Tracheal surgery in children: outcome of a 12-year survey

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INTRODUCTION

Paediatric tracheal surgery encompasses the management of a wide spectrum of diseases, both congenital and acquired. The reconstruction of paediatric airways requires strong technical and management skills due to the fact that infants, and neonates all the more, have an extremely reduced tracheal diameter [1, 2]. In recent years, the introduction of innovative surgical techniques, such as slide tracheoplasty, and the development of a multidisciplinary approach by a dedicated tracheal team have dramatically improved the survival rate of patients [3, 4]. Nevertheless, the postoperative course of infants undergoing tracheal surgery is still characterized by the need to follow specific postoperative procedures, such as balloon dilatation, stent positioning or reoperation for residual stenosis [5]. For this reason, different countries have equipped themselves with organized referral centres that treat a large number of cases [6–8]. At the same time, several institutional reports on the subject have been published worldwide [9].

Tracheal surgery is highly complex because it frequently involves heart, abdominal and lung malformations beyond those related to genetic syndromes [1, 2, 10]. These comorbidities have been shown to potentially aggravate the outcome of the operation even when the surgical repair of the trachea is perfect [10].

In this study, we reviewed the results, spanning the last 12 years, concerning the surgical management of congenital and acquired tracheal defects in children, with particular attention paid to the principle of multidisciplinary management in the treatment of tracheal and associated malformations.

CONCLUSIONS: The result of paediatric tracheal surgery depends on several factors. The number of cases treated at a particular centre is an important one, but our experience, although limited, can be compared with that at centres with a higher volume of cases. We emphasize the need for applying a multidisciplinary approach to master the surgical command of different reconstructive tracheal procedures, to manage associated defects, particularly cardiovascular defects, and to manage complications under endoscopic guidance. These can be considered the mainstays of building a successful tracheal programme.

Keywords: Pediatric tracheal surgery • Congenital heart disease • Endoscopic management • Tracheal team
Table 1: Summary of patient characteristics and outcomes

<table>
<thead>
<tr>
<th>Variables</th>
<th>Age group</th>
<th></th>
<th></th>
<th></th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>0–30 days</td>
<td>30 days–1 year</td>
<td>Over 1 year</td>
<td></td>
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<tr>
<td>Boys</td>
<td>15 (50.0)</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Girls</td>
<td>15 (50.0)</td>
<td></td>
<td></td>
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<tr>
<td>Weight, n (%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>≤3 kg</td>
<td>3 (75.0)</td>
<td>4 (22.2)</td>
<td>0 (0.0)</td>
<td></td>
<td>7 (23.3)</td>
</tr>
<tr>
<td>&gt;3 kg</td>
<td>1 (25.0)</td>
<td>14 (77.8)</td>
<td>8 (100.0)</td>
<td></td>
<td>23 (76.7)</td>
</tr>
<tr>
<td>Congenital tracheal stenosis, n (%)</td>
<td>3 (75.0)</td>
<td>17 (94.4)</td>
<td>5 (62.5)</td>
<td></td>
<td>25 (83.3)</td>
</tr>
<tr>
<td>Acquired lesions, n (%)</td>
<td>1 (25.0)</td>
<td>1 (5.6)</td>
<td>3 (37.5)</td>
<td></td>
<td>5 (16.6)</td>
</tr>
<tr>
<td>Associated malformations, n (%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cardiac</td>
<td>4 (100.0)</td>
<td>7 (38.8)</td>
<td>5 (62.5)</td>
<td></td>
<td>16 (53.3)</td>
</tr>
<tr>
<td>Bronchopulmonary</td>
<td>1 (25.0)</td>
<td>8 (44.4)</td>
<td>2 (25.0)</td>
<td></td>
<td>11 (36.6)</td>
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<tr>
<td>Operations, n (%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Slide tracheoplasty</td>
<td>4 (100.0)</td>
<td>16 (88.9)</td>
<td>3 (37.5)</td>
<td></td>
<td>23 (76.7)</td>
</tr>
<tr>
<td>End-to-end anastomosis</td>
<td>0 (0.0)</td>
<td>1 (5.6)</td>
<td>3 (37.5)</td>
<td></td>
<td>4 (13.3)</td>
</tr>
<tr>
<td>Complex reconstruction</td>
<td>0 (0.0)</td>
<td>1 (5.6)</td>
<td>2 (25.5)</td>
<td></td>
<td>3 (10.0)</td>
</tr>
<tr>
<td>Associated cardiac procedure, n (%)</td>
<td>4 (100.0)</td>
<td>8 (44.4)</td>
<td>4 (50.0)</td>
<td></td>
<td>16 (53.3)</td>
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<tr>
<td>Follow-up (months)</td>
<td></td>
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<tr>
<td>Median (minimum–maximum)</td>
<td>29.1 (11.3–39.4)</td>
<td>50.3 (2.2–88.7)</td>
<td>18.3 (2.8–49.7)</td>
<td>38.5 (2.2–88.7)</td>
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<tr>
<td>In-hospital mortality, n (%)</td>
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<td>Overall mortality, n (%)</td>
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PATIENTS AND METHODS

