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Brief Report

Rapid formation of collateral arteries in a neonate with interruption of the aortic arch

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Abstract In a neonate born prior to term with a weight of 1825 grams, and diagnosed prenatally as having atrioventricular septal defect and Down's syndrome, we found the aortic arch to be interrupted between the left carotid artery and the left subclavian artery, with the arterial duct being the only route of distal perfusion. Three days later, however, echocardiographic interrogation revealed marked collateral connections between the aortic arch and the descending aorta, the picture then mimicking coarctation rather than interruption of the aortic arch. The rapid development of the collateral arteries was confirmed by magnetic resonance imaging and during cardiac surgery.

Keywords: Down's syndrome; atrioventricular septal defect; aortic arch interruption; magnetic resonance imaging

Interruption of the aortic arch is often associated with a ventricular septal defect, usually with the muscular outlet septum deviated to produce subaortic obstruction.[1–3] The pattern of interruption between the left carotid artery and the left subclavian artery, so-called type B, is a typical defect in patients with 22q11 deletion. In patients with Down's syndrome, the most frequent associated cardiac lesion is atrioventricular septal defect with common atrioventricular junction.[4] The combination of atrioventricular septal defect with interruption of the aortic arch, however, is extremely rare. To our knowledge, it has never been reported in a patient with Down's syndrome.

When diagnosed, patients with interruption of the aortic arch are currently referred for surgery after evaluation by echocardiography alone. Some patients may survive through infancy without the need for surgical treatment and, in addition to a patent arterial duct, usually exhibit a markedly developed collateral circulation across the interruption. Nevertheless, some patients have been known to survive in the absence of a patent arterial duct.[5] In this setting, there is a prominent collateral supply to the area distal to the obstruction, which develops over a period of time. When choosing surgical intervention based on echocardiography, it may be difficult to distinguish between interruption of the aortic arch and coarctation produced by a long, narrow segment of the hypoplastic aortic arch and isthmus, with significantly reduced antegrade flow. Catheterisation is now only rarely required for the precise delineation of this anatomy. In this report, we describe our use of magnetic resonance imaging to confirm the echocardiographic suggestion of the early development of the collateral circulation across an interrupted segment of the aortic arch.

Case report

A premature female, born during the thirty-fifth week of gestation, with a weight of 1825 grams and measuring 44 cm in length, had been diagnosed in fetal life as having an unbalanced atrioventricular septal defect with common atrioventricular valve in the setting of trisomy 21. The course of the aortic arch could not be defined. Shortly after birth, echocardiography using a 10 mHz transducer, confirmed the fetal diagnosis, revealing a small right ventricle, a large ventricular component, with only a small remnant of the muscular ventricular septum, and

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complete absence of the interatrial septum. Additionally, the investigation showed that the aortic arch was interrupted between the left carotid artery and the left subclavian artery. The length of the missing segment was measured at 3.5 mm. The aortic valve was bifoliate, but there was no evidence of deviation of the muscular outlet septum. No antegrade flow was detectable between the aortic arch and the descending aorta, which was perfused exclusively via the arterial duct. Alprostadil was started at a dose of 0.01 μg/kg/min, this being increased on the fourth day to 0.02 μg/kg/min because increasingly turbulent flow was seen in the arterial duct. The arterial duct remained open, and the clinical condition stable. Artificial ventilation was not required. Two days later, prior to the electively planned surgical procedure, repeated echocardiography demonstrated an obvious connection between the aortic arch and the descending aorta (Fig. 1). Two narrow vessels flow could be visualized, mimicking antegrade flow through a hypoplastic aortic isthmus. Magnetic resonance imaging using a 1.5 Tesla magnet and an electrocardiographically triggered turbo-spin-echo and gradient echo technique, performed under oral sedation, demonstrated convincingly the persisting interruption of the arch, and confirmed the presence of two newly developed small collateral arteries (Fig. 2).

