Late-presenting complete heart block after pediatric cardiac surgery

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Late-presenting complete heart block after pediatric cardiac surgery is a rare complication and its management is well defined once the initial diagnosis is made timely and appropriately. In this report we described a child who underwent atrioventricular septal defect repair with a normal sinus rhythm during the postoperative period, as well as during the first 2 years of follow up.

She subsequently developed complete heart block with bradycardia that required insertion of a pacemaker. Here we discuss this unusual late-presenting complication, possible risk factors, and management.

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Introduction

Complete heart block (CHB) following open-heart surgery for congenital heart disease is uncommon but a well-known complication. Its incidence ranges between 1% and 3% [1], with more occurrences during surgeries or interventions adjacent to the atrioventricular node [2]. Early-presenting heart block (HB) associated with cardiac surgery or intervention usually appears during surgery or shortly thereafter. However, any HB that develops after 30 days of surgery or intervention, is considered to be late onset [2]. It has been described in children following cardiac surgical repair or cardiac intervention with an interim period of normal rhythm followed by late development of HB that may occur months to years after initial repair [2]. In this report we present a case of a child with trisomy 21 syndrome who underwent atrioventricular septal defect (AVSD) and then presented with CHB 2 years after surgery. The report will review some of the available literature that focuses on late-presenting CHB after pediatric cardiac surgery, try to early identify high-risk groups, and suggest strategies for the follow up of high-risk groups for this potential life-threatening complication.
Case report

A 4-year-old girl with Down syndrome diagnosed at the age of 1.5 years was found to have complete balanced AVSD. She underwent a total repair of the AVSD at the age of 2 years with the usual postoperative course that included normal sinus rhythm with a heart rate ranging between 110 beats/min (bpm) and 150 bpm (Fig. 1). The child was discharged 10 days after surgery with good condition and with no reported arrhythmia during her entire hospital course. After discharge, the patient had satisfactory repair and normal sinus rhythm throughout 2 years of follow up (Fig. 2). However, at 4 years of age during a routine follow-up visit she was found to have CHB with a heart rate ranging between 50 bpm and 76 bpm with junctional escape rhythm (Fig. 3). Although she was clinically asymptomatic, her Holter monitor confirmed the diagnosis of persistent CHB with a junctional rhythm that slowed periodically to 50 bpm. She underwent permanent pacemaker insertion with ventricular pacing – ventricular sensing – inhibit (VVI) mode and was discharged with good condition.

Discussion

The incidence of CHB after pediatric cardiac surgery ranges from 1% to 3% [1]. Most
postoperative CHBs are consequence of procedures near the atrioventricular node [2]. They usually appear immediately during surgery or in the early postoperative period. A high number (43–92%) of patients with transient postoperative CHB recover shortly after surgery [1] and regain their normal sinus rhythm within 10–14 days after surgery. On a rare occasion, HB may occur many months or years after the initial surgical repair with a reported incidence of 0.3% to 0.7% [3,4].

Little has been mentioned regarding the etiology and risk factors of late-presenting HB in postoperative cardiac patients. It is postulated that late presentation of a conduction defect is due to progressive fibrosis in the surgical site near the atrioventricular node with slow sclerosis extending over the conduction pathway that is already fragile [5]. The time for late-presenting HB is variable and ranges from a few months to as late as 19 years after surgery [5]. Typically, it develops within 2–6 years after surgery. The possibility of this late complication development calls for at least yearly follow-up examinations with electrocardiogram for all postoperative cardiac surgical cases who are at risk. Some patients may show subtle warning signs which should be recognized quickly either intraoperatively or postoperatively. Villian et al. [6] studied the predictive factors of late presentation of CHBs and concluded that all patients who have transitional HBs immediately postoperatively lasting more than 48 h and then develop prolonged PR, different P, or QRS morphology compared with preoperative patients are at an increased risk of late developing atrioventricular blocks (AVBs) and should have an electrophysiology study during their follow up. Lin et al. [3] concurred with the same recommendations that transient AVBs postoperatively increase the risk of delayed block.

