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Sensorineural Hearing Loss and Cochleovestibular Malformations in Children in Côte d'Ivoire

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Abstract

Objective: to find the distribution of various malformations and to report the clinical features of commonly encountered IEMs in our practice.

Methods: This was a retrospective cross-sectional study, over a period of 14 months (January 2019 to March 2020), in the ENT departments of University Hospitals and the Mother and Child Hospital in Bingerville. All patients, aged up 15 years with computed tomography and/or MRI available, who had a cochleovestibular malformation and sensorineural or predominantly sensorineural mixed hearing loss confirmed were included.

Results: The prevalence of sensorineural hearing loss related to cochleovestibular malformations was 3.75% (6 cases out of 160 patients). Patients ranged in age from 2 to 14 years. The diagnosis was made before the age of 3.5 years in 5/6 cases. The sex ratio was 1. Language delay was noted in 5/6 cases. No contributing risk factors were identified. All patients had profound and bilateral hearing loss. Associated malformations were present in 2/6 cases. The most common abnormalities were semicircular canal hypoplasia, internal auditory canal abnormalities and cochlear malformations. There was an association between sensorineural hearing loss linked to malformation and the profound degree of hearing loss.

Conclusion: Sensorineural deafness linked to cochleovestibular malformations in children appears to be uncommon. However, they are associated with a risk of profound sensorineural hearing loss. It is therefore important, in the face of any sensorineural hearing loss in the child, to systematically carry out an imaging assessment, even in our low income's countries.

Keywords: Sensorineural Deafness; Malformations; Cochlea Abnormalities; Children; Côte d'Ivoire

Abbreviations: IEMs: Inner Ear Malformations; CT: Computed Tomography; SNHL: Sensorineural Hearing Loss; MRI: Magnetic Resonance Imaging; LE: Left Ear; RE: Right Ear; SCC: Semicircular Canal; IAC: Internal Auditory Canal; SCCs: Superior Semicircular Canal; EVA: Enlarged Vestibular Aqueduct; PLF: Peri Lymph Fistula.

Introduction

Deafness is one of the most common sensory disabilities in children. It is estimated that one child in a thousand has severe or profound deafness at birth [1]. Most congenital hearing loss (90%) is caused by sensorineural impairments

in the inner ear, cochlear nerve, and/or central auditory pathway [2].

Detectable inner ear malformations (IEMs) and cochlear nerve deficiencies, identifiable with computed tomography (CT scan) and/or magnetic resonance imaging (MRI), are responsible for 15–40% of pediatric sensorineural hearing loss (SNHL) cases [2,3]. The etiology of these malformations is usually unknown [4]. Taking these IEMs into account in the assessment is essential because they present specific challenges in therapeutic management.

The lack of epidemiological data related to cochleovestibular malformations causing sensorineural hearing loss in our context justifies this study. The aim of our study was to find the distribution of various malformations and to report the clinical features of commonly encountered IEMs in our practice.

Methods

The study was conducted in the ENT departments of the Abidjan University Hospitals (CHU), the Dominique Ouattara Mother and Child Hospital in Bingerville and a private medical group in Abidjan. This was a retrospective cross-sectional study. The study took place over a period of 14 months (January 2019 to March 2020).

All patients aged up 15 years, who had a cochleovestibular malformation related to sensorineural hearing loss and/or predominantly sensorineural mixed hearing loss confirmed during the study period were included. Other types of hearing loss and malformations of outer or middle ear were not included. Sampling was non-probability for convenience, with an exhaustive collection of patients followed during the study period and meeting the selection criteria.

Data collection was carried out using the medical records of children, consultation registers and imaging registers.

All studied variables, reported in the documents used, have been listed. The total number of consultants in ENT departments for sensorineural hearing loss and/or mixed sensorineural hearing loss during the study period was recorded.

The variables to be studied were age at diagnosis, gender, ethnicity, level of education. Clinically: the reason for consultation, associated signs (language delay, hearing loss, behavioral problems, balance disorders), risk factors and pathological history (embryo-fetopathies, gestational age less than 34 weeks, congenital malformations of the head and neck, meningitis, neonatal jaundice, consanguineous marriage), clinical data (ENT examination), etiologies of deafness (acquired, congenital), results of objective or subjective audiometry, characteristics of the deafness (side and degree according to the BIAP classification [5]), IEMs detected by imaging (CT scan and MRI of inner ear and brain) were classified according to the side (right, left) and the site (cochlea, vestibule, internal auditory canal).

