Presentation, diagnosis and management of congenital long-segment tracheal stenosis: a single-centre experience.

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Abstract

Congenital long-segment tracheal stenosis (CLSTS) is a rare airway malformation, caused by absence of the pars membranacea in part of the trachea.

The aim of this study is to provide our single-centre experience with CLSTS and a literature review on clinical presentation, diagnostic workup, treatment methods and outcome. Special attention will be given to the relatively new technique for anatomic visualisation, using anatomic optical coherence tomography (aOCT).

Methods: Single-centre retrospective cohort study of children diagnosed with CLSTS and treated at the University Hospital of Leuven between January 2010 and July 2021. A literature review was performed using PubMed.

Results: Six children were included, age at diagnosis between 11 days and 12 years. Three children had associated pulmonary artery sling and one had unilateral agenesis of the lung, both requiring additional surgery. Presenting symptoms were mainly stridor and respiratory insufficiency during lower airway infections. Diagnosis was made by a combination of bronchoscopy, chest CT, bronchography and aOCT. Four patients have been treated surgically and one is awaiting surgery; one was treated conservatively. One patient suffered from important cardiac ischemia postoperatively. Three patients required balloon dilation and one required additional pericardial patch tracheoplasty. Follow-up after diagnosis ranged between two and eight years. One patient still has stridor with exercise, two still have intermittent stridor with respiratory tract infections after surgery.

Interpretation: Outcome in our study population was consistent with data from the literature. CLSTS requires a dedicated and multidisciplinary approach to obtain optimal results. aOCT has proven its added value for diagnosis.

Introduction

Congenital tracheal stenosis is a rare type of airway malformation in children, occurring in one in 64500 live births and is caused by absence of the pars membranacea of one or more tracheal rings resulting in complete cartilaginous rings (1). Depending on the percentage of trachea involved, it is classified as congenital short-segment (< 30-50% of tracheal length) or congenital long-segment tracheal stenosis (CLSTS) (> 30-50% of tracheal length) (1,2,3). The presence and severity of symptoms is dependent on the diameter of the lumen: symptoms are often only present when there is more than 50% narrowing of the lumen, and symptoms at rest when there is more than 75% narrowing (1,4). Stenosis is generally considered severe when the diameter of the lumen is ≤ 3 mm, and in that situation surgical intervention is required in most cases (5). Resection with an end-to-end anastomosis of the trachea is not advisable if more than 30% of the trachea is affected, as the risk for dehiscence becomes too high due to traction on the anastomosis (2,6,7).

For surgical treatment of CLSTS slide tracheoplasty is the preferred technique. After sternotomy, the anterior trachea is denuded to identify the entire length of the stenosis by inspection and placement of markers visualised by combined bronchoscopy. The trachea is then completely mobilised until normal trachea and/or bronchi are seen. In the absence of combined cardiovascular repairs, normothermic cardiopulmonary bypass is started and the trachea is transected halfway through the stenotic part, in combination with a posterior vertical incision in the proximal part of the stenotic trachea and an anterior vertical incision in the distal part. Subsequently, an oblique sliding anastomosis is performed with interrupted mattress polydioxanone (PDS) sutures, resulting in a doubling of the diameter of the lumen and a shortening of the trachea by half the length of the stenotic part (3).

This technique remains a challenging intervention with need for cardiopulmonary bypass, high morbidity and a mortality rate between 5% and 12% in larger studies (1,3,4,8). Therefore, this treatment should be performed by an experienced multidisciplinary team through centralisation of care. For countries with a smaller population, collaboration with an experienced foreign team can be an added value.

Materials and methods

All patients under the age of 18 years diagnosed with and/or treated for CLSTS at the University Hospital of Leuven between January 2010 and July 2021 were included and data were collected retrospectively from their medical files and pseudonymized. The study was approved by the local ethics committee (MP016961).

A literature review was conducted using PubMed with search terms 'congenital tracheal stenosis', 'slide tracheoplasty' and 'optical coherence tomography', reviewing relevant articles written in English from the year 2000 onwards and their most important references.

