

Percutaneous fetal balloon valvuloplasty for aortic stenosis

1 Guidance

- 1.1 Current evidence on the safety and efficacy of percutaneous fetal balloon valvuloplasty for aortic stenosis 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 aortic stenosis 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 (available from www.nice.org.uk/IPG175publicinfo).
 - Audit and review the clinical outcomes of percutaneous fetal balloon valvuloplasty for aortic stenosis.
- 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 (www.aepc.org), and clinicians are encouraged to enter all cases into this registry.
- 1.5 Further publication on the criteria for patient selection 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 aortic valve stenosis. The severity of aortic valve stenosis ranges from mild to severe; severe cases are rare but carry a high rate of postnatal morbidity and mortality.

- 2.1.2 Severe aortic stenosis in early fetal life causes left ventricular dysfunction which may initially produce left ventricular dilatation. However, myocardial damage arrests left ventricular growth which can lead to hypoplastic left heart syndrome (HLHS). Obstruction to the left ventricular outflow tract increases pressure in the left ventricle and atrium. The foramen ovale allows blood to flow from left to right. However, if the foramen ovale closes before birth the resulting high left heart pressure is associated with fibrosis of the myocardium and pulmonary venous hypertension with arterialisation of the pulmonary veins. This condition is known as aortic stenosis with a restrictive interatrial communication, and it has a very poor prognosis. In addition, in HLHS the mitral valve and aortic arch may be underdeveloped.
- 2.1.3 Many fetuses with severe aortic stenosis will survive until birth. However, about 10% will die in utero either from hydrops associated with a restrictive interatrial communication or from a chromosomal abnormality. At birth, most neonates with severe aortic stenosis will not be eligible for biventricular heart repair; approximately 50% die during the first year of life in spite of surgical treatment. This prognosis can lead many parents to request termination of pregnancy.
- 2.1.4 For babies born with an adequate biventricular heart and aortic valve disease, postnatal balloon valvuloplasty is the initial preferred option to encourage remodelling and growth of the left ventricle. Further balloon valvuloplasty is often required with later valve replacement.
- 2.1.5 Staged reconstruction for HLHS requires up to three operations over 3 or more years and involves complex high-risk open-heart surgery.
- 2.1.6 The aim of fetal aortic balloon valvuloplasty is to prevent progressive damage to the ventricular muscle and development of pulmonary vascular hypertension in utero. This may allow a greater chance of surgical success postnatally.
- 2.1.7 Fetal aortic balloon valvuloplasty may be considered where there is a high risk of deterioration before delivery, with an increased likelihood of postnatal mortality and morbidity. Improvements in fetal imaging have assisted with the identification of suitable cases.

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This guidance is written in the following context

This guidance represents the view of the Institute which 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.

Interventional procedures guidance is for healthcare professionals and people using the NHS in England, Wales and Scotland.

This guidance is endorsed by NHS QIS for implementation by NHSScotland.

2.2 Outline of the procedure

- 2.2.1 Fetal aortic 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 left ventricle of the fetus. A guidewire is inserted through the needle and across the aortic valve. A balloon catheter is then inserted and 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 are limited published data on this procedure. The largest published series reports on 20 fetuses who underwent in utero aortic valvuloplasty. Technical success was achieved in 70% of fetuses (14/20), with a significant difference demonstrated in the growth of the mitral valve, aortic valve and ascending aorta compared with those whose parents declined the procedure or in whom it was technically unsuccessful (n = 10). Of the 14 technical successes, three babies were born with a biventricular heart, six were born with HLHS, two died in utero and a further three were unborn at the time of writing. In a second series of 12 fetuses with severe obstructions of the aortic valve, there were seven technical successes with only one still alive at the time of writing.
- 2.3.2 Few studies reported on maternal outcomes. In one study, maternal hospital stay was 2 days (range 1–7 days). For more details, refer to the 'Sources of evidence' section.
- 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.

2.4 Safety

- 2.4.1 There were three deaths from the procedure in a study of 20 fetuses. Two fetuses died within 24 hours of having the procedure, one due to severe hydrops and the other as a result of fetal stress. The third fetus died 3 days after the procedure due to severe bradycardia. In a second study of 12 fetuses, four died within 24 hours of the procedure. Two died from persistent bradycardia, one from bleeding and one at valvotomy after emergency delivery. Both studies listed balloon rupture and intraoperative fetal bradycardia as further complications. In the study of 20 fetuses, bradycardia occurred in 15 cases, with medication required in 73% (11/15).

- 2.4.2 Maternal morbidity was primarily related to the need for emergency caesarean section following fetal valvuloplasty. Three emergency caesareans were undertaken in a study of 12 fetuses, two for sustained bradycardias and one for chorioamnionitis. 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 (www.nice.org.uk/IPG078), balloon dilatation of pulmonary valve stenosis (www.nice.org.uk/IPG067), balloon angioplasty of pulmonary vein stenosis in infants (www.nice.org.uk/IPG075), balloon dilatation with or without stenting for pulmonary artery or non-valvar right ventricular outflow tract obstruction in children (www.nice.org.uk/IPG076), balloon dilatation of systemic to pulmonary arterial shunts in children (www.nice.org.uk/IPG077), radiofrequency valvotomy for pulmonary atresia (www.nice.org.uk/IPG095) and percutaneous fetal balloon valvuloplasty for pulmonary atresia with intact ventricular septum (www.nice.org.uk/IPG176).

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May 2006

Information for the public

NICE has produced information describing its guidance on this procedure for pregnant women, their partners and families, and those with a wider interest in healthcare. It explains the nature of the procedure and the decision made, and has been written with patient consent in mind. This information is available from www.nice.org.uk/IPG175publicinfo

Sources of evidence

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

'Interventional procedure overview of percutaneous fetal balloon valvuloplasty for aortic stenosis', September 2005.

Available from: www.nice.org.uk/ip322overview

Ordering information

Copies of this guidance can be obtained from the NHS Response Line by telephoning 0870 1555 455 and quoting reference number N1041. *Information for the public* can be obtained by quoting reference number N1042.

The distribution list for this guidance is available at www.nice.org.uk/IPG175distributionlist

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