Partial Atrioventricular Septal Defect Associated With Wolff-Parkinson-White Syndrome: Perioperative Dysrhythmias During the Intracardiac Repair

Anand Mammen, Prabhat Tewari, Pallavi Horo, Shantanu Pande

Departments of Anaesthesiology and Cardiovascular and Thoracic Surgery, SGPGIMS, Lucknow, Uttar Pradesh, India

ABSTRACT

Wolff-Parkinson-White syndrome (WPW) is rarely seen in association with atrioventricular septal defect. Although paroxysm’s of palpitation due to supraventricular tachycardia can occur in these patients, rare, fatal, ventricular dysrhythmias can also occur. Herein, we report the case of a 20-year-old male patient with partial atrioventricular septal defect and WPW syndrome for intracardiac repair, developing intraoperative Torsades de pointes and postoperative cardiac arrest, adding to the difficulty in overall patient management.

Keywords: Atrioventricular septal defect, Torsades de pointes, WPW syndrome

INTRODUCTION

Atrioventricular septal defect (AVSD) is characterized by incomplete development of the atrioventricular septum along with abnormalities of the atrioventricular valve. With an incidence of 4–5.3 per 10,000 live birth, AVSD comprises 7% of all congenital heart disease.[1]

Abnormalities in the conduction system may be seen in patients with AVSD. Injury to the conduction system can also occur during surgical repair of an AVSD since the AV node and the bundle of His are displaced inferiorly.[2] Supraventricular tachyarrhythmias have been reported in the postoperative period after AVSD correction. Kharbanda et al. have reported the incidence of late life-threatening ventricular arrhythmias and sudden cardiac death (SCD) after AVSD correction as 0.5% and 1.7%, respectively.[3]

However, there is overall little data on the incidence of ventricular arrhythmias and SCD after AVSD correction.

The prevalence of electrocardiographic preexcitation is between 0.1 and 3.1 per 1000 person.[4] Nearly, 7–20% of patients with WPW syndrome have other associated congenital cardiac anomalies of which Ebstein anomaly is the most common.[5] Association of AVSD with WPW syndrome is rare.

We report the case of a patient with partial AVSD and WPW syndrome posted for intracardiac repair of the AVSD developing intraoperative supraventricular tachycardia that progressed to polymorphic ventricular tachycardia (VT)/Torsades de pointes and postoperative cardiac arrest in the cardiac surgical intensive care unit (ICU) but revived.
CASE REPORT

A 20-year-old male, weighing 45 kg presented with exertional breathlessness associated with palpitation for 10 years and occasional episodes of paroxysmal palpitation associated with syncope for 3 years. He also gave history of recurrent lower respiratory tract infection since childhood. On evaluation, his electrocardiogram (ECG) showed preexcitation pattern – WPW syndrome and 2D echo showed 11 mm ostium primum atrial septal defect (ASD), cleft mitral valve with moderate mitral regurgitation (MR), severe tricuspid regurgitation, and moderate pulmonary artery hypertension.

Catheterization study confirmed partial AVSD with >2:1 left to right shunt, posteriorly and inferiorly displaced left atrioventricular (AV) valve annulus with a cleft in left AV valve anterior leaflet and grade 1 MR with normal biventricular function. During the catheterization study, the patient developed atrioventricular reentrant tachycardia (AVRT) followed by irregularly irregular broad complex tachycardia, suggestive of atrial fibrillation with conduction through the accessory pathway that reverted to sinus rhythm with defibrillation and amiodarone infusion. The patient was started on the Tab. diltiazem extended-release (ER) 90 mg OD and elective surgical repair of the AVSD was planned.

At the time of preanesthetic evaluation, patient was hemodynamically stable with a pulse rate of 64 bpm, blood pressure (BP) of 120/70 mmHg, and oxygen saturation (SpO2) of 98% on room air. On examination of the cardiovascular system (CVS), S2 was loud, the mid systolic murmur of AVSD and pansystolic murmur of MR could be heard.

His chest X-ray showed cardiomegaly with a cardiothoracic ratio of 0.6. ECG showed regular sinus rhythm with a rate of 75 bpm, normal QT interval, short PR interval, wide QRS, and delta waves that is negative in inferior leads suggestive of preexcitation syndrome – WPW pattern with posteroseptal accessory pathway [Figure 1].

The patient was properly counseled regarding the anesthesia technique and procedure, prescribed premedication as per the institute protocol, and fasted till 6 am. It was ensured that the morning dose of Tab. diltiazem ER 90 mg was taken.

In the OT, intravenous (IV) access was secured, induction was done using Inj. etomidate 10 mg, Inj. fentanyl 250 mcg, Inj. midazolam 2 mg, Inj. pancuronium 8 mg and the patient was intubated with 8 mm internal diameter (ID), cuffed, oral endotracheal tube (OETT). Intraoperative monitoring included ECG, pulse oximetry, end‑tidal CO2 (EtCO2), invasive blood pressure (IBP), central venous pressure (CVP), transesophageal echocardiography (TEE), arterial blood gas (ABG), temperature, blood glucose, urine output, activated clotting time (ACT) and bispectral index (BIS). Anesthesia was maintained using propofol infusion, isoflurane inhalation, and intermittent boluses of fentanyl and midazolam.

