Case Study

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Complete atrio-ventricular septal defect and Wolf-Parkinson-White syndrome

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CASE PRESENTATION

A full term female infant was born after 41 weeks of gestation, with a birth weight of 3815g and normal Apgar scores (9-10-10). Immediately after birth tachycardia was noted. An electrocardiogram (ECG) showed a heart rate of 300 beats per minute (bpm) and narrow QRS complexes.

The newborn was clinically stable. She was admitted to the neonatal intensive care unit and an intravenous line was inserted for treatment with Adenosine. While preparing for Adenosine infusion her heart rate spontaneously decreased.

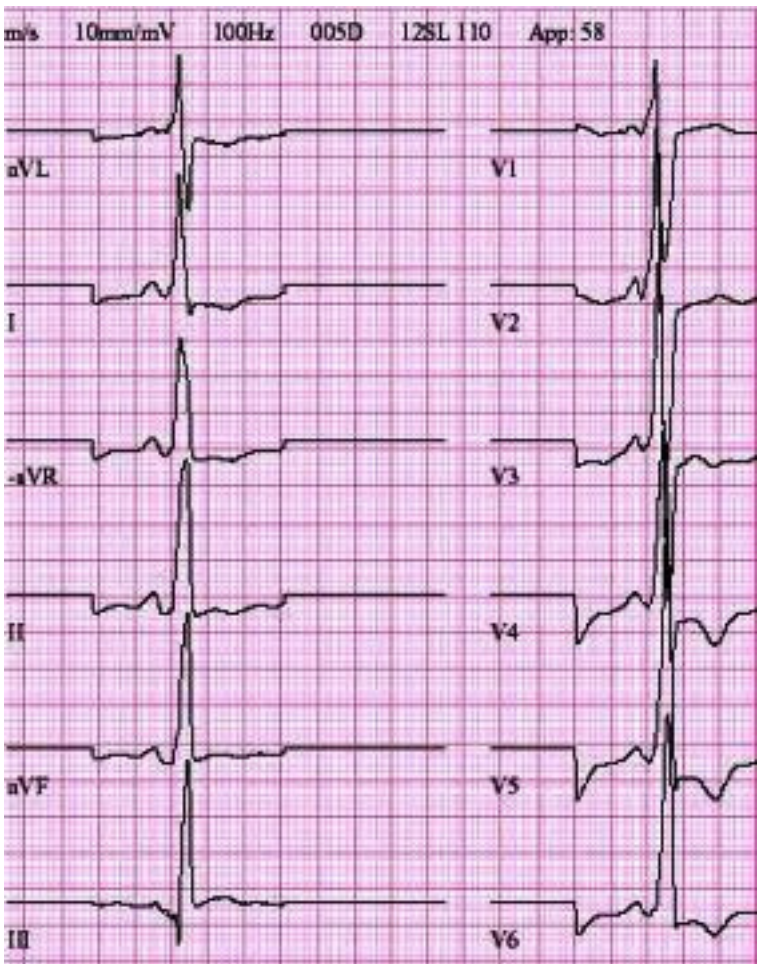
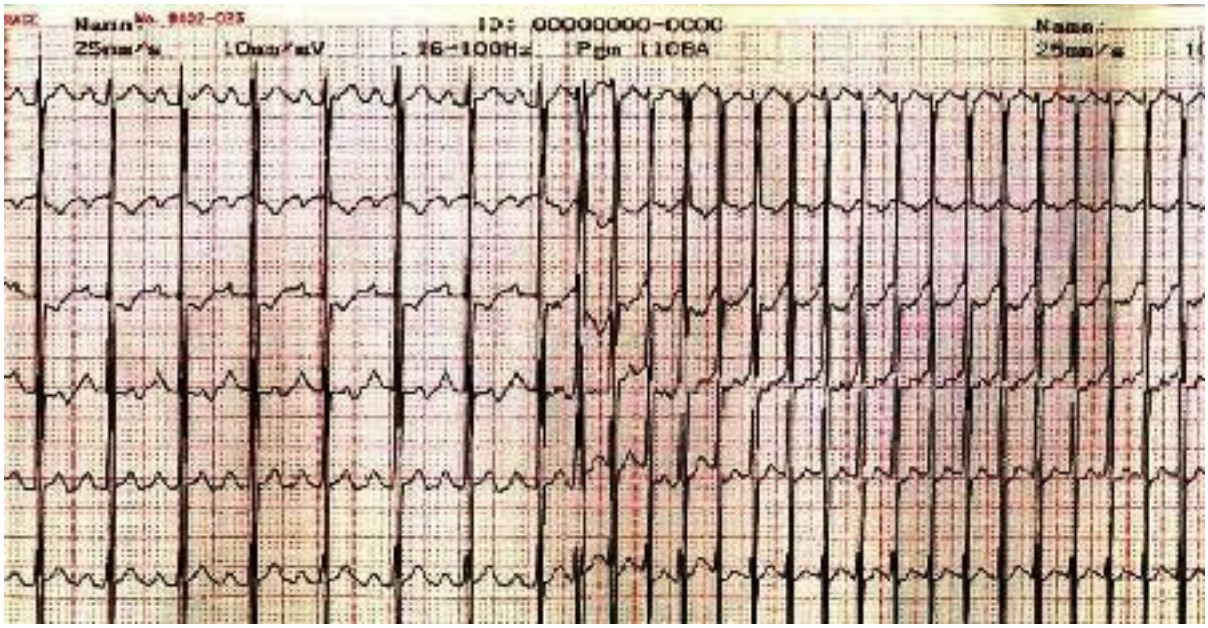


