Heart Block in Dextrocardia and Situs Inversus: A Case Report

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The authors report a case of a 39-year-old woman with dextrocardia and situs inversus who presented with episodes of complete heart block, managed successfully with a permanent dual chamber endocardial pacemaker.
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Dextrocardia may be accompanied by other congenital cardiac and extracardiac abnormalities.¹ There are only a few reports in the literature describing valvular and ischemic heart diseases² and only one report on complete atrioventricular block in isolated dextrocardia and ventricular septal defect.³ Only one report was found on dextrocardia with situs inversus in association with heart block.⁴

CASE REPORT

A 39-year-old woman with negative family and personal history of heart disease, diabetes mellitus, or hypertension was admitted to the hospital because of symptoms of low cardiac output and syncope. On initial physical examination, she was conscious and well oriented. Her blood pressure was 100/70 mmHg. She had a regular pulse at 37 beats/min. There were no rales in the lungs. The heart sounds were normal; no murmurs, gallop, or rub were heard. Examination of the abdomen and extremities was normal. The resting electrocardiogram showed complete heart block at a rate of 40 beats/min.

The two-dimensional and Doppler echocardiogram revealed dextrocardia with situs inversus without any other anomaly. A biochemical profile, complete blood count, serum electrolytes, thyroid

function test, and lipid, rheumatologic, and immunologic profiles, as well as urinalysis, all were within normal limits. There was no obvious cause of the heart block. The patient had a permanent dual chamber endocardial pacemaker.

DISCUSSION

By definition, dextrocardia represents a mirror image of the heart and the abdominal viscera, for which the term *situs inversus* is applied. Dextrocardia with complete situs inversus occurs in approximately 2 per 10,000 live births, among which 3% are associated with congenital heart disease. In situs inversus, there are two possible cardiac positions—mirror image dextrocardia (apex on the right) and levoversion (apex on the left). The primitive cardiac tube in the fetus begins as a straight line structure that normally undergoes an initial bend to the right (d-loop) and subsequent rotation in the leftward direction, so that the ventricular portion of the heart comes to lie in the left chest.⁵ In situs inversus, the initial bend is to the left (l-loop) and subsequent rotation is in a right direction, so that the ventricular portion of the heart comes to lie in the right chest.

Two basic variations of this pattern can occur: (1) the apex may remain on the right (dex-

Figure 1. Schematic illustration of the anatomic relationship of the descending aorta, left atrium, apex and stomach in the four basic cardiac positions. In situs solitus (normal position) the descending aorta, left atrium, apex, and stomach are all on the left. In situs inversus (mirror image dextrocardia) the descending aorta, left atrium, apex, and stomach are all on the right. In dextroversion the descending aorta, left atrium, and stomach are on the left (as in the normal) but the apex is on the right. In levoverision the descending aorta, left atrium and stomach are on the right (as in situs inversus) but the apex is on the left. (Reprinted with permission from Perloff JK: *The Clinical Recognition of Congenital Heart Disease*, Ed.2, Philadelphia, W.B. Saunders Co., 1978.)

troversion of the ventricles), while the descending aorta, left atrium and stomach remain in their normal left side positions; or (2) the apex may remain on the left (levoverision of the ventricles), while the descending aorta, left atrium and stomach remain in the situs inversus positions (Fig. 1). The situs solitus heart can be modified by dextroversion of the apex, and the situs inversion heart can be modified by levoverision of the apex. In situs solitus (and dextroversion) the atria are normally positioned, while in situs inversus (and levoverision) they are in mirror image positions. The left atrium is virtually always on the same side as the descending aorta and stomach. The most common and familiar form of dextrocardia is mirror image dextrocardia, in which the anterior-posterior relationships of the various parts

of the heart are normal but their right-to-left orientation is reversed. Its electrocardiographic changes are diagnostic, with inverted P, QRS, and T waves in lead I. The mean T vector is often rightward and the QRS loop is counterclockwise in the frontal plane. In this case report, during complete AV block, the QRS complexes are negative in AVR. In the precordial leads there are progressive increases of the R waves, which indicates the ventricle impulse originally comes from the left ventricle, positioned in the right side (Fig. 2).

During pacemaker implantation, a left subclavian vein puncture was used to introduce an atrial J-lead and the "ventricular" lead (Fig. 3). The ventricular electrode could be placed rather easily in a stable position in the right ventricle apex. The endocardial electrogram measured 9.6 mV and the

Figure 2. A 12-lead resting ECG showing complete AV block at a rate of 40 beats/min. The QRS complex in AVR is negative and in the precordial leads there are progressively increasing R waves.

Figure 3. Lateral and posteroanterior chest x-ray views showing dextrocardia with sinus inversus with a permanent dual chamber endocardial pacemaker.

threshold was 0.4 V. The atrial electrode was inserted in the right atrial appendage and the endocardial electrogram was 3 mV and the threshold was 0.9 V.

CONCLUSION

In our literature review, only one case report on heart block in dextrocardia with situs inversus was found. We report a case of dextrocardia with situs inversus that presented with episodes of prolonged complete heart block and syncopal episodes due to idiopathic degeneration of the conduction system, managed successfully with a permanent dual chamber endocardial pacemaker.

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