Results

Literature review

Clinical presentation

The clinical presentation of CLSTS is variable: some children present with severe neonatal respiratory distress, stridor and/or cyanotic spells, others develop symptoms of stridor and respiratory insufficiency at the time of an airway infection during infancy. Some present in adolescence or adulthood with exercise-induced stridor, respiratory distress or cyanosis (1,4).

A high proportion (70-90%) of patients have associated congenital malformations: cardiovascular anomalies in 70% of the cases and other congenital malformations such as respiratory and gastro-intestinal tract, anorectal, renal and skeletal abnormalities in 40% (1,3,4,8). In around 6% of cases, these malformations are part of a VACTERL/VATER association (1). The most frequently associated malformation is a pulmonary artery sling (abnormal position of the left pulmonary artery arising from the right pulmonary artery and going to the left lung between the trachea and oesophagus), present in 50% of patients (1,3,8). Important are also additional respiratory tract malformations such as abnormal bronchial arborisation (30-35%) and unilateral agenesis of the lung (7-9%) (1,3,7).

Diagnostic workup

It is suggested in the literature that diagnostic workup for CLSTS should include contrast computed tomography (CT) of the thorax as well as bronchoscopy with bronchography and anatomic optical coherence tomography (aOCT) (1,3). Which of these investigations is performed first, depends on the presenting symptoms and the availability of the techniques.

However, both CT and bronchoscopy have important limitations. Due to the dynamic nature of the stenosis and the difficulties getting detailed images in young dyspnoeic children, CT may underestimate the degree of luminal narrowing and does not allow a proper evaluation of the tracheobronchial cartilaginous structures (4). Bronchoscopy on the other hand provides limited possibilities for obtaining quantitative measurements and the distal airways can be inaccessible with the bronchoscope when severe proximal stenosis is present. The addition of bronchography to the evaluation will accommodate to some of these limitations (1). It can visualize the anatomy of the respiratory tract distal to the stenosis even if the bronchoscope cannot pass it, which is especially useful to map the anatomy of the main stem bronchi to detect possible involvement in the stenotic disease. While ventilated using a laryngeal mask, a guide wire is placed through the vocal cords by bronchoscopy guidance. Then, a small catheter is placed over the wire and the wire is removed. Contrast is injected through the catheter and fluoroscopy images are made in 2 planes.

aOCT is a more recently implemented light-based imaging modality, using a technique similar to ultrasound, but overcoming several limitations of a soundwave-based method such as its inability to image through air-filled spaces and its suboptimal spatial resolution (9). The minuscule probe (as small as 0.36 mm) can be inserted through the bronchoscope to acquire a series of real-time ultra-high detail (spatial resolution 20 µm) cross-sectional and longitudinal images of the airway and the tracheobronchial wall with the possibility of quantitative measurement of the luminal diameter and characterisation of surrounding tissue, and allowing three-dimensional reconstruction of the images (fly-through) (3). In addition to bronchoscopy and bronchography, aOCT thus provides more information about the degree and characteristics of the stenosis and the airways distal to the stenosis and can confirm the presence of complete cartilaginous rings (see figure 1b) (3,7).

Finally, to screen for associated anomalies, a thorough history and physical examination as well as a cardiac ultrasound are indispensable to detect associated abnormalities (4).

Treatment and outcome

The treatment strategy is highly dependent on the severity of symptoms, individual patient characteristics and associated malformations and should therefore always be discussed multidisciplinary (1,4). Patients with mild symptoms, mostly presenting at a later age, can sometimes be treated conservatively (4).

In the past, several treatment options have been used when surgery is needed. Insertion of a cartilaginous, pericardial or donor homograft and tracheal stenting resulted in major complications including increased risk of granulation tissue formation, restenosis, anastomotic leakage and graft necrosis, and are therefore only recommended as a salvage technique (1,2,4,7).

Slide tracheoplasty is currently the method of choice, as the risk of complications is significantly lower than with other methods (1,4). The technique can be adapted in case of abnormal arborization or bronchial stenotic disease, although some complex cases will require a different technique, and can be combined with cardiovascular surgery (4,7). Tracheal growth after tracheoplasty was found to be satisfactory (4,8,10). Still, it remains a complex and invasive procedure with need for cardiopulmonary bypass and mortality rates between 5 and 12% in larger studies (1,3,4,8).

