Cardiology in the Young

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A child with spontaneous recovery of normal sinus rhythm from idiopathic complete atrioventricular block

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Brief Report

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Abstract

Atrioventricular block in children is not common but is a life-threatening disease. As no spontaneous regression of conductive disruption was reported, those sustaining idiopathic atrioventricular blocks are difficult to manage and often require pacemaker implantation. In this study, we presented the first case of a child who surprisingly recovered from idiopathic complete atrioventricular block without intervention 4 years after initial presentation.

An 8-year-old girl was transferred to our institution due to bradycardia. The patient remained asymptomatic yet was informed with slow heart rate at health checkup. Her birth history was unremarkable and the maternal pregnancy course was uneventful. There was no delay in the developmental milestones. Additionally, family history thereof was negative for dilated cardiomyopathy, conduction disturbance, Brugada syndrome, or infiltrative disease. She had no history of myocarditis or sarcoidosis.

In outpatient clinic, the girl was fair-looking and haemodynamically stable. However, chest X-ray showed borderline cardiomegaly with a cardiothoracic ratio of 51.5% (Fig 1a). Electrocardiogram (ECG) documented left ventricular hypertrophy, atrioventricular dissociation, and ventricular rate at 49 bpm without QTc prolongation or QRS complex widening (Fig 2a). Mean heart rate was 53 bpm and maximal RR interval was 1.82 seconds from Holter monitoring. Transthoracic echocardiography illustrated borderline enlarged left ventricular dimension (43.3 mm at end-diastole) with preserved systolic function (ejection fraction 77.6%). Survey upon biochemistry revealed normal autoimmune panel and thyroid function. She was eventually diagnosed as idiopathic complete atrioventricular block. Due to the lack of symptoms and parental reluctance for permanent pacemaker implantation, outpatient follow-up was determined.

Three years after initial presentation, ECG exhibited regression of complete atrioventricular block to 2nd degree atrioventricular block (Mobitz type I) (Fig 2b). Subsequently, improvement to 1st degree atrioventricular block (Fig 2c) was observed 4 years after the first checkup. Latest chest X-ray (4.5 years interval from the 1st visit) also suggested a reduced heart size (cardiothoracic ratio: 48.4%) (Fig 1b). Additionally, a yearly 24-hour Holter monitoring recorded an increasing tendency of mean heart rate, and maximal RR interval was less than 3 seconds in every test (supplementary Table S1). During such period, the patient was free from any episode of fainting or syncope and under no medication.

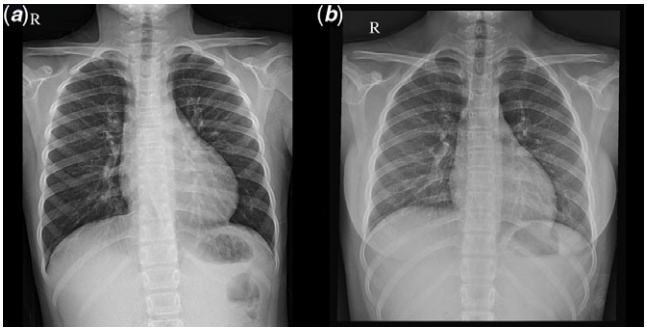

Figure 1. Chest X-ray at initial presentation (***a***) and 4 years from baseline (***b***) Cardiothoracic ratio decreased from 51.5 to 48.4%.