Cardiac surgery was performed on the fifth day of life. Because the intracardiac anatomy was deemed unsuitable for biventricular correction, the aortic arch was anastomosed directly to the descending aorta, along with banding of the pulmonary trunk. The presence of two small collateral arteries between the aortic arch and the descending aorta was confirmed at surgery. Despite the good function of the anastomosis, the patient died several weeks later due to intractable heart failure caused by persistent high pulmonary blood flow and atrioventricular valvar insufficiency.

Comment

Congenital cardiac malformations are common in patients with Down's syndrome, especially atrioventricular septal defect. Combinations with other cardiac anomalies are well recognised, such as persistent arterial duct and tetralogy of Fallot, although rare. The association of interruption of the aortic

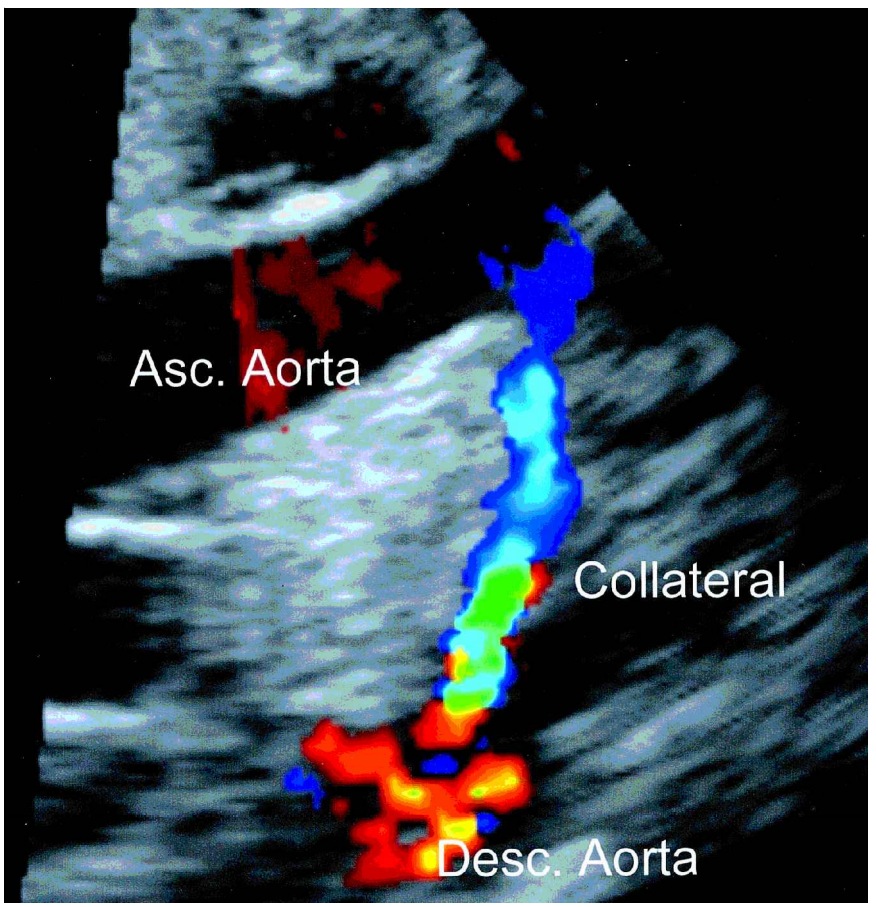

Figure 1.
Suprasternal echocardiography with Doppler color flow showing collateral arteries spanning the interrupted aortic arch between the ascending and descending segments of the aorta. Antegrade flow is visualized in blue, with turbulence in the distal part of the collateral vessel shown in green. Asc. Aorta = Ascending Aorta; Desc. Aorta = Descending Aorta.

