CASE PRESENTATION

Situs inversus totalis and cardiac rhythm/conduction disorders

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Abstract: Introduction – Situs inversus totalis is a rare congenital malformation, diagnosed at about 1 from 10.000 of patients. It's diagnosis is usually made incidentally. **Case report –** Female patient, 46 years old, diagnosed with dextrocardia at the age of 16 based on chest X ray, is referred to our clinic for assessement of one sincopal episode. The ECG trace at admission revealed sinus bradycardia, a heart rate of 35 beats per minute, negative T waves in precordial leads V1,V2. Transthoracic echocardiography revealed concordant atrioventricular and ventriculo-arterial connections, normal size for both ventricles, normal biventricular function and mild tricuspid regurgitation. A dual pacemaker was performed, with favorable evolution. **Disscusions –** Limited data about patients with situs inversus totalis and rhythm/conduction disorders, makes more difficult their management, being fundamental to make a correct diagnosis. The implantation technique of a patient with dextrocardia can be a very challenging one.
Keywords: situs inversus totalis, rhytm/conduction disorders, dextrocardia.

Rezumat: Introducere – Situs inversus totalis este o malformaţie congenitală rară, diagnosticată la aproximativ 1 din 10.000 de pacienţi. Diagnosticul este de obicei unul accidental. **Prezentare de caz –** Pacientă, 46 de ani, diagnosticată la vârsta de 16 ani cu situs inversus totalis, este adresată clinicii noastre în urma unui episod sincopal. Pe traseul ECG efectuat la internare se observă: bradicardie sinusală, FC=35 b/min, unde T negative în derivaţiile V1, V2. Ecocardiografia transtoracică a evidenţiat concordanţă atrioventriculară şi ventriculo-arterială, ventricul stâng şi ventricul drept cu dimensiuni normale, o funcţie sistolică păstrată şi regurgitare tricuspidiană uşoară. S-a realizat implant de stimulator cardiac bicameral cu evoluţie ulterioară favorabilă. **Discuţii –** Existenţa unor date limitate în literatură despre pacienţii cu situs inversus şi tulburări de conducere sau aritmii face dificil managementul acestor cazuri, fiind esenţială ilustrarea unui diagnostic complet. Tehnica de implant la pacienţii cu dextrocardie reprezintă o adevărată provocare.
Cuvinte cheie: Situs inversus totalis, tulburari de conducere sau aritmii, dextrocardie.

INTRODUCTION

Situs inversus, a rare congenital malformation, described for the first time by Matthew Baille. It is an autosomal recessive genetic condition, but sometimes it can be X-linked. The term situs inversus, short form of the Latin "situs inversus viscerum" describes the inverted position of abdominal and chest organs. When associated with dextrocardia is referred as situs inversus totalis[1]. There are also cases of situs inversus with levocardia, these are rare and 95% of them are associated with congenital heart diseases, most commonly transposition of great vessels[2]. About 25% of individuals with situs inversus can have primary ciliary dyskinesia, the so called Kartagener syndrome, characterized by bronchiectasis and chronic sinusitis[3]. Generally individuals are asymptomatic, unaware of their anatomical variation, being diagnosed by mistake.

CASE REPORT

A 46 year old, female patient, diagnosed with dextrocardia at the age of 16 based on X-ray fluoroscopy, was reffered to our clinic for assessment of one syncopal episode. In addition she was diagnosed with situs inversus totalis 3 years ago. Her physical examination

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was normal, with resting blood pressure in left arm of 120/60 mmHg and a heart rate of 35 beats per minute at rest. On cardiovascular examination, the apex was present in the 6th intercostal space on the right side, lateral to mid clavicular line, rhythmic heart sounds, without murmurs.

The ECG trace at admission revealed sinus bradycardia, a heart rate of 35 beats per minute, a QRS axis of 0 grd, narrow QRS complexes and negative T waves in precordial leads V1-V2 (Figure 1). The biologic investigations revealed high levels of triglyceride. Transtoracic ecocardiography showed mirror image dextrocardia, concordant atrioventricular and ventriculo-arterial connections, normal size for both ventricles, EF 55%, normal function of the right ventricle (TAPSE of 23 mm) and mild tricuspid regurgitation (Figure 2). During her hospitalization we made two Holter monitoring that revealed severe bradycardia with junctional escape rythm; a medium heart of 30 beats/minute, numerous sinus pauses grater than 3000 ms, the maximum of 5000 ms, numerous isolated premature ventricular contractions and systematized, with maximum length no more than 5 complexes. It is important to emphasize that the patient was paucysimptomatic during her monitorization. At the stress test, the patient achieved 125 W, maximum heart rate was 76 beats/minute (only 44% of her target), that showed a chronotropic incompetence; without induction of nonsustained or sustained tachyarrithmias during all stages.

