INTERRUPTED AORTIC ARCH

Synonyms:
- Aortic arch interruption

Definition:
From the transverse portion of the normal aorta 3 large arteries emerge that nourish the head and arms: the brachiocephalic arterial trunk or innominate artery (which shall provide blood flow to the right subclavian artery and the right carotid artery), the left carotid artery and left subclavian artery, in that order (See Cardiac Anatomy in Normal Heart).

The interruption of the aortic arch is a rare congenital malformation characterized by anatomical discontinuity between the segments of the aortic arch.
The ascendent aorta and some branches of the transverse aorta remain connected to the left ventricle, while the remaining branches of the transverse aorta and the descendent aorta remain connected to the ductus arteriosus and, therefore, to the pulmonary artery and the right ventricle. According to the place where the aorta is interrupted, 3 subtypes of interruption are defined: types A, B and C. The most frequent one is type B (60%). Type A (39%) and type C (1%) follow.

**Classification of IAA**

**Normal**

**Type A**

**Type B**

**Type C**

Ao: aorta, PA: pulmonary artery, Inn: innominate artery, LC: left coronary artery, LS: left subclavian artery, d: ductus arteriosus
In most cases, there is another anatomic defect related to this aorta interruption, which is a **ventricular septal defect**. This implies the presence of an orifice connecting both ventricles, allowing the passage of blood abnormally from one side of the heart to the other.

Other related malformations are aberrant right subclavian artery and subaortic stenosis. The first of them is the abnormal location of the right subclavian artery. Instead of coming from the innominate artery, it arises alone from the aorta and as the last branch, after the left subclavian artery. In order to arrive from that origin to the right arm, it shall cross behind the esophagus, which sometimes produces a certain type of oesophageal obstruction with digestive symptoms such as vomiting.

Subaortic stenosis is the decrease in the diameter (with obstruction to blood flow) in left ventricular outflow tract, just before the origin of the aorta. This malformation has serious implications in the choice of the most convenient surgical approach for the patient, as we shall see later.

**Clinical Picture:**

In the newborn with interrupted aortic arch, blood supply to the lower half of the body depends on the ductus arteriosus. While it remains open, the baby is clinically stable. However, its normal closure days after birth triggers severe heart failure (see **Clinical picture** in Diagnosis and Treatment) and shock. This carries a high risk of death if not treated immediately with prostaglandins (to open the ductus) and aggressive measures such as diuretics, inotropes and mechanical ventilation.

Rarely, a different color can be seen between the upper and lower body, the latter being more bluish.

**Diagnosis:**

Clinical suspicion arises when a baby shows heart failure during the first days of life (See **Diagnosis** in Diagnosis and Treatment). The difference in the intensity of pulses between the 4 extremities can be useful to localize the interruption. The chest x-ray shows severe cardiomegaly (enlarged heart) and blood excess in the lungs.

Echocardiography is the best diagnostic method for these cases. It defines the anatomy of the aortic arch, the intracardiac anatomy (presence and location of the ventricular septal defect and other defects) and the presence or absence of subaortic stenosis.

Given the usefulness of the echocardiography, there is usually no need to carry out complementary studies such as catheterization, computed tomography or magnetic resonance imaging. These are reserved for specific cases in which the anatomy is not entirely clear.

**Treatment:**

Initial treatment should be aimed at compensating the patient clinically. This includes connecting the patient to a ventilator. Intravenous medication (prostaglandins to open the ductus, inotropes to improve heart function and diuretics to reduce the amount of fluid in the lungs) must also be administered.

Once compensated, the patient should undergo surgery (See **Surgery** in Diagnosis and Treatment). A few years ago, the repair was performed in two stages. The first one included the repair of the aortic arch and the partial closure of the pulmonary artery (banding) through an incision below the left scapula, without cardiopulmonary bypass. In a second surgery, the chest was opened through a sternotomy, the pulmonary artery banding was removed and the ventricular septal defect was closed using cardiopulmonary bypass.
Currently, the global trend is to correct everything in a single stage through sternotomy. A critical issue for the surgical strategy is to determine whether once operated the left ventricular outflow tract will be too small, a defect known as subaortic stenosis.

There is no clear division between patients who will develop it and those who will not. In general, it can be assumed that if the diameter (measured in millimeters) of the left ventricular outflow tract is smaller than the value of the patient's weight, the postoperative obstruction will be important. Conversely, if the diameter is larger than the weight + 2 mm there shall probably be no postoperative obstruction.

As an example, let us assume a 3 kg baby. If the left ventricular outflow tract is more than 5 mm, there shall probably be no postoperative obstruction, but if it were less than 3 mm, it shall probably develop. Intermediate values represent a great challenge for medical teams and should be evaluated promptly.

**Decision-making in surgical treatment of interrupted aortic arch**

- **Likely postoperative subaortic stenosis**
  - **NO**
    - Aortic arch repair + VSD closure
  - **YES**
    - **Enlarge LVOT?**
      - 1.- LVOT muscle resection
      - 2.- VSD patch
    - **Bypass LVOT?**
      - 1.- Biventricular: Yasui operation
      - 2.- Univentricular: Norwood operation

LVOT: left ventricular outflow tract, VSD: ventricular septal defect

If obstruction is unlikely, the aortic arch shall be repaired and the ventricular septal defect shall be closed (See picture below).

If the obstruction is likely after correction, there are two possible strategies. The first one is to enlarge the left ventricular outflow tract by removing muscle and placing the ventricular septal defect patch in such a way that it pulls from the tissues, thereby relieving the obstruction.

The second strategy is to bypass the left ventricular outflow tract. In turn, there are 2 options for this situation. If the left ventricle is too small, the Norwood operation shall be carried out (See Hypoplastic left heart syndrome in Congenital Heart Diseases). This operation reconstructs the aorta and connects it to the pulmonary artery so that both ventricles send their blood to all the body. As there is no other source of blood flow to the
In case the size of the left ventricle is adequate, a biventricular repair might be attempted by means of the **Yasui operation**, in which the aorta is reconstructed in the same way as in the Norwood procedure. The left ventricle is connected to the pulmonary valve and the reconstructed aorta with a patch in the ventricular septal defect. In that way, both ventricles are separated. Finally, the right ventricle is connected to the pulmonary
artery branches with a conduit, so as to send blood to the lungs. This procedure is extremely complex and of high risk, therefore it is carried out in very few situations.

**Prognosis:**

The first challenge is to overcome the surgery and the postoperative period. Both of them represent an important risk of death. The patient typically remains in intensive care for many days, connected to a ventilator and with a lot of medications (See *Postoperative* in Diagnosis and Treatment). If there are no residual defects such as obstruction of the reconstructed aortic arch, ventricular septal defect or subaortic stenosis, the prognosis is typically favorable, with a mortality rate close to 5% in the most experienced centers. In the absence of these residual defects, it is unusual that the patient needs another operation and the quality of life shall be excellent (See *Follow-up and control* in Diagnosis and Treatment).