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Mini-Review

Anatomical Variations and Surgical Challenges in Pediatric Cochlear Implantation: A Single-Center Experience of 489 Cases

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ABSTRACT

Background: Cochlear implantation (CI) is the gold standard for severe-to-profound sensorineural hearing loss in children. However, temporal bone anatomical variations and inner ear malformations can significantly increase surgical complexity.

Objective: To evaluate the prevalence of anatomical variations in pediatric CI candidates, identify associated surgical challenges, and describe technical adaptations implemented to achieve safe and complete electrode array insertion.

Methods: A retrospective single-center study was conducted between 2014 and 2025 at Ibn Rochd University Hospital, including 489 pediatric patients (<18 years) who underwent primary CI. Preoperative radiological evaluation included high-resolution temporal bone computed tomography (HRCT) and 3T magnetic resonance imaging (MRI).

Results: The mean age was 59 months, with a male-to-female ratio of 51% to 49%. Significant anatomical variations/anomalies were identified in 18.6% of cases. Facial nerve anomalies included tympanic segment dehiscence (7.2%), bifid nerve (0.2%), and aberrant course (0.2%). Vascular variations were dominated by sigmoid sinus procidence (10.5%). Round window (RW) access required surgical modification (extended RW or cochleostomy) in 30% of patients. Surgical adaptations-such as diamond drill usage, bone dust/cartilage obliteration (1.2%), and scala vestibuli insertion in complete tympanic scala ossification (2 cases)-enabled successful insertion across the cohort.

Conclusions: Pediatric anatomical variability necessitates comprehensive preoperative radio-anatomical mapping. Systematic surgical adaptation strategies are crucial to optimize surgical outcomes and prevent major neurovascular complications.

Keywords: Cochlear implantation, Pediatric hearing loss, Anatomical variations, Facial nerve, Round window, Mastoidectomy, Temporal bone CT

Introduction

Congenital and early-onset severe-to-profound sensorineural hearing loss (SNHL) affects 1 to 3 per 1,000 live births globally. Deprived of auditory input during critical neurodevelopmental windows, affected children experience severe impairments in speech perception, language acquisition, and cognitive development. Cochlear implantation (CI) has established itself as the gold standard intervention for auditory rehabilitation in this population[1,2].

While the surgical technique-comprising cortical mastoidectomy, posterior tympanotomy, and intracochlear electrode placement-is well standardized, pediatric otologic surgery presents unique anatomical challenges. These stem from incomplete temporal bone pneumatization, dynamic postnatal skull base growth, and a higher incidence of congenital inner ear malformations or post-meningitic labyrinthine ossification.

Understanding the spectrum and frequency of anatomical variations is paramount for surgical planning and complication avoidance. This single-center study evaluates a cohort of 489 pediatric cochlear implant recipients to categorize anatomical variations, assess surgical difficulties, and outline tailored surgical strategies[3,4].

Materials and Methods

Study Design and Patient Selection

This retrospective descriptive and analytical study was conducted at the Department of Otorhinolaryngology and Head and Neck Surgery, 20 August 1953 Hospital, Ibn Rochd University Hospital Center, Casablanca, Morocco. The study period spanned 11 years (2014-2025).

Inclusion criteria were: (1) age < 18 years at the time of primary cochlear implantation; (2) bilateral severe-to-profound SNHL confirmed by behavioral audiometry and auditory brainstem responses (ABR); and (3) complete preoperative radiological documentation including high-resolution temporal bone computed tomography (HRCT) and 3-Tesla inner ear magnetic resonance imaging (MRI). Exclusion criteria comprised incomplete medical/radiological records and revision surgical cases.

Radiological Assessment and Data Collection

Preoperative HRCT (sub-millimeter axial and coronal reconstructions) and 3T MRI (3D CISS/FIESTA sequences) were systematically reviewed by a consensus panel of two senior otologic surgeons. Evaluated anatomical parameters included:

- **Mastoid and Middle Ear:** Degree of pneumatization, anterior/medial positioning of the sigmoid sinus, height of the dura/jugular bulb, and status of the ossicular chain.
- **Facial Nerve (CN VII):** Bony canal integrity (dehiscence), nerve course anomalies (bifurcation, anterior displacement, or ectopia).
- **Inner Ear & Cochlea:** Congenital malformations (incomplete partition, cochlear hypoplasia/aplasia), RW niche visibility/accessibility, and labyrinthine obliteration (fibrosis or ossification).

Results

Demographic characteristics

A total of 489 pediatric patients met the eligibility criteria. The cohort comprised 249 boys (50.9%) and 240 girls (49.1%). The overall mean age at surgery was 59 months (range: 8 months to 17.5 years). The largest age group represented was 2-5 years (58.1%), followed by 6–12 years (19.0%) **(Table 1)**.

Table 1: Demographic breakdown of 489 pediatric cochlear implant recipients.

Age Category	Number (n)	Proportion (%)	Male (%)	Female (%)
< 12 months	18	3.70%	52%	48%
12–24 months	48	9.80%	49%	51%
2–5 years	284	58.10%	51%	49%
6–12 years	93	19.00%	48%	52%
> 12 years	46	9.40%	50%	50%
Total	489	100.00%	50.90%	49.10%

Prevalence of anatomical variations and technical adaptations

Anatomical variations affecting the inner ear, facial nerve, or surrounding vascular structures were encountered in 18.6% of the cohort.

