

Chiari Network: Not Always a Harmless Structure an Unusual Case of Cyanosis in a Neonate and Review of the Literature

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1. Abstract

Chiari's network is an embryological vestige of the right valve of the sinus venosus. It is described as a fenestrated membranous or reticular net-like structure occasionally present in the right atrium, near the opening of the inferior vena cava and coronary sinus. While often considered a normal anatomical variant and clinically insignificant, it might cause cardiological manifestations in infants and adults. We present the case of a newborn with a Chiari network causing persistent cyanosis due to an interatrial right-to-left shunting through the patent foramen ovale, followed later by supraventricular tachycardia.

2. Introduction

Chiari's network was initially described in the literature by von Rokitsky in 1875. Yet earned its name from the pathologist Hans Chiari, due to his publication in 1897, exposing an autopsy series [1]. It is an embryological vestige of the right valve of the sinus venosus and is described as a reticulated network of fibers originating from the Eustachian valve or Thebesian valve, connecting different parts of the right atrium [2].

The estimated incidence of the Chiari network varies between 1.3 to 4% in post-mortem findings and 0.3 to 9.5% with the use of transthoracic echocardiography. The majority of medical textbooks mention a prevalence of 2% [1].

The Chiari network is in most cases asymptomatic and incidentally revealed by cardiac imaging performed for other reasons. Considered thereby as a normal anatomical variant. However, it can be rarely associated with a patent foramen ovale (in approxi-

mately 83% of patients), intra-auricular thrombus, thromboembolic events, formation of atrial septal aneurysms, infective endocarditis, supraventricular arrhythmias, and catheter entrapment. Also, in cases of severe ventricular inflow obstruction, congenital heart diseases may be associated, such as tricuspid atresia, pulmonary atresia, right ventricular hypoplasia, and right ventricular bipartite [3,4].

Management varies from close monitoring to surgical resection, depending on the severity of the right ventricular filling obstruction, the importance of the symptoms, and the associated cardiac anomalies.

In this report, we describe and discuss an unusual case of neonatal cyanosis and supraventricular tachycardia due to a prominent Chiari network, drawing the practitioner's attention to this uncommon cause.

3. Case Report

A 1-day-old male neonate was transferred to our department for the management of refractory hypoxemia.

He was born at term weighing 3960g with a good Apgar score, by spontaneous vaginal delivery from a healthy 27-year-old mother. The pregnancy was uneventful; however, a 34-hour premature rupture of the membranes and a stained amniotic fluid at birth were noticed, for which prophylactic antibiotic therapy per partum was given.

The newborn presented postnatally an expiratory grunting without signs of respiratory distress and cyanosis with low oxygen saturation (SaO₂) at 80% that did not improve with oxygen supplement-

tation. The chest x-ray and blood gas returned to normal. Empirical intravenous antibiotic therapy was started.

On physical examination on the first day of life, we noticed hemodynamic stability and absence of signs of respiratory distress with SaO₂ around 80 to 85%. Heart sounds were well perceived without murmurs and there were no signs of heart failure. The electrocardiogram study showed a right atrial enlargement with no other abnormalities.

Transthoracic echocardiography showed no transposition of the great arteries, total anomalous pulmonary venous return, or right outflow obstruction. Instead, it showed a thin, mobile and echogenic structure in the right atrium moving freely and protruding through the tricuspid valve during diastole (Figure 1; Video clip 1), which caused an intermittent and moderate right ventricular inflow obstruction in late diastole (Figure 2; Video clip 2) and a right-to-left atrial shunting across the patent foramen ovale (Figure 3; Video Clip 3). This latter corresponds to a prominent Chiari network. The sizes of the tricuspid, pulmonary valve, and right ventricle were normal.

After a few days of close monitoring only, we have noticed a clear improvement: SaO₂ (90 %). Control ultrasound showed improvement in the right ventricular filling and the initial right-to-left atrial shunting became bidirectional.

On day 10 after birth, the newborn presented an episode of supraventricular tachycardia which lasted for one hour and disappeared spontaneously. Echocardiographic findings have not changed. Therefore, he was treated with Amiodarone and was discharged home in stable condition while continuing a close follow-up.

The newborn was seen again two weeks later at our clinic. He was asymptomatic and the echocardiogram showed the persistence of the Chiari network, intermittently obstructing the right ventricular inflow; however, the ventricular filling pressures were low. We also observed the persistence of minimal interatrial shunting, but this time, predominantly left-to-right.

We decided to continue to follow up in consultation and to maintain the medical treatment with Amiodarone.

