Complete heart block in a 64-year-old female patient with unrepaired tetralogy of Fallot with AV leadless pacemaker implant

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Introduction

Tetralogy of Fallot, the most common cyanotic congenital heart disease, accounts for 7%–10% of all congenital heart diseases and overall occurs in 4–5 per 100,000 live births.[1] It is characterized by anterior deviation of the conal septum, which causes 4 anomalies: pulmonary stenosis (valvular and/or subvalvular), ventricular septal defect, hypertrophy of the right ventricle, and aorta overlapping the ventricular septum by up to 50%.[2] Post-repair of tetralogy of Fallot, can be associated with rare complete heart block but this is not typically seen in younger unrepaired patients.[3,4] We present a rare case of a 64-year-old female patient with unrepaired tetralogy of Fallot and complete atrioventricular (AV) septal defect who developed spontaneous complete AV block. She subsequently underwent successful placement of a Medtronic Micra AV leadless pacemaker (Minneapolis, MN).

Case report

A 64-year-old female patient, born in Mexico, with a past medical history of attempted but unsuccessful repair of tetralogy of Fallot, complete AV canal defect, and seizure disorder presented to an outside hospital after syncope and fall at a store that resulted in intracranial hemorrhages and a leftward midline shift in the frontal region. She decompensated neurologically, was intubated, and was given levetiracetam, lorazepam, and hypertonic saline. Atropine was also given owing to bradycardia in the 30 beats/min range. She was then transferred to our institution. Upon arrival, she was found to have a complete AV block with a wide complex escape rhythm (posterior fascicular escape, Figure 1A). She was hemodynamically unstable, requiring a dopamine infusion en route to the operating room. She subsequently developed an episode of torsades de pointes and coded in the operating room. Upon return of spontaneous circulation, a temporary transvenous pacing lead was emergently and successfully placed by anesthesiology into the right internal jugular vein. Once the patient was stabilized, evacuation of the bleed was performed. Echocardiogram revealed tetralogy of Fallot with severe pulmonary stenosis and left-to-right shunting ventricular septal defect. Given her ventricular septal defect, transvenous pacemaker was a suboptimal option given potential for embolic stroke. Both epicardial and leadless pacemaker placement were discussed with patient's family and the decision was made to proceed with leadless pacemaker implantation. The right and left femoral veins were stenotic, measuring 6 mm and 7 mm, respectively. Her left internal jugular vein measured a maximum diameter of 13 mm.

KEY TEACHING POINTS

- Unrepaired congenital heart disease can be related to eventual heart block.
- Adults with unrepaired congenital heart disease may be candidates for leadless pacemakers.
- Adequate atrioventricular synchrony can be achieved in unrepaired congenital heart disease patients with the Micra AV (Medtronic).

Pacemaker placement method

Access for the Medtronic Micra sheath challenged by stenotic right (6 mm) and left (7 mm) femoral veins was obtained via the left internal jugular vein, which was serially dilated to an outer diameter of 27F. The sheath was then positioned in the mid right atrium. Next, the Micra AV was deployed, and the deployment catheter and sheath were moved across the tricuspid valve into a proximal right ventricular/upper apex/septal position (under fluoroscopy and transvenous thoracic echocardiographic guidance). Deployment of the Micra AV into the septal location was successful

