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Damus-Kaye-Stansel surgery in a patient with tricuspid atresia, transposition of the great arteries, and type A aortic arch interruption

Cirugía de Damus-Kaye-Stansel en un paciente con atresia tricuspídea, con transposición de grandes arterias e interrupción de arco aórtico tipo A

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The tricuspid atresia is described as an absence of connection between the right atrium and right ventricle, it's classified based on the relationship of the great vessels, the existence or absence of pulmonary stenosis, and the characteristics of the ventricular septal defect[1]. The association with interrupted aortic arch type A is rare. The actual treatment for these patients is palliative, specifically in those with D-transposition of the great vessels and left obstructive outflow tract, the Damus–Kaye–Stansel procedure (DKS) associated

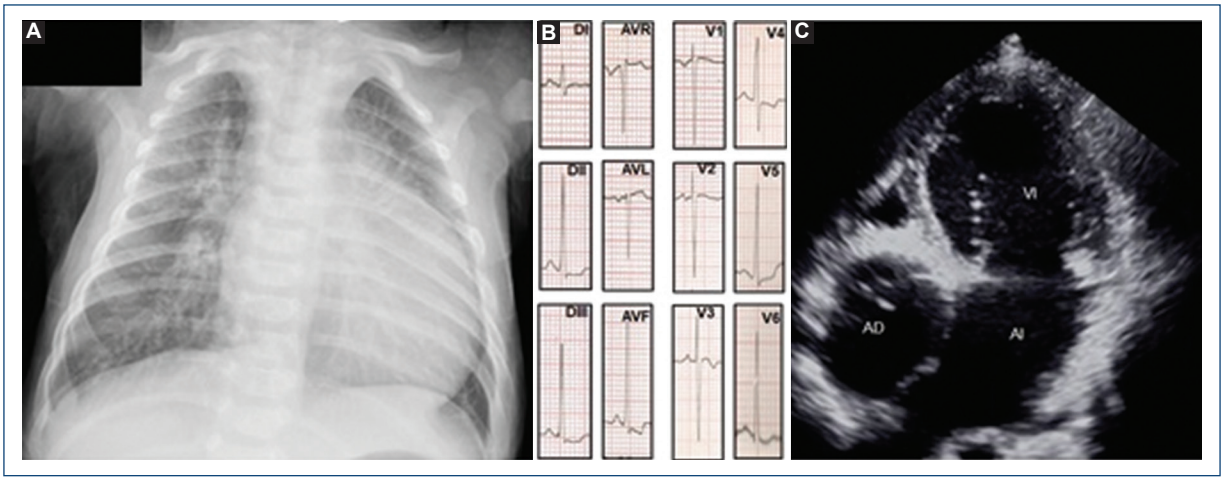

Figure 1. A: x-ray with cardiomegaly at the expense of the left cavities and increased pulmonary blood flow. **B:** electrocardiogram: Sinus rhythm, rS pattern in V1 and V2, left ventricular hypertrophy and left ventricular diastolic overload. **C:** echocardiogram: apical four-chamber image showing absence of the right AV connection.

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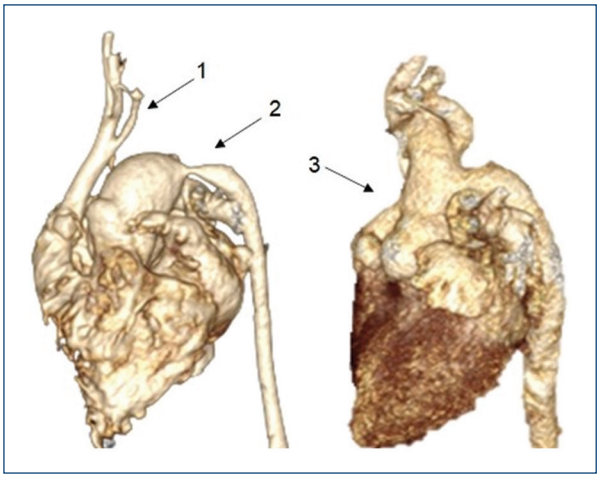

Figure 2. 3D computed tomography reconstruction. On the left: the type A aortic arch interruption (arrow 1) and the descending aorta connected to the highly dilated pulmonary aorta by a small patent ductus arteriosus (arrow 2) can be seen. Right: 3D post-surgery Damus–Kaye–Stansel reconstruction (arrow 3) 2 years after the procedure.

with Blalock–Taussig shunt is indicated, a surgery with high morbidity and mortality[2]. In this procedure, the pulmonary trunk is sectioned and anastomosed laterally (or posteriorly, depending on the position of great vessels) to the aorta to create a single ventricular outflow pathway, and pulmonary flow is ensured by performing a Blalock–Taussig shunt, this is the first step to take the patient to univentricular physiology in the future[3-5].

We present the case of a 4-year-old male patient with a history of dyspnea and diaphoresis since the age of 2 months. The physical examination showed respiratory distress, left parasternal regurgitant systolic murmur, single and intense second sound, hepatomegaly, and wide pulses. The X-ray with cardiomegaly and increased pulmonary blood flow. The electrocardiogram showed sinus rhythm, left ventricular hypertrophy, and decreased J point in V5-V6. Echocardiography and tomography diagnosed absence of the right atrioventricular connection with wide atrial septal defect, restrictive ventricular septal defect, ventriculoarterial discordance, interruption of the aortic arch type A, and a non-restrictive, 3 x 4 mm patent ductus arteriosus with the right-to-left shunt, which translates suprasystemic pulmonary pressure (Fig. 1). He was taken to correction of aortic arch interruption with end-to-end anastomosis, atrioseptectomy, and DKS with a 5 mm systemic-pulmonary shunt, with adequate postoperative evolution without post-surgical pulmonary hypertension, and discharged 3 weeks after (Fig. 2). This extremely rare association of congenital heart disease treated with DKS, to our knowledge, has been reported in less than 10 times in the literature[2,3,6-8]. Despite the complexity in the management, he has remained stable in the 2-year follow-up with good quality of life.

Funding

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Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript, nor for the creation of images, graphics, tables, or their corresponding captions.

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