Sample population

We conducted a retrospective review of the records of all patients requiring tracheal surgery over a 12-year period (2005–17). Patients’ records were collected in a specific database; the institutional review boards approved this study and waived the need for individual consent.

A dedicated tracheal team managed all the patients. During the study period, 32 children underwent tracheal reconstruction for congenital or acquired tracheal lesions. However, the hospital and follow-up records were complete for only 30 patients, who thus represented our final study sample.

Patients were equally distributed between boys and girls (15 boys and 15 girls). The median age at operation was 7 months (15 days–9.6 years), and the median weight was 5.2 kg (2.8–34 kg). Four patients (13.3%) were neonates (0–30 days); overall, 22 (73.3%) patients underwent surgery before 1 year of age. The detailed characteristics of the sample population and of each age group are described in Table 1. Congenital tracheal lesions were defined as tracheal stenosis with complete tracheal rings. Acquired tracheal lesions that required surgery were identified in all other cases that were not characterized by complete tracheal rings. Twenty-five patients (83.3%) were diagnosed with congenital tracheal stenosis (CTS), whereas acquired lesions were found in 5 cases (16.5%). In the group of acquired lesions, 1 child (3.3%) had a double aortic arch with tracheal rupture and tracheovascular-oesophageal fistula; 2 patients (6.6%) were diagnosed with postintubation tracheal stenosis and 2 children (6.6%) showed the sequelae of compression by vascular rings (1 child with a double aortic arch and 1 with a right anomalous subclavian artery that had already been operated on in a different institute). All of them showed, at bronchoscopic evaluation, reduction in the tracheal diameter associated with softness of the tracheal wall. In all acquired cases, the involved tracheal tract appeared markedly damaged (confirmed intraoperatively) and was not susceptible to conservative treatment.

The trachea was involved in 60% of cases for its full length and in 40% of cases in the proximal-medium tract. In cases of CTS, the mean internal diameter was 1.5 mm with complete tracheal rings present in all patients. Associated malformations, when present, were bronchopulmonary in 11 patients (36.7%) and cardiovascular in 16 patients (53.3%, Supplementary Material Tables S1 and S2). A pulmonary artery sling was present in 8 patients (26.7%), and 2 children presented a double aortic arch (6.6%). Eight patients (26.7%) were on mechanical ventilation before tracheal surgery and 4 (13.2%) had preoperative tracheostomy.

Instrumental preoperative assessment

Patients were diagnosed with tracheal defects by well-trained operators. The examination was performed by a flexible (Karl Storz®; Germany; Ø 2.5/3.7/5.2 mm) or rigid bronchoscope (Karl Storz, Germany; Ø 3.5/4.5/5.5 mm). Bronchoscopy was used as the first diagnostic tool, and the variables measured were the following: the internal tracheal diameter and the length of the stenosis, defined by the number of tracheal rings involved and their characteristics, in particular, the presence of complete or incomplete tracheal rings. Finally, the presence of pulsatility on the tracheal wall was also described.

A chest computed tomography scan (contrast-mediated) was performed in all children to confirm the results of the endoscopic examination. The computed tomography (Philips Brilliance i-Dose 64 slice) scan was performed according to age and weight and adjusted following a low-dose protocol (120 or 80 kV; 120 or 80 mA; collimation 1.5 or 0.75 mm and gantry rotation 0.5 s). When required, a 3D reconstruction and print of the trachea were made for a better anatomical definition (Fig. 1). A preoperative chest radiograph, transthoracic echocardiogram and abdominal echogram completed the diagnostic assessment to...
determine the type of associated malformations. All cases were discussed in a preoperative meeting by all the medical personnel involved in the dedicated tracheal team.

Operative techniques

Operations were performed through a median sternotomy with the incision extending to the neck when the tracheal stenosis involved the proximal tracheal tract. Cardiopulmonary bypass with moderate hypothermia (rectal temperature 30°C) was used in all operations. Venous cannulation was accomplished by a single- or double-venous cannula, depending on the presence of associated cardiovascular malformations requiring simultaneous correction. All combined cardiovascular anomalies were treated simultaneously with tracheal surgery (Supplementary Material, Table S3), except for a single neonate in whom the diagnosis of CTS was made following the correction of the cardiac defect. If the estimated cross-clamp time to correct the cardiac anomalies was more than 60 min, we performed a single infusion in the aortic root using the Bretschneider histidine-tryptophan-ketoglutarate (HTK) solution, commercially known as Custodiol (Custodiol HTK, Köhler Chemie GmbH, Bensheim, Germany).

