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ORIGINAL ARTICLE

Incidence of congenital tracheal stenosis in left pulmonary artery sling diagnosed by bronchoscopy

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Abstract

Purpose Congenital tracheal stenosis (CTS) has been reported to occur in 50–65% of cases of left pulmonary artery sling (LPAS), but the exact incidence rate is unknown. This study aimed to determine the actual rate using bronchoscopy and to elucidate morphological features in computed tomography (CT) diagnosis.

Methods We performed a single institutional retrospective review of all patients with LPAS between January 2010 and March 2022. The percentage of complete tracheal rings in patients with LPAS was evaluated using bronchoscopy. The anteroposterior/lateral diameter ratios at the smallest and largest diameters of each CTS patient's trachea were measured on CT. The Wilcoxon signed-rank test was used to analyze the differences between the two parts.

Results Fifty-two patients with LPAS were enrolled. All patients had complete tracheal rings on bronchoscopy. CT analysis of 32 patients with CTS was performed. The median anteroposterior/lateral diameter ratio at the smallest diameter was 1.05 (interquartile range [IQR] 0.95–1.15); the median ratio at the largest diameter was 0.94 (IQR 0.89–0.99). There was a significant difference between the two parts ($p = 0.013$).

Conclusion CTS might be universally associated with LPAS. The circular tracheal cross-section on CT might imply the existence of a complete tracheal ring.

Keywords Left pulmonary artery sling · Congenital tracheal stenosis · Bronchoscopy · Computed tomography

Introduction

Left pulmonary artery sling (LPAS) is a rare malformation in which the left pulmonary artery originates from the right pulmonary artery, runs between the trachea and esophagus, and then follows the dorsal aspect of the left main bronchus to the left pulmonary hilar region. LPAS is often associated with congenital tracheal stenosis (CTS), and to emphasize this association, the term "ring-sling complex" was used in a previous study [1].

CTS is an intractable condition associated with complete tracheal rings with a defective membranous trachea, resulting in narrowing of the tracheal lumen and consequent severe respiratory compromise. Past reports have indicated that 50–65% of patients with LPAS have concurrent CTS [1, 2]. However, some of these reports are old, so the actual incidence of CTS in patients with LPAS has been questioned [3]. CTS is ideally diagnosed endoscopically based on the identification of a complete tracheal ring [3]. However, because bronchoscopy requires general anesthesia, computed tomography (CT) is often used as an alternative to make a definitive diagnosis.

Our policy is to perform bronchoscopy in all patients with LPAS. In this study, we investigated the presence or absence of complete tracheal rings with posterior membranous defects, namely the CTS incidence by bronchoscopy for patients with LPAS over a 12-year period at a single children's hospital. We also evaluated CT scans, which are frequently used to diagnose CTS. CTS is called the "stovepipe"

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trachea because the tracheal cross-section at the complete tracheal ring is circular instead of the normal U-shape [4]. Therefore, we focused on the morphology of the tracheal cross-section at the complete tracheal ring and performed CT analysis.

The purposes of this study were to clarify the actual incidence rate of CTS in LPAS by bronchoscopy and review commonly used CT findings for diagnosis.

Methods

Ethics statements

The present study was approved by the institutional review board.

Study design and population

All patients diagnosed with LPAS from January 2010 to March 2022 at a single-center pediatric hospital in Japan were included in this retrospective study. This study had no exclusion criteria. We examined the incidence rate of complete tracheal rings during bronchoscopy (i.e., CTS). In our hospital, bronchoscopy was performed with a rigid bronchoscope, but a flexible bronchoscope was used for patients who had already been intubated at the time of admission to our hospital and for those with a clear diagnosis of CTS on CT. Bronchoscopy was performed by six board-certified pediatric surgeons. The treatment policy for LPAS and CTS at our hospital is as follows: in principle, all patients with LPAS are operated on, while the pediatric surgeon determines the indication for surgery for CTS based on the degree of stenosis on imaging evaluation and symptoms. In patients who underwent surgery for CTS, the presence of cartilage in the posterior tracheal wall on histological examination was also examined.

