International Journal of Medical and Health Research

Volume: 1, Issue: 2, 30-31
Sep 2015
www.medicalsjournals.com
ISSN: 2454-9142

Prachi Naragude
Senior residents, Department of Pediatrics, Grant Government Medical College and JJ Hospital Mumbai, Maharashtra, India.

Md Ashfaque Tinmaswala
Senior residents, Department of Pediatrics, Grant Government Medical College and JJ Hospital Mumbai, Maharashtra, India.

Sushant Mane
Assistant Professor Department of Pediatrics, Grant Government Medical College and JJ Hospital Mumbai, Maharashtra, India.

Amin Kaba
Assistant Professor Department of Pediatrics, Grant Government Medical College and JJ Hospital Mumbai, Maharashtra, India.

Anurag chandel
Senior residents, Department of Pediatrics, Grant Government Medical College and JJ Hospital Mumbai, Maharashtra, India.

Lekhraj Talmale
Senior residents, Department of Pediatrics, Grant Government Medical College and JJ Hospital Mumbai, Maharashtra, India.

Correspondence
Md Ashfaque Tinmaswala
Senior residents, Department of Pediatrics, Grant Government Medical College and JJ Hospital Mumbai, Maharashtra, India.

Chiari Network causing central Cyanosis in newborn

Prachi Naragude, Md Ashfaque Tinmaswala, Sushant Mane, Amin Kaba, Anurag chandel, Lekhraj Talmale

Abstract
The Chiari network is mobile redundant structure sometimes seen in right atrium near the opening of inferior vena cava and coronary sinus. Usually it is asymptomatic and of no clinical significance. However in some cases it may present with signs and symptoms of right to left shunt like cyanosis, arrhythmias, endocarditis and thromboembolism. Here we present case of a neonate born to mother with type II diabetes and hypothyroidism. On day 2 of life baby presented with central cyanosis. A 2D Echo was done which showed presence of chiari network along with Atrial Septal Defect (ASD). She was treated with supportive care and was discharged with an advice to remain in regular follow up.

Keywords: Chiari Network, Left to right shunt, Central cyanosis

1. Introduction
In Fetal life approximately 50% of the umbilical venous blood enters the hepatic circulation, whereas the rest bypasses the liver and joins the inferior vena cava via the ductus venosus, where it partially mixes with poorly oxygenated inferior vena cava blood derived from the lower part of the fetal body. This combined lower body plus umbilical venous blood flow enters the right atrium and is preferentially directed by a flap of tissue at the right atrial–inferior vena caval junction, the Eustachian valve, across the foramen ovale to the left atrium [1]. After birth when foramen ovale closes this Eustachian valve does not have any specific function and is no longer needed hence this valve regresses by 9-15 weeks. Incomplete or nonregression of this Eustachian valve results in the condition termed as chiari network. This condition was named after an Australian pathologist named Hans Chiari who in 1897 first described it [2]. Chiari network sometimes may be associated with other congenital structural heart diseases like right sided valvular stenosis or atresia [3]. Majority of the patients with chiari network remain asymptomatic [4]. But rarely the patients may present with symptoms like cyanosis [5]. Here in this case report we describe a neonate born to diabetic and hypothyroid mother. Baby presented with central cyanosis and was later diagnosed to be having chiari network.

Case Report
A full term Female child delivered by normal delivery born to mother with type II diabetes and hypothyroidism. Mother was on daily insulin and thyroid replacement therapy. Baby didn't cry immediately after birth and was given bag and mask ventilation for 30 seconds. The baby was admitted in NICU. On admission the baby was haemodynamically stable. Respiratory rate was 42/min. Heart rate was 136/min. Peripheral pulses were well felt. Routine investigations like CBC, Sr calcium and Blood glucose levels were within normal limits. Chest X ray was within normal limits. Sepsis screen was negative. Baby was started on 1 hourly breast feeding and blood sugar levels were monitored according to NICU protocol. On Day2 of admission the baby suddenly developed central cyanosis. Cyanosis has not responded to oxygen administration. In view of cyanosis a 2D Echo was done which showed " Excessive Redundant Eustachian valve" 2D echo also confirmed presence of other structural congenital heart disease in the form of 2mm Ostium Secundum ASD. This chiari network was causing obstruction to the flow of blood from right atrium to right ventricle and consequently was responsible for right to left shunt across ASD. Baby was given supportive care in the form of thermo neutral environment and supplemental oxygen while nasogastric feeding was continued. Later baby was again shifted to breastfeeding and eventually was discharged. As Chiari network is known to regress on its own a regular follow up was adviced.

