Baird Samantha (Orcid ID: 0000-0002-5274-8761)

Article Title: Inner ear and retrocochlear pathology on magnetic resonance imaging for sudden and progressive asymmetrical sensorineural hearing loss

Running Head: Inner ear MRI abnormalities in sensorineural hearing loss

Samantha M. Baird MBBS(Hons) GDipSurgAnat[1],

Kevin Nguyen MBBS BMedSc, GDipSurgAnat FRACS[1]

Daman D. S. Bhatia BMed MS[1]

Benjamin P. C. Wei MBBS FRACS PhD[1,2]

[1]Department of Otology, The Royal Victorian Eye and Ear Hospital, Melbourne

[2]Department of Otolaryngology, University of Melbourne

Acknowledgements – Nil **Financial Disclosures –** Nil **Conflict of Interest** – Nil

Abstract word count: 245 words

Manuscript Word Count (including references): 2583 words

Number of Tables: 1 **Number of Figures**: 5

Corresponding Author: Dr Samantha M. Baird

dr.sm.baird@gmail.com

Address: Department of Otolaryngology

32 Gisborne Street

East Melbourne, VIC, 3002

Australia

Tel: +61 03 9929 8666

Abstract

Background: In sudden and asymmetrical progressive sensorineural hearing loss (SNHL), magnetic resonance imaging (MRI) is required to evaluate for retrocochlear pathology and, with recent advances in MRI techniques, inner ear pathology. Given the limited literature regarding inner ear pathology associated with SNHL, we aimed to assess the incidence of retrocochlear and inner ear pathology, and congenital malformation on MRI in sudden and progressive SNHL.

Methods: 987 acoustic neuroma (AN) protocol MRI internal acoustic meatus (IAM) studies performed at our institution to investigate SNHL between January 2013 to December 2015 inclusive were identified. Following categorisation for indication of sudden versus progressive asymmetrical SNHL, MRIs with retrocochlear or inner ear abnormality, congenital malformation or other otology-related abnormality were identified, and further data was collected for these patients including patient demographics, associated symptomatology, management and outcomes.

Results: In sudden SNHL (SSNHL), aetiological abnormality on MRI was identified for 6.9% patients with AN present on 4% overall. 3.2% of MRIs for progressive asymmetrical SNHL identified a causative lesion with 2.3% of scans overall diagnosing AN. The incidence of congenital inner ear malformation on MRI in the setting of SSNHL and progressive asymmetrical SNHL are 1.7% and 0.6% respectively.

Conclusion: This is the first retrospective study of inner ear MRI abnormalities in both sudden and progressive asymmetrical SNHL in Australia and one of the largest cohorts published in the literature to

date. MRI must be performed in the setting of SNHL to ensure aforementioned and rarer causative lesions are identified.

INTRODUCTION

In the presence of sudden or asymmetrical progressive sensorineural hearing loss (SNHL), magnetic resonance imaging (MRI) is widely accepted as standard of care and is a cost-effective investigation in the evaluation of retrocochlear pathology. However, with recent advances in MRI techniques there has been an increase in inner ear findings in many of these patients, hence increasing its diagnostic role[1].

Sudden sensorineural hearing loss (SSNHL) is defined as >30 decibels SNHL in at least three contiguous frequencies occurring over 72 hours or less[2]**.** Despite continuous efforts to clarify the pathophysiologic characteristics, the cause remains largely unclear with approximately 90% of cases idiopathic[2]. However, SSNHL can also be a consequence of various conditions including viral infection or labyrinthritis, tumour of the internal auditory canal (IAC) or cerebellopontine angle (CPA), infarction, multiple sclerosis, rupture of an inner ear membrane, or rarely congenital malformations[2,3]. As such imaging of SSNHL is required to rule out CPA masses as well as to detect inner ear abnormalities.

Asymmetrical SNHL is commonly defined as the presence of a difference of 15 or more decibels at two or more frequencies, or a difference of 15% or greater in word recognition scores[4]. Imaging for asymmetrical progressive SNHL is performed with the same techniques and for the same reasons as described for SSNHL.

In this retrospective study, we identified inner ear and retrocochlear abnormalities on MRI for patients investigated for SSNHL and asymmetrical progressive SNHL at The Royal Victorian Eye and Ear Hospital (RVEEH) in Melbourne, Australia from January 2013 to December 2015. We aimed to assess the incidence of retrocochlear and inner ear pathology, and congenital malformation on MRI in sudden and progressive SNHL, and to compare these two groups.