After opening the pericardium, patient went into supraventricular tachycardia (SVT) that quickly progressed to polymorphic VT/Torsades de pointes [Figure 3] but reverted to sinus rhythm after 20 J direct DC shock. ABG was normal. After complete heparinisation, the patient was put on cardiopulmonary bypass (CPB). Intracardiac repair of AVSD was done by a single patch technique with the autologous pericardium, anterior mitral leaflet (AML) cleft was closed and TV repaired by pericardial felt annuloplasty. MgSO₄ infusion at 1 g/h was started on rewarming. TEE evaluation after filling on partial bypass showed residual tricuspid regurgitation and TV was repaired again after going back on full support. The patient was successfully weaned off CPB with ionotropic support’s adrenaline at 0.04 mcg/kg/min and noradrenaline at 0.02 mcg/kg/min. Total CPB time was 106 min and aortic cross-clamp (ACC) time was 60 min. The patient was shifted to ICU with ionotropic support’s adrenaline at 0.05 mcg/kg/min and noradrenaline at 0.03 mcg/kg/min with a BP of 84/40 mmHg and paced HR of 90 bpm.

On arrival in the ICU, patient went into another episode of ventricular tachycardia that soon progressed to ventricular fibrillation. CPR was initiated, 200 J DC shock, and Inj.
adrenaline 1 mg IV was given. As there was no response, sternum was immediately reopened. The dysrhythmia reverted after intracardiac massage and direct DC shock of 20–50 Joules, 4 times. On post-op day 2, the patient had another episode of VT [Figure 4] that responded to a loading dose of amiodarone 150 mg, and oral diltiazem ER 90 mg was restarted. The general condition gradually improved and the patient was discharged after 6 days of ICU stay and advised follow-up in cardiology.

**DISCUSSION**

AVSD includes a spectrum of cardiac anomalies with a common atrioventricular junction. It can be complete or partial. Partial AVSD’s can have an isolated atrial-level shunt or an isolated ventricular-level shunt, though the former is more common. In AVSD’s, surgical repair is usually accomplished with good long-term survival.

WPW syndrome is the most common form of ventricular preexcitation. Durre et al. has defined it as a “condition in which all or some portion of the ventricular muscle is activated earlier, in reaction to atrial events, than would be expected had the impulse reached the ventricle by way of the normal atrioventricular conduction system.” A short PR interval (<0.12 s), prolonged QRS complex (>0.12 s), and a slurred, slow rising onset of the QRS complex known as delta wave are the 3 main electrocardiographic features of WPW syndrome. Based on the polarity of the QRS complex on ECG, the accessory pathway may be localized to left free wall (41.2%), right free wall (19.3%), posteroseptal (27.5%), anteroseptal (6.4%), and mid septal pathway (5.5%). QRS complex that is predominantly negative in inferior leads, as in our case, is suggestive of the posteroseptal accessory pathway.

WPW syndrome may be seen in association with other congenital cardiac anomalies like tricuspid atresia, hypertrophic obstructive cardiomyopathy, mitral valve prolapse, atrial septal defect, ventricular septal defect, transposition of the great artery, coartation of the aorta, and dextrocardia, to name a few. Association of partial AVSD with WPW syndrome is rare.

Management of symptomatic patients with WPW syndrome includes antiarrhythmic drugs or catheter ablation of the accessory pathway. Electrophysiology (EP) study for locating the accessory pathway and radiofrequency (RF) ablation of the pathway is the definitive treatment. In the present case, EP study was not done preoperatively, but the patient was started on oral diltiazem.

Atrial fibrillation in the presence of an accessory pathway that conducts rapidly is potentially lethal because the rapid ventricular response may lead to ventricular fibrillation. Our patient had one such episode during the catheterization study done preoperatively. Intraoperatively, soon after opening the pericardium and handling of the heart, the patient developed SVT that got conducted through the accessory pathway and degenerated into polymorphic VT/torsades de pointes. Postoperatively, soon after arrival in the ICU, the patient had another episode of VT that progressed into VF and on post-op day 2, the patient had another episode of VT.

Preoperative EP study and RF ablation of the accessory pathway, in this case, would have probably prevented fatal ventricular dysrhythmias in the perioperative period. In the future, hybrid operating rooms with facility for EP study may benefit such patient’s in whom epicardial mapping and RF ablation of the accessory pathway can be done.
CONCLUSION

Association of AVSD with WPW syndrome is rare and management of these patients can be challenging as they are prone to fatal dysrhythmias in the perioperative period that is often resistant to conventional antiarrhythmic drugs.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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