CLINICAL COMMENTARY

Middle ear implant for mixed hearing loss with malformation in a 9-year-old child

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Summary

Objective: The aim was to report the results of the first case in France of pediatric auditory rehabilitation with a middle ear implant and to discuss the putative indications with this new therapeutic option in children.

Patient and methods: A prospective study over 18 months on clinical and audiometric results after a middle ear implantation with a Vibrant Med-El® implant in a 9-year-old child with mixed hearing loss.

Results: Postoperative unaided pure tone audiometry (PTA) was unchanged by the surgical procedure. After 18 months of implant use, the mean PTA loss in free-field warble tone audiometry was 33.75 dB and the intelligibility threshold was 30 dB. After 18 months of follow-up, the intelligibility threshold was improved by 25 dB in comparison with the preoperative results with two hearing aids. The implant worked perfectly well and the child did not show any complication during this period.

Conclusion: The reliability of the implant and the quality of the auditory results obtained in this case and in a limited number of cases in the world make the Vibrant Med-El® a new therapeutic option in hearing loss in children with bilateral auricular atresia.

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Introduction

Rehabilitation of permanent deafness in children diagnosed before the age of 3 months with early adaptation of hearing aids can provide significantly better levels of oral language development [1,2]. In the most frequent cases in which the outer ear anatomy is normal, conventional behind-the-ear amplifying hearing aids are proposed. In cases of bilateral major auricular atresia, the diagnosis of deafness is suspected at birth. The level of pure or mixed conductive hearing impairment is quickly confirmed with auditory evoked potentials and infant audiometry. Once the diagnosis has been made, it is possible to provide these children with bone conduction hearing aids. Since 2002, beginning at the age of 1 month, these hearing aids can use the BAHA® softband, which, in cases of pure conductive hearing impairment, provides a mean aided threshold of 27 ± 6 dB HL [3].

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Bone stimulation with a headband remains an effective alternative with a mean aided threshold of $25 \pm 6$ dB HL, but wearing this device is often uncomfortable because of the constant pressure on the head [3]. A BAHA® can be suggested before the age of 6 years, but the possible cutaneous and fixture complications are such that during this period this is not advised [4]. The US Food and Drug Administration recommends the BAHA® after 5 years of age.

After 6 years of age, a BAHA® or functional aural surgery in cases of type I or IIa atresia according to the Cremers’ classification [3] is currently being debated. Beyond these situations that are a priori well codified, the advances of middle ear implant (MEI) technologies are opening new perspectives for restoring hearing. In 2007, for the first time in France a team in Lyon implanted such a device in a 14-year-old child presenting bilateral auricular atresia in a context of Franceschetti’s syndrome with the fully implantable Otologics Carina MEI [5]. Another MEI, the vibrant Med-El®, extended its indications in September 2007 to mixed or conductive deafness in the adult [6] and to children in June 2009.

The objective of this case study was to report the results with more than 18 months of follow-up of the first implantation in France of a Vibrant Med-El® in a 9-year-old child with bilateral auricular atresia.

**Case study**

The child was 9 years old at the time of implantation and presented stenosis-type atresia of the external auditory canal (EAC). The diagnosis of hearing loss had been confirmed at the age of 1.5 years with an early auditory evoked potential (EAEP) study (right and left ear threshold, 70 dB) in accordance with the infant audiometry study. Conventional amplifying hearing aids were initiated 1 month after diagnosis. A bone conduction hearing aid with a headband was suggested but refused by the family. In the child’s neonatal history, there had been septicemia following vesicourethral reflux. A pyelocaliceal malformation required corrective surgery. Karyotype genetic testing identified monosomy 18. This child received initial orthophonic and psychomotor care. He then attended preschool and primary school in an inclusion class for disabled children with multidisciplinary care including orthophonic and psychomotor work, psychological support, and special education. For 7 years, hearing aids were worn regularly despite the recurrent Larsen-effect problems and the many absences because of recurring episodes of eczema in both EACs.

The child consulted in 2007 enquiring about the possibilities of hearing restoration. The results of the preoperative exams were as follows.

Functional auditory testing showed bilateral nonevolving type II intermediate mixed symmetrical deafness. The mean air conduction thresholds (measured at 500, 1000, 2000, and 4000 Hz) were 65 dB on the right and 60 dB on the left. The mean bone conduction thresholds were 23.75 dB on the right and left (Fig. 1). The mean aided thresholds measured with warble tones were 43.75 dB for a mean overall aided gain of 23 dB (Fig. 2). The intelligibility thresholds evaluated based on the Lafon French standardized children’s disyllabic word lists were 70 dB on the right and 65 dB on the left for air conduction and 35 dB on the right and 30 dB on the left for bone conduction. The aided intelligibility thresholds with two hearing aides were at 55 dB (Fig. 3). The electrophysiological thresholds obtained with auditory evoked potentials of the brainstem were 60 dB on both the right and left.

The CT scan of the petrosal bone showed recessed EACs with no differentiation of the tympanic membrane. The normally ventilated tympanic cavities were small in volume and there was bilateral incudomallear fusion with a normal descending branch of the incus and normal stapes. The cochleae, the vestibular aqueducts, and the internal auditory canals showed no morphological deformities (Fig. 4).
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The therapeutic options proposed to the parents and the child included functional ossiculoplasty surgery with meato-plasty or BAHA® or a Vibrant Med-El® implant. The third option, the MEI Vibrant Med-El® implant, was retained after informing the parents on the risks involved (i.e., VIIth nerve injury, cochlear risk, the possibility that implant fixation would not be achievable, and unknown results in children) and acceptance by the parents and the child.

