NATIONAL INSTITUTE FOR HEALTH AND CLINICAL EXCELLENCE

INTERVENTIONAL PROCEDURES PROGRAMME

Interventional procedure overview of thoracoscopic aortopexy for severe primary tracheomalacia

Tracheomalacia is a weakness and floppiness of the main airway (the trachea) and may cause breathing difficulties. It usually presents at birth but may develop later in life. Aortopexy involves attaching the aorta to the sternum with sutures. This pulls the front wall of the trachea forwards to prevent it from collapsing. The thoracoscopic procedure is performed using special instruments inserted through several small incisions in the chest and with the help of a camera ('keyhole' surgery).

Introduction

This overview has been prepared to assist members of the Interventional Procedures Advisory Committee (IPAC) in making recommendations about the safety and efficacy of an interventional procedure. It is based on a rapid review of the medical literature and specialist opinion. It should not be regarded as a definitive assessment of the procedure.

Date prepared

This overview was prepared in April 2007.

Procedure name

- Thoracoscopic aortopexy
- Thoracoscopic aortocardiosternopexy
- Thoracoscopic aortosternopexy

Specialty societies

- Society of Cardiothoracic Surgeons of Great Britain and Ireland
- British Association of Otorhinolaryngologists, Head and Neck Surgeons
- British Association of Paediatric Surgeons
- Royal College of Paediatrics and Child Health
- British Thoracic Society

Description

Indications

Severe primary tracheomalacia

The trachea is normally a rigid structure which remains open throughout respiration. In tracheomalacia, the walls of the trachea are weak and floppy, collapsing during respiration resulting in obstruction to normal airflow. It may be either primary (intrinsic) or secondary. Primary tracheomalacia is a congenital condition that occurs when the cartilage of the trachea does not develop properly. It is often associated with other congenital abnormalities such as oesophageal atresia and tracheo–oesophageal fistula. Secondary tracheomalacia is an acquired condition that occurs when normal cartilage in the trachea degenerates, either due to external pressure (for example, from a tumour) or as a result of prolonged intubation or chronic infection involving the trachea. Symptoms of tracheomalacia can range from mild to severe and include breathing problems that get worse with coughing, crying or feeding, wheezing, high-pitched breathing (stridor), coughing, respiratory tract infections and reflex apnoea or ‘dying spells’.

Current treatment and alternatives

Mild-to-moderate symptoms in infants usually improve with age. Treatment is therefore conservative and includes 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.

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

What the procedure involves

Thoracoscopic aortopexy is performed under general anaesthesia. Intraoperative bronchoscopy is usually also used. Several small ports are inserted into the mediastinum through the left or right chest wall. One trocar is used to insert a fine telescope to visualise the procedure. The left lobe of the thymus is dissected and mobilised or excised to expose the aortic arch. A narrow spinal needle is passed through the sternum to determine proper alignment of the suture and aortic arch. The needle is removed and a small incision is made at the site. A suture is passed through the incision, through the sternum, into the mediastinum and around the aortic arch. The suture is then brought back out through the sternum near the original pass and out through the same incision. Two or three additional sutures are placed in a similar fashion, moving slightly laterally on the aortic arch. The aorta is elevated by pulling tension on the sutures, which are then tied individually. The trachea lies immediately behind the aortic arch and its anterior wall is drawn forward by this realignment of the aorta, so it is less likely to collapse

on inspiration. Adequacy of the aortopexy can be verified by visualisation of an enlarged tracheal lumen on intraoperative bronchoscopy.

Efficacy

The Specialist Advisers listed the key efficacy outcomes as improvement in symptoms of tracheomalacia (including cyanotic episodes, stridor and lower respiratory tract infections), the ability to breathe unsupported, degree of correction as assessed at bronchoscopy and growth (assessed from standard growth charts).

In a case series of six children, there were two recurrences of life-threatening events after the procedure (at two and four weeks respectively). Both children underwent redo thoracoscopic aortopexies. After a mean follow-up of 27 months, all six children were described as doing well with no further life-threatening events.[1]

In a case report, 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 27 and 17 months after the operation, respectively.[2]

A second case report stated that a child treated with thoracoscopic aortopexy showed appropriate growth and had no feeding or respiratory difficulties at follow-up (period not stated).[3]

A third case report stated that follow-up bronchoscopy 7 months after surgery showed a widely patent airway with no significant collapse. At 22 months’ follow-up the child was well, no longer fatigued easily with activity and had experienced no more ‘dying spells’.[4]

Safety

The Specialist Advisers listed potential adverse events as life-threatening haemorrhage, temporary or permanent injury to one or both phrenic nerves, pneumothorax and mediastinitis.

