

High Degree AV Block in Infants

Agus Cahyono¹

Abstract

Background: Atrioventricular (AV) block in children may pose a challenge for physicians. However, it can be detected with careful physical examination.

Case illustration: A 4-month-old infant presented with bradycardia that did not improve during the observation period. Her electrocardiography (ECG) showed a 3:1 high degree atrioventricular block and her echocardiography showed secundum atrial septal defect (ASD) and patent ductus arteriosus (PDA). Her father's ECG showed a first-degree AV block. The patient recovered well after pacemaker implantation and PDA ligation.

Conclusion: An infant who suffered from a 3:1 high-degree AV block was successfully treated with a pacemaker.

¹ Department of Clinical
Medicine, Faculty of Medicine,
Universitas Surabaya, Surabaya,
Indonesia.

Correspondence:

Agus Cahyono
Department of Clinical Medicine,
Faculty of Medicine, Universitas
Surabaya, Surabaya, Indonesia.
Email: agus_jsc@yahoo.co.id

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Introduction

Atrioventricular (AV) block in children, albeit rare, is a serious problem that requires special attention. In a multicenter study, this case was found only in 141 children in 28 years. However, if undetected, it can lead to sudden death.¹ In children, this disease can go unnoticed by parents due to no visible symptoms.² Further, in some instances, it can be detected by chance after a careful physical examination during a visit to healthcare facilities for other diseases.³

There are several AV block classification degrees. First-degree AV block is the mildest and shows in electrocardiogram (ECG) as a prolonged PR interval. Second-degree AV block begins to show asynchronous contractions of the atria and ventricles and presents in the ECG as P waves that are not always followed by a QRS complex. In third-degree or complete AV block, the heart ventricles experience bradycardia with a very low frequency of contractions at less than 60 beats per minute (bpm), appearing as independent contractions of the atria and ventricles in the ECG.⁴ Clinically, AV block in children can be asymptomatic or presented as syncope, fatigue, and heart failure.⁵

Even though third-degree AV block is severest, it can be asymptomatic in children and may have been overlooked by healthcare workers. This can be attributed to various reasons, including the extremely rare prevalence that makes it the least possible diagnosis and, thus, not screened during medical consultation.⁵ Another cause is that children under three years of age are unable to fully communicate the signs and symptoms they feel so the disease is often unidentified.⁶

Case illustration

A 4-month-old female infant coughing for three consecutive days was brought by her parents to the hospital's emergency room. She presented with a high fever (since a day before admission) and cough, without nausea, vomiting, diarrhea, and seizures. Her older sibling also had a cough. This was her episode of respiratory symptoms. Parents informed the emergency department that the patient had been given formula milk and sometimes seemed startled for a few seconds. On physical examination, the patient was conscious,

pulse rate of 130 bpm, respiratory rate of 55x/minute, the body temperature of 37.4°C, fine wet crackling sounds in both lung fields, S2 splitting, no murmur, and weight of 4500g. According to the mother, the patient was able to lift her head (appropriate for a 4-month-old developmental milestone), and her birthweight 3000g (with only 1500g increment of body weight within 4 months indicating failure to thrive) Laboratory results showed Hb 12.3 g/dL, leukocytes 8930/mm³, platelets 510,000/mm³, random blood sugar 112mg/dL, Sodium 135 mmol/L, Potassium 4.9 mmol/L, and IgG, IgM negative for SARS-CoV-2. Upon chest radiological examination, infiltrates were found in both lung fields, with prominent pulmonary conus and cardiothoracic ratio (CTR) of 61% (**Figure 1**).

