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Forum for Discussion

Successful closure of the arterial duct in the setting of rubella syndrome

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Abstract A 9-year-old boy, with significant left-to-right shunting across a large duct in the context of rubella syndrome, was tested during catheterization to establish the feasability of occluding the duct with a device. The testing, including temporary closure of the duct and monitoring of pulmonary vascular reactivity to vasodilative substances, lead to the decision to implant an Amplatzer occluder. Sixteen months later, there was no residual shunting across the duct, and pulmonary arterial pressures had normalised. It remains unclear why the patient had not developed irreversible pulmonary vascular disease.

Keywords: Eisenmenger syndrome; pulmonary vascular disease; ductus arteriosus; Amplatzer duct occluder

Pulmonary hypertension, when encountered in children with congenital cardiac disease and left-to-right shunting, may produce the morphological alterations of the pulmonary vasculature known as pulmonary plexogenic arteriopathy.[1] This change is characterised by increased muscularisation of the small pulmonary arteries, cellular intimal proliferation, intimal fibrosis, dilation lesions, fibrinoid necrosis, and plexiform lesions. Advanced pulmonary vascular disease is associated with reversal of shunting so that the blood flows from right-to-left, a change known as the Eisenmenger syndrome. In most instances, this reaction is irreversible, obviating the potential to close the defect. It is currently considered that, when the shunting is across the arterial duct, then the smaller the duct, the lower is the left-to-right shunt, and the lower the risk of developing irreversible pulmonary vascular disease. By the same token, it is usually assumed that, with significant left-to-right shunting across the duct, the patient will finally develop the Eisenmenger syndrome.[2] For unknown reasons, nonetheless, some patients with large arterial ducts will continue to present with left-to-right shunting even in adulthood.[3] We present here a child with intrauterine acquired rubella syndrome who presented with pulmonary hypertension due to a patent duct. We invite further discussion on our findings in the light of the above considerations.

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Case report

A 9-year-old male presented with the typical symptoms of intrauterine acquired rubella syndrome, namely deafness, mental and motor disabilities, and patency of a large arterial duct. At catheterization, the pulmonary arterial pressure was elevated, with systolic levels of 61 mmHg and a mean pressure of 47 mmHg. The ratio of the pulmonary to the aortic pressure was 0.8. There was significant shunting across the duct, with ratios of pulmonary to systemic flow greater than 4. The duct was displayed angiographically, and sized at 7–8 mm (Fig. 1). In order to test the feasibility of closing the duct by catheter intervention, we occluded it temporarily using a balloon (Meditec, Boston Scientific), while monitoring aortic and pulmonary pressures. The pulmonary pressure decreased to 45 mmHg systolic during occlusion, while the aortic pressure increased from 90 mmHg to 145 mmHg.

Testing for acute pulmonary vascular reactivity showed selective pulmonary vasodilation after

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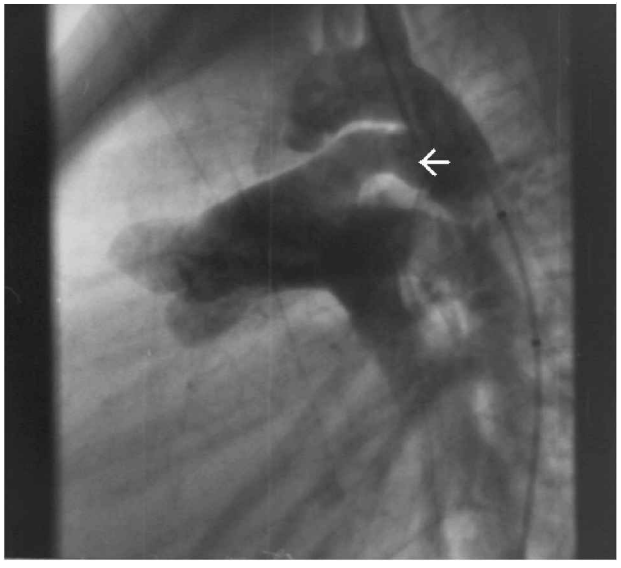

Figure 1.
Angiography of the aortic arch in 90 degree lateral projection. The aorta gives rise to the large duct (arrow), which fills the pulmonary arteries because of the left-to-right shunting.

