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Long-term clinical outcomes of cochlear implantation in children with symptomatic epilepsy

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ABSTRACT

Objectives: To describe long-term clinical outcomes of cochlear implantation in deaf children with symptomatic epilepsy.
Materials and methods: A retrospective data analysis review of patients implanted at the Cochlear Implant Center of the University of Parma, Italy, was performed, searching for implanted children with a confirmed diagnosis of symptomatic epilepsy. Clinical data, imaging findings, pre- and post-operative epilepsy pattern and EEG traces were analyzed; communicative skills were assessed using the Profile of Actual Linguistic Skills.
Results: Search retrieved two patients affected by profound bilateral sensorineural hearing loss and symptomatic epilepsy (associated respectively with methylmalonic acidemia and cerebral palsy). After careful parental counselling both patients were offered and underwent cochlear implantation. Activation and use of cochlear implant did not determine substantial changes of pre-existing seizure pattern and EEG traces. Both patients showed substantial development of their communicative abilities.
Conclusions: Cochlear implantation in children with symptomatic epilepsy did not determine variations in seizure pattern or EEG traces. Both patients experienced substantial benefit from cochlear implantation.

1. Introduction

Epilepsy is a brain disorder characterized by recurrent, unpredictable interruption of neural function, as defined by the International League Against Epilepsy (ILAE) [1,2]. Epilepsy is divided in four etiologic categories: idiopathic, symptomatic, provoked and cryptogenetic [3]. Symptomatic epilepsy is associated with central nervous system (CNS) abnormalities and clinical features of an underlying CNS disease. Symptomatic epilepsy may complicate several hereditary or acquired neurological disorders, including developmental anomalies and neurocutaneous syndromes [4].

Recent studies focused on the benefit of cochlear implantation (CI) on deaf children with additional disabilities (i.e. cerebral palsy, CNS and inner ear malformations, CHARGE syndrome) [5–10] while only few papers focused on results of CI in children with epilepsy [11–13].

CI may give significant benefits in rehabilitating young patients with profound sensorineural hearing loss (SNHL) and associated symptomatic epilepsy, although electrical stimulation of the inner ear carries a theoretical risk of triggering or worsening seizures.

Purpose of this study was to report our experience in CI in deaf children with symptomatic epilepsy and to investigate whether electrical stimulation of the auditory pathways may determine seizures or exacerbate the natural course of epilepsy.

2. Materials and methods

After institutional review board approval, a retrospective data analysis of patients implanted at the Cochlear Implant Center of the University of Parma, Italy, was performed, searching for implanted children with a confirmed diagnosis of symptomatic epilepsy. Children were examined by developmental paediatricians, paediatric neurologists, audiologists and speech pathologists and careful counselling was given to parents before obtaining informed consent for CI. Preoperative assessment included

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auditory brainstem responses, otoacoustic emissions, acoustic immittance, behavioural and routine audiometric procedures under earphone and in free sound field. Children underwent high resolution computed tomography (HRCT) of the temporal bones and brain magnetic resonance imaging (MRI). Mentally retarded children were evaluated using the Profile of Actual Linguistic Skills (PALS). The PALS is a tool that allows evaluation of verbal and non-verbal communicative abilities assigning the child to one of three different linguistic categories: preverbal, transitional and functional language (Table 1) [14,15]. Performances are measured and rated for five linguistic levels: communicative, receptive, expressive, voice and speech skills. The profile is completed by gathering observations from the speech pathologist as well as from other people involved with children rehabilitation (i.e. parents, teachers). An Italian version of PALS was used in the preoperative evaluation and to assess communication skills after surgery [16].

3. Results

Complete medical records with a follow-up time of at least 60 months after CI were available for two children; both patients presented with profound SNHL, mental retardation and symptomatic epilepsy.

