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Case Report

Atrial flutter with slow ventricular response revealing a double discordance: A case report ☆

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ABSTRACT

Congenital corrected transposition of the great arteries also called double discordance (DD) is a rare cardiac malformation characterized by the combination of discordant atrioventricular and ventriculoarterial connections. It is rarely diagnosed in the antenatal period, and is often found in adults. Clinical recognition can be challenging, so echocardiography is often the means by which definitive diagnosis is made. Most patients have associated cardiac diseases, and conduction anomalies. We reported a case of DD diagnosed at the age of 57 years. The patient spent most of his life asymptomatic until the age of 50 years old, and started presenting heart failure symptoms. Electrocardiogram showed an atrial flutter with slow ventricular rate. Echocardiography and injected CT scan confirmed the diagnosis of DD. Patient was under medical treatment for heart failure.

Introduction

Double discordance (DD) also called congenital corrected transposition of the great arteries is an uncommon congenital heart defect.

Classically, DD implies both ventriculoarterial discordance (embryologic L-looping resulting in an anterior and leftward aorta arising from the right ventricle [RV], with pulmonary artery posterior and rightward from the left ventricle [LV]) and atrioventricular (AV) discordance (ventricular inversion, meaning right atrium emptying into the LV through a mitral valve, and left atrium emptying into the RV through a tricuspid valve).

The defect is therefore « corrected » because of the physiologic flow of blood through the body.

Ventricular septal defect (VSD), subvalvular and/or valvular pulmonary stenosis, or abnormalities of the systemic atrioventricular valve with coexisting accessory pathways are the main abnormalities met in DD.

As a consequence of the unusual position of the atrioventricular node and course of the atrioventricular conduction bundle, heart rhythm and conduction disorders and supraventricular tachycardias have been reported.

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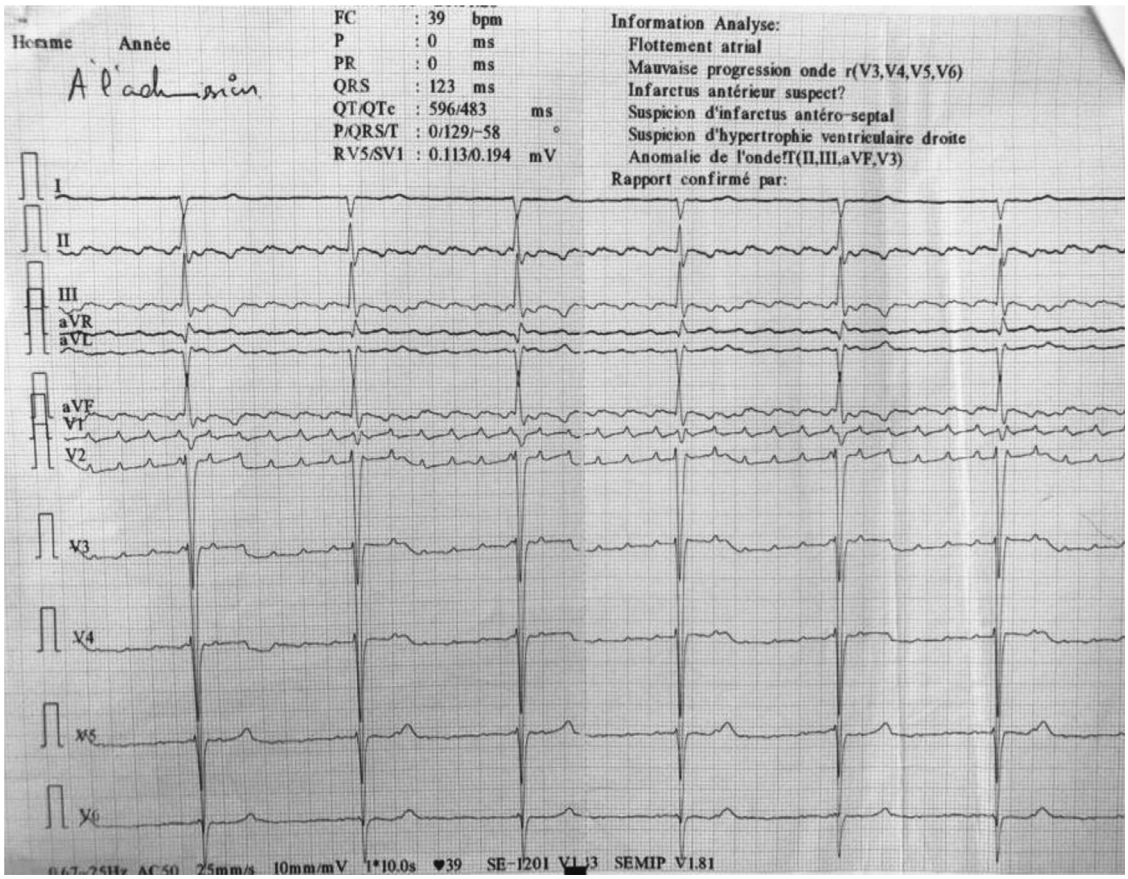

