IMJ Official Journal of the Irish Medical Organisation

Ir Med J; March 2025; Vol 118; No. 3; P41
March 20th, 2025

Intra-temporal and intracranial radiological abnormalities in cochlear implantation: Radiologic findings and audiological outcomes

E. Farrell[1,2], H. Jones[1,2], C. Simoes-Franklin[1,2], L. Viani[1,2], F. Glynn[1,2], P. Walshe[1,2]

1. National Hearing Implant & Viani Research Centre, Department of Otology and Cochlear Implantation, Beaumont University Hospital, Dublin, Ireland.
2. Royal College of Surgeons in Ireland, St. Stephen's Green, Dublin 2, Ireland.

Abstract

Aim

Radiological investigations are an important part of the work up for cochlear implantation candidacy. Inner ear anomalies and intracranial anomalies are frequently found in the preoperative investigation phase. This study aims to investigate the frequency of such abnormalities, the complication rates and the outcomes associated with such findings in adult and paediatric cohorts at a national tertiary referral centre.

Methods

A retrospective review of a prospectively maintained database at a national referral centre for cochlear implantation was undertaken. We included all patients from 2013 to 2020 who had radiological abnormalities identified on review by consultant radiologists and who proceeded to cochlear implantation. Audiological outcomes were measured using BKB scores in the adult group and LiP, SIR and CAP scores for the paediatric group. Rates and types of complications were also recorded.

Results

There were 898 cases reviewed at the MDT during the time frame. There were 90 (10%) radiological abnormalities noted, 57 (63%) paediatric and 33 (37%) adult. The most common type of abnormality in adult and paediatric groups was incomplete partition type 2. White matter changes associated with CMV was the most common paediatric brain abnormality and old infarction was the most common in the adult group. CSF gusher was encountered in 2/57 (3.5%) cases. Mean BKB results in adults was 68% for temporal and 46.6% for brain abnormalities. Mean LiP, SIR, CAP scores in the temporal bone (38.2, 4.6, 5.9) and brain abnormality (35.7, 3, 4.7) groups were acceptable.

Discussion

IMJ
Official Journal of the Irish Medical Organisation

Ir Med J; March 2025; Vol 118; No. 3; P41
March 20th, 2025

Radiological abnormalities are detected in a significant number of patients with profound hearing loss who are undergoing candidacy work up for cochlear implantation. Results may be variable particularly in children with CMV related changes detectable on MRI. Complication rates are low and should not preclude these patients from undergoing implantation.

Introduction

The criteria for patients who benefit from cochlear implantation has increased since the introduction of implantable hearing devices first became available to those with profound sensory neural hearing loss. While initially cochlear implantation was only recommended in patients with normal middle and inner ear anatomy, implantation candidacy has expanded in recent years. Furthermore, the outcomes of patients with these abnormalities have been reported as comparable to patients who do not demonstrate such abnormalities, albeit inconsistently and dependent on individual study methodology.

The benefits of cochlear implantation in the paediatric cohort are most beneficial during early years of life when speech and language development are emerging. Complicating this important phase of development in children with hearing loss is the fact that paediatric sensory neural hearing loss is thought to be associated with anatomical abnormalities in as many as 20%-40% of cases[1,2]. The classification of inner ear abnormalities is also evolving as shown by Sennaroglu's analysis of incomplete partitions of the cochlea[3]. In tandem with radiological examinations producing increasingly higher definition images, standardised reporting and measuring methods[4], it is probable that the number of abnormalities detected and operated on will increase with time.

Cochlear implantation is not unique to the paediatric population. The benefits of implanting adults with acquired hearing loss an audiological and cognitive outcomes perspective are only beginning to be understood[5]. With such promising new indications for implantation emerging it is likely there will be a corresponding rise in the number of patients with brain and temporal bone abnormalities that receive cochlear implantation as they are detected or acquired later in life. The outcomes from such cases are still a topic of debate and the merits of implantation are not yet part of official guidance.

The complication rates for cochlear implant are low but much of this data is based on operations performed on patients with normal anatomy[6]. The complication rates for procedures carried out on patients with abnormal anatomy is therefore not determined. While the limits of implantable cases are constantly being tested, the implantable community

and body of literature will benefit from publication of outcomes of implanted abnormal cochleae.

