Inner Ear Malformations in Cochlear Implantation Candidates at Mother and Child Hospital of Bingerville (Côte d'Ivoire)

Marie-Josée Tanon-Anoh[1] Esther K. Badou-Nguessan[2] Olivier-Didier Zamblé Tra Bi[1]
Corine M.A. Assebian-Houenou[1] Raïssa Michelle Kabas[1] Yannick James Tatsitsa Langouo[2]

[1]Department of Pediatric ENT, Mother and Child Hospital of Bingerville, Université Félix Houphouët-Boigny, Abidjan Riviera, Côte d'Ivoire
[2]Department of ENT, Yopougon Teaching Hospital, Côte d'Ivoire

Address for correspondence Marie Josée Tanon-Anoh, MD, Department of Pediatric ENT, Mother and Child Hospital of Bingerville, BP 725 Cidex 3 Abidjan Riviera, Côte d'Ivoire, (e-mail: anohjose@ymail.com, josee.tanon@hme.ci).

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Abstract

Objective The aim of this study was to determine the prevalence and types of inner ear and/or cochlear nerve anomalies in children who are candidates for cochlear implantation.

Methods This was a descriptive cross-sectional study with a **r**etrospective review of medical and imaging records performed in a tertiary care children's hospital. All cochlear implants candidates under 15 years old with imaging assessment between January 2019 and December 2021 were concerned. The studied parameters were reason for consultation, risks factors, inner ear malformations (IEMs) classified and stratified by sensorineural hearing loss (SNHL) degree, gender, and age at diagnosis.

Results In total, 81 children (162 ears) were included in the study. Abnormalities of the inner ear were found in nine children representing a prevalence of 11.1%. These children were aged between 2 and 14 years. The average age at diagnosis of SNHL was 3.5 years. Seven children had bilateral anomalies and two unilateral anomalies, that is, 16 ears presenting 40 malformations. These included 1 complete labyrinthine aplasia (2.5%), 12 cochlear malformations (30%), 1 common cavity (2.5%), 17 vestibular/squamous cell carcinoma (SCC) malformations (42.5%), and 5 internal auditory canal malformations (12.5%). Three ear out of 16 with abnormalities demonstrated a deficient cochlear nerve. There was an association between IEMs and profound deafness, and age at diagnosis of SNHL less than 2 years.

Conclusion This study shows that 11.1% of children with profound deafness have IEMs. SCC malformations and cochlear hypoplasia were the most common. A precise description of these malformations during the imaging assessment is particularly useful for cochlear implantation to better plan this surgery.

Keywords
- cochlear implantation
- hearing loss
- inner ear malformations
- imaging
- radiology

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Introduction

Inner ear and vestibulocochlear nerve malformations may lead to congenital sensorineural hearing loss (SNHL).[1] Hearing loss is the most common sensory disability in children. It is considered that 1 in 1,000 children has severe or profound deafness at birth and 90% of birth hearing loss are sensorineural.[2]

The reported prevalence of inner ear malformations (IEMs) in individuals with congenital hearing loss varies from 2.3 to 28.4%.[3] A variety of congenital anomalies are seen, syndromic and nonsyndromic.[4] These anomalies have been grouped according to various classifications and the most used is proposed by Sennaroglu.[5,6]

In cases of severe-to-profound deafness, cochlear implant is proven to be the most beneficial in children. For this management, IEMs may not only affect the decision to perform the implant procedure but also increase the risk of complications.[1] In Côte d'Ivoire, our cochlear implantation experience is recent (December 2015), and the first cochlear implantations at Bingerville Mother-Child Hospital were performed in 2020. Moreover, no study has been carried regarding the inner ear anomalies in our setting. The lack of data on malformations therefore justifies this study. Thus, the objectives were to determine the prevalence and the types of inner ear and/or cochlear nerve abnormalities in children with severe or profound SNHL in Côte d'Ivoire.

