

CASE REPORT

Dextrocardia and Situs inversus as part of Situs inversus totalis with Pulmonary and Sub-Pulmonary Stenosis, in a patient with Sinus Node Dysfunction

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Abstract:

Dextrocardia with a normal abdominal situs (situs solitus) is a rare condition. Dextrocardia with situs inversus, also known as Situs Inversus Totalis, is even rarer. It has a high incidence of associated congenital cardiac anomalies including: transposition of the great arteries, atrial septal defects (ASDs), and ventricular septal defects (VSDs) in 90 to 95% of cases.

We report a case of dextrocardia as part of situs inversus totalis with severe pulmonary and subpulmonary stenosis associated with severe bradycardia due to sinus node dysfunction and junctional bradycardia, and describe our approach in management.

Keywords: Dextrocardia. Situs inversus totalis, Pulmonary Stenosis, Atrioventricular block, Permanent pacemaker

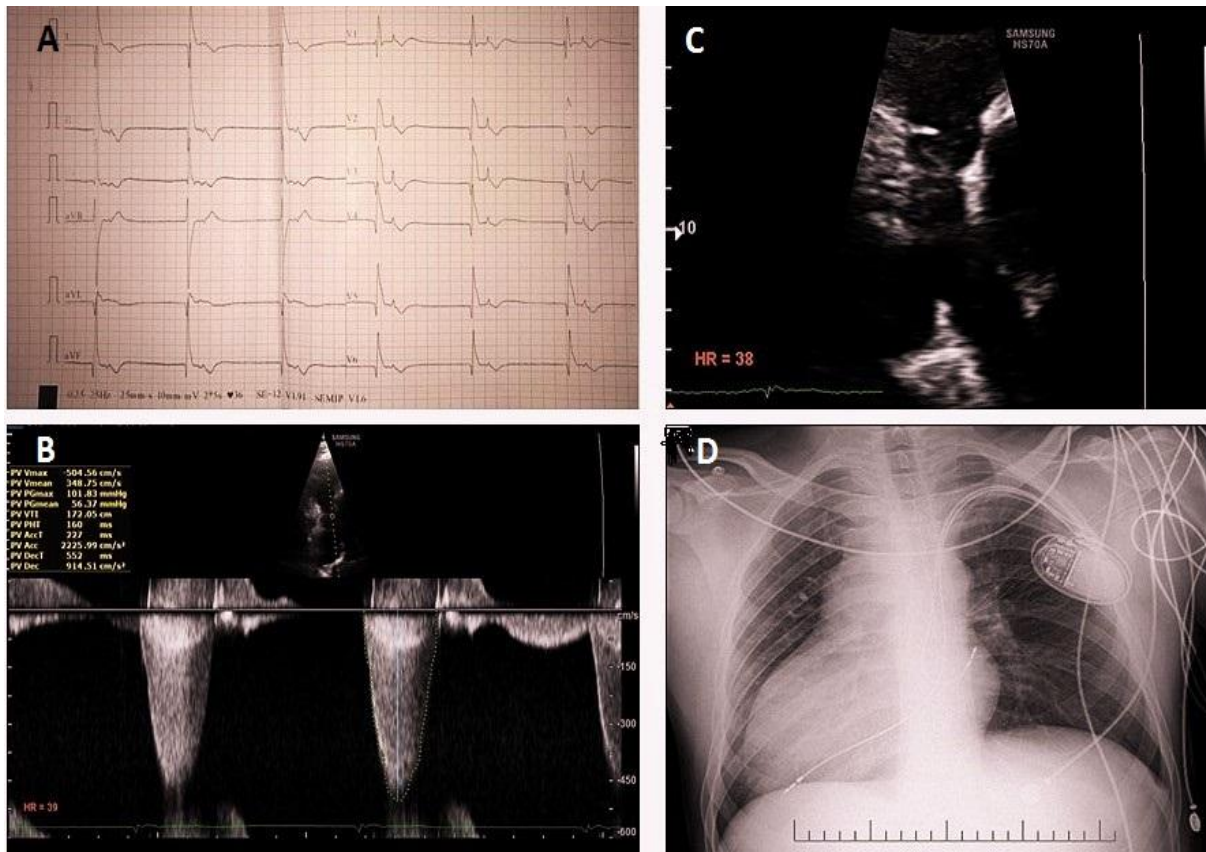


Figure 1: Panel A B C and D

Introduction:

Dextrocardia is a cardiac positional anomaly in which the heart is located in the right hemi thorax with its base-to-apex axis directed to the right and inferiorly (1). This disorder may occur independently or as part of situs inversus Totalis. Dextrocardia with a normal abdominal situs (situs solitus) has a high incidence of associated congenital cardiac anomalies (2) (3). Examples include: congenital sinus node dysfunction and junctional bradyarrhythmias, Ventricular Septal Defect (VSD), Atrioventricular Septal Defect, Patent Ductus Arteriosus (PDA), Mitral Regurgitation, Patent Foramen Ovale (PFO), Coarctation of the

Aorta, Transposition of the Great Arteries, and Ebstein's anomaly (4). The association of Dextrocardia with Pulmonary Stenosis or Membranous subpulmonary Stenosis is rare (5). To our knowledge, this is first case to report the association of Dextrocardia with situs inversus totalis, subvalvular pulmonary Stenosis and sinus node dysfunction.

Case report:

35-year-old previously healthy male presented to our hospital with 2 months history of recurrent episodes of dizziness and generalized weakness, precipitated by physical activities. He had no family history of congenital or acquired

cardiovascular disease. On examination, he had a pulse rate of 36 bpm, BP130/80 mm of Hg, the apex was located in the right 6th intercostal space (ICS), lateral to mid clavicular line. There was a systolic ejection murmur best heard at the second right intercostal space. ECG showed sinus node dysfunction with junctional escape bradycardia at 36 beats per minutes [Panel A]. Chest x-ray showed a right-sided cardiac shadow with cardiomegaly and the liver dome on the left side. Transthoracic

Echocardiography confirmed Dextrocardia; with normal left ventricular size and function and mild concentric left ventricular wall hypertrophy. There was a severely increased gradient across the pulmonary valve (peak/mean pressure gradient 101/56 mmHg [Panel B]) with suspicion of subvalvular Stenosis [Panel C] and mild to moderate pulmonary regurgitation. The right ventricle was moderately dilated with normal function and severe right ventricular wall hypertrophy. There were no intracardiac masses or thrombi, pericardial effusion or transposition of the great arteries.

Due to severe bradycardia, a temporary pacing wire was inserted via right femoral vein approach with difficulty as the wire favored entry into the left sided superior vena cava. Transesophageal Echocardiogram confirmed subvalvular pulmonary stenosis with a 3cm ridge below the pulmonary valve, the valve itself was thickened, with restricted mobility. Pulmonary trunk was dilated. There was no intracardiac shunt on agitated saline injection. Surgical Repair was recommended, which the patient chose to have done in his home country. A dual-chamber permanent pacemaker

was implanted (Biotronik®) via the right axillary vein prior to discharge with relative ease, after performing venography to select access site. [Panel D].

Discussion:

Dextrocardia has been commonly associated with sinoatrial node dysfunction (6). Successful management with dual-chamber pacemaker implantation has been reported in several cases where atrioventricular blocks were associated with dextrocardia along with other cardiac anomalies (7). Our patient had symptomatic bradycardia due to sinus node

dysfunction and junctional bradycardia and likewise underwent successful pacemaker implantation. Right femoral vein is a convenient site for temporary pacemaker placement for patient needing future permanent pacemaker insertion, but is more challenging in patients with Dextrocardia. We performed a venogram to construct an angiographic road map to determine the best access point, and selected the right axillary vein for needle puncture. Similar challenges in permanent pacemaker placement in patients with Dextrocardia have been reported (8).

To the best of our knowledge, this is the first case report of the presence of two congenital anomalies of Dextrocardia and pulmonary/subpulmonary Stenosis with sinus node dysfunction and junctional escape bradycardia. Temporary and permanent pacemaker could be challenging in Dextrocardia and might need venogram to exclude any associated venous anomalies that would preclude access to the right side of the heart for permanent pacemaker placement.

Conclusion:

Dextrocardia as part of situs inversus totalis can exist with subvalvular and valvular pulmonary Stenosis and sinus node

dysfunction and junctional bradycardia. Management in such cases is challenging and needs multidisciplinary approach.

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