METHODS AND MATERIALS

Ethical approval to conduct the study was granted by RVEEH Human Research Ethics Committee (Reference number: 15/1242HS). Since all patients at our institution undergo outpatient MRI scans at St Vincent's Hospital Melbourne radiology department, their radiology database was used to identify study participants. All patients who underwent an acoustic neuroma (AN) protocol MRI internal acoustic meatus (IAM) study requested by RVEEH for investigation of SNHL between January 2013 to December 2015 inclusive were identified. On clinical record review, duplicate MRIs for an individual patient, follow up MRIs for previously identified AN, and MRIs performed for other indications were excluded.

MRI was conducted using a 1.5 or 3.0-Tesla superconducting magnet system (Siemens Skyra 3.0T, Siemens Avanto 1.5T) with a phased-array head coil. The scanning encompassed the region from the mastoid to the upper edge of the petrous bone. The protocol consists of a dual echo sequence of the whole head, acting as a localiser, which is followed by T2 weighted high resolution CISS 0.85mm slice thickness with 1mm thick reconstructions in axial images. If pathology is identified, contrast is administered and T1 weighted imaging is performed with fat saturation.

Patients were categorised into groups who clinically presented with SSNHL versus progressive asymmetrical SNHL. MRIs that showed a retrocochlear or inner ear abnormality, congenital malformation or other otology-related abnormality were identified. Further data was then collected for these patients including patient demographics, associated symptomatology at presentation, management and outcomes.

RESULTS

A total of 987 MRI scans were identified for our 3-year study period. The mean age was 55 years (range 9 to 91 years) and there was a near even gender split with 484 (49%) females and 503 (51%) males. 174 (17.6%) MRI scans were ordered for investigation of SSNHL compared with 813 (82.4%) for progressive asymmetrical SNHL. All identified abnormalities are summarised in Table 1.

Twelve of the 174 patients with SSNHL (6.9%) had abnormalities on MRI. The most common finding was an AN, newly diagnosed in seven (4.0%) patients. Amongst this subgroup, the mean age was 51 years (range 29 to 71 years) and five (71.4%) patients were male. Six (85.7%) patients presented with right-sided SSNHL with only two (28.6%) patients presenting with ipsilateral tinnitus. All tumours were small (<1cm) to medium-sized (1.1-2.9cm) with the largest tumour noted at 12x14x11mm. Six of the ANs (85.7%) were

lateral, arising from or extending into the IAM. Interestingly, in one case, the diagnosed AN was on the contralateral side to the SSNHL. All patients received a high-dose weaning course of oral prednisolone with two (28.6%) obtaining complete resolution of their SNHL. Three patients were referred for neurosurgical opinion regarding surgical excision, two for radiotherapy and the remaining two were observed.

A congenital malformation was identified for three (1.7%) patients, which was bilateral enlarged vestibular aqueducts (EVAS) in all cases (Figure 1). All three patients were female, aged 23, 36 and 75 years. Two of the three patients also experienced vertigo and all had severe to profound SSNHL on audiogram. Both the 23- and 75-year-old patients subsequently underwent cochlear implantation; the third is being monitored.

Interestingly, a 60-year-old male who presented with left SSNHL was found to have lacunar infarcts within the pons, left anterior medulla, left temporal stem and bilateral thalami. Also, a 51-year-old male who presented with right mild to moderate SSNHL had a 1.4cm right petrous apex mass eroding into the superior and posterior semi-circular canals on MRI. This was diagnosed as Langerhans' Cell Histiocytosis on histopathology following surgical excisional biopsy.

Of the 813 MRIs performed for progressive asymmetrical SNHL, 99 (12.2%) were performed as part of our cochlear implantation assessment protocol, and 26 (3.2%) patients had abnormalities on MRI. In contrast to the SSNHL subgroup, there was a lower incidence of congenital abnormalities amongst patients investigated for progressive SNHL. Five (0.6%) patients had congenital abnormalities evident on MRI. Two male patients, a 37- and 55-year-old had EVAS with Mondini malformation, one of which was in the setting of known Pendred syndrome. Two others, a 25-year-old male and a 67-year-old female displayed Mondini

malformation alone and one patient, a 69-year-old male had congenital right cochlear hypoplasia with only a small remnant of the basal turn present (Figure 2). All five patients received cochlear implantation.