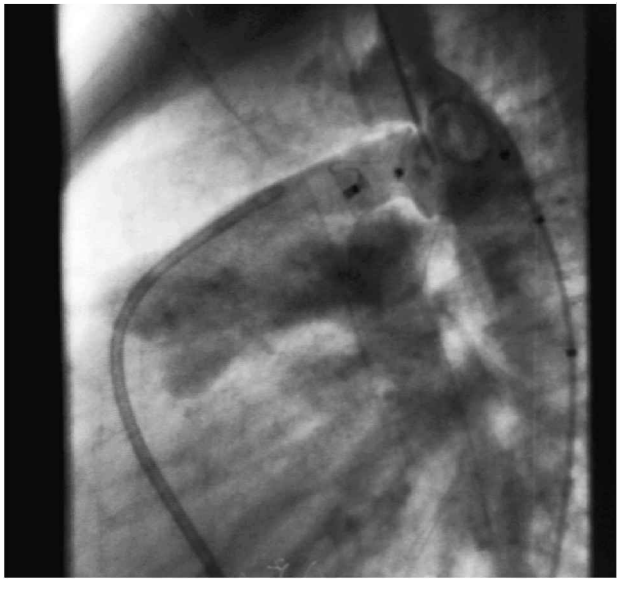

Figure 2.
An Amplatzer duct occluder has been placed in the duct, but there is still significant residual shunting (same angulation as Fig. 1).

inhalation of oxygen, with the mean pulmonary arterial pressure decreasing by 10 mmHg. Inhalation of nitric oxide at 20 parts per million did not significantly lower the mean pulmonary pressure, which did not move more than 20% from baseline.[4] Our protocol of testing vasoreactivity has been described previously.[5] Levels of endothelin-1 in the plasma

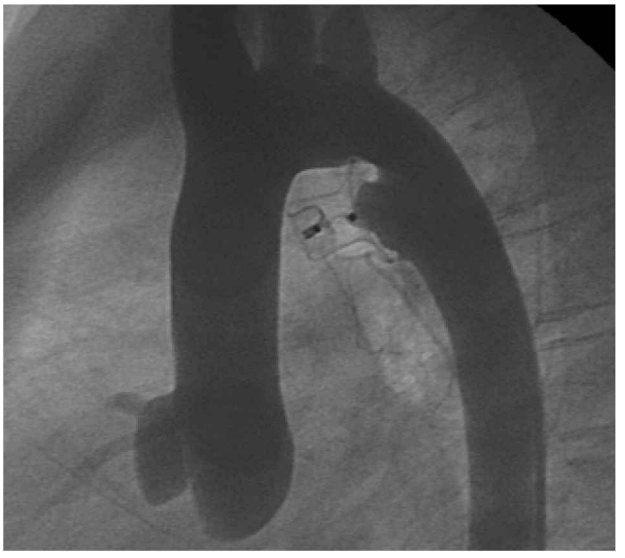

Figure 3.
Complete occlusion is noted after a period of follow-up of 1½ years (same angulation as Fig. 1; different magnification).

Table 1. Pressure values from hemodynamic measurements before (1998) and after (2000) interventional closure of the ductus.

	PA syst mmHg	PA mean mmHg	Aorta syst mmHg	Aorta mean mmHg	PA/aorta mean ratio
1998	61	47	90	60	0.78
2000	38	20	80	59	0.34

obtained from samples of blood from the pulmonary arteries (2.4 pg/ml) and the aorta (2.5 pg/ml) were not elevated compared with control values obtained in our laboratory.[6]

After testing, the duct was closed using a 12/10 Amplatzer duct occluder. We introduced the device via a 7 French delivery system designed for closure of atrial septal defects. The size and morphology of the duct justified the use of an Amplatzer duct occluder, rather than an occluder designed for closure of ventricular septal defects. At the end of the procedure, there was still significant residual shunting (Fig. 2). Follow-up catheterization 16 months later showed no residual shunt (Fig. 3). Pulmonary pressures had dropped significantly, with a mean pressure of 20 mmHg and a ratio to aortic pressure of 0.34 (Table 1).

Comment

It is very unusual to be able to close a large duct permitting a left-to-right shunt of sufficient magnitude to produce pulmonary hypertension. In such a setting,

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it is crucial to test the acute reactivity of the pulmonary vascular bed. In our patient, testing using vasoactive substances, as well as temporary occlusion of the duct, convinced us that interventional closure would have a beneficial effect, and this proved to be the case.

The question posed by our experience is: why did the patient retain a reactive pulmonary vascular bed after 9 years of shunting? Does the condition of rubella acquired during intrauterine life play a particular role? We have been unable to find any literature indicating that, in the setting of rubella, the pulmonary vasculature reacts differently from any other situation. Was our patient just lucky? We would welcome comments, which can be posted on the website of the Journal (http://www.greenwich-medical.co.uk).

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