Pacemaker Implant in Dextrocardia with Right Superior Vena Cava and Persistence of Situs Inversus Viscerum - Case Report

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Summary

Dextrocardia with situs inversus viscerum totalis occurs in only two over 10-000 patients and it may be associated to a persistence of the right superior vena cava which drains in a dilated coronary sinus. Only few cases of pacing by endocardial approach in patients with such anatomic anomaly are reported by literature. A 71-year-old woman showing a situs inversus viscerum totalis with persistence of the right superior vena cava and a total AV block was implanted with a unipolar endocardial lead connected to a VVI pulse generator (Biotronik mod. PIKOS 01). The endocardial approach was complicated by the difficulty to go with the lead beyond the tricuspid valve, since the lead was proceeding in the accessory right superior vena cava coming into the right atrium through the coronary sinus. To overcome the obstacle due to the presence of an acute angle between the coronary sinus and the tricuspid valve, it was necessary to shape the lead mandrel in order to describe a wide "a " in the right atrium. Cardiac pacing by endocardial approach is feasible, but requires complex lead insertion maneuvers in patients with dextrocardia with right superior vena cava and persistence of situs inversus viscerum totalis.

Key Words

Dextrocardia, pacemaker

Introduction

One of the most frequent vitiums of cardiac position is dextrocardia, situation in which the heart is located in the right side of the hemithorax. In present casuistic, its incidence is of 1 over 10,000 live births [1]. In the majority of cases the dextrocardia is associated with cardiac malformations, most of them of cyanotic type [3, 5, 6]; but it may also be combined associated with extra-cardiac defects, of which the situs viscerum inversus is the most frequent as it occurs in the 3% of cases [2, 3].

Great relevance holds the concomitant presence of a venous malformation, as the persistence of right superior vena cava draining in dilated coronary sinus, the difficulties it may generate to permanent endocardial pacing. The main problems are: difficulty in positioning the lead in right ventricle and risks of lead dislocation, either in acute and chronic phase, with the possibility of an epimyocardial implant. The article describes an exceptional case of dextrocardia with persistence of right superior vena cava and situs viscerum inversus, the sole case over 6,700 pacemaker implants performed at the Cardiac Pacing Center of the Institute of Cardiac Surgery of the Bari University.

Case Report

A 71-year-old woman was sent for hospitalization in our Center with a diagnosis of syncopal episodes in patient with rhythm disorders. In anamnesis, we found arterial hypertension treated with drugs for long time; episodes of intense dyspnea and palpitations; periods of bradycardia and two syncopes during last year.

The patient's conditions at the arrival were: blood pressure 140/90, good hemodynamic compensation; complete BAV with ventricular rate of 32 bpm, narrow QRS, negative P-wave in D1 and AVL leads. Diagnosis of dextrocardia associated with situs viscerum inversus totalis (right gastric bubble and left

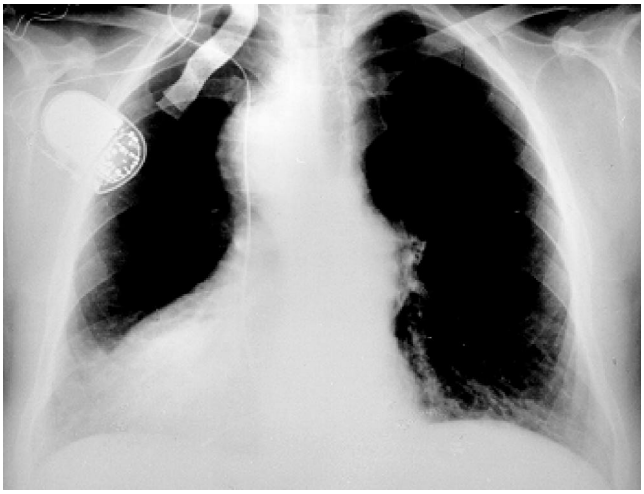

Figure 1. The anteroposterior radiogram shows the course of the lead inside the right superior vena.

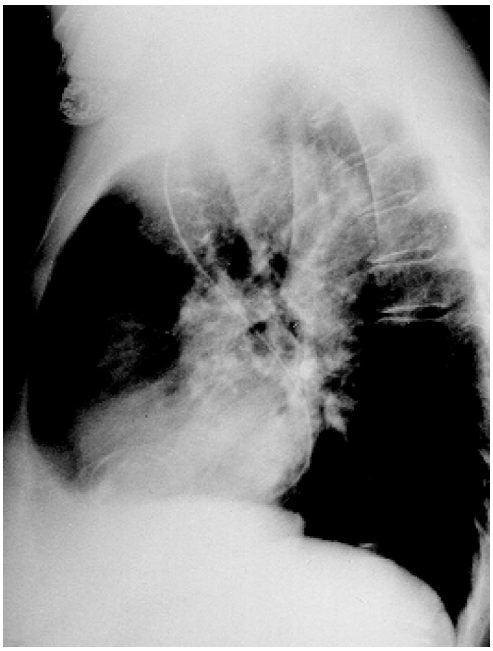

Figure 2. The left lateral radiogram shows the loop formed by the lead inside the right atrium.

hepatic shadow) was made using X-ray fluoroscopy. To avoid further risks of syncope, the patient was immediately implanted with a unipolar endocardial lead connected to a VVI pulse generator (Biotronik mod. PIKOS 01). Before the implantation, the patient was informed of her conditions and her anatomic anomaly she did not know.