The case series of six children reported that no adverse events occurred.[1] Two of the three case reports stated that there were no complications associated with the procedure and one stated that there was ‘no notable blood loss’.[2,4] One case report stated that extubation was delayed because of upper airway oedema (which resolved after a short course of intravenous steroid).[3]

Literature review

Rapid review of literature

The medical literature was searched to identify studies and reviews relevant to thoracoscopic aortopexy for severe tracheomalacia. Searches were

conducted via the following databases, covering the period from their commencement to 29/03/2007: Medline, PreMedline, EMBASE, Cochrane Library and other databases. Trial registries and the Internet were also searched. No language restriction was applied to the searches. (See appendix A for details of search strategy.)

The following selection criteria (Table 1) were applied to the abstracts identified by the literature search. Where these criteria could not be determined from the abstracts the full paper was retrieved.

Table 1 Inclusion criteria for identification of relevant studies

Characteristic	Criteria
Publication type	Clinical studies were included. Emphasis was placed on identifying good quality studies. Abstracts were excluded where no clinical outcomes were reported, or where the paper was a review, editorial or laboratory or animal study. Conference abstracts were also excluded because of the difficulty of appraising methodology.
Patient	Patients with severe tracheomalacia
Intervention/test	Thoracoscopic aortopexy
Outcome	Articles were retrieved if the abstract contained information relevant to the safety and/or efficacy.
Language	Non-English-language articles were excluded unless they were thought to add substantively to the English-language evidence base.

List of studies included in the overview

This overview is based on one case series of six thoracoscopic aortopexies, three case reports and a case series of 28 aortopexies, one of which was performed thoracoscopically .[1–5]

Existing reviews on this procedure

No published reviews on thoracoscopic aortopexy were identified at the time of the literature search. A Cochrane Review on *Interventions for primary (intrinsic) tracheomalacia in children* was published in 2005.[6] No randomised controlled trials (RCTs) were found. The review briefly summarised eight recent non-controlled studies, none of which included thoracoscopic aortopexy.[7–14] Four case series reported on a total of 113 patients who underwent open aortopexy for tracheomalacia. One of these studies reported that symptoms improved markedly or disappeared within the first 3 months in 97% (28/29) of patients, and there was no early or late mortality. Morbidity associated with surgery included reversible lesion of the phrenic nerve in 2 patients and pneumothorax in 3 patients. Another study reported no intraoperative or postoperative mortality, and permanent relief of symptoms in 81% (13/16) of children. The third study reported improvement in airway obstruction in 96% (46/48) of children. The fourth case series of 54 children with airway problems (20 with tracheomalacia) reported that there were no intraoperative deaths. Of the remaining four non-controlled studies summarised in the review, two described stenting procedures, one described surgical management that was not otherwise defined and one described CPAP.

The authors concluded that there is a lack of evidence to support any of the therapies currently used for the management of intrinsic tracheomalacia but that there is unlikely to be an RCT on surgically based management for children with severe life-threatening illness associated with tracheomalacia. RCTs are needed for patients with less severe disease and outcomes should include measurements of the trachea and physiologically based outcomes in addition to clinical outcomes.

Related NICE guidance

There is no NICE guidance related to this procedure.

Table 2 Summary of key efficacy and safety findings on thoracoscopic aortopexy for severe primary tracheomalacia

Abbreviations used:			
Study details	**Key efficacy findings**	**Key safety findings**	**Comments**
Van der Zee (2007)[1] **Case series** The Netherlands Study period: 2002–2005 **n = 6** Population: children with oesophageal atresia and life threatening events attributable to tracheomalacia 4 girls, 2 boys Mean age: 5 months (range 14 days to 12 months) Indications: inclusion and exclusion criteria not described in abstract Technique: thoracoscopic tracheoaortopexies. **Mean follow-up: 27 months (range 10–45)** Conflict of interest: none stated	All tracheoaortopexies could be performed thoracoscopically. There were two recurrences of life-threatening events (at 2 and 4 weeks respectively), which could be treated using thoracoscopy. In one patient, two additional sutures were placed. In the second patient, reexploration found only one suture. The other sutures seem to have dissolved and could not be retrieved. Three new sutures were placed. At follow-up, all the patients were doing well and had had no more life-threatening events.	The report states that 'all the children tolerated the procedure well, and besides the recurrence, no other adverse effects occurred.'	The authors state that this is the largest thoracoscopic series to date. The authors note that there is a learning curve.

