NICE guidance

Thoracoscopic aortopexy for severe primary tracheomalacia

Interventional procedures guidance
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www.nice.org.uk/guidance/ipg243

1 Guidance

1.1 Current evidence on the safety and efficacy of thoracoscopic aortopexy for severe primary tracheomalacia is limited to small case series and reports. Therefore, clinicians wishing to undertake this procedure should make special arrangements for clinical governance, consent and for audit or research.

1.2 Clinicians wishing to undertake thoracoscopic aortopexy for severe primary tracheomalacia 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. They should explain the alternative management options and the likely need for additional treatments. They should provide parents or carers with clear, written information. Use of the Institute's information for patients ('Understanding NICE guidance') is recommended.

- Audit and review clinical outcomes of all patients having thoracoscopic aortopexy for severe primary tracheomalacia (see section 3.1).

1.3 Patient selection and treatment should be carried out by a multidisciplinary team, which should include a surgeon experienced in paediatric thoracoscopic procedures (see also section 2.5).

2 The procedure

2.1 Indications

2.1.1 Primary tracheomalacia is a congenital condition in which the tracheal wall is weak. When the weakness is severe, the trachea can collapse during inspiration, obstructing the normal airflow. It is often associated with congenital abnormalities of the oesophagus. Symptoms may include breathing problems, such as coughing, wheezing, high-pitched breathing (stridor), respiratory tract infections and reflex apnoea/bradycardia ('dying spells'). In addition, there may be feeding difficulties. Mild to moderate symptoms in children usually improve with age. Treatments for children with this condition include the use of humidified air, chest physiotherapy and medication to control infection. Supplemental oxygen is sometimes required and continuous positive airway pressure (CPAP) may be used to treat short-term respiratory distress.

2.1.2 Surgery may be required if conservative management fails or if symptoms are severe (such as in reflex apnoea/bradycardia). Surgical options include endoluminal or extraluminal stenting of the trachea, tracheostomy, segmental tracheal resection, and open aortopexy.

2.2 Outline of the procedure

2.2.1 Thoracoscopic aortopexy is performed under general anaesthesia. A telescope and other instruments are inserted into the mediastinum through small incisions on the chest wall and used to visualise the operative field and to carry out the procedure. The left lobe of the thymus is moved aside or removed to expose the aortic arch. Several polypropylene sutures are passed through the sternum and into the

mediastinum in close proximity to the aortic arch. These are tightened so that the aortic arch and the trachea that lies behind it are both pulled forward, making the tracheal wall less likely to collapse on inspiration. Adequacy of the procedure can be verified by intraoperative bronchoscopy, which shows an enlarged tracheal lumen.

2.3 Efficacy

2.3.1 In a case series of six children, there were two recurrences of life-threatening events after thoracoscopic aortopexy (at 2 and 4 weeks respectively), after which both children underwent a repeat procedure. At a mean follow-up of 27 months, all six children were described as 'doing well' with no further life-threatening events.

2.3.2 A case report stated that two children improved 'dramatically' after the procedure and all stridor disappeared. Oxygen saturation improved from less than 85–90% before surgery to 96–100% postoperatively. Both children gained weight and were well at 17-month and 27-month follow-up.

2.3.3 A second case report of a child treated with thoracoscopic aortopexy stated that a follow-up bronchoscopy 7 months after surgery showed the child had a patent airway with no significant collapse. At 22-month follow-up the child was well, no longer fatigued easily with activity and had experienced no more reflex apnoea/bradycardia spells.

2.3.4 A third case report of a child treated with thoracoscopic aortopexy reported appropriate growth and no feeding or respiratory difficulties at follow-up (period not stated).

2.3.5 A fourth case report stated that there was immediate resolution of tracheomalacia after thoracoscopic aortopexy and long-term resolution of clinical respiratory symptoms at 1 year. For more details, refer to the 'Sources of evidence' section.

2.3.6 The Specialist Advisers listed key efficacy outcomes as improvement of symptoms (cyanotic episodes, stridor and lower respiratory tract infections), the ability to breathe unsupported, the degree of correction

as assessed at bronchoscopy, and growth (assessed using standard growth charts).

2.4 Safety

2.4.1 The case series of six children reported that no adverse events occurred. Two of the four case reports stated that there were no complications associated with the procedure and one stated that there was 'no notable blood loss'. One case report stated that extubation was delayed because of upper airway oedema (resolved with steroid treatment). For more details, refer to the 'Sources of evidence' section.

2.4.2 The Specialist Advisers listed potential adverse events as life-threatening haemorrhage that cannot be controlled without opening the chest, temporary or permanent injury to one or both phrenic nerves, pneumothorax, and mediastinitis.

2.5 Other comments

2.5.1 The Committee noted that the National Commissioning Group (NCG), which has a remit to commission highly specialised national services for very rare conditions for the population of England, has designated and commissioned Great Ormond Street Hospital to provide a service for complex tracheal disease, including severe tracheomalacia. Scottish residents also have access to the service under an agreement between the NCG and the National Services Division, Scotland. Health Commission Wales has a separate agreement with the provider for Welsh residents.

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 has developed an audit tool (which is for use at local discretion).

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 thoracoscopic aortopexy for severe primary tracheomalacia', April 2007.

Information for patients

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

4 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.

Changes since publication

14 January 2012: minor maintenance.

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.