Fig. 1 – Admission ECG showed atrial flutter with a ventricular rate of 39 bpm.

We describe a case of DD diagnosed at the age of 57 years. The patient spent most of his life asymptomatic until the age of 50 years old, and started presenting heart failure symptoms.

Electrocardiogram showed an atrial flutter with slow ventricular rate. Echocardiography and injected CT scan confirmed the diagnosis of DD.

Case report

The patient was a 57-year-old male who presented to the emergency room with asthenia and lower-extremity edema. He had a medical history of several hospitalizations for heart failure.

He was conscious, with shortness of breath stage 4 of the NYHA, and reported no chest pain. His blood pressure was 12/8 cmHg, heart rate was 41 bpm. Respiratory rate was 18/min and oxygen saturation was 99% on ambient air.

Admission electrocardiogram showed an atrial flutter with slow ventricular rate of 39 bpm (Fig. 1).

Transthoracic echocardiography revealed a situs solitus, levocardia configuration. Functional LV (RV) is dilated to 38 mm/m^2. Ejection fraction of the functional LV (RV) was 43% by simpson. RV (LV) function was altered on TAPSE and the tricuspid S wave. Left ventricular outflow tract cannot be seen on the standard long axis parasternal view. With apicalization of the left-sided systemic AV valve compared to the right-sided AV valve, representing the tricuspid and mitral valve, respectively. The valvular structures are slightly altered with mild regurgitation in both of them. The atria are dilated but empty of echoes. No venous return abnormalities were noted (Figs. 2 and 3).

Computed tomography showed congenitally corrected transposition of great arteries with AV, ventriculoarterial discordance (Figs. 4 and 5).

The patient managed medically with heart failure treatment. After discussion with the cardiac team, surgery was not an option. Then the patient scheduled for flutter ablation but unfortunately succumbed before the procedure.

Discussion

Double discordance is a complex condition that accounts for 1% of congenital heart disease [1]. Anatomically, DD is defined by an aberrant connection between a right (left) ventricle with a left (right) morphology, without trabeculations. Then, from this right ventricle, anterior, comes out the pulmonary artery (PA). From the left ventricle, posterior, with right morphology (trabeculated), comes out a large non-dividing vessel (aorta) [2]. The situs is most often solitus, but it can be inversus, dex-

