Combined Surgical and Endoscopic Approach for Ring–Sling Complex

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Abstract

Background Left pulmonary artery (LPA) sling (PAS) is a vascular ring, which is frequently associated with long-segment tracheal stenosis (TS). Mortality rate in operated children is still high, especially in cases of severe tracheal hypoplasia and/or associated congenital heart defects (CHDs). We report our experience of treatment and follow-up in a pediatric cohort of patients affected by PAS with severe tracheobronchial involvement.

Methods From 2005 to 2017, we enrolled 11 children diagnosed with PAS and congenital TS requiring surgical intervention. Echocardiography, computed tomography, and bronchoscopy were performed in all patients. Associated CHD were present in 5 (45%) patients. Tracheal reconstruction techniques included slide tracheoplasty (7/11; 63%), slide tracheoplasty and costal cartilage graft (2/11; 18%), and Hazekamp technique (2/11; 18%).

Nine patients underwent LPA direct reimplantation and concomitant tracheoplasty; concomitant surgical repair for CHD was performed in three children.

Results Over a mean follow-up of 30 months (range: 3–75 months), a late mortality of 18% was registered; no early death occurred. Good flow through LPA could be documented in all patients. Ten children required operative bronchoscopies (mean: 16/patients) aimed at stent positioning/removal, treatment of granulomas, and tracheobronchial dilatation.

Conclusions Severe tracheobronchial stenosis and associated CHD were the main determinants for hospitalization time, intensive assistance, and repeated endoscopic procedures.

Patients affected by PAS/TS complex require a careful management at high-specialized centers providing multidisciplinary team.

Respiratory endoscopy may play a central role both in preoperatory assessment and in postoperative management of patients showing severe tracheobronchial involvement.

Keywords
- congenital heart disease
- CHD
- pulmonary arteries
- trachea

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Introduction

Pulmonary artery sling (PAS) is a rare congenital disease defined by anomalous origin of the left pulmonary artery (LPA) distally from the right pulmonary artery. Throughout its pathway, abnormal LPA passes between trachea and esophagus, configuring a vascular ring around distal trachea and right main stem bronchus. Beside external tracheobronchial compression, respiratory symptoms may be worsened by intrinsic tracheal anomalies. More than 75% of affected patients suffer from associated congenital tracheal stenosis (TS), as consequence of complete cartilage rings narrowing inner tracheal diameter.[1] Affected children may be occasionally asymptomatic; otherwise, they develop during the first months of life respiratory symptoms as stridor, wheezing, or respiratory distress requiring mechanical ventilation. Parenchymal air trapping, atelectasis, and/or pneumonia may occur, depending on the degree of tracheobronchial stenosis. In up to 85% of cases, association with other congenital cardiac defects and extracardiac abnormalities has been reported; among these, ventricular septal defect (VSD), atrial septal defect (ASD), patent ductus arteriosus (PDA), persistent left superior vena cava, abnormal branching of the right pulmonary artery, coarctation of the aorta, and abnormalities of aortic arch branching seem to be the most common.[1–4] The expected prognosis in nonoperated children is unfavorable, leading to death for respiratory insufficiency due to airway obstruction within infancy or childhood. Surgical intervention is the gold standard treatment; since slide tracheoplasty has been introduced, a significant reduction in precocious mortality rates and perioperative complication has been registered. However, data concerning long-term follow-up, late mortality rates, and the role of endoscopy for recurrent stenosis and secondary complications are still limited. We report our experience of surgical and endoscopic treatment of PAS associated with congenital TS.

Materials and Methods

The Ethics Committee of our hospital approved this study in January 2017 as a retrospective chart analysis. The Committee waived the need for patient consent.

