Post binaural cochlear implant fluctuating hearing loss - A case study

Author(s): Ahmad H. Salahaldin, Khalid A Hadi, Razan Alfakir, Abdulbari Bener

Vol. 14, No. 1 (2010-01 - 2010-06)

Ahmad H. Salahaldin¹, Khalid A Hadi¹, Razan Alfakir¹, Abdulbari Bener²,³

(1) Department of ENT, Audiology Unit, Rumailah Hospital and Hamad General Hospital, Hamad Medical Corporation, Doha, State of Qatar
(2) Department of Medical Statistics and Epidemiology, Hamad General Hospital, Hamad Medical Corporation, Weill Cornell Medical College, Doha, State of Qatar
(3) Department, Evidence for Population Health Unit, School of Epidemiology and Health Sciences, The University of Manchester, Manchester, United Kingdom

Abstract

So far, there seems to be no sufficient therapy for stopping the progression of hearing loss. It has been shown that a cochlear implantation is a worthwhile procedure if the patient is almost deaf. The aim of current study is to review literature and compare them with our case study report of some of the common conditions related to the etiology of Fluctuating Sensorineural Hearing Loss (FSNHL). This is supported by similar clinical picture, outcome and is commonly considered to involve cochlear implant as a possible form of their treatment.

Design: Case reporting study.
Setting: Ear Nose Throat (ENT) Audiology Unit, and Endocrinology outpatient clinics of the Hamad General and Rumailah Hospitals, Hamad Medical Corporation.
Case study: Full clinical, audiological and laboratory evaluation and patient follow up with comprehensive review of literature.

The baby’s hearing, speech, social and mental development were reviewed and was decided that, in order to improve the state of hearing, speech performance and future education, the baby is considered for cochlear implant. Thus he started to have preparatory speech and audiological rehabilitation sessions for Cochlear implantation (CI) prior to surgery and found to score well in them. The baby was fitted with cochlear implant in both ears at consecutive dates. Following the first CI intraoperative electrical check was normal and postoperative telemetry and programming was done properly at later date and hearing remarkably improved with discrimination score (DS) score of 100% at 40 dB HTL FF. The patient continued to have the same evaluation figures till early 2007 and start to deteriorate after.

The patient started medication with Steroid treatment on the account of diagnosis of AIED. He was started with prednisolone 20 mg, 4 times daily for one day followed by 20mg tds (60mg) for 3 days. The patient showed remarkable improvement that hearing improved to normal in the right ear with DS of 100% at 40 dB but to HTL of 50 dB with poor DS 40% at 100dB in left ear. All treatment showed a remarkable improvement of hearing and speech discrimination. An improvement in addition of methotrexate is done on the basis of reducing steroid dosage without hearing deterioration progressing with no registered serious side effects.

All performed treatment has been revealed a remarkable improvement of hearing and speech discrimination with no registered serious side effects.

Key Words: Post binaural, cochlear implant, fluctuating hearing loss, case study

Accepted July 06 2009

Introduction
Fluctuating sensorineural hearing loss (FSNHL) is a well reported condition and is widely described and discussed in the literature. But as far as we know, the occurrence of the condition in association with binaural cochlear implant (CI) is interesting and is scarcely reported in the literature. Hence, it is worth describing and discussing.

The aim of current study is to review literature and compare them with our case study report of some of the common conditions related to the etiology of Fluctuating Sensorineural Hearing Loss (FSNHL). This is supported by similar clinical picture, outcome and is commonly considered to involve cochlear implant as a possible form of their treatment. These include; fluctuating autoimmune sensorineural hearing loss (FSNHL), Auditory Neuropathy and Enlarge vestibular aqueduct syndrome. Other conditions causing FSNHL like; Meniere’s disease and other causes of Endolymph hydrops, Luetic labyrinthitis, Perilymph fistula, Multiple Sclerosis.

An autoimmune reaction ensues when the immune system’s ability to distinguish between self and nonself is disturbed, [1] who describes it as Fluctuating Autoimmune Inner Ear Disease (AIED). It is clinically characterized by the presence of a rapidly progressive, often fluctuating and mostly bilateral SNHL over a period of weeks to months. It is associated with poor speech discrimination (79% cases), tinnitus and aural fullness (25-50%), vestibular symptoms (50%) and occasionally existing symptoms and signs of systemic autoimmune condition such as Rheumatoid Arthritis, ulcerative colitis systemic lupus erythematosis and poly arthritis nodosa(15-30%). This is commonly seen in women of 20 – 50 years of age. Its clinical diagnoses, based on its distinct clinical course, immune test results and treatment response, which can be regarded as the most important diagnostic finding which brings improvement in hearing observed with a trial of steroids or immunosuppressant [2,3,4].