The aim of this study was to examine the radiological abnormalities encountered in the only national tertiary referral centre for cochlear implantation in the Republic of Ireland across adult and paediatric patient cohorts, to measure outcomes in terms of speech and audiological outcomes and finally, to measure the frequency and long term sequelae of surgical complications.

Methods

A retrospective review of a prospectively maintained database of all patients implanted at a national tertiary referral centre in the Republic of Ireland between 2016 and 2020 was reviewed. The year 2020 was chosen as a cut off to allow sufficient time for follow up assessments to be conducted and recorded in the data base.

Permission to undertake this work was sought and granted by the hospital board.

Radiological abnormalities were recorded following identification by consultant radiologists with specialist training in neuroradiology. All scans and patient cases were discussed at a multi-disciplinary team meeting allowing for further scrutiny of each case in the presence of the operating surgeons. Abnormalities were documented on a consensus multi disciplinary team proforma following discussion. Candidates deemed appropriate for implantation underwent investigations and supportive counselling prior to surgery in keeping with best practice guidelines. All cases of radiological abnormalities that went on to have cochlear implantation were recorded. Abnormalities were classified as either primarily temporal bone or brain abnormalities. Subsequent classifications of subsite of abnormalities were also recorded.

Sub types of incomplete partition were described congruent with the conventional description outlined by Sennaroglu, where type 1 demonstrates a cystic appearance in conjunction with an absent modiolus and interscalar septae and type 2 represented a cystic apex and a normal basal turn[3].

All procedures were conducted by consultant surgeons or higher specialist trainees under the direct supervision of a consultant. Operative notes and demographics were retrieved from charts. Information on intra operative difficulties or post-operative complications were recorded from clinic notes and records.

Outcomes were recorded where available and included BKB scores for adult patients and scores from a variety of tests including Listening Progress Profile (LiPP), Categories of Auditory Perception (CAP) and Speech Intelligibility Rating (SIR) for paediatric patients.

Results

There were 898 patients included in the database. All of these patients underwent MDT discussion prior to implantation. There were 90 abnormalities detected during the course of the work up and MDT discussion.

Of the 90 radiological abnormalities that were detected, 33 were detected in the adult group and 57 were found in the paediatric group. The average age in the adult group was 55.3 years of age, the median was 57.5 and the range was 19-83 years. The mean age of bilateral cochlear implantation was 3.1 years, the average was increased owing to the implantation of some patients in their teenage years. The median was 1.5 years. The mean age of implantation in the unilateral group was 5.4 years and median was 5 years. (Table 1)

The primary abnormality in the adult group was localised to the temporal bone in 18 cases while 14 were localised to the brain and 1 case demonstrated both brain and temporal bone abnormalities. (Table 2)

The most commonly occurring abnormalities within the temporal bone for the adult cohort were incomplete partition type 2 followed by an enlarged endolymphatic sac. There were 14 brain abnormalities detected in the adult group, the most common of which were old infarctions followed by atrophy. (Table 3)

The most common abnormality in the paediatric group was incomplete partition type 2. There were 23 cases of this type of defect identified. This was followed by an enlarged vestibular aqueduct and incomplete partition type 1. In the group of paediatric patients with abnormal brain scans, 4 were due to white matter changes consistent with CMV and one patient was identified as having abnormal gyri. There were 4 cases in the paediatric cohort that had both brain and temporal bone abnormalities. (Table 4)

Neural response telemetry for 22 electrodes as recorded at the end of an operation was assessed. Bamford, Kowal and Bench (BKB) scores were used to measure adult hearing outcomes. The total number available for analysis was 16 out of the cohort of 18. The mean BKB score was 68.1% and ranged from 0 to 96%. There were 11/14 data points available in the adult patients with brain abnormalities group. They had an average BKB score at 1 year follow up of 46.6% and a range of 0-96%. (Table 5) Spearman's rank correlation coefficient

did not reveal a correlation between NRT at the time of insertion and BKB scores at 1 year follow up (r_s=0.35671).