19 (2.3%) MRIs newly diagnosed an AN, three (15.8%) of which were intracochlear (Figure 3). Amongst this subgroup the mean age was 61 years (range 40 to 87 years) and nine (57.9%) patients were female. Six (31.6%) of the diagnosed ANs were referred for operative management or radiotherapy. All three intracochlear ANs were managed conservatively. One MRI diagnosed a cerebellopontine angle epidermoid cyst in a 32-year-old female that required surgical excision (Figure 4). Another MRI for an 80-year-old male revealed a 7x5x6mm meningioma above the right IAC extending to the superior aspect of the porous acoustic with contact of the superior vestibular nerve.

DISCUSSION

To our knowledge, this is the first retrospective study of inner ear MRI abnormalities in both sudden and progressive asymmetrical SNHL in Australia and the largest cohort published in the literature to date. Our study demonstrated that 6.9% (12 of 174) patients with SSNHL showed abnormalities on MRI IAM, which is consistent with previous studies which report an incidence of 4.4% to 13.75%[3,5-9]. This contrasts with progressive SNHL, in which MRI IAM abnormalities were detected in 3.2% (26 of 813 patients).

AN, or vestibular schwannoma, is the most commonly encountered mass in the IAC or CPA, accounting for approximately 85% CPA masses[10]. Historically ANs are known to present with varying degrees of progressive sensorineural hearing loss, dizziness or vertigo and tinnitus. Our study showed that 2.3% MRIs performed for progressive asymmetrical SNHL newly diagnosed an AN. However, SSNHL has also

been recognised as a presenting symptom of AN in around 10-20% of cases[11-14]. Therefore, it was not unexpected that AN was the most common abnormality detected on MRI in our SSNHL population with an incidence of 4%.

Whilst it is believed that tumour size does not correlate with severity of hearing loss[15], an isolated study has reported that SSNHL more frequently occurs with small tumours (<1cm) as opposed to medium (1.1-2.9cm) and large (>3cm) tumours[12]. Our data were consistent with this as all seven ANs diagnosed in the setting of SSNHL in our study were of small to medium size. Lateral ANs arising from or extending into the IAM are more frequently associated with SSNHL[9,13,14,16] and this is thought secondary to cochlear nerve compression and elevated IAM pressure induced by the tumour. This was also reflected in our data with 85.7% (6 of 7) ANs situated laterally.

Primary inner ear schwannoma (PIES) is a very rare variant of AN with around 260 cases reported to date[17]. It is associated with a 99% rate of sensorineural hearing loss[17], of which 61% is progressive, 39% is sudden and the remaining 7% usually fluctuating[18]. In contrast to CPA ANs, these tumours show intense, focal homogenously enhancing masses on postgadolinium MRI[1,19]. In our population, 15.8% (three out of seven) of the ANs diagnosed in our progressive SNHL cohort were intracochlear, which is the most common type of PIES usually accounting for around 31-51% of cases[17,19]. There appears to be a higher than reported incidence of PIES in our cohort compared to other studies. This may be due to the size of our cohort and the role of RVEEH as a tertiary referral centre for SNHL. Improvements in MRI technology and protocols with regard to MRI IAM as well as maintaining a high index of suspicion have also contributed to increased detection of this pathology[17-19]. Other CPA masses causing SNHL include meningioma (3% to 8% cases) and

epidermoid cyst, both of which were represented in our study population with one of each identified within the progressive SNHL cohort.

In our data the incidence of congenital inner ear malformation identified on MRI in the setting of SSNHL and progressive asymmetrical SNHL were 1.7% and 0.6% respectively. EVAS is the most common congenital inner ear abnormality detected on imaging[20]. Among the congenital malformations noted in our study, EVAS was present in all cases in the SSNHL cohort (3 of 3) and 80% of those in the progressive SNHL cohort (4 of 5). 50% of EVAS cases in the progressive SNHL population also displayed Mondini malformation, one of which was in the setting of known Pendred syndrome, which is associated with these abnormalities. To our knowledge this is the first study in the literature to date which reports on the incidence of congenital abnormalities identified on MRI IAM in the investigation of SSNHL and progressive asymmetrical SNHL. Cochlear hypoplasia and aplasia are rarer congenital inner ear malformations which have important implications for cochlear implantation. This condition was represented in our data whereby one patient with progressive asymmetrical SNHL (representing 0.1% of the total cohort) showed cochlear hypoplasia on MRI. The patient subsequently underwent a successful cochlear implantation.