Abbreviations used:			
Study details	**Key efficacy findings**	**Key safety findings**	**Comments**
Schaarschmidt K (2002)[2] **Case reports** Germany Study period: not stated **n = 2** Population: two young children with severe upper respiratory obstruction A 4.5 year old boy presented with extreme expiratory stridor, barking unproductive cough and respiratory pauses of 4 to 8 missing inspirations. Tracheoscopy showed complete collapse of the trachea. He was born with ventricular septal defect, spinal deformation, oesophageal atresia with proximal fistula and high imperforate anus. A 2-year old girl had extreme expiratory stridor at birth caused by a double aortic arch and right descending aorta. Open aortosternopexy was performed after birth but she sustained a left laryngeal nerve palsy and developed recurrent tracheomalacia with loud expiratory stridor. Technique: thoracoscopic aortopericardiosternopexy; innominate artery and pericardium were also fixed to the sternum. **Follow-up: 17 and 27 months respectively** Conflict of interest: none stated	In both children, Intraoperative and postoperative tracheoscopy after 1 year showed the trachea to be widely open throughout its length and remained so on forced suction. Operating time: 110–130 min After the operations, both children improved dramatically, coughed productively and all stridors disappeared. Oxygen saturations improved from less than 85–90% preoperatively to 96–100% postoperatively. It was reported that both children started to play happily, lost the permanent facial expression of grief or fear and started to speak loudly. They gained weight and learned to walk and speak without respiratory pauses, even on exertion. The children were well at 27 and 17 months after the operation, respectively.	Both operations were described as ‘uneventful’ and there was ‘no notable blood loss’.	The authors state that, to their knowledge, this is the first report of a redo thoracoscopic aortosternopexy.

Abbreviations used:			
Study details	**Key efficacy findings**	**Key safety findings**	**Comments**
Jensen A (2004)[3] **Case report** USA Study period: not stated **n = 1** Population: 55-day-old boy with a history of oesophageal atresia and tracheoesophageal fistula repair in first week of life and subsequent gastro-oesophageal reflux. The infant presented with stridor after a life-threatening apnoeic event associated with feeding. Clinical investigation, including oesophagogram and rigid bronchoscopy, showed severe tracheomalacia and oesophageal narrowing with proximal dilatation. Technique: thoracoscopic aortopexy with a four-segment fixation of the ascending aorta and the base of the innominate artery. The child then underwent a laparoscopic Nissen fundoplication. **Follow-up: not stated** Conflict of interest: none stated	The child was discharged home 9 days after surgery. Follow-up revealed 'appropriate growth and no feeding or respiratory difficulties' (follow-up period not stated).	Postoperative course: extubation delayed because of upper airway oedema; performed on postoperative day 6 after a short course of intravenous steroid).	

Abbreviations used:			
Study details	**Key efficacy findings**	**Key safety findings**	**Comments**
DeCou J (2001)[4] **Case report** USA Study period: not stated **n = 1** Population: 13-month-old girl with Down's syndrome who presented with recurrent 'dying spells' and easy fatigue. Bronchoscopy revealed tracheomalacia that extended along the entire length of the trachea and into the major bronchi. Technique: thoracoscopic aortopexy, in which the aortic sutures were passed directly through the sternum and then tied extracorporeally, burying the knots in the subcutaneous tissue. **Follow-up: 22 months** Conflict of interest: none stated	Postoperative hospital stay = 6 days Follow-up bronchoscopy 7 months after surgery showed a widely patent airway with no significant collapse. At 22 months' follow-up, the child was well, had experienced no more 'dying spells' and no longer fatigued easily with activity.	There were no postoperative complications.	

Abbreviations used:			
Study details	**Key efficacy findings**	**Key safety findings**	**Comments**
Dave S (2006)[5] **Case series** Australia Study period: 1981–2004 **n = 1 thoracoscopic aortopexy (part of a case series of 28 open aortopexy procedures)** Population: children with severe and localised tracheomalacia 16 boys, 12 girls Median age at diagnosis: 5 months (range 1–84) Indications: evidence of significant tracheomalacia on bronchoscopy associated with acute life-threatening events in 22; failure to extubate in 5; recurrent pneumonia in 1. Patients were excluded if the tracheomalacia was caused by vascular sling or double aortic arch anomaly. 15 patients had associated oesophageal atresia and 13 had primary tracheomalacia (including isolated aberrant innominate artery compression). Technique: 27 patients had aortopexy by thoracotomy (12 had muscle-sparing left thoracotomy) and 1 had a thoracoscopic aortopexy. **Mean follow-up: 8.4 years (range 6 months–12.6 years)** Conflict of interest: none stated	Outcomes for the patient treated thoracoscopically were not reported separately. Resolution of symptoms = 93% (26/28) Two children had persistent stridor but no further acute life-threatening events and the stridor resolved within 6 months of surgery. Recurrent chest infections and asthma-like symptoms were noted in 5 children on follow-up; 3 of them required more than one admission to hospital.	Outcomes for the patient treated thoracoscopically were not reported separately There were no deaths. **Early complications** • Lung collapse/consolidation = 11% (3/28) • Phrenic nerve palsy requiring diaphragmatic plication = 4% (1/28) • Inability to extubate due to engorged thymus = 4% (1/28) (redo thoracotomy was performed and the left thymic lobe was excised. Since then in this centre, in small infants and neonates, the left lobe of the thymus is usually excised). There were no cases of scoliosis or chest-wall deformity on follow-up.	Retrospective review of patient notes An additional patient (not included in this series) had a double aortic arch that was missed on preoperative barium swallow and echocardiography. After aortopexy, the child could not be extubated and further surgery was performed before the aortopexy was redone. The single patient treated with thoracoscopic aortopexy is not discussed separately. The authors state that the thoracoscopic approach shows promise and can be attempted in older infants and children.