Outcomes were available for 45% of the paediatric cohort with radiologically abnormal temporal bones. Audiological outcomes for paediatric patients with both temporal bone and brain anomalies are recorded as Listening Progress Profile (LIP), Categories of Auditory Performance (CAP) and Speech Intelligibility Rating (SIR) scores. As before NRT for 22 electrodes as recorded at the end of an operation was also assessed. (Table 6)

The most common surgical approach was the cortical mastoidectomy with posterior tympanotomy and round window approach (n=66). Deviations from this approach were all within the paediatric group and were adopted due to anatomical topography which included; cochleostomy (n=1) which was performed in a case of CHARGE syndrome and hypoplastic mastoid cavities; combined approach (n=1) was performed in a case of hypoplastic cochlea and facial recess approach (n=1) performed in a case of incomplete partition type 2. Data were not available for 21 cases.

Complication rates for the paediatric cohort was low with 7 documented complications out of a total of 57 cases. None of these complications had adverse effects on the child's overall health and were dealt with at the time of surgery. Three gushers were encountered, two procedures were abandoned, one implant was incompletely inserted and one tegmen breach. One documented complication was encountered in the adult group. This was due to hardware failure and required explant and reimplant of the device.

Discussion

Previously published data on cochlear implantation in anatomical abnormalities have shown that outcomes are comparable between anatomically normal and abnormal patients[7,8]. As up to 20%-40% of paediatric sensory neural hearing loss can be associated with a structural abnormality[9] of the inner ear, it is not surprising that the balance of abnormalities in the paediatric group was in favour of temporal bone anomalies. In this dataset the most common temporal bone abnormality in the paediatric group was incomplete partition type 2 which is in keeping with other published reports[10]. Enlarged vestibular aqueduct (EVA) was the next most common abnormality encountered and occurred bilaterally where it was identified. Although these EVAs were diagnosed in isolation, they are commonly found with cochlear anomalies that may not be detectable radiologically, including interscalar abnormalities[11]. In that regard, an isolated EVA could in the future be interpreted as probable abnormal cochlea, however we have insufficient data to definitively draw this conclusion. There is some evidence to suggest that cochlear nerve deficiencies may be a hallmark for other nerve

abnormalities including vestibular, facial and/or olfactory nerves[12], however this was not found in the present study.

The abnormalities detected intra cranially in the paediatric group were primarily due to white matter changes in the cortex associated with CMV infection. The average age at implantation in the intracranial abnormalities group was 2.6 years which was similar to the non CMV related group. The speech outcomes and audiological outcomes for this cohort of patients was also similar to the general abnormalities group which would indicate that they benefited equally as well from implantation. The outcomes for patients following cochlear implantation for CMV related deafness have been investigated by other authors and yielded varying results. One such study compared 16 CMV related cases to 131 congenital hearing loss cases and found 50% of children implanted scored better or the same while 50% of the cohort of 16 scored less than pre implantation[13]. Similar data which compared CMV related hearing loss to a cohort of Connexin 26 mutation patients found that those CMV related patients with MRI changes lagged behind those without such changes in terms of speech production and developmental milestones but may have similar results for speech perception[14]. These findings were echoed in a later study which compared 25 cases of congenital CMV deafness to 23 cases of non CMV related congenital deafness and found adequate function in the CMV group despite lagging behind their non CMV comparators[15]. The neurological and cognitive effects of CMV infection is often not accounted for in these studies and may provide an explanation for the wide variety of audiological outcomes. There were no complications encountered in these studies and no complications recorded in this cohort. While sample sizes in these studies are quite small, the tide of evidence leans towards implantation in patients with CMV related deafness in those, with and without MRI detectable brain changes, as the benefits of implantation outweigh the risks. Variable results should be expected however and this should be incorporated in to the consent and counselling process.

The pattern of abnormalities in the adult temporal bone group mirrored that of the paediatric group. Incomplete partition type 2 was the most commonly recorded abnormality followed by an enlarged endolymphatic sac. These abnormalities which would have been present since birth are a feature of the expanding criteria for cochlear implantation and the inclusion of these patients in to candidacy. The variety of abnormalities detected in the adult cohort was more varied in the intracranial group. Atrophy and previous infarction is expected in the aging adult and acquired abnormalities such as trauma and microangiopathic change are also more likely in older patients. None of these would have been contraindications to implantation at any stage during the work up process. The benefits of cochlear implantation in the adult cohort is only beginning to be understood. However, the cost effectiveness and benefits to quality of life scores, speech and audiology scores, cognitive function and social isolation are apparent from previously published studies[16-18]. Furthermore, the positive impact on

cognitive function and cognitive decline has been demonstrated through maintaining auditory ability through cochlear implantation in older adults[5]. As candidacy guidelines expand to incorporate older adults, the variety and number of intracranial abnormalities will surely increase and these should not be a contraindication to surgery.