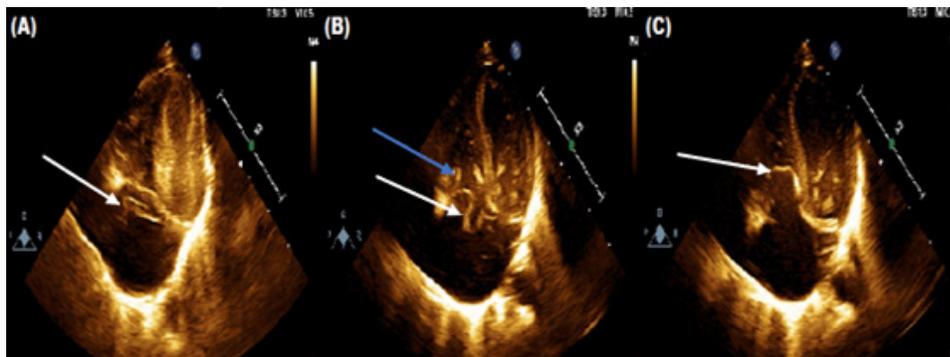


Figure 1: Apical four-chamber view of transthoracic two-dimensional echocardiography showing the position of the Chiari network (White arrows) during the cardiac cycle.

(A) Chiari network in the right atrium during systole. (B) Chiari network near the tricuspid valve (Blue arrow) in mid-diastole. (C) Chiari network in the right ventricle, protruding through the tricuspid valve in late-diastole.

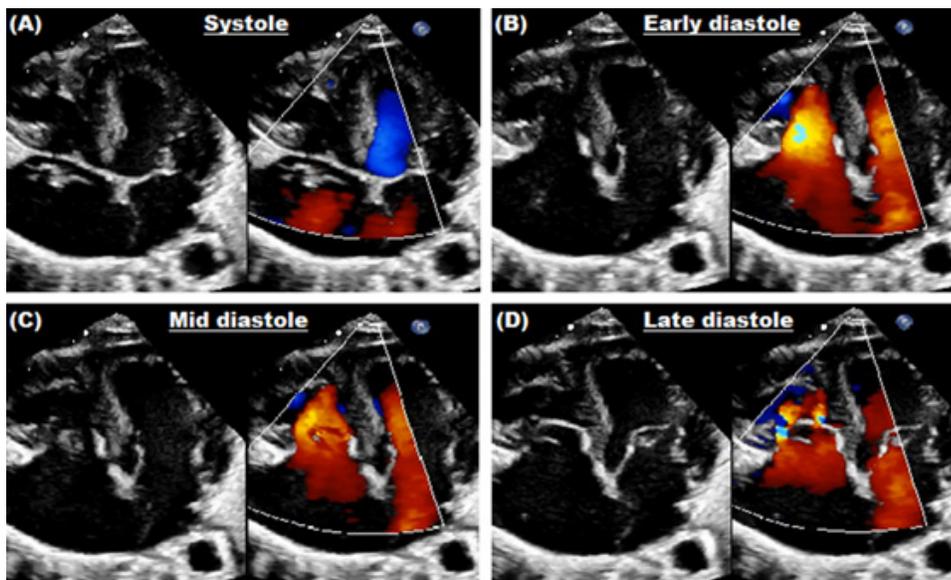


Figure 2: Apical four-chamber view of transthoracic two-dimensional echocardiography with color Doppler showing the Chiari network causing moderate obstruction in the late diastolic phase.

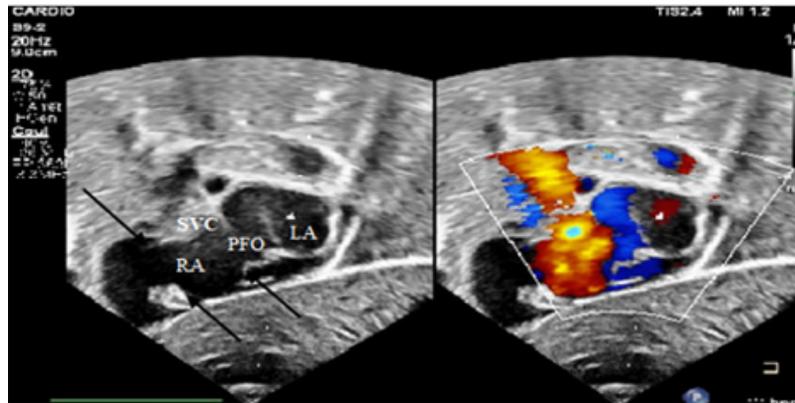


Figure 3: Subcostal view of transthoracic two-dimensional echocardiography with color Doppler showing the different attachment sites of the Chiari network in the right atrium (Black arrows) and the interatrial right-to-left shunting across the patent foramen ovale. RA right atrium. LA left atrium. PFO patent foramen ovale. SVC superior vena cava.