AIED is rare accounting for less than 1% of all cases of hearing loss or dizziness [2]. The precise incidence is controversial. About 16% of persons with bilateral Meniere’s disease, and 6% of persons with Meniere’s disease of any variety may have symptoms due to immune dysfunction.

Treatment of AIED involve a number of protocols of which include the use of predneslone or dexam男篮honesone for four weeks followed by maintenance dose which is usually quite effective and may need to be stopped because of the side effect of steroid treatment.

Recently, Transtympanic steroid therapy had been tried with good results [6]. A trial of low molecular weight heparin (enoxaparin) was found to be effective [7], yet it is recommended that larger trial is necessary before the therapy can be widely practiced. Genetic manipulation and application of inner ear cell therapy [8] and surgical treatment using cochlear implant had recently gained good place in treatment especially in bilateral cases [9]. Auditory neuropathy/Auditory Dyssynchrony (AN/AD) is a hearing disorder characterized by an absent or severely abnormal auditory brainstem response (ABR), with preservation of the cochlear microphonics (CM) and otoacoustic emissions (OAEs). Clinically using OAE and ABR makes it possible to identify, if the function of the VIII nerve were distorted while the function of cochlear outer hair cells was normal [10]. A lesion caused by an insult that may result in injury to synaptic junction between inner hair cells and dendrite of spiral ganglion neurons, or in damage to dendrite of spiral ganglion itself or in axonal damage to the nerve itself [11].

A number of risk factors have been speculated to contribute to the etiology of AN/AD including; Neonatal anoxia, neonatal hyperbilirubinemia, admission to NICU, congenital brain abnormalities, low birth weight and severe prematurity (<28 wk). The use of gentamycin sulphate and other ototoxic medications, cerebral palsy, and genetics or family history of auditory neuropathy in 36% with a possible recessive inheritance pattern also documented [12]. The association with viral infection, seizures and high fever were also reported [11]. AN/AD was observed in many syndromic peripheral neuropathies, like Friedreich’s ataxia and hereditary sensorimotor neuropathy (HSMN) formerly known as Charcot-Marie-Tooth disease [13].

Complete data regarding the incidence of auditory neuropathy in the pediatric population come from an Australian study of a newborn hearing screening program [14]. The study conducted screening in 5199 infants and 12 out of them met the criteria of auditory neuropathy, that concluded to have affected with 0.23% of the general population. In a 2002 review [15], suggested that approximately 1/10 children reported to have hearing loss who are severely affected found to have AN/AD. In conclusion overall AN/AD is rare and can be found in an estimated 1-3 children per 10,000 births irrespective of sex, age or racial differences [11].

AN/AD is managed through a protocol of close watch once diagnosed during the first 3 months of life. There had been a lot of controversy regarding the use of hearing aids. The danger to damage the outer hair cells of the cochlea (OHC) by loud noise was tried to be solved through careful use of amplification level, frequent monitoring of the hair
cells functions through the use of OAE and also by the use of FM system which improves the signal to noise ratio [16]. In a study by [17] only 17% of the 50% of children with AN/AD who tried amplification received benefit.

Cochlear implant (CI) has been considered and attempted by many authors as a form of treatment for AN/AD. It seemed logical if the causative pathology was in the inner hair cells of the synapse. It is mostly like bypasses the lesion; on the other hand if the cause is in the cochlear nerve itself the outcome would be doubtful. Thus determining suitability of patient for CI would require careful evaluation of speech discrimination ability and careful determination of the site for etiopathology [18,19].

Fluctuating hearing loss can be due to enlarged vestibular aqueduct (EVA) is another form of inner ear dysplasia and is regarded by [20] as one of the commonest inner ear abnormality. It is rather unique, because it has been associated with delayed onset of SNHL. The EVA syndrome is typically bilateral. Children are usually born with normal or mildly impaired hearing that gradually deteriorates through childhood into adolescence and early adulthood. Hearing levels are variable, although at least 40% eventually develop profound SNHL [21]. As with other inner ear malformations, there is a tendency for the baby to suffer sudden decrements of hearing, particularly after head trauma. Fluctuation of hearing is common and usually affects one ear at a time, and disequilibrium symptoms might occur [20]. Although EVA has been reported to be inherited in a recessive manner, little is known about the genetic basis of this hearing loss.