The optimal age for surgery is thought to be between 10 and 24 months (8). Patients presenting in the neonatal period are often the most difficult to treat, sometimes even requiring extracorporeal membrane oxygenation prior to surgery, and have a significantly higher risk of mortality (4,8). From a surgical point of view, older children and adolescents also pose an important challenge: mobilization of the trachea becomes more difficult, the stenotic part is longer with more risk of traction on the suture, shortening of the trachea caused by the slide tracheoplasty causes more anatomic and functional issues and operation time is longer (8).

Restenosis with need for balloon dilation and/or CO_2 lasering occurs in 33-48% of children after slide tracheoplasty, need for additional stenting or pericardial patch insertion in around 21% (1,3,4). Other frequent postoperative complications are residual tracheomalacia in 20-25% and granulation tissue formation (1,3). One study showed an overall incidence of granulation tissue formation of 13.6% in their study population, with a significant decrease from 31.6% in the first three years to 8.1% in the following three years. It also demonstrated an age-related difference with significantly less granulation tissue formation when surgery was performed between 10 and 24 months of age (5.7%) compared to surgery before 10 months (45.5%) or after 24 months (17.6%) (8).

Less frequent complications include anastomotic dehiscence resulting in mediastinitis or pneumothorax, -mediastinum or -pericardium, laryngeal nerve paralysis, chylothorax, pulmonary hypertension and swallowing difficulties, occurring in 2-4% of patients each (1,3,4,8).

Patient characteristics

Five patients were diagnosed with CLSTS at our institution during the inclusion period and one was referred after diagnosis elsewhere. The patient characteristics, treatment and outcome were summarized in table 1 and 2.

Clinical presentation and diagnosis

The age at diagnosis ranged from 11 days to 12 years and was mostly dependent on the severity of symptoms. One patient presented in the neonatal period with recurrent episodes of severe desaturation, four patients presented during infancy or early childhood with respiratory insufficiency during viral airway infections, and one had similar but less severe symptomatology in childhood, but was only diagnosed during adolescence because of exercise-induced stridor.

Four patients had associated congenital malformations. Three patients had a pulmonary artery sling, one of which had several other cardiovascular anomalies. One patient had an atrial septal defect. Two patients had skeletal anomalies, one had a unilateral agenesis of the lung and one patient was born with an imperforate anus.

Diagnostic workup

Diagnosis was made in all patients by a combination of contrast chest CT and bronchoscopy with bronchography. The added value of bronchography was illustrated in several cases, providing better visualization of the respiratory tract anatomy distal to a particularly narrow stenosis and getting more detailed information about the anatomy of the left main stem bronchus in the patient with unilateral agenesis of the lung. The two most recently diagnosed patients were also examined with aOCT during diagnostic

Table 1: Patient characteristics and treatment method (M= male, F= female)

Patient	Sex	Age at diagnosis (days or months)	Presenting symptoms	Associated congenital malformations	Minimal luminal diameter (mm)	Treatment method	Age at surgical intervention (months)
1	M	34 months	Persistent stridor, respiratory insufficiency with airway infections	Pulmonary artery sling, persistent left vena cava superior Dysmorphism Skeletal anomalies	2.9-3.5	Slide tracheoplasty and pulmonary artery reimplantation	52 months
2	F	11 days	Neonatal severe desaturations Respiratory insufficiency with airway infections	Agenesis left lung Skeletal anomalies Accessory spleen ASD	1.7	Tissue expander Awaiting slide tracheoplasty	Tissue expander at 12 months
3	F	60 days	Respiratory insufficiency with infections	Pulmonary artery sling Anal imperforation	3	Slide tracheoplasty and pulmonary artery reimplantation	18 months
4	F	4 months	Respiratory insufficiency with infections	Pulmonary artery sling	1.3	Slide tracheoplasty and pulmonary artery reimplantation	7 months
5	F	73 months	Almost constant stridor, exercise-induced dyspnoea, frequent lower airway infections, feeding difficulties	/	3	Slide tracheoplasty Pericardial patch tracheoplasty Awaiting correction of a tracheal wall dehiscence	117 months 123 months
6	F	148 months	Respiratory distress with infections Exercise-induced stridor and respiratory distress	/	9	Conservative Avoid intense physical activity	/

