CASE REPORT

Aortopulmonary window: Direct closure? A rare procedure.

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The aortopulmonary window is a rare congenital heart disease, characterized by a communication between the pulmonary artery and the ascending aorta in the presence of two separate semilunar valves. Early recognition in mandatory and surgical closure is indicated at the time of diagnosis, before the onset of irreversible pulmonary vascular disease. A case of type I aortopulmonary window without pulmonary vascular disease in childhood, repaired by direct closure without extracorporeal circulation is described in the present report.

Key words: **Aorta; Aortopulmonary window; Congenital heart disease; Direct closure; Pulmonary artery.**

La ventana aortopulmonar es una cardiopatía congénita rara, caracterizada por una comunicación entre la arteria pulmonar y la aorta ascendente en presencia de 2 válvulas semilunares separadas. La intervención temprana y el cierre quirúrgico están indicados al momento del diagnóstico, antes de la aparición de una enfermedad vascular pulmonar irreversible. A continuación, presentamos el caso de un escolar con ventana aortopulmonar tipo I sin enfermedad vascular pulmonar, corregido mediante cierre directo sin circulación extracorpórea.

***Palabras clave*: Aorta; Ventana aortopulmonar; Cardiopatía congénita; Cierre directo; Arteria pulmonar.**

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The aortopulmonary window (APW) is a rare congenital heart disease, representing between 0.1% and 0.6% of all congenital heart disease [1,2]. It is characterized by a communication between the pulmonary artery and the ascending aorta in the presence of 2 separate semilunar valves [1]. It can be present as an isolated lesion, although it is more commonly associated with other cardiovascular abnormalities, especially disruption of the aortic arch [2]. Clinical symptoms are related to pulmonary blood flow and the size of the defect between the ascending aorta and the pulmonary artery [3]. APW can be classified into three main types: type I (proximal), above the sinus of Valsalva; type II (distal), in the distal ascending aorta and type III (total), in the entire ascending aorta; the most common being type I [1-4]. Surgical repair is indicated at the time of diagnosis, before the onset of irreversible pulmonary vascular disease [1, 2, 5].

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CLINICAL CASE

We present herein a 6-year-old male with a history of patent ductus arteriosus closure at one year of life and a one-year history of dyspnea. On physical examination, he showed generalized cyanosis, hyperdynamic precordium, intense second noise and continuous murmur in the left parasternal region. The chest radiograph showed pulmonary hyperflow and the right pulmonary branch enlarged. The echocardiogram reported a type I APW with a diameter of 20 mm, with dilation of the left chambers, ascending aorta and left branch of the pulmonary artery, with systolic pressure of the pulmonary artery at the systemic level. The study protocol was completed with computed tomography identifying the defect and coinciding with the echocardiographic diagnosis (**Fig. 1**). Surgical correction was decided through conventional sternotomy and without cardiopulmonary bypass (CPB) support. When identifying the vascular structures (**Fig. 2A**), dissection and lateral clamping of the ascending aorta and the pulmonary artery trunk were performed followed by APW section (**Fig. 2B**) (**Fig. 2C**), the edges of each vessel were sutured with polypropylene 4-0 USP (**Fig. 2D**) (**Fig. 2E**), finally hemostasis and the location of the structures were verified (**Fig. 2F**). The patient had a satisfactory postoperative evolution and is being followed by the outpatient clinic with a good prognosis