So far, there seems to be no sufficient therapy for stopping the progression of a loss of hearing. It has been shown that a cochlear implantation is a worthwhile procedure, if the patient is almost deaf and found to be effective for both adults and children with EVA and specifically successful treatment in cases of bilateral very severe progressive sensorineural hearing loss due to bilateral EVA, [22,23,9].

This case study including above review of literature, intend to discuss and explore diagnostic possibilities which seems to involve possible combination of one or more of the above condition in their production, complaint and their progression to magnify the validity of treatment and outcome.

**Subject and follow up**

QA (DOB: 11th DEC 1993) a non Qatari boy seen for evaluation in the Audiology clinic for the first time at 13 March 2004 accompanied by his parents with Delayed Language Development (DLD), and difficult to produce intelligible speech. The patient is a Caucasian, with no gross bodily abnormality, conscious and cooperative. He is the first son of upper mid class socioeconomic family of four with only a younger sister and have a history of being born prematurely early at 34 weeks gestation with clearly uneventful pregnancy and no significant prenatal problem. The boy was born at home urgently, before reaching the hospital. He did not cry immediately soon after the birth and started to turn blue. Cyanosis continued for about 45 minutes. Urgent measures were taken and were admitted to the Neonatal intensive care unit (INCU) for resuscitation and further management. He was admitted for 12 days, during which he developed Jaundice that eventually reached high level that warrant exchange blood transfusion twice. At one session he developed septicaemia and treated accordingly. The baby’s weight at the time of delivery date was 2200 g and reduced to 1500 g during his discharge from NICU. There was no family history of congenital hearing abnormality. The baby sent home with a cover of high dosage of (Gentamycin) and kept well insulated in an incubator for the next 2 months. During that time and the following months he was under the follow up of a pediatrician. Then he was referred to a pediatrics neurologist for further evaluation of neurological and mental development. Electroencephalography (EEG) and auditory brainstem evoked (ABR) testing were done and showed normal. There was no clear response to maximum stimulation of 90 dB nHL indicating that he had bilateral profound sensorineural hearing loss with hearing threshold level worse than 90 dB nHL. Behavioral testing revealed no response to maximum free field stimulation in both ears. Accordingly it was decided to be fitted with powerful binaural hearing aids and to start him on auditory rehabilitation and speech therapy. The hearing aids prescribed were Bernafon powerful BTE type. At the age of one year and four months he was taken by his family to USA and another full checkup was done and eventually fitted with new binaural hearing aids. The results of hearing tests were the same and hearing aids were changed into Phonak type PP-C-L-4. The auditory rehabilitation and speech therapy continued till the age of 1 year and 10 months and as a result he started to produce early words.

**Motor Development**

During this period the baby showed definite delay in motor development, he was unable to control or raise his head, and could not even sit or walk until he reached the age of 3 years, despite dedicated efforts by his mother and occupational therapist.
Language and Speech Development:

The baby did not produce early sounds till the age of 3 months. With the help of hearing aids and with hard and persistent effort, he started to develop spoken words to the extent of producing 200 words at the age of five. The following months he was enrolled for auditory training and speech therapy in specialized training center in his home town till the age of seven years. At the age of 5 years, using pictures articulation test the baby was able to recognize all pictures included in the test, but with changes of sounds like /b/ into /p/ in all sentences.

Receptive Language Development and comprehension

Despite the fact that the baby did not have standardized tests for language evaluation, testing with certain pictures available for routine speech training at the speech therapy room and by using play tools and events it was clear that the baby appears to have simple but clear delay in understanding the meaning of some commonly in use words (like: Suff [class] and he appeared to have a vocabulary of good number of other words meanings.

Expressive Language Evaluation (ELE) was done using locally used tests. The baby appeared to have good deal of expressive meaningful words with definite flight of meaning of some words used during speech (like Suff (class) etc). Articulation seems to be fair and he is able to tell a short story. Grammatically he faced problem in changing singular into plural, without any problem with communication whether person to person or else.

