

Case Report

Cochlear Implant in a Child with Microcephaly for Congenital Zika Virus Syndrome: A Case Report

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Abstract

Background: Congenital Zika syndrome (CZS) is characterized by microcephaly with calcifications between cortical and subcortical zones, and other abnormalities. Like other congenital infections, CZS can cause hearing loss, and the authors have previously demonstrated this association with a 5.8% of prevalence.

Objective: To report a case of cochlear implant in a child with severe to profound hearing loss due to CZS.

Case report: A 2-year-old boy with microcephaly and profound bilateral sensory-neural hearing loss due CZS was submitted to cochlear implantation on his right ear. The surgery was performed by conventional transmastoid and round window insertion with standard electrodes and no intra or postoperative complications occurred. One month after activation he smiles when the device is turned on and pay attention to his mother's voice.

Conclusion: Cochlear implantation could be done in microcephaly patients with standard technique and would probably be a reasonable option to improve hearing in cases of severe to profound hearing loss due to CZS and also benefit children interaction with surrounding environment, collaborating with the neurodevelopment.

INTRODUCTION

Since April 2015, an increasing number of cases of autochthonous infection by the Zika virus have been reported in South America, specially, in Brazil's northeast. By August 2016, almost two thousand cases of microcephaly due to CZS were confirmed throughout the entire country [1].

The totality of CZS's characteristics are still unknown but a recent review delineated a pattern of five features that are commonly found in congenital Zika virus infection such as severe microcephaly with partially collapsed skull; thin cerebral cortices with subcortical calcifications; macular scarring and focal pigmentary retinal mottling; congenital contractures and marked early hypertonia with extra pyramidal involvement [2]. Other reported features are swallowing disorders, seizures, visual impairment and 5.8% risk of sensorineural hearing loss [3-5].

Concerning hearing loss, no study addresses the topic of rehabilitation. This issue is of particular interest because of the multiple impairments of these children. If, on the one hand, the

severe central disorders limit considerably the chances of a good outcome from an auditory rehabilitation, on the other hand, one must consider that any additional sensory input could minimize his (her) handicap and benefit the neural development.

The first years of life area critical period for the maturation of the central auditory system, being considered ideal for the development of auditory and language skills. This fact points to the benefits of early intervention in the first years of life in children with sensory-neural hearing loss [6,7].

Patient information

The patient was born in 2015 in Recife, Pernambuco, Brazil, from a dizygotic twin pregnancy. His mother had an episode of skin rash associated with itching and fever at 1 month of gestation. At the 25th week of pregnancy, microcephaly was diagnosed by obstetric ultrasonography, but no anomaly was detected on the twin brother by this method. Delivery occurred at gestational age of 37 weeks, with the newborn weighting 2100g; the head circumference (HC) was of 28 cm (classified as severe microcephaly according to Fenton curve).

Clinical findings

In additions to reduced head circumference, the patient presented craniofacial disproportion, closed anterior fontanel, exuberant occipital protuberance, redundant scalp skin and right clubfoot. Brain computerized tomography (CT) revealed diffuse bilateral reduction of cerebral parenchyma, ventriculomegaly, cortical underdevelopment, multiple calcifications predominantly in the basal ganglia and cortical/subcortical white matter regions and hypoplasia of the brainstem and cerebellum (Figure 1). Zika virus-specific immunoglobulin M (IgM) capture enzyme-linked immunosorbent assay (ELISA) performed on cerebrospinal fluid was positive. All other viral agents that could cause congenital hearing loss (toxoplasmosis, rubella citomegalovirus, herpes and syphilis) as well as other flavi-virosis (dengue and chikungunya) were tested and excluded. The twin brother was completely normal.

In the first months of life, the child was diagnosed with hearing and visual impairment and submitted to glaucoma surgical correction after the third month of life. By the age of 7 months, he was diagnosed with epilepsy and controlled with valproate.

A severe neurodevelopment delay was obvious, with no interaction with the environment and no head control at 21 months of age. The head circumference at this moment was 37.5 cm (severe microcephaly) and an X-ray of the hips showed left hip dysplasia.

Diagnostic assessment

Audiological evaluation with transient otoacoustic emissions (TOEA) and auditory brainstem response (ABR) to click stimuli

at 35 decibels normal hearing level (dB nHL) were performed in the first month of life with no response in any of them. Further, sensory-neural hearing loss was confirmed by frequency-specific ABR, with tone bursts, at frequencies 500 and 2.000 Hz and revealed a response only at 85 dB nHL for 2.000Hz in the right ear. Behavioral auditory evaluation was done using musical instruments (complex sounds) of known frequency range at 60cm from the pinna, with no response even for high intensity stimulus. Cochleo-palpebral reflex was absent and there was no response to voice stimulus at 100dB in free field audiometric stimulation (inside of an acoustical booth).

Brain magnetic resonance image (MRI) at this point revealed diffuse bilateral reduction of cerebral parenchyma, increased ventriculomegaly compared with the previous exam, malformation of cortical development with simplification of gyros pattern and polymicrogyria hypoplasia of the brainstem and cerebellum (Figure 2).

herapeutic intervention

After an unsuccessful attempt of rehabilitation with hearing aids and speech therapy, a cochlear implantation was performed at 24 months of age in the right ear with standard electrodes inserted trough the round window without any complications. Intra-operative neurotelemetry demonstrated a normal impedance for all electrodes and good neural response for the basal and midturn's electrodes.

