Abstract

We present a case of an 8-year-old boy referred to our department due to speech and learning delays at school. Hearing investigations revealed severe mixed hearing loss on the left with normal hearing on the right. A CT scan was performed which revealed multiple middle ear malformations with Persistent Stapedial Artery (PSA) on the left. Although several therapeutic options had been considered, the patient was implanted with a bone-anchored hearing aid without surgical complications and with a subsequent hearing gain. To our knowledge, this is the first recorded case of bone-anchored hearing aid implantation in a pediatric patient with PSA.

Keywords: hypoacusis, persistent stapedial artery, bone-anchored hearing aid.

Resumo

Apresentamos o caso de um menino de 8 anos de idade referenciado ao serviço de otorrinolaringologia por atraso na linguagem e dificuldades

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de aprendizagem. A investigação audiológica revelou uma surdez mista à esquerda e acuidade auditiva normal à direita. A tomografia computorizada revelou múltiplas malformações do ouvido médio e Persistência da Artéria Estapédica (PSA) à esquerda. Foram deliberadas várias opções terapêuticas, mas optou-se por colocar um implante osteointegrado que proporcionou um ganho auditivo considerável sem registo de complicações. Até à data, este parece ser o primeiro caso descrito na literatura sobre a experiência da utilização de implante osteointegrado em doentes pediátricos com PSA.

P**alavras-Chave:** hipoacusia, persistência da artéria estapédica, implante osteointegrado.

**Introduction**

Persistent Stapedial Artery (PSA) is a rare congenital vascular anomaly with prevalence calculated to be of 0.02% - 0.05% in surgical series. The Stapedial Artery is a primitive vessel that connects the future external carotid artery to the internal carotid artery and normally disappears during the third month of fetal life.

In most times, patients with PSA are asymptomatic, but reports about cases of conductive hypoacusis, sonitus, retrotympanic pulsatile mass and, rarely, sensorineural hearing loss are found in the literature.

In this article, we report a case of a child with PSA and mixed hearing loss that we successfully treated using bone-anchored hearing aid (BAHA) implantation.

**Case report**

An 8-year-old boy was referred to our institution with an history of learning delay and suspicion of hypoacusis for the last three years. The patient denied tinnitus and dizziness. There was no significant prenatal, birth, or postnatal history, or any family history of hearing loss. Physical examination evidenced normal ears and adenotonsillar hypertrophy. Hearing investigations revealed severe mixed hearing loss with an air-bone gap of 45dB on the left (Fig.1) and bilateral type B tympanogram. He underwent adenotonsillectomy with insertion of ventilation tubes at the age of 9 years old but without hearing improvement on the left side. By the age of 10 years old, the patient underwent revision of adenoidectomy with left tympanostomy tube placement. However, mixed hearing loss persisted on the left side. Given the persistence of hearing loss a high resolution CT scan was performed, which revealed left PSA (Fig.2) and facial nerve canal dehiscence. Absence of left round window, stapes and incus lenticular apophysis with left oval window narrowed were also found. The right ear showed no significant alterations.

After a period of three years, during which time the patient’s hearing remained stable, he underwent a test period with a bone-anchored hearing aid softband. Then, the surgery was performed in a single-stage procedure and the patient was fitted with the Cochlear™ BAHA BI300 implant.
Figure 1: Audiogram showing mixed hearing loss with an air-bone gap of 45dB on the left.

Figure 2: High – resolution computed topographic images demonstrating a soft tissue tubular structure with an osseous canal in petrous segment of the temporal bone (A) that enters the middle ear (B) travelling superiorly along the medial wall (partially inside an osseous canal in the hypotympanum). In the epitympanum the structure enters the anterior tympanic segment of the facial nerve (D) and immediately leaves, reaching the middle cranial fossa, where it continues within a sulcus in the squamous part of temporal bone (C). The foramen spinosum is absent (B).
The sound processor was fitted six weeks after surgery. Since that time, the sound processor has been used on a daily basis, without complications and with very good audiological results.

**Discussion**

The PSA is a very rare, congenital, vascular anomaly of the middle ear³.

The embryonic stapedial artery persists because of a failure in its involution⁴. Such artery is transiently present in fetal life and connects the branches of the future external carotid artery to the internal carotid artery⁴. The stapedial artery arises at 4-5 weeks of fetal life from the hyoid artery that is a branch of the internal carotid artery. Then, it extends cranially and passes through the mesenchymal primordium of the stapes, forming the obturator foramen of the stapes⁵, becoming the middle meningeal artery, in the extradural region of the middle cranial fossa⁶. When the stapedial artery fails to involute, it originates the middle meningeal artery, and the spinous foramen that normally includes the middle meningeal artery will be absent⁴.