Reviewing the baby's hearing, speech, social and mental development, it was decided that in order to improve the baby's state of hearing, speech performance and future education needs, the baby should be considered for cochlear implant. Thus he started to have preparatory speech and audiological rehabilitation sessions for Cochlear implantation prior to surgery and found to score well in them. No attempt to review the diagnosis of his condition was made. The baby was fitted with cochlear implant (MedEL C40+ device) in the left ear on (March 2003). The interesting findings during the left CI was, normal middle ear structure, including facial nerve course and intratympanic muscles and ossicles normally looking medial wall structures. A gash of perilymph fluid were noticed with cochleostomy in the inner ear with reduced cochlear turns and shorter intra cochlear course noticed that led 3 or 4 electrode points non inserted confirmed with intraoperative radiological monitoring. Yet CI intraoperative electrical check was normal and post-operative telemetry and programming was done properly at later date and hearing remarkably improved with discrimination score (DS) score of 100% at 40 dB HTL FF. The patients’ condition remained stable till 2007 and following that the patient hearing started to have deterioration and developed speech discrimination problem with DS of down to 50% score at 100dB FF which persisted and got worse that derives the treating team to implant him with another CI device in the right ear (MedEl pulsar) on 23/9/2007. Full preparation and investigation was done for the second CI surgery, with uneventfully full insertion of electrodes. Post operative improvement in hearing and DS was disappointing as there was mild or little improvement in speech discrimination in both ears that warrant trial of FM system on both ears in addition to existing CI which improves DS mostly in the left ear by small fraction +/- 10%. The warrant revision of the diagnosis, thus decided that, he should start with the steroid treatment as a possible candidate with AIED. Two month after the surgery (ie on 9/11/07) he was started with prednisolone 20mg, 4times daily for one day followed by 20mg tds (60mg) for 3 days.

The patient showed remarkable improvement with hearing reached to normal in the Left ear with DS of 100% at 40 dB but in the right ear to HTL of 50 dB with poor DS 40% at 100dB. Steroid therapy was then reduced to 30mg bid for another three days till 16/11/2007 and then to 40mg Bid for next 10dys. The steroid therapy was further reduced to 20mg OD from 2/12/07 till 30/12/07. All treatment was under the supervision of immunology specialist. During the period of medical treatment there has been a remarkable improvement of hearing and speech discrimination.

Table 1: CT Scan investigation

CT scan result – preoperative
CT scan report 15/9/2002 (before the 2nd cochlear implant): Finding: left and right EAC are unremarkable, pneumatization of the visualized mastoid air cells noted bilaterally. The semi circular canals are developed bilaterally, both cochleae were demonstrated with bilaterally developed copular, intermediate and ground cochlear turns. No hypoplasia or sclerosis was seen. No destructive osseous lesion was seen. Impression: – bilaterally developed cochlea. – No mal osseous development or sclerosis. No enlarged vestibular aqueduct.

Company evaluation of cochlear implant device[ Medel Pulser]
1. Implant assessment – good result
2. Expert telemetry – good result
3. Advanced fitting – same result
4. Post operative ESRT measurement, Objective fitting at university clinic, same results
5. CT scan – facial nerve quite close to middle turn of cochlea

Conclusion: No technical problem possibly medical issue after reimplant there could be a risk for the same situation.

Table 2: Recent Clinical investigation

<table>
<thead>
<tr>
<th>CBC count result</th>
<th>Differential count:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin 15.5 G/100ml,</td>
<td>Poly neutrophils 6930 /c.mm, (55%),</td>
</tr>
<tr>
<td>Basic biochemistry</td>
<td>Lymphocytes 4914/c.mm (39%)</td>
</tr>
<tr>
<td>Creatin 70.7 mmol/l (0.8mg/dl),</td>
<td>Monocytes 630/c.mm (5%).</td>
</tr>
<tr>
<td>Blood Urea Nitrogen 3.81 mmol/l (23 mg/dl),</td>
<td>Albumin 4.3 g/dl</td>
</tr>
<tr>
<td>Calcium 2.47 mmol/l (9.9 mg/dl),</td>
<td>Globulins 3.6 g/dl</td>
</tr>
<tr>
<td>Phosphate 1.80 mmol/l (5.6 mg/dl).</td>
<td>A/G Ratio 1:19</td>
</tr>
<tr>
<td>Hormonal study</td>
<td>Transaminase SGPT (alt)23 U/L</td>
</tr>
<tr>
<td>Thyroid function result</td>
<td>Alkaline Phosphatase 165.</td>
</tr>
<tr>
<td>Free Thyroxin (FT4) 1.8 ng/dl</td>
<td></td>
</tr>
<tr>
<td>TSH (Ultrase nsitive) 2.80 m IU/L</td>
<td></td>
</tr>
</tbody>
</table>

During tapering of the steroid dosage hearing deteriorated rapidly and dosage had to be increased back to 20mg for next 15 days till 15/1/08. The dosage was again reduced to 30mg per day along with an additional treatment with cytotoxic drug Methotrexate 2.5 mg raised to tds under the supervision of immunologist. Improvement was made in hearing with the addition of Methotrexate, and is taken on the basis of reducing steroid dosage without hearing deterioration that progressed with no registered serious side effects.

