Arterial duct angioplasty as an adjunct to dilatation of the valve for critical pulmonary stenosis

Kevin P Walsh, Simon E Abrams, Robert Arnold

Abstract

Objective—To assess the efficacy of arterial duct angioplasty in maintaining adequate ductal patency in neonates with critical pulmonary valve stenosis.

Patients—Two neonates presenting with cyanosis due to critical pulmonary valve stenosis with severe right ventricular hypoplasia underwent percutaneous balloon dilatation of the pulmonary valve. Despite successful dilatation, both remained cyanosed while receiving prostaglandin E₂ infusions.

Results—Angioplasty of the arterial duct resulted in increased arterial oxygen saturations. Adequate arterial duct patency was maintained for three months during which time right ventricular dimensions increased enough to allow sufficient pulmonary circulation without the duct.

Conclusions—Arterial duct angioplasty is an effective additional treatment for patients with critical pulmonary valve stenosis whose right ventricle is too small to provide adequate forward flow after balloon dilatation of the valve. Arterial duct angioplasty sustains adequate patency of the duct while the right ventricle grows sufficiently to undertake the pulmonary circulation.

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Neonates with critical pulmonary valve stenosis and an intact ventricular septum present soon after birth with cyanosis due to inadequate pulmonary blood flow.¹ Current management consists of balloon dilatation of the critically stenosed pulmonary valve.²⁻⁴ Although many of these patients have significant associated right ventricular hypoplasia, these hypoplastic ventricles can grow if continuity between the right ventricular cavity and pulmonary artery is established.⁵ Patients whose right ventricle is too small alone to provide adequate pulmonary blood flow after a technically adequate dilatation of the pulmonary valve, require a systemic to pulmonary shunt or prolonged administration of prostaglandin (PG) E₁ or E₂.⁴ Unfortunately, PGE₂ infusions produce sustained clinical improvement in only 60% to 88% of patients with cyanotic heart disease.⁶

We have recently shown that angioplasty of the arterial duct restores the systemic circulation of neonates with severe left heart obstruction who are unresponsive to infusion

of PGE₂.⁷ We now report the use of angioplasty of the arterial duct in two neonates with critical pulmonary stenosis and severe right ventricular hypoplasia who remained cyanosed after balloon dilatation of the pulmonary valve despite prostaglandin infusion.

Patients and methods

Two neonates (aged five and four days; weight 4.0 and 3.9 kg) presented with cyanosis. A PGE₂ infusion had been started at their referring hospital. A continuous murmur at the left infraclavicular area was easily heard in both patients. Chest x ray film showed normal heart size and pulmonary oligoemia. Electrocardiograms showed sinus rhythm with right atrial enlargement, and a QRS axis of +70° with upright T waves in V3R and V1. Cross sectional echocardiography showed critical pulmonary valve stenosis with a peak Doppler jet of 4.7 and 4.5 m/s. There was severe right ventricular hypoplasia in both patients. The arterial duct was the main source of pulmonary blood flow with a continuous aorta to pulmonary artery Doppler profile. Colour flow mapping showed predominantly right to left shunting across the foramen ovale.

Figure 1 Right ventriculogram (patient 1, left lateral projection) performed before dilatation of the valve showing a tiny jet of contrast through the critically stenosed pulmonary valve and the associated severe right ventricular hypoplasia.

Heart Clinic, Royal
Liverpool Children's
Hospital, Alder Hey,
Liverpool
K P Walsh
S E Abrams
R Arnold

Correspondence to:
Dr K P Walsh,
Heart Clinic, Royal
Liverpool Children's
Hospital, Alder Hey, Eaton
Road, Liverpool L12 2AP.

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Figure 2 Angiogram of the arterial duct after angioplasty (patient 2, left lateral projection). A widely patent and slightly irregular arterial duct fills the main pulmonary artery and contrast can be seen regurgitating into the hypoplastic right ventricle.