Methods

This is a descriptive cross-sectional study with a review of the medical files of children under 15 years old, candidates for cochlear implantation, who presented to our hospital for a preimplantation imaging assessment (computed tomography [CT] and magnetic resonance imaging [MRI]), from January 2019 to December 2021. All cochleovestibular or cochlear nerve malformations, isolated or associated, were included in the study. Malformations isolated from the windows, abnormal hyperdensity, or ossified lesions were not retained. They were categorized by side (unilateral, bilateral), site (cochlea, vestibule, semicircular canal, entire labyrinth), and Sennaroglu classification.[5,6]

The variables to be studied were reason for consultation, risk factors, types of inner ear abnormalities classified and analyzed according to degree of SNHL, gender, and age at diagnosis. Significant risk factors and history corresponded to the presence of parameters that could impact the occurrence of cochleovestibular malformations: TORCH (TOxoplasmosis, Rubella, Cytomegalovirus, Herpes simplex virus) infection during pregnancy and other microorganisms; gestational age less than 34 weeks, birth defects of the head and neck.

After collection, data was tabulation using Microsoft Excel Worksheet and analyzed statistically using Statistical Package for Social Studies (SPSS) Version-23. Results were analyzed using descriptive statistic including frequencies, percentages, mean, and standard deviation. Chi-squared and Fischer's exact tests were used to see association (p-value < 0.05).

The study was conducted after administrative approvals were obtained; and the anonymity of the tools and data collected was ensured.

Results

Prevalence

During this study, we identified 81 patients (162 ears) with available clinical information and imaging results. Among these children, nine (9) had cochleo-vestibular malformations, that is, a prevalence of 11.1%.

Sociodemographic and Clinical Data of Malformations Cases

They are summarized into ►**Table 1**.

The patients were unschooled except for one. Case 6 had a pathological history such as diffuse brain damage. No significant risk factor was identified in these nine patients.

The age of the patients ranged from 2 to 14 years, with mean age of 5.2 years (62.4 months) and a standard deviation of 4.6 years. There were five males for four females (sex ratio: 1.25).

Table 1 Sociodemographic and clinical data

	Age	Sex	Sibling rank	Consultation reason	Age at diagnosis of deafness	ENT and general examination
Case 1	25 mo	Female	1st	Language delay	12 mo	Auricular anomalies + cervical fistulas + congenital PFP[a]
Case 2	42 mo	Male	1st	Language delay	20 mo	Normal
Case 3	29 mo	Male	1st	Language delay	12 mo	Normal
Case 4	55 mo	Male	1st	Language delay + hearing loss	24 mo	Normal
Case 5	168 mo	Female	5th	Hearing loss	141 mo	Normal
Case 6	39 mo	Male	1st	Hearing loss	36 mo	Normal
Case 7	53 mo	Male	1st	Language delay	45 mo	Autism spectrum disorder
Case 8	112 mo	Female	1st	Language delay + hearing loss	60 mo	Normal
Case 9	39 mo	Female	2nd	Language delay	30 mo	Normal

Abbreviation: PFP, peripheral facial paralysis.

The reason for consultation was a language delay in 7/9 cases. The age at diagnosis ranged from 1 to 12 years with a mean age of 3.5 years (42.2 months). Among the 16 ears with malformations, 11 had profound hearing loss, 4 had severe hearing, and 1 had moderate hearing loss. A branchio-oto-renal syndrome (auricular abnormalities, bilateral prehelical and second branchial arch fistulas, small right kidney) with congenital deafness was identified (case 1). A persistence of the ductus arteriosus was noted in case 5.

Radiological Assessment

A petrous CT scan was performed in all patients (100%) and an MRI of the inner ear and brain in eight cases (88.9%). All patients had multiple IEMs except case 8. Seven children had bilateral anomalies of the inner ear and/or cochlear nerve and two had unilateral anomalies, for a total of 16 malformed ears. A total of 40 types of IEM were identified in these 16 ears (►**Table 2**). Abnormalities were on the right side in 23 cases (57.5%) and on the left side in 17 cases (42.5%). In four cases, brain MRI also showed frontal and/or biparietal leukomalacia, predominantly on the left side.

