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CASE REPORT

Late presenting complete heart block after surgical repair of ventricular septal defect

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ABSTRACT

Late onset of complete heart block is a life-threatening uncommon complication after surgical repair of congenital heart diseases. In this report, we discuss two cases of Perimembranous ventricular septal defect (VSD) that had late presentation of complete heart block after surgical repair. We are aiming to highlight this unusual complication for more awareness, better understanding and management of this unusual complication after pediatric cardiac surgery.

1. Introduction

Despite the advances in management of congenital heart diseases, injury to the conduction system during surgical repair may still occur leading to different degrees of heart block. The incidence of conduction system injury during cardiac surgery ranges between 1% and 3%.[1,2] Post-operative heart block occurs mainly in children who had cardiac repair near the conduction system (Figs. 1 and 2) or when the conduction system is manipulated intra-operatively.[3] Local edema, inflammation and direct insult to atrioventricular node (AVN) or adjacent conduction system contribute to development of heart block during or after surgical repair. Certain types of cardiac surgeries such as VSD repair, atrioventricular septal defect (AVSD) repair and tetralogy of fallout (TOF) repair are associated with higher incidence of heart block during early post-operative stage.[1,4] Heart block usually appears during or shortly after surgery and it is often a transient complication. Majority of children who develop heart block after surgery recover within a few days and the conduction system reclaims its normal sinus rhythm thereafter. Few published papers reported late presentation of heart block following cardiac repair of congenital heart disease, which is defined as a heart block that occurred 30 days after cardiac surgery with a temporary period of normal rhythm.[2]

Late presenting heart block may appear after months or years of impermanent normal sinus rhythm.[5] In general, incidence of late presenting heart block after cardiac repair was described between 0.3% and 0.7% in most of reported pediatric studies.[6] There are, however, a few reports describing higher incidence reaching up to 1–2% in children undergoing cardiac repair.[5] Most of the patients who developed late presenting heart block were noted to have early transient block intra or post-operatively.[6] The causes of this late presenting heart block are related to progressive fibrosis and slow sclerosis extending over conduction pathways, which are congenitally fragile.[5]

Symptoms leading to diagnosis of late presenting heart block vary according to the type of heart block ranging from asymptomatic presentation where the diagnosis is incidentally discovered during routine exam to a more striking symptomatic presentation that may include syncope, dizziness, shortness of breath, signs of congestive heart failure and even sudden death.[5,7] There are scarce case reports describing late presenting complete heart block in children after cardiac repair. In this report we present 2 pediatric cases who underwent surgical repair of VSD and were discharged home with normal sinus rhythm then presented later on with picture of heart block that required immediate management and intervention.

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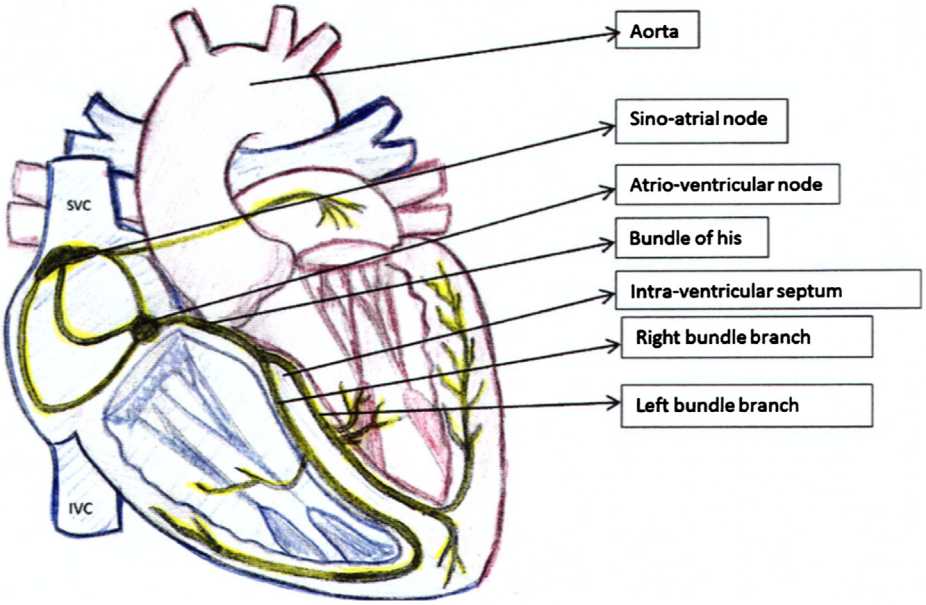

