International Journal of Pediatric Otorhinolaryngology 79 (2015) 1896–1900

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Expanding pediatric cochlear implant candidacy: A case study of electro-natural stimulation (ENS) in partial deafness treatment

Henryk Skarzynski [a], Artur Lorens [a,*], Beata Dziendziel [a], Piotr H. Skarzynski [a,b,c]

[a] World Hearing Center, Institute of Physiology and Pathology of Hearing, Kajetany, Mokra 17, 05-830 Nadarzyn, Poland
[b] Heart Failure and Cardiac Rehabilitation Department, Medical University of Warsaw, Banacha 1a, 02-097 Warszawa, Poland
[c] Institute of Sensory Organs, Kajetany, Mokra 1, 05-830 Nadarzyn, Poland

ARTICLE INFO

Article history:
Received 16 June 2015
Received in revised form 27 August 2015
Accepted 29 August 2015
Available online 6 September 2015

Keywords:
Cochlear implant
Partial deafness treatment
Hearing preservation
Electric complementation
Electric-acoustic stimulation
Electro-natural stimulation

ABSTRACT

Background: Some adolescents have hearing impairments characterized by normal or slightly elevated thresholds in the low and mid-frequency bands (below 1500 Hz) and nearly total deafness in the high frequency range. These patients often remain beyond the scope of effective hearing aid treatment.
Case report: This study presents the case of a 16-year-old adolescent with good hearing in the range 125–1500 Hz and deafness at other frequencies. An implant was used to restore hearing at high frequencies, while preserving low and mid frequency acoustic hearing in the implanted ear. This is described as electro-natural stimulation (ENS) of the inner ear.
Conclusions: The results demonstrate that low and mid frequency hearing (up to 1500 Hz) can be preserved using the round window surgical technique. A substantial improvement in speech discrimination was also observed when electrical stimulation on one side was combined with acoustic stimulation on both sides. There is scope to extend qualifying criteria for cochlear implantation to include adolescents who are suited to ENS.

1. Introduction

Traditionally, children are considered candidates for a cochlear implant (CI) if they have bilateral severe to profound sensorineural hearing loss (SPHL). It has been well established that early cochlear implantation leads to many benefits for deaf children, including an almost normal language development [1–3]. Children implanted in infancy outperform their peers who receive implants later, and often approach the abilities of normal-hearing children of the same age [4,5]. This means that adolescents with prelingual deafness are now regarded as a special population that are only reluctantly considered suitable for a CI [6,7].

However, recent work with prelingual adolescents who have residual hearing and oral communication, and who have been using hearing aids (HA) since childhood, has shown that their speech perception can be markedly improved after cochlear implantation [8]. There are now dedicated adolescent cochlear implant programs which address the unique challenges faced by adolescents with SPHL who struggle with hearing aids [9].

Notably, however, there is another group of adolescents whose hearing impairment is characterized by normal or slightly elevated thresholds in the low and mid-frequency bands with nearly total deafness in the high frequency range. This type of partial deafness remains beyond the bounds of effective treatment by hearing aids. The only practical way to improve hearing is to complement the normal hearing at low and mid frequencies with electric stimulation at higher frequencies using a cochlear implant. This arrangement is described as electro-natural stimulation (ENS) of the inner ear [10].

In previous reports, we have demonstrated the efficacy of applying electric stimulation to totally (or almost totally) inactive regions of the inner ear using a cochlear implant, thereby complementing the preserved low-frequency hearing [11–20]. It has been called partial deafness treatment–electric complementation (PDT–EC) [16,18,19]. In the conception introduced by Skarzynski in 2002 in adults and in 2004 in children, frequencies up to 500 Hz are complemented [12,14], providing a significant extension of the previously accepted indications for cochlear implantation. Subsequently, the underlying theory behind PDT has been further developed, and a thorough description was published in 2010 [16] (Fig. 1).

In summary, PDT has three approaches for dealing with three distinct groups of patients [15,17,18]. (1) EC in patients who have normal or slightly elevated thresholds at low frequencies and

* Corresponding author.
E-mail address: a.lorens@ifps.org.pl (A. Lorens).