Reporting to the 16 ears with IEMs, there are 1 ear with complete anomaly of the labyrinthine (6.25%), 1 ear with common cavity (6.25%), 8 ears with abnormalities of the cochlea (50%), 8 ears with vestibular abnormalities (50%), 5 ears with abnormalities of the internal auditory canal (31.25%), 1 vestibular aqueduct abnormality (6.25%), and 3 ears with cochlear nerve abnormalities (18.75%). One ear had at least one abnormality at each site on the same side (case 6).

The IEMs distribution according to Sennaroglu and Bajin classification[5] is summarized in ►**Table 3**

Malformations of the Inner Ear according to Degree of Deafness, Age, and Sex of Patients

The *p*-value compares the rate of child with age at diagnosis inferior to 2 years in those with malformations and those

Table 2 Distribution of malformations according to their frequency

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7	Case 8	Case 9	(*n*) % of 40 IEM	
Enlarged IAC					1					(1) 2.5	(5) 12.5%
IAC abnormalities[a]	1			2						(3) 7.5	
Vascular-nervous conflict in IAC				1						(1) 2.5	
Hypoplastic cochlear nerve						1		2		(3) 7.5	(3) 12.5%
Complete labyrinthine aplasia		1								(1) 2.5	(1) 2.5%
Cochlear aplasia		1								(1) 2.5	(12) 30%
Cochlear hypoplasia			2		1		1			(4) 10.0	
Modiolus malformations[b]			2		1				2	(5) 12.5	
Cochlea with less than 2 turns	1									(1) 2.5	
Dilated cochlear duct					1					(1) 2.5	
Common cavity (cystic cavity)	1									(1) 2.5	(1) 2.5%
Bone vestibular dysplasia (utricle and saccule)			1		1					(1) 2.5	(3) 7.5%
Dilated vestibule			1		1					(2) 5.0	
SCCl hypoplasia			2		1					(3) 7.5	(11) 27.5%
SCCs hypoplasia or aplasia			2		1				2	(5) 12.5	
SCCp hypoplasia or aplasia			1			1			1	(3) 7.5	
SCCs bone capsule dehiscence					1		1			(3) 7.5	(3) 7.5%
Enlarged (or dilated) vestibular aqueduct				1						(1) 2.5	(1) 2.5%
Outer and middle ears abnormalities associated											
–Prolapse of jugular vein	1 Yes	Yes							Yes	1	2
–Moderate atresia of EMAs										2	
–Ossicular dysmorphism (stapes) and narrow oval window										Yes	
Brain abnormalities											
Leukomalacia											

Abbreviations: IEMs, inner ear malformations; SCC, squamous cell carcinoma.
[a]Internal auditory canal (IAC): 1 horizontalization, 2 narrows.
[b]Modiolus: 4 incomplete, 1 short.

without, and the rate of male in those with malformations and those without (►**Table 4**).

There was a significant association (p-value < 0.05) between age at diagnosis and IEM. The occurrence of IEMs was not gender related (p-value > 0.05).

The association between the degree of hearing loss and IEMs is presented in ►**Table 5**.

There was a significant correlation (p-value < 0.05) between IEM and degree of deafness: IEM was associated with severe and profound hearing loss.

Discussion

We found a prevalence of 11.1% of malformations in children's candidates for cochlear implantation, which is within the range of prevalence reported worldwide: 7.5 to 20%.[7–9] With a prevalence of 3.7% in cases of unilateral SNHL, the study by Masuda and Usui[10] showed a significant increase in the prevalence of malformations in the case of bilateral SNHL.

Gender does not seem to be a predisposing factor for ear malformations either in our study or in the literature.[4,10] However, studies concerning the prevalence by sex of the different malformations in general are lacking, apart from those on the anomalies of the external ear that note a male predominance.[11] More than the age of the patients included, the present study demonstrated a significant correlation between IEMs and early age at diagnosis. The children were mostly the first born in the family (7/9 cases), like in other studies on deafness.[12,13] Children were mostly out of school, perhaps because of the hearing loss?