Discussion

Central cyanosis in a newborn is usually due to cyanotic congenital heart disease like Tetrology of Fallot, Transposition of great arteries and total anomalous pulmonary venous connections. But rarely a redundant Eustachian valve may be responsible for these symptoms. In fetal life, blood from inferior vena cava is directed towards foramen ovale by presence of Eustachian valve which usually is present in the superior portion of right sinus venosus. This valve regresses up to around 15 weeks of gestation. Incomplete regression of eustachian network results in a persistent or prominent eustachian valve. This usually appears as a thin ridge or a crescent shaped fold of endocardium arising from the anterior portion of rim of the inferior vena cava orifice. The incidence of chiari network is different according to different authors. Schneider B *et al* conducted transesophegeal echocardiography of 1436 Patients and reported the prevalence of chiari network to be 2 %. [6] Patients with chiari network are usually asymptomatic but may present with cyanosis. Mi Akcaboy [7] and Ho-Shun Ko [8] described patients of chiari network presenting with cyanosis. There is no reported case of chiari malformation in case of maternal diabetes. The diagnosis of chiari malformation is usually by 2 D Echo. With advances in diagnostic modalities cardiac MRI is being frequently used to diagnose this pathology [9]. Antenatal diagnosis of this condition, though extremely difficult, is possible [10]. The clinical features are expected to resolve as chiari network is known to regress on its own and rarely needs any treatment. Very rarely surgical intervention may be needed depending upon the nature of abnormality and severity of symptoms. Trento *et al* [11] described patients in whom surgeries were performed because of significant obstruction to the blood flow.

Conclusion

Though the most common cause of central cyanosis in newborn is structural congenital heart diseases like Tetrology of Fallot, Transposition of great arteries and total anamolous pulmonary venous connection etc the possibility of Chiari network must be taken into account when baby presents with unexplained post natal cyanosis. Also it must be kept in mind that this pathology is known to regress on its own and hence in stable babies doesn't need any intervention. Antenatal diagnosis may aid in counseling of parents and allaying the fears associated with congenital heart diseases. As there is no previous reported case of chiari malformation in babies of diabetic or hypothyroid mother it is difficult to comment on the role these diseases might have played in the pathogenesis of chiari malformation in this case.

Conflict of Interest: None

References

1. Daniel Bernstein the Fetal to Neonatal Circulatory Transition In: Kliegman RM, Behrman RE, Jenson HB, editors.Nelson Textbook of Paediatrics. 19th ed. Philadelphia: Saunders, 2011, e415-2
2. Chiari H. Über Netzbildungen im rechten Vorhof des Herzens. Beitr Pathol Anat. 1897; 22:1-10.
3. Trento A, Zuberbuhler JR, Anderson RH, Park SC, Siewers RD. Divided right atrium (prominence of the eustachian and thebesian valves) J Thorac Cardiovasc Surg. 1988; 96:457-463
4. Lanzarini L, Lucca E. Persistence of the right sinus venosus resulting in an unusually prominent Chiari network remnant mimicking cor triatriatum dexter. Pediatr Cardiol. 2002; 23:103-105
5. Ko HS, Chen MR, Lin YC. A huge Chiari network presenting with persistent cyanosis in a neonate. Pediatr Cardiol. 2011; 32(2):239-40.
6. Schneider B, Hofmann T, Justen MH, Meinertz T. Chiari's network: normal anatomic variant or risk factor for arterial embolic events? J Am CollCardiol. 1995; 26(1):203-10.
7. Akçaboy MI, Ekici F, Tutar E. Unusually redundant Chiari network. Pediatr Cardiol. 2006; 27(4):525-6
8. Ko HS, Chen MR, Lin YC. A huge Chiari network presenting with persistent cyanosis in a neonate. Pediatr Cardiol. 2011; 32(2):239-40
9. Altbach MI, Squire SW, Kudithipudi V, Castellano L, Sorrell VL. Cardiac MRI is Complementary to Echocardiography in the Assessment of Cardiac Masses. Echocardiography, 2007; 24:286-300.
10. Bendadi F, van Tijn DA, Pistorius L, Freund MW. Chiari's network as a cause of fetal and neonatal pathology. Pediatr Cardiol. 2012; 33(1):188-91
11. Trento A, Zuberbuhler JR, Anderson RH, Park SC, Siewers RD. Divided right atrium (prominence of the eustachian and thebesian valves) J Thorac Cardiovasc Surg. 1988; 96:457-463.