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Case Report
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Congenital Complete Heart Block in Congenitally Corrected Transposition of the Great Arteries

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Abstract

Background: Congenital complete Heart Block (CHB) in patient with Congenitally Corrected Transposition of the Great Arteries (CCTGA) is extremely rare.

Case Summary: A premature 2-days-old baby was referred with hypotension and bradycardia detected in utero. Electrocardiograph showed complete heart block with junctional escape rhythm of 71/min and features of ventricular inversion. Cardiomegaly was seen on chest X-ray and the laboratory findings suggesting sepsis with hyperbilirubinemia. Two-dimensional echocardiography revealed CCTGA. Transvenous Temporary Pacemaker (TPM) was inserted immediately in the Right Ventricle (RV) endocardium, followed by VVI pacemaker implantation at the RV epicardium. The patient was discharged in full recovery.

Discussion: A planned antenatal monitoring, delivery and early referral can lead to favorable outcomes, however, a lifelong follow up for RV function worsening, progressive tricuspid regurgitation, and epicardial pacemaker side effects is important, this might be a problem for patients who live in countries with limited resources.

Keywords: Complete heart block; Congenitally corrected transposition of great arteries; Neonate; Epicardial pacemaker

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Introduction

Complete Heart Block (CHB) is classified as congenital if diagnosed in utero, at birth, or within the first month of life, it can be transient or permanent, the estimated prevalence is 1 per 15,000 to 20,000 live births. Congenital CHB (CCHB) in normal hearts mostly caused by lupus, other causes of CHB include myocarditis and genetic disorders, this group of patients hold an increased risk of late-onset cardiomyopathy. Only a small percentage of CCHB cause by Congenital Heart Disease (CHD), the most common are Congenitally Corrected Transposition of the Great Arteries (CCTGA), Atrioventricular (AV) discordance or heterotaxy syndrome with AV septal defect [1,2]. CCTGA is characterized by the combination of discordant atrioventricular and ventriculoarterial connections [3]. The incidence is around 1 per 33,000 live births, accounting for approximately 0.05% of CHD [4]. We present an extremely rare case of CCHB associated with CCTGA who was successfully managed despite the limitations of resources in developing country like Indonesia.

Case Presentation

A 2-days-old premature baby was referred from a rural area due to bradycardia. The mother is a thirty-two years old lady, who has a bad history of previous pregnancies. Her eldest child died at the age of eight days with history of heart murmur without any further follow up, her second pregnancy ended with spontaneous abortion. Because of these reasons, she went to an obstetrician clinic for prenatal care. Toxoplasma infection was detected and Spiramycin was given until 20 weeks of gestation; she has no other chronic diseases. At 33 weeks of gestation, the obstetrician detected fetal HR of 90 bpm, but the fetus was growing well. The baby was delivered by section cesarean at 36 weeks of gestation due to threatening single-nuchal cord, with APGAR score 8/9 and birth weight

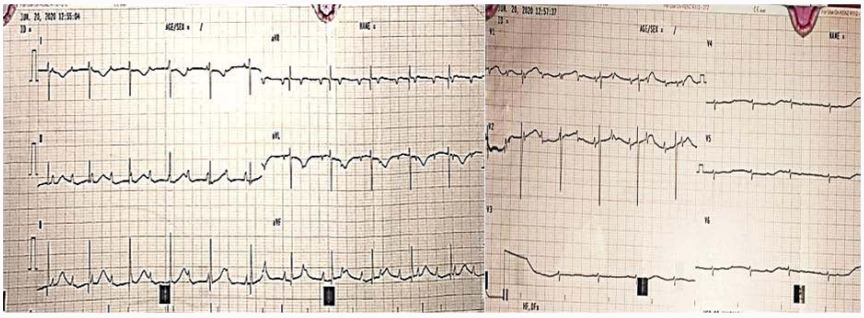

Figure 1: ECG showed normal sinus rhythm 120/min, complete heart block with junctional escape rhythm of 71/min, narrow QRS, superior axis, rS pattern in left precordial leads with absent transitional QRS complex.

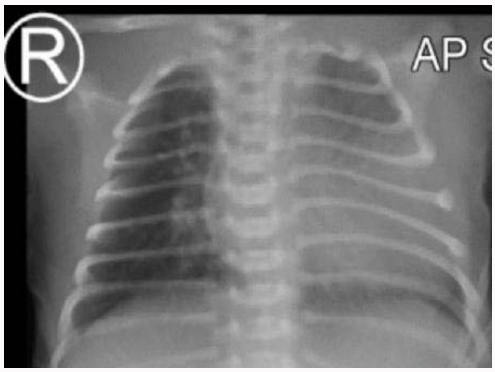

Figure 2: The chest X-ray.

2670 gram.

On physical examination no specific signs was detected, a part of HR 70 bpm and BP 49/34(39) mmHg on dopamine 5 ug/kg/min. CHB with narrow junctional escape rhythm and features of ventricular inversion were seen on electrocardiogram. The Chest X-Ray revealed cardiomegaly, narrow cardiac waist and left lung infiltrate (Figure 2). Laboratory findings showed increase septic markers, hyperbilirubin and hypoalbuminemia. Echocardiography confirmed diagnosis of situs solitus, CCTGA, patent foramen ovale, small patent ductus arteriosus and mild TR (Figure 3). Dopamine infusion was continued, antibiotic was adjusted according to blood culture result and transvenous temporary pacemaker was inserted with the lead placed at the endocardial RV. Two weeks later a permanent VVI mode pacemaker was implanted at the RV epicardial, the patient was discharged in full recovery.