CT scan analysis of patients with CTS

Patients with LPAS from January 2010 to March 2022 were analyzed. Patients with only CT imaging during intubation, those who could not be analyzed using the SYNAPSE VINCENT system (Fujifilm, Tokyo, Japan) because CT was performed at another hospital, and those in whom the entire length of the trachea was not imaged were excluded from this analysis. If CT was performed multiple times, preoperative imaging was conducted. Two experienced reviewers assessed the preoperative CT images and came to a consensus. Patients were imaged in the supine position using 160- detector or 320-detector row CT scanner (Aquilion PREMIUM and Aquilion ONE; Cannon Medical Systems, Otawara, Japan). All the images were acquired using a mediastinal window. Using the SYNAPSE VINCENT system, the tracheal center was traced to avoid oblique cut distortion and to define the site of the smallest and largest diameter of the trachea. The anteroposterior/lateral diameter ratio of the tracheal cross-section is an indicator of shape of the tracheal cross-section. Therefore, the anteroposterior/lateral diameter ratios of the tracheal cross-section at the largest diameter of the trachea (i.e., a normal part of the trachea) and the smallest diameter of the trachea (i.e., a complete tracheal ring) in patients with CTS were measured on CT. The smallest diameter was measured at the site excluding the LPAS area to remove the factor of tracheal diameter change due to external compression of the trachea caused by LPAS.

Statistical analysis

The Wilcoxon signed-rank test was used to analyze the differences between the anteroposterior/ lateral diameter ratios in the smallest and largest parts of the trachea. SPSS software (IBM Corp., Armonk, NY, USA) was used to perform statistical analysis, and p values < 0.05 were considered to be statistically significant.

Results

Patient characteristics

Fifty-two patients (male/female ratio = 26:26) had LPAS during the study period, and all of them were analyzed. The chief complaint of LPAS was respiratory disorder in 42 patients and 10 patients were asymptomatic. Four asymptomatic patients were diagnosed with LPAS at fetal diagnosis, three at postnatal screening, and three at close examination for cardiac disease. All cases were diagnosed by ultrasound examination and contrast-enhanced CT. Forty-nine patients were referred from other hospitals because of the diagnosis of LPAS and CTS in 37 patients, LPAS in 7 patients, CTS in 2 patients, worsening respiratory status after LPAS repair in 2 patients, and critical respiratory management in 1 patient. Patient characteristics are shown in Table 1.

CTS complication rate in patients with LPAS

Of the 52 bronchoscopy procedures, 37 (71.2%) were performed using a rigid bronchoscope and 15 (28.8%) were performed using a flexible bronchoscope. The results showed that 52 (100%) of the 52 eligible patients had complete tracheal rings on bronchoscopy. The histological examination revealed the presence of cartilage in the posterior wall in all 43 patients who underwent surgery. The complete tracheal rings of the 9 nonoperative cases are shown on bronchoscopy in Fig. 1. The types of CTSs were generalized type in

Table 1 Patient characteristics ($n=52$)

Characteristics	Value
Age, month, median [IQR]	6.5 [3.75–11]
Sex, no. (%)	
Male	26 (50)
Female	26 (50)
Weight, g, median [IQR]	6105.5 [4791–7705]
Respiratory anomalies (%)	28 (53.8)
Tracheal bronchus	11 (21.2)
Single lung or unilateral lung hypoplasia	15 (28.8)
Others	2 (3.8)
Cardiovascular anomalies (%)	
Patent ductus arteriosus	11 (21.2)
Ventricular septal defect	10 (19.2)
Atrial septal defect	6 (11.5)
Coarctation of the aorta	5 (9.6)
Double outlet right ventricle	3 (5.8)
Total anomalous pulmonary venous connection	2 (3.8)
Tetralogy of Fallot	1 (1.9)
Partial anomalous pulmonary venous connection	1 (1.9)
Pulmonary atresia with ventricular septal defect	1 (1.9)
Pulmonary valve stenosis	1 (1.9)
Aortopulmonary window	1 (1.9)
Treatment	
Simultaneous repair	34 (65.4)
Staged repair	6 (11.5)
Only PA sling repair	9 (17.3)
Only tracheoplasty	3 (5.8)
Surgical approaches for CTS	
Slide tracheoplasty	43 (82.7)
Prognosis (%)	
Alive	51 (98.1)
Death	1 (1.9)

IQR interquartile range, *no*. number, *PA* pulmonary artery, *CTS* congenital tracheal stenosis

21 (40.4%), funnel type in 19 (36.5%) and segmental type in 12 (23.1%).