Fig. 1. Conduction system of the heart.

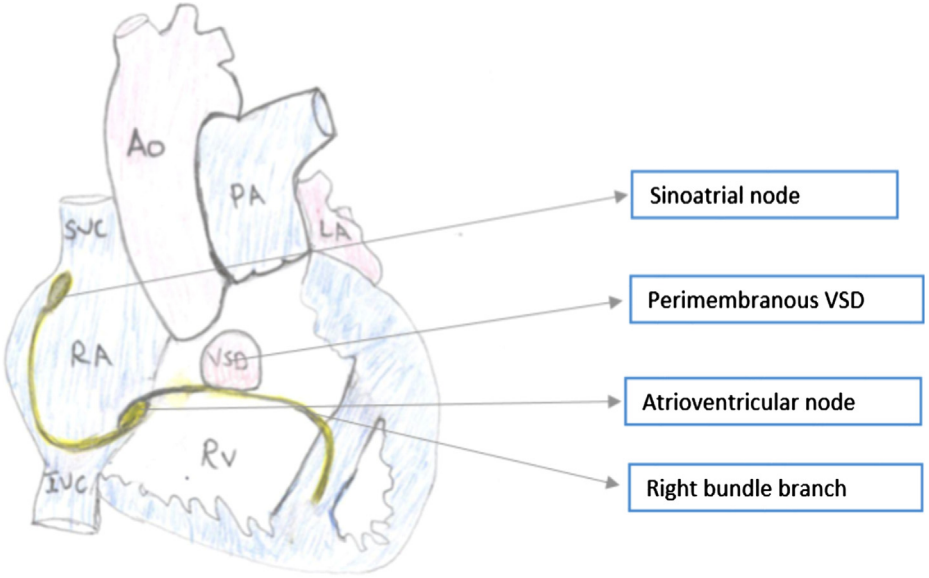

Fig. 2. A view through Right ventricle showing relation between Perimembranous VSD and conduction system.

2. Case report

First case was a 6 months old male weighing 3.8 kg, had surgical repair for Perimembranous VSD. He developed complete heart block immediately after surgery that was managed by temporary pacemaker for two days. Patient recovered and was discharged home on the 7th day post surgery with normal sinus rhythm. Two years later and during routine follow up examination he was noted to have asymptomatic second-degree heart block (Fig. 3). With subsequent follow-ups he was noted to progress slowly to persistent complete heart block with bradycardia that was associated clinically with exertion intolerance. A permanent pacemaker was inserted for him seven years after surgery (Fig. 4).

Second case was a 4 months old female, weighing 3.5 kg, underwent surgical repair for Perimembranous VSD closure. She developed complete heart block early after surgery (Fig. 5) managed by temporary pacemaker for 5 days and discontinued when normal sinus rhythm was detected (Fig. 6). Two month after discharge, patient presented to emergency room for respiratory distress. During her assessment she was discovered to have severe bradycardia due to complete heart block (Fig. 7). She was hospitalized and permanent pacemaker was inserted for her.

3. Discussion

While incidence and management of early post-operative heart block have been studied extensively, only few reports described late presenting complete heart block in children.