Table 2: Postoperative course, need for additional interventions and outcome (ETT = endotracheal tube; ECMO = extracorporeal membrane oxygenation; ASD = atrial septal defect)

Patient	Age at surgery (months)	Treatment method	Days of invasive airway management (ETT)	Days of hospitalization at paediatric ICU postoperatively including day of surgery	Need for additional interventions after surgical repair	Morbidity and outcome after surgery
1	52	Slide tracheoplasty and pulmonary artery re-implantation	19	68	3 balloon dilations	Severe cardiac ischemia with impaired cardiac function requiring ECMO, partially recovered over time Mild dyspnoea with exercise No hospitalizations for respiratory tract infections
2	12	Insertion tissue expander	1	6	Awaits slide tracheoplasty	8 days after surgery reduction of volume of tissue expander because of suspicion of compression on surrounding tissues Since diagnosis 3 hospitalizations because of respiratory insufficiency during airway infections
3	18	Slide tracheoplasty and pulmonary artery re-implantation	4	11	/	Residual tracheo/bronchomalacia Recurrent episodes of respiratory distress/insufficiency with respiratory infections, factor asthma Need for maintenance treatment with azithromycin and inhaled corticosteroids during winter months 1 hospitalization because of respiratory insufficiency
4	7	Slide tracheoplasty and pulmonary artery re-implantation	1	11	2 balloon dilations	Mild residual tracheo- and bronchomalacia with residual stridor mostly with exercise and infections Need for maintenance therapy with azithromycin prophylaxis and inhaled corticosteroids during winter months 3 hospitalizations because of respiratory insufficiency
5	117	Slide tracheoplasty	4	5	16 balloon dilations 2x CO_2 lasering Patch tracheoplasty 6 months after slide tracheoplasty	Residual severe stenosis with tracheal wall dehiscence Dyspnoea and stridor with moderate exercise Intermittent dysphagia and subjective swallowing difficulties
6	/	No surgical intervention	/	/	/	/

bronchoscopy, which provided important additional information about the degree and length of stenosis in both cases.

Figure 1a shows the round aspect of the trachea during bronchoscopy. Figure 1b-e show aOCT images of the same patient. Figure 2A shows a CT reconstruction of the trachea of patient 3 before tracheoplasty.

Treatment and outcome

Four patients have been treated surgically with slide tracheoplasty, three of them combined with pulmonary artery reimplantation. Table 2 shows treatment and outcome results of our study population. Age at surgical repair ranged between 7 and 117 months. Need for ventilatory support varied between 24 hours and 19 days, including the day of surgery. The patients were discharged from the paediatric ICU after 5 to 68 days. One patient was readmitted to the paediatric ICU shortly after discharge because of acute deterioration caused by restenosis. One patient with combined CLSTS and pulmonary agenesis was still awaiting slide tracheoplasty after initial insertion of a tissue expander in the left thoracic cage at the end of the study period. One patient was not treated surgically because of limited symptoms at the age of 12 years. All treatment strategies were discussed multidisciplinary between the paediatric pulmonologists and cardiothoracic surgeons of our institution, with additional advice of other health care professionals depending on the individual characteristics of the cases, as well as consultation of highly experienced international colleagues.

Figure 3 shows the evolution of the tracheal diameter in patient 3 on bronchography before and after tracheoplasty.

One patient suffered from severe perioperative cardiac ischemia, with prolonged need for extracorporeal membrane oxygenation and residual left ventricular dysfunction. Three patients required postoperative balloon dilations because of restenosis or excessive granuloma formation. One of these patients required additional CO_2 lasering and a pericardial patch tracheoplasty because of insufficient effect of minimally invasive treatment of the restenosis. Follow-up investigations showed tracheal wall dehiscence with the formation of a fibrotic band, for which redo surgery will be planned.

Three patients had residual tracheo- and/or bronchomalacia. One patient suffered from intermittent dysphagia and subjective swallowing difficulties. Two patients experience intermittent respiratory distress or stridor with respiratory tract infections. No other complications were observed in the study population, nor was there any mortality perioperatively or during follow-up. In July 2021, the length of follow-up after surgery ranged between 34 months and 6 years. For privacy reasons in this small patient population, the length of follow-up is not reported for each individual case.