Considering that the patient had a family history of sudden death (father deceased at age 49), the suspicion of arrhytmogenic right ventricular dysplasia (ARVD) was rised, so we had to establish whether the patient has indication for an implantable cardioverter defibrillator or a pacemaker. We so performed an MRI, the sistolic and diastolic function of both ventricles were normal, also the absence of fibro-fatty inflammation afecting the right ventricle helped us to rule out the diagnosis of ARVD. The MRI showed the total transposition of abdominal and thoracic viscera (the so called mirror image of the internal organs normal positioning) (Figure 3). We managed also to exclude different structure cardiac anomalies which can be associated with dextrocardia such as interventricular defects, transposition of the great vessels[4]. Therefore according to „2015 ESC Guidlines on cardiac pacing and resyncronization therapy" our patient has class I indication, level of evidenc C for pacemaker (PM) implant. The next day, under antibiotic protection, a dual chamber PM was implanted, using the left subclavicular vein access. Through the left superior vena cava, two pasive leads were implanted, one in the right ventricular apex and one in the right atrial appendage; the senses and threshold parameters were appropriate (Figure 4). Regarding the implantation technique,

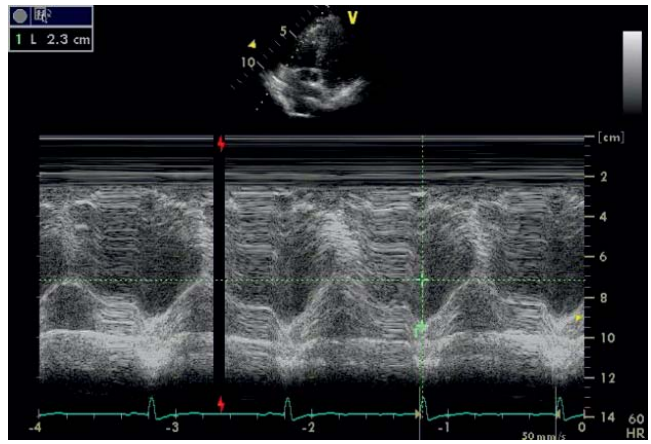

Figure 2. Tricuspid annulus systolic excursion of 23 mm.

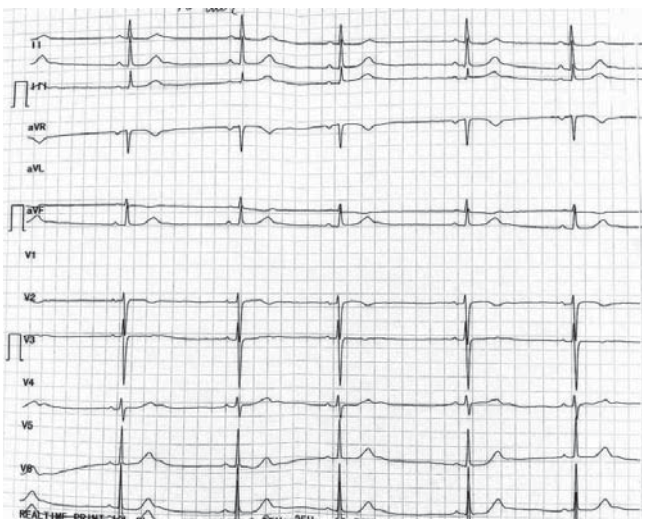

Figure 1. The electrocardiogram performed at admission-sinus bradycardia, narrow QRS complexes and negative T waves in precordial leads V1-V2.