CT scan analysis of patients with CTS

Of the 52 patients with CTS, 32 were eligible for CT analysis. The remaining 20 patients were excluded from the analysis for various reasons: eight underwent CT imaging only during intubation, nine patients' CT images were not compatible with the SYNAPSE VINCENT system as the CT scans were performed at another hospital, and three patients did not have their entire length of the trachea imaged. CT analysis of 32 patients with CTS revealed that the median of the anteroposterior and lateral diameters at the maximum diameter of the trachea was 6.35 mm and 6.65 mm, respectively. The median anteroposterior and lateral diameters at the smallest diameter of the trachea were 3.45 mm and 3.3 mm, respectively. The results showed that the median anteroposterior/lateral diameter ratio at the largest diameter of the trachea was 0.94 (interquartile range [IQR] 0.89–0.99). The median ratio at the smallest diameter of the trachea was 1.05 (IQR 0.95–1.15). There was a significant difference in the median anteroposterior/lateral diameter ratio between these two parts ($p=0.013$) (Table 2).

Discussion

The treatment strategy for LPAS can vary depending on the existence of complete tracheal rings and the severity of airway symptoms. Recently, some authors advocated conservative LPAS repair alone as the treatment strategy [5–8]. However, the surgical indication for CTS in patients with LPAS should be carefully determined because airway symptoms immediately after LPAS repair alone tend to worsen due to increased secretion and tracheal edema. Mechanical stimuli by endotracheal suction and the endotracheal tube might cause granulation at the narrow segment and subsequent respiratory deterioration. In fact, of the 52 cases observed in this study, 2 infants were transferred from another hospital because of respiratory failure after LPAS surgery. Therefore, simultaneous surgery (LPAS and CTS repair) was advocated by the centers with substantial experience in tracheal surgery [9–11].

Surgical decision making for tracheoplasty in patient with LPAS and mild CTS is difficult and should be carefully determined. Huang et al. recommended the consideration of concomitant tracheoplasty in patients with a tracheal diameter of < 3 mm [5]. Choi and colleagues retrospectively reviewed 22 patients who underwent LPAS repair only and identified 3.4 mm at the narrowest preoperative diameter of the trachea as the cut-off value for predicting hospital readmission for respiratory symptoms. They also demonstrated that the growth rate of the narrowest segment of the trachea after LPAS repair was 0.24 mm per year [6].

As aforementioned, because the presence or absence of CTS is related to the treatment strategy for patients with LPAS, it is essential to know the presence of CTS before LPAS repair is performed. The most reliable method for the definitive diagnosis of CTS is rigid bronchoscopy, which is performed by experts [3]. Bronchoscopy detects a complete tracheal ring; however, to our best knowledge, there is only one report of bronchoscopy being performed in all LPAS cases to identify complete tracheal rings. Herein, 52 LPAS cases over a 12-year period at a single pediatric hospital were evaluated for the presence of