Data collection and statistical analysis were done in CSPRO VERSION 6 and exported to SPSS 23 for data analysis. All p-values less than 0.05 were considered statistically significant.

Results

Prevalence

During the study period, we identified 160 patients with sensorineural hearing loss, including 6 cases of cochleovestibular malformations, i.e., a prevalence of 3.75%.

Socio-Demographic Data

These are grouped in Table 1.

	Age	Sex	Sibling rank	Ethnic group	Nationality	Schooling
Case 1	3 years	Feminine	2nd	Baoule	Ivorian	Not in school
Case 2	2 years 1 month	Feminine	1st	Mossi	Non-Ivorian	Not in school
Case 3	3 Years 6 Months	Masculine	1st	Abbey	Ivorian	Not in school
Case 4	2 Years 5 Months	Masculine	1st	Senufo	Ivorian	Not in school
Case 5	4 Years 7 Months	Masculine	1st	Senufo	Ivorian	Not in school
Case 6	14 years old	Feminine	5th	Avikam	Ivorian	School

Table 1: Socio-Demographic Data.

Patients ranged in age from 2 to 14 years. The median age was 3.5 years and an interquartile range of 2–7.3 years. The sex ratio was 1. The Akan ethnic group (Baoule, Abbey, Avikam) accounted for half of the cases and the Mande from the north (Mossi, Senufo) the other half.

Clinical Data

Clinical data are summarized in the Table 2.

	Reason for consultation	Age at diagnosis	Related Signs	Pathological history	ENT Physical Examination Data
Case 1	Language Delay	2 years	None	None	Normal
Case 2	Language Delay	1 year	None	None	Bilateral Cervical Fistula+ Right Congenital PFP
Case 3	Language Delay	2 years	None	None	Normal
Case 4	Language Delay	1 year	None	None	Normal
Case 5	Language Delay	2 years	Hearing loss	None	Normal
Case 6	Hearing loss	12 years	None	None	Normal

Table 2: Summary Table of Clinical Data.

The reason for consultation was language delay in 5 out of 6 cases. The age of SNHL diagnosis ranged from 1 to 12 years with a mean age of 2 years. No history has been found in this study. A non-ENT malformation was associated in case 6: it was the persistence of the ductus arteriosus.

Paraclinical Data

Audiometric Assessment: objective audiometry was performed in 5 cases and subjective audiometry in one case.
Degree of Deafness: 5 patients had profound and bilateral deafness (5 / 6 cases), including 6 cases on the right and 5 on the left.
Radiological Assessment: a CT scan of temporal bone was performed in 6 cases and an MRI of the inner ear in 2 cases. Several types of inner ear malformations were identified (Table 3).

	Case 1		Case 2		Case 3		Case 4		Case 5		Case 6		%
	RE	LE	RE	LE	RE	LE	RE	LE	RE	LE	RE	LE	
Oval window (small, rounded structure)	1	1											16,7
Cochlea hypoplasia			1			1							16,7
Common cavity (cystic cavity)				1							1		16,7
Complete labyrinthine aplasia					1								8,3
SCCl hypoplasia							1				1		16,7
SCCs hypoplasia							1	1			1		25
SCCp Hypoplasia								1			1		16,7
Bone dysplasia of the utricle								1					8,3
Modiolus malformation								1			1		16,7
Atretic IAC					1				1	1			25
Vascular-nervous conflict in IAC										1			8,33
SCCs bone capsule dehiscence									1				8,3
Hypoplastic cochlear nerve					1								8,3
Reduction in the number of turns											1		8,3
Enlarged (or dilated) vestibular aqueduct											1		8,3
Enlarged IAC											1		8,3

LE: Left Ear; RE: Right Ear; SCC: Semicircular Canal; IAC: Internal Auditory Canal.
Table 3: Distribution of Malformations According to their Frequency.

Abnormalities and Deafness

We identified 8 vestibular abnormalities, 5 cochlea abnormalities, 4 internal auditory canal abnormalities, and one complete labyrinth abnormality.