Although majority of patients who develop a forewarning early post-operative heart block have high chance (42–93%) to recover their sinus rhythm within 7–14 days after cardiac surgery, they

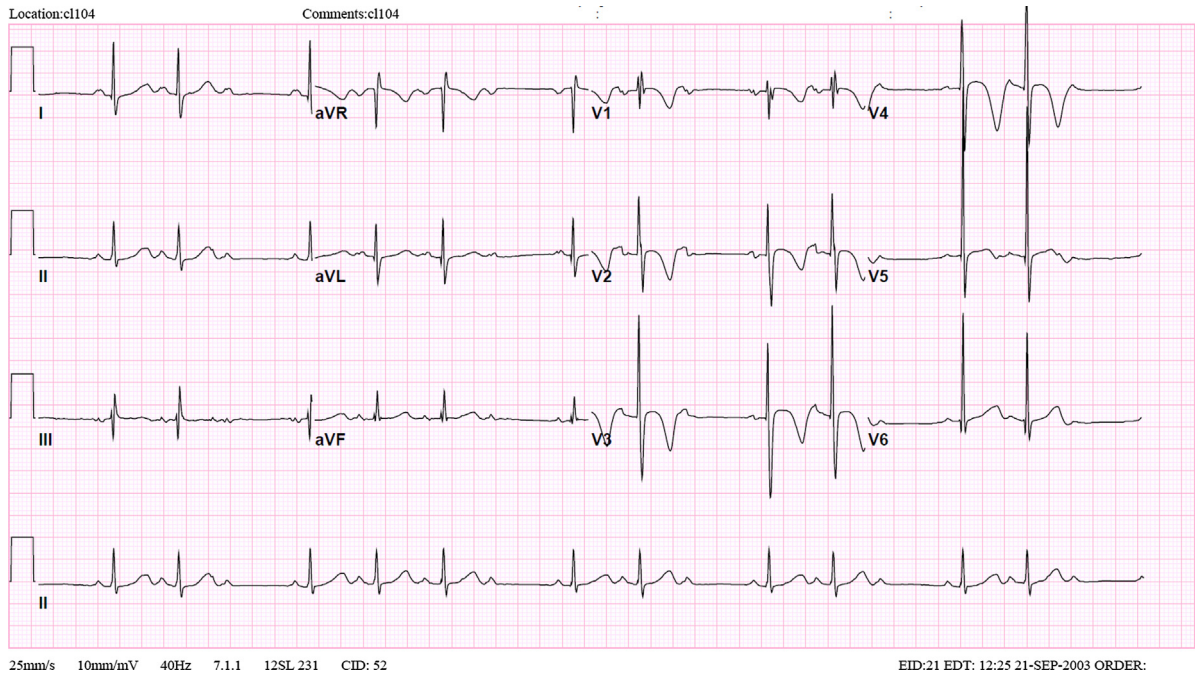

Fig. 3. Second degree heart block two years after surgery in patient (1).

