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Short Communication

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Hearing loss associated with patent ductus arteriosus

Abstract

The Ductus Arteriosus is a connection between the aorta and pulmonary artery. The essential foetal structure closes instantly after the birth. If the ductus arteriosus fails to close after the birth it is considered to be an abnormal. To date, there are very few studies which have reported hearing loss as an associated feature of Patent Ductus Arteriosus. Disruption of blood supply to the auditory system is reported to be a possible cause for hearing loss in children with Patent Ductus Arteriosus. Autosomal recessive inheritance has also been found to play a vital role in the genetic predisposition of Patent Ductus Arteriosus and hearing loss. Here in we report a 3year old female child with a confirmed diagnosis of Patent Ductus Arteriosus associated with gross developmental delay, hearing loss and bilateral cataract. *"Hearing loss thus seems to co-exist in children with Patent Ductus Arteriosus which in turn warrants detailed audiological assessment in every patient with Patent Ductus Arteriosus to facilitate development of regular speech and language development"*

Keywords: audiological assessment, congenital heart disease, hearing loss, patent ductus arteriosus, behavioural observation audiometry, carpenter's syndrome, holt-oram syndrome, echocardiography, aids, cytomegalovirus, toxoplasmosis, congenital rubella

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"What is known"

Hearing loss as an associated feature in cases with PDA has been reported earlier by Dahl & Boesen,[1] but the cause for the same has been correlated to maternal rubella in their subjects. However, there is no detailed information accessible on the methodology and reasoning for the findings obtained by the investigators.

"What is new"

Detailed investigation with a battery of audiological tests is potential enough to delineate presence of hearing loss in cases with PDA, thereby facilitating early identification and intervention in these patients. Also, the subject concerned did not present with any history of maternal rubella as reported in literature.

Introduction

The Ductus Arteriosus is a cardinal for foetal vascular circulation which closes immediately after birth and consistently within 48hours of birth. It is considered abnormal if it remains open till 3months of age.[2] The clinical sign and symptoms of PDA depend primarily on the size of shunt and the basic cardiovascular condition of the patient. The prevalence of PDA accounts 5 to 10% of all inborn heart diseases and believed to be 1 in 2000 births.[3]

The components accountable for firm patency of ductus arteriosus after few hours of new-borns life are still implicit. The prevalence of PDA increased with physiological factor related to prematurity instead hereditary anomaly of the ductus.[4] In full term neonates, PDA occurs occasionally. Genetic syndromes may also cause PDA. The most common genetic syndromes, which may cause PDA are chromosomal abnormalities (i.e., 4p syndrome and trisomy 21), X-linked mutations (i.e., incontinentia pigmenti), single-gene mutations (i.e., Holt-Oram syndrome and Carpenter's syndrome), and autosomal recessive inheritance with incomplete penetrance.[5] The definite causes through which these genetic anomalies bring about persistency of PDA are not clear. However, Satoda et al.,[6] recommend that the anomaly in Char syndrome associate with PDA, facial dismorphism, and upper extremity malformations is due to derangement of neural crest cell derivatives.[6]

The incidence of PDA highly increases due to infection of rubella virus in first trimester of pregnancy especially in first month.[7] Anoop et al.,[8] noted that environmental factors, such as fetal valproate syndrome may also cause PDA,[8] but the mechanism has not been cleared. This case report aims to show case such a patient presenting with PDA associated with asymmetrical hearing loss.

Case report

A 3year old female child with confirmed diagnosis of PDA was referred from an advanced pediatric center to an audiology and speech rehabilitation unit, Department of Otolaryngology and Head & Neck Surgery, PGIMER, Chandigarh with a chief complaint of inadequate speech & language and gross developmental delay. Other observation included a bilateral cataract operated on at the age of 5months. Cardiovascular examination, conducted at Advanced Cardiac Centre revealed that the child had a normal S1 and S2, but that a clinically holosystolic murmur of IV grade intensity was audible, indicating that the child had a PDA. The Echocardiography showed a tiny PDA of 1mm with mild pulmonary stenosis of 20mm Hg. The chest X-ray explains the cardiomegaly and fullness of pulmonary bay. Systemic examination of central nervous system, abdominal system and renal system did not reveal any abnormality. A complete case history of the child was taken with mother as the informant. Prenatally, there was no history of any maternal infections including-AIDS, Cytomegalovirus, Toxoplasmosis, Congenital Rubella, and Herpes I and II. Perinatal history reported Full Term Normal Vaginal Delivery. There was no history of respiratory distress or anoxia. Postnatally, there was no history of neonatal jaundice, pneumonia, hyperbilirubinemia, high grade fever, ototoxic medications, bacterial meningitis, craniofacial anomalies (including pinna and ear canal), family history of congenital

