

MRI Imaging in an NF2 Patient with a Novel MRI Compatible Auditory Brain Stem Implant

INTRODUCTION

- Neurofibromatosis Type II (NF2) is the only FDA approved use for auditory brainstem implant (ABI) in the United States.
- NF2 is an autosomal dominant disorder characterized by aberrant growth along Schwann cells of the central nervous system, particularly pathognomonic for bilateral vestibular schwannomas. Secondary to NF2 patients propensity to develop multiple cranial nerve schwannomas, close surveillance in the form of yearly MRI's is necessary.
- The only ABI that is currently FDA approved in the US is Nucleus 24 ABI (Cochlear Corporation). For surveillance a separate surgical procedure for removal of the internal magnet for every MRI or external compression device to try and minimize magnet displacement is necessary[2].
- New ABI devices have been introduced that are MRI compatible, particularly MED EL Mi1200 Synchrony ABI, which allows routine MRI surveillance. The device works through a freely rotating and self-aligning diametric magnet, thus preventing torque pressure or demagnetization from the surrounding magnetic field.

CASE REPORT

The patient is a 27-year-old female who was initially referred for poor balance; subsequent work up led to bilateral vestibular schwannomas along with other peripheral nerve tumors along the spinal cord leading to the diagnosis of NF2. Our patient 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 discrimination. At that point she elected to undergo left ABI placement with MED EL Mi1200 Synchrony ABI. While not FDA approved in the US, exemption was sought because of its MRI compatibility up to 1.5 Tesla, 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 MR imaging 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 homogeneously enhancing cerebellopontine angle mass filling and expanding the internal auditory canal measuring 3.7 x 2.9 cm along with mass effect on the fourth ventricle and upper pons without evidence of obstruction (Figure 1). Furthermore MRI with the ABI in place clearly demonstrated the right jugular foramen and upper cervical spinal schwannomas without distortion.

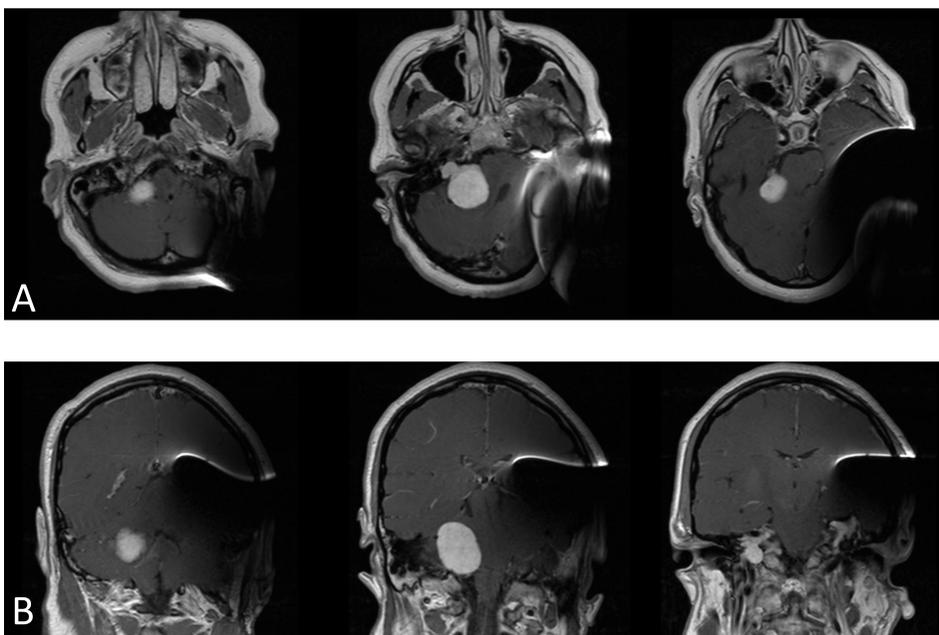


Figure 1. MRI with a MedEl Synchrony auditory brainstem implant (ABI) demonstrates clear and quality images of the contralateral homogeneously enhancing cerebellopontine mass. The ABI creates moderate metallic artifact distortion which limits evaluation of the ipsilateral cerebral and cerebellar hemispheres. A) Axial view sequence from inferior to superior (left to right). B) Coronal view sequence from anterior to posterior (left to right).

DISCUSSION

To our knowledge, this is the first case report of a MRI compatible auditory brainstem implant (ABI) in a NF2 patient in the United States. Studies have shown the most devastating impact of NF2 is deafness and overcoming communication barriers that leads to not only strain on social and personal relationships but mood and self-confidence [3, 4]. These findings emphasize the importance 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. Currently the gold standard for disease surveillance is with MRI.

The only FDA approved ABI currently available in the US is the Nucleus 24 ABI (Cochlear Corporation), 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]. Recently there has been a push for securement with an external compression device for cochlear implants that contain an internal magnet, alleviating the need for separate surgical procedure. Gubbels et al (2006) examined 16 cadaver heads with Nucleus 24 cochlear implant, in which they demonstrated without proper securement 14 of the 16 had moderate to severe displacement of the magnetic device while undergoing MRI[9]. However in this same study they found that if properly secured with a compression device minimal displacement was seen [9]; this eventually led to studies that solidified Nucleus 24 Cochlear implant as FDA approved for the use of MRI at 1.5Tesla when properly secured with a compression device [10, 11]. On the other hand, there exists no FDA approved ABI that is MRI compatible in the US.

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 MRI's of the head, C and T spine without any issues or demagnetization to the device while still providing quality images (Figure 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 1.5T is safe. Walton *et al* (2014) most recently examined 10 patients with NF2 who underwent Nucleus ABI placement in the UK, and they found no altered implant function of demagnetization while undergoing MRI at 1.5T[12]. 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 1.5T, the same device design with a freely rotating and self-aligning magnet for its cochlear implant (CI) model is the only approved CI for up to 3T[13]. Use of a 3T 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 which offers a significant advantage to patient care.

CONCLUSION

- The MED-EL is MRI compatible ABI because it has a freely rotating and self-aligning diametric magnet, thus preventing torque pressure or demagnetization from the surrounding MRI field.
- While not approved in the US, the MED-EL ABI provides significant advantage for patient care because of its ability to undergo multiple surveillance MRI's without magnet removal, minimal patient discomfort, and minimal risk of displacement or demagnetization.
- Our patient has undergone seven MRI's to date without any issues or problems. She continues to receive a tremendous benefit from the device.

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