Owing to the risk of late developing HBs, patients should be followed up for life after repair of congenital heart disease with special attention to the conduction system, particularly after repair of perimembranous ventricular septal defects, Tetralogy of Fallot, or AVSD [4]. Some investigators discussed the effectiveness of electrophysiologic studies in identifying high-risk cases and recommended this diagnostic tool in detecting conduction disturbance after correction of Tetralogy of Fallot. Applying this diagnostic modality may help to detect patients at risk for late-presenting HB and justify prophylactic pacemaker insertion for them [7]. Another consideration should be taken while following patients after discharge is that AVSD per se is a possible risk factor for AVBs. Some authors consider endocardial cushion defect as a risk factor for congenital HB development even without surgery [8,9]. Kugler et al. [9] reported nonsurgical acquired CHB in a child with endocardial cushion defect at the age of 8 years. Craig [10] found that late-developing HBs and the need for pacemaker insertion is one of the causes of reoperation in patients after complete repair of AVSD.

The authors explained the development of CHB as a result of fibrosis near the bundle of His at the edge of the endocardial cushion defect. They hypothesized that constant swirling of blood between the septal leaflet of the tricuspid valve and the ventricular septal defect as a possible cause of the damage to the bundle of His [10]. Other studies suggested that the formation of the atrioventricular node and the ventricular conduction system in AVSD and Down syndrome differ from normal development and that can be a possible contributing factor in the vulnerability to develop atrioventricular conduction disturbance [11].
Conclusion

The development of late CHB is a rare serious complication that requires life-long follow up of patients at risk. An increase in awareness of this complication will enable the physician to identify high risk groups such as those who have had transient heart block after surgery for more than 48 h, prolonged PR interval after surgery, evidence of bundle of His damage, or tri-fascicular damage which can be suspected by a change in QRS morphology postsurgery, or by electrophysiologic study in selected cases. Furthermore, patients with Down syndrome and AVSD should be considered to have a greater risk of developing CHB. Recognizing this risk will lead to early detection and promote immediate treatment.

References

[1] Gross GJ, Chiu CC, Hamilton RM, Kirsh JA, Stephenson EA. Natural history of postoperative heart block in congenital heart disease: implications for pacing intervention. Heart Rhythm 2006;3:601–4.

[2] Kumar Das M. Modern pacemakers – present and future. USA: InTech; 2011.

[3] Lin A, Mahle WT, Frias PA, Fischbach PS, Kogon BE, Kanter KR. Early and delayed atrioventricular conduction block after routine surgery for congenital heart disease. J Thorac Cardiovasc Surg 2010;140:158–60.

[4] Liberman L, Pass RH, Hordof AJ, Spotnitz HM. Late onset of heart block after open heart surgery for congenital heart disease. Pediatr Cardiol 2008;29:56–9.

[5] Laurens P, Gavelle P, Piwnica A, Farge C, Dubost C, Maurice P. Severe postoperative heart blocks appearing late. 16 cases. Arch Mal Coeur Vaiss 1983;76:1132–9.

[6] Villain E, Ouarda F, Bleyer C, Sidi D, Abid F. Predictive factors for late complete atrio-ventricular block after surgical treatment for congenital cardiopathy. Arch Mal Coeur Vaiss 2003;96:455–8.

[7] Friedli B, Bolens M, Taktak M. Conduction disturbance after correction of tetralogy of fallot: are electrophysiologic studies of prognostic value? J Am Coll Cardiol 1988;11:162–5.

[8] Somerville J. Ostium primum defect: factors causing deterioration in the natural history. Br Heart J 1965;27:413–9.

[9] Kugler JD, Gillette PC, Gutgesell HP, McNamara DG. Nonsurgically-acquired complete atrioventricular block in endocardial cushion defect. Cardiovasc Dis 1981;8:205–9.

[10] Craig B. Atrioventricular septal defect from fetus to adult. Heart 2006;92:1879–85.

[11] Blom NA, Ottenkamp J, Deruiter MC, Wenink AC, Gittenberger-de Groot AC. Development of the cardiac conduction system in atrioventricular septal defect in human trisomy 21. Pediatr Res 2005;58:516–20.