

NATIONAL INSTITUTE FOR CLINICAL EXCELLENCE

INTERVENTIONAL PROCEDURES PROGRAMME

Interventional procedure overview of balloon angioplasty of pulmonary vein stenosis in infancy

Introduction

This overview has been prepared to assist members of the Interventional Procedures Advisory Committee (IPAC) advise on the safety and efficacy of an interventional procedure previously reviewed by SERNIP. It is based on a rapid survey of published literature, review of the procedure by Specialist Advisors and review of the content of the SERNIP file. It should not be regarded as a definitive assessment of the procedure.

Date prepared

This overview was prepared by Bazian Ltd in March 2003.

Procedure name

Balloon angioplasty of pulmonary vein stenosis

Specialty society

British Paediatric Cardiology Association

Indications

Pulmonary vein stenosis.

Pulmonary vein stenosis (narrowing) may be congenital, or acquired following surgery to correct other congenital cardiac anomalies. It is rare. Untreated, it leads to severe lung damage. It is often associated with other cardiac abnormalities.

Balloon angioplasty of pulmonary vein stenosis, sometimes combined with stenting, is a palliative treatment of children with a very poor prognosis, or sometimes a temporary measure for children awaiting further interventions.

The procedure involves inserting a catheter into a large blood vessel, and passing it into the narrowed area under X-ray control. A balloon is then inflated to relieve the narrowing. A stent may be inserted following dilatation to maintain patency. There is no reliable alternative treatment.

Efficacy

Weak evidence was found from small case series, showing that balloon angioplasty to pulmonary vein stenosis is efficacious in the immediate term. We found no evidence of efficacy after the immediate post-procedure period.

According to the Specialist Advisors, pulmonary veins are often resistant to dilatation. However, there is almost no role for surgical treatment of pulmonary vein stenosis, so even a partial relief of stenosis may have clinical benefits in the short term.

Safety

Very limited evidence was found that balloon angioplasty to pulmonary vein stenosis may cause venous tears.

According to the Specialist Advisors, complications of balloon angioplasty of pulmonary vein stenosis include death, rupture of myocardium, rupture of pulmonary vein, cerebral or other systemic embolism, arrhythmias, and sepsis.

Literature review**Appraisal criteria**

Studies of balloon angioplasty in children (including babies) with congenital or acquired pulmonary venous stenosis were included.

List of studies found

No systematic reviews or controlled studies were found.

Four small case series and a number of single case reports were found.

The table give details of the four case series.¹⁻⁴

Summary of key efficacy and safety findings (1)

Study details	Key efficacy findings	Key safety findings	Key reliability, generalisability and validity issues
Lock, 1984 ¹ USA Case series n=5 children with pulmonary venous stenosis received balloon angioplasty All had other severe cardiac anomalies	None had clinical improvement	Venous tear leading to mediastinal haemorrhage: 1 Haemoptysis: 1 4/5 died within 4 months, though not likely to be due to procedure	Small case series
Dupuis, 1994 ² Location not clear, probably France Case series n=4 children with pulmonary venous stenosis Some received balloon angioplasty, not clear how many	'None of the treatments were satisfactory'	None provided	Published in French Data extracted from abstract
Driscoll, 1982 ³ USA Case series n=3 babies with pulmonary venous stenosis received balloon angioplasty All had other severe cardiac anomalies	All had immediate reduction in pulmonary vein pressure Clinical improvement: 1/3 (angiographic evidence of relief of stenosis – same patient that dies of infection)	Died of infection within 36 hours: 1	Small case series
Mendelsohn, 1993 ⁴ USA Case series n=3 babies with pulmonary venous stenosis received balloon angioplasty followed by stenting	Fall in right ventricular systolic pressure, mean pressure gradient, and wedge pressure	Puncture of distal vein: 1/3, sealed spontaneously Restenosis: 3/3	Small case series

Validity and generalisability of the studies

All the studies found were small case series. These provide limited evidence about efficacy and safety of the balloon dilatation in pulmonary vein stenosis.

Specialist advisor's opinion / advisors' opinions

Specialist advice was sought from consultants who have been nominated or ratified by their Specialist Society or Royal College.

- Complete success rare
- Children with pulmonary vein stenosis are often chronically ill with a very uncertain prognosis
- Should be undertaken in specialist centre by trained personnel, with on site paediatric cardiac surgeon

References

1. Lock JE, Bass JL, Castaneda-Zuniga W, Fuhrman BP, et al. Dilation angioplasty of congenital or operative narrowings of venous channels. *Circulation* 1984;70:457-64.
 2. Dupuis C, Rey C, Godart F, Vliers A, et al. Scimitar syndrome complicated by stenosis of the right pulmonary vein. Apropos of 4 cases. [French] *Archives des Maladies du Coeur et des Vaisseaux* 1994; 86: 607-613.
 3. Driscoll DJ, Hesslein PS, Mullins CE. Congenital stenosis of individual pulmonary veins: Clinical spectrum and unsuccessful treatment by transvenous balloon dilation. *American Journal of Cardiology* 1982; 49: 1767-72.
 4. Mendelsohn AM, Bove EL, Lupinetti FM, Crowley DC, et al. Intraoperative and percutaneous stenting of congenital pulmonary artery and vein stenosis. *Circulation* 1993; 88: 210-17.
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