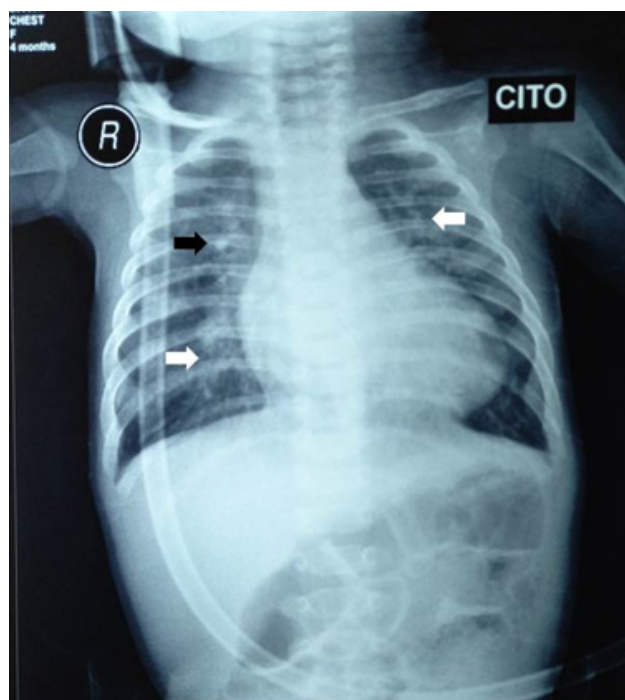


Figure 1. Thoracic radiology. The image shows infiltrates in both lung fields (white arrow), prominent pulmonary conus (black arrow), and cardiomegaly with a CTR of 61%.

The patient was diagnosed with bronchopneumonia and received oxygen mask therapy at 4 liters per minute, ampicillin sulbactam 3x150mg intravenously, and paracetamol 3x50mg orally. The bronchopneumonia and shortness of breath improved during the treatment; then, the oxygen mask therapy was discontinued on the third day. The patient had developed bradycardia

on the fourth day, and an ECG examination revealed a 3:1 high-degree AV block (**Figure 2a and 2b**), although no symptoms were observed. She was hemodynamically stable with a blood pressure of 90/44 mmHg and a capillary refill rate of < 2 seconds. Nevertheless, she was closely observed and attached to a bedside vital sign monitor, and the parents were given educational counseling regarding her disease for potential pacemaker implantation if symptoms appeared and the condition on treatment remained.

After one day of observation, the parents complained that their child's vision occasionally went blank, however, no repetitive movement from extremities or facial muscles was observed. On physical examination when this occurred, the pulse rate was 50 bpm, and the oxygen saturation was 94%. An echocardiographic

examination showed cardiac function was within the normal limits (i.e., ejection fraction (EF) 72% and fraction shortening (FS) 39%) and found left atrial and left ventricular dilatation, patent ductus arteriosus (PDA) 0.7 cm (**Figure 3**), and a secundum atrial septal defect (ASD) with a diameter of 0.5 cm and a left-to-right shunt (**Figure 4**). ECG tests of both parents and three siblings of the patient were also performed to determine if the AV block was hereditary. The electrocardiography of the mother and three siblings was normal, while that of the father showed first-degree AV block in lead V1. The patient was then referred to a higher-level healthcare facility for pacemaker implantation and PDA ligation. After the insertion of a pacemaker, she was hemodynamically stable, and her heart rate was 95 bpm.

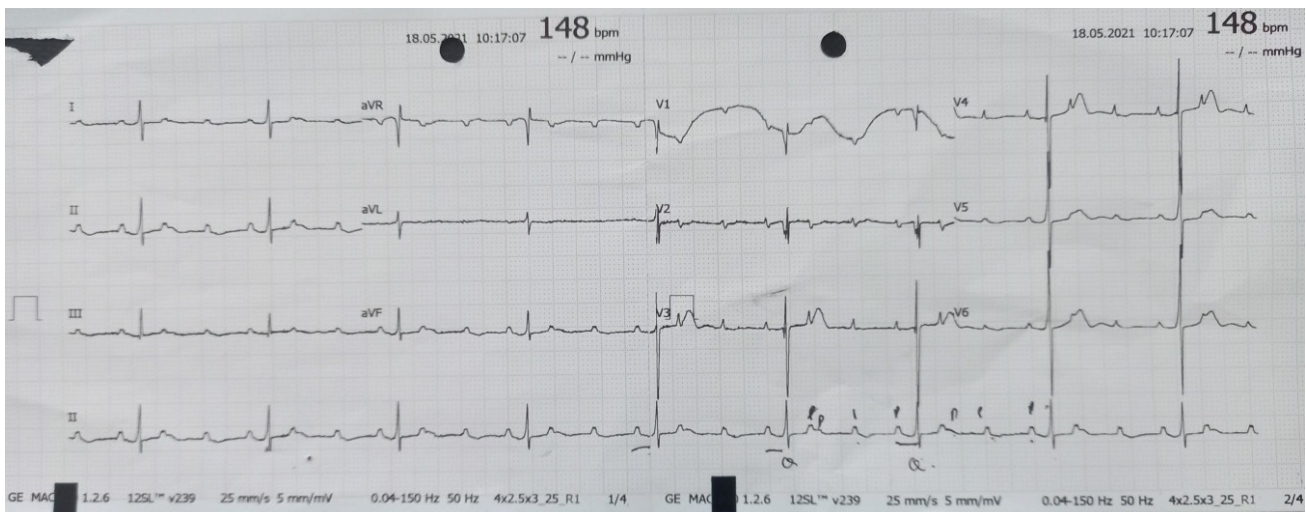


Figure 2a. 12 lead electrocardiography of the patient. The recording shows 3:1 high degree AV block.