Both patients underwent cardiac catheterisation and balloon dilatation of the pulmonary valve within 24 hours of admission. Both had suprasystemic right ventricular pressures with right ventricle to femoral artery systolic pressure ratios of 1.51 and 1.84. Right ventriculography in both showed a very hypertrophied ventricle with small cavity and a dysplastic doming pulmonary valve with an extremely small central jet (fig 1). On the lateral projection the tricuspid valves measured 11 and 10 mm, and the pulmonary valves measured 6 and 5.3 mm in diameter. In each patient the valve was crossed with an 0.018 inch (William Cook) or 0.025 inch (Terumo Corporation) guide wire that was advanced to the distal left pulmonary artery or through the arterial duct into the descending aorta. The pulmonary valve was dilated serially with 6, 8, and 10 mm diameter balloons (Balt). After dilatation the peak systolic pulmonary valve gradient measured 26 mm Hg in both patients with a right ventricle to femoral artery systolic pressure ratio of 0.72 and 0.7. In view of the severe hypoplasia of the right ventricles both patients remained on PG E₂ infusions but became progressively more cyanosed with arterial PO₂ of 28 and 24 mm Hg. Cross sectional echocardiography with colour flow mapping showed the arterial ducts to be considerably narrowed at their pulmonary ends. Therefore five and six days later both patients underwent arterial duct angioplasty. With a trans-venous approach, a 4 F Gensini or multipurpose catheter was passed through the arterial duct and used to position an 0.018 inch or 0.032 inch exchange guide wire in the abdominal aorta. A 10 mm diameter 2 cm long balloon on a 5.5 F shaft was positioned across the arterial duct and rapidly inflated with carbon dioxide gas and deflated, and then with dilute contrast. Aortic saturation increased from 78% to 86% and from 70% to 96% respectively and pulmonary artery pres-

sure increased from 26/23 to 52/20 mm Hg and from 21/17 to 46/21 mm Hg. Angiography in patient 2 showed a widely patent arterial duct after angioplasty (fig 2). A loud continuous murmur was easily heard in both patients who were no longer clinically cyanosed after treatment. In both patients the arterial duct was noted as becoming more restrictive on follow up echocardiograms and closed without clinical consequences after three months. Both have exhibited right ventricular growth on follow up echocardiography and neither has required further intervention during six months follow up.

Discussion

Neonates with critical pulmonary valve stenosis and intact ventricular septum are currently treated by balloon dilatation of the critically stenosed pulmonary valve.²⁻⁴ The degree of associated right ventricular hypoplasia is variable.¹ The hypoplastic right ventricle has considerable potential for long-term growth after relief of the pulmonary valve stenosis, although its end diastolic volume and stroke volume decrease transiently after decompression.⁸ The right ventricle is unable to provide adequate pulmonary blood flow unassisted over this period and a systemic to pulmonary shunt,⁹ or prolonged administration of PG E₁ or E₂ is often required.⁴ Administration of PG to maintain arterial duct patency over this period may avoid a surgical shunt and a further intervention to occlude it when it is no longer required. Unfortunately, PGE₂ infusions produce sustained clinical improvement in only 60% to 88% of patients with cyanotic heart disease,⁶ and a surgical systemic to pulmonary shunt may be necessary. Arterial duct angioplasty provides an alternative to a surgical shunt in such patients.

We have recently shown that angioplasty of the arterial duct in neonates with severe left heart obstruction produces a widely patent duct and restoration of the systemic circulation where PG had failed.⁷ This report shows that angioplasty of the arterial duct is also effective in neonates with critical pulmonary stenosis and severe right ventricular hypoplasia who remain cyanosed after balloon dilatation of the pulmonary valve despite PG infusion. Arterial duct angioplasty resulted in an immediate increase in pulmonary blood flow. The ducts remained open for at least three months but showed evidence on Doppler echocardiography of gradual constriction. In these particular patients increasing forward flow through the growing right ventricle seemed to run parallel with the gradual narrowing of the duct thus avoiding over-perfusion of the lungs. Formal duct closure was not required. In other types of cyanotic congenital heart disease such as pulmonary atresia where the pulmonary blood supply is entirely duct dependent, arterial duct angioplasty may not provide a secure long-term source of pulmonary blood supply. These patients require a surgical systemic to pulmonary shunt or possibly maintenance of

duct patency with a balloon expandable endoluminal stent.¹⁰

We have used large diameter balloons for the angioplasty procedure safely despite previous reports of the friable nature of the duct after giving PG. Indeed, histological studies have shown that PG infusion results in an oedematous media with clear spaces and an increased number of breaks in the internal elastic lamina.¹¹ It is possible that these PG induced alterations of the wall of the arterial duct render it more compliant and therefore safer to dilate.

In this particular subset of patients with critical pulmonary valve stenosis and severe right ventricular hypoplasia arterial duct angioplasty provides adequate sustained pulmonary blood supply while right ventricular growth is occurring. It also avoids a surgical systemic to pulmonary shunt that would require a further operation to either take it down surgically or occlude it with a detachable balloon when it is no longer required.¹²

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