Last evaluation was done on 10/2/2008, and found that both tympanic membranes were intact. FF testing revealed threshold of RE CI device (45, 40, 25, 35, 40 dB) at(0.25k,0.5k,1k,2k,4kHz) and DS score of 50% at 90 dB level. LE with CI device (10, 15, 20, 25) at(0.25k,0.5k,1k,2k,4kHz) with excellent reliability and DS 100% at ordinary 70 dB level. Tympanometry using 265 Hz probe tone showed type A tympanograms, normal middle ear pressure, canal volume, and compliance in both ears acoustic reflexes were negative. It is worth mentioning that following cochlear implant improvement or deterioration is considered if HTL was shifted by a minimum of -/+ 10 dB from the previous tests conducted at regular follow up periods Table 1 and 2 shows latest investigation results.

Discussion

Having a young male patient with very severe to profound sensorineural fluctuating hearing loss which respond to treatment with steroid at first and then to Methotrexate complies with the diagnosis of AIED [1,2,3]. Although hearing fluctuation usually should involve one side and then affect the other [4] this could not be determined in our case, and the patient is male and the condition involves a younger age candidate, in contradiction with AIED which usually affect females at their 20-50 year of age, usually with the history of other systemic autoimmune disease in 15-30% of cases [3].

This put the diagnosis of AIED in doubt in addition to the fact that this patients was born prematurely with low birth weight, cyanosis, hyperbilirubinemia and had exchange transfusion followed by septicemia and exposure to treatment with ototoxic drug gentamycin. All the above factors can account for the possible diagnosis of auditory neuropathy [12]. Yet the immediate and dramatic response to steroid therapy and worsening of hearing loss followed on stopping or dose reduction of steroid or Methotrexate therapy can divert the possibility toward the diagnosis of AIED. The response of the patient to first cochlear implant therapy as a treatment and deterioration of the condition afterwards to
the extent that warranted a second Cochlear implant on the other side, makes it another puzzling category specially after improvement with medical treatment with steroid therapy and decrease hearing on reduction of the dosage of prednolone and then improvement with increasing the dose after a remarkable improvement with the cytotoxic therapy (Methotrexate) is another point in favor of the diagnosis AIED.

Mallur et al [24] reported in a case study presented a 12 years old girl with clinical picture, OAE and ABR test results suggestive of AN/AD with fluctuating response to steroid therapy. But with MRI, Gadolinium contrast and stereotactic biopsy revealed the diagnosis Multiple sclerosis, and they suggest that the clinical picture were suggestive of central nervous system pathology. There was no abnormality with MRI with contrast in our patient and so far there was no central nervous system abnormality on clinical ground.

A point of strength in support of auditory neuropathy is the onset of hearing loss with fast reduction of speech discrimination one year following cochlear implant despite no technical electronic fault or program error in the instrument as witnessed by the implant manufacturer check and despite continuous speech therapy. Another point that does not support the diagnosis is the presence of normal hematological, biochemical, immunological and serological investigation. Immunological investigation shows that Anti 68 kd antibodies and all other anti bodies are negative although results are mostly unspecific yet it may put the diagnosis of autoimmune inner ear disease as a slightly far possibility which does not conform with clinical diagnosis possibility of AIED. The diagnosis of Enlarged Vestibular aqueduct Syndrome was excluded with CT scan and MRI study.

**Conclusion**

All performed treatment has been revealed a remarkable improvement of hearing and speech discrimination with no registered serious side effects.

**References**

15. Sininger Y, Oba S. Patients with auditory neuropathy: who are they and what can they hear? Auditory neuropathy a new perspective on hearing disorders 2001; 15 - 36

Correspondence:
Abdulbari Bener
Department of Medical Statistics and Epidemiology
Hamad Medical Corporation and Weill Cornell Medical College
PO Box 3050, Doha, State of Qatar
e-mail: abener(at)hmc.org.qa