

Hybrid procedure for interim management of hypoplastic left heart syndrome in neonates

Interventional procedures guidance

Published: 12 December 2007

www.nice.org.uk/guidance/ipg246

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, and specifically any special arrangements relating to the introduction of new interventional procedures. 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.

All problems (adverse events) related to a medicine or medical device used for treatment or in a procedure should be reported to the Medicines and Healthcare products Regulatory Agency using the [Yellow Card Scheme](#).

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. Providers should ensure that governance structures are in place to review, authorise and monitor the introduction of new devices and procedures.

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

The hybrid procedure to which these recommendations apply consists of pulmonary artery banding, stenting of the ductus arteriosus and, if necessary, atrial septostomy.

- 1.1 Current evidence on the safety and efficacy of the hybrid procedure for interim management of hypoplastic left heart syndrome (HLHS) in neonates does not cover sufficiently all the parts of the procedure (see italicised text above) when used in combination and synchronously. The procedure should therefore only be used with special arrangements for consent, audit or research, and clinical governance.
- 1.2 Clinicians wishing to undertake the hybrid procedure for interim management of HLHS in neonates should take the following actions.
 - Inform the clinical governance leads in their Trusts.
 - Ensure that parents or carers understand the uncertainty about the procedure's safety and efficacy, and understand that the child will require further operations. They should provide parents or carers with clear written information. In addition, use of NICE's information for the public is recommended.
 - Audit and review clinical outcomes of all patients having the hybrid procedure for interim management of HLHS in neonates
- 1.3 The procedure should only be undertaken in paediatric cardiology centres specialising in the treatment of HLHS.

- 1.4 The [National Institute for Cardiovascular Outcomes Research \(NICOR\)](#) runs the [UK Central Cardiac Audit Database \(UKCCAD\)](#) and clinicians are encouraged to enter all patients into this database.
- 1.5 Further publication about criteria for patient selection and on the particular combination of techniques used in the hybrid procedure would be useful. The Institute may review the procedure upon publication of further evidence.

2 The procedure

2.1 Indications

- 2.1.1 Hypoplastic left heart syndrome (HLHS) is a combination of congenital abnormalities of the left side of the heart. Without surgical intervention it is fatal in the first weeks of life.
- 2.1.2 The standard treatment for HLHS is a staged reconstruction requiring up to three complex open heart operations over three or more years (stages 1 to 3). Some children who survive staged reconstruction will reach adulthood in good health. However, some may need other cardiac procedures or a heart transplant at a later stage.

2.2 Outline of the procedure

- 2.2.1 The hybrid procedure is performed under general anaesthesia or conscious sedation as soon as possible after birth. The aim is to improve the systemic circulation, while restricting the blood flow to the lungs (to prevent the development of pulmonary hypertension). This delays the need for high-risk open heart surgery reconstruction until the patient is older. It involves:
- banding of the right and left branches of the pulmonary artery either by open surgery, via a median sternotomy, or by a percutaneous endovascular technique

- endovascular insertion of a stent into the ductus arteriosus, via a percutaneous approach
- atrial balloon septostomy by a percutaneous endovascular technique.

2.2.2 The individual components of the hybrid procedure may be carried out synchronously or may be staged depending on individual patient need.

2.3 Efficacy

- 2.3.1 In three case series including HLHS patients treated by the hybrid procedure, 90% (52 out of 58), 52% (15 out of 29) and 57% (8 out of 14) of patients survived to undergo stage 2 reconstruction (two and three patients in the first two studies were waiting for the stage 2 procedure at publication). Survival after the stage 2 operation was 88% (46 out of 52) and 75% (6 out of 8) in the first and third series, respectively.
- 2.3.2 In a case series of 40 patients, 88% (15 out of 17) of patients treated by the hybrid procedure survived the procedure, of whom 27% (4 out of 15) subsequently died before further surgical treatment, and 67% (10 out of 15) were treated with a heart transplant.
- 2.3.3 A study of 22 patients compared 5 high-risk HLHS patients treated by the hybrid procedure with 17 standard- and high-risk patients treated with stage 1 reconstruction. All five patients treated by the hybrid procedure and 65% (11 out of 17) of patients treated by stage 1 reconstruction survived their respective operations. Two of the five hybrid procedure survivors and 9% (1 out of 11) of stage 1 reconstruction survivors died before undergoing a stage 2 operation. For more details, see the [overview](#).
- 2.3.4 The Specialist Advisers stated that the efficacy of the hybrid procedure is uncertain because only small numbers of patients have been reported. One stated that the long-term efficacy, specifically survival following the hybrid procedure compared with the standard open heart procedure without the hybrid procedure, is not clear in the literature.

2.4 Safety

- 2.4.1 The case series of 58 patients reported 30-day mortality of 3% (2 out of 58) after the hybrid procedure. The case series of 29 patients reported five hospital deaths (17%).
- 2.4.2 In the case series of 40 patients, 35% (6 out of 17) had complications relating to an intraluminal banding device used to band the pulmonary artery. In two patients the banding device was larger than required, resulting in excessive restriction of blood flow, two further patients developed acute pulmonary artery occlusion at the site of the device and in a further two patients the device was either placed or embolised further into the pulmonary circulation. For more details, see the [overview](#).
- 2.4.3 The Specialist Advisers stated potential, theoretical events to include death, brain damage, bleeding, infection, heart failure, damage to pulmonary arteries, stent migration, stent stenosis, stent thrombosis, migration of the pulmonary artery bands and perforation of the ductus arteriosus. One Specialist Adviser suggested that the procedure may shift mortality from the neonatal period to the infant period.

3 Further information

- 3.1 NICE has issued [interventional procedures guidance on balloon dilatation of systemic to pulmonary arterial shunts in children](#), [balloon dilatation with or without stenting for pulmonary artery or non-valvar right ventricular outflow tract obstruction in children](#), [balloon angioplasty of pulmonary vein stenosis in infants](#), [endovascular atrial septostomy](#), [endovascular closure of patent ductus arteriosus](#) and [percutaneous balloon valvuloplasty for fetal critical aortic stenosis](#).

Sources of evidence

The evidence considered by the Interventional Procedures Advisory Committee is described in the [overview](#).

Information for patients

NICE has produced [information for the public on this procedure](#). It explains the nature of the procedure and the decision made, and has been written with patient consent in mind.

ISBN: 978-1-4731-6265-5

Endorsing organisation

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