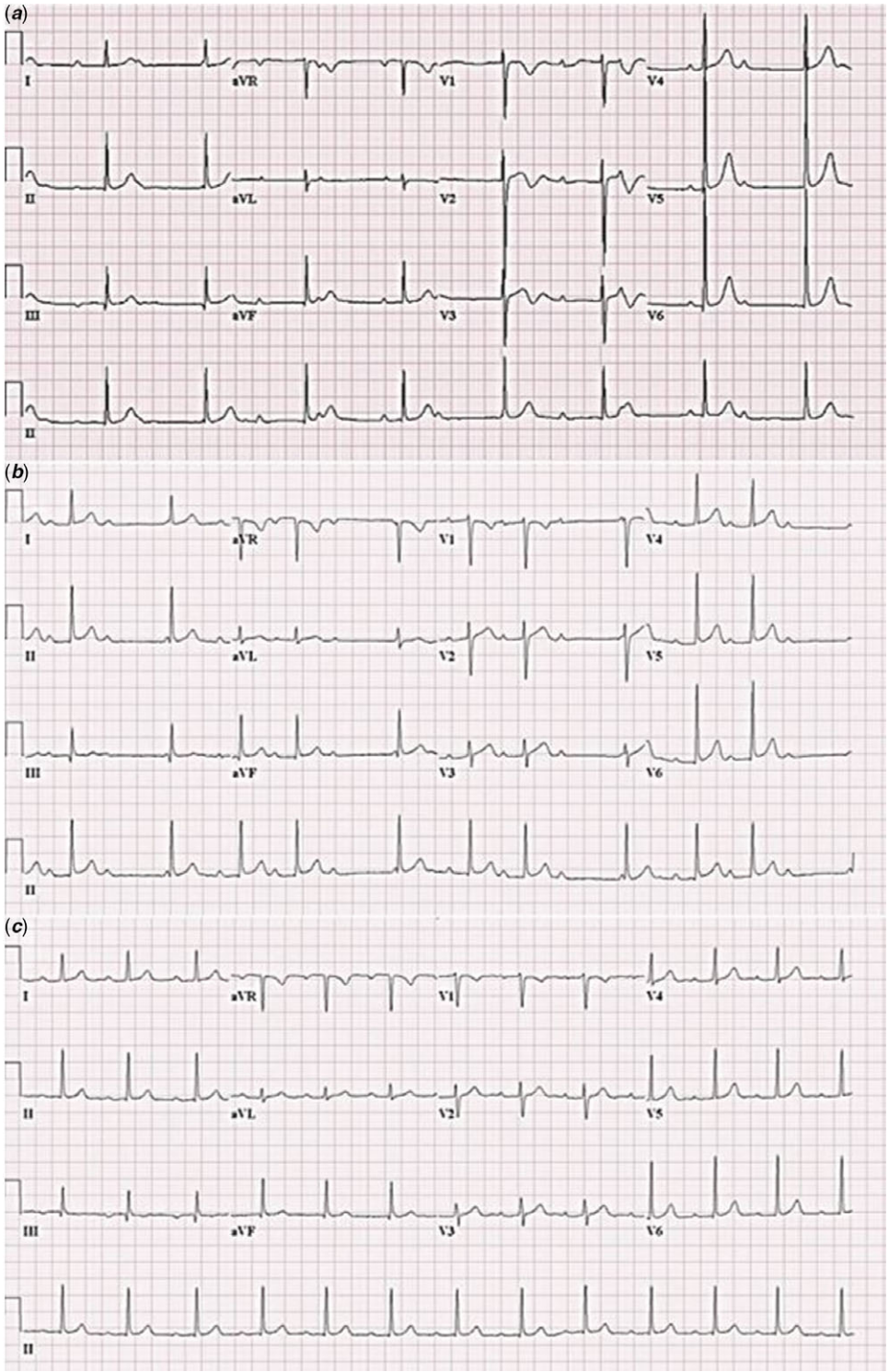

Figure 2. (***a***) Electrocardiogram (ECG) at the first visit documented left ventricular hypertrophy, complete atrioventricular block with ventricular rate at 49 bpm. (***b***) Three years after the first visit, the follow-up ECG changed to 2nd degree atrioventricular block (Mobitz type I). (***c***) Four years after the first presentation, the baseline rhythm recovered to normal sinus rhythm with 1st degree atrioventricular block and mean heart rate at 75 bpm.