Intra-axial lesions can also cause SSNHL, hence MRIs investigating SSNHL should include central auditory pathways. In rare cases, sudden hearing loss may be the presenting symptom of stroke, and this phenomenon has been previously linked to lacunar[3] and anterior inferior cerebellar artery[21] infarction. This is concordant with our data whereby one patient with SSNHL revealed lacunar infarction on MRI.

Strengths of the study include a large cohort size, unique reporting on the incidence of congenital malformations in SNHL, and the demonstration of differences in MRI findings between unilateral sudden

and progressive SNHL. The role of RVEEH as the sole otolaryngology specialist hospital in Australia and a tertiary referral centre for SNHL across Victoria represents both strengths and limitations. The volume of referrals facilitated a large cohort whilst reflecting an accurate demographic representation of the Victorian population. However, our institution's position as a tertiary centre may render certain aetiologies underrepresented in our cohort. For example, we did not report any cases of trauma or inner ear haemorrhage as a cause for SNHL. These aetiologies may have been more prominent in a trauma centre or general emergency department setting, where patients with severe labyrinthine symptoms consistent with inner ear haemorrhage may be more likely to present. We also acknowledge that a small number of patients undergo MRI at external institutions and hence these patients would not be captured in the study. Lastly, the reliance on clinical documentation, particularly in relation to presenting symptomatology is another limitation of the study.

CONCLUSIONS

We present the first retrospective study of inner ear MRI abnormalities in both sudden and progressive asymmetrical SNHL in Australia and the largest cohort published in the literature to date. Our findings demonstrate that 6.9% patients with SSNHL and 3.2% patients with progressive asymmetrical SNHL had an aetiological abnormality on MRI. AN was diagnosed on 4% MRIs performed for SSNHL and 2.3% for progressive asymmetrical SNHL. The incidence of congenital inner ear malformation on MRI in the setting of SSNHL and progressive asymmetrical SNHL were 1.7% and 0.6% respectively, with EVAS the most common finding. The significant proportion of abnormalities identified on MRI in this study reinforces the necessity for MRI in the investigation of sudden and asymmetrical progressive SNHL.

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Table 1: Summary of Abnormal MRI Findings in Patients with SSNHL and Progressive Asymmetrical SNHL

Abnormality	**SSNHL cohort (total *n* = 174)**	**Progressive SNHL cohort (total *n*= 813)**
Vestibular schwannoma (n (%))	7 (4.0)	19 (2.34)

Primary inner ear schwannoma (n (%))	–	3 (0.37)
Congenital malformation	3 (1.7)	5 (0.62)
Enlarged Vestibular Aqueducts (EVAS) (n (%))	3 (1.7)	2 (0.25)
EVAS with Mondini malformation (n (%))	–	2 (0.25)
Cochlear hypoplasia	–	1 (0.12)
Other		
Petrous apex lesion (n (%))	1 (0.6)	–
Epidermoid cyst (n (%))	–	1 (0.12)
Meningioma (n (%))	–	1 (0.12)
Lacunar infarction (n (%))	1 (0.6)	–
Total Abnormalities (n (%))	**12 (6.9)**	**26 (3.2)**

Figures and Legends:

Figure 1: Axial T2 weighted MRI showing bilateral enlarged vestibular aqueducts (white arrows)

Figure 2: Axial T2 weighted MRI showing non-visualisation of the right cochlear apart from tiny remnant of the basal turn (arrow).

Figure 3: Axial post contrast T1 weighted MRI with fat saturation showing enhancing soft tissue density within the cochlear consistent with an intracochlear primary inner ear schwannoma (arrow).

Figure 4: Axial T2 weighting MRI revealing an intermediate signal right cerebellopontine angle mass that appears inseparable from the 7^{th} and 8^{th} cranial nerves.

