CASE REPORT

Interruption of Type A aortic arch with severe hypoplasia of the aorta. A case report

Carlos Jiménez-Fernández, Dalia Feliz-Alcántara, Jaime López-Taylor, David Ramírez-Cedillo, Miguel A. Medina-Andrade, Italo Massini-Aguilera, and Rocio A. Peña-Juárez.

Department of Cardiovascular and Thoracic Surgery, Hospital Civil de Guadalajara "Fray Antonio Alcalde". Guadalajara, Jalisco, MÉXICO.

The interruption of the aortic arch is a rare pathology, with an incidence of 0.003 per 1000 newborns. Type A presents an interruption between the origin of the left subclavian artery and the descending aorta. We present the case of a newborn with cardiogenic shock secondary to type A aortic arch interruption with severe aortic hypoplasia. The patient underwent aortic advancement. It was found severe hypoplasia of the aorta, requiring placement of a Dacron graft.

***Key words:** Aortic Arch; Congenital heart disease; Pediatrics.*

La interrupción de arco aórtico es una patología infrecuente, con una incidencia de 0.003 por 1000 recién nacidos. El tipo A presenta interrupción entre el origen de la arteria subclavia izquierda y aorta descendente. Presentamos el caso de un recién nacido con choque cardiogénico secundario a interrupción de arco aórtico tipo A con hipoplasia severa de aorta. Se sometió a cirugía para realizar avance aórtico. Se encontró una hipoplasia severa de la aorta, requiriendo colocación de injerto de Dacrón.

***Palabras clave**: Arco aórtico; Cardiopatía congénita; Pediatría.*

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Interruption of the aortic arch is an infrequent pathology, with an incidence of 0.003 per 1000 newborns, associated with 14% of patients with Di George syndrome [1]. It is characterized by a complete separation between the ascending and descending aorta. In many cases it is accompanied by ventricular septal defects and patent ductus arteriosus as an obligate defect. In addition to aortic arch hypoplasia, bicuspid aortic valve, mitral valve anomalies, truncus arteriosus, and obstructions in the left ventricular outflow tract have been described. There are three types. However, regardless of the type of aortic arch interruption, it is important to remember that this is a critical heart disease; that is, dependent on the ductus arteriosus with high mortality if it is not diagnoses in the first days of life [2].

CLINICAL CASE

A newborn of 2 days of life which began with data of cardiogenic shock. At the physical examination, a second unique and strong sound stands out, significant decrease in pulses in the lower limbs with respect to the upper ones. Imaging studies confirmed type A aortic arch interruption with severe aortic hypoplasia (**Fig. 1**). In a medical surgical session, it was decided to perform aortic advancement. During the surgical procedure, severe hypoplasia of the aorta with ductal descending aorta was found out (**Fig. 2**). A 2mm remnant was cut that goes to the left subclavian artery to the descending aorta. Therefore, a side to end anastomosis was performed with a 6mm Dacron graft (**Fig. 3**). Aortic cross-clamping time was of 18 minutes. In the immediate postoperative period, he presented hemodynamic instability, with signs of cardiogenic shock. The postoperative echocardiogram confirmed a patent anastomosis with no obstructive gradient due to the descending aorta.

COMMENT

It is considered that of the 130 million newborns born worldwide each year, more than one million are born with some type of congenital heart disease, which constituted a serious public health problem worldwide [3]. The incidence of severe congenital heart disease is approximately 2.5 to 3 per 1000 births [4]. The interruption of the aortic arc is an infrequent pathology, representing 1% of heart diseases, being associated with high early mortality [5]. The first reported case with this anomaly dates back to 1818 by Seidel et al [6].

Celoria y Patton described anatomical variants of this pathology based on the distal or proximal relationship of the interruption of the aortic arch with the origin of the 3 great

Corresponding author: Dra. Rocío Alejandra Peña Juárez.
email: alepejz@gmail.com

Figure 1. Cardiac tomography with reconstruction showing the interruption between the communication of the left subclavian artery and the descending aorta.