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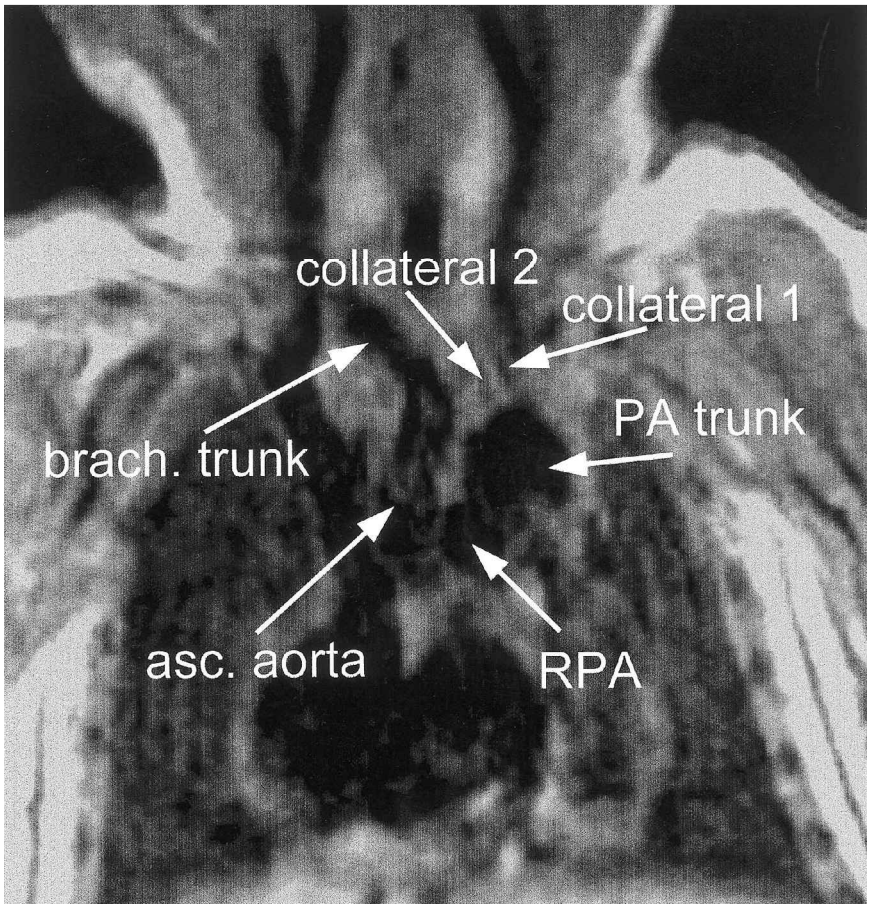

Figure 2.
Magnetic resonance imaging in the coronal plane on the fourth day of life confirms the interruption, and shows two arteries (collaterals 1 and 2) running between the ascending and descending aorta. Both vessels insert to the upper part of the descending aorta, which is connected via the arterial duct to the pulmonary trunk (PA trunk). Asc. Aorta = ascending aorta; brach. trunk = brachiocephalic trunk; RPA = right pulmonary artery.

arch and atrioventricular septal defect in a child with Down's syndrome, however, is extremely rare, and to our knowledge has not been previously described.

In adults, it is known that collateral arteries can span an area of complete interruption of the aortic arch in absence of an arterial duct.[5] Also, in older babies, formation of collateral arteries has been described, though most have been recognised at autopsy.[6] At a later age, restriction of the ductal patency may produce a difference in pressure between the segments of the arch proximal and distal to the interruption. Consequently, pre-existing communications become more important under the changed hemodynamic conditions. In our patient, the reason for such a rapid development of collateral bypass of the interruption of the aortic arch is not entirely clear. The dosage of alprostadil had to be increased to maintain the ductal patency. It is possible that this potential closure of the arterial duct changed the pressures in the descending aorta, at the same time enhancing the formation of collateral flow.

Whereas echocardiography is undoubtedly accepted as the best non-invasive diagnostic tool in small pre-term neonates, experience with magnetic resonance imaging in neonates of this age and weight is still limited. The babies may be exposed to the risks associated with transportation to another department, such as hypothermia, unstable hemodynamics, and other problems. Nevertheless, with experience, resonance imaging can become just as safe as bedside echocardiography performed in the neonatal intensive care unit. The limitations of magnetic resonance imaging are attributed to the specifications of the equipment and the experience of the staff. Our experience demonstrates that reliable distinction between interruption of the aortic arch with early development of collateral circulation, and a tight coarctation of the aorta can be performed with echocardiography and magnetic resonance imaging even in small premature neonates.

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