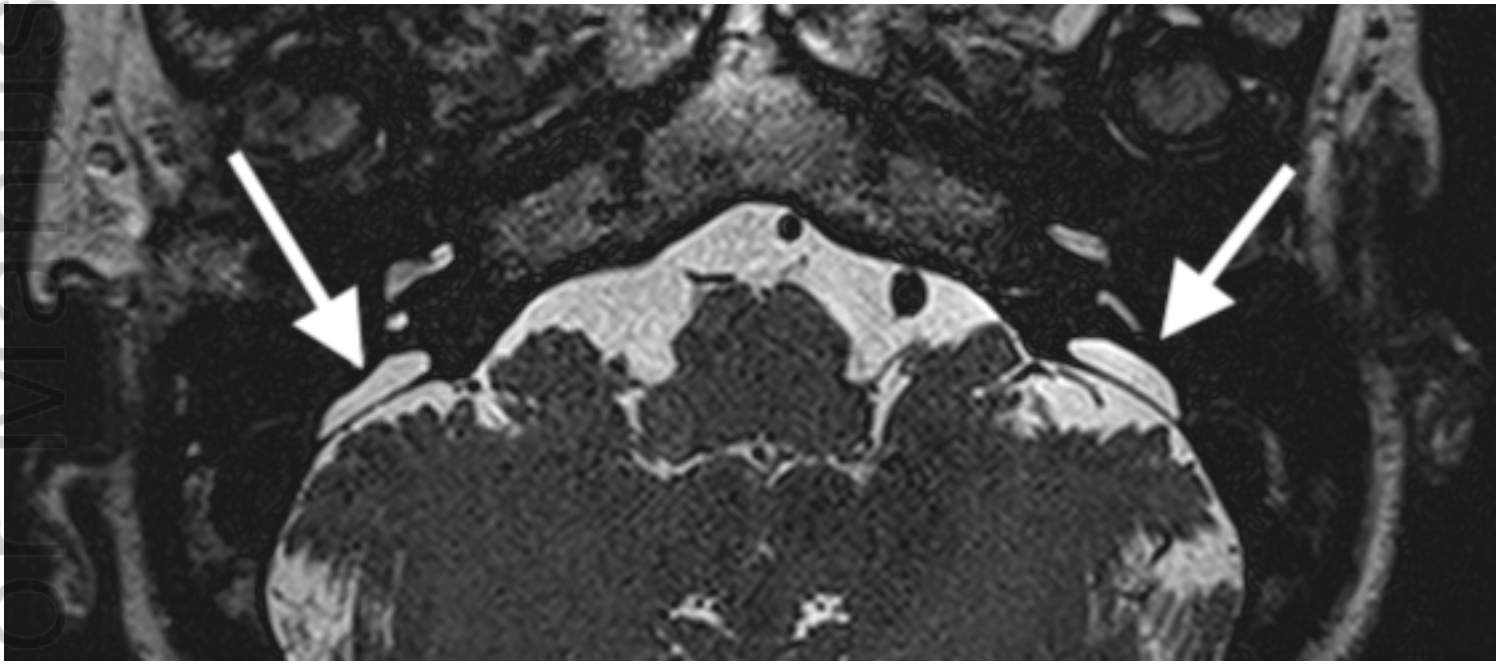

Figure 1 EVAS annotated.tiff

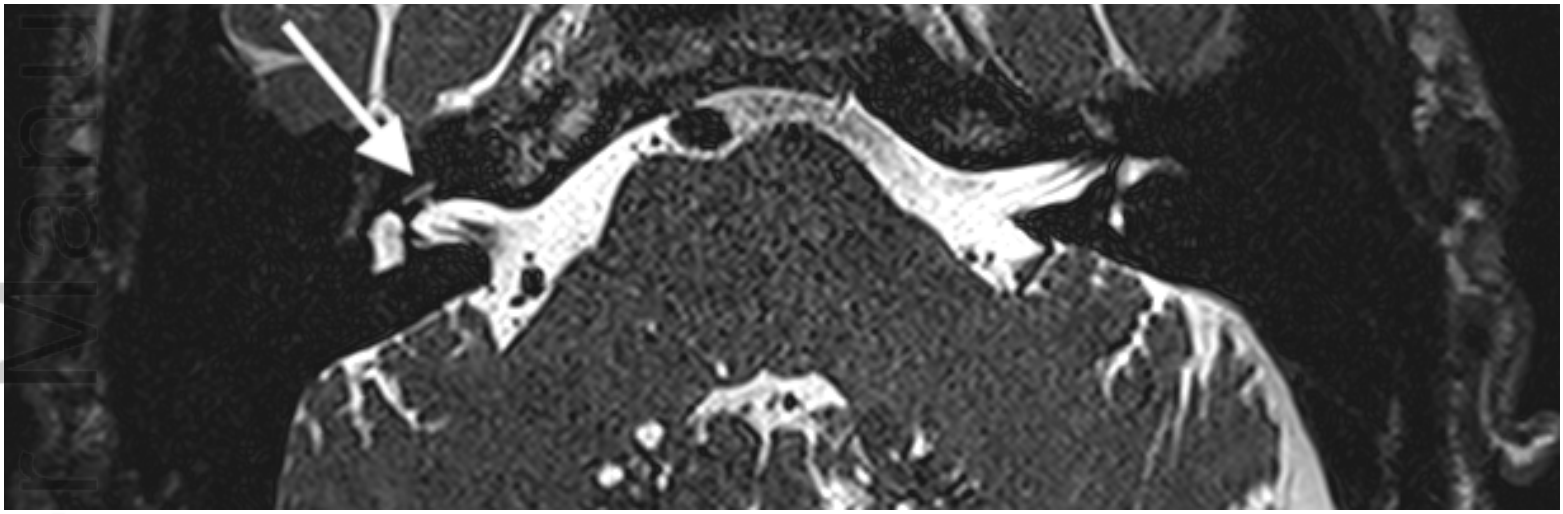

Figure 2 Remnant cochlea annotated.tiff

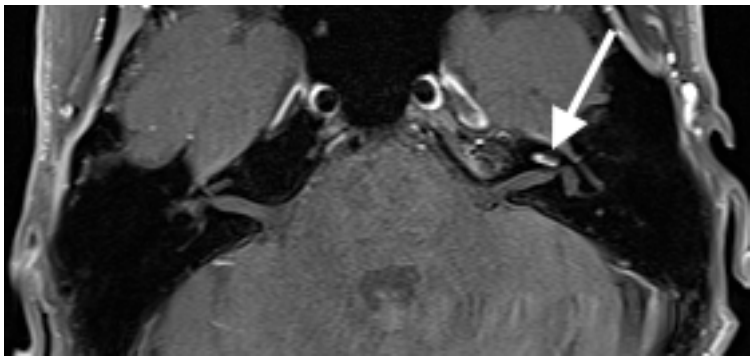

Figure 3 PIES annotated.tiff

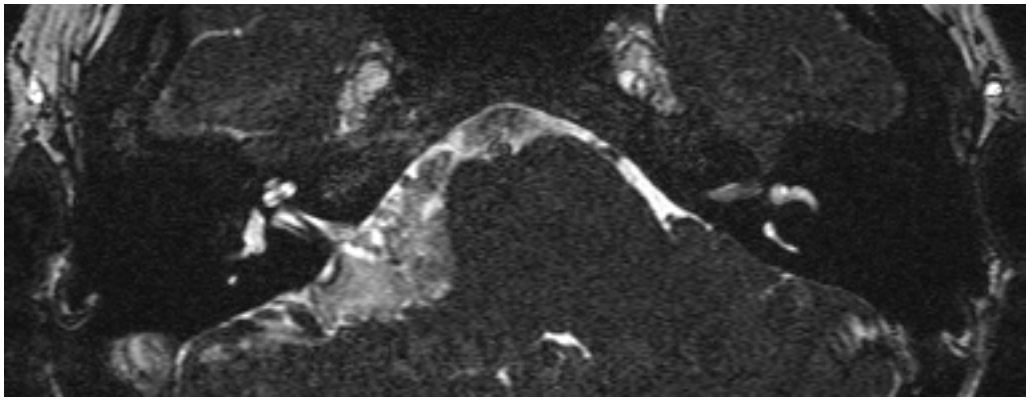

Figure 4 CPA mass.tif

Author/s:
Baird, SM; Nguyen, K; Bhatia, DDS; Wei, BPC

Title:
Inner ear and retrocochlear pathology on magnetic resonance imaging for sudden and progressive asymmetrical sensorineural hearing loss

Date:
2019-06-01

Citation:
Baird, S. M., Nguyen, K., Bhatia, D. D. S. & Wei, B. P. C. (2019). Inner ear and retrocochlear pathology on magnetic resonance imaging for sudden and progressive asymmetrical sensorineural hearing loss. ANZ JOURNAL OF SURGERY, 89 (6), pp.738-742. https://doi.org/10.1111/ans.14957.