Aortic atresia with interrupted aortic arch: a combination incompatible with life?

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CLINICAL PROBLEM

Aortic valve atresia is a common feature of hypoplastic left heart syndrome, which consists of various degrees of underdevelopment of the left ventricle/aorta complex; nevertheless it can also be associated with a large ventricular septal defect (VSD) and a rather developed left ventricle.

Atresia of the aortic valve prevents antegrade blood flow from the left ventricle to the ascending aorta, so perfusion of the coronary arteries, ascending aorta and aortic arch is provided by the ductus arteriosus in a retrograde fashion.

Interrupted aortic arch (IAA) is quite often associated with aortic or subaortic stenosis; however, very few cases of IAA have been reported in association with aortic valve atresia.

Aortic valve atresia with IAA is an extremely rare association which would be incompatible with life, unless blood flow is provided to the ascending aorta and coronary arteries from (i) the ductus/descending aorta through ‘collaterals’ [1], or (ii) from the pulmonary trunk and pulmonary arteries through an aorto-pulmonary window [2] or bilateral ductus [3]. In the absence of a direct connection, blood flow to the coronary arteries is exclusively provided by the Circle of Willis through both carotid arteries, in a reverse direction.

Figure 1: (A) Angiography shows Willis’ circle coronary dependent circulation. (B) Angiography shows large subclavian and vertebral arteries and diminutive carotid arteries and ascending aorta visualized by retrograde filling.


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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


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.