4. Discussion and Literature Review

The cardiac origin of the cyanosis was suspected by the absence of other clinical conditions with a negative hyperoxia test [5]. In such situations, the usual early postnatal cyanotic congenital heart diseases are suspected, namely transposition of the great vessels, abnormal total blocked pulmonary venous return, hypoplasia of the left heart with intact interatrial septum, tetralogy of Fallot as well as the obstacles of the right heart: tricuspid atresia, pulmonary stenosis and atresia and Ebstein's anomaly of the tricuspid valve [6].

The echocardiography findings ruled out all these usual causes and demonstrated a prominent Chiari network hindering atrial systole and causing a right-to-left shunting via a patent foramen ovale, explaining the mechanism of the cyanosis.

In the early embryonic period, the right valve of the right horn systemic venous sinus nearly divides the right atrium into two separate chambers [1]. The persistence or the incomplete involution of this valve instead of its natural regression, which must take place between the 9th and 15th week of gestation, leads in the simplest form, to a prominent or giant Eustachian valve. Chiari network represents a more incomplete involution. Divided right atrium is no more than the more severe form with no or minimal involution of tissues of the venous valves. In the most severe cases, the tissue forms a windsock spinnaker-like membrane across the tricuspid valve which causes severe right ventricular outflow tract obstruction [7]. Moral et al. (2016) have proposed interesting tools allowing the distinction between the different vestiges of the right valve of the venous sinus by describing the anatomical and radiological aspects of each of them [2].

A reported case (Aypar et al., 2013) has described the detection of the Chiari network in a neonate with a heart murmur and another (Aljemali et al., 2020) with hypoxemia without cyanosis [8,9], both of which had no apparent symptoms, emphasizing the importance of postnatal screening such as the systematic examination of the newborn and the pulse oximetry screening in the maternity ward.

Bendadi et al. (2014) reported two interesting symptomatic prominent Chiari network cases, demonstrating different clinical consequences of the Chiari network, both prenatally and postnatally. One case of fetal hydrops, and a second of postnatal cyanosis with abnormal cardiac auscultation [10].

The Chiari network has been correlated with supraventricular tachycardia. Indeed, the cardiac muscle strands in the Chiari network have the potential to promote the alteration of heart rhythm and the occurrence of arrhythmias [3]. Clements et al. (1982) published a case of persistent fetal arrhythmia after birth due to a Chiari network that disappeared after the surgical resection of the network, underlining the possibility of antenatal diagnosis and recommending ultrasound study in the evaluation of fetal cardiac arrhythmias [11]. Chesi et al. (1989) also reported the detection of supraventricular tachycardia in five of the ten patients who had the Chiari network without other cardiac pathologies [12].

In a recent study, Irdem et al. (2020) analyzed P-wave differences on electrocardiograms to investigate changes in atrial conduction in 71 children with a Chiari network and no other heart problems compared to a control group of 60 healthy children. This study showed that children with Chiari may be at risk of atrial arrhythmia [13].

Echocardiography remains the most useful mean in the diagnosis of the Chiari network with a described superiority of transesophageal ultrasound over the transthoracic [2]. The Chiari network appears as a free-floating, curvilinear structure, highly reflective echo target, and highly mobile that waves with blood flow in the right atrium [2,14]. It may be confused for tricuspid vegetation, flail tricuspid valve, free right atrial thrombus, or pedunculated tumours. Careful tracing to identify its attachment to the orifice of the inferior vena cava makes the differential diagnosis [12,14].

The curative treatment of choice for the Chiari network is surgical resection. However, its indications remain rare and reserved only for cases with complete and severe ventricular inflow obstruction resulting in the underdevelopment of right heart structures, cases

of persistent cyanosis, and atrial arrhythmias unresponsive to medical treatment. It has been demonstrated that the natural evolution of the Chiari network tends towards regression, and the surgical choice from the outset is not necessary. Nevertheless, regular monitoring of these patients remains mandatory [2,7,15]

Percutaneous catheterization can also be proposed as a therapeutic choice but remains difficult to perform because of the risk related to the entrapment of the catheter with the network [2].

Based on all these data, we opted only for medical treatment for his arrhythmia which we managed to control, and we planned to follow up on the newborn clinically and sonographically in our consultation.

5. Conclusion

Despite the fact that the presence of the Chiari network is a very rare condition and the consideration of its possible existence as being a normal anatomical variant, it can make a differential diagnosis of cardiac cyanosis and also leads to other serious clinical manifestations and complications.

The diagnosis of this condition can be assured by echocardiography which is considered the tool of choice.

The usual evolution tends towards the regression of the network and the disappearance of clinical signs, requiring only close monitoring. Nevertheless, surgical treatment is sometimes necessary in case of the persistence of the clinical signs.

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