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Triple valvular attack in situs inversus totalis

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SUMMARY

This clinical case is about a triple valve replacement, in a patient with a Situs Inversus Totalis (SIT), intervened at the University Hospital of Caracas, Venezuela, with a satisfactory result. It is a very rare case, and currently few cases were published in the world literature. A mitro-aortic valve replacement with biological valves and implantation of a semi-rigid ring (MC3) in the tricuspid annulus previous valvular plastia was performed under extracorporeal circulation. The result of the surgery was successful with the patient referred to its locality of origin, for definitive cardiological control.

Key words: *Situs inversus totalis, mitro-aortic valve.*

RESUMEN

Este caso clínico trata sobre un reemplazo de válvula triple, en un paciente con un situs inversus totalis (SIT), intervenido en el Hospital Universitario de Caracas, Venezuela, con un resultado satisfactorio. Es un caso muy raro, actualmente se han publicado en la literatura mundial algunos casos. Se realizó un reemplazo valvular mitro aórtica con válvulas biológicas y la implantación de un anillo semirrígido (MC3) en el anillo tricuspídeo, previa plastia valvular bajo circulación extracorpórea. El resultado de la cirugía fue exitoso con el paciente referido a su localidad de origen, para el control cardiológico definitivo.

Palabras clave: *Situs inversus totalis, válvula mitro aórtica.*

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INTRODUCTION

Situs inversus totalis (SIT) is a rare clinical entity characterized by completely reverse anatomy of the organs of the thoracic and abdominal cavities, finding these in a mirrored image (1). Its estimated incidence is 1 to 2 per 20 000-50 000 live births (2), affecting approximately 0.01% of the population. All organs are involved, but it is more evident in impair organs. Dextrocardia was first described by the Italian physician Marcus Aurelius Severinus in 1 643 (3). However, the situs inversus was later described by the English physician Matthew Baillie in 1793, as with the first identification of the transposition of large vessels (TGV) (4). The exchange of psychomotor brain laterality may also be a common case of Situs Inversus (Figure 1) (4).

This congenital condition may go unnoticed until it is discovered during emergency surgery or in the investigation of symptoms of a pathological condition that requires immediate

Figure 1. Anatomical drawing of Situs inversus totalis. Source: Alfred López | @yelqtls | Madrid

attention. Dextrocardia, on the other hand, is a situation in which the heart is in the right half of the chest due to the wrong rotation of the heart tube, within the second month of pregnancy. It has an embryological cause and is more common than SIT.

Patients with this congenital situation cannot be considered as carriers of defects or abnormalities since they do not have any symptoms or discomfort and their diagnosis is the product of clinical findings at the time of presenting a true pathology in their body, unrelated to their congenital state, needing to consult a physician.

Only a few cases of inversus situs that came to require valve replacements for acquired heart disease are reported in the world literature, with single, one-valve, the most frequent replacements. Those reported to have required double valve replacement are less common; Such is the case of a 38-year-old male patient in India, who was a carrier of dextrocardia and situs inversus totalis, that required double mitro-artic replacement because of a rheumatic etiology valvulopathy (5), with another similar case in a 43-year-old male patient, reported by Haldar et al in 2018 (6). In the reviewed literature, only one case of inversus situs with intervention in three valves was found, in a 79-year-old male patient who had a double mitro-aortic replacement and a tricuspid ring annuloplastia, published in Japan in 2017 (7). In most reported cases, the authors claim an inverse positioning of the principal surgeon (left side) from patient habitus.

Of 3 393 patients intervened by the lead author with cardiovascular pathologies, 1 282, were for valve replacements. Of these, 25 patients had triple valve surgery, five (5) a mitro-aortic-tricuspid valve replacement, and the other twenty (20), had a mitro-aortic valve replacement and a De Vega tricuspid plastia or annuloplasty with an implant of rings or bands designed for a diameter reduction on the tricuspid annulus.