The majority of patients with PSA reported in the literature have been asymptomatic²,⁵. However, PSA may present as a pulsatile mass in the middle ear cavity, and it is almost always an incidental finding to pulsatile tinnitus or discovered during middle ear surgery⁷. It can also manifest as a conductive hearing loss probably attributed to stapes ankylosis because of pulsations of the PSA⁸. In rare instances, PSA may even erode the otic capsule and result in sensorineural hearing loss⁹. An association with dizziness or vertigo has also been described⁵.

Predicting the disease is not easy and the diagnosis of PSA is usually accidental⁷. However, knowledge about PSA is relevant in order to have this vascular anomaly into account in the management of patients with hearing loss. Imaging identification of this variant may obviate unnecessary surgery and may help in planning surgical or endovascular interventions⁵.

The imaging diagnosis may be made by means of multidetector computed tomography or by angiography. Magnetic resonance imaging as well as magnetic resonance angiography play a limited role in the diagnosis of PSA because of the reduced dimensions of the Stapedial Artery⁵. The diagnosis can be suggested by a soft – tissue prominence in the region of the proximal tympanic segment of the facial nerve, arising of the PSA from the petrous internal carotid artery and by the absence of the foramen spinosum on computed tomography scans, however this last finding is nonspecific⁵,⁶,⁷.

PSA does not require treatment, except in cases of intense tinnitus, where endovascular ligation can be performed⁴. Although no ill effects have been reported from ligation of the PSA, experience is limited.

More than 10 coagulations of PSA have been published since 1990 without any complications⁹,¹⁰. Hitier insists that the PSA can be coagulated¹² but these cases do not guarantee that coagulation of the stapedial artery is safe in all cases⁷.

In theory, disruption of this vessel could lead to ischemia near the geniculate ganglion and medial lemniscus clinically leading to crossed hemiplegia and hearing loss, because of this possibility, some have recommended extreme care when manipulating this artery¹³. Another surgical risk is facial palsy⁷. The facial nerve is partially vascularized by the superior petrous artery which rises from the middle meningeal artery
and can therefore arise from a PSA\textsuperscript{12}. Thus, there is a possibility of causing facial palsy by coagulating the PSA\textsuperscript{7}. Moreover, there is a risk to cause facial palsy by coagulating the PSA because the facial canal is always dehiscent where the artery penetrates\textsuperscript{7}. In the case of an aberrant internal carotid with PSA, we have to take care to avoid bleeding due to carotid artery injury during surgery\textsuperscript{7}.

The clinical case presented concerns a child with absence of left round window, oval window narrowed and PSA which correspond to Teunissen and Cremers class 4, the rarest and the most advanced stage of middle ear malformations. In pediatric patients, these changes in the middle ear limit the conduction of sound to the inner ear and could lead to language and learning delays. Surgical intervention like exploratory tympanotomy could be an option. However, it has a high risk of facial nerve and inner ear injury in patients with complex abnormalities of the structures of the middle ear. Consider that, this option was not performed in the case reported.

Our therapeutic option was BAHA implantation which is a good option for the child who is not a candidate for reconstructive surgery. BAHA is a prosthesis implanted in the temporal bone that allows the transmission of sound waves to the cochlea by direct bone conduction\textsuperscript{14}. In our opinion, it appeared to be the most secure and has provided significant improvement in the hearing level.

At present, no generally accepted guidelines or recommendations are available as to improve hearing in young patients with unilateral hearing loss (UHL). Children with congenital UHL are rarely fitted with hearing aids or are rarely subject to reconstructive ear surgery. However, a published meta-analysis\textsuperscript{15} on the consequences of UHL in children showed a higher risk of delayed language acquisition and school difficulties.

In the case presented, hearing loss was only noticed when the child attended the school and expressed speech and learning delays. It was due to this fact that the child had only been referred by the age of 8 years old. After BAHA implantation, the hearing level showed significant improvement and the rate of grade failures had decreased with no need of additional educational assistance. To our knowledge, our case represents the first report of successful BAHA implantation in a pediatric patient with PSA and severe unilateral mixed hearing impairment.

Children with UHL should be properly considered as medically impaired and to benefit from appropriate steps in evaluation and management at an early age. The ideal treatment modality for each individual may vary depending upon the specific needs of the patient but with any treatment modality, early intervention and appropriate treatment may be the keys to maximum benefit.

**Conflict of Interest:** The authors have no conflict of interest to declare.

**References**
