fashion; in these situations, the presence of an aberrant subclavian artery could be crucial [4].

To the best of our knowledge only 16 cases of aortic valve atresia and IAA have been reported in the literature to date; only 3 of these cases had no direct connection to the ascending aorta and all had an aberrant right subclavian artery [5], thus proving that an aberrant subclavian artery receiving blood directly from the ductus arteriosus is indispensable for survival.

CASE DESCRIPTION

A full-term female newborn of an uncomplicated pregnancy was transferred to our unit shortly after birth with a prenatal diagnosis of aortic valve atresia. Prostaglandin-E1 infusion was started promptly and echocardiography confirmed [S,D,S] aortic valve atresia, a large cono-ventricular VSD and a rather developed left ventricle; moreover it unexpectedly revealed type B interrupted (left) aortic arch and an aberrant right subclavian artery. Blood flow through the ascending aorta and aortic arch was clearly inverted as well as that through both carotid arteries. Angiography confirmed the diagnosis and showed blood flowing from the ductus to the descending aorta, to both subclavian (left and aberrant right subclavian artery) and vertebral arteries, and from the Circle of Willis to both carotid arteries downward to the ascending aorta and coronary arteries (Fig. 1A and B).

In conclusion, coronary blood flow was assured by bilateral retrograde carotid artery flow in a sort of Circle of Willis dependent circulation, exclusively supplied by both vertebral arteries. Consequently we believe that the presence of an aberrant subclavian artery was necessary for survival.

Moreover, if the presence of a large VSD led to a rather developed left ventricle, a hypoplastic ascending aorta and small carotid arteries would have made cannulation for cardiopulmonary bypass extremely challenging and unsafe.

REFERENCES


Treatment solution by Careddu et al.

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The treatment of choice for aortic valve atresia with interrupted aortic arch was a biventricular repair at 12 months.

On Day 6 of life, the patient underwent selective pulmonary arteries banding and ductus stenting with the aim to promote ascending and aortic arch growth for further repair. The hybrid procedure was preferred because it was judged to be safer

Figure 1: (A) Left carotid artery division and connection to the main pulmonary artery (proximally), and to the right carotid artery with azygos vein segment interposition (distally). (B) Biventricular repair. Right ventricle to pulmonary artery conduit (Contegra). Dacron conduit connection of the main pulmonary artery (neo aorta) to the aortic arch.
in the neonatal period, considering (i) the uncertainty of biventricular repair and (ii) the difficulties in achieving a good cannulation and reliable perfusion for such a long and unpredictable procedure.

Three months later, after ischaemic changes were detected on electrocardiography (ECG), the patient was taken to the Cath-lab for angiography that confirmed the presence of a reverse flow from the Circle of Willis to the ascending aorta and coronary arteries, and a very small left carotid artery (LCA). Thus, a 3-mm polytetrafluoroethylene shunt was implanted between the main pulmonary artery and the LCA in order to increase coronary perfusion.

After 3 months, despite further changes seen on the ECG, we still considered a biventricular repair unsafe at that point in time. Instead, we decided to divide the LCA and transpose it directly to the pulmonary trunk in order to increase blood flow to the coronary arteries. The shunt was removed and the LCA patency was re-established with azygos venous segment interposition (Fig. 1A).

At 12 months of age, the patient successfully underwent biventricular repair: (i) under deep hypothermic circulatory arrest, main pulmonary artery was transected and the ductus partially removed; the stent was cut open and enlarged because it was impossible to remove; (ii) anastomosis between a 12-mm Dacron conduit and the aortic isthmus was performed; (iii) the pulmonary arteries were debanded and detached from the main pulmonary artery; (iv) intraventricular left ventricle to pulmonary artery patch baffling through the VSD was performed; (v) anastomoses between the main pulmonary artery and the Dacron conduit and (vi) between the right ventricle and a pulmonary artery valved conduit (Contegra 12 mm) (Fig. 1B) were carried out.

The use of a Dacron conduit was necessary because it was impossible to remove the stent from the aortic isthmus and because of the shortness of the remaining main pulmonary artery trunk that served as the ascending aorta. There was not enough autologous tissue for an end-to-end direct anastomosis or a combined end-to-end procedure with patch completion.

The postoperative period was uneventful and the baby was discharged home on the 13th postoperative day. After 18 months, she is doing fine, asymptomatic and growing normally with no aortic pressure gradient.