

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

Interventional procedures guidance

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[nice.org.uk/guidance/ipg199](https://www.nice.org.uk/guidance/ipg199)

## 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. However, 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.

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.

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

- 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.
- 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 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.

## 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).
- 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](#).

Andrew Dillon  
Chief Executive  
December 2006

### *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.

### *Information for patients*

NICE has produced [information on this procedure for patients and carers](#) ('Understanding NICE guidance'). It explains the nature of the procedure and the guidance issued by NICE, and has been written with patient consent in mind.

### 4 Changes since publication

The guidance was considered for reassessment in December 2009 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](#).

18 January 2012: minor maintenance.

## 5 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](#). Tools to help you put the guidance into practice and information about the evidence it is based on are also [available](#).

### Your responsibility

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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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### Contact NICE

National Institute for Health and Clinical Excellence  
Level 1A, City Tower, Piccadilly Plaza, Manchester M1 4BT

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

[nice@nice.org.uk](mailto:nice@nice.org.uk)

0845 033 7780

### *Endorsing organisation*

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