In a previous publication from this institution the revision rates for cochlear implantation were recorded at 5% with device failure accounting for 3.1% and wound infection and electrode migration accounting for majority of the remainder[6]. None of the failures or revision cases were due to anatomical abnormalities. The previously described benefits combined with the low risk nature of surgery in the adult cohort place the risk benefit balance firmly in favour of implantation in appropriate candidates. In the present series the only adult complication encountered was not due to the associated radiological abnormality which was, in this case, an aberrant vessel looping under the internal carotid artery. This finding did not influence the hardware failure.

In the paediatric group of patients there were 3 gushers recorded which would be in keeping with previous reports by Papsin and Suri who reported a CSF gusher rate of 6-7% in paediatric patients with cochlear anomalies[10,19]. Of the 3 cases where gushers were encountered, each had dilated vestibules as part of incomplete partition abnormalities which has been a recognised risk for CSF gushers. A similar rate of CSF gusher was found in a smaller study than the present paper which compared complication rates between inner ear anomalies and congenitally deaf patients with no abnormalities[20]. A systematic review of cochlear implantation in children with anomalous cochleovestibular anatomy found a much higher rate of gushers, however the authors rightfully point out that heterogeneity amongst papers limited the strength of their conclusions [21]. Only 2 out of the 57 cases of abnormalities were abandoned intraoperatively owing to unfavourable anatomy and increasing risk to the patient of complications. The abandoned cases were cases of type 1 mondini dysplasia and hypoplastic cochlea respectively. Although these abnormalities raise awareness for potential complications, they are not in themselves contraindications as demonstrated by the cases in this series and others, where successful implantation occurred in the presence of such abnormalities. The single tegmen breach that occurred in this series was repaired intraoperatively, this was in a case of white matter changes related to CMV and not related to the complication encountered. In this sense, it is not possible to draw any conclusions on mitigation of complications in the presence of these abnormalities. However, the presence of such abnormalities should alert the surgeon to be extra vigilant. The complication rate recorded in the present study is in keeping with previously published literature and lower than that reported by a systematic review on the topic. While the MDT should be mindful of the higher risk of complications in cases of inner ear malformation, they should not be precluded.

Results from this study demonstrate that adult outcomes when measured with BKB results are favourable even in those with brain and temporal bone abnormalities. Initial electrode function was measured with NRT at the time of insertion for all patients and did not correlate with ultimate BKB scores indicating that faulty electrodes or incomplete insertion was not a causative factor in BKB results. The average and median BKB results were slightly worse in the brain abnormality group which may be accounted for by a degree of auditory processing difficulties that could not be measured. There have been many studies examining outcomes in the adult population but none have focused specifically on radiologically detected abnormalities pre op. Although the sample size is modest, the outcome profile paired with the minimal complication rate serves to strengthen the argument for adult implantation even in the presence of these types of abnormalities.

The outcomes for the paediatric cohort with temporal bone abnormalities is in keeping with other studies that suggest that these children do benefit from implantation even though average scores may lag behind their normal anatomy counterparts[20]. While patients with brain abnormalities scored less on average than the temporal bone group this can be explained by the larger number of CMV related changes detected and the previously described reasons. Findings from the current study are supported by ample evidence demonstrating that children with inner ear malformations can make improvements in speech perception and production following implantation[7,22-25]. What we are not able to account for in the present study is the individual goals that patients who opt to undergo implantation aim for. In some cases sound localisation may be the goal whereas in others, speech discrimination and production may be the expected outcome.

The optimal imaging modality for preoperative planning has been debated before with proponents of dual modality pointing out that important findings that can explain hearing loss aetiology can be missed if only one is used. Detractors from dual modality imaging would argue that resource limitations as well as the need for keeping unnecessary doses of radiation at a minimum, particularly in a paediatric patient cohort, necessitate only one form of imaging.