The main reason for consultation in children's SNHL was a language delay according to Ridal et al.[14] The

Table 3 IEMs distribution according to Sennaroglu and Bajin classification

		Effective	Percentage
Complete anomaly of the labyrinthine (or Michel deformity)		1	6.25
Common cavity		1	6.25
Cochlear aplasia		1	6.25
Incomplete partition	IP type II (Mondini deformity)	1	6.25
	IP type I	1	6.25
Cochlear hypoplasia	CH type II	3	18.75
	CH type III	1	6.25
Isolated SCC hypoplasia		2	12.50
Isolated narrow IAC		1	6.25
Narrow IAC associated with SCCs bone capsule dehiscence		1	6.25
Hypoplastic cochlear nerve associated with SCCs bone capsule dehiscence		1	6.25
Isolated hypoplastic cochlear nerve		2	12.50

Abbreviations: CH, cochlear hypoplasia; IAC, internal auditory canal; IEMs, inner ear malformations; IP, Incomplete partition; SCC, semicircular canal.

Table 4 Age of diagnosis/sex and presence of inner ear malformations

	Age of diagnosis		Sex	
	≤ 2 years *n* (%)	> 2 years *n* (%)	Male	Feminine
Malformations, yes	8 (50)	4 (50)	9 (56.3)	7 (43.7)
Malformations, no	24 (16.4)	122 (83.6)	76 (52.1)	70 (47.9)
	Fisher's exact test *p*-Value = 0.004; RR = 4.06		Chi-squared test = 0.10 < 3.84 *p*-Value = 0.75	

Abbreviation: RR, relative risk.

Table 5 Degree of SNHL and presence of inner ear malformations

	Profound and severe deafness, *n* (%)	Other level of deafness, *n* (%)	*p*-Value[a]
Malformations, yes	15 (93.7)	01 (06.3)	0.008
Malformations, no	88 (50.3)	58 (39.7)	

Abbreviations: RR, relative risk; SNHL, sensorineural hearing loss.
[a]Fisher's exact test; RR: 8.59 > 1.

profound deafness would favor early diagnosis, even in under-equipped countries, because of the main impact on the language development of young children. We did not note any pathological history in our work, due to the size of our sample. However, the role of pregnancy and childbirth conditions as well as prenatal infections, in particular TORCH, are known to be responsible of congenital craniofacial malformations, including auricular. Indeed, ear malformations may have not only an acquired background (infections, chemical agents, irradiation, etc.) but also a genetic origin. Theses malformations can affect outer/middle ant inner ear, sometimes in combination. But the different embryogenesis of the outer/middle ear and the inner ear explains malformations in outer and/or middle ear without IEMs.[3] An association with other malformations (cervical, renal, cardiac, etc.) is noted in two patients, which may be part of so-called syndromic genetic deafness described in 10 to 30% of cases of genetic deafness.[3] The IEMs were associated with profound and/or severe degree sensorineural deafness in majority of cases, with varying rates.[8,10]

CT is the first-line examination in the exploration of the ear in children, but CT and MRI must be complementary in the preoperative assessment. MRI essentially makes it possible to look for an anomaly of the cochlear nerve or of the labyrinthine membranous structures and of the brain.[1,4] In most cases, the underlying disorders involve the membranous labyrinth at a microscopic level and therefore radiological examinations are entirely normal.[1] The most used classification is that in eight groups.[1,5,6] Majority of patients demonstrated multiple abnormalities.[5] The prevalence of any type of IEMs is variable with large differences in the proportions reported by studies. In our series, the most common abnormalities were SCC malformations, including SCC hypoplasia and dehiscence of superior SCC bone capsule. In second position, we had cochlear malformations with cochlear hypoplasia type II or III (25%) while incomplete partitions were the most frequent malformations in studies by Sennaroglu and Sun.[7,15] Conversely, the most common malformations were dilated vestibules for other authors.[9,16] An enlarged vestibular aqueduct has been reported at high rates (40–56%) in several studies, whereas they represent only 2.5% of the malformations in our series.[9,15,17] No documented explanation could justify these significant differences.

Conclusion

This study shows that 11.1% of children with profound SNHL have inner ear malformations. SCC anomalies and cochlear hypoplasia were the most common. A precise description of these malformations during the imaging assessment is particularly useful for cochlear implantation, for better planning of this surgery.

Conflict of Interest
None declared.

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