Figure 2b. Lead II electrocardiography of the patient. Every three P wave is followed by the QRS complex, indicating a 3:1 high degree AV block.

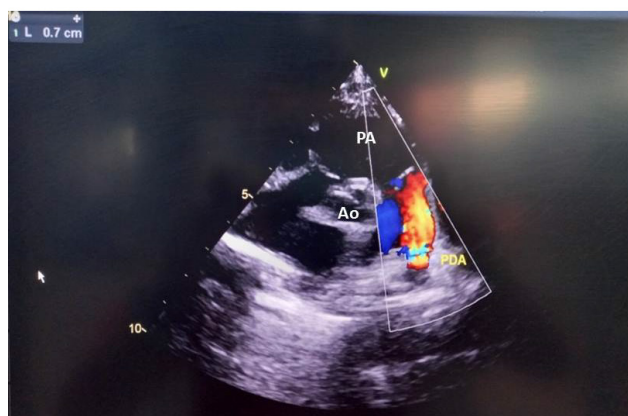


Figure 3. Short axis echocardiography view. The image shows a PDA with a diameter of 0.7 cm and a left-to-right shunt. PA pulmonary artery, Ao aortic valve.

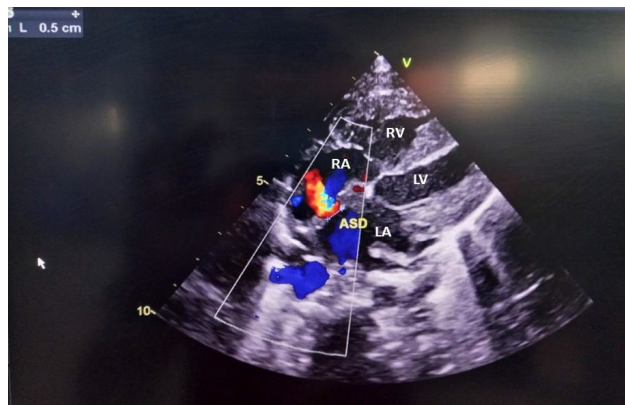


Figure 4. Subcostal echocardiography view. The image shows a secundum ASD with a diameter of 0.5 cm and a left-to-right shunt. RA right atrium, LA left atrium, RV right ventricle, LV left ventricle.

Discussion

AV block is a rare disease in children and, thus, a frequently overlooked diagnosis. This study reported a 4-month-old with AV block after several days of hospitalization. The possibility of AV block was examined after the patient developed bradycardia because there were no clinical symptoms. A fitting symptom, occasionally blank stares, appeared for the first time a day after the diagnosis. A multicenter study reported that, for almost 30 years, there were only 141 cases of AV block in fetuses and pediatric patients of up to 15 years old, and merely 15.6% were symptomatic at the time of diagnosis.⁵ Another case study found a 9-month-old infant experiencing complete AV block without showing symptoms.² Children, especially those under three years old, often rely on expressive language inadequate to communicate their feelings, making the diagnosis difficult. Moreover, four-month-old infants can only express their feelings with vocal games.⁶ Overall, diagnosing cases of AV block in pediatric patients is inevitably challenging.

The conduction disorder found in this case is a 3:1 high-degree AV block. ECG examination found bradycardia (pulse rate 55 bpm) and every three P waves were followed with the QRS complex. To determine the severity (degree) of the AV block, physicians can observe the PR interval and synchronization between

the P wave and the QRS complex. Although the patient, in this case, did not show symptoms of AV block on admission, further observation revealed that the three P waves were followed by the QRS complex, an indicator of a 3:1 high-degree AV block. Advanced, high-grade, or high-degree atrioventricular block: ≥ 2 consecutive P waves at a constant physiologic rate that do not conduct to the ventricles with evidence for some atrioventricular conduction.⁴ Clinically, in pediatric patients, this condition can be asymptomatic or presented as syncope, fatigue, and heart failure.⁵