Figure 1a: ECG at birth



Her ECG at presentation with normal heart rate is shown in figure1a.

On auscultation, a soft systolic murmur was heard.

Echocardiography was performed and demonstrated a complete atrio-ventricular septal defect (AVSD) with a large primum defect, large ventricular septal defect, common atrio-ventricular orifice (AV-valve), and balanced size of the right and left ventricles.

At around two hours of age the infant started to get frequent, short episodes of supraventricular tachycardia alternating with sinuous rhythm (figure 1b).

Figure 1b: ECG at normal heart rate and at tachy-arrythmia

Prophylactic treatment against recurrent tachy-arrythmia with Digoxin was started (full digitalisation within the first 24 hours and then with a maintenance dose of 5 µg/kg, B.I.D).

The infant was observed for several days and continued to be free of symptoms. She was discharged to her home at five days of age.

She was seen five days later and was found to be clinically stable with no tachy-arrhythmia. Her ECG then is shown in figure 2.

QUESTIONS

1. What are the ECG findings and the ECG diagnosis at presentation?
2. Compare the ECG on day one (with normal heart rate, Fig 1a) and on day ten (figure 2). How can the differences be explained?
3. At what age should this infant preferably be operated on?
4. What are the expected intra-operative and post-operative complications because of her arrhythmia and what are the needed pre-operative precautions?