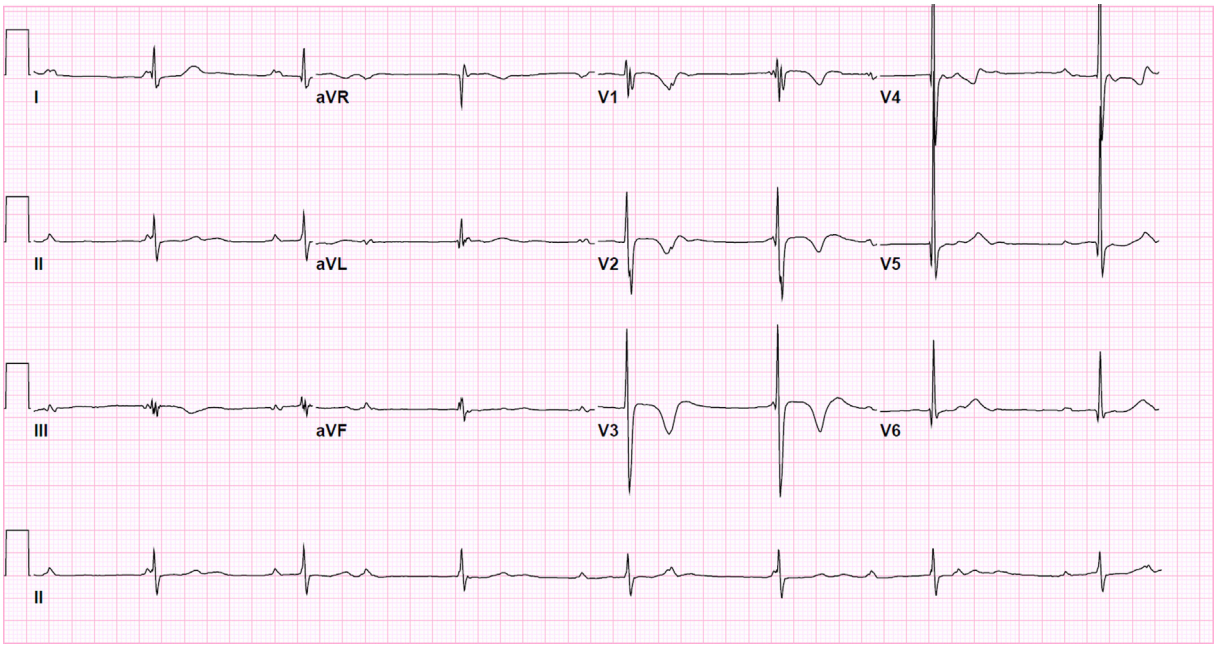

Fig. 4. Complete heart block seven years after surgery in patient (1).

are at higher risk for developing late presenting complete heart block.[2] There are even some occult cases that were blamed for sudden death after cardiac surgery.[8] That may imply a higher actual incidence of late presenting heart block than what have been reported. Both cases that we are reporting developed early postoperative complete heart block and recovered within 3 days in the first case and 5 days in the second case. Premonition signs such as a residual bundle branch block, change of QRS axis, or bifascicular block may alert one for future block, however it is difficult to predict the time of late onset AVB after initial complete recovery. Fukuda et al.[9] reported complete AVB 15 years after ventricular septal defect surgery. Villian et al.[8] studied the predictive factors of late presenting heart block and concluded that patients who have interim heart block early post operatively lasting more than 48 h and then develop prolonged PR, different P morphology, or different QRS morphology compared with preoperative findings