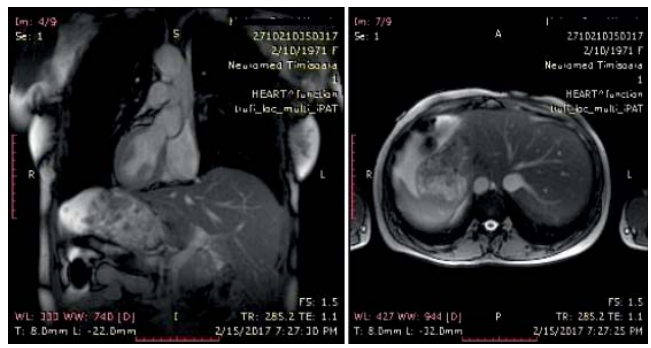

Figure 3. Mirror image of the internal organs normal positioning.

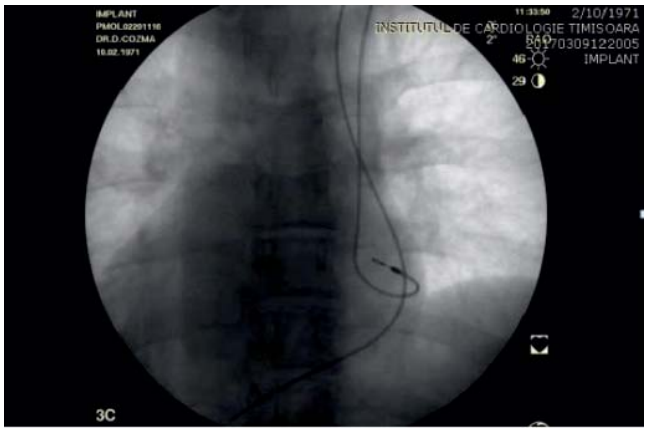

Figure 4. Through the left superior vena cava, two leads were implanted, one in the right ventricular apex and one in the right atrial appendage.

there is no clear recommendation in this case, being more a choice of the interventional cardiologist. There were no complications during and after the intervention. The pacemaker was programmed DDDR 60 beats per minute, we optimized the atrioventricular interval (a long AV delay) in order to allow the intrinsec conduction and to minimize the right ventricular pacing. A post procedural 12 lead ECG trace showed atrial pace and ventricular sensed complexes (Figure 5). As treatment we recommended antihypertensive therapy with a combination of converting enzyme inhibitor and diuretic tiazidic like (Perindoprilum/Indapamidum 5/1,25 mg a day), fenofibrate 145 mg a day.

CONCLUSIONS

Limited data about patients with situs inversus totalis and rhythm/conduction disorders, makes more difficult their management, being fundamental to make a correct diagnosis because of their individual anatomy.

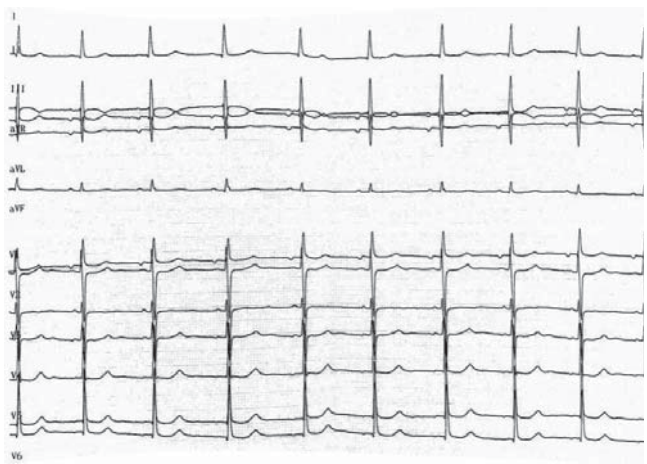

Figure 5. Post procedural 12 lead ECG trace.

The implantation technique of a patient with dextrocardia can be a very challenging one, that is why before the procedure it is important to gather informations about the anatomy and the pressence or absence of congenital abnormalities, very common at this patients[5,6]. As far as we know, few cases with situs inversus totalis who were implanted using the left vascular access were reported in literature, our patient being one of them.

Conflict of interest: none declared.
Authors' contributions: Concept/design: OM, CV, DC, LP; Data collection: OM, CV, CN, OP Data analysis/interpretation: DC, CM, SC; Drafting article: OM, CV, CN, EVG, SC; Critical revision of article: LP, CM, DC.

Abbreviations

ARVD	arrhytmogenic right ventricular dysplasia
MRI	magnetic resonance imaging
PM	pacemaker
TAPSE	tricuspid annulus planse systolic excursion
EF	ejection fraction

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