Discussions

Herein, we characterised the clinical course of the first child who endorsed spontaneous recovery from complete atrioventricular block. Advanced atrioventricular block is a potentially fatal interruption of the cardiac conduction system, with a global incidence of 1 in 20,000 live births.[1] Mortality rate, albeit in the absence of associated congenital heart disease on most occasions, was estimated to be 8~16% in children with congenital complete atrioventricular block if left untreated.[2] Most patients are symptom-free until later stage of disease progression. Therefore, the diagnosis is

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primarily based on ECG and Holter monitoring, in conjunction with echocardiography to assess cardiac structure and function.

Previous literature has interrogated a plethora of possible triggers for paediatric atrioventricular block, including but not limited to prior infection, infiltrative disease, non-ischemic cardiomyopathy, autoimmunity, Bezold–Jarisch reflex, iatrogenic effect after the administration of β-blockers, calcium channel inhibitors, and antiarrhythmia medications. Although the identification of secondary cause and corresponding specified management are pivotal to restore conductive fluency, those who were eventually classified into idiopathic complete atrioventricular block principally required permanent pacemaker implantation to prevent consequent cardiovascular collapse, Torsade de pointes, or asystole. The risk factors of atrioventricular block ranged from maternal lupus and anti-SSA(Ro)/anti-SSB(La) antibody to elevated systolic blood pressure and hyperglycaemia.[3] Genetically, NKX2.5 mutation was related to conduction defects,[4] while SCN5A variant to atrial standstill and conductive suspension.[5] Hence, the pathogenesis is intertwined with junctional or ventricular escape machinery secondary to the fibrosis of the conductive system. Still, the entire mechanistic understanding is in its infancy.

The diagnostic algorithm to evaluate young patients with advanced atrioventricular block has been well constructed and was practiced when assessing the present case. Notably, no overt cause was delineated after the exhaustion of every inquiry toward the common etiologies, hampering the establishment of further intervention aside from close observation. Moreover, American College of Cardiology/American Heart Association guidelines does not recommend permanent pacemaker implantation on patient sustaining 3rd degree atrioventricular block (class III) unless in the presence of symptomatic bradycardia, comorbid with atrial fibrillation, >3 seconds asystole, escape rhythm <40 bpm, or status post recent cardiac surgery.[6] The management in acute stage as well as the frequency or duration of future follow-ups remained ambiguous. Besides, pattern of exacerbation or subsiding of atrioventricular block has yet been elucidated, making the alteration of the natural clinical course arduous.

To our knowledge, no spontaneous improvement of advanced atrioventricular block has been published. Even though multiple reports documented the recovery of baseline atrioventricular block or post-operative atrioventricular block after congenital heart surgery, no single clinical index was eligible to predict possible regression or of prognostic implication.[7] Unfavourable outcome was associated with atrioventricular block onset unless the reversible cause was eliminated. Other anecdotal case reports described the abatement of complete atrioventricular block due to primary giant cell myocarditis[8] or sarcoidosis[9] after corticosteroid therapy. Besides, one patient recovered from anterior wall myocardial infarction complicating complete atrioventricular block after coronary reperfusion by thrombolysis.[10] Whilst in the present case, no clinical indicator was available at presentation to dictate subsequent therapeutic arrangement. In addition, long-term monitoring is warranted to analyse whether regressed atrioventricular block would recur and the development of future complications, if any. This in turn may retrospectively facilitate the identification of original aetiology.

Conclusions

Scrutinising the secondary cause for advanced atrioventricular block in children thoroughly is paramount to determine the aetiology and potential reversal of conductive abnormality. The paediatric population with idiopathic complete atrioventricular block ought to be probed with the indication for permanent pacemaker implantation. Moreover, the arrangement of a regular follow-up schedule and education on self-monitoring toward subjective conditions are the cornerstones for the prevention of further sequelae, and possibly to anticipate spontaneous recovery.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951120004485

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Conflicts of interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human medical regulations and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by the ethical committee of Seoul Nation University Children's Hospital.

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