The surgery proceeded via a posterior approach with a retroauricular incision. The musculoperiosteal flap was cut and the periosteum was elevated from the mastoid process, thus creating a pocket. After a canal wall-up mastoidectomy, a posterior tympanotomy was performed. Despite the small size of the tympanic cavity, the malleus and the incus were cleared and the float mass transducer was clipped onto the long process of the uncus, which had a reduced mobility.

The child left the unit 2 days after the surgery and the implant was activated 2 weeks later using a No.2 magnet. Cutaneous healing was perfect with no sign of irritation. No modification of cochlear function was noted and the device was set, with immediate results. The child’s first reaction was surprise once audition had become clear, without the disadvantage of the EAC occlusion.

After 1 month of experience with the implant, the mean aided thresholds at conversational frequencies were 37.5 dB with 100% vocal intelligibility at 50 dB with the Lafon French standardized children’s disyllabic word lists (Fig. 3).

After 12 months of wear, the device was set again. The aided thresholds remained stable at 37.5 dB with a slight improvement in vocal intelligibility: 100% at 45 dB (Fig. 3). The child wore the implant continuously and had abandoned the contralateral hearing aid. Reliability was perfect.

After 18 months of wear, the device was readjusted. The aided thresholds improved to 33.75 dB, as did vocal intelligibility: 100% at 40 dB (Fig. 3). The mean aided tone gain was 23.75 dB. The child wears the implant continuously. Reliability is perfect. During the last adjustment, the child requested a contralateral MEI.

Discussion

This first French experience of a Med-El® MEI in a 9-year-old child with the purpose of correcting mixed congenital deafness confirmed the feasibility of this technique for this indication.

After 18 months of follow-up, this technique has demonstrated its stability and reliability. The child had undergone no repair of the external processor and the adjustments of the processor were proposed systematically every 6 months. The adjustments were minimal each time. The results in terms of auditory perception were deemed perfect by the child and his family with the request for bilateral implantation because the contralateral hearing aid had been abandoned, which was appropriate because of the auditory asymmetry, the new hearing quality, the comfort of the absence of EAC occlusion, as well as the recurring problems of eczema on the side aided by the classic hearing aid.

In this type of clinical presentation, other therapeutic options, including functional surgery with meato-plasty and ossiculoplasty and BAHA® were discussed with the parents and the child.

The Cremers’ classification of congenital auricular atresia [3] distinguishes four types. In typeI, the tympanic membrane is present but hypoplastic. TypeIIa consists in partial atresia of the EAC, whereas typeIIb is full atresia. In typeIII, atresia of the EAC is full, with malformation of the tympanic cavity. According to Cremers et al., reconstructive surgery of the EAC is not possible in types I and IIa [3]. In our clinical case, the malformation was type I or IIa, which therefore required analyzing the functional results as well as the follow-up and potential complications, notably meato-plasty. Studies have shown the surgical possibilities of canaloplasty, with their results summarized in the study by Evans and Kazahaya [7]. The prerequisite to any analysis is to define the criteria for audiometric success in relation to the schoolchild’s needs in terms of auditory perception. The perception threshold should be 15 dB HL or less if the bone conduction is normal or at the level of bone conduction if sensory participation has been added [7]. According to this same study, in cases of auditory canal reconstruction, the mean gain in audition was 17.3 dB and more than 93% of the subjects needed complementary auditory amplification. Late complications such as stenosis recurrence, recurrent external otitis, and cholesteatoma were noted. The conclu-
sions favored BAHA® as regards the quality of the functional results at the short-, intermediate-, and long-terms, with a mean gain in hearing of 31.8 dB. In our clinical case, no complication was noted in 18 months and the intelligibility obtained with the Vibrant Med-El® was equivalent if not slightly better than in direct bone conduction.

BAHA® “guarantees” the functional results, which explains its priority indication in the majority of congenital auricular atresia cases. However, the esthetic aspect, the psychological apprehension for a transcutaneous implant, the risks of local inflammation, and the consequent need for daily local hygiene makes some parents reluctant to choose this option.

The indication for the Vibrant Med-El® in mixed deafness and adult conduction made this option possible in the clinical case reported herein. According to the study conducted by Coletti et al., there exist different vibroplasty techniques in relation with the ossicular anatomy encountered [8]. Notably, the Float Mass Transducer can be placed in the round window. The stability of this technique’s long-term results have not been clearly established; consequently, in children it is more suitable to favor the surgical technique aiming to attach the FMT to the descending branch of the incus or even the stapes, but additional results in adults would solidify this indication.

The MEI in children now has its place among the therapeutic options for bilateral congenital conductive or mixed hearing loss. This indication has been in the validation phase for the Vibrant Med-El® since June 2009. The possibilities for this type of implant continue to be explored. For example, in 2009 Frenzel et al. [9] reported seven cases of hearing rehabilitation using the Vibrant Med-El® in cases of unilateral hearing loss stemming from major unilateral aplasia. In one case, the FMT was placed on the descending branch of the incus, in three cases on the stapes, in two cases on the base of the stapes, and in two cases in the round window. The bone curves were unchanged between the pre- and post-operative phases. The functional results at 8 months were 23.8 dB HL for the mean aided thresholds, for a mean aided gain of 45.5 dB HL. Aided intelligibility was 64% at 50 dB.

The therapeutic options for congenital bilateral mixed or conductive deafness due to auricular atresia in children should be discussed in light of this clinical case. Perspectives will undoubtedly open up in the management of this type of unilateral hearing loss in the near future.

Conflicts of interest statement

The authors have not communicated conflicts of interest.

References