Validity and generalisability of the studies

- Only one case series and four case reports of thoracoscopic aortopexy were identified, describing a total of 11 patients.
- All of the patients were young children.
- The technique used varied between studies.

Specialist advisers' opinions

Specialist advice was sought from consultants who have been nominated or ratified by their Specialist Society or Royal College.

Mr D Crabbe, Mr G MacKinlay, Mr G Morrison

- One Specialist Adviser described this procedure as novel with uncertain safety and efficacy and another described it as a novel method of performing an established operation. A third Adviser described it as a minor variation on an existing procedure which is unlikely to affect that procedure's safety and efficacy.
- Aortopexy is an uncommon procedure in the UK.
- The National Specialist Commissioning Advisory Group (NSCAG) has commissioned Great Ormond Street Hospital for Children, London, UK to provide a service for children with complex tracheal abnormalities, including severe tracheomalacia.
- Quantification of the severity of tracheomalacia is subjective and difficult.
- Tracheomalacia has a tendency to improve spontaneously as the child grows.
- Expertise and an established practice in open aortopexy is necessary, along with expertise in bronchoscopy and advanced thoracoscopic surgery in children.
- The procedure should be carried out in specialist centres.
- Outcome measures of benefit include symptomatic improvement, appropriate growth as measured on centile growth and weight charts, avoidance of tracheostomy or CPAP, percentage reduction on airway collapse seen on rigid bronchoscopy, and improvement in blood gases.
- The potential impact of this procedure on the NHS is minor in terms of numbers of patients and use of resources.

Issues for consideration by IPAC

- A variant of the procedure including 'sternopexy' of the pericardium and innominate artery (in addition to aortic arch) has been described.
- No evidence for thoracoscopic aortopexy in acquired / degenerative tracheomalacia was found.

References

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Appendix A: Literature search for thoracoscopic aortopexy for severe tracheomalacia

IP: 396 Thoracoscopic aortopexy for severe tracheomalacia		
Database	**Date searched**	**Version searched**
Cochrane Library	29/03/2007	2007, Issue 1
CRD databases (DARE & HTA)	29/03/2007	2007, Issue 1
Embase	29/03/2007	1980 to 2007 Week 12
Medline	28/03/2007	1950 to March Week 2 2007
Premedline	29/03/2007	March 27, 2007
CINAHL	29/03/2007	1982 to March Week 4 2007
British Library Inside Conferences	29/03/2007	-
NRR	29/03/2007	2007, Issue 1
Controlled Trials Registry	29/03/2007	-

Search strategy used in Medline

The search strategy was adapted for use in the databases above

1 exp Thoracoscopy/
2 thoracoscop$.tw.
3 Thoracotomy/
4 Thoracoto$.tw.
5 (Endoscop$ adj3 pleur$).tw.
6 Pleuroscop$.tw.
7 ((Thoro$ adj3 surg$) or Aortop$).tw.
8 Aortopex$.tw.
9 Aortocardio$.tw.
10 or/1-9
11 Tracheal Diseases/
12 (trachea$ adj3 diseas$).tw.
13 Tracheomalac$.tw.
14 (trache$ adj3 (compress$ or collap$ or obstruct$)).tw.
15 (Collaps$ adj3 airwa$).tw.
16 tracheobronchomalac$.tw.
17 (Flacc$ adj3 trache$).tw.
18 (trachea$ adj3 wall collap$).tw.
19 or/11-18
20 10 and 19
21 animals/
22 humans/
23 21 not (21 and 22)
24 20 not 23