At X-ray fluoroscopy, the cardiac image was located in the right side of hemithorax with apex in anterior-right side. The lead, first inserted by right cephalic access, was not able to proceed over 10-12 cm, thus, never arriving in the succlavian vein. After this, the right jugular vein was isolated and the lead was introduced till the heart but with unexpected course, since it ran on the right side of the cardiac shadow (Figure 1) and it described a complete 360° loop with very acute angle in the right atrium before proceeding inside the ventricle (Figure 2). After routine test of pacing threshold and pacemaker functions, an angiography was performed, in which it was possible to see presence of a right superior vena cava draining in a dilated coronary sinus, in dextrocardia, with right atrium and ventricle in back side.

The bi-dimensional four-chambers echocardiogram confirmed the lead course from coronary sinus to right atrium and then to ventricle (Figure 3). The patient was discharged after 5 days and now, two years later, a correct functioning of the implanted pacemaker is observed.

Discussion

Data concerning the occurrence of position vitiums of the heart and its chambers, especially dextrocardias, are discrepant, since they change in depending to the detection methodology. Some mispositions do not cause evident disease and so produce a valence lower than the normal incidence.

Nowadays the dextrocardia incidence vary from 1 over 10,000 [1] to 5 over 20,000 [2] live births; 4/5 of these patients present situs viscerum inversus, with associated heart diseases in 12-18% of cases, while 1/5 patient shows a situs viscerum solitus or ambiguus, with associated congenital heart diseases in 90% of cases [7].

The most frequent intra-cardiac malformations are: great veins and arteries transposition, pulmonary stenosis or atresia, inter-ventricular defect, single ventricle, single ventricle with double outlet [3].

The most frequent extra-cardiac malformations are: breast lobation anomalies, nasal polyps associated with bronchiectasis (Kartagener Syndrome), asplenia (Breschet Syndrome), presence of a double superior vena cava, absence of inferior vena cava [3-12].

Friedbergh and Braunwald describe three types of dextrocardia [2-14]:

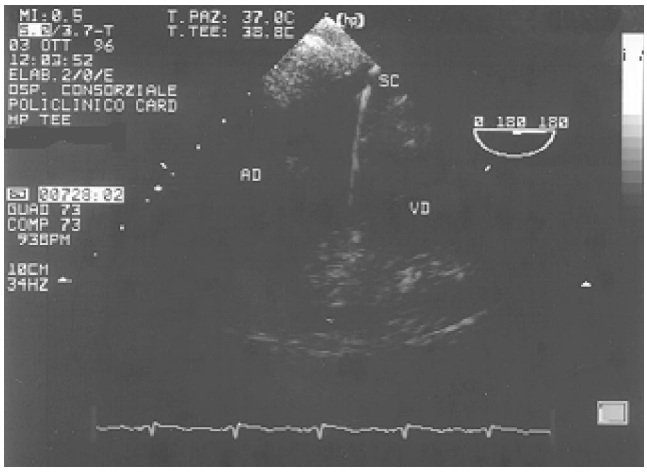

Figure 3. Four-chambers bi-dimensional echocardiography. Lead course from coronary sinus (SC) to right atrium (AD) and into right ventricle (VD) is visible in the frame. Negative P-wave electrogram detected in lead D1 are shown in the bottom of the frame.

- Type 1: dextrocardia with situs viscerum inversus totalis. In this case, there is a specular heart transposition with left atrio-ventricular cavities and apex on the right side, and right cavities on the left side;
- Type 2: insulated dextrocardia. The cardiac cavities are in the same position than type 1, and the abdominal viscera are in their normal position.
- Type 3: correct dextrocardia (dextroversion). In this case, the heart is located in the right side of the hemitorax, left cavities are anterior while right cavities are posterior. This type may be associated with congenital diseases both extra-cardiac or intra-cardiac (most of them of cyanotic type).

The presence of dextrocardia with an associated complete A-V block in an elder patient is, according to literature, a very rare occurrence and represents the sole alteration in heart location we have seen in over 6,700 patients implanted by endocardial approach. The presence of a superior vena cava draining in coronary sinus make this case extraordinary. The implant of a permanent pacemaker, in presence of a right superior vena cava (especially if insulated) is anyhow possible but it presents complications sometime serious [8]. Possible negative events are: recurrent lead dislodgements, thrombosis of the coronary sinus, occlusion of the posterior coronary vein, serious rhythm alterations during transit of the lead through the coronary sinus.

The use of endocardial coronary leads longer than 62 cm, with porous electrode tip or with active fixation, may reduce the occurrence of dislocations. If, after several attempts, it will result impossible to maintain the ventricular tip electrode in its correct location, the sole remaining possibility is the epimyocardial implant.

To perform a proper endocardial approach, an angiographic investigation is mandatory to verify the anatomic situation, i.e. to identify the position of cardiac cavities and the presence or absence of intra- or extra-cardiac anomalies.

The occurrence of a second or third degree AV Block in dextrocardia is very rare aligned with few reports in literature, while Sick Sinus Syndromes with consequent needs of permanent pacing are more frequent [10-13].

Conclusion

The implant of an endocardial pacing system in patient affected by dextrocardia, with or without venous anomalies, is possible. Some major difficulties are encountered, when a persistent right superior vena occurs.

In every case, it is necessary to perform an angiographic investigation to make implant more safe and easy, and to prevent eventual complications.

We advise to effect a bi-dimensional echocardiography before the discharge of the patient, in order to verify the real location of the electrode in right ventricle.

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