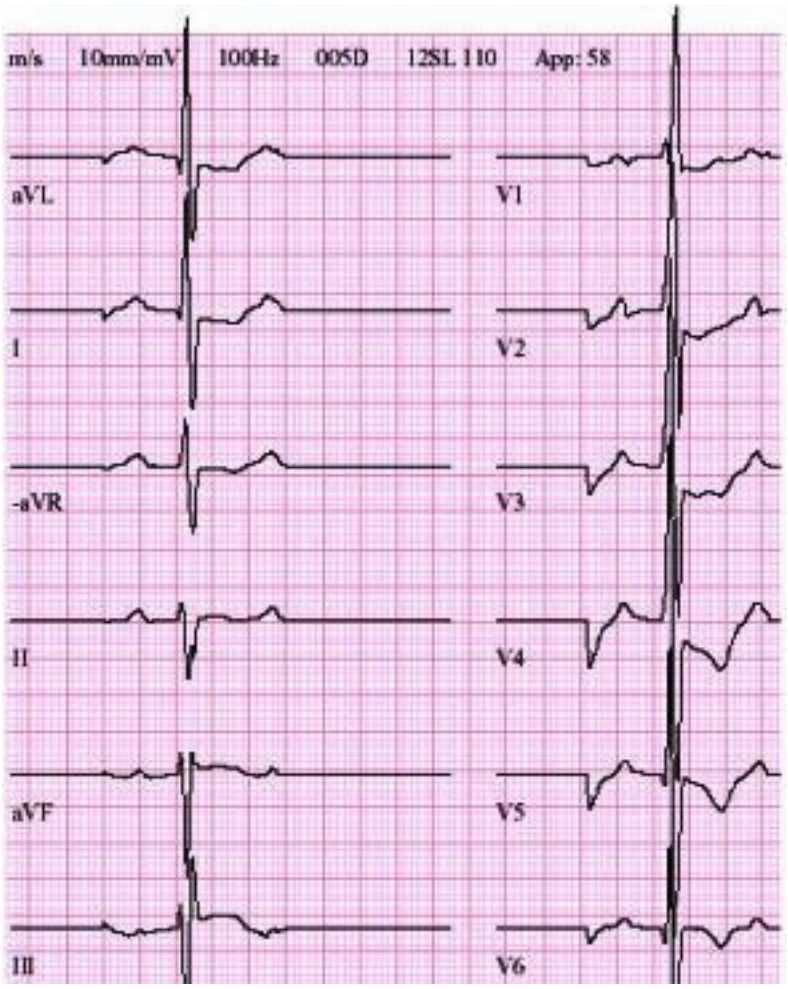


Figure 2: ECG at ten days of age

ANSWERS

1. This infant was born with complete AVSD and presented immediately after birth with supraventricular tachycardia with a heart rate of 300 bpm.
2. The ECG at normal heart rate (figure 1a) shows short P-R interval with delta wave, and prolonged QRS complex, a typical finding in Wolf-Parkinson-White (WPW) syndrome (1). The infant continues then to have short attacks of supraventricular techycardia with stable circulation. The prophylaxis with Digoxin was started with a good effect, as she continues to be free of arrhythmias.

3. The ECG at ten days of age (figure 2) shows no delta wave as seen in the ECG at birth (figure 1a). This demonstrates the intermittent nature of this finding, which is a known phenomenon in patients with WPW syndrome. The delta wave is due to the premature conduction of the atrial impulse through the accessory pathway to the ventricle at the time of ECG registration. If the atrial impulse conduction occurs through the AV-node, the delta wave was not seen in the ECG. There are changes also seen in the electrical axis of the QRS complex, as seen on the ECG without delta wave. There is a left axis deviation in the presence of AV-septum defect and no pre-excitation (figure2). This is due to postero-inferior displacement of the AV-node and early depolarisation of the ventricle in children with this cardiac malformation (2, 3). However, if the impulse is conducted through the accessory pathway, the electrical axis of the heart can be normal (figure 1a).
4. The complete AVSD comprises 2% of congenital cardiac diseases (CHD) with markedly increased incidence in children with Down´s syndrome. The surgical management of this condition is preferred to be within the first 6 months of age, to avoid the development of permanent high vascular resistance. In the presence of heart failure very early treatment may be considered. The presented infant underwent surgical treatment with total correction of her cardiac defect at three and half month of age with good result. During the induction phase of the anaesthesia she developed three attacks of supraventricular tachycardia, which were controlled with intravenous Adenosine infusion.
5. In the presence of history of arrhythmias in infants with complex CHD, the risk for difficult arrhythmias during and after surgical correction is high. One should consider this risk and properly prepare for prevention and treatment if needed. In our case this issue was discussed thoroughly. The possibility to treat infants with WPW syndrome with radiofrequency catheter ablation and the risks of such procedure, in such small babies, was studied and discussed in details (4). If needed, it can be performed before surgical correction as

a separate procedure or at the time of surgical operation (5). This was not performed to our presented infant because her type of arrhythmia can easily be treated with Adenosine and the risk for prolonged and serious arrhythmias was considered small in comparison to treatment with radiofrequency ablation in such a small child.

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