CASE REPORT

This is a 63 years old female patient, nulliparous, with an acquired valvular heart disease, very symptomatic, that consults a primary medicine practice of her region of origin in the interior of the country, in a very clinically advanced pathological situation and despite this, it went months before being referred to the University Hospital of Caracas (UHC), specifically to the Cardiology Service. The only study conducted in the center of origin was a chest X-ray, which because unidentified (right or left) was misinterpreted (Figure 2).

Figure 2. Patient chest X-ray, highlighting the reverse situs.

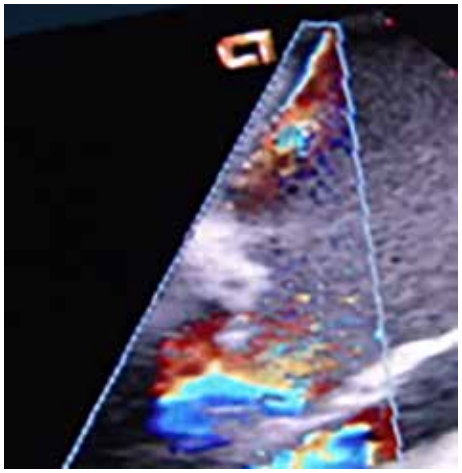

Figure 4. Mitral valve in long-axis view, with an eccentric regurgitation jet.

At UHC, the necessary studies were conducted which were conclusive. A severe calcified bivalva aortic stenosis is seen in echocardiographic findings with max 5.43 m/sec, an average gradient of 86 mmHg, and an orifice of 0.2 cm^2 (Figure 3). A severe mitral insufficiency with thickened and calcified valves whose flow slide through the atrial septum with an eccentric regurgitation jet towards the free wall of the left atrium, reaching the roof of the auricle (Figure 4), dilated tricuspid ring (Figure 5) with a moderate pulmonary hypertension of 53 mmHg.

Figure 5. Tricuspid valve in the long-axis view. Dilated tricuspid ring and insufficiency jet.

Figure 3. Aortic valve in long-axis view, severely calcified.

The case is reviewed and is suggested as surgical resolution with amitro-aortic valve replacement and tricuspid valve replacement or repair, according to intraoperative findings of the latter, following surgical criteria. The surgery took place on June 18th of 2012, positioning of the main surgeon on the left side of the patient was proposed, which is routinely done on the right side because it would make it difficult to space the spatial sense of the structures to intervene since

these were distributed in mirror image. This is an important recommendation, as it facilitates surgical development in this rare situation.

In addition to this deference, from anesthetic and perfusion point of view, there was no indication or suggestion other than conventional surgery of this category. It was performed in normothermia, with direct retroplegia through the coronary sinus. The anatomically left atrium was evident. For the mitral approach, an auriculotomy was performed through the anatomically right and subsequently transeptal atrium, being exposed to the coronary sinus for the transient fixation of the cannula for the administration of retroplegia as well as the tricuspid annulus for valvuloplasty and implantation of the reductive ring. The aortic valve replacement was made by classic technique way by italic Saortotomy. There was no surgical event to highlight. The anoxic time was 2 hours with 43 minutes, although the implant of the tricuspid ring was made at beating heart. The postoperative evolution was satisfactory; she had an uneventful postoperative course, with extubation at 10 hours, and discharged from the Intensive Care Unit (ICU) at 24 hours and on the tenth postoperative day from in-hospital care.

