CASE REPORT

COCHLEAR IMPLANTATION IN A PATIENT WITH LARGE VESTIBULAR AQUEDUCT SYNDROME: A CASE REPORT

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ABSTRACT

Large vestibular aqueduct syndrome (LVAS) is a distinct clinical entity characterized by fluctuative sensorineural hearing loss associated with isolated enlargement of the vestibular aqueduct. In this report, we present our experience with cochlear implantation in a patient with LVAS.

Keywords: Cochlear implant, large vestibular aqueduct syndrome, enlarged vestibular aqueduct, hearing loss

INTRODUCTION

The association of congenital sensorineural hearing loss with enlarged vestibular aqueducts was initially determined from histopathologic studies of inner ear malformations. The large vestibular aqueduct syndrome (LVAS) was named by Valvassori and Clemis in 1978, when they described radiologic detection of this anomaly. Vestibular aqueduct is defined as “large” if the anteroposterior diameter is larger than 1.5mm. LVAS often associates with profound, nonprogressive sensorineural hearing loss.

Sudden sensorineural hearing loss attacks and fluctuative hearing loss have been reported in some cases. The incidence of sudden hearing loss was found to be rare in other reports. Hearing loss is found to be generally sensorineural but Jackler and De la Cruz found a conductive component in 27% of the ears.

Hearing level may be from normal to profound deafness. Only a few patients were reported to have normal hearing with LVA.

There is no treatment to prevent hearing loss in patients with LVAS. Avoidance of head trauma is important, and hearing aids are symptomatic treatment.

The following case report describes a patient with LVAS who received a cochlear implant. A review of the patient’s audiological assessments before and after the cochlear implantation will be presented.

CASE REPORT

The patient is a 21-year-old female with LVAS. The family suspected hearing loss at the age of 3, she had an audiological evaluation elsewhere and fitted a hearing aid on the left ear.
She was a 9-year-old girl with normal speech development when referred to our clinic. She was the product of a full term pregnancy without any complication. There is no consanguity between the parents but there is a history of hearing loss. Her sister also has hearing loss and benefits from hearing aids.

The patient’s first audiological evaluation demonstrated profound sensorineural hearing loss in the right ear, and moderate to severe mixed hearing loss in the left ear (Fig. 1). Acoustic immitancemetry indicated type A tympanograms bilaterally (Fig. 2). Acoustic reflexes were present only in the left ear contralaterally at the maximum intensity levels at 500, 1000 and 2000 Hz.

Three years later, her hearing in the left ear progressed to severe then to profound, and the hearing thresholds in the right ear improved (Fig. 3). A new hearing aid appropriate to her hearing loss was recommended and scheduled for frequent follow-ups. Two years later, although her hearing
thresholds did not change, significantly, her speech discrimination score in the left ear deteriorated to 12% from 68%. Fluctuations in hearing were determined in both ears in the following assessments.

Fig. 3: Second audiogram obtained three years after the first audiological assessment

Fig. 4: The latest audiogram before cochlear implantation. The patient was evaluated for the cochlear implantation at the age of 15 and found to be a good candidate.

Fig. 4: shows the final audiogram before the implantation.
The otorhinological and neurological evaluations were normal. Routine blood chemistries, urinanalysis and complete metabolic work up were also normal.

A temporal bone high resolution computed tomography (HRCT) scanning demonstrated bilateral LVA. Her sister was assessed with HRCT, because of the LVA diagnosis. She was also diagnosed as LVA.

Cochlear implantation to the right ear was performed without any complication. Full insertion of Nucleus 24M electrode array was achieved. Electrically evoked stapedius reflexes were recorded and electrically evoked compound action potentials were reliably measured using Neural Response Telemetry (NRT) software intraoperatively.

The postoperative performance is encouraging. Her free field hearing thresholds with the cochlear implant were found to be 35 to 45 dB HL from 250 to 8000 Hz (Fig. 5)

Her speech performance after cochlear implantation was evaluated with closed set three syllable words and monosyllabic words and open set three syllable words, monosyllabic words and open set sentences. Table I illustrates the speech perception test results of the patient with cochlear implant. The improvement in speech discrimination was significant. She continues to use her hearing aid on the opposite ear because of better sound lateralization.

Table I: Speech perception test results after implantation.

<table>
<thead>
<tr>
<th>Speech test</th>
<th>6 months</th>
<th>1 year</th>
<th>2 years</th>
</tr>
</thead>
<tbody>
<tr>
<td>Closed set 3 syllable words test</td>
<td>100%</td>
<td>100%</td>
<td>100%</td>
</tr>
<tr>
<td>Closed set monosyllabic words test</td>
<td>82%</td>
<td>98%</td>
<td>98%</td>
</tr>
<tr>
<td>Open set 3 syllabic words</td>
<td>60%</td>
<td>86%</td>
<td>88%</td>
</tr>
<tr>
<td>Open set monosyllabic words</td>
<td>56%</td>
<td>60%</td>
<td>76%</td>
</tr>
<tr>
<td>Open set sentences</td>
<td>72%</td>
<td>76%</td>
<td>80%</td>
</tr>
</tbody>
</table>
**DISCUSSION**

In LVAS, a conductive component has occasionally been reported although hearing loss is predominantly sensorineural \(^8,11,12\). In our case, although the patient has normal (type A) tympanograms on both ears there was an unexplained conductive component in the left ear. According to Valvassori, the conductive component is probably caused by a decreased mobility of the stapes due to increased perilymphatic or endolymphatic pressure \(^11\). The conductive component had disappeared in subsequent audiograms.

The hearing loss usually begins in early childhood, often with an acute onset and fluctuating or progressive in course in relation to head trauma \(^8,5,10\). In our case, the beginning age of hearing loss is similar to most of the LVAS cases in literature; there were fluctuations in the hearing of both ears; but there was no significant head trauma.

Cochlear implantation in patients with LVAS have been reported since 1995 \(^4,13,14\). The audiometric improvement is a common feature in all of them. In this case, sound field warble tone thresholds with the implant were within nearly normal limits and the patient also displayed valuable improvement in speech discrimination.

Patients with LVAS usually have an acquired hearing loss, so they may be accepted as good cochlear implant candidates when they can no longer benefit from hearing aids.

**REFERENCES**