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Cochlear implantation and vestibular function in children

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ABSTRACT

Objectives: To analyze vestibular function Q2 (VF) after cochlear implantation (CI) in children.
Methods: Retrospective cohort study at a tertiary pediatric referral center. 43 children that had vestibular testing (VT) after unilateral cochlear implantation, from 2001 to 2010, were included. CT scan of the temporal bone was done systematically before surgery. VT included three tests: head-impulse test, caloric tests, vestibular evoked myogenic potentials. VF was graded in: normal (type 1), partial dysfunction (type 2), severe dysfunction and areflexia (type 3). In 12 cases, VT was done before and after CI. Vestibular function was analyzed looking to side, etiology and preoperative status.
Results: Mean age at CI was 2.9 years. Bilateral inner ear malformation were retrieved in 16%. Before surgery, 50% of children had normal vestibular responses, 4/12 had bilateral type 2, two had asymmetrical VF. In this group, after surgery, 2 children had VF worsening, none on the CI side only. Considering all 43 patients, post operative VT showed normal response in 48.8% and type 2&3 in 16.2%. Children had asymmetrical poorer vestibular function on the side of CI in 19%. Among them, 75% had normal contralateral VF. Sensorineural hearing loss etiologies known to be associated with vestibulopathy (Usher/Meningitis/Inner Ear Malformations/CMV) were associated to abnormal vestibular function more frequently than in other causes ($p = 0.01$).
Conclusion: Half of the children had initial vestibular dysfunction. In our study, 20% of cochlear implantation could have worsened vestibular function. As vestibular function should be part of the choice in cochlear implantation, side of implantation and survey; and vestibular tests are uneasy to achieve in pediatric population, we propose a three-steps evaluation and gradation which allow easier comparison.

1. Introduction

In congenitally deaf children, the main goal of rehabilitation is to enable communication and development of oral language. Balance deficits that are frequently associated to deafness, can impact on motor and language development [1–6]. Indeed vestibular dysfunction is an underestimated cause of associated disabilities, such as dyslexia, and should be assessed in all hearing-impaired children [7,8].

Vestibular function is more often tested in adults than in children. The surgical impact of CI on vestibular function has been first reported in children in 2009 [9] and more recently in few other series [10,11]. Comparison between studies on implanted children remains difficult because of the variability of the protocols. This study reports the experience of our department on vestibular results after CI, and aims to define the basic vestibular examination and grading in view to help the CI team as a decision-making tool.

2. Patients and methods

This retrospective study included all children who had received a unilateral CI in our department between 2001 and 2010, and who had undergone a postoperative CI vestibular assessment including both canalar and otolithic testing. All children that had partial testing were excluded. Some children had both pre- and post-operative vestibular assessment. The CI was switched off during all tests. The conditions of the vestibular test were the following.

2.1. Assessment of horizontal SCC function

2.1.1. High frequency canalar test

The head-impulse test (HIT), subjective test of the horizontal semi-circular canal, was done manually. When the head is turned

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to one side in the plan of the semi-circular canal to be tested, the vestibulo-ocular reflex maintains the visual fixation. The breaking of the visual fixation, shown by a refixation saccade, is due to the quick eye movement back to the target and indicates a canalar disorder. Presence of the refixation saccade is pathologic. As the repetition of this movement is not well tolerated, this test requires an experimented operator. This test is possible as soon as the child can hold his head steady.

2.1.2. Low frequency canalar test

The bithermic caloric test was conducted using 30 °C and 44 °C water irrigation to assess the function of the horizontal semicircular canals at low frequencies. As caloric tests are known to be more reliable when using water, we excluded children that had an air-testing condition. The vestibule ocular responses (VOR) were measured through videoscopy. The VOR was analyzed for 30 s, starting at 60 s after the beginning of stimulation. The caloric response was quantified for each ear based on the summation of number of nystagmus of the warm and the cold irrigation response. Relative valence was quantified using the Jonkees formulae: Relative valence = [(right caloric response − left caloric response)/(right caloric response + left caloric response)] * 100. Only absolute values were used.

2.2. Assessment of saccular function

Otolithic-function, which tests vestibulospinal responses, was evaluated with vestibular-evoked myogenic potentials test (VEMP). The VEMP was done with bone stimulation to optimize the myogenic response, and to avoid limiting factors such as transmission and hearing level. The stimulus was a 750Hz-6ms tone burst delivered by bone conduction at a rate of 4.1 Hz through a Radio Ear vibrator [12]. Contralateral head turn was used to activate SCM. Vestibular evoked myogenic potentials were judged as present or absent. A range of acceptable latencies of the PINI waveform were used based on published age-appropriate values [2,13]. Presence of a myogenic potential at 110 dB bone conduction stimulation was considered as normal response.

We classified the VF into 3 grades: normal (type 1), partial dysfunction (type 2), severe dysfunction and areflexia (type 3) (Table 1).

2.3. Inner ear imaging

All patients underwent CT scan before cochlear implantation. Inner ear malformations were reported under: Semi-Circular Canal (SCC) malformation, Dilated Vestibular Aquaduct (DVA), dilated vestibula (DV), cochlea malformation.