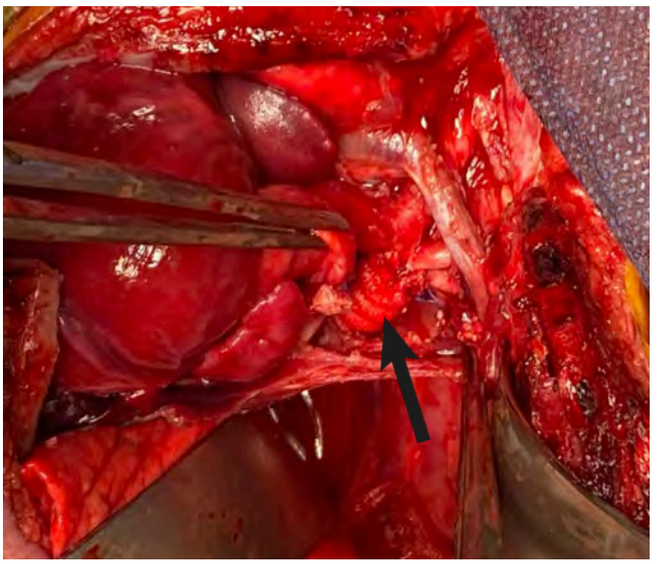

Figure 3. Dracon 6mm graft placement. Arrow indicates the Dacron graft.

vessels. Type A (15%), the interruption occurs distal to the left subclavian artery an many sometimes it can be confused with a critical coartation. As in our case, where the lack of communication between the left subclavian artery and the descending aorta can be observed in the Computed Tomography (CT) angiography (**Fig. 1**). Type B (80%), the interruption occurs between the left common carotid artery and the left subclavian artery, commonly associated with Di George syndrome. Type C (5%), the interruption is located between the origin of the innominate and the left common carotid [7].

The main associated cardiac anomaly is the interventricular defect due to posterior desviation of the conal septum (which can contribute to the obstruction of the left ventricular outflow tract). Other anatomical anomalies include hypoplasia of the aortic ring and/or bicuspid aortic valve with fusion of the commissures, and atrial septal defects [8]. In our case, it is associated with severe hypoplasia of the long, tubular descending aorta, which makes surgical repair difficult.

Surgical mortality depends of the associated anomalies, weight, type of interruption and if the type of correction is made in one single or two steps. Currently, one stage correction is considered to be preferred. However, the decision depends on the experience of the surgical team and the patient´s anatomy. In our case, it was decided to perform aortic advancement with the objective of making an incision in the proximal aortic arch, and performing an end to side anastomosis between the descending aorta and the arch.

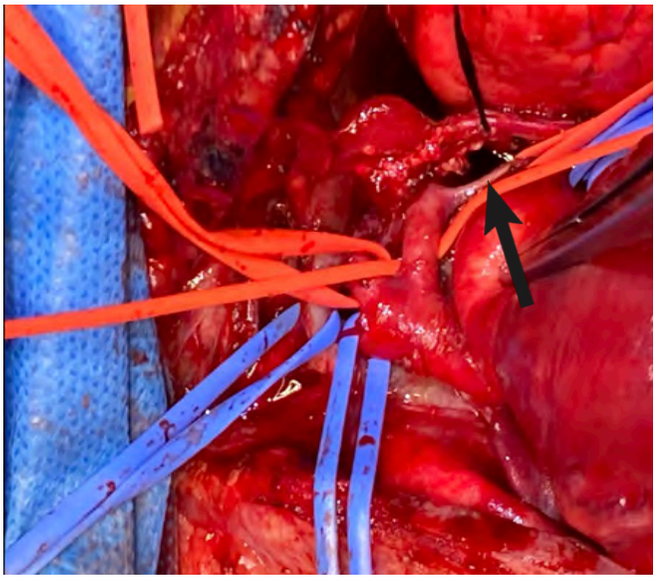

Figure 2. Severe hypoplasia of the aorta. Arrow indicates descending aorta

The intraoperative findings were severe hypoplasia of the aorta with a wide ductal descending aorta. Thus, it was decided to perform a side-to-end anastomosis with a 6mm Dacron graft (**Fig. 3**). The placement of a graft in the neonatal stage is extremely rare. In fact, this is a debatable issue since some authors such as Sell et al. [9] reported that direct anastomosis and early surgery increase the risk of persistent stenosis of the aortic arch. Brown et al. [10] published that using the left carotid artery as an autologous conduit for aortic arch continuity can be safe without observing neurological abnormalities in the intermediate-term follow-up. As we previously mentioned, the type of procedure depends on the experience of the surgical groups. In our center, it was decided to place a Dacron graft, since it has been seen that using the left carotid artery as a graft increases the risk of neurological alterations. In addition, no early reoperations with Dacron graft have been observed.

Although it is a rare pathology and its management is carried out in third level hospitals. It should be noted that it is a critical heart disease, that is, a pathology that if not detected and treated in time, patients have a high risk of dying in the first 10 days of life.

Despite the great technological advances in the field of pediatric cardiology and heart surgery, it is extremely important to perform a complete physical examination for the timely detection of these heart disease prior to discharge from birth. Finally, although it is not the objective of this study, it should be noted that currently at the national level, attempts are being made to implement mandatory cardiological screening. We recognize that it is not the perfect test, but it is definitely better than doing nothing for the timely detection and management of congenital heart disease.

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