Surgical outcome of slide tracheoplasty in patients with long congenital segment tracheal stenosis and single lung

Simone Speggiorin a,*, Michele Torre a,b, Derek J. Roebuck a, Clair A. McLaren a, Martin J. Elliott a

a The National Service for Severe Tracheal Disease in Children, The Great Ormond Street Hospital for Children NHS Trust, London, WC1N 3JH, UK
b Pediatric Surgery Department, Istituto G. Gaslini, 16148, Genoa, Italy

Received 20 December 2010; received in revised form 25 January 2011; accepted 31 January 2011; Available online 2 April 2011

Abstract

Objective: The aim of the study is to assess the surgical outcome of slide tracheoplasty in patients with congenital tracheal stenosis and single lung. Methods: Pre, intra- and postoperative data were collected. Anatomy and associated anomalies were described. Results: Seven patients (median age 5 months; range 39 days – 1 year) with single lung underwent slide tracheoplasty. Single right lung was present in four patients. Associated anomalies were present in four patients (56%) including vascular ring, left pulmonary artery (LPA) sling, VACTERL (abnormalities of the vertebrae, anus, cardiovascular tree, trachea, oesophagus, renal system, and limb buds) syndrome, atrial septal defect (ASD) and aberrant left subclavian artery. Six patients (85%) needed preoperative ventilation and two (28%) needed preoperative extracorporeal membrane oxygenation (ECMO). Median postoperative ventilation was 7 days (6 – 35 days). Two patients needed postoperative ECMO, one of whom required preoperative ECMO. Complications occurred in five patients (71%): reintubation due to pneumothorax, pneumonia and several tracheal dilatations due to recurrent tracheal stenosis, which was eventually stented. There was one hospital death in a patient, who could not be weaned off ECMO due to severe distal malacia. At a median follow-up of 16 months (7 days – 7 years), all survivors are in good clinical condition and without additional stenting. Conclusion: Slide tracheoplasty can be performed in patients with single lung and tracheal stenosis with a good surgical outcome.

Keywords: Lung agenesis; Slide tracheoplasty

1. Introduction

Long-segment congenital tracheal stenosis (LSCTS) is a rare and life-threatening malformation, usually characterised by the presence of complete cartilaginous rings along more than two-thirds of the length of the trachea [1]. The incidence of this anomaly is unknown, and the clinical presentation can vary from almost asymptomatic patients to severe or fatal respiratory failure.

Slide tracheoplasty (STP) is currently considered the surgical repair of choice for LSCTS because it is associated with lower morbidity and mortality than other techniques [1–5]. It is applicable to all anatomical variants of LSCTS. One such extreme variant is LSCTS associated with agenesis or severe hypoplasia of one lung [6]. Very few articles have documented this association, and these report a mortality ranging from 18% to 65% [7–9].

The aim of our article is to analyse the outcome of STP on this highly selected group of patients in our tracheal surgery population to determine the current era mortality and morbidity.

2. Material and methods

A retrospective review of medical notes was undertaken of patients with LSCTS and lung agenesis or severe hypoplasia of one lung, who had an STP between January 1995 and May 2010. Patients were identified from the database of the UK National Service for Severe Tracheal Disease in Children at the Great Ormond Street Hospital for Children NHS Trust London.

Demographic data, airway anatomy, cardiac and non-cardiac associated anomalies, radiological investigations, the use of preoperative mechanical ventilation, preoperative and postoperative extracorporeal membrane oxygenation (ECMO), surgical airway and cardiac procedures, postoperative management, complications and postoperative length of stay were recorded. Follow-up data, including the