There was a statistical association between the degree of sensorineural hearing loss and cochleovestibular malformations (Table 4).

	Degree of Deafness			
	Profound deafness n (%)	**Other level of deafness n (%)**	**OR (CI: 95%)**	**Value P**
Yes	6 (100)	0 (0)	1,9 (1,6 - 2,2)	0,03
No	93 (60,4)	61 (39,6)		

Table 4: Association between the Degree of Ear Hearing Loss and the Presence of Cochleovestibular Malformations.

Discussion

Out of the 160 patients identified for this study, we found 6 patients with cochleovestibular malformations (3.75%). This rate remains below those reported in the literature: 40% for Sarioglu, et al. [6]; 20% for Sennaroglu, et al. [4] and 10% for Adibelli, et al. [7]. A bias might be created by the small sample size in our study. Sensorineural hearing loss of malformative origin does not appear to be related to sex: sex ratio 1 in our study and 0.95 in Masuda, et al. study [8]. The size of the sample does not allow us to conclude that there is a link between ethnic origin and the existence of IEMs. However, the presence of the Akan's group, an indigenous population of Abidjan and its surroundings, would probably be linked to the location of the study. Unlike the group from Northern Mande, for which ethnic origin could be incriminated, due to the frequent existence of consanguineous marriage. According to an observation made in several studies on deafness, the hearing-impaired child is the eldest of the siblings in most cases (46 to 53% of cases) [9,10]. The patients were mostly under 4 years old, so they were not in school.

These SNHL with malformative origin are sometimes syndromic (2 cases in our study). Petit [11] states that 30% of genetic deafness is associated with other symptoms or malformations, therefore called syndromic, when Denoyelle, et al. [12] report that genetic SNHL is syndromic in 10 -15% of cases. To explore deafness, objective audiometry was mainly performed in our study, probably related to the young age of the patients. Some authors note that behavioral audiometry, generally gives reliable answers in children over 4 years old [13]. We noted a preponderance of profound deafness and bilateral deafness (5/6 cases); while Masuda, et al. [8] had identified profound deafness in 46% of cases, bilateral in 49% of cases. Adibelli, et al. [7] found different levels of hearing loss (severe and profound) in their study. The difference in the size of the study populations could explain these results, even though our study seems to link cochleovestibular malformation and the degree of deafness. The risk of having profound deafness is estimated at 1.9.

Once the diagnosis of sensorineural deafness had been made, an imaging assessment was carried out in some patients. This assessment most often consisted of CT scans. This is consistent with data from the literature and in the work done by Er-Raji I [14] especially. According to him, computed tomography should be the first-line examination in the exploration of the ear in children, whatever the type of malformation; MRI is only complementary. Essentially, it is used to look for an abnormality of the cochlear nerve or labyrinthine structures. The proportions of the different abnormalities identified vary from one study to another. In Masuda, et al. [8] report the authors cite in order of frequency, IAC abnormalities, cochlear malformations then the vestibule anomalies. In our study, these are mainly the SCC hypoplasia, then IAC abnormalities and finally, cochlear malformations. The description and classification of these abnormalities also vary. Some studies separate inner ear malformations from third window abnormalities [6]. In this context, some malformations including EVA are classified for certain authors as IEMs [4] and for others, as abnormalities of the third window [6]. These abnormalities of the third window bring together disorders which result in abnormal communications between the inner and the middle ear, as well as cerebrospinal fluid and vascular structures. Superior semicircular canal (SCCs) dehiscence, posterior SCC (SCCp) dehiscence, enlarged vestibular aqueduct (EVA), X-linked stapes gusher, perilymph fistula (PLF), and bone dyscrasias of the temporal bone comprise the third window abnormalities [15].

Three cases are reported in our study: SCCs dehiscence, enlarged vestibular aqueduct, bone dysplasia of the utricle. Different types of hearing loss were considered for third window abnormalities [6].

Conclusion

Sensorineural deafness linked to cochleovestibular malformations in children appears to be uncommon in

our practice, sometimes associated with malformations of other organs. However, they are associated with a risk of profound SNHL, for which the only alternative is cochlear implant surgery. It is therefore important, in the face of any sensorineural hearing loss in the child, to systematically carry out an imaging assessment, including at least a CT scan of the inner ear, even in our countries.

Conflict of Interest: None

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