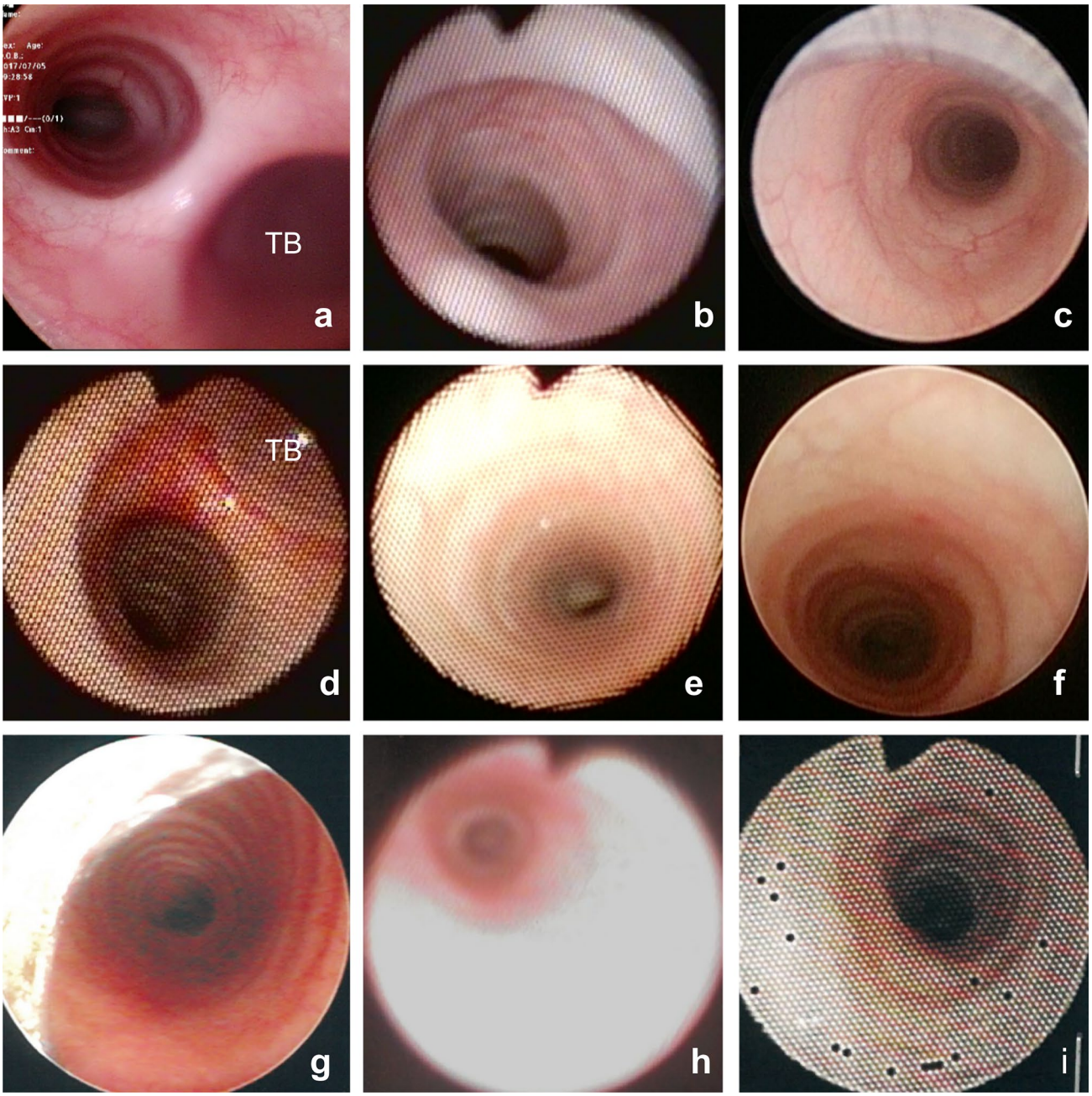

Fig. 1 Bronchoscopy findings of nonoperative patients (Patients **a**–**i**)

complete tracheal rings on bronchoscopy; complete tracheal rings were observed in all LPAS cases. This rate is a significant deviation from those in previous reports. For several decades, the incidence of CTS in patients with LPAS has been reported to range from 50 to 65% [4, 6, 12]. However, those exact percentages were not described in the cited literatures [1, 2]. In a case series by Cohen et al., complete tracheal ring was confirmed by autopsy in all 3 patients [2]. Berdon and colleagues reported that 5 patients with LPAS autopsy studies revealed complete tracheal ring in all 3 fatal cases, and high-kV radiography delineated the anatomical features of CTS in 2 surviving patients [1]. Rigid bronchoscopy clearly depicts the tracheal cartilages. However, this procedure is invasive and requires general anesthesia. Thus, the indication for rigid bronchoscopy in patients with minimal airway symptoms might be controversial. In addition, rigid bronchoscopy poses a risk for respiratory catastrophe in patients with extreme severe CTS whose narrowest segment was stented by an endotracheal tube. In those patients, the narrowest