We retrospectively analyzed clinical, instrumental, and surgical records of all patients consecutively admitted for PAS and associated TS. From 2005 to 2017, 32 patients underwent tracheal reconstruction for congenital or acquired TS; among these, 11 patients (6/5 = males/females) presented with associated PAS and were therefore enrolled in our study population. Diagnosis of PAS was suspected at echocardiography (►**Fig. 1**); this technique is currently considered the gold standard for identifying LPA origin and to rule out associated intracardiac defects. Preoperative flexible (Karl Storz, Tuttlingen, Germany) and rigid (Karl Storz, Germany) bronchoscopy and chest computed tomographic (CT) angiography have been performed in all patients, aimed at assessing the severity and length of TS and to better define patient's anatomy before, during, and after surgical intervention. Anatomic characterization was implemented with three-dimensional (3D) reconstruction in selected cases. During surgical correction,

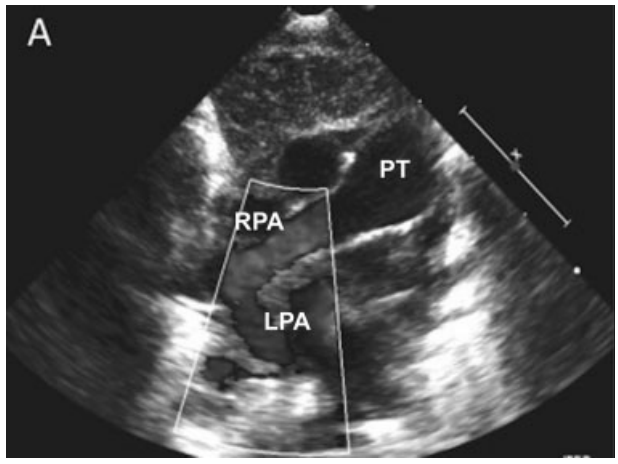

Fig. 1 Echocardiograms from Patient 9 : abnormal origin of left pulmonary artery distally from right pulmonary artery can be documented. Abbreviations: LPA, left pulmonary artery; PT, pulmonary trunk; RPA, right pulmonary artery.

bronchoscopy allowed a better definition of the upper limit of the stenotic tract, while, at the end of the intervention, it endorsed a real-time assessment of the surgical outcome in terms of airway patency obtained. In the postoperative time, the first bronchoscopic evaluation was performed at the time of extubation. Stenting was executed in symptomatic patients with moderate or severe fixed stenosis, according to the classification reported by Burden et al.[5] Silicon or covered nitinol stents were placed in the operating theater using a rigid bronchoscope under general anesthesia (sevoflurane inhalation ± intravenous propofol) and radiological guidance. A closed clinical and bronchoscopic follow-up (every 2, 4, or 6 months) was scheduled, depending on child's clinical conditions and granulation tissue development. To minimize the risk of granulation, all patients received inhaled corticosteroids; in case of airway obstruction, laser therapy was required to limit excessive tissue overgrowth. In case of residual scar stenosis, laser incision and tracheobronchial balloon dilatation were the first-line treatments, to widen the airway: diode laser (Dornier MedTech) was used to make radial cuts and, if necessary, a balloon dilatation was performed, with a dilating catheter passed into the narrowed segment of the tracheobronchial tree. Repeated balloon dilatations were usually required to achieve a sufficiently wide airway lumen. Stenting was used in case of relapsing stenosis needing a measure to better stabilize the airway. Endoscopic procedures for tracheal dilatation are showed in ►**Video 1**. Before stent placement, a dilatation was often performed; only in case of severe and relapsing TS treated with hour glass shaped stents, the stenotic segment was not dilated and the narrowed part of the device was placed in the stenotic segment. When airway patency improved, in presence of stable clinical conditions, the stent was removed.

Nonabsorbable silicone stent (Dumon) or covered nitinol stent was chosen in most of the cases; the employment of bare metal coronary stent was limited, given the high risk of mucosal ischemia and necrosis. Absorbable stents were

considered unsuitable for small children affected by severe tracheal and/or bronchial stenosis and were therefore avoided.

Surgical Techniques

All patients underwent LPA direct reimplantation on main pulmonary artery; concomitant tracheoplasty was performed in 9/11 (81%) patients; in the remaining two cases, it was postponed since the inner tracheal diameter was considered as not severely compromised. Tracheal reconstruction techniques included slide tracheoplasty (7/11; 63%), slide tracheoplasty, and costal cartilage graft (2/11; 18%), and tracheal reconstruction according to the Hazekamp technique (2/11; 18%). Concomitant surgical repair for associated congenital heart disease (CHD) was performed in four children, comprising VSD repair with autologous pericardial patch, VDS repair associated with aortic arch reconstruction, ASD direct suture, and PDA closure (►**Table 1**). The patient presenting with double outlet right ventricle (DORV) underwent tracheoplasty and LPA reimplantation at first (age: 4 months, weight: 2000 g); pulmonary binding was postponed until the age of 7 months for persistent failure to thrive, and definitive correction performed at the age of 10 months.