A study that compared 92 high resolution CT and FIESTA MRs of the temporal bone and FLAIR MR of the brain found that HRCT was inadequate for identifying early obliterative labyrinthitis and the presence of a cochlear nerve within the IAM, while MR demonstrated difficulties in identifying enlarged vestibular aqueducts and narrow cochlear canals and lead to the conclusion that dual modality should be employed in the presence of craniofacial abnormalities, CHARGE syndrome, a history of meningitis or cochlear dysplasia[26]. Although there were some abnormalities not detected, the most common abnormalities including EVAs

and various types of incomplete partition were all detected by both CT and MRI. On the other extreme end of the argument, a study group claimed that in their cohort of 118 adults in which 23% had radiological abnormalities, no deviation from the surgical plan occurred leading them to conclude that in adult cases of progressive sensory neural hearing loss, pre operative imaging was not necessary[27].

The surgical approach was altered based on pre operative imaging in at least two cases in the present study, although these were both in the paediatric cohort. The optimal balance is likely in between these two extremes. All children undergoing candidacy assessment should have imaging to exclude aberrant anatomy or absence of a cochlear nerve. Adult patients may require either CT or MR depending on history and clinical findings.

The detection rate for inner ear abnormalities appears to be consistent across multiple centres and makes up a small but not insignificant number of patients who undergo cochlear implant. Specialist centres who hold MDT conferences and have access to multi-disciplinary expert opinion are adept at identifying abnormalities pre operatively and customising treatment accordingly. The detection of inner ear or brain abnormalities in the paediatric and adult cohort of patients undergoing cochlear implantation should not be a contraindication. Complication rates are higher compared to normal anatomy counterparts but still remain in the acceptable range. The outcomes are less predictable in the adult group and the paediatric group of patients with CMV related changes detectable on MRI however these patients still benefit from receiving cochlear implants. Standardisation of outcome measurement across centres would allow amalgamation of results for more powerful findings.

Declarations of Conflicts of Interest:
None declared.

Corresponding Author:
Holly Jones,
National Hearing Implant & Viani Research Centre,
Department of Otology and Cochlear Implantation,
Beaumont University Hospital,
Beaumont Rd.,
Dublin 9,
Ireland.
E-Mail: hollyjones@rcsi.com

References:

1. Jensen J. Malformations of the inner ear in deaf children. A tomographic and clinical study. *Acta Radiol Diagn (Stockh)*. 1968:Suppl 286:3+.
2. Wentland CJ, Ronner EA, Basonbul RA, et al. Utilization of diagnostic testing for pediatric sensorineural hearing loss. *International Journal of Pediatric Otorhinolaryngology*. 2018/08/01/ 2018;111:26-31. doi:https://doi.org/10.1016/j.ijporl.2018.05.024
3. Sennaroglu L, Saatci I. A new classification for cochleovestibular malformations. Article. *Laryngoscope*. 2002;112(12):2230-2241. doi:10.1097/00005537-200212000-00019
4. Pamuk G, Pamuk AE, Akgöz A, Bajin MD, Özgen B, Sennaroğlu L. Radiological measurement of cochlear dimensions in cochlear hypoplasia and its effect on cochlear implant selection. *J Laryngol Otol*. Jun 2021;135(6):501-507. doi:10.1017/s0022215121000979
5. Sarant J, Harris D, Busby P, et al. The Effect of Cochlear Implants on Cognitive Function in Older Adults: Initial Baseline and 18-Month Follow Up Results for a Prospective International Longitudinal Study. *Front Neurosci*. 2019;13:789. doi:10.3389/fnins.2019.00789
6. Wijaya C, Simões-Franklin C, Glynn F, Walshe P, Reilly R, Viani L. Revision cochlear implantation: The Irish experience. *Cochlear Implants Int*. Nov 2019;20(6):281-287. doi:10.1080/14670100.2019.1647372
7. Buchman CA, Copeland BJ, Yu KK, Brown CJ, Carrasco VN, Pillsbury HC, 3rd. Cochlear implantation in children with congenital inner ear malformations. *Laryngoscope*. Feb 2004;114(2):309-16. doi:10.1097/00005537-200402000-00025
8. Luntz M, Balkany T, Hodges AV, Telischi FF. Cochlear implants in children with congenital inner ear malformations. *Arch Otolaryngol Head Neck Surg*. Sep 1997;123(9):974-7. doi:10.1001/archotol.1997.01900090090013
9. Sennaroglu L. Cochlear implantation in inner ear malformations--a review article. *Cochlear Implants Int*. Mar 2010;11(1):4-41. doi:10.1002/cii.416
10. Suri NM, Prasad AR, Sayani RK, Anand A, Jaychandran G. Cochlear implantation in children with Mondini dysplasia: our experience. *The Journal of Laryngology & Otology*. 2021;135(2):125-129. doi:10.1017/S0022215121000372
11. Jackler RK, Luxford WM, House WF. Congenital malformations of the inner ear: a classification based on embryogenesis. *Laryngoscope*. Mar 1987;97(3 Pt 2 Suppl 40):2-14. doi:10.1002/lary.5540971301
12. Morlet T, Pazuniak M, O'Reilly RC, Kandula V, Choudhary AK. Cochlear Nerve Deficiency and Brain Abnormalities in Pediatric Patients. *Otol Neurotol*. Mar 2017;38(3):429-440. doi:10.1097/mao.0000000000001308
13. Ramirez Inscoe JM, Nikolopoulos TP. Cochlear implantation in children deafened by cytomegalovirus: speech perception and speech intelligibility outcomes. *Otol Neurotol*. Jul 2004;25(4):479-82. doi:10.1097/00129492-200407000-00014

