Issue date: **July 2004**

NHS
National Institute for Clinical Excellence

Balloon angioplasty of pulmonary vein stenosis in infants

1 Guidance

1.1 Current evidence on the safety and efficacy of balloon angioplasty of pulmonary vein stenosis in infants does not appear adequate for this procedure to be used without special arrangements for consent and for audit or research. The available evidence suggests that the procedure is not efficacious. However, there are no special concerns about the safety of the procedure, especially in the context of very ill infants for whom it is used.

1.2 Clinicians wishing to undertake balloon angioplasty of pulmonary vein stenosis in infants should take the following actions:

- Inform the clinical governance leads in their Trusts.
- Ensure that the parents of patients understand that the limited available evidence indicates a lack of efficacy. Parents should be given clear written information. Use of the Institute's *Information for the Public* is recommended.
- Audit and review clinical outcomes of all patients having balloon angioplasty of pulmonary vein stenosis in infancy.

1.3 This procedure should only be offered to gravely ill infants with a very poor prognosis, and in the setting of a specialist paediatric cardiology unit.

1.4 The Department of Health runs the UK Central Cardiac Audit Database (UKCCAD) and clinicians are encouraged to enter all patients onto this database (www.ccad.org.uk).

1.5 Publication of safety and efficacy outcomes will be useful in reducing the current uncertainty. The Institute may review the procedure upon publication of further evidence.

2 The procedure

2.1 Indications

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

2.1.2 There is currently no reliable alternative treatment.

2.2 Outline of the procedure

2.2.1 Balloon angioplasty of pulmonary vein stenosis, sometimes combined with stenting, is a palliative treatment for children with a very poor prognosis, or is sometimes a temporary measure for children awaiting further interventions. The procedure involves inserting a catheter into the narrowed area under radiological guidance. A balloon is then inflated to relieve the narrowing. A stent may be inserted after dilatation to maintain patency.

Interventional Procedure Guidance 75

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. Health professionals are expected to take it fully into account when exercising their clinical judgement. This guidance does not, however, override the individual responsibility of health 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 health professionals and people using the NHS in England, Wales and Scotland.

2.3 Efficacy

2.3.1 The evidence was limited to four very small, poor-quality case series, the largest including only five patients. The two largest studies found no benefit from the procedure in any patients. Another study of three patients found an immediate reduction in pulmonary vein pressure in all the patients, as well as angiographic evidence of relief of stenosis in one patient. However, this patient died of infection within 36 hours of surgery. For more details, refer to the Sources of evidence (see right).

2.3.2 The Specialist Advisors considered that this procedure may have only short-term efficacy (if any at all), and that recurrence rates may be high. They also noted, however, that there was almost no role for surgery in this condition, and that even a partial result from this procedure may offer palliation in this group of patients.

2.4 Safety

2.4.1 Some of the main adverse events reported in the studies included: venous tear leading to mediastinal haemorrhage in 20% (1/5) of patients; haemoptysis in 20% (1/5) of patients; death caused by infection in 33% (1/3) of patients; and puncture of the distal vein in 33% (1/3) of patients. For more details, refer to the Sources of evidence.

2.4.2 The Specialist Advisors considered the main potential adverse events to be death, rupture of myocardium, rupture of pulmonary vein, cerebral or other systemic embolism, arrhythmias and sepsis.

Andrew Dillon
Chief Executive
July 2004

Information for the Public

The Institute has produced information describing its guidance on this procedure for patients, carers 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, in English and Welsh, from www.nice.org.uk/IPG075publicinfo

Sources of evidence

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

Interventional procedure overview of balloon angioplasty of pulmonary vein stenosis in infants, March 2003

Available from: www.nice.org.uk/ip153overview

Ordering information
Copies of this guidance can be obtained from the NHS Response Line by telephoning 0870 1555 455 and quoting reference number N0643. *Information for the Public* can be obtained by quoting reference number N0644 for the English version and N0645 for a version in English and Welsh.

The distribution list for this guidance is available on the NICE website at URL www.nice.org.uk/IPG075distributionlist

Published by the National Institute for Clinical Excellence, July 2004 ISBN: 1-84257-710-7

National Institute for Clinical Excellence
MidCity Place, 71 High Holborn, London WC1V 6NA, website: www.nice.org.uk

N0643 1P 20k July 04 (OAK)