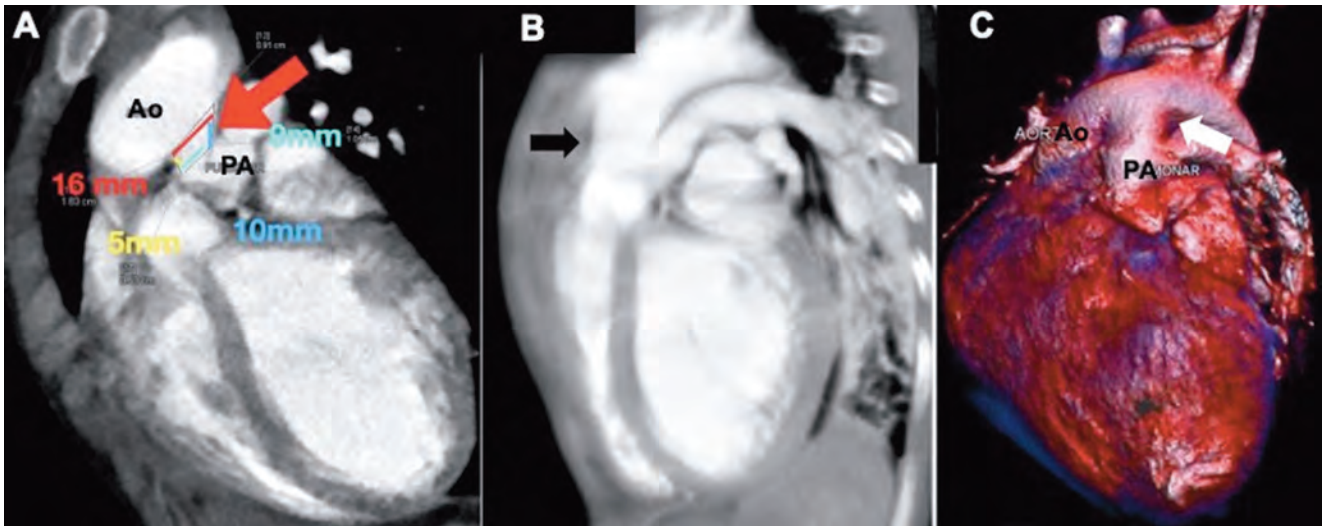

Figure 1. Computed tomography with sagittal reconstruction, shows: A) APW measurements (red arrow) and B) the relationship with the valves and bifurcation of pulmonary branches (black arrow). C) 3D reconstruction shows type I morphology (white arrow). Ao: Aorta, PA: Pulmonary artery.

COMMENT

APW was first described by Ellioston in 1830 [1], and it was until 1952 when Gross performed the first surgical correction through a ligation and 16 years later Wright performed the first surgery with CPB [1,3]. Since then, a spectrum of surgical techniques that evolved according to the needs and technologies of each era have been described, from simple ligation, through section and suture without and with CPB support, transpulmonary closure with CPB, trans-window closure (anterior sandwich patch closure) and direct transaortic closure with CPB [4,5].

The main objective of surgical repair is to avoid pulmonary vascular injury that leads to irreversible pulmonary hypertension, being the main cause of mortality in the first year of life in these patients, reaching 50% [1,3]. In this sense, in most cases the procedure is performed with CPB,

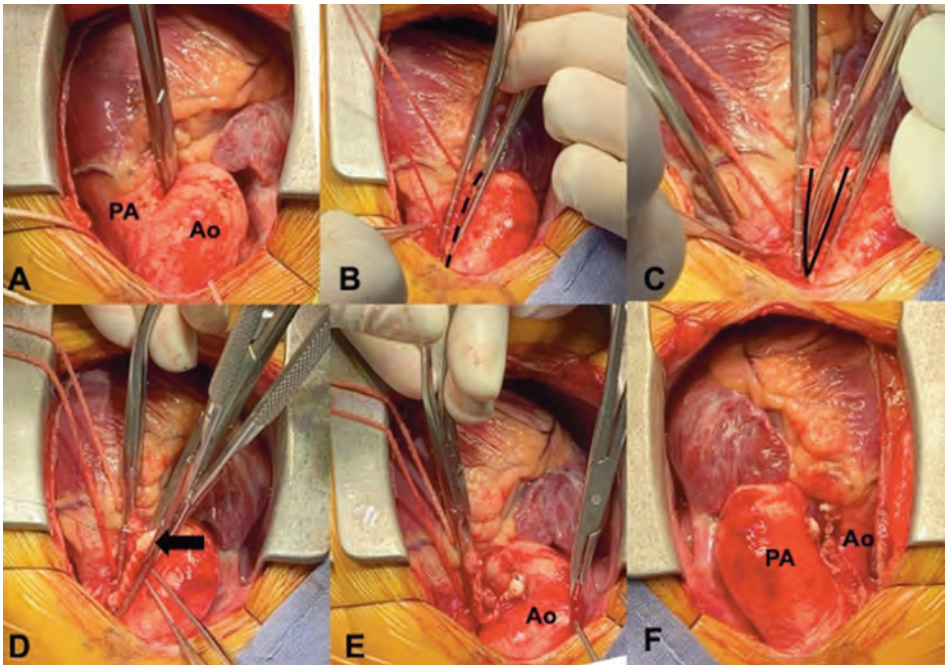

Figure 2. Surgical sequence: A) identification of vascular structures, B) lateral clamping of the ascending aorta and pulmonary artery (APW in dotted line), C) section of the structures (black line), D) closure of the lateral wall of the aorta (black arrow), E) pulmonary artery lateral wall closure, F) separated aorta and pulmonary arteries. Ao: Aorta, PA: Pulmonary artery.

I feel the transaortic technique is the one of choice [1,3-5]. However, in selected cases where the defect is considered type I, with a relatively small size and located at a safe distance from the bifurcation of the pulmonary arteries and semilunar valves, closure with section and suture without CPB can be chosen [1, 2].

The main advantage of direct closure of the aortopulmonary window is to avoid the effects and complications that the use of CPB may bring. Nasir et al. in a period of 5 years demonstrated an excellent surgical and follow-up result in a dozen of patients operated for a period of 5 years using direct closure and without CPB [1]. A similar article was published by Chen et al. [2] and Bhan et al. [5], in whom this type of correction was made [2,5]. However, the latter suggests sectional and suture closure rather than simple ligation due to the potential for residual defects and the likelihood of pulmonary artery distortion [5].

In our case, we performed the direct closure in a school patient, who was older than that reported in the literature, who underwent an adequate trans-surgical evolution and had a follow-up without complications, as commented by other groups that used the closure with section and suture.

In conclusion, we consider that direct closure is safe and with excellent results in the short and long term in selected patients, demonstrating an alternative in patients who cannot use CPB.

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