Discussion

Our patient series, albeit small, illustrates the broad range of signs and symptoms that can be caused by CLSTS, and therefore the different ways these patients present in clinical practice. Some children were referred because of unexplained respiratory symptoms, some already with a tentative diagnosis of tracheal stenosis elsewhere, others after diagnosis of a pulmonary artery sling, and one child because of difficult intubation. Also the broad range of age at diagnosis and presenting symptoms is consistent with data from the literature, as is the rate of associated congenital anomalies (1,3,4,8). In our population, four out of six patients (66%) had associated congenital anomalies, three of which (50%) had a pulmonary artery sling and three of which had involvement of other organ systems: two (33%) had skeletal abnormalities, one (16%) had unilateral agenesis of the lung and one (16%) had an anal imperforation. These rates correlate well with the results from literature.

Most frequently, the diagnosis of CLSTS is made unsuspectedly when investigations are issued because of respiratory symptoms. When stridor is a prominent symptom, laryngotracheobronchoscopy is often the investigation of choice, as it can identify a wide variety of possible causes, like laryngo- and tracheomalacia, subglottic stenosis, airway haemangiomas, foreign body aspiration and vocal cord paralysis. When investigations are issued because of recurrent or persistent respiratory infections or when a vascular ring is suspected, a tentative diagnosis is often made by contrast enhanced chest CT.

In our study, all patients underwent a contrast enhanced chest CT as well as a bronchoscopy with bronchography. The two most recently diagnosed patients were also examined with aOCT during diagnostic bronchoscopy. As the measurement is three-dimensional and real-time, both longitudinal and transverse measurements can be performed. It also has the best resolution to evaluate the presence of complete cartilaginous tracheal rings compared to bronchoscopy or CT, as it can differentiate between different types of tissue underneath the surface of the respiratory epithelium. Additional advantages are the lack of radiation exposure and the possibility to use it during diagnostic bronchoscopy.

When decisions are made concerning surgical intervention, several factors must be taken into account. The surgery is complex, needs cardiopulmonary bypass and has a mortality rate between 5 and 12% (1,3,4,8). Therefore, only patients with severe symptoms are eligible for tracheoplasty and the decision should always be made by a multidisciplinary team. The ideal age for surgical repair is estimated between 10 and 24 months of age (8). When surgery is deemed necessary, slide tracheoplasty is the best technique available, as it has several advantages compared to other surgical strategies and a lower risk of complications like restenosis and excessive granulation tissue formation (1,4). The original blood supply to the tracheal wall is preserved, the doubling of the diameter of the trachea leads to quadrupling of the cross-sectional area and the suture is not circular, thereby decreasing the risk of restenosis. Also, the length of the trachea is only diminished by half the length of the stenosis, which makes it possible to correct even very long segments of diseased trachea, with sufficient tracheal growth afterwards (8).

The decision to treat conservatively should also be discussed multidisciplinarily and close follow-up with the possibility of reconsideration of surgical treatment should be organized if needed.

The most frequently occurring complications after surgery are restenosis and granulation tissue formation, which we encountered in three out of four operated patients, with one patient suffering from persistent granulation tissue formation requiring more than three interventions. A large single-centre study showed a need for additional endoscopic interventions of 48% (3). This high rate of reintervention is an important aspect to keep in mind, both to give patients and parents realistic expectations and to understand the need for intensive follow-up, especially in the first weeks to months after surgery. Whether reintervention is necessary or not, many children have residual symptoms after surgery. Three out of four operated patients had some degree of postoperative airway malacia, compared to 20-25% in larger studies, possibly partially because of different definitions of the degree of airway collapse needed for the diagnosis (1,3). Our patient with recurrent need for endoscopic interventions was known to develop excessive granulation tissue formation and keloid formation on her skin after surgery, possibly illustrating an underlying vulnerability to complications due to improper healing of damaged tissues. It could therefore partially explain her complicated postoperative course. Another possible explanation for the increased need for additional endoscopic interventions when comparing to the literature is the small size of the study population, making it more prone to skewing of results.