VF was analyzed looking to each ear, to the etiology of the hearing loss and the results were compared when possible pre- and post-operatively. Statistical analysis used Chi^2 test.

3. Results

During the study period, 577 children were implanted unilaterally; among them, 43 patients (Table 2) had full post-operative VT criteria and could be included. Twelve also received a full criteria preoperative VT.

In the studied population, gender ratio was 1.1 and mean age at CI was 2.9 years (0.6–15.1). The surgical technique was the same for all patients and was performed by a senior surgeon. All patients had a single-side implant using the standard cochleostomy technique (anterior–inferior to the round window). Forty-two patients received a Cochlear®® (CI24RE/RST) implant and one patient received an Advanced Bionics ® (HiREs). The implant was placed in the right ear in 65.1% of cases (28/43).

Etiologies of deafness were: Connexin gene mutation ($n = 7$), Usher syndrome ($n = 6$), Waardenburg syndrome ($n = 2$), PDS mutation and other inner ear malformation (IEM) ($n = 7$), cytomegalovirus maternal infection (CMV) ($n = 3$), meningitis ($n = 2$), Kallman syndrome ($n = 1$). No etiology was found in 15 cases (Table 2).

Bilateral malformation of the inner was found in 7 patients. Cochlea malformation and DVA were the most frequently reported.

Mean age at VT was 4.3 years (1.2–17.2). Mean delay between CI and post-operative VT was 1.4 years (0.8–10.8).

Twelve children had both pre- and post-operative VT. (Table 3) the mean age at first VT was 1.9 years (0.2–8.9). The mean delay at post-implant VT was 1.4 years (0.1–10.9). Pre-operatively, six had

Table 1
Vestibular function and classification. HIT, head impulse test. VEMP, vestibular-evoked myogenic potentials.

	HIT	Reflectivity	Relative valence	VEMP
Type 1	No catch-up saccade	$R > 30$	RV < 15	Normal response
Type 2	Variable	$30 < R < 10$	–	Variable
Type 3	Catch up saccade	$R < 10$	–	No response

Table 2
Population. CI, cochlear implantation; IEM, inner ear malformation; CMV, cytomegalovirus; VT, vestibular test.

Patient	Age at CI (in years)	Etiology	Side of implantation	Age at VT (in years)	VT
SI.LI	5.2	Unknown–IEM	Right	4.8/8.8	Pre/post
VI.EL	3.2	Unknown–IEM	Right	2.5/4.5	Pre/post
TR.HA	4.3	Unknown	Right	4.2/4.4	Pre/post
AI.ER	2.4	Unknown	Right	2/2.6	Pre/post
BA.BA	9.0	Connexin	Left	8.9/10	Pre/post
BE.AN	2.8	Unknown	Left	0.6/4	Pre/post
GA.NA	0.9	Usher	Right	0.9/1.2	Pre/post
GO.PI	1.2	Waardenburg	Right	1.2/3.6	Pre/post
RA.AD	0.6	Unknown	Right	0.2/1.9	Pre/post
BA.OL	1.9	CMV	Left	1.8/1.9	Pre/post
RA.AL	1.7	Unknown	Right	1.5/2.6	Pre/post
CH.EL	3.5	PDS	Left	3/3.8	Pre/post
MA.AV	2.2	Unknown	Left	9.1	Post
MA.DA	1.7	Waardenburg	Left	2.9	Post
GU.HU	2.4	Connexin	Left	4.9	Post
VO.CE	3.0	Connexin	Right	5.3	Post
HE.MA	2.9	Connexin	Right	4.5	Post
OU.KY	1.4	Connexin	Right	3.6	Post
PO.NI	2.1	Unknown–IEM	Right	5.3	Post
OL.MI	1.0	Unknown	Left	2.3	Post
LE.MA	1.2	Connexin	Left	2.6	Post
HA.LO	4.1	Unknown–IEM	Left	4.4	Post
BO.SA	8.1	Unknown–IEM	Left	8.5	Post
HE.EV	2.9	Kallman	Right	8.0	Post
GO.CE	15.2	Unknown	Right	17.2	Post
LE.MA	3.7	Usher like	Right	7.0	Post
ZA.HU	3.4	Unknown	Right	4.0	Post
CO.AL	1.4	Meningitis	Right	1.5	Post
IN.PI	1.9	Unknown	Right	2.4	Post
KE.LA	2.4	Unknown	Right	3.4	Post
TH.GU	3.1	Usher	Right	4.6	Post
SH.NA	4.1	Unknown	Left	7.4	Post
BA.CE	1.4	Usher like	Right	4.1	Post
GO.LO	1.5	Meningitis	Left	2.3	Post
VO.LU	3.5	Connexin	Left	7.5	Post
GR.RO	3.1	Unknown	Right	4.6	Post
AR.BE	2.9	CMV	Right	10.5	Post
DU.AL	3.6	PDS	Right	7.1	Post
MB.KE	3.7	Unknown	Right	6.5	Post
CO.LE	3.9	CMV	Right	4.1	Post
JA.ZO	3.4	Unknown	Left	4.0	Post
BA.CE	3.6	Usher like	Right	14.5	Post
GO.SO	4.6	Usher	Right	6.1	Post