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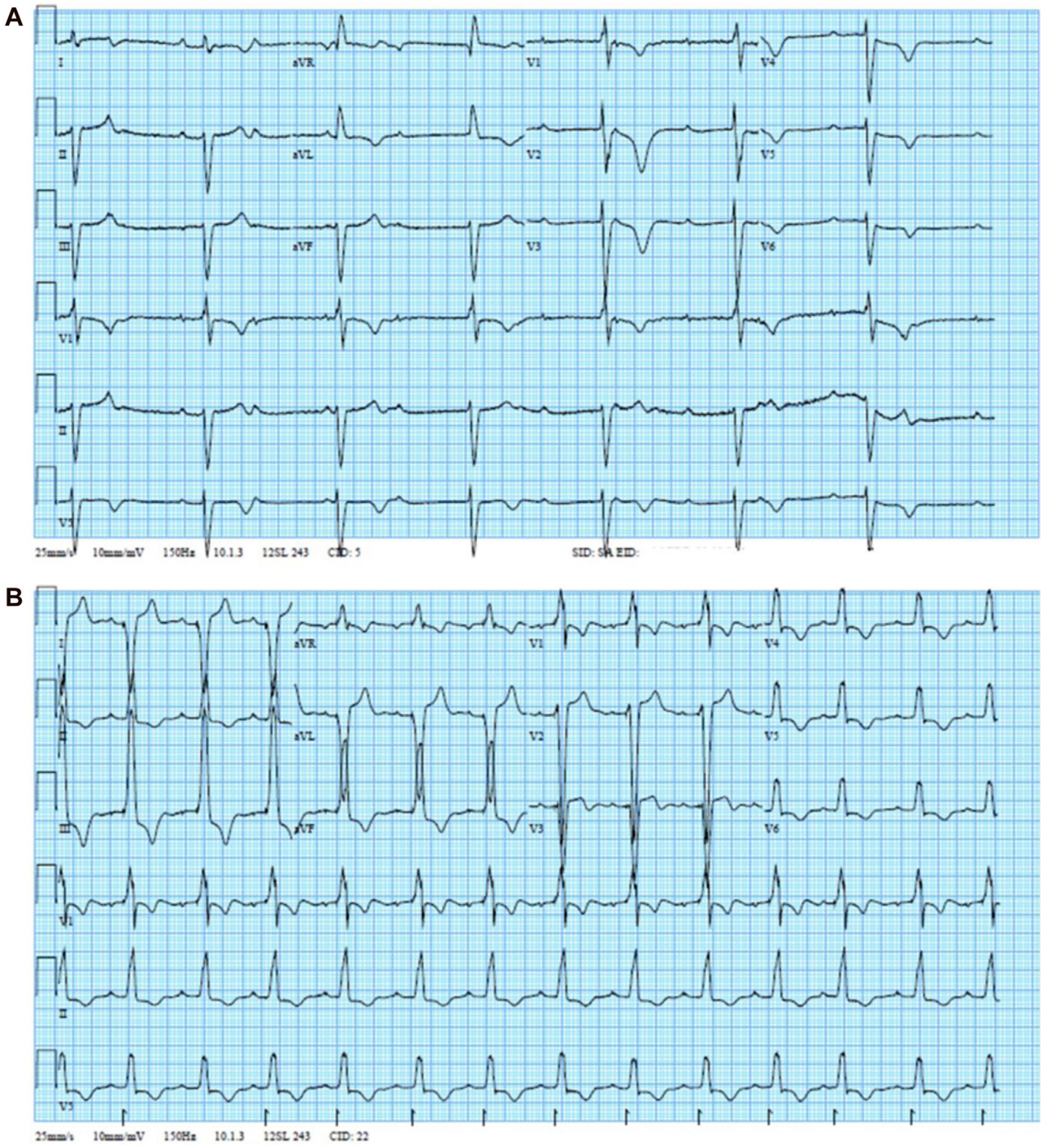

Figure 1 **A:** Complete heart block with fascicular escape at baseline. **B:** Atrioventricular synchronous pacing.

on the first attempt, with a suitable threshold of 0.25 V at 0.24 ms, R wave of 20 mV, and impedance of 920 ohms. Tug test revealed that 3 tines were connected to the right ventricular muscle, and no recaptures were needed. She was placed on anticoagulation therapy for 3 months postimplant. Post–device implant testing at 6 months revealed similar numbers, with the following being the last check: ventricular threshold 0.38 V at 0.24 ms, R wave 20 mV, impedance 920 ohms, and 92% AV synchrony and without ventricular arrhythmias or further syncope noted. Figure 1B demonstrates pacing with AV synchrony. Figure 2 demonstrates the device on radiography.

Discussion

It is well known that the risk of developing sinus node dysfunction with requirement for a pacemaker is ~29% in congenital heart disease. Complete heart block is much more rate, except in cases of discordant AV chamber connections, endocardial septal defects and with surgical repair of