Follow-up and outcomes

Device's activation showed immediate response of attention to the sound. One month after activation he smiles when the device is turned on and pays attention to his mother's voice.

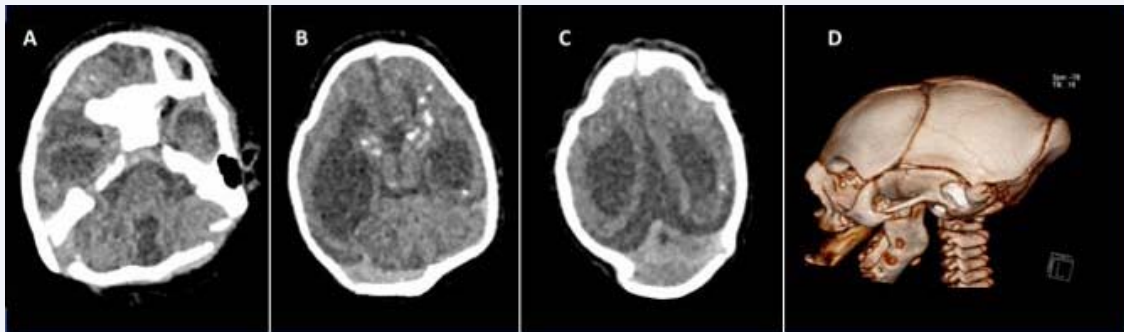


Figure 1 Cranial imaging evaluation revealed the typical appearance of CZS.

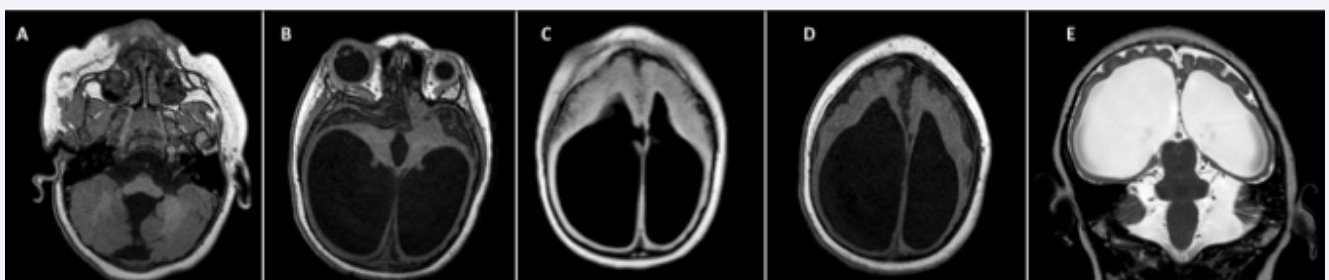


Figure 2 Zika virus-specific immunoglobulin M (IgM) capture enzyme-linked immunosorbent assay.

He's now able to detect Ling's sounds at 50dB in open field and shows more self-confidence and is able to stay without maternal physical contact.

DISCUSSION

This case evokes the dilemma of auditory rehabilitation in children with multiple deficiencies, including central nervous system anomalies. As in other examples of congenital neurological diseases, there is a great chance of a poor outcome from cochlear implantation in these cases, because of the limitations of auditory signal processing due to the encephalic involvement. However, the decision must take into account the fact that a neurologically handicapped child should benefit from any additional sensorial input that one could offer him or her. Several studies have demonstrated that, although it is quite difficult to achieve a gold standard outcome, cochlear implant can allow these patients to improve their quality of life, self-confidence, independence and social integration [8-10].

CZS cases represented a peculiar challenge not only because of the severity of the neural damage but also because its pathology is still far from being completely elucidated. So, until now, it was uncertain if auditory rehabilitation would be of significant help for these children or not.

We presented the case of a child with severe microcephaly secondary to CZS who, in addition to important ocular alterations of the posterior eye compartment and surgically corrected congenital glaucoma, presented profound bilateral sensory-neural hearing loss, severely limiting the infant's contact with the environment and making his neuropsychomotor rehabilitation even more difficult. In this way, we perceive the need to devise strategies to optimize social interaction and improve quality of life in cases where neurological impairment is associated with incapacitating handicaps.

As there was no response to acoustic amplification by hearing aids, cochlear implantation was performed at two years of age, respecting the best window of neuronal plasticity. The choice for unilateral rather than bilateral implantation was based on the fact that the child had severe encephalic lesions that need to be periodically followed by imaging exams, and two devices on a small skull could jeopardize this evaluation even in CT's due to artifacts. Moreover, the perspective of a limited outcome and the uncertainty about the feasibility of the procedure led us to choose to implant only one side. The usual transmastoid technique was performed without complications, demonstrating that, despite cranial malformations, it is a feasible procedure. The benefits

could be noticed already at the moment of activation, when the child reacted readily to sound stimulus. And after one month, one could notice a progressive increasing in sound perception and a remarkable behavioral improvement.

CONCLUSION

Cochlear implantation could be done in microcephaly patients with standard technique and would probably be a reasonable option to improve hearing in cases of severe to profound hearing loss due to CZS and also benefit children interaction with surrounding environment, collaborating with the neurodevelopment.

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