childhood sensorineural hearing loss, stigmata or any history of syndrome which is known to be associated with a sensorineural and/or conductive hearing loss. The child was subjected to both subjective and objective audiological investigations. Subjective assessment included behavioural observational audiometry (BOA).

Results

Behavioral Observation Audiology (BOA) was done using noisemakers of different frequencies and intensities. No responses were observed at loud intensity stimuli for both low and high frequencies. Auropalpebral reflex was observed for a loud drum beat. This was followed by Impedance audiometry. Tympanometry showed bilateral type 'A' Tympanograms. Both ipsilateral, as well as contralateral reflexes, were absent on 500Hz, 1000Hz, 2000Hz and 4000Hz. The child was then referred for Otoacoustic Emissions (OAE). Both Transient Otoacoustic Emissions and Distortion Product Otoacoustic Emission were done and found to be absent. Auditory brainstem response audiometry findings showed Wave V at 70dB nHL in the right ear. No wave V was observed in the left ear until 99 dB nHL. This led to the diagnosis of moderately severe hearing loss in the right ear, and profound hearing loss in the left ear.

Discussion

Although early identification is known to be vital for the development of normal speech and language development in children, early hearing identification and intervention (EHDI) programs are legislatively mandated in only half of the European countries. These programs are implemented in 60% of European countries. In the United States, this program covers 98% of newborn babies. In the United Kingdom, 99.8% screened for hearing.[9] However in South-East Asia, there is no organized attempt to start newborn and infant hearing screening programs. PDA is a prevailing.

PDA is a prevailing dilemma occurs in premature neonates and mostly associated with necrotizing enterocolitis chronic lung disease, and intraventricular hemorrhage. Ductus arteriosus is a vital vascular channel between the aorta and pulmonary artery in a developing fetus. The pressure of a pulmonary artery at birth is high and there is restricted diversion from the aorta to the pulmonary arteries via the ductus arteriosus. During first few hours to days of life, a decline of pulmonary artery pressures takes place because systemic circulation is changed to the pulmonary circulation. This changes results in reduction of blood flow towards the skin, muscles, kidneys, gastrointestinal tract and the cochlea. The pulmonary circulation due to the left to right shunting across the PDA can also cause pulmonary edema and deteriorate the adjusting respiratory status of the infant. Hence the presence of PDA lays negative effect on cerebral perfusion and lessens the oxygen supply to the brain.[10] This can lead to significant damage to the brain which might have led to gross developmental delay and hearing loss.

Genetic predisposition is found to be the most important key factor in PDA. Despite most cases of PDA would seem to occur intermittently, multifactorial inheritance is considered to determine many cases. Nora JJ[10] speculate that the environmental factor trigger the genetic predisposition during vulnerable time of pregnancy and results PDA.[10] The concept behind how these genetic abnormalities lead to PDA is still not clear yet. It can thus be predicted that the genetic causes which might have led to PDA might also have resulted in hearing loss.

Hearing loss thus seems to be present as an associated feature with PDA, which could be pertaining to reduced blood supply to the auditory system. Also, the genetic predispositions for both PDA and Hearing loss might be the same, which could be stated as a reason for association of PDA with hearing loss. Thus, all the neonates who have been diagnosed with PDA should be referred for a complete audiological assessment, to verify for risk factor for sensorineural hearing loss in order to facilitate early diagnosis and intervention. If the diagnosis of sensorineural hearing loss is delayed in children, the critical period to develop normal speech perception and production get lost. Hence this case report highlights the importance of early audiological screening universally.

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Conflicts of interest

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