Table 2 Anteroposterior/lateral diameter ratios of the tracheal cross-section

	Anteroposterior/lateral diameter ratio of the largest diameter of the trachea	Anteroposterior/lateral diameter ratio of the smallest diameter of the trachea	*p* value
Median [IQR]	0.94 [0.89–0.99]	1.05 [0.95–1.15]	0.013

The Wilcoxon signed-rank test was used to analyze the differences between the ratio of the largest diameter of the trachea (i.e., the normal part of the trachea) and the smallest diameter of the trachea (i.e., a complete tracheal ring)

IQR interquartile range

segment might be completely occluded soon after tracheal extubation for bronchoscopic examination. Thus, reports of rigid bronchoscopic surveillance in patients with LPAS have been limited. In 2012, Backer et al. from Children's Memorial Hospital in Chicago reported on 34 patients with LPAS [9]. All 34 patients underwent rigid bronchoscopy in the operating room immediately before surgical repair, and 27 patients (79%) had CTS with complete tracheal rings [9]. In 2020, Muthialu and colleagues also reported on 79 consecutive patients who underwent surgery for LPAS at the Great Ormond Street Hospital [13]. Although diagnostic modalities were not described in their study, the incidence of CTS with complete cartilaginous ring was 87% (69/79) [13].

It is often difficult to identify the complete tracheal ring on CT owing to its limited density resolution. Therefore, generally, the diagnosis of CTS on CT is made based on the presence of "stenosis" of the trachea. Stenosis is usually determined by the caliber change in the tracheal diameter and absolute small diameter of the trachea. However, there are cases in which the caliber change is unclear in the generalized type, where stenosis extends the entire length of the trachea. There are also cases in which a complete tracheal ring exists but the tracheal diameter is preserved. Therefore, we speculate that these CTS cases are difficult to diagnose using CT. In our pediatric hospital, we perform bronchoscopy in all patients with LPAS regardless of the presence of symptoms or CT findings; consequently, we may be diagnosing CTS patients with mild symptoms who have been previously missed or cases that are difficult to diagnose with CT.

To prevent missing CTS cases caused by complete tracheal rings on CT, we focused on the fact that the tracheal cross-section is circular rather than U shaped [4]. In the 32 cases analyzed using CT in this study, the median anteroposterior/lateral diameter ratio of the tracheal cross-section at the complete tracheal ring was significantly different from that of the normal part of the trachea in each patient ($p = 0.013$). Therefore, when the diagnosis of CTS is made based on CT findings instead of rigid bronchoscopy because of its invasiveness, it may be important to focus on the morphology of the tracheal cross-section and look for a round trachea in addition to stenosis of the trachea.

Although this study included a relatively large number of patients with LPAS, the most important limitation of this study is the small study population; LPAS is rare, and individual centers have limited experience. In addition, most of the LPAS patients included in this study were found with respiratory symptoms or on close examination for cardiac disease. Therefore, this study did not include all asymptomatic, undiagnosed LPAS patients, which may represent a selection bias. Furthermore, although bronchoscopy should be performed with a rigid bronchoscope with good resolution, some patients are diagnosed using flexible bronchoscopy. However, all patients diagnosed with flexible bronchoscopy underwent surgical treatment for CTS, and the pathology showed a complete tracheal ring. Additionally, CT is not respiration-synchronized, and the aspect ratio of the tracheal cross section at normal sites may not be appropriate as a value for statistical comparison. On the other hand, the tracheal cross-sectional aspect ratio at the complete tracheal cartilage ring site is close to 1, making it unlikely that tracheal cross-sectional diameter is affected by respiration. A final limitation of this study is the definition used for normal and complete tracheal ring sites on CT analysis. The largest diameter site was defined as the normal part of the trachea and the smallest diameter site was defined as the complete tracheal ring. However, this is a theoretical definition and not necessarily reflective of actual bronchoscopic findings. The height of the tracheal cartilage could not be accurately determined on CT, making it impossible to precisely match the tracheal diameter with the bronchoscopic findings. Therefore, the definition is based on theoretical assumptions, where the smallest diameter site, excluding the LPAS site, is considered the complete tracheal ring, and the largest diameter site is considered the normal part of the trachea.