14. Philips B, Maes LK, Keppler H, Dhooge I. Cochlear implants in children deafened by congenital cytomegalovirus and matched Connexin 26 peers. *International Journal of Pediatric Otorhinolaryngology*. 2014;78(3):410-415. doi:10.1016/j.ijporl.2013.11.009
15. Bolduc SH, Bussières R, Philippon D, Côté M. The Correlation of Congenital CMV Infection and the Outcome of Cochlear Implantation. *J Int Adv Otol*. May 2021;17(3):190-194. doi:10.5152/iao.2021.9335
16. Gumbie M, Olin E, Parkinson B, Bowman R, Cutler H. The cost-effectiveness of Cochlear implants in Swedish adults. *BMC Health Services Research*. 2021;21(1)doi:10.1186/s12913-021-06271-0
17. Gaylor JM, Raman G, Chung M, et al. Cochlear Implantation in Adults. *JAMA Otolaryngology–Head & Neck Surgery*. 2013;139(3):265. doi:10.1001/jamaoto.2013.1744
18. Maharani A, Pendleton N, Leroi I. Hearing Impairment, Loneliness, Social Isolation, and Cognitive Function: Longitudinal Analysis Using English Longitudinal Study on Ageing. *Am J Geriatr Psychiatry*. Dec 2019;27(12):1348-1356. doi:10.1016/j.jagp.2019.07.010
19. Papsin BC. Cochlear implantation in children with anomalous cochleovestibular anatomy. *Laryngoscope*. Jan 2005;115(1 Pt 2 Suppl 106):1-26. doi:10.1097/00005537-200501001-00001
20. Melo AS, Martins J, Silva J, Quadros J, Paiva A. Cochlear implantation in children with anomalous cochleovestibular anatomy. Review. *Auris Nasus Larynx*. 2017;44(5):509-516. doi:10.1016/j.anl.2017.02.003
21. Pakdaman MN, Herrmann BS, Curtin HD, Van Beek-King J, Lee DJ. Cochlear implantation in children with anomalous cochleovestibular anatomy: a systematic review. *Otolaryngol Head Neck Surg*. Feb 2012;146(2):180-90. doi:10.1177/0194599811429244
22. Karamert R, Tutar H, Altinyay Ş, et al. Cochlear Implantation in Inner Ear Malformations: Considerations Related to Surgical Complications and Communication Skills. *ORL*. 2021:1-8. doi:10.1159/000517562
23. Gaurav V, Rajguru R. Effects of radiological abnormalities in temporal bone and brain on auditory outcomes in cochlear implant recipient children. Article. *Indian Journal of Otology*. 2019;25(2):85-89. doi:10.4103/indianjotol.INDIANJOTOL_122_18
24. Ehrmann-Müller D, Kühn H, Matthies C, Hagen R, Shehata-Dieler W. Outcomes after cochlear implant provision in children with cochlear nerve hypoplasia or aplasia. *Int J Pediatr Otorhinolaryngol*. Sep 2018;112:132-140. doi:10.1016/j.ijporl.2018.06.038
25. Arnoldner C, Baumgartner WD, Gstoettner W, et al. Audiological performance after cochlear implantation in children with inner ear malformations. *Int J Pediatr Otorhinolaryngol*. Apr 2004;68(4):457-67. doi:10.1016/j.ijporl.2003.11.018
26. Trimble K, Blaser S, James AL, Papsin BC. Computed tomography and/or magnetic resonance imaging before pediatric cochlear implantation? Developing an investigative strategy. Article. *Otology and Neurotology*. 2007;28(3):317-324. doi:10.1097/01.mao.0000253285.40995.91