In this case, the 3:1 high-degree AV block was found together with pneumonia. In its clinical course, the pneumonia improved while the heart block signs began to appear. The patient's ECG did not reveal low voltage, prolonged PR interval, ST elevation, T inversion, or QT prolongation, which are signs of myocarditis. Also, her antibody test results for SARS-CoV-2 were negative. Based on these reasons, an echocardiographic examination was performed. The results showed good heart function, dilated left ventricle and left atrium, PDA with a diameter of 0.7 cm, a secundum ASD with a diameter of 0.5 cm and a left-to-right shunt, and no pericardial effusion. The likelihood of myocarditis as a cause of AV block is small. Myocarditis is a condition that is not easy to diagnose because of its wide range of clinical manifestations, ranging from subclinical to fulminant heart failure. Its common signs and symptoms include acute chest pain, dyspnea on exertion, palpitations, arrhythmias,

syncope, and cardiogenic shock. Myocarditis can appear as first to third-degree AV blocks, bundle branch block, ST/T wave changes, sinus arrest, ventricular tachycardia, atrial fibrillation, reduced R-wave height, widened QRS complex, abnormal Q waves, low voltage, frequent premature beats, and supraventricular tachycardia in the electrocardiogram. Also, the myocardiocytolysis markers show elevated TnT/TnI. Further ECG examination will reveal functional abnormalities of the left ventricle and/or right ventricle, and cardiac magnetic resonance (CMR) imaging will show edema and/or a classic form of myocarditis.⁷ Some viruses like Coxsackie, Adenovirus, and Echovirus and bacteria like Diphtheria, Salmonella, and Leptospira can cause myocarditis.⁸ However, during the current SARS-CoV-2 pandemic, this virus is also clinically suspected to be responsible for myocarditis.⁹ Although Indonesia is a tropical country with a high incidence of dengue virus infection—a probable cause of myocarditis,³ it can be ruled out because the patient in the reported case did not show signs of bleeding or thrombocytopenia.

The patient was found with structural defects, namely ASD and PDA. In this case, the ostium secundum ASD can result in AV blocks because of the increased right atrial load that disrupts cardiac conduction, and the PDA adds pressure to the left ventricle and atrium. ASD is the most common congenital heart disease, and the ostium primum defect most often results in high-degree AV blocks.¹⁰ The AV blocks can occur at any time without surgical interventions or procedures.¹¹ In adults, chronic right atrial stretch causes electrical remodeling with a prolonged right atrial effective refractory period (RA ERP), slower conduction at the crista terminalis, and sinus node dysfunction.¹² Meanwhile, in PDA, cardiac rhythm disturbances can occur in the form of sinus tachycardia or atrial fibrillation in moderate or large shunts.¹³

Although the AV block can be attributed to ASD, there is a possibility of hereditary cardiac conduction disorder. On ECG examination, the patient's father had prolonged PR interval, while the mother and the three siblings were normal. There are at least two genes to be investigated, namely NKX2.5 and SCN5A. In addition to ASD, mutations in NKX2.5 are responsible for postnatal AV block¹⁴, which potentially leads to the loss of conduction system myocytes, a mechanistic pathway for AV block.¹⁵ These mutations

can be autosomal dominant inheritance.¹⁶ Meanwhile, mutations in SCN5A can also cause AV block through the channelopathy mechanism.¹⁷

On admission, the AV block was managed by close observation because the patient was hemodynamically stable. When symptoms such as daydreaming appeared, she was given an oxygen mask and considered for immediate insertion of a pacemaker. Postnatal AV block management is medical, including administering drugs (isoproterenol, atropine, epinephrine, and intravenous dopamine) and implanting a pacemaker. A pacemaker is recommended for symptomatic second or third-degree AV blocks,¹⁸ and in its absence, children with AV block will have a different prognosis depending on the age at diagnosis. The mortality rate can be as high as 43% if the condition is diagnosed in utero (26.1±5.1 weeks, fetal life) and decrease to 6% and 0% if diagnosed within the first month of life (≤28 days, infancy) and during early childhood (5.7±4.8 years).¹ Fortunately, the pacemaker was placed in this patient and the response was remarkable. **(Figure 5)** shows every impulse from the pacemaker was followed by a QRS complex.