In conclusion, this study's findings suggest that the incidence rate of CTS in LPAS can occur more frequently than previously reported, possibly with an incidence rate of 100%. The possibility of CTS can be considered to prevent respiratory failure after LPAS surgery. In addition, the morphology of the tracheal cross-section may be useful when diagnosing CTS on CT. Continued rigid bronchoscopy evaluation of additional LPAS cases for CTS is warranted.

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Author contributions All authors contributed to the study conception and design. Material preparation, data collection and analysis were performed by KM, ET and HM. The first draft of the manuscript was written by ET and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

Data availability The data that support the findings of this study are available from the corresponding author upon reasonable request.

Declarations

Conflict of interest All authors declare no conflicts of interest.

Ethics approval This retrospective study was conducted in accordance with the principles of the Declaration of Helsinki and was approved by the Ethics Committee of Hyogo Prefectural Kobe Children's Hospital with opt-out consent.

References

1. Berdon WE, Baker DH, Wung JT et al (1984) Complete cartilage-ring tracheal stenosis associated with anomalous left pulmonary artery: the ring-sling complex. Radiology 152:57–64. https://doi.org/10.1148/radiology.152.1.6729137
2. Cohen SR, Landing BH (1976) Tracheostenosis and bronchial abnormalities associated with pulmonary artery sling. Ann Otol Rhinol Laryngol 85:582–590. https://doi.org/10.1177/000348947608500504
3. Torre M (2017) Left pulmonary artery sling and congenital tracheal stenosis: to slide or not to slide? J Thorac Dis 9:4881–4883. https://doi.org/10.21037/jtd.2017.11.76
4. Fiore AC, Brown JW, Weber TR, Turrentine MW (2005) Surgical treatment of pulmonary artery sling and tracheal stenosis. Ann Thorac Surg 79:38–46. https://doi.org/10.1016/j.athoracsur.2004.06.005
5. Huang SC, Wu ET, Wang CC et al (2012) Surgical management of pulmonary artery sling: trachea diameter and outcomes with or without tracheoplasty. Pediatr Pulmonol 47:903–908. https://doi.org/10.1002/ppul.22516
6. Choi ES, Park CS, Kim DH et al (2022) Outcomes of pulmonary artery sling repair without tracheoplasty. J Thorac Cardiovasc Surg S0022–5223(22):01255–01257. https://doi.org/10.1016/j.jtcvs.2022.11.017
7. Binsalamah ZM, Thomason A, Ibarra C et al (2021) Midterm outcomes of pulmonary artery sling repair with and without tracheoplasty. Cardiol Young 31:52–59. https://doi.org/10.1017/s1047951120003212
8. Kwak JG, Kim WH, Min J, Lee C, Jang W, Lee CH (2013) Is tracheoplasty for all patients with pulmonary artery sling and tracheal stenosis? Pediatr Cardiol 34:498–503. https://doi.org/10.1007/s00246-012-0481-7
9. Backer CL, Russell HM, Kaushal S, Rastatter JC, Rigsby CK, Holinger LD (2012) Pulmonary artery sling: current results with cardiopulmonary bypass. J Thorac Cardiovasc Surg 143:144–151. https://doi.org/10.1016/j.jtcvs.2011.09.038
10. Oshima Y, Yamaguchi M, Yoshimura N et al (2008) Management of pulmonary artery sling associated with tracheal stenosis. Ann Thorac Surg 86:1334–1338. https://doi.org/10.1016/j.athoracsur.2008.04.020
11. Manning PB, Rutter MJ, Lisec A, Gupta R, Marino BS (2011) One slide fits all: the versatility of slide tracheoplasty with cardiopulmonary bypass support for airway reconstruction in children. J Thorac Cardiovasc Surg 141:155–161. https://doi.org/10.1016/j.jtcvs.2010.08.060
12. Hong X, Li R, Zhao Z et al (2020) Tracheal development after left pulmonary artery reimplantation: an individual study. Sci Rep 10:17702. https://doi.org/10.1038/s41598-020-74890-4
13. Muthialu N, Martens T, Kanakis M et al (2020) Repair of pulmonary artery sling with tracheal and intracardiac defects. Asian Cardiovasc Thorac Ann 28:463–469. https://doi.org/10.1177/0218492320943342