- **Facial Nerve Anomalies:** Tympanic/mastoid segment dehiscence was detected in 35 patients (7.2%). Rare anomalies included a bifid facial nerve (0.2%, n=1) and an aberrant facial nerve course traversing the round window niche (0.2%, n=1).
- **Vascular & Dural Variations:** Anterior procidence of the sigmoid sinus was present in 51 cases (10.5%), narrowing the mastoidectomy cavity. Low-hanging tegmen/dural procidence was identified in 6 patients (1.2%).
- **Cochlear Access & Labyrinthine Pathology:** Direct visualization of the RW membrane through a standard posterior tympanotomy was impaired in 147 cases (30.0%), requiring extended RW drilling or formal anterior-inferior cochleostomy. In 2 post-meningitic patients with total obliteration of the scala tympani, array insertion was successfully achieved via the scala vestibuli.

Discussion

Global prevalence and comparative analysis

Anatomical findings in pediatric cochlear implantation vary substantially across published series, partly because studies use different definitions and inclusion criteria. In our single-center cohort of 489 patients, significant anatomical variations were observed in 18.6% of cases. This figure should therefore be interpreted in the context of the specific anatomical criteria used in the present study[5,6].

Table 2: Comparison of anatomical variation rates in major pediatric CI cohorts.

Study / Author	Cohort Size (n)	Reported Variation Rate (%)
Current Study (Ibn Rochd, Casablanca)	489	18.60%
London et al. (2020)	743	28.00%
Papsin (2005)	395	42.00%
Pradhananga et al. (2022)	186	35.00%

Differences between published series should be interpreted cautiously because the cohorts and definitions are not directly comparable. Papsin's series evaluated cochleovestibular

anomalies in children undergoing implantation, whereas Loundon et al. focused specifically on inner ear malformations. The present study used a broader operative-anatomical classification and therefore should not be compared directly with studies using different endpoints.

Facial nerve aberrations and intraoperative risk management

The facial nerve is the most critical structure at risk during posterior tympanotomy. In pediatric temporal bones, incomplete mastoid pneumatization and delayed Fallopian canal ossification increase susceptibility to iatrogenic trauma. Dehiscence of the tympanic or mastoid segment was the most common neural anomaly in our series (7.2%).

Extremely rare variations-such as bifid facial nerve (0.2%) or aberrant facial nerve courses crossing the RW niche (0.2%)-render standard posterior tympanotomy impossible or extremely hazardous. In such scenarios, continuous intraoperative electromyographic (EMG) nerve monitoring is indispensable. When an aberrant nerve completely obstructs the facial recess, alternative surgical routes must be considered, including the suprameatal approach or a transcanal/endoscopic-assisted technique.

Vascular and dural contraints: Sigmoid sinus and tegmen procidence

Anterior displacement of the sigmoid sinus (10.5% in our series) significantly compromises the surgical corridor, narrowing the distance between the sinus wall and the external auditory canal. Unprepared drilling in this region risks catastrophic venous hemorrhage or sinus thrombosis.

Management strategies involve creating a surgical corridor tailored to the vascular anatomy. This requires preserving an ultrathin eggshell layer of cortical bone over the sinus (*skeletonization*) to protect the vessel wall, or using soft dural/venous retractors and neurosurgical cottonoids when gentle refolding is required to gain surgical trajectory toward the facial recess.

Similarly, a low-hanging tegmen tympani (1.2%) narrows the superior boundary of the facial recess. In these cases, diamond burrs must be utilized exclusively under continuous irrigation to prevent thermal injury or accidental dural tear leading to cerebrospinal fluid (CSF) leakage.

Cochlear access, Labyrinthine ossification, and Alternative insertion routes

RW insertion is universally favored to minimize intracochlear trauma and preserve residual acoustic hearing. However, in 30% of our cases, anatomical overhang of the RW niche or unfavorable facial recess trajectories mandated extended drilling of the crista fenestrae or a formal anteroinferior cochleostomy.

In post-meningitic labyrinthitis ossificans, osteogenesis preferentially affects the basal turn of the scala tympani near the round window. When drill-out of the basal turn (up to 6-8 mm) fails to identify a patent lumen, forcing the electrode array risks complete array buckling or extracochlear displacement.

In two cases featuring complete basal scala tympani obliteration, electrode placement was achieved via the scala vestibuli. These cases demonstrate the feasibility of this alternative route in selected patients. Published pediatric series also support scala vestibuli insertion as a viable option when the scala tympani is ossified, although outcomes depend on the extent of ossification and the individual cochlear anatomy.

Dural defect management and CSF gusher prevention

Certain congenital inner ear malformations can be associated with an increased risk of intraoperative cerebrospinal fluid (CSF) leakage. Preoperative CT and MRI are therefore important for identifying malformations that may alter the surgical approach and the risk profile[7,8].

In our cohort, dural procidence was encountered in 1.2% of cases. Prevention of postoperative CSF leaks and secondary bacterial meningitis relies on meticulous sealing techniques. Following electrode insertion, the cochleostomy or RW opening was routinely packed with autologous fascia, cartilage grafts, or bone dust combined with fibrin glue. No postoperative CSF leaks or intracranial infections occurred in our series.

Conclusion

Pediatric cochlear implantation in the presence of anatomical variations is a demanding procedure requiring tailored surgical expertise. In this large single-center study of 489 children, 18.6% presented with significant anatomical variations or anomalies.

Key determinants of surgical success include:

- Systematic preoperative radio-anatomical mapping using HRCT and 3T MRI to identify neurovascular anomalies and cochlear patency;
- Universal intraoperative facial nerve monitoring;
- Mastery of surgical adaptation protocols, including scala vestibuli insertion for ossified cochleae, skeletonization of procident vascular structures, and robust soft-tissue seal of the labyrinth.

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