Surgical Intervention Description

PAS and TS were approached by complete median sternotomy. The thymus was resected, and a patch of autologous pericardium or a patch was harvested and soaked in glutaraldehyde if necessary. In the case of necessity of cartilage patch, a 3-cm incision was made at the level of the sixth costal rib preparing a small cartilage patch. Cardiopulmonary bypass and moderate hypothermia were used. Venous cannulation was accomplished by single or double venous cannula depending on the presence of intracardiac associated malformations. If necessary, when the estimated cross-clamp time was considered to be more than 60 min, the Bretschneider histidine–tryptophan–ketoglutarate solution was used. Isothermic blood cardioplegia was adopted when a shorter aortic cross-clamp time could be presumed.

The LPA was identified and mobilized until the hilar branches avoiding phrenic nerve injury. Using a C-shaped occluding clamp, the LPA was divided from the right pulmonary artery and the stump was closed with continuous 6/0 polypropylene sutures. The LPA was disconnected from the right pulmonary artery and sewed to the main pulmonary trunk using a partial occluding clamp with continuous 6/0 polypropylene sutures. The slide tracheoplasty was made according to the principle described by Tsang et al[6] and by Beierlein and Elliot.[7] Trachea was fully mobilized taking care of the neurovascular structures from the cricoid cartilage to the carina, enclosing the main bronchi. The mid portion of the stenotic segment was divided and the upper and lower parts of the trachea were incised posteriorly and anteriorly, respectively. If right upper lobe bronchus or main bronchial stenosis was present, a lateral slide incision was made. The corners of both sides of the transacted trachea were trimmed and after the sliding the anastomosis was made by interrupted 5/0 or 6/0 polydioxanone sutures. Subsequently, the patient was ventilated with trachea submerged in water to rule out air leaks. Two patients (18%) underwent tracheal reconstruction according to the technique described by Hazekamp et al.[8] In these patients, the trachea was opened in the midline. An autologous pericardial patch was used to enlarge the anterior wall and hence to increase tracheal diameter by interrupted 6/0 PDS sutures (Ethicon Endo-Surgery, Inc. Johnson & Johnson, New Jersey, United States). Subsequently, a series of costal cartilage strips previously harvested from the sixth costal rib were attached to the pericardial patch by separate stiches positioned one in the midline and laterally to confer more stabilization to the trachea–patch strips complex.

Two children, respectively, 6 and 5 years old underwent tracheal reconstruction by a combination of conventional slide tracheoplasty for the lower two third of the stenosis and enlargement of the upper stenotic third with a costal cartilage patch. This approach was required in three children where a complete mobilization of the trachea (condition necessary for a successful slide tracheoplasty) was not obtained despite the careful splitting from the surrounding structures. In this case, avoiding anastomotic tension was preferred instead to extend slide's incisions. At the end of the procedure, the endotracheal tube was repositioned acting as a stent and left in place for at least 5 days.

Results

Baseline Clinical Feature

The median age at intervention was 7 months (range: 1–78; mean: 25.4); median weight of patients 4 kg (range: 2.7–24; mean: 8.5).

At the baseline evaluation, 10 patients (90%) showed clinical respiratory symptoms: five patients developed respiratory failure, four patients presented with stridor and recurrent respiratory infections, whereas in one patient episode of apnea was reported. Intubation and mechanical ventilation due to severe respiratory failure and carbon dioxide retention were required in three cases; one child required preoperative tracheotomy due to intubation difficulties. The last patient did not present with typical respiratory symptoms, but only with moderate tachypnea, initially attributed to a large PDA.

Associated CHD were present in 5/11 (45%) cases; one patient diagnosed with VSD and aortic arch hypoplasia, a case with isolated VSD, one patient presented with ASD, and one with large PDA. A case of DORV associated with PDA was also documented (►**Table 1**).

Concerning endoscopic and radiologic findings, the mean length of TS was 45 mm (range: 20–90 mm) and the mean internal diameter was 2.0 mm (range: 1.2–3.5). Carina involvement was confirmed in 8/11 patients (72.7%). All the patients show complete cartilage trachea rings (O-rings) at the bronchoscopy. The mean LPA diameter was 3.8 mm, and the mean right diameter was 6.1 mm.