3.1. Case number 1

Patient 1 was a 4 years old boy affected by *Methylmalonic Acidemia* (MMA), an inherited disorder involving the metabolic pathway of cobalamin. Seizures presented for the first time one month after birth and were treated with vigabatrin and adrenocorticotropic hormone. Epilepsy was classified as West syndrome [2,17]. At time of evaluation in our department, the patient showed severe mental retardation with generalized hypotonia. Brain MRI showed diffuse demyelination, ventricular dilatation and slight brain atrophy. Inner ear and cranial nerve VII–VIII were normal. Temporal bones HRCT was normal. Audiological evaluation was consistent with bilateral profound SNHL; the child had been fitted with hearing aids since 15 months of age with poor results on hearing abilities and language development. Preoperative electroencephalography (EEG) during sleep showed absence of the hypsarrhythmic pattern associated with West syndrome in the context of a generalized slowed rhythm for patient's age. A Nucleus C24 Contour Advance device (Cochlear Corporation, Melbourne, Australia) was implanted in the right ear at age 4 without complications. The implant was activated after one month and fitted using the Advanced Combination Encoder strategy with 12 maxima and a stimulation frequency of 900 Hz. No episodes of implant related seizures were evident at activation and with subsequent implant use. One year after surgery the patient developed partial epileptic seizures arising at awakening with clonic movements of the right arm and leg. Seizures were treated by sodium valproate as supporting therapy. Last EEG during sleep, performed 60 months after CI, showed a slowed rhythm with a spike-wave-pattern and no modifications related to cochlear implant use.

Table 1
Profile of actual linguistic skills.

Categories of linguistic skills		
Preverbal	Functional	Transitional
Child are in a pre-lexical stage or may use signs to communicate	Recognizable words and simple expressions are reported by caregivers and may be elicited during assessment	Spoken language is the primary mean of communication

3.2. Case number 2

The second patient, a 6 years old girl, was admitted to our Department for profound bilateral deafness in the context of cerebral palsy (CP) following maternal cytomegalovirus (CMV) infection during pregnancy; she suffered from generalized epilepsy since 4 years of age. The patient showed a right spastic hemiplegia type of CP. She had been diagnosed with eyelid myoclonia with absences [18] and had only one episode of generalized tonic–clonic seizures at age 5. Epilepsy was controlled by ethosuximide and no episodes of seizure/absence were reported in the 12 months before surgery. Brain MRI showed subhemispheric polymicrogiria, atrophy of the left cerebral hemisphere and white matter gliosis. Inner ear and cranial nerves VII–VIII were normal. Temporal bones HRCT was unremarkable. Audiological evaluation was consistent with bilateral profound SNHL. The child had been fitted with hearing aids since 12 months of age with no substantial benefit. Preoperative sleep-recorded EEG showed paroxysmal abnormalities with spikes over central and parietal brain regions. A Digisonic SP device (MXM Corporation, Antibes, France) was implanted in the left ear at age 6. The implant was activated and fitted one month later using the Main Peak Interleaved Sampling strategy with 9 maxima and a stimulation frequency of 600 Hz [19]. No episodes of implant related seizures were evident at activation or with subsequent implant use. The patient developed generalized seizures at a temporal distance of 5 and 12 months after activation. *As in Case 1 postoperative EEG traces under background therapy (etohosuximide) performed 60 months after surgery showed persistence of spike wave discharge over central and parietal regions but no evidence of reproducible anomalies related to CI.*

Figs. 1 and 2 illustrate communicative abilities of the two patients as evaluated by PALS; all competences evolved from a preoperative preverbal level to a postoperative transitional and functional level in both cases.

4. Discussion

Epilepsy and severe to profound SNHL may coexist in various clinical syndromes; CI may give significant benefits in profoundly deafened patients with symptomatic epilepsy in which CNS alterations may determine additional handicaps. Electrical stimulation of the auditory pathway in patients with symptomatic epilepsy carries a theoretical risk of triggering epileptic episodes as these patients are affected by structural or metabolic CNS conditions associated with an increased risk of developing seizures.

There are two possible ways by which CI could worsen/trigger seizures: mechanical irritation of brain cortex [i.e. epidural bleeding during mastoid surgery] and uncontrolled activation of the cortex due to retrograde cranial nerve electrical stimulation [13]. In the last case, structural CNS alterations as in children with symptomatic epilepsy, may theoretically favour de novo seizure development or modification of a preexisting epileptic syndrome.

When defining a seizure as related to CI, a temporal relationship with CI use should be obvious; CI may elicit seizures at activation and with subsequent implant use. *In both circumstances, EEG traces should demonstrate alterations which should be reproducible with cochlear implant use (*i.e. *discharge from temporal area).*

Literature review retrieved 2 case reports and a case series including 7 patients (Table 2) [11–13]. Six patients had symptomatic seizures before CI while 3 developed seizures after a variable amount of time from CI. In patients with symptomatic seizures before CI [6/9], there were no changes in the EEG traces or epilepsy pattern after surgery. In 3 patients seizures appeared 3 months, 2 and 5 years after surgery; one of these patients had meningitis 3 months after CI but seizures appeared 5 years after surgery. In