Our patients are currently 34 months, 46 months, 47 months, and 6 years in follow-up after slide tracheoplasty and all but one experience none to mild symptoms in daily life. One patient suffers from recurrent granulation tissue formation and still experiences respiratory distress with exercise and intermittent swallowing difficulties. Two patients experience respiratory distress and more frequent need of antibiotic treatment with respiratory tract infections, for which they are treated with inhaled corticosteroids, prophylactic antibiotics and respiratory physiotherapy during winter months. The larger studies available in literature do not clearly describe the frequency of this symptom in their population.

The patient who was treated conservatively, was referred to her own primary care physician for follow-up, with no new referrals to a paediatric pulmonologist or hospitalisations since diagnosis. One patient was still awaiting further treatment at the end of the study period and has been hospitalized

Figure 1a: shows the round aspect of the trachea during bronchoscopy, resulting from the absence of the pars membranacea, in patient 5.

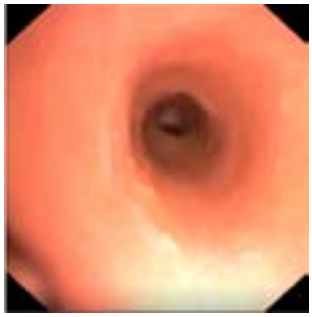
Figure 1b: shows an aOCT image of the same patient, with transverse imaging of the trachea in the top half of the figure and the longitudinal image in the lower half. Note the black circle on the transverse image (yellow arrow) illustrating how aOCT confirmed the presence and extent of complete tracheal rings. The green arrow shows at which point the transverse image was made. Moreover, the diameter and minimal cross-sectional area can be measured along the entire length of the trachea.

Figure 1c: shows the same image as figure 1b, with the gray circles visualising the edges of the cartilaginous ring.

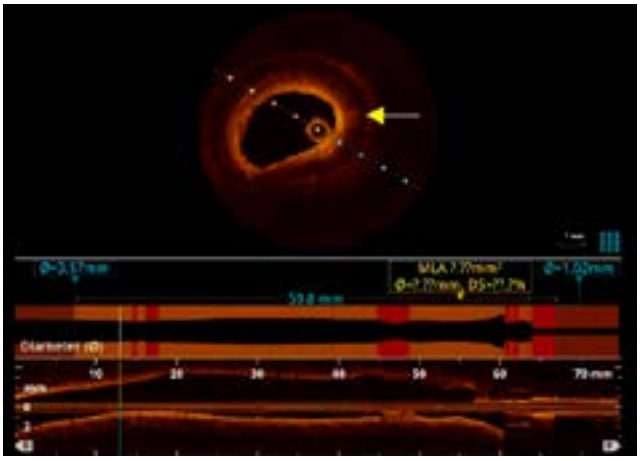
Figure 1d shows a normal part of the trachea with incomplete cartilaginous ring.

Figure 1e: shows a transverse image of the trachea in between two cartilaginous rings.

