

# Septostomy with or without amnioreduction for the treatment of twin-to-twin transfusion syndrome

## 1 Guidance

- 1.1 Current evidence on the safety and efficacy of septostomy, with or without amnioreduction, for the treatment of twin-to-twin transfusion syndrome (TTTS) 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 septostomy, with or without amnioreduction, for the treatment of TTTS should take the following actions.
- Inform the clinical governance leads in their Trusts.
  - Ensure that the parents understand the uncertainty about the safety and efficacy of the procedure, the range of treatment options available and that one or both twins may not survive. The parents should also understand that in spite of amnioreduction, there is still a risk of serious abnormalities in the development of the nervous system among survivors of TTTS. Clinicians should provide parents with clear written information. In addition, use of the Institute's information for patients ('Understanding NICE guidance') is recommended (available from [www.nice.org.uk/IPG199publicinfo](http://www.nice.org.uk/IPG199publicinfo)).
  - Audit and review clinical outcomes of all patients having septostomy with or without amnioreduction for the treatment of TTTS (see section 3.1).
- 1.3 Clinicians should consider case selection carefully because there is uncertainty about the stages of TTTS for which this procedure is appropriate.
- 1.4 This procedure should only be performed in centres specialising in fetal medicine and by an appropriately constituted multidisciplinary team.
- 1.5 Clinicians are encouraged to collaborate on longer-term data collection across the centres performing septostomy for the treatment of TTTS. The Institute may review the procedure upon publication of further evidence.

## 2 The procedure

### 2.1 Indications

- 2.1.1 Approximately 70% of monozygotic twins are monochorionic/diamniotic (one placenta with two amniotic sacs). TTTS affects approximately 15% of these pregnancies and perinatal mortality is up to 80% if the syndrome is left untreated. TTTS results from abnormal shunting of blood between the circulations of the unborn twins through anastomoses of the vessels of the shared placenta. Blood is transfused from the donor twin, whose growth becomes restricted and who develops oligohydramnios or anhydramnios (too little or absent amniotic fluid), to the recipient, who develops circulatory overload, cardiac compromise and polyhydramnios (too much amniotic fluid). The combination of polyhydramnios in the recipient and oligo/anhydramnios in the donor squashes the donor twin against the wall of the uterus. The syndrome is generally associated with high morbidity and mortality for both twins. Morbidity among survivors includes cardiac, renal and serious neurological impairment, such as cerebral palsy. About 15% of survivors have long-term neurological sequelae.
- 2.1.2 The options for managing TTTS include expectant management, amnioreduction, septostomy, laser ablation, and selective fetal termination using techniques such as umbilical cord occlusion. In some cases the treatment aim is to enable one twin to survive, as the chances for both surviving are extremely poor. Some parents may choose to terminate the pregnancy because of the high risk of perinatal morbidity and mortality in both twins and the risks of serious long-term morbidity in survivors.

### 2.2 Outline of the procedure

- 2.2.1 Under local anaesthesia and ultrasound guidance, an amniocentesis needle is inserted through the maternal abdominal wall and uterine wall. The

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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.

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

needle is then used to make a small hole in the membrane between the twins, allowing the fluid around the recipient twin to move into the donor twin's sac. Amnioreduction may also be performed before and/or after the septostomy; this is an established procedure in which excess amniotic fluid is removed via a needle passed through the uterine wall and into the amniotic sac.