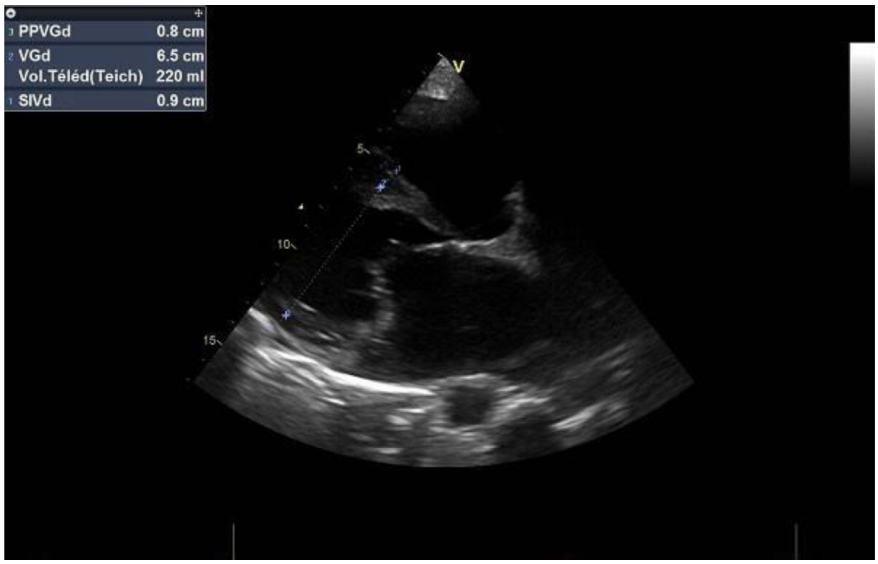

Fig. 2 – Transthoracic echocardiography, standard PLAX where left ventricular outflow tract cannot be seen.

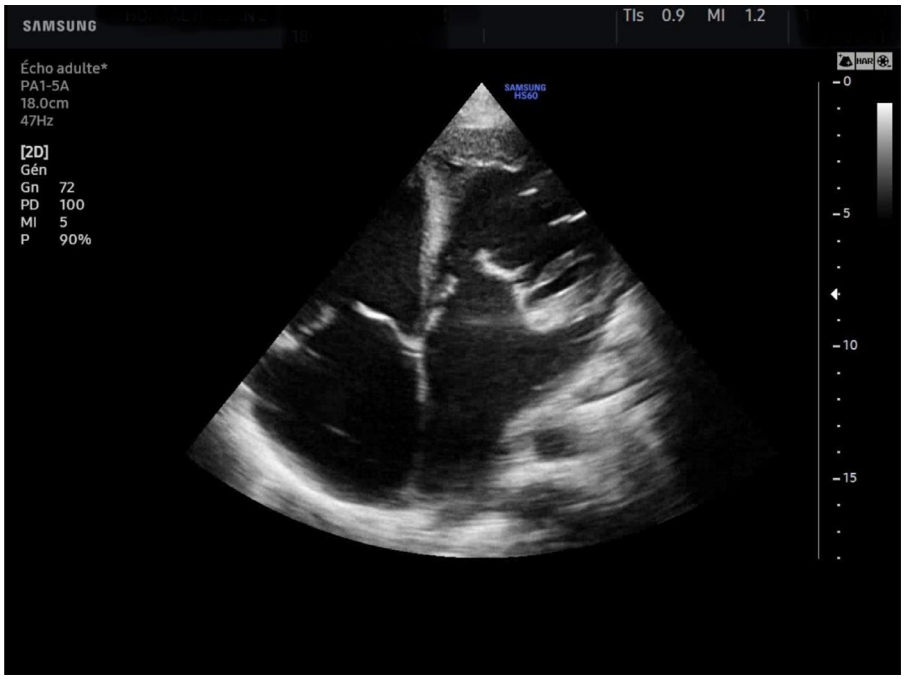

Fig. 3 – Apical 4-chamber view: situs solitus, levocardia configuration, and double discordance.

trocardia can be found in 20% of cases and represents a reason for discovery in adults.

In 90% of cases, there are associated anomalies such as VSDs (60%), pulmonary valvular or subvalvular stenosis (50%), complete atrioventricular blocks (2%), and anomalies of the tricuspid valve (tricuspid insufficiency, Ebstein's disease) [3].

In our observation, the double discordance was associated with cardiac conduction disorder (AV block), without valve abnormalities or septal defects.

The absence of associated anomalies allows prolonged survival into adulthood [4]. Several cases of DD described in the literature have been discovered in octogenarians [5].

In the Mayo study [4], involving 44 patients with DD, the mean age was 44 years old, and the circumstances of discovery were variable. The initial presentation was dextrocardia in 16% of cases, and a conductive disorder was present in 32% of cases.