Impact on Outcome

The mean follow-up was 30 months (range: 1–72 months). The mean postoperative intubation time was 26.1 days (range: 5–70 days). We recorded overall mortality of 18% (two patients); both children presented whit long-segment stenosis

Table 1 Presenting symptoms, clinical features at intervention, and degree of tracheal involvement in enrolled patients

Patient no.	Age at intervention (months)	Weight (Kg)	Symptoms	Associated diagnosis	Length of stenosis (mm)	Cardiac procedures	Tracheal procedure	Postoperative intubation (days)	Tracheostomy or stenting	Outcome
1	5	7.8	Respiratory failure	–	40	LPA reimplant	Hazekamp technique	22	Tracheal stent	Moderate tracheal stenosis
2	78	24	RRI, stridor	–	30	LPA reimplant	Slide	5	–	Spontaneous breathing
3	2	4	Respiratory distress	–	50	LPA reimplant	Slide	29	Tracheal stent	Moderate tracheal stenosis
4	7	4	RRI, wheezing	VSD	20	LPA reimplant, VSD surgical repair	Slide	6	–	Spontaneous breathing
5	75	17	RRI, wheezing	–	90	LPA reimplant	Slide + costal graft	10	Tracheal stent	Death
6	48	22	RRI, stridor	–	80	LPA reimplant	Slide + costal graft	10	Tracheal stent	Death
7	1	2.8	RRI, respiratory distress	Aortic coarctation, VSD	30	LPA reimplant, VSD surgical repair, aortic arch reconstruction	Slide	53	Tracheostomy, tracheal stent	Spontaneous breathing
8	23	2.8	None–	Patent ductus arteriosus	n/a	LPA reimplant, PDA closure	Slide	n/a	Tracheostomy, tracheal stent	Mechanical ventilation
9	11	6.5	Respiratory distress	ASD	n/a	LPA reimplant, ASD surgical repair	Slide	n/a	Tracheal stent	Spontaneous breathing
10	4	2.7	Apnea	DORV	25	LPA reimplant	Slide, tracheopexy	70	Bronchial stent	Mechanical ventilation
11	1	3	Respiratory distress	–	n/a	LPA reimplant	Hazekamp technique	30	Tracheostomy, tracheal stent	Mechanical ventilation

Abbreviations: ASD, atrial septal defect; DORV, double outlet right ventricle; LPA, left pulmonary artery; RRI, recurrent respiratory infections; VSD, ventricular septal defect.
Note: Data concerning surgical approach, further tracheobronchial procedures, and final outcome have also been reported.

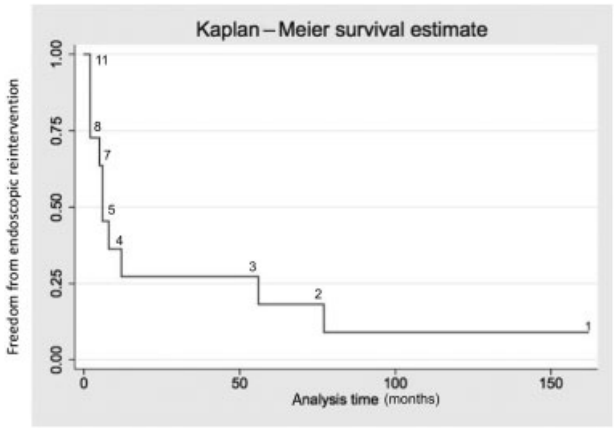

Fig. 2 Kaplan–Meier analysis estimating survival from endoscopic procedures after surgical treatment; the risk to require operative bronchoscopy is elevated, especially within the first 2 months from intervention.

and underwent combined slide tracheoplasty and cartilage costal graft. One patient developed acute hemoptysis 6 months after the procedure, whereas the second child died after 3 months from the intervention as consequence of foreign body inhalation. No early death occurred. A first attempt of hybrid procedure involving laser division of complete tracheal rings and tracheal endoscopic balloon dilatation was performed in two children; however, given the unsatisfactory result they were both redirected to surgery. Notably, the peculiar presence of complete rings and the length of stenosis imply a high risk of complications (esophageal damage, tracheal leak, and perforation, bleeding) without allowing a sufficient enlargement of inner tracheal diameters.

