory-speech rehabilitation.

Authors' contributions: N. T. Tunyan did the literature search and analysis, wrote the text, and carried out the research; E. V. Bychkova wrote the text and carried out the research.

The authors of this article confirm that there are no conflicts of interest to disclose.

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