Associated malformation such as VSD was present in 20% of cases, subvalvular pulmonary stenosis in 32%, and systemic atrioventricular valve anomalies in 59% of cases [4].

In our observation, the disease was discovered at the age of 57 and was isolated.

The electrocardiogram can show supraventricular rhythm disorders (fibrillation, flutter, atrial tachycardia) during DD,

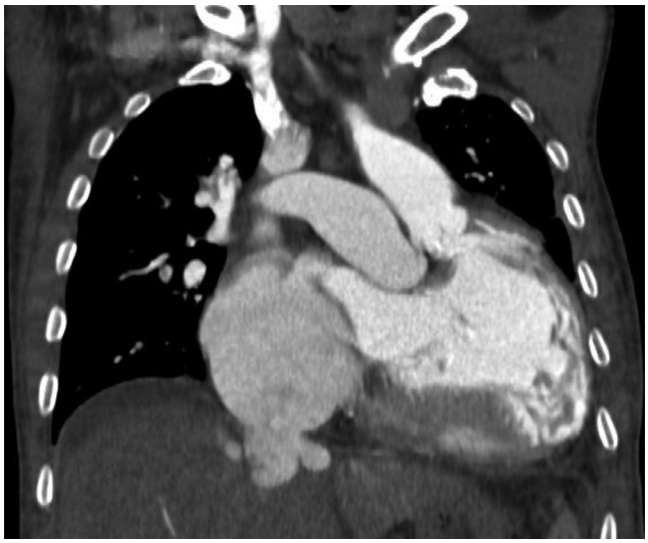

Fig. 4 – Injected thoracic CT scan: the origin and the path of aorta to the left of the pulmonary artery.

generally appearing from the age of 50 years if the DD is isolated [6], which is the case of our patient.

Conduction abnormalities in the DD are not related to the age of the patient. They can affect the entire conduction system.

In the absence of associated malformations, the patient may remain asymptomatic for many years, and not manifest until much later in life with progressive AV-block.

In our case, we don't know if the AV block was present at birth or was acquired with time.

Besides a clinical suspicion of DD, made by data of medical history, physical findings, chest X-ray, and ECG provided information. Non-invasive imaging is indicated for the final diagnosis.

Echocardiography is the key diagnostic modality, demonstrating DD and identifying associated anomalies.

A systematic approach is necessary for the echocardiographic examination. The study should specify several elements to confirm the diagnosis:

- Cardiac position.
- Identify the atrioventricular discordance.
- Morphology of the ventricles, great arteries, atrioventricular, and ventriculoarterial connections.
- Coexisting anomalies: Ebstein-like malformation of the TV, tricuspid regurgitation, ventricular septal defects, left ventricular outflow tract obstruction (LVOTO), and pulmonary stenosis.
- Systolic RV and LV function.
- Evaluation of tricuspid and mitral regurgitation.
- Assessment of TR severity.

Transesophageal echocardiography is helpful, in adult patients with suboptimal transthoracic windows, for the mitral and tricuspid valve morphology, any atrial septal communication, LVOTO, or endocarditis vegetations.

According to Jimnez [7], evaluation of the RV systolic function is based on the longitudinal velocity of the tricuspid annulus (RV S') by tissue Doppler. In our case, there was RV dysfunction (Fig.).