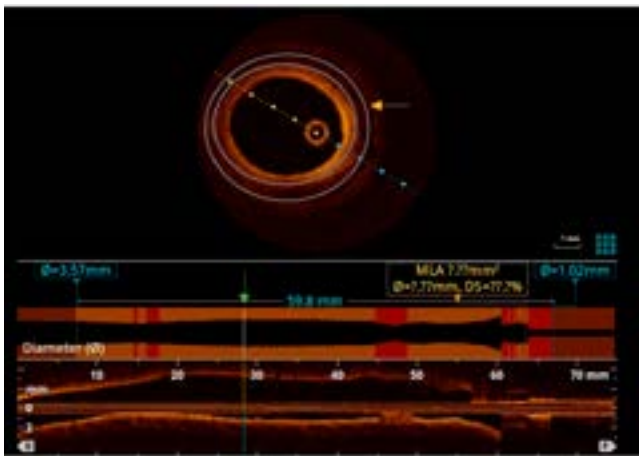
1a



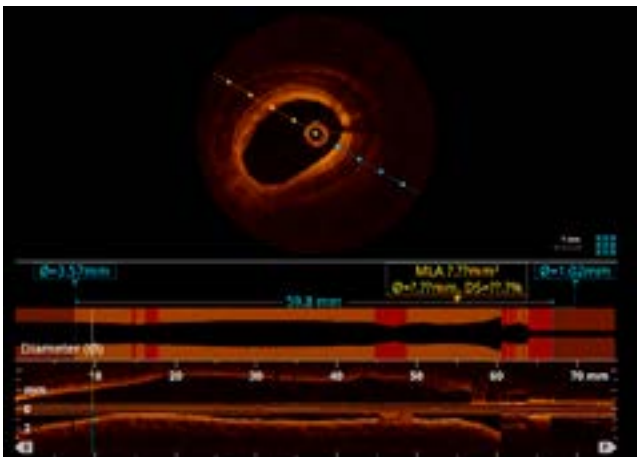
1a



1b



1c



1d

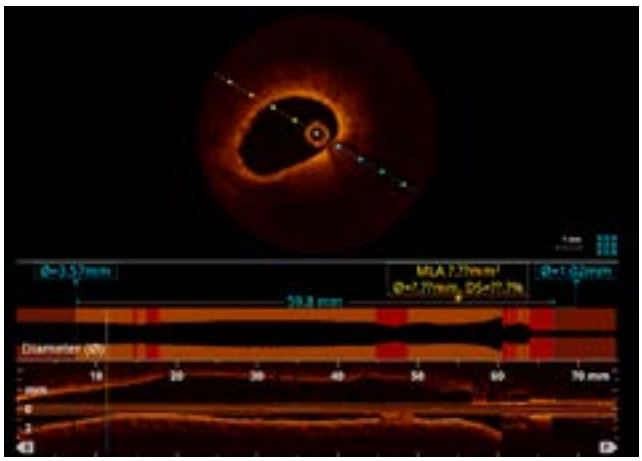
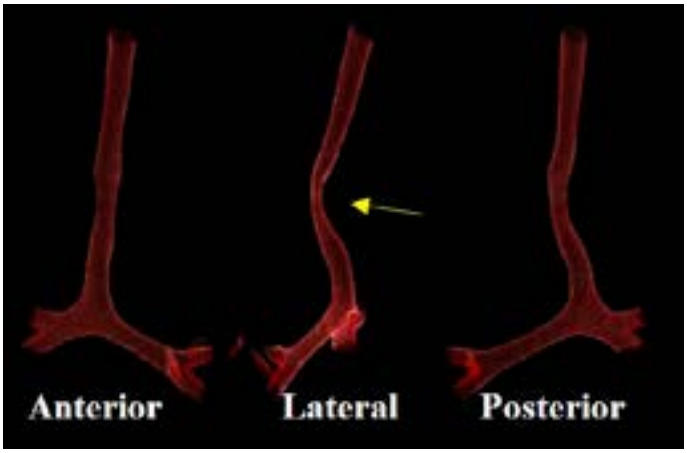


Figure 2a: shows a CT reconstruction of the trachea of patient 3 before tracheoplasty. The lateral view (middle panel) shows the external compression on the trachea from a pulmonary artery sling (yellow arrow).

Figure 2b: shows a transverse CT image showing both the narrow trachea (yellow arrow) and the presence of the pulmonary artery sling (red arrow) crossing the distal trachea posteriorly.