Discussion

Although the sinus node is normal, in CCTGA the AV node and bundle of his have an unusual location and course, therefore cardiac conduction disorders are frequent [1-3,5]. Bradycardia in our patient was detected through handheld Doppler ultrasound; no fetal echocardiography was done due to limited resources. Fetus with isolated CCTGA may be overlooked by routine ultrasound screening. A left sided or posterior RV, identified by reversed differential insertion or the moderator band, in association with abnormal orientation of the great arteries should alert a sonographer to the diagnosis of CCTGA [6].

Isolated CCTGA account for approximately 10% of CCTGA, typically be asymptomatic early in life, those patients may become symptomatic due to systemic RV dysfunction, progressive TR, congestive heart failure, heart block, or ventricular arrhythmia by the fourth or fifth decade of life. Transthoracic echocardiogram using a segmental approach will confirm the diagnosis and will determine the presence of any associated malformations. Cardiac MRI is particularly important for assessment of ventricles volume and function. Cardiac catheterization typically reserved for the post-surgical intervention [3,7].

Recent indications for cardiac pacing in CCHB are: 1) symptomatic patients, 2) asymptomatic CHB with ventricular dysfunction, prolonged QTc, complex ventricular ectopic, wide QRS escape, abrupt pauses in ventricular rate that are two or three times the basic cycle length, HR <55 bpm or <70 bpm associated with CHD such in our patient [1,2,8]. Our patient has HR of 70 bpm with PDA and cardiomegaly on chest X-ray, for these reasons

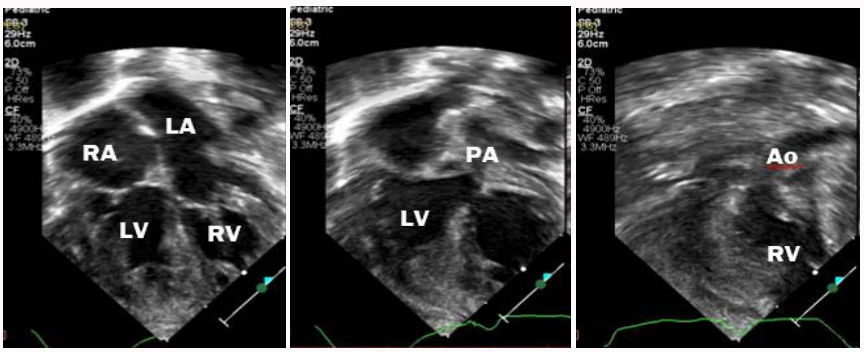

Figure 3: Echocardiography showed CCTGA: The right AV valve insertion higher than the left AV valve, and the chordae attached to the free wall (typical for mitral valve) with smooth Left Ventricle (LV) where the main Pulmonary Artery (PA) originated. Beside had lower insertion the left AV valve chordae attached to the septum, (typical for tricuspid valve) with coarse Right Ventricle (RV) where the Aorta (Ao) originated.

pacemaker was implanted. It is now recognized that a subset of paced patients develops dilated cardiomyopathy and heart failure, therefore regular follow-up is mandatory. Epicardial pacemaker is indicated in children weighing <15 kg, in the presence of intracardiac shunts, absence of appropriate cardiac cavity, and absent or limited venous access to the heart. The disadvantage of epicardial leads is the shorter longevity of these leads compared with endocardial leads. Reports have demonstrated an association between single chamber pacemaker implantation and the deterioration of systemic RV function related to ventricular dyssynchrony, septal activation causing a septal "shift" and secondary dilatation of the TV annulus and worsening TR in patients with CCTGA. These are common indications for conversion to a dual-lead pacing system [8]. Asymptomatic CCTGA patients with evidence of declining RV function and worsening TR should be operated. The decision toward physiologic operation or anatomic operation depends on function of both ventricles and age of the patient. A multi-institutional study still required to confirm the superiority of anatomical repair.

Conclusion

Although CCHB in CCTGA may associated with high morbidity and mortality, a planned antenatal monitoring, delivery and early referral can lead to favorable outcomes. However, lifelong follow up related to CCTGA natural history and epicardial pacemaker negative effects is important; this may become a problem for patients who live in countries with limited resources.

References

1. Baruteau AE, Pass RH, Thambo JB, Behaghel A, Le Pennec S, Perdreau E, et al. Congenital and childhood atrioventricular blocks: Pathophysiology and contemporary management. Eur J Pediatr. 2016;175(9):1235-48.
2. Bordachar P, Whinnett Z, Ploux S, Labrousse L, Haissaguerre M, Thambo JB. Pathophysiology, clinical course, and management of congenital complete atrioventricular block. Heart Rhythm. 2013;10(5):760-6.
3. Wallis GA, Debich-Spicer D, Anderson RH. Congenitally corrected transposition. Orphanet J Rare Dis. 2011;6:22.
4. van der Linde D, Konings EEM, Slager MA, Witsenburg M, Helbing WA, Takkenberg JJM, et al. Birth prevalence of congenital heart disease worldwide: A systematic review and meta-analysis. J Am Coll Cardiol. 2011;58(21):2241-7.
5. Anderson RH, Becker AE, Arnold R, Wilkinson JL. The conducting tissues in congenitally corrected transposition. Circulation. 1974;50(5):911-23.
6. Sharland G, Tingay R, Jones A, Simpson J. Atrioventricular and ventriculoarterial discordance (congenitally corrected transposition of the great arteries): Echocardiographic features, associations, and outcome in 34 fetuses. Heart. 2005;91(11):1453-8.
7. Presbitero P, Somerville J, Rabajoli F, Stone S, Conte MR. Corrected transposition of the great arteries without associated defects in adult patients: Clinical profile and follow up. Br Heart J. 1995;74(1):57-9.
8. Floris EA. Udink ten Cate and Narayanswami Sreeram. Pacing therapy in infants and children with congenital and acquired complete atrioventricular block: Optimal pacing strategies, management, and follow-up.