1010-7940/$ – see front matter © 2011 European Association for Cardio-Thoracic Surgery. Published by Elsevier B.V. All rights reserved.
doi:10.1016/j.ejcts.2011.01.075
Table 1. Overview of the patients’ characteristics.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex/age (months)</th>
<th>Morphologic single lung</th>
<th>TB anatomy</th>
<th>Other anomalies</th>
<th>Pre-surgery ventilation</th>
<th>Pre-surgery ECMO</th>
<th>CPB (min)</th>
<th>Associated surgical procedures</th>
<th>Postoperative complications</th>
<th>Post-surgical TB interventions</th>
<th>Postoperative ECMO</th>
<th>Ventilation time (days)</th>
<th>Hospital death</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F/12</td>
<td>Left</td>
<td>LCTS, vascular compression at carina</td>
<td>Vascular ring</td>
<td>Yes</td>
<td>No</td>
<td>90</td>
<td>Vascular ring division</td>
<td>—</td>
<td>—</td>
<td>No</td>
<td>6</td>
<td>—</td>
</tr>
<tr>
<td>2</td>
<td>F/5</td>
<td>Right</td>
<td>LCTS</td>
<td>PDA</td>
<td>Yes</td>
<td>Yes</td>
<td>70</td>
<td>PDA ligation and division</td>
<td>—</td>
<td>—</td>
<td>Yes</td>
<td>17</td>
<td>—</td>
</tr>
<tr>
<td>3</td>
<td>F/2</td>
<td>Right</td>
<td>LCTS</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>51</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>No</td>
<td>35</td>
<td>—</td>
</tr>
<tr>
<td>4</td>
<td>M/1</td>
<td>Left</td>
<td>LCTS, severe distal malacia</td>
<td>Imperforate anus (s/p gastrostomy), VACTERL syndrome, aberrant left subclavian artery</td>
<td>Yes</td>
<td>No</td>
<td>103</td>
<td>PTP of the distal bronchus, aortoectomy</td>
<td>Unable to ventilate due to malacia</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>—</td>
</tr>
<tr>
<td>5</td>
<td>F/4</td>
<td>Right (left lung hypoplasia)</td>
<td>LCTS, small malacic LMB</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>76</td>
<td>Aortoectomy</td>
<td>Pneumonia after inspiration (GOR), unable to swallow</td>
<td>—</td>
<td>No</td>
<td>7</td>
<td>—</td>
</tr>
<tr>
<td>6</td>
<td>F/7</td>
<td>Right</td>
<td>LCTS</td>
<td>Situs viscerum inversus, LPA sling</td>
<td>Yes</td>
<td>No</td>
<td>65</td>
<td>Aortoectomy</td>
<td>Reintubation, pneumothorax</td>
<td>—</td>
<td>No</td>
<td>7</td>
<td>—</td>
</tr>
<tr>
<td>7</td>
<td>M/7</td>
<td>Left</td>
<td>LCTS, CTR from cricoid to upper lobe bronchus</td>
<td>Situs viscerum inversus, GOR, ASD</td>
<td>Yes</td>
<td>Yes</td>
<td>65</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>No</td>
<td>10</td>
<td>—</td>
</tr>
</tbody>
</table>

ASD: atrial septal defect; CTR: complete tracheal ring; CPB: cardiopulmonary bypass; ECMO: extracorporeal membrane oxygenator; GOR: gastro-oesophageal reflux; LCTS: long congenital tracheal stenosis; LMB: left main bronchus; LPA: left pulmonary artery; PDA: patent ductus arteriosus; PTP: patch tracheoplasty; and TB: tracheo-bronchial.
need for interventional radiological (IR) procedures (tracheal or bronchial dilatations and/or stenting) were also collected.

In the presence of situs viscerum inversus (SVI), the single lung has been named according to the anatomic morphology (i.e., right-sided anatomical left lung).

Preoperative investigations included chest computed tomography (CT) scan, bronchoscopy, bronchography, echocardiogram and optical coherence tomography (OCT), which uses long-wave-length light to identify the presence of complete tracheal rings.

The perioperative management we employed in these patients has been previously described [2].

Associated cardiovascular anomalies were corrected during the same surgical session (Table 1).