Conclusion

3:1 High-degree AV block in infants is rare and requires careful examination for identification. In these cases, the possibility of it being inherited genetically needs to be considered. Appropriate management, such as the insertion of a pacemaker, is necessary for a better prognosis.

Acknowledgements

None

Conflict of interest

None

References

1. Jaeggi ET, Hamilton RM, Silverman ED, Zamora SA, Hornberger LK. Outcome of children with fetal, neonatal or childhood diagnosis of isolated congenital atrioventricular block. *JACC*. 2002;39(1):130–7.

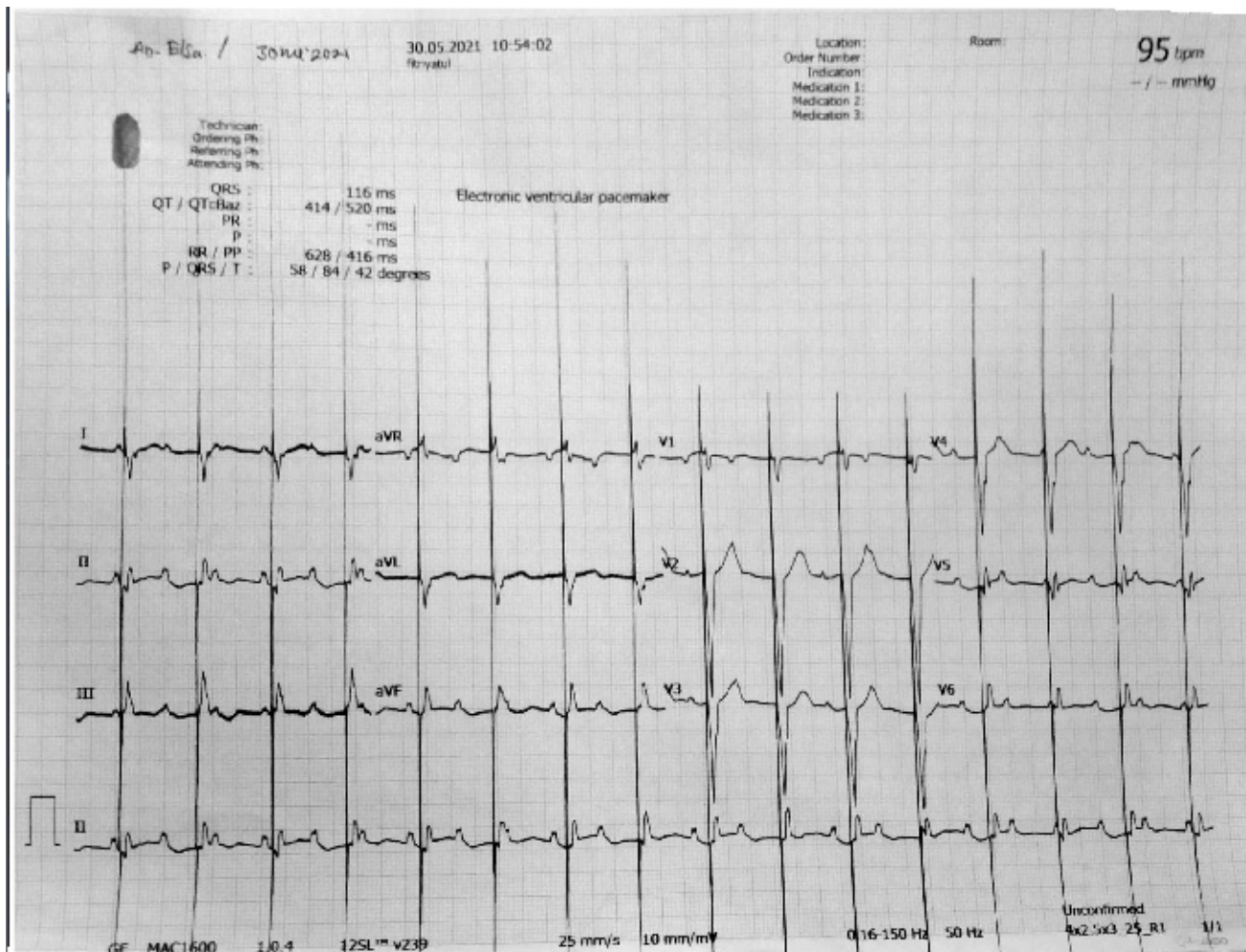


Figure 5. ECG after pacemaker placement. Every pacemaker impulses was followed by QRS complex.