Tracheal reconstruction was performed using 1 of the 3 different techniques: a slide tracheoplasty was performed in 23 of 30 cases (76.7%), resection and end-to-end anastomosis were performed in 4 cases (13.3%) and complex reconstruction in 3 cases (10%).

Slide tracheoplasty was performed according to the principle described by Tsang et al. [3] and by Beierlein and Elliot [11] and was done in cases of lesions involving more than 50% of the tracheal length. The trachea was carefully mobilized from the cricoid cartilage to the carina including the main bronchi, taking care to avoid the nearest vascular structures as much as possible. Under the bronchoscopic view, the mid-portion of the stenotic segment was transected, and the upper and lower parts of the trachea were incised posteriorly and anteriorly, respectively. In the presence of the right upper lobe bronchus or in the presence of bronchial stenosis, a lateral incision was made in each of the 2 tracheal segments [11]. The corners of both sides of the transected trachea were trimmed, and the 2 opened ends were then slid onto each other and anastomosed by multiple interrupted 5/0 or 6/0 polydioxanone sutures (PDS, Ethicon, Johnson & Johnson, Somerville, NJ, USA). On completion of the anastomosis, the patient was ventilated with the trachea submerged in water to rule out air leaks.

The tracheal resection and end-to-end anastomosis were performed according to the principle described by Grillo et al. [12] and were done in cases of lesions involving less than 50% of the tracheal length. In this group, 2 patients presented postintubation tracheal stenosis, whereas the other 2 showed tracheal damage
due to the sequelae of chronic vascular compression. In the first of the latter 2 children, vascular malformations comprised a double aortic arch. This patient underwent left aortic arch division, tracheal resection and reanastomosis. The same approach was followed for the other patient who had evidence of tracheal stenosis after surgical correction and of an anomalous right subclavian artery diagnosed in a different hospital.

Complex reconstruction included combining a standard slide tracheoplasty for the lower two-thirds of the stenosis with an enlargement of the upper stenotic third by means of a costal cartilage patch (Fig. 2). This approach was followed in 3 children because a careful complete mobilization of the trachea was not possible. In these cases, unfortunately, we did not identify any anatomical condition or adherences from previous operations that could hamper tracheal mobilization. We felt a great resistance in the tracheal pulling considering that this situation was potentially at risk of leakage. At the end of each procedure, the endotracheal tube was repositioned to act as a stent and left in place for at least 5 days.

**Statistics**

Data were presented as mean, median and range. The statistical analyses were performed using the software STATA 11 (Stata Corp. LLC, College Station, TX, USA). The Kaplan–Meier curves were used to describe the overall survival rate and the freedom from reinterventions for each of the 3 age groups (neonates, children who underwent tracheal surgery between 30 days and 1 year of age and children who underwent tracheal surgery after 1 year of age) and for the surgical technique used (Figs. 3 and 4; Supplementary Material, Tables S4 and S5). A log-rank test was used to show statistically significant differences between the groups with a P-value set at <0.05.

**RESULTS**

No early deaths or reoperations occurred in our sample population. Overall mortality was 4 of 30 cases (13.3%). Details on the characteristics of the patients who died are provided in Table 2. No significant difference in terms of survival or freedom from reinterventions was observed among the 3 age groups studied. However, a statistically significant difference was observed in terms of survival rate when the results were compared by the surgical technique adopted (Fig. 3B). In this case, in fact, the children who were exposed to complex reconstruction showed a significantly higher mortality rate than the patients undergoing the other 2 surgical procedures (P = 0.000).

The median ventilation time was 11.5 days (5–53 days), and the median hospital length of stay was 35 days (21–124 days).
Extubation was performed according to the standard weaning methods and usually following a fibre-optic evaluation. We left an intratracheal tube as a 'temporary' stent to allow healing of the tracheal surgical wound for further endoscopic treatments, if required.