2a



2b

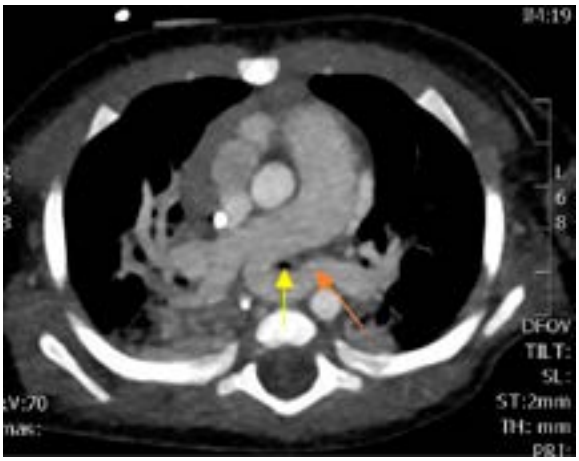
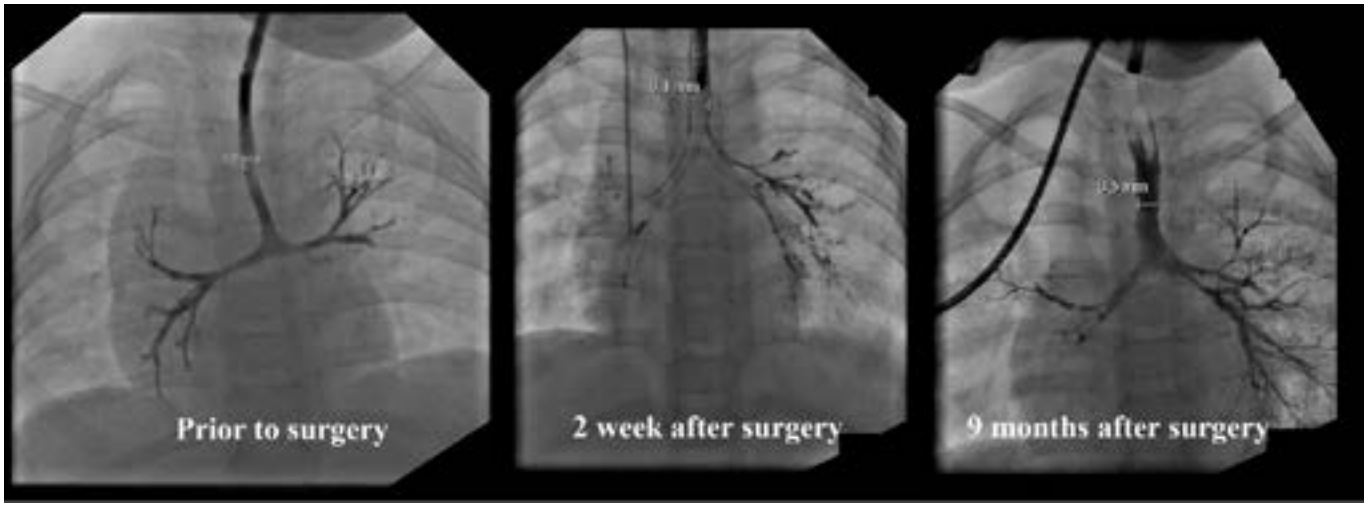


Figure 3: shows the evolution of the tracheal diameter in patient 3 on bronchography before (3.5 mm), 2 weeks after (8.1 mm) and 9 months after tracheoplasty (8.5 mm).



three times because of respiratory insufficiency with respiratory infections since diagnosis. During follow-up, some tracheal growth was seen with the narrowest luminal diameter of 3 mm at 32 months of age. Her slide tracheoplasty was eventually performed at the age of 41 months and was successful without perioperative complications. She was extubated the day after surgery and was hospitalized at the paediatric ICU for 8 days including day of surgery. So far, one bronchoscopic dilatation was performed during follow-up.

Strengths and limitations

Limitations of the study are the retrospective nature of the data collection and the small patient population. On the other hand, this is the first Belgian study about this topic and includes all patients with CLSTS diagnosed and/or treated at our hospital. Follow-up data are available for all but one patient. Another asset of the study is the use of aOCT in the most recently diagnosed patients, an investigational tool that allows more detailed visualisation of the stenosis when added to the diagnostic workup.

Conclusion

CLSTS is a rare but potentially life-threatening condition requiring an experienced multidisciplinary team of paediatric pulmonologists, cardiologists, cardiothoracic surgeons, ear-nose-throat surgeons, intensive care specialists and specialized paramedic healthcare professionals for diagnosis, treatment and follow-up. Dedicated care for these patients is important to optimize quality of care. Diagnosis is made by CT and bronchoscopy, with additional information gathered by bronchography and aOCT. Treatment with slide tracheoplasty is the method of choice when conservative treatment is insufficient. The most frequent complications are restenosis, granuloma formation and residual airway malacia.

This study displays our experience with the diagnosis and treatment of CLSTS, with no mortality, perioperative cardiac ischemia in one patient and need for postoperative interventions because of restenosis in three patients, of which one needed redo tracheoplasty with a pericardial patch.

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Conflicts of Interest

The authors have no conflict of interest to declare with regard to the subject discussed in this manuscript.

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