

Percutaneous fetal balloon valvuloplasty for pulmonary atresia with intact ventricular septum

Interventional procedures guidance

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[nice.org.uk/guidance/ipg176](https://www.nice.org.uk/guidance/ipg176)

Your responsibility

This guidance represents the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, healthcare professionals are expected to take this guidance fully into account. However, the guidance does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or guardian or carer.

Commissioners and/or providers have a responsibility to implement the guidance, in their local context, in light of their duties to have due regard to the need to eliminate unlawful discrimination, advance equality of opportunity, and foster good relations. Nothing in this guidance should be interpreted in a way that would be inconsistent with compliance with those duties.

Commissioners and providers have a responsibility to promote an environmentally sustainable health and care system and should assess and reduce the environmental impact of implementing NICE recommendations wherever possible.

1 Guidance

- 1.1 Current evidence on the safety and efficacy of percutaneous fetal balloon valvuloplasty for pulmonary atresia with intact ventricular septum does not appear adequate for this procedure to be used without special arrangements for consent and for audit or research.

- 1.2 Clinicians wishing to undertake percutaneous fetal balloon valvuloplasty for pulmonary atresia with intact ventricular septum should take the following actions.
- Inform the clinical governance leads in their Trusts.
 - Ensure that parents understand the uncertainty about the procedure's safety and efficacy. Clinicians should provide parents with clear written information, and with counselling and support both before and after the procedure. In addition, use of the Institute's [information for the public](#) is recommended.
 - Audit and review the clinical outcomes of percutaneous fetal balloon valvuloplasty for pulmonary atresia with intact ventricular septum.
- 1.3 This procedure should only be performed in centres specialising in invasive fetal medicine and in the context of a multidisciplinary team including a consultant in fetal medicine, a paediatric cardiologist, a neonatologist, a specialist midwife and a paediatric cardiac surgeon.
- 1.4 An intention-to-treat registry has been developed by the [Association for European Paediatric Cardiology](#), and clinicians are encouraged to enter all cases into this registry.
- 1.5 Further publication on the criteria for selecting patients for this procedure rather than treating them conservatively until delivery will be useful. The Institute may review the procedure upon publication of further evidence.

2 The procedure

2.1 *Indications*

- 2.1.1 Congenital heart defects are the most common type of birth defect and include critical pulmonary stenosis, pulmonary atresia with a ventricular septal defect and pulmonary atresia with intact ventricular septum (PAIVS). In pulmonary atresia the pulmonary valve orifice is completely closed, thereby obstructing the outflow of blood from the heart to the lungs. Before birth oxygenated blood can still reach the systemic circulation by passing through the foramen ovale and any ventricular septal defect (VSD) to the left side of the heart. After birth the foramen ovale, VSD and ductus arteriosus must remain open to allow blood to

pass to the lungs to become oxygenated. In PAIVS the absence of blood flow at ventricular level can result in severe hypoxia soon after birth if the flow through the foramen ovale and ductus arteriosus are not maintained.

- 2.1.2 PAIVS is associated with underdevelopment of the tricuspid valve and the right ventricle but relatively normal growth of the pulmonary vessels and vascular bed. About 3% of fetuses with PAIVS will die in utero from hydrops. Severe cases of PAIVS are rare but carry a high rate of postnatal morbidity and mortality.
- 2.1.3 Many fetuses diagnosed with PAIVS will survive until birth, and treatment is then possible. About 50% of children with PAIVS have a functional biventricular heart, and overall survival from birth to 5 years is about 85%. Given the complex staged open cardiac surgery that some infants and children will require, some parents will request termination of pregnancy.
- 2.1.4 For babies born with PAIVS a staged approach to treatment is undertaken. Postnatal balloon valvuloplasty is the preferred option initially to encourage remodelling and growth of the right ventricle. Further balloon valvuloplasty is often required with later valve replacement. If balloon valvuloplasty is unsuccessful then open cardiac surgical options are available.
- 2.1.5 The aim of fetal pulmonary balloon valvuloplasty is to prevent progressive damage to the ventricular muscle in utero, the development of hydrops and subsequent fetal death. This may also allow a greater chance of surgical success postnatally along with the preservation of a biventricular heart.
- 2.1.6 Fetal pulmonary balloon valvuloplasty may be considered where there is a high risk of deterioration before delivery, with an increased likelihood of postnatal mortality or morbidity. Improvements in fetal imaging have assisted in the identification of suitable cases.