Postoperative seriate bronchoscopy was performed according to clinical status. Endoscopic reinterventions were required for 19 children (63%). Minor endoscopic reinterventions consisted of tracheal balloon dilatation (minimum 1 dilatation; maximum 12 dilatations), which was performed in 5 children (16.7%), and granulation removal, which was conducted in 4 patients (13.3%). Major endoscopic reinterventions were performed in 15 children (50%). More specifically, 13 children (43.3%) required postoperative positioning of an intratracheal stent. In this subgroup, excluding the 4 children who died, the stent was successfully removed from 7 children (7 of 9, 77.8%). The type of stent was selected according to endoscopic indication. The silicone (Poliflex, Rusch\textsuperscript{10}), Dumon (Novatech\textsuperscript{10}) or covered nitinol stents (Silmet\textsuperscript{10}) were the preferred choice in cases of tracheomalacia, whereas stainless steel metallic stents were preferred in cases of bronchomalacia because the mesh of the metallic stents allows the air to move through its wires inside the bronchial orifice. Postoperative tracheostomy was performed in 2 patients (6.7%) and is still in place. These 2 patients were operated on during the neonatal period. They both presented a pulmonary sling associated with tracheal stenosis and had a mean weight at operation of 2.9 kg. They underwent slide tracheoplasty and left pulmonary artery repositioning. In contrast, the children requiring a preoperative tracheostomy were all successfully decannulated.

The median follow-up period was 38.5 months (minimum 2 months to maximum 12 years) and was complete for all the patients of our sample. Of the survivors (26 of 30, 86.7%), 11 children (42.3%) showed an adequate tracheal diameter related to age and were free from symptoms. For this reason, they did not require further bronchoscopic check-ups after a median follow-up period of 3.5 years or until they developed new symptoms.

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**Table 2:** Characteristics of the patients who died

<table>
<thead>
<tr>
<th>Patients</th>
<th>Age at surgery (years)</th>
<th>Diagnosis</th>
<th>Associated malformations</th>
<th>Type of surgery</th>
<th>Complications</th>
<th>Endoscopic procedures</th>
<th>Time of death (months)</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>6.2</td>
<td>CTS</td>
<td>PS</td>
<td>Complex</td>
<td>Anastomatic dehiscence</td>
<td>Stent positioning</td>
<td>8</td>
<td>a</td>
</tr>
<tr>
<td>2</td>
<td>4</td>
<td>CTS</td>
<td>PS-Di George syndrome</td>
<td>Complex</td>
<td>Residual tracheomalacia</td>
<td>Stent positioning</td>
<td>3</td>
<td>b</td>
</tr>
<tr>
<td>3</td>
<td>2.3</td>
<td>CTS</td>
<td>AVSD-down syndrome</td>
<td>Slide</td>
<td>Residual tracheomalacia</td>
<td>Stent positioning</td>
<td>10</td>
<td>c</td>
</tr>
<tr>
<td>4</td>
<td>0.2</td>
<td>CTS</td>
<td>No</td>
<td>Slide</td>
<td>No</td>
<td>Stent positioning</td>
<td>6</td>
<td>c</td>
</tr>
</tbody>
</table>

\(^{a}\)Haemoptysis from arteriovenous pulmonary malformations.
\(^{b}\)Unknown, autopsy did not revealed tracheal alterations.
\(^{c}\)Unknown, the children came from foreign countries where a monitoring programme for tracheal disease does not exist, and an autopsy was not performed.

AVSD: atrioventricular septal defect; CTS: congenital tracheal stenosis; PS: pulmonary sling.

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Figure 4: The Kaplan–Meier curves for freedom from reinterventions by age groups (A) and surgical techniques adopted (B).
Two patients (6.7%) still showed mild tracheal stenosis that did not affect the air flow.

**DISCUSSION**

Paediatric tracheal surgery may be required for several types of lesions. The most frequent congenital tracheal malformation is CTS, which can cause life-threatening respiratory insufficiency in neonates and infants and is associated with complete cartilaginous tracheal rings [1, 2]. Acquired lesions are frequently represented by complications of chronic compression on the tracheal wall, by vascular rings/anomalies or by prolonged intubations that produce a softness of the tracheal wall, which is described as tracheomalacia [1, 7, 13].

Tracheal surgery has evolved considerably over the last 20 years. Today, there is general consensus that 2 prerogatives are necessary for a tracheal surgery programme to be established: the use of slide tracheoplasty and its application to the wide spectrum of tracheal diseases and the management of patients by a dedicated multidisciplinary tracheal team [7, 11]. The need for a multidisciplinary approach is explained by the frequent association of cardiovascular, abdominal and bronchopulmonary malformations in children undergoing tracheal surgery (other than the presence of genetic syndromes) [1, 2, 6, 7].

However, despite the present technical and management improvements in the field, the mortality rate following tracheal surgery is still 10–18%; the incidence of further complications is as high as 40% [6, 7].

