Percutaneous transcatheter myectomy of subvalvar pulmonary stenosis in tetralogy of Fallot: a new palliative technique with an atherectomy catheter

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Abstract
A male infant with tetralogy of Fallot was given a modified left Blalock-Taussig shunt at 11 days of age because of duct dependence. Twenty months later, because of increasing cyanosis and polycythaemia, he was given palliative treatment with balloon dilatation of the right ventricular outflow tract. Seven months later a right ventricular angiogram showed further progression of the infundibular stenosis with poor anterograde opacification of the pulmonary arteries. He underwent percutaneous transcatheter myectomy of the infundibulum with an atherectomy catheter. One month later he was well and less cyanosed.

Pulmonary atresia associated with the tetralogy of Fallot is usually congenital. Occasionally, atresia may develop at the infundibular level after a systemic to pulmonary artery shunt operation.1,2 A valve conduit is sometimes required for complete repair in this situation which may then be associated with an increased morbidity and operative mortality.3,4 Although it is not possible to identify patients with tetralogy of Fallot who may develop pulmonary atresia, any intervention that prevents this complication may, ultimately, reduce the risks associated with complete surgical repair. Brock achieved effective palliation through closed infundibular resection or dilatation.4,5 Similarly, balloon dilatation of the right ventricular outflow tract has recently been shown to be of value in infants and children with tetralogy of Fallot.6,7

We report the successful use of a new technique of transcatheter myectomy of the infundibulum in a child with tetralogy of Fallot.

Case report
Tetralogy of Fallot was diagnosed in a one day old boy who weighed 2.5 kg. Cross sectional echocardiography showed confluent, but small, pulmonary arteries and severe pulmonary valvar and infundibular stenosis. When he was 11 days old he was given a modified (5 mm polytetrafluoroethylene) left Blalock-Taussig shunt because of severe cyanosis and dependence upon prostaglandin E1 to maintain the patency of the ductus arteriosus.

At 14 months of age he was moderately cyanosed with a haemoglobin concentration of 19.3 g/l. Cardiac catheterisation showed that the pressure in the pulmonary artery (entered retrogradely through the shunt) was 10/0 (mean 8) mm Hg and the aortic pressure was 120/70 (mean 90) mm Hg. The aortic saturation was 81% in air. A right ventricular angiogram showed a trivial amount of contrast flowing through the infundibulum into the pulmonary trunk. The right ventricular outflow tract was dilated with a 5 mm balloon catheter and then a 8 mm balloon catheter. He was reinvestigated seven months later. Angiography showed severe infundibular stenosis. He underwent further balloon dilatation of the right ventricular outflow tract followed by transcatheter myectomy of the infundibulum with an atherectomy catheter.

Technique
Cardiac catheterisation was performed under general anaesthesia. The left femoral artery was cannulated with an 18 G Abbocath. Catheters were also introduced percutaneously into the right and left femoral veins. The right ventricular systolic pressure was 110 mm Hg, the femoral artery pressure 125/60 (mean 80) mm Hg, and the main pulmonary artery pressure 17/11 (mean 13) mm Hg. Saturations in the femoral artery were 78% and in the pulmonary artery 69%. A selective injection of contrast with the catheter in the infundibulum showed severe subvalvar pulmonary stenosis caused by hypertrophy of the infundibulum (fig 1). To allow an 8 French transseptal sheath (Bard International) to be passed into the pulmonary trunk, the outflow tract was dilated with an 8 mm diameter balloon catheter by the standard technique. The level of the infundibulum was marked with a needle positioned externally on the patient’s chest. The 8 French
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Figure 1 Selective angiogram in the infundibulum (anteroposterior projection) taken at start of procedure, showing gross hypertrophy of the infundibulum.

Figure 2 (A) Atherectomy catheter (on anteroposterior x-ray screening) in position during myectomy. The balloon was inflated (hollow arrow), the cutting window (small arrows) was in contact with the infundibulum, and the drill, which was being advanced, can be seen (large arrow). (B) Selective angiogram in the infundibulum (anteroposterior projection) after myectomy. The outflow tract was wider and the surface of the infundibulum (arrows) was irregular.

transseptal sheath and its dilator were advanced over an 0.035 inch guide wire and positioned in the proximal pulmonary trunk. The 6 French, Simpson coronary atherectomy catheter (Devices for Vascular Intervention) with a “nose cone” distal to the cutting window was prepared by purging the lumen port and 1.5 mm diameter balloon. The guide wire and the dilator were removed, leaving the sheath across the pulmonary valve. We introduced an 0.014 inch exchange guide wire through the sheath and advanced the atherectomy catheter over the wire until it was positioned across the infundibulum, using the landmarks from the marker needle. The sheath was then withdrawn and the catheter was rotated until the cutting window pointed postero-medially. The balloon was then fully inflated to a pressure of 4 atmospheres and the drill was activated by the motor drive unit and advanced manually (fig 2A). Nine cuts were made with the cutting window in different orientations. The balloon was deflated and the sheath advanced over the atherectomy catheter which was then removed. A further balloon dilatation with an 8 mm diameter balloon was then performed. All of the pieces of resected tissue were removed from the “nose cone”. Microscopic examination confirmed that they were myocardium and fibrous tissue. A selective angiogram in the infundibulum showed a wider and irregular outflow tract (fig 2B). At the end of the procedure, the pressure was 16/7 (mean 10) mm Hg in the pulmonary artery, 125/80 (mean 100) mm Hg in the descending aorta, and 110 mm Hg in the right ventricle during systole. Oxygen saturation was 67% in the pulmonary artery and 81% in the descending aorta. There were no complications during or after the procedure. One month later he was well, less cyanosed, and the saturation by pulse oximetry was 85%.

Discussion

Many centres adopt a staged surgical approach in the treatment of tetralogy of Fallot. This consists of the creation of a systemic to pulmonary artery shunt followed later by complete repair. This approach has the advantage of allowing continued growth of the pulmonary valve ring and pulmonary arteries, thus reducing the need for a transannular patch and decreasing the operative risk. Occasionally, however, atresia may be developed at the
infundibular level after a previous shunt operation.\textsuperscript{2,3} Such a situation requires an extensive outflow patch or a conduit for complete repair.\textsuperscript{11} Brock first reported the technique of infundibular resection and dilatation in 1948.\textsuperscript{4} He believed, as did Matthews and Belsey,\textsuperscript{12} that increasing anterograde flow across the right ventricular outflow tract not only achieved adequate palliation but was more likely to promote the growth of the pulmonary valve ring, trunk, and arteries.\textsuperscript{6} Recently balloon dilatation of the right ventricular outflow tract has been used as a non-surgical method of increasing anterograde pulmonary blood flow\textsuperscript{4,7} and has produced adequate palliation.

A further report has indicated that an increase in the size of the pulmonary valve ring, trunk, and arteries may also occur after balloon dilatation.\textsuperscript{11}

In our patient palliation after balloon dilatation, though initially effective, was limited by further increase in the infundibular obstruction. After another balloon dilatation to facilitate the passage of a large sheath through the infundibulum, we were able to proceed with myocardial resection. Histology confirmed that the resected pieces were myocardium and fibrous tissue.

Atherectomy catheters are currently under evaluation in peripheral vascular and coronary artery disease.\textsuperscript{12,13} We showed the successful application of an atherectomy catheter for the resection of the infundibulum in a child with tetralogy of Fallot.

We suggest that transcatheter resection of the infundibulum may be important in the palliation of the tetralogy of Fallot and possibly other conditions characterised by fibromuscular stenosis. However, further modifications of the atherectomy catheter are needed for this technique to become widely accepted.

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