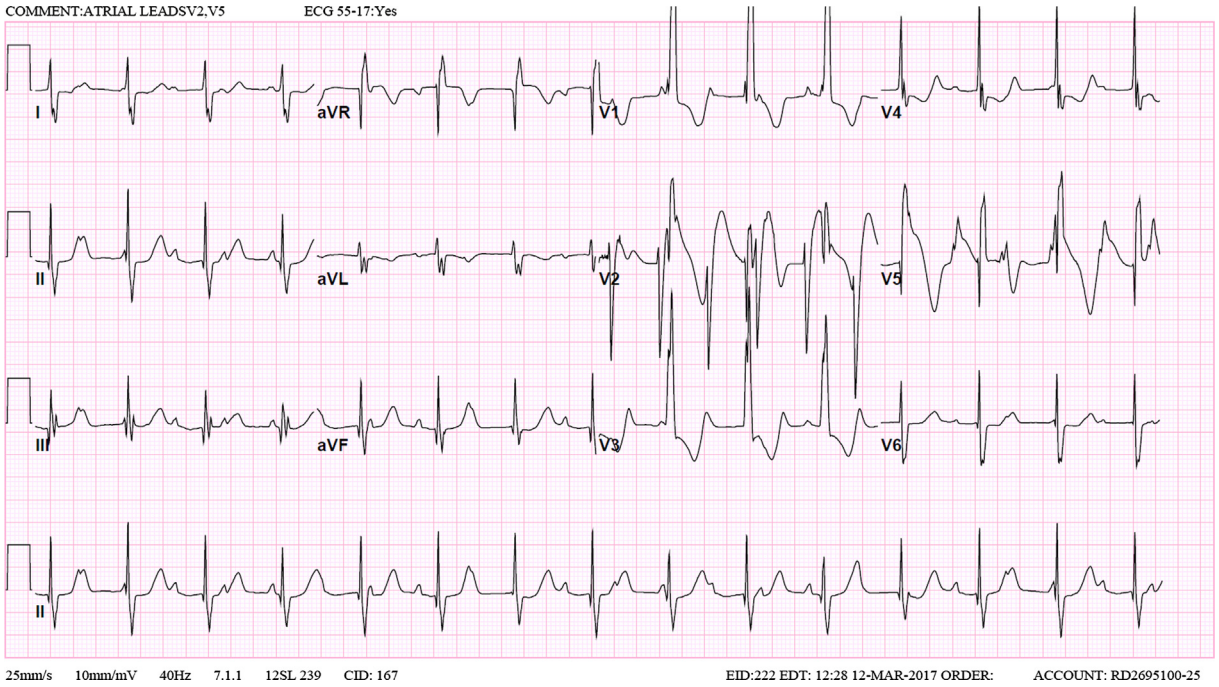

Fig. 5. Complete heart block early after surgery in patient (2).

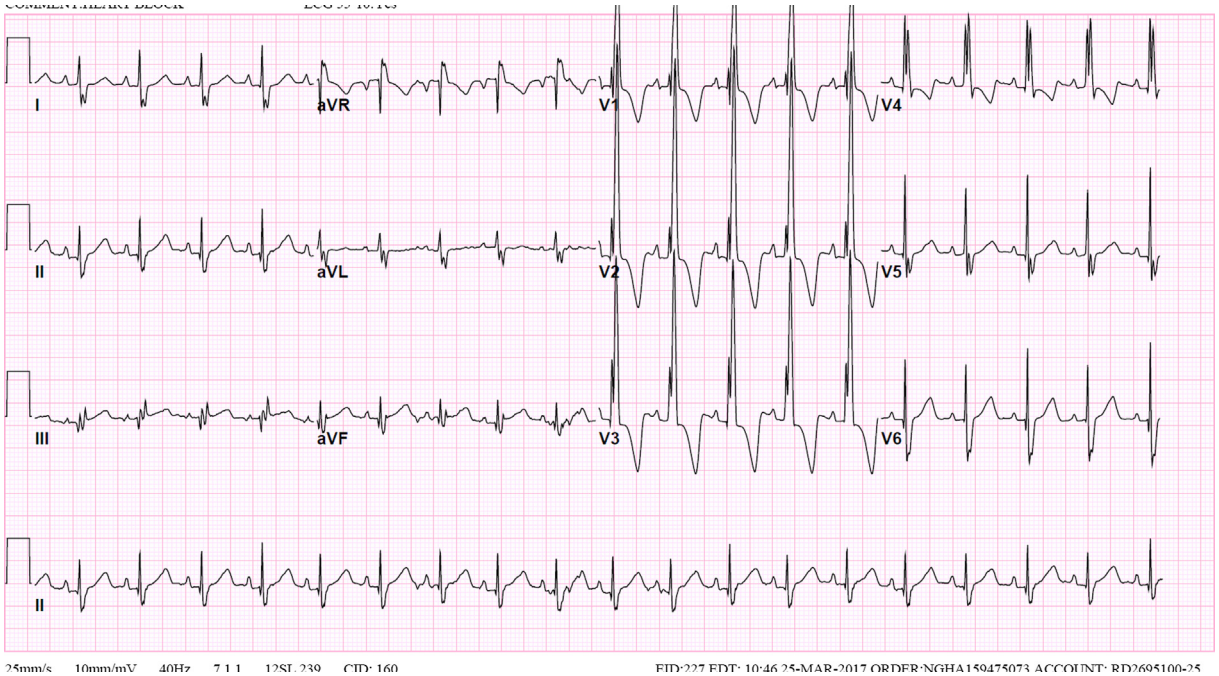

Fig. 6. Recovery of conduction 1:1 with residual RBBB in patient (2).

are at an increased risk of delayed presenting heart block and should have an electrophysiology study during their follow up.

