Transcatheter umbrella closure of aorto-pulmonary window

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Abstract
Aorto-pulmonary window (aorto-pulmonary septal defect) is an uncommon congenital cardiac malformation which is repaired using cardiopulmonary bypass. A case is described of an infant with a small aorto-pulmonary window which was closed by transcatheter insertion of a double umbrella device. Complete occlusion of the defect was achieved without complications. Transcatheter umbrella closure of a small aorto-pulmonary window is feasible in infancy and the technique is likely to be applicable in a few cases.

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The first report of an aorto-pulmonary window was that of Elliotson in 1830. The window is usually a large oval defect between the ascending aorta and pulmonary trunk, but in about 10% of cases it is small. The defect is often short and is usually a single orifice.

There may be associated anomalies such as anomalous coronary artery origin, anomalous origin of the left or right pulmonary artery from the ascending aorta, or even pulmonary atresia or aortic atresia. Because in most cases the defect is large, infants usually present with congestive heart failure. Other potential problems include the development of pulmonary vascular disease and bacterial endocarditis. In most cases, surgical repair is undertaken during infancy.

We report here the use of the Rashkind double umbrella (Bard) to close an aorto-pulmonary window in an infant.

Case report
A 9 week old infant was symptom free but presented with a loud continuous murmur that was loudest at the upper left sternal edge. The initial echocardiogram did not show an arterial duct but did show continuous turbulent flow in the pulmonary trunk and a dilated left atrium and left ventricle. The electrocardiogram and chest radiograph were normal.

At cardiac catheterisation at the age of six months, the pulmonary artery pressure was normal. A selective ascending aortogram showed a small aorto-pulmonary window 3 mm in diameter (fig 1). His parents gave informed consent to transcatheter closure with a 12 mm Rashkind double umbrella four months later, when the infant weighed 8 kg. A retrograde arterial approach was used to pass an end-hole catheter from the aorta through the defect and into the pulmonary artery and then to the superior caval vein through the right ventricle and right atrium. An 0.025 inch
wire introduced through the catheter was snared and retrieved to the femoral vein. A 12 mm Rashkind double umbrella device was modified by bending the arms medially, in order to allow the device to lie flat against the aorto-pulmonary septum. An 8F Mullins sheath was then used to deliver the umbrella device. The distal arms were opened in the aorta and the proximal arms were opened in the pulmonary trunk. The procedure was uncomplicated and lasted 65 minutes with a fluoroscopy time of 26 minutes. After the procedure there was complete occlusion of the defect as assessed by angiography and echocardiography (fig 2). There was no interference with pulmonary or aortic valve function.

The child was reviewed six months after closure. At that time there was no murmur or other complication. Echocardiographic assessment showed complete occlusion of the defect, with resolution of the left atrial and left ventricular enlargement. After Doppler interrogaton there was no evidence of stenosis induced by the umbrella in either the pulmonary trunk or the ascending aorta.

Discussion
There have been several reports of the surgical ligation of an aorto-pulmonary window, initially with a closed technique which could be applied to small defects. The introduction of cardiopulmonary bypass allowed for the correction of smaller defects with division, however, in most cases the defect is large so that patch closure from the aorta is the best treatment.

The use of the Rashkind double umbrella for the closure of a persistent arterial duct is well established, and it has now been used to close atrial septal defects, baffle fenestrations after total cavopulmonary anastomoses, and ventricular septal defects.

This case demonstrates a novel use of the double umbrella delivery system to close a small aorto-pulmonary window. There is one previous case report of such a technique, but in that case the child was 3 years old and there was residual left to right shunt after the procedure, which meant that the risk of endocarditis was not avoided.

Most infants presenting with an aorto-pulmonary window will require conventional surgical repair with the aid of cardiopulmonary bypass. However, we have shown that when the defect is small, distant from the semi-lunar valves, and not associated with anomalous origin of the right or left pulmonary arteries from the ascending aorta, transcatheter umbrella closure can be effective and safe. It is probably the best treatment in a few cases.

1 Elliotton J. Case of malformation of the pulmonary artery and aorta. Lancet 1830;E:247–50.
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