3. Results

Seven patients (two boys and five girls) with LCSTS and functional single lung were observed. All patients presented with complete tracheal rings creating an LSCTS. These patients represent 9% of the patients, who have undergone STP in our institution to date. The demographic, anatomical and perioperative data are shown in Table 1 and Fig. 1.

The STP was performed involving the single main bronchus in all cases, as previously described [1,5].

Median cardiopulmonary bypass time was 70 min (range 51–103 min), and median postoperative mechanical ventilation time was 8.5 days (range 6–35 days).

Patient #2 required postoperative venovenous ECMO (V-V-ECMO) for increasing end-tidal CO₂ (6–8 mmHg) despite adequate oxygenation, and was successfully weaned off 48 h later.

In Patient #3, the postoperative period was characterised by recurrent hypercapnic episodes with prolonged expiratory time. Bronchoscopy showed a tight narrowing (1 mm) of the airway distal to the repair. In the following 30 days, the patient remained intubated and ventilated, whilst the narrow segment was progressively dilatated by balloon to increasing diameters. As the maximal accepted dilatation diameter was 5 mm, and the distal part of the trachea appeared floppy, a stent (Palmaz Genesis 7 x 18 mm, Cordis Corporation, Miami, FL, USA) was implanted. The patient was successfully weaned from the ventilator and extubated after 2 further days.

Patient #4 was the only one who did not survive. The preoperative investigations revealed severe malacia of the single left bronchus. The STP was thus anticipated to be high risk. After the STP was performed, severe malacic flattening of the distal bronchus was evident. The distal part of the STP was converted into a patch tracheoplasty (PTP) by a longitudinal incision of the trachea extending through the repair into the left main bronchus (LMB). An autologous pericardial patch was sewn with interrupted ‘U’ 5/0 polydioxanone (PDS) (PDS II, Ethicon, Inc., Edinburgh, UK) stitches. The endotracheal tube was placed in the distal part of the repair to keep the PTP patent. It was considered that the aorta might have been compressing the distal malacic bronchus, allowing only an intermittent clearance of CO₂ from the lung. An anterior aortopexy was attempted to relieve the compression. However, a fibre-optic bronchoscopy in the operating room revealed a patent trachea, but a flat and severely malacic LMB. Venoarterial ECMO (V-A-ECMO) was commenced but treatment was electively withdrawn after 48 h, as ventilation proved impossible.
On postoperative day 11, Patient #5 developed pneumonia after aspiration due to gastro-oesophageal reflux whilst on the ward. Antibiotic treatment was commenced with good result.

Patient #6 required reintubation for acute respiratory failure due to a tension pneumothorax. A chest drain was inserted promptly, and the patient was extubated after 24 h.

With the exception of Patient #4, all patients were discharged home in a stable clinical condition and spontaneous breathing in room air.

4. Follow-up

The median follow-up was 1.7 years (range 5.8 months—7.1 years). Only one patient (Patient #3, who had the stent) has required further balloononing of the airways after developing intra-stent stenosis. After the stent has been implanted, a routine follow-up bronchoscopy has been performed every 6 months for the first 2 years, and then every 1 year. All others are in good clinical condition and free of respiratory symptoms.

5. Discussion

The presence of LCSTS and single lung or severe hypoplasia of one lung is a rare association connected with a high mortality (18—65%) [6—8]. Backer and associates reviewed the literature and reported a total of 28 patients with functional single lung [8], forming 10—20% of the total population of patients with LCSTS undergoing surgical repair. We have identified a comparable incidence in our cohort (7/77, 9%).

The association of single lung with LCSTS and other anomalies, such as cardiovascular, gastrointestinal or genetic, is not uncommon [5,8,10—11].

Left pulmonary artery (LPA) sling has been reported to occur in nearly 30% in these patients [8], similar to the incidence in the two-lung population where it varies from 33% to 48% [5,8]. In our experience, an LPA sling was present in only one patient (14%).

