

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

Interventional procedures guidance

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[www.nice.org.uk/guidance/ipg246](http://www.nice.org.uk/guidance/ipg246)

## 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 the Institute's [information for patients](#) ('Understanding NICE guidance') 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 Clinicians undertaking this procedure should enter all patients onto the Department of Health's [UK Central Cardiac Audit 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 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% (52/58), 52% (15/29) and 57% (8/14) 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% (46/52) and 75% (6/8) in the first and third series, respectively.

2.3.2 In a case series of 40 patients, 88% (15/17) of patients treated by the hybrid procedure survived the procedure, of whom 27% (4/15) subsequently died before further surgical treatment, and 67% (10/15) 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% (11/17) of patients treated by stage 1 reconstruction survived their respective operations. Two of the five hybrid procedure survivors and 9% (1/11) of stage 1 reconstruction survivors died before undergoing a stage 2 operation. For more details, refer to the 'Sources of evidence' section.

- 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% (2/58) 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% (6/17) 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, refer to the 'Sources of evidence' section.
- 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 The Institute has issued the following guidance: '[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 fetal balloon valvuloplasty for aortic stenosis](#)'.

Andrew Dillon  
Chief Executive  
December 2007

## Sources of evidence

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

'Interventional procedure overview of hybrid procedure for interim management of hypoplastic left heart syndrome (HLHS) in neonates', June 2007.

## Information for patients

NICE has produced information describing its guidance on this procedure for patients and their carers ('Understanding NICE guidance'). It explains the nature of the procedure and the decision made, and has been written with patient consent in mind.

## 4 Changes since publication

The guidance was considered for reassessment in January 2011 and it was concluded that NICE will not be updating this guidance at this stage. However, if you believe there is new evidence which should warrant a review of our guidance, please [contact us](#).

8 May 2012: minor maintenance.

## 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.

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.