27. Roberts DM, Bush ML, Jones RO. Adult progressive sensorineural hearing loss: is preoperative imaging necessary before cochlear implantation? *Otol Neurotol*. Feb 2014;35(2):241-5. doi:10.1097/MAO.0b013e3182a437b3

Table 1. Patient Demographics

Program	
Adult • Average age at implantation • Median • Range	n=33 55.3yrs 57.5yrs 19-83yrs
Paediatric • Average age at B/L implantation • Median • Average age at U/L implantation • Median	n=57 3.1yrs 1.5yrs 5.4yrs 5yrs

Table 2. Imaging abnormalities by site

Site of primary abnormality	
Adult	
• Temporal Bone	20
• Brain	14
• Both	1
Paediatric	
• Temporal Bone	48
• Brain	5
• Both	4

Table 3. Adult imaging abnormalities

Adult abnormal temporal bones		**Adult brain abnormalities**	
Incomplete Partition type 2	6	Atrophy	2
Early Sclerosis	1	Old Infarct	5
Scala deficiency	1	Other	7
IAC Meningioma	1	• Acoustic schwannoma	1
Incomplete cochlear turn	1	• Previous trauma	1
Bilateral labyrinthitis ossificans	1	• Superficial siderosis	1

Enlarged endolymphatic sac	4	• Aberrant vessel	1
Hypoplastic cochlear nerve	1		
Intracanalicular vestibular schwannoma	1		
Cochlear signal loss	1		
Mastoid Effusion	1		
Adult brain and temporal bone abnormalities			
Incomplete partition type 2 and Vasculitic changes			1

Table 4. Paediatric imaging abnormalities

Paediatric Temporal Bone		**Paediatric Brain abnormalities**	
Incomplete Partition type 2	23	White matter changes due to CMV	4
Incomplete Partition type 1	4	Abnormal gyri	1
Unilateral ossification	1		
Enlarged Vestibular aqueduct	8		
Enlarge vestibule	2		
Hypoplastic Cochlear Nerve	1		
Anterior Jugular bulb	1		
Vestibular cochlear dysplasia	1		
Hypoplastic cochlea	1		
Hypoplastic semi-circular canal	1		
Endolymphatic sac dilation	1		
Basal turn cochlear inflammation	1		
Hypoplastic cochlear nerve	2		
Mastoid effusion	1		
Paediatric brain and temporal bone abnormalities			
IP 1 + hydrocephalus			1
IP II + Ectopic white matter			1
Absent + hypoplastic cochlear nerve + ANSD (T21)			1
Dilated cochlear aqueduct + previous medulloblastoma excision			1

Table 5. Adult audiological outcomes

IMJ
Official Journal of the Irish Medical Organisation

Ir Med J; March 2025; Vol 118; No. 3; P41
March 20th, 2025

	Adult temporal bone abnormalities		Adult Brain abnormalities		Adult brain and temporal bone abnormality	
	BKB (%)	NRT	BKB (%)	NRT	BKB (%)	NRT
Range	0-96	4-22	0-96	15-22	96	22
Mean	68.1	20.5	46.6	20.2		
Median	80		50			
Available for analysis (n)	16/18	16/18	11/14	13	1/1	1/1

Table 6. Paediatric audiological outcomes

	Paediatric temporal bone abnormalities				Paediatric brain abnormalities				Paediatric temporal bone and brain abnormalities			
	NRT	**LiP**	**SIR**	**CAP**	**NRT**	**LiP**	**SIR**	**CAP**	**NRT**	**LiP**	**SIR**	**CAP**
Mean	18.2	38.2	4.6	5.9	21.8	35.7	3	4.7	15.5	30.25	4.25	4
Range	0-22	0-42	1-6	0-7	21-22	25-42	2-5	3-6	9-22	0-42	2-6	0-7
Available for interpretation	40/48	21/48	21/48	21/48	4/5	3/5	3/5	3/5	2/4	4/4	4/4	4/4