2. Mall P, Shah I. A 9 months old child with asymptomatic bradycardia. *Pediatric Oncall Journal*. 2020;17(2):68–9.
3. Yantie NPVK, Gunawijaya E, Suradipa IW, Gustawan IW. Asymptomatic cardiac rhythm abnormality in children with dengue virus infection. *Bali Med J*. 2016;5(2):351–4
4. Kusumoto FM, Schoenfeld MH, Barrett C, Edgerton JR, Ellenbogen KA, Gold MR, Goldschlager NF, Hamilton RM, Joglar JA, Kim RJ. Guideline on the evaluation and management of patients with bradycardia and cardiac conduction delay. *J AM Coll Cardiol*. 2018. <http://www.onlinejacc.org/guidelines/bradycardia>
5. Baruteau AE, Fouchard S, Behaghel A, Mabo P, Villain E, Thambo JB, Marcon F, Gournay V, Rouault F, Chantepie A, et al. Characteristics and long-term outcome of nonimmune isolated atrioventricular block diagnosed in utero or early childhood: a multicentre study. *European Heart Journal*. 2012;33:622–9.
6. Lust B. *Child Language*. Cambridge University Press;2006
7. Hazebroek MR, Everaerts K, Heymans S. Diagnostic approach of myocarditis: strike the golden mean. *Neth Heart J*. 2014;22:80–4.
8. Dancea AB. Myocarditis in infants and children: A review for the paediatrician. *Paediatr Child Health*. 2001;6(8):543–5.
9. El-Assaad I, Hood-Pishchany MI, Kheir J, Mistry K, Dixit A, Halyabar O, Mah DY, Meyer-Macaulay C, Cheng H. Complete heart block, severe ventricular

- dysfunction, and myocardial inflammation in a child with COVID-19 infection. *J Am Coll Cardiol Case Rep.* 2020;2:1351–5.
10. Goodman DJ, Harrison DC, Cannom DS. Atrioventricular conduction in patients with incomplete endocardial cushion defect. *Circulation.* 1974;49:631–7.
 11. Mehta AV, O’Riordan AC, Sanchez GR, Black IFS. Acquired nonsurgical complete atrioventricular block in a child with endocardial cushion defect. *Clin. Cardiol.* 1982;5:603–5.
 12. Morton JB, Sanders P, Vohra JK, Sparks PB, Morgan JG, Spence SJ, Grigg LE, Kalman JM. Effect of chronic right atrial stretch on atrial electrical remodeling in patients with an atrial septal defect. *Circulation.* 2003;107:1775–82.
 13. Douglas J, Schneider, MD; John W. Moore. Patent ductus arteriosus. *Circulation.* 2006;114:1873–82.
 14. Benson DW, Silberbach GM, Kavanaugh-McHugh A, Cottrill C, Zhang Y, Riggs S, Smalls O, Johnson MC, Watson MS, Seidman JG, Seidman CE, Plowden J, Kugler JD. Mutations in the cardiac transcription factor NKX2.5 affect diverse cardiac developmental pathways. *J. Clin. Invest.* 1999;104:1567–73.
 15. Pashmforoush M, Lu JT, Chen H, St. Amand T, Kondo R, Pradervand S, Evans SM, Clark B, Feramisco JR, Giles W, Yen Ho S, Benson DW, Silberbach M, Shou W, Chien KR. Nkx2-5 pathways and congenital heart disease: loss of ventricular myocyte lineage specification leads to progressive cardiomyopathy and complete heart block. *Cell.* 2004;117:373–86.
 16. Gutierrez-Roelens I, De Roy L, Ovaert C, Sluysmans T, Devriendt T, Brunner HG, Vikkula M. A novel CSX/NKX2-5 mutation causes autosomal dominant AV block: are atrial fibrillation and syncope part of the phenotype? *European Journal of Human Genetics.* 2006;14:1313–6.
 17. Wang DW, Viswanathan PC, Balser JR, George AL, Benson DW. Clinical, genetic, and biophysical characterization of SCN5A mutations associated with atrioventricular conduction block. *Circulation.* 2002;105:341–6.
 18. Baruteau AE, Pass RH, Thambo JB, Behaghel A, Le Pennec S, Perdreau E, Combes N, Liberman L, McLeod CJ. Congenital and childhood atrioventricular blocks: pathophysiology and contemporary management. *Eur J Pediatr.* 2016;175:1235–48.