Magnetic Resonance Imaging in a Neurofibromatosis Type 2 Patient with a Novel MRI-Compatible Auditory Brainstem Implant

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Introduction

Auditory brainstem implantation has become a key technique for the rehabilitation of hearing in patients with neurofibromatosis type 2. The nature of this devastating genetic disease requires ongoing MRI for the patient’s lifespan. Today, most auditory brainstem implants require removal of the magnet that connects the internal device to the external speech processor to undergo imaging as their disease progresses. Patients have the option of having a short procedure to have the magnet taken out and replaced each time, or alternately using a headband to secure the processor over the receiver coil of the internal device. Novel magnet technology has led to the development of a freely rotating magnet that can be used inside the magnetic field of an MRI scanner without losing magnet strength and without being displaced from the body of the device. We report one of the first patients implanted with a Med-El Synchrony ABI in the United States who subsequently underwent successful imaging with MRI 1.5 tesla to follow for other existing schwannomas.

Abstract

Auditory brainstem implantation has become a key technique for the rehabilitation of hearing in patients with neurofibromatosis type 2. The nature of this devastating genetic disease requires ongoing MRI for the patient’s lifespan. Today, most auditory brainstem implants require removal of the magnet that connects the internal device to the external speech processor to undergo imaging as their disease progresses. Patients have the option of having a short procedure to have the magnet taken out and replaced each time, or alternately using a headband to secure the processor over the receiver coil of the internal device. Novel magnet technology has led to the development of a freely rotating magnet that can be used inside the magnetic field of an MRI scanner without losing magnet strength and without being displaced from the body of the device. We report one of the first patients implanted with a Med-El Synchrony ABI in the United States who subsequently underwent successful imaging with MRI 1.5 tesla to follow for other existing schwannomas.

Keywords

► neurofibromatosis type 2
► auditory brainstem implant
► magnetic resonance imaging

Case Report

The patient is a 27-year-old woman who was initially referred for poor balance; subsequent workup led to bilateral vestibular
schwannomas along with other peripheral nerve tumors along
the spinal cord leading to the diagnosis of NF2. Our patient
who underwent observation for some time, however, started
developing hydrocephalus secondary to compression along
the brainstem from increasing left vestibular schwannoma
growth. She underwent left translabyrinthine resection with
sacrifice of the vestibulocochlear nerve complex. Over time she
developed increasing contralateral tumor growth, ultimately
leading to profound hearing loss with 0% speech discrimina-
tion. At that point she elected to undergo left ABI placement
with Med-El Mi11200 Synchrony ABI. While not FDA approved
in the United States, exemption was sought because of its MRI
compatibility up to 1.5 tesla (T), which was necessary for
surveillance of her right vestibular schwannoma, right jugular
foramen schwannoma, and myriad of spinal schwannomas.

She underwent placement of ABI and activation without
difficulty or complications. Subsequent follow-up, she noted
improvement in perception of environmental sounds and
improvement in understanding others; however, she still
struggled with clarity of speech. Twelve months post ABI
placement, she underwent MRI scanning to monitor her
other tumors using multiplanar and multisequence MRI
before and after gadolinium contrast. While metallic artifact
secondary to the ABI limited examination of the left cerebral
and cerebellar hemispheres, MRI with the ABI successfully
and clearly demonstrated large homogenously enhancing
cerebellopontine angle mass filling and expanding the inter-
nal auditory canal measuring 3.7 × 2.9 cm along with mass
effect on the fourth ventricle and upper pons without evi-
dence of obstruction (Fig. 1). Furthermore, MRI with the ABI
in place clearly demonstrated the right jugular foramen and
upper cervical spinal schwannomas without distortion.

Discussion

To our knowledge, this is the first case report of an MRI-
compatible ABI in an NF2 patient in the United States. Studies
have shown that the most devastating impacts of NF2 are
deafness and overcoming communication barriers that lead to
not only strain on social and personal relationships but mood
and self-confidence.3,4 These findings emphasize the impor-
tance of hearing rehabilitation in patients with NF2. However,
hearing rehabilitation goals have to strike a balance with
practitioner’s ability to safely monitor disease progress. Cur-
cently the gold standard for disease surveillance is with MRI.

The only FDA approved ABI currently available in the
United States is the Nucleus 24 ABI, which contains an
internal magnet within the implant receiver. Traditionally
the internal magnet has been a contraindication to MRI
because of the torque introduced to the device by the coil
of the magnetic resonance imager putting the device and
patient at risk.5,6 This often necessitated separate surgical
intervention with magnet removal and replacement, which
puts the device at high risk for damage or infection.7,8

Fig. 1  MRI with a Med-El Synchrony auditory brainstem implant (ABI) demonstrates clear and quality images of the contralateral homogenously
enhancing cerebellopontine mass. The ABI creates moderate metallic artifact distortion that limits evaluation of the ipsilateral cerebral and
cerebellar hemispheres. (A) Axial view sequence from inferior to superior (left to right). (B) Coronal view sequence from anterior to posterior (left
to right).
Recently there has been a push for securement with an external compression device for cochlear implants (CIs) that contain an internal magnet, alleviating the need for separate surgical procedure. Gubbels and McMenomey (2006) examined 16 cadaver heads with Nucleus 24 CI, in which they demonstrated that without proper securement 14 of the 16 had moderate to severe displacement of the magnetic device while undergoing MRI. However, in this same study they found that if properly secured with a compression device, minimal displacement was seen; this eventually led to studies that solidified Nucleus 24 CI as FDA approved for the use of MRI at 1.5 T when properly secured with a compression device. On the other hand, there is no FDA-approved ABI that is MRI compatible in the United States. Unique to the Med-El Synchrony ABI, a compression device is not necessary, thus eliminating any inconvenience or hesitation for emergent MRI. Our patient and device has undergone seven MRIs of the head, C spine, and T spine without any issues or demagnetization to the device while still providing quality images (Fig. 1). The Med-El is MRI compatible because it has a freely rotating and self-aligning diametric magnet, thus preventing torque pressure or demagnetization from the surrounding MRI field. While the ABI is not FDA approved for MRI use, there is some evidence that external securement for MRI up to 1.5 T is safe. Walton et al (2014) most recently examined 10 patients with NF2 who underwent Nucleus ABI placement in the United Kingdom, and they found no altered implant function of demagnetization while undergoing MRI at 1.5 T. However, they did not investigate device displacement, thus questioning the cumulative effect on the device after multiple MRI procedures. Furthermore, while the Synchrony is approved in Europe for up to 1.5 T, the same device design with a freely rotating and self-aligning magnet for its CI model is the only approved CI for up to 3 T. Use of a 3 T scanner would result in a larger metal artifact, potentially decreasing the advantage of this device. Wearing the external device is easier when an internal magnet can be used to position the external device over the receiver coil. While the Synchrony device does not in itself reduce artifact, the mobile internal magnet allows for repeated MRI without magnet removal, improves patient comfort, and decreases the risk of displacement that offers a significant advantage to patient care.

Note
The authors have no funding, financial relationships, or conflicts of interest to disclose.

References
9 Gubbels SP, McMenomey SO. Safety study of the Cochlear Nucleus 24 device with internal magnet in the 1.5 tesla magnetic resonance imaging scanner. Laryngoscope 2006;116(6):865–871
10 Crane BT, Gottschalk B, Kraut M, Aygun N, Niparko JK. Magnetic resonance imaging at 1.5 T after cochlear implantation. Otol Neurotol 2010;31(8):1215–1220