2.2 *Outline of the procedure*

- 2.2.1 Fetal pulmonary balloon valvuloplasty is performed at 21–32 weeks' gestation under maternal local anaesthesia and sedation, by inserting a needle through the mother's abdominal wall into the uterine cavity under ultrasound guidance. Fetal analgesic is then injected before advancing the needle through the fetal

chest wall into the right ventricular infundibulum of the fetus. A guidewire is inserted through the needle and across the pulmonary valve. A balloon catheter is inserted and then inflated to dilate the stenotic valve. The catheter and needle are then withdrawn.

2.2.2 Fetal positioning is critical for success of the procedure.

2.3 *Efficacy*

2.3.1 There is limited published evidence on this procedure. The total number of reported cases in the published literature is less than 10 and the patients were highly selected and heterogeneous. The largest series reports on the outcomes of five fetuses following fetal pulmonary valvuloplasty. Technical success was achieved in three fetuses, with some improvements in fetal haemodynamics. All three children survived to have postnatal surgery and were alive at 2, 3 and 4.5 years, respectively. The two fetuses in whom the procedure failed were both delivered: one had surgery after birth, but both died in the newborn period. For more details, refer to the 'Sources of evidence' section.

2.3.2 None of the studies specifically reported on maternal outcomes.

2.3.3 The Specialist Advisors noted the lack of data on this procedure and the difficulty in basing judgements about efficacy purely on survival, when the condition is rare and patients are carefully selected for this procedure. Specialist Advisors also noted that the criteria for case selection are not yet clear.

2.4 *Safety*

2.4.1 There is limited published evidence on this procedure. In one report of two fetuses, both had pericardial effusions that resolved spontaneously. There was also one instance of transient pericardial effusion in a report of five fetuses.

2.4.2 None of the studies specifically reported on maternal outcomes. For more details, refer to the 'Sources of evidence' section.

2.4.3 The Specialist Advisors listed fetal death, bleeding, bradycardia, pericardial effusion and balloon rupture as potential complications. They also noted that

there was a risk of premature labour and possible maternal morbidity associated with the use of anaesthesia.

3 Further information

3.1 The Institute has issued interventional procedures guidance on [balloon valvuloplasty for aortic valve stenosis in adults and children](#), [balloon dilatation of pulmonary valve stenosis](#), [balloon angioplasty of pulmonary vein stenosis in infants](#), [balloon dilatation with or without stenting for pulmonary artery or non-valvar right ventricular outflow tract obstruction in children](#), [balloon dilatation of systemic to pulmonary arterial shunts in children](#), [radiofrequency valvotomy for pulmonary atresia](#) and [percutaneous fetal balloon valvuloplasty for aortic stenosis](#).

Andrew Dillon
Chief Executive
May 2006

Sources of evidence

The evidence considered by the Interventional Procedures Advisory Committee is described in the following document.

['Interventional procedure overview of percutaneous fetal pulmonary balloon valvuloplasty for pulmonary atresia with intact ventricular septum \(PAIVS\)'](#), September 2005.

Information for patients

NICE has produced [information on this procedure for patients and carers](#). It explains the nature of the procedure and the guidance issued by NICE, and has been written with patient consent in mind.

4 Changes since publication

As part of NICE's work programme, the current guidance was considered for review in June 2009 but did not meet the review criteria as set out in the IP process guide. This guidance therefore remains current.

19 January 2012: minor maintenance.

5 About this guidance

NICE interventional procedure guidance makes recommendations on the safety and efficacy of the procedure. It does not cover whether or not the NHS should fund a procedure. Funding decisions are taken by local NHS bodies after considering the clinical effectiveness of the procedure and whether it represents value for money for the NHS. It is for healthcare professionals and people using the NHS in England, Wales, Scotland and Northern Ireland, and is endorsed by Healthcare Improvement Scotland for implementation by NHSScotland.

This guidance was developed using the NICE [interventional procedure guidance](#) process.

It has been incorporated into the [NICE pathway on antenatal care](#), along with other related guidance and products.

We have produced a [summary of this guidance for patients and carers](#). Information about the evidence it is based on is also [available](#).

Your responsibility

This guidance represents the views of NICE and was arrived at after careful consideration of the available evidence. Healthcare professionals are expected to take it fully into account when exercising their clinical judgement. This guidance does not, however, override the individual responsibility of healthcare professionals to make appropriate decisions in the circumstances of the individual patient, in consultation with the patient and/or guardian or carer.

Implementation of this guidance is the responsibility of local commissioners and/or providers. Commissioners and providers are reminded that it is their responsibility to implement the guidance, in their local context, in light of their duties to avoid unlawful discrimination and to have regard to promoting equality of opportunity. Nothing in this guidance should be interpreted in a way which would be inconsistent with compliance with those duties.

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Endorsing organisation

This guidance has been endorsed by [Healthcare Improvement Scotland](#).