Most of the delayed presenting heart blocks are of an advanced degree.[5] One of our two cases of complete life threating heart block was fortunately preceded with second-degree heart block, which was an alert for parents and physician about this significant complication and the need for frequent follow up and close monitoring. While complete heart block in the second case has led to only respiratory distress without significant cardio hemodynamic embarrassment giving the child enough time to seek medical attention and to be managed appropriately.

It is important to recognize as well that late presenting complete heart block is reported not only after surgical repair of VSD, but also after VSD device closure. Because of proximity of perimembranous VSD to AVN and conduction system, direct mechanical compression by the deployed device on conduction system and local inflammatory response are suggested mechanisms for this rare complication.[10,11]

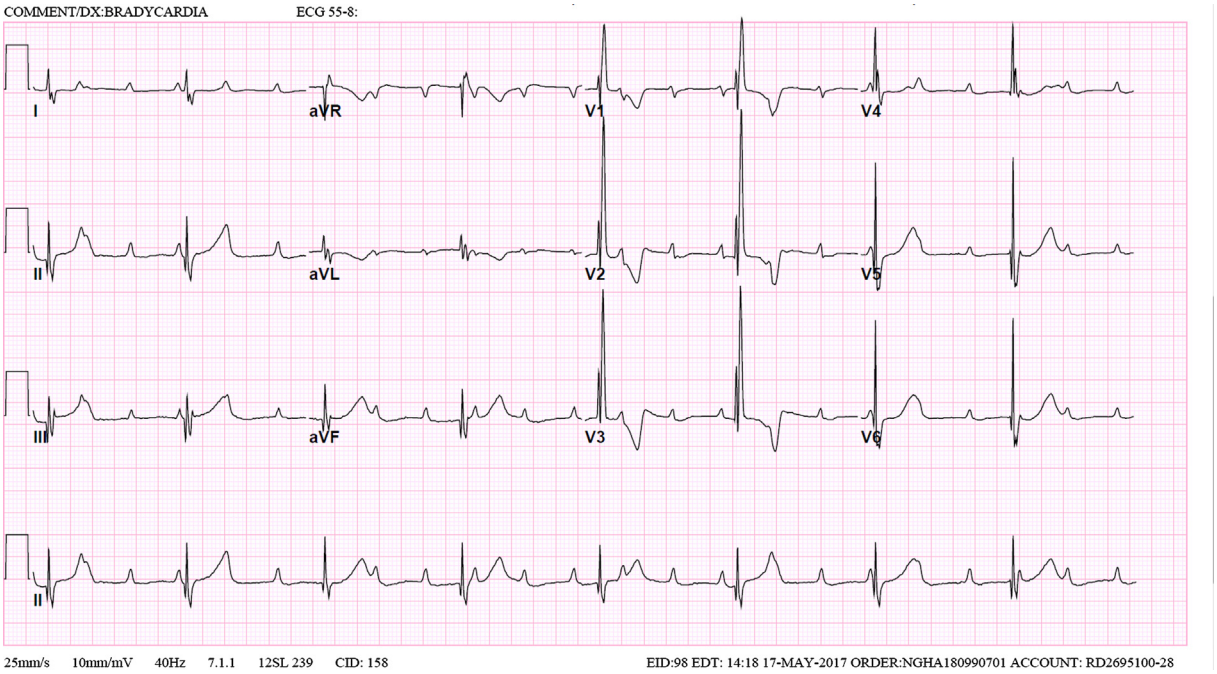

Fig. 7. late presenting complete heart block in patient (2).

4. Conclusion

Late presenting complete heart block is a life threating uncommon complication with higher incidence in children who develop transient early post-operative complete heart block lasting more than 48 h. It is vital to do periodic electrocardiography follow-up studies for those cases and to alert both primary health care providers and parents about this infrequent but potentially significant complication.

Conflict of interest

The authors declare that there are no conflicts of interest.

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