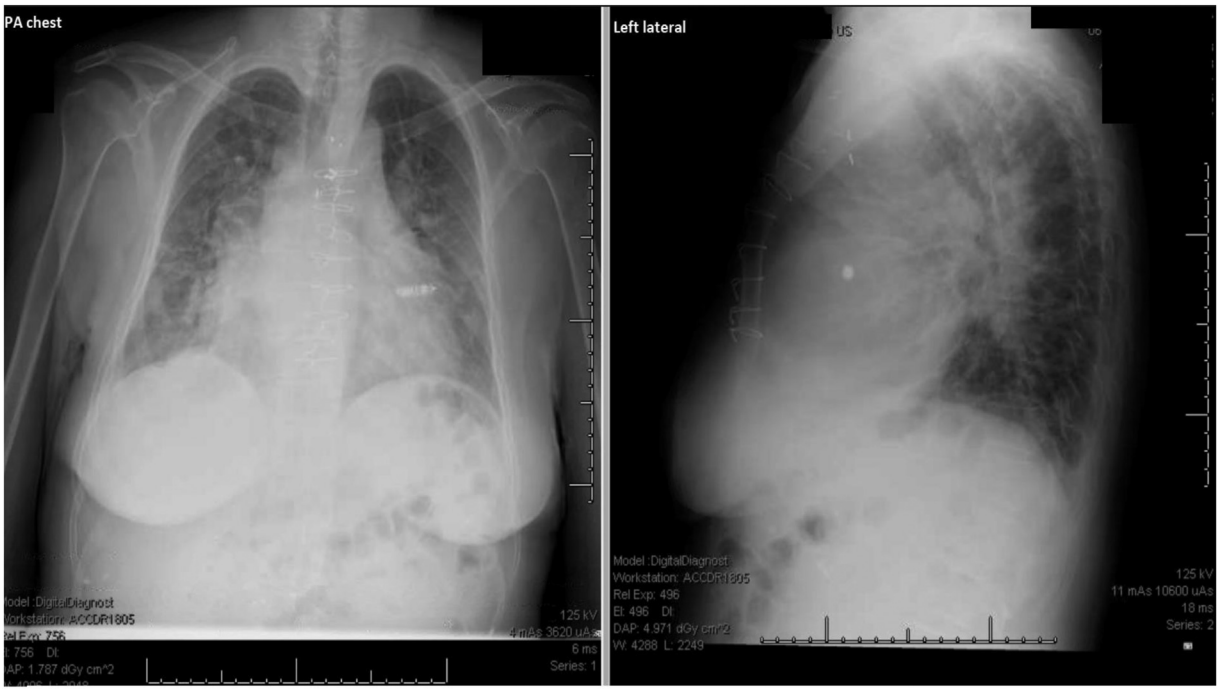

Figure 2 Chest radiograph of leadless device via posteroanterior and left lateral views.

congenital heart diseases, given that the repair involves manipulation and suturing occur at or near the AV conduction system. The reported incidence of developing complete AV block after repair of the isolated complete AV septal defect is 1.5%. In contrast, complete AV block in patients undergoing repair of tetralogy of Fallot is rarer and more unknown because only case reports have been reported.[2,4] As expected, the combined pathology of tetralogy of Fallot with complete AV canal defect, while a rare diagnosis, results in a higher incidence of patients developing complete AV block, 12.5%.[5] In comparison, the incidence of developing idiopathic complete AV block in patients with unrepaired tetralogy of Fallot with an associated complete AV septal defect is unknown because it has never been reported. In 2011, our patient had a Holter monitor that revealed sinus bradycardia with rare premature atrial contractions / premature ventricular contractions and 1 episode of ventricular couplet but did not show signs of heart block. Subsequent electrocardiograms revealed a right bundle branch block with an eventual bifascicular block but never a complete AV block. It is unclear why she later developed a complete AV block. It has been shown histologically and via computed tomography imaging that the AV conduction system is displaced inferiorly and caudally in patients with complete AV septal defect, and the simple location of the AV conduction system could have predisposed her to develop complete AV block.[6] Otherwise, regarding anticoagulation, we presumed endothelialization within 3 months of implanting the device and thus treated the patient for this course. This is of course debatable, but seemed to be reasonable after group and patient family discussion. Furthermore, given the urgency of the situation, a cardiac catheterization was not performed at the time; however, catheterization 3 years prior demonstrated right ventricular end-diastolic pressure of 10 mm Hg with systolic pressure of 63 mm Hg, while the left ventricular end-diastolic pressure was 8 mm Hg with systolic pressure of 79 mm Hg, and with QP:QS of 3. Otherwise, long-term continuing to assess for right ventricular synchrony in the setting of pacing will be important.

This is one of the oldest cases of tetralogy of Fallot described. We treated with an AV synchronous leadless pacemaker owing to the natural development of complete heart block in the setting of a sizeable intraventricular connection.

Conclusions

This is one of the oldest cases of tetralogy of Fallot described, and we treated it with an AV synchronous leadless pacemaker owing to the natural development of complete heart block in the setting of a large intraventricular connection.

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