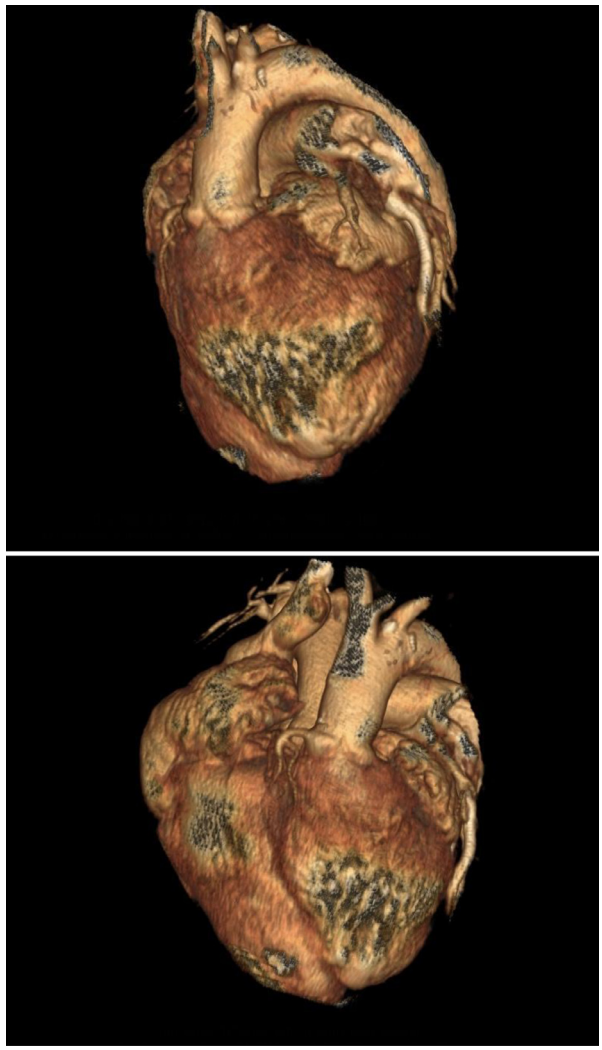

Fig. 5 – 3D reconstruction showed the anterior position of the aorta.

Cardiac MRI (CMR) provides excellent anatomic detail, intracardiac and great vessel anatomy, and is indicated for quantification of ventricular volumes, mass, and EF.

For adult patients in whom CMR is not feasible, cardiovascular computed tomography is helpful as an alternative modality for the diagnosis, and particularly useful for coronary artery anatomy.

Our patient underwent a cardiac injected CT with 3D reconstruction, which confirmed the echocardiographic data.

Patients with isolated DD have variable natural history. It depends on 3 factors [8].

- RV dysfunction and failure.
- Progressive tricuspid regurgitation.
- Arrhythmias (complete AV block, ventricular tachycardia).

Therefore, without surgical intervention, RV that is anatomically mismatched to generate systemic flow, so it will fail in the longer term.

These profoundly abnormal RV loading conditions also predispose to tricuspid valve regurgitation which tends to worsen with time.

Progressive congestive heart failure is due to decompensation of the RV and/or tricuspid valve in DD.

The mortality rate is approximately 20% in subjects over 18 years old with a mean age of death between 30 and 35 years [3,4]. RV dysfunction and heart failure are frequent from the third decade and their occurrence depends on the associated anomalies.

In addition, the risk of heart failure around the age of 45 is 67% in case of DD with associated anomalies versus 25% in isolated forms. RV dysfunction appears most often in the fourth decade and is assessed to be about 50% in complex forms versus 30% in isolated forms [3,4]. Our patient already had RV dysfunction.

Conclusion

CcTGA or DD is an uncommon congenital heart defect, often coupled to numerous cardiac abnormalities and conduction disease.

Clinical recognition can be challenging, and echocardiography is the means by which diagnosis is made.

Natural history varies, isolated form may go unnoticed until adolescence or late adulthood.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

And the patient stated that he gives his full permission for the publication, reproduction, broadcast, and other use of photographs, recordings, and other audio-visual material of himself (including of my face) and textual material (case histories) in all editions of the above-named product and in any other publication (including books, journals, CD-ROMs, online, and internet), as well as in any advertising or promotional material for such product or publications.

He declared, in consequence of granting this permission, that he has no claim on ground of breach of confidence or any other ground in any legal system against—author's/developer's name—and its agents, publishers, successors, and assigns in respect of such use of the photograph(s) and textual material (case histories).

He hereby agreed to release and discharge (author's/developer's name), and any editors or other contributors and their agents, publishers, successors and assigns from any and all claims, demands or causes of action that he may now have or may hereafter have for libel, defamation, invasion of privacy, copyright or moral rights or violation of any other rights arising out of or relating to any use of my image or case history.

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