## 2.3 Efficacy

- 2.3.1 In one randomised controlled trial that included 36 twin pregnancies treated with amnioreduction and 35 treated with septostomy with or without amnioreduction, perinatal survival of at least one twin (measured until hospital discharge) was similar in both groups (78% [28/36] in the amnioreduction group versus 80% [28/35] in the septostomy group; relative risk [RR] 0.94, 95% confidence interval [CI] 0.55 to 1.61,  $p = 0.82$ ). Survival of both twins was 50% (18/36) in the amnioreduction group compared with 60% (21/35) in the septostomy group (RR 0.82, 95% CI 0.52 to 1.30,  $p = 0.40$ ). In a cohort study, perinatal survival of both twins was 43% (3/7) in the amnioreduction group compared with 57% (4/7) in those who underwent septostomy with or without amnioreduction. Survival of at least one twin was 86% (6/7) in both groups. Overall survival for amnioreduction alone was 9/14 (64%), versus 10/14 (71%) in the septostomy group; this difference was not statistically significant.
- 2.3.2 In four case series, overall perinatal survival (the total number of babies born alive divided by the total number of affected fetuses) ranged from 46% (12/26) to 83% (20/24). The differences in survival reported among the studies may be attributed to varying severity of TTTS, the studies that included more severe cases reporting poorer results.
- 2.3.3 Mean gestational age at delivery ranged from 27 to 31 weeks. Both comparative studies reported that pregnancy was prolonged following septostomy compared with amnioreduction alone (31 and 30 weeks compared with 28 and 30 weeks,  $p = 0.24$  and  $p = 0.08$  respectively). Long-term neurological outcomes were not reported in the studies. For more details, refer to the 'Sources of evidence' section.
- 2.3.4 The Specialist Advisers commented that the best treatment for early-stage TTTS is still unclear. They noted that results from the two comparative studies indicate no significant advantage of septostomy and amnioreduction over amnioreduction alone.

## Ordering information

Copies of this guidance can be obtained from the NHS Response Line by telephoning 0870 1555 455 and quoting reference number N1166. 'Understanding NICE guidance' can be obtained by quoting reference number N1167.

The distribution list for this guidance is available at [www.nice.org.uk/IPG199distributionlist](http://www.nice.org.uk/IPG199distributionlist)

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Interventional procedures guidance makes recommendations on the safety and efficacy of a procedure. The guidance does not cover whether or not the NHS should fund a procedure. Decisions about funding are taken by local NHS bodies (primary care trusts and hospital trusts) after considering the clinical effectiveness of the procedure and whether it represents value for money for the NHS.

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## 2.4 Safety

- 2.4.1 The safety evidence is based on one randomised controlled trial and two case series. In general, safety data were not well reported in the studies.
- 2.4.2 In the randomised controlled trial, there were two cases of disruption of the intertwin membrane resulting in a monoamniotic twin gestation (one in each study arm) which is associated with increased risk of cord entanglement and/or cord prolapse. This complication has also been reported in two case reports. In a case series of 13 pregnancies, one woman went into spontaneous labour following septostomy and amnioreduction. This was thought to be related to placental damage. For more details, refer to the 'Sources of evidence' section.
- 2.4.3 One Specialist Adviser considered septostomy and amnioreduction to be a safe procedure, and noted that septostomy is often performed unintentionally during standard amnioreduction. All three Specialist Advisers highlighted that cord entanglement is a theoretical complication of septostomy.

## 3 Further information

- 3.1 This guidance requires that clinicians undertaking the procedure make special arrangements for audit. The Institute has identified relevant audit criteria and developed an audit tool (which is for use at local discretion) available from [www.nice.org.uk/IPG199](http://www.nice.org.uk/IPG199)
- 3.2 The Institute has issued interventional procedures guidance on intrauterine laser ablation of placental vessels for the treatment of twin-to-twin transfusion syndrome ([www.nice.org.uk/IPG198](http://www.nice.org.uk/IPG198)).

Andrew Dillon  
Chief Executive  
December 2006

## 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. This information is available from [www.nice.org.uk/IPG199publicinfo](http://www.nice.org.uk/IPG199publicinfo)

## Sources of evidence

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

'Interventional procedure overview of septostomy with or without amnioreduction for the treatment of twin-to-twin transfusion syndrome', March 2006.

Available from: [www.nice.org.uk/IP337overview](http://www.nice.org.uk/IP337overview)