http://dx.doi.org/10.1016/j.ijporl.2015.08.040
0165-5876/

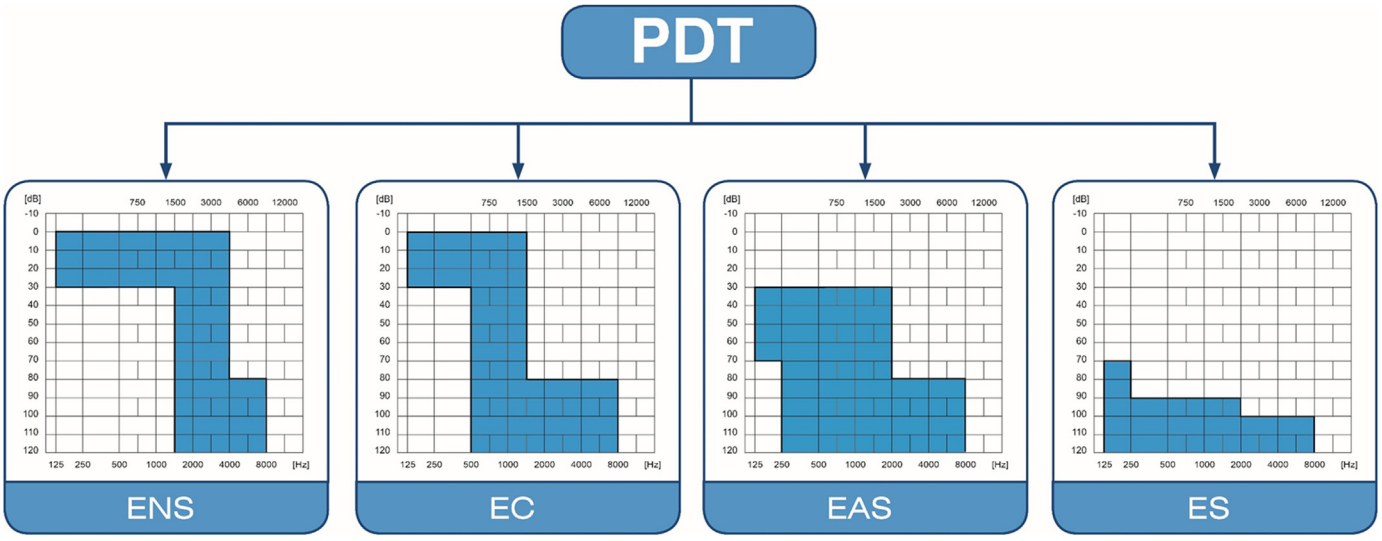

Fig. 1. Skarzynski's original partial deafness conception with the addition of electro-natural stimulation (ENS).

almost total deafness at higher frequencies. Here, non-amplified low frequency hearing is complemented by electric stimulation with a cochlear implant. (2) Electric-Acoustic Stimulation (EAS) in patients with mild-to-severe hearing loss at low frequencies and profound hearing loss at high frequencies. In this group, low frequency hearing is amplified and combined with electric stimulation in the same ear. (3) Electric Stimulation (ES) in cases with non-functional hearing.

This report presents the results of treating an adolescent with a hearing deficit diagnosed as partial deafness, in whom the natural hearing below 1500 Hz was complemented by means of electric stimulation in the frequency range above 1500 Hz. We describe the electric complementation of the almost natural hearing up to 3000 Hz as ENS [10]. This represents a new step forward, opening up the possibility of extending current indications for cochlear implantation.

2. Case description

The case was a 16-year-old boy who, at the time of implantation, had fully efficient hearing in the frequency range 125–1500 Hz and deafness at other frequencies (Table 1; Fig. 2). The bilateral hearing loss was prelingual, probably caused by an ototoxic drug (gentamicin) used shortly after birth.

2.1. Surgery

Since the patient needed only complementation of his natural low and medium frequency hearing, implantation presented a major challenge to the surgeon because the procedure needed to preserve pre-operative hearing. Despite the relatively good extant natural hearing, the decision to use a cochlear implant on this patient was based on his poor level of speech understanding. The surgery was performed in accordance with the six-step surgical procedure developed by Skarzynski [11–19]:

- Step 1: Conservative antromastoidotomy, preceded by harvesting a bone chip from the mastoid cortex, which was later used to close the middle ear space.
- Step 2: Typical posterior tympanotomy with exposure of the round window.
- Step 3: Puncture and central incision of the round window membrane.
- Step 4: Careful insertion, at first by hand and then with forceps, of a flexible active electrode (of the Med-El Flex EAS type) through the round window into scala tympani to a depth of 18 mm.
- Step 5: Fixation of the electrode in the round window niche with tissue glue and a piece of fascia.
- Step 6: Fixation of the internal part of the implant into a drilled well, closing the mastoid cavity with the bone chip harvested at the beginning of the procedure, and suturing the incision.

A steroid prescription was given, with 0.1 mg/kg/day of dexamethasone administered intravenously in two equal doses per day (on the first day about 30 min before the surgery and 3 h after surgery). Dosing with steroids was continued for 3–4 days. The dexamethasone solution was also administered locally into the middle ear before opening a round window when the bed for an implant was being prepared.

Selection of the optimal length of electrode considered the effect of an electrode according to the outer hair cells (OHC) gain predictions described by Skarzynski et al. [19]. Loss of OHC function could have an important effect on properties of residual hearing such as frequency selectivity and dynamic range. To avoid such effect, we routinely prescribe an insertion depth according to the relatively good level of preoperative residual hearing at 1500 Hz.

Over a 7-year follow-up period, there have been regular evaluations of (1) pre-operative hearing preservation in both the operated and non-operated ear; and (2) improvements in speech discrimination.

2.2. Hearing preservation

All audiometric tests were performed using calibrated audiometers with maximum outputs of 90 dB HL at 125 Hz, 105 dB HL at

Table 1
Case description: patient summary.

Sex:	Male
Age at diagnosis:	4
Etiology:	Perinatal load and ototoxic medication
Age at operation:	16
Ear	Left
Implant type	Med-El Pulsar CI100
Surgical method	Round window
Insertion	Partial