No hospital deaths were observed in our dataset. Overall, our results may be considered very satisfying, considering the heterogeneity of our sample population, when compared with previously published reports with comparable numbers of subjects, the age distribution, the types of lesions treated and the surgical techniques adopted [9, 14]. Our experience also confirms the fact that tracheal surgery is especially indicated during the neonatal period or early infancy [2, 6]. In our sample population, 22 patients (73.3%) underwent surgery before 1 year of age. Overall mortality, which in our experience was long-term mortality, was in line with previously published reports conducted on large patient series [1, 6, 7]. A high mortality rate was observed in our analysis in patients who underwent complex reconstruction (2 of the 3 who underwent complex reconstruction died, 75% mortality rate for those having the technique). Considering the small sample population and the fact that this technique has not been described by other research groups, we cannot compare our results with those of others nor are we in a position to suggest that other authors should adopt this technique instead of slide tracheoplasty or patch enlargement. We have reviewed the ‘failure’ of this technique with the help of a 3D printing reconstruction of the trachea of one of our patients. We have ‘suggested’ that, in these cases, the patients face the cumulative risk of 2 surgical techniques, with dehiscence as the most dangerous one, but unfortunately we do not have objective data to back up this statement. Furthermore, the children who died after undergoing complex reconstruction were aged 4–6 years, respectively, which is in contrast with data in the current literature that indicates that increased risk of death seems to occur during the first year of life [7]. Slide tracheoplasty, on the contrary, was the surgical technique adopted for two-thirds of our patients (76.7%); it was used for different types of lesions, both CTS and acquired lesions. We agree with other authors that slide tracheoplasty offers the advantage of enlarging the tracheal diameter by using autologous tracheal tissue [1–4], giving the reconstructed trachea the possibility to grow [12, 15]. However, although slide tracheoplasty can be extended to several anatomical patterns, including restenosis following primary reconstruction with different techniques [16], we are still not convinced that ‘one slide fits all’ [7]. In our opinion, the complete absence of tension on the anastomotic site, which in the case of slide tracheoplasty is long, is an indispensable condition for slide tracheoplasty to be successful. For this reason, careful harvesting of the trachea should be mandatory for its mobilization.

As results from the literature indicate, complications following tracheal surgery are frequent and require further tracheobronchial interventions [5–7]. Anastomotic leakage is one of the most dramatic problems. Butler et al. [6] and Manning et al. [7] reported an incidence of further tracheal reinterventions in 48% and 28% of patients, respectively. In our work, we considered all the types and numbers of additional interventions, including patients who were exposed to a single balloon dilatation.

As pointed out by previously cited authors, the use of cardio-pulmonary bypass for tracheal reconstruction is helpful, safe and necessary for long lesions [6, 7]. Cardiopulmonary bypass is reputed to have a high level of safety and is not considered a risk factor by many authors [7]. Neither neonatal/infant age at surgery nor associated cardiac malformations worsened our outcomes, contrary to previously reported results [17–19]. With the exception of 1 child, whose tracheal stenosis was discovered following neonatal palliation for univentricular physiology, associated cardiac malformations were treated simultaneously with tracheal reconstruction. Once again, neither patients with bronchial arborization abnormalities, such as right upper lobe bronchus (broncus suis), nor patients with a single lung showed a negative outcome in our experience. Our results are in agreement with those recently described by Spaggiari et al. [20] in the case of a single lung, showing that this pre-existing malformation cannot be considered a limitation for surgery.

**Limitations**

We are fully aware that the retrospective nature of our research, together with the heterogeneity of our sample population, makes the results difficult to interpret and produces an analysis that is still preliminary. This can be considered one of the limitations of our work.

**CONCLUSION**

In conclusion, paediatric tracheal surgery continues to be a challenging field in fighting congenital and acquired tracheal malformations that can cause life-threatening respiratory insufficiency in neonates and infants. The management of children requiring cardiac surgery should include the following:

i. A preoperative assessment of the anatomy of the trachea and of the associated malformations. Bronchoscopy remains the gold standard for preoperative (also intraoperative and postoperative) assessment as well as chest computed tomography [1, 2, 5–7, 21]. The value of bronchoscopy is demonstrated not only in the preoperative stage but also in the management of post-surgical complications to the point that
iv. A well-established policy of follow-up is needed with particular attention to the education of parents regarding the characteristics of this complex disease [4, 23].

These 4 points have been shown to be indispensable in improving the results and reducing costs. In an era like the one we are living in, resource optimization is crucial for the development of surgical programmes.

SUPPLEMENTARY MATERIAL

Supplementary material is available at ICVTS online.

Conflict of interest: none declared.

REFERENCES