Ten patients (90%) required operative bronchoscopy treatments after surgical correction, with a mean of 16 endoscopic procedures for each patient (standard deviation score [SDS] ± 11.24; range: 1–43) (►**Figs. 2** and **3**). More in detail, affected patients required a mean of seven procedures (SDS ± 5.6; range: 1–21) for stent positioning/removal and a mean of 5.8 procedures (SDS ± 5.0; range: 1–14) for the treatment of granulomas. Nine out of eleven patients required multiple endoscopic balloon calibration of trachea, main left, and/or main right bronchus (mean: 11.4, SDS ± 10; range: 4–28) due to persistent or relapsing stenosis.

Video 1

Severe tracheal hypoplasia with complete cartilage rings approached with endoscopic treatment. The first phase of therapy included laser separation of complete rings and subsequent balloon dilatation of stenotic tract. Stent positioning under radiologic control followed, completed with stent balloon dilatation. Good increase in tracheal inner diameter was documented at the end of the procedure. Online content including video sequences viewable at: www.thieme-connect.com/products/ejournals/html/doi/10.1055/s-0039-1678670.

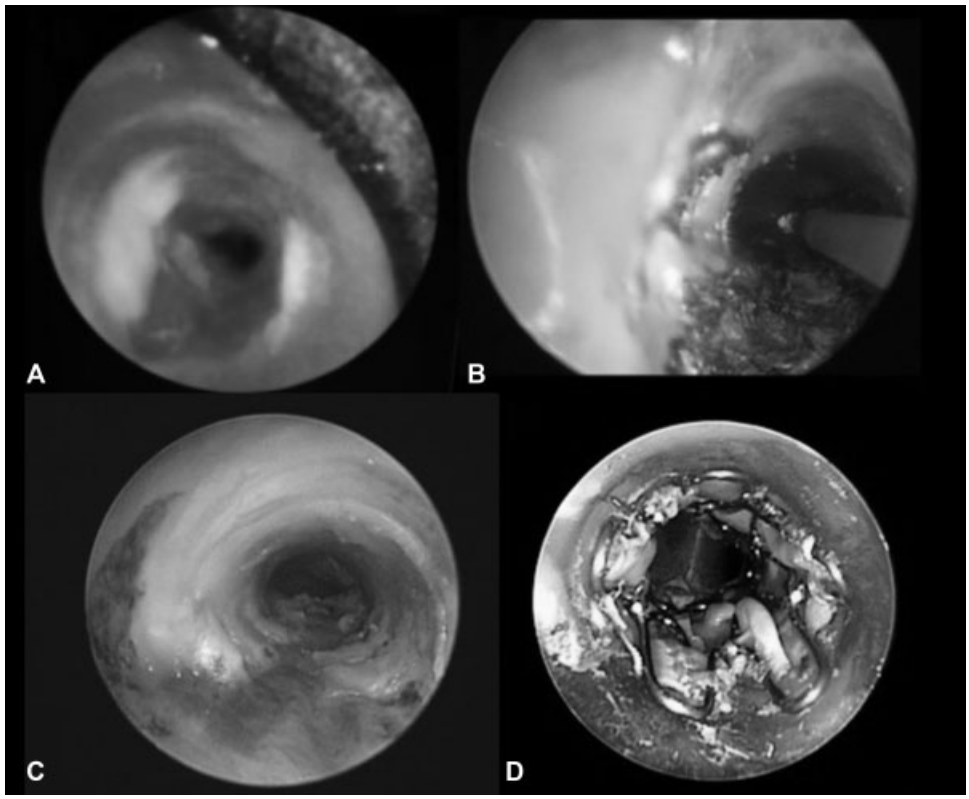

Fig. 3 Severe congenital tracheal stenosis documented at baseline bronchoscopy (**A**). Combined endoscopic treatment including laser therapy (**B**), balloon dilatation (**C**), and stent positioning (**D**) led to a satisfactory increase in tracheal inner diameter.

At last available follow-up, good flow through the LPA could be documented at echocardiography in all patients. The mean final inner tracheal diameter was 7.4 mm (range: 5–12 mm); in 8/11 (72.5%) patients, we could achieve an inner tracheal diameter adequate for age.