The association of single lung and chromosomal anomalies has been already reported by Cunningham and Mann [12], who identified that the majority of the patients with single lung had anomalies of the first and second branchial arches as well as radial ray defects [12]. This finding led them to speculate that the presence of single lung represents a feature of VACTERL (abnormalities of the vertebrae, anus, cardiovascular tree, trachea, oesophagus, renal system, and limb buds) anomaly [12]. In our experience, only one patient was diagnosed as having frank VACTERL syndrome. This patient (Patient #4) had the association of LCSTS and severe distal malacia, which strongly influenced the outcome.

In the past 2 years, the use of OCT has become a routine preoperative investigation performed at the time of bronchoscopy to inspect the presence of airway cartilage as complete tracheal rings or total absence as in malacia, as suggested by Ridgway and associates [14]. In our experience, this technique gives valuable and useful information on the anatomical structure of the airway. In retrospect, we believe that if OCT had been performed on this patient (Patient #4), we might not have proceeded to STP due to the severity of the malacia. SVI is a very rare anatomical condition occurring in less than 0.01% of the population. To our knowledge, no previous cases of LSCTS and SVI have been previously reported. SVI can be associated with Kartagener syndrome and ciliary defects, but in only one case was this reported in association with SL [13]. Interestingly, two of the patients we reviewed had SVI. These findings highlight the fact that, in the presence of a single lung, accurate genetic testing might help us to better understand the cause of these pathologies.

Another important issue is the preoperative ventilatory status. In this particular group of patients, there is a significantly higher necessity of preoperative mechanical ventilation in patients with one rather than two lungs (73% vs 25%) [8]. Our results are in line with Backer and associates, where all except one required preoperative mechanical ventilation. In two patients, ECMO was required to stabilise the haemodynamic status.

In the past decade, the outcome of surgery for congenital tracheal stenosis in patients with two lungs has improved considerably, due to a better pre- and postoperative management and evolution of the surgical technique [2]. LSCTS associated with single-lung patients still represent a population particularly challenging, with a reported mortality up to 65% of cases [6—8]. The difficulties are related not only to the preoperative clinical presentation or the surgical procedure, but also to morbidity with concern about long periods of intensive care management in the pre- and postoperative periods. For this reason, the decision to treat these patients is often challenged.

Few articles report the surgical outcome in this unique population. Backer and associates, reported 11 patients of LSTS with single lung, who underwent a variety of surgical procedures, including PTP in two patients, tracheal autograft in four, STP in three and tracheal resection in three [8]. Post-surgical early mortality, intended as mortality before discharge, was comparable with the population with two lungs (18% vs 17%) [8].

The series reported here represents the biggest group of patients with functional single lung, who underwent STP for LCSTS. The mortality rate (14%) that we have observed confirms that, in many cases, STP can be successful in these patients.

It is noteworthy that, within our population, no patient required pre- or postoperative tracheostomy, whereas in Chicago’s experience, as published by Backer and associates, it was necessary in 36% of the cases (4/11) [8].

STP is an excellent treatment for patients with single lung, and early follow-up suggests no difference in outcome from their two-lung peers. It will be important to report the medium- and long-term outcomes and quality of life in due course.

6. Limitations

This study reports only a small number of patients, and is a single-centre, single-surgeon experience. Follow-up is short, albeit encouraging, and we cannot find reason to decline STP in patients with a single lung.

7. Conclusion

Single or severely hypoplastic lung and LCSTS is a rare and life-threatening association. This unique population presents in many cases with associated anomalies, such as SVI and others. Despite the complexity of the perioperative management of these patients, STP has proven to be a reliable treatment for the relief of the tracheal stenosis, with excellent surgical outcome, comparable with the two-lung patients’ population. As a consequence, we suggest the presence of unilateral lung agenesis should not be considered a contraindication for STP.

Further effort should be put into understanding the quality of life of this group of patients.

References