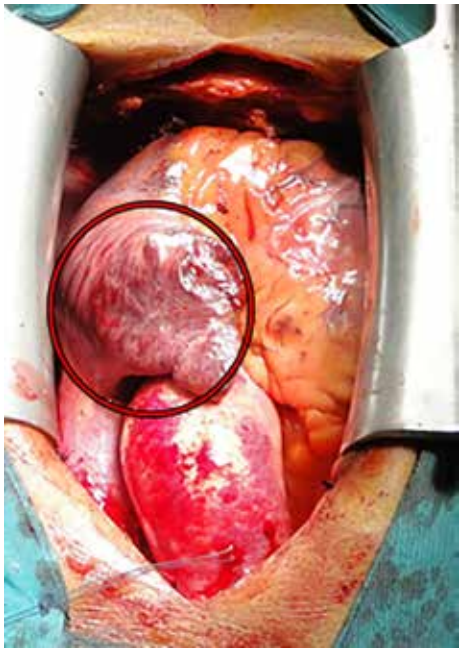

Figure 6. Taken during surgical act from the position of the anesthesiologist (Superior: Caudal; Lower: Cephalic).

Figure 6 corresponds to a picture taken during the surgical act, where the top is caudal and the lower part is cephalic. The right atrium morphologically and anatomically is located on the left side, on the side where the main surgeon is positioned in this case, corresponding to a mirror image.

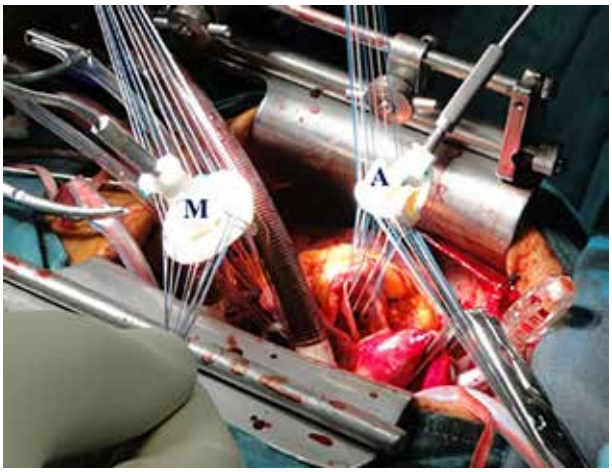

Figure 7. Left: Caudal, Right: Cephalic. It allows seeing the cardiotomies performed; with the alignment to be implanted in its natural annulus the grafts chosen that replaces the native mitral valve (M) on the left, and aortic to the right (A). Without SIT, the orientation is inverse.

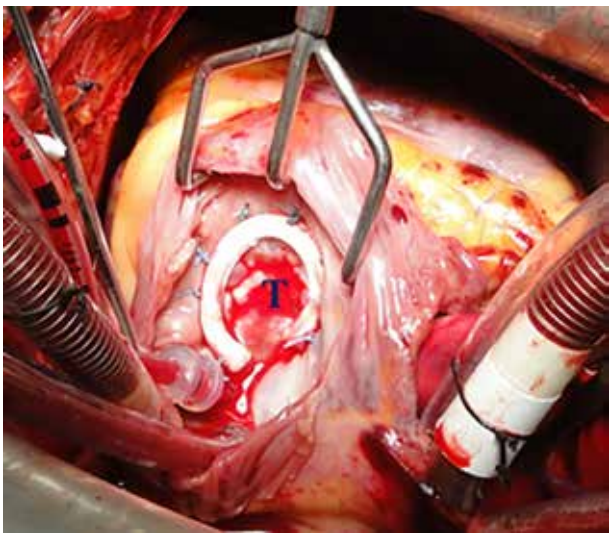

Figure 8.

Finally, Figure 8 shows the synthetic ring implanted in the tricuspid orifice (T) before a valvular plastia, decreasing the diameter, this allows the cusp in the systole to make contact with each other and the preoperative valvular insufficiency is corrected.

DISCUSSION

The objective pursued and achieved with this procedure as with any patient, is a successful result. This was the first intervention of this type achieved in the country and one of the few reported worldwide, coinciding in the published literature the reversal of the usual position of the surgeon and the rest of the surgical team. The novelty is not to have achieved the triple replacement that is already not a routine intervention but having done it in a patient with a situs inversus totalis (SIT).

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