Two patients (18%) did not require tracheal stent or tracheostomy and were therefore discharged on spontaneous breathing. Three patients (27.2%) required tracheal/bronchial stent positioning due to residual stenosis. Four patients (36%) necessitated both tracheostomy and tracheal stent; two of them were still hospitalized in critical care unit at the end of the follow-up, requiring mechanical ventilation. Notably, they both suffered from associated CHD (DORV, large PDA). The other two children were discharged with domiciliary overnight ventilation assistance.

Overall, less favorable course was documented in children showing carina involvement; 75% of them (6/8 patients) necessitated tracheostomy, ventilator assistance, or died precociously.

Discussion

We report our experience concerning surgical, endoscopic, and cardiologic management of 11 patients diagnosed with PAS and associated congenital severe TS.

PAS management with LPA reimplantation is currently considered a safe and effective procedure, whether performed alone or concomitantly to tracheal surgery.[1,4,9] According to this, at last available follow-up, we could document adequate LPA patency in all children, regardless the timing of intervention and the characteristic of affected patients (age and weight at intervention, associated CHD, and degree of tracheobronchial involvement).

The degree of associated airways involvement may be variable, from mild asymptomatic tracheal narrowing to severe tracheobronchial stenosis leading to acute respiratory failure. The entity of tracheobronchial involvement has already been reported as one of the main determinants of outcomes, together with the presence of associated CHD.[9,10]

Over the last decades, progress in tracheal surgery has allowed limiting surgical mortality rate, whereas postoperative complications remain common.[6–8,10,11] In the large cohort of 127 patients affected by congenital TS published by Hawett et al, the elevated survival rate was reported; however, further requirement of postoperative endoscopic procedure was assessed in more than 40% of patents, with airway malacia persisting in 25% of cases.[12] Arcieri et al documented the need of reintervention in more than 60% of patients surgically treated for TS; the most common complications included restenosis requiring stent positioning and balloon dilatation, and granulomas removal.[13] Nowadays, LPA reimplantation and slide tracheoplasty are considered procedures of choice for patients affected by PAS/TS in presence of favorable anatomy.[14] Slide tracheoplasty has been proved to be successful also in case of severe tracheal involvement with long-segment TS.[15] We performed different tracheal reconstruction techniques depending on patient's anatomy (slide tracheoplasty, slide tracheoplasty and costal cartilage graft, and the Hazekamp technique). Concomitant LPA reimplantation was performed in 9/11 cases and the concomitant repair for associated CHD was performed in four children. We reported a mortality rate of 18% (two patients); no early death occurred. Both patients underwent slide tracheoplasty associated with cartilage costal graft, due to long TS. Our data concerning that mortality rates are consistent with previous findings; Backer et al reported a case series of 34 patients, who underwent PAS repair using median sternotomy, cardiopulmonary bypass, and LPA reimplantation;[1] they described an overall mortality of 11.7%, that is, four late decease. With regard to TS, better results were achieved after slide tracheoplasty compared with different surgical techniques. Pericardial patch tracheoplasty and tracheal autograph were associated with higher mortality rate and surgical revision requirement. The authors suggested performing tracheal resection for short segment of TS (<6 tracheal rings) and slide tracheoplasty for long-segment TS. Similar results have been described in further study by Oshima et al[16] and Yong et al;[17] left pulmonary reimplantation with concomitant repair of TS and minor intracardiac anomalies led to positive results in term of operative mortality, LPA patency, and postoperative complication.

Beside mortality rate, the final outcome should also consider the requirement of intensive cares, postoperative procedures, tracheostomy, and/or stent positioning.

According to Elliot et al,[18] despite a significant reduction in early mortality has been achieved through slide tracheoplasty, late-mortality rates are still considerable and management of recurrent stenosis remains challenging. Patients requiring tracheal stent positioning are keener to necessitate to multiple interventions and recurrent hospitalizations. In our cohort, a mean of 16 postoperative endoscopic procedures were required for each patient, primary aimed at treating residual stenosis, removing granulomatous tissue and allow a growth of tracheobronchial tree adequate to patient's age and weight. Four patients necessitated tracheostomy and prolonged intensive unit hospitalization since they were dependent from ventilatory support.

The apparently less favorable results in term of postoperative procedures, hospitalization time, and ventilator assistance requirement reported in our work are likely ascribable to different conditions. First, published experience from high qualified and skilled centers[14,19,20] report a risk for reintervention ranging from 26% to 44%, with mortality rates of 5 to 13%. However, these papers address to congenital TS of any etiology, whereas few large studies specifically focus on the subgroup of patients affected by congenital TS and associated PAS.[4,9,16] In these cases, overall mortality is still higher (up to 21%) and postoperative complications, including residual stenosis, granulomas, and requirement of tracheostomy, are not infrequent (up to 57%).

Furthermore, our sample was subject to self-selection bias: the presence in our hospital of a high specialized department for respiratory endoscopy led to collect cases presenting with moderate to severe airway involvements, that is, most of the children have been referred from other

Italian hospitals due to complications with patient intubation/extubation or ventilator support. Given the presence of severe clinical symptoms, many children required surgical treatment at a very low weight and young age (median age: 7 months). Despite there is no consensus regarding whatever could be the optimal age for surgical intervention, some authors[4] reported higher mortality and morbidity rates among children younger than 1 year. We did not register an increased mortality in this subgroup of patients; however, the requirement of postoperative endoscopic procedures was undoubtedly considerable.

Recent evidences suggest the possible role of tracheal stenting in case other therapeutic options have failed and further surgical approach is unfeasible.[21]

We opted for nonabsorbable stent since they are less deformable and supply the radial force required to guarantee an adequate and stable patency of tracheal lumen.[22] This seems particularly true in presence of associated tracheomalacia, postoperative stenosis, or extremely narrow inner tracheal diameter. Moreover, fragments derived from absorbable stent degradation may accumulate in the patient's airways creating obstructive problems and facilitating inflammatory processes. Finally, the mean degradation time is of nearly 3 months, which is in our opinion insufficient to allow the advisable trachea-bronchial growth.

According to the literature, severest course was observed in younger children presenting with low body weight, long TS and associated complex cardiac heart disease.

Given the high risk of postoperative complications, several papers suggest a conservative approach to PAS/TC complex.[9,23,24] The rationale of conservative management relies on the possibility of spontaneous growth in tracheal inner diameter, with progressive amelioration in clinical symptoms.

Cheng et al suggested only clinical and radiological follow-up as a safe alternative in patients presenting with short-segment TS and the tracheal inner diameter at least 60% of normal for age.[23] Beside anatomic features evaluated at CT and bronchoscopy, functional tests may provide complementary assessment in patients treated conservatively.[25] Brouns et al proposed computational flow dynamic study as possible noninvasive tool in patient's monitoring. This technique is aimed at assessing flow patterns and pressure drops over TS artificially inserted into a 3D CT-based upper airway model; the role of this method in pediatric cohort needs still to be ascertained.[26]

As previously addressed, we managed a selected cohort of patients in which treatment options were limited by the presence of severe respiratory symptoms; therefore, conservative treatment resulted unfeasible. This was one of the main limits to our study, since the unavailability of patients eligible to conservative management prevented us to identify a control group of comparison.

Moreover, statistical significance on our findings was limited by the small observational sample. In addition to this, our cohort was heterogeneous under different aspect (surgical approach, age, and weight at intervention, presence and kind of associated CHD), resulting unsuitable for possible comparison and for the identification of possible prognostic factor.

Given the rarity of the disease, further multicentric studied are advocated to define the most appropriate treatment options and identify possible prognostic factors.

Conclusions

Patients affected by PAS/TS complex require careful management at high-specialized centers providing multidisciplinary team involving pediatric cardiac surgery, pediatric cardiology, respiratory endoscopy, intensive care unit, and neonatology.

Given the heterogeneous degree of airway involvement and the possible association with CHDs, preoperative anatomic characterization is mandatory to plan appropriate surgical approach; in this clinical setting, combined information obtained through bronchoscopy, CT scan, and even 3D reconstruction may help surgeons and endoscopists to choose the best strategy of treatment. The spectrum of this malformation is so wide that the management can be very different case by case. A conservative approach in asymptomatic patients without severe tracheal narrowing (e.g., tracheal diameter < 2 mm) can be discussed.

Respiratory endoscopy may play a central role both in preoperatory assessment and in postoperative management of patients showing severe tracheobronchial involvement.

Conflict of Interest
None.

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