Surgical management of pulmonary artery sling in children

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Objectives: Pulmonary artery (PA) sling is a rare vascular anomaly associated with congenital tracheal stenosis. The natural history is poor and these patients often require early surgical intervention. We describe our experience with repair of this condition.

Methods: From 1984 to 2011, 21 patients with PA sling underwent repair at the Royal Children's Hospital (median age, 5.9 months). PA sling was associated with compression of the trachea in all patients. Tracheal surgery was required in 12 (57.1%) patients. All patients had an echocardiogram, and concomitant repair of coexisting cardiac anomalies was performed in 6 (28.6%, 6/21) patients.

Results: Operative mortality was 14.3% (3/21), occurring at 19 days, 4.4 months, and 5 months after surgery. Operative mortality for the first 10 years was 22.2% (1984-1993; 2/9), the next 10 years was 14.3% (1994-2003; 1/7), and 0% for the most recent 7 years (2004-2011; 0/5). All deaths occurred in patients requiring tracheal repair (25%, 3/12). No deaths have occurred since 2004 with introduction of the slide tracheoplasty technique. One (5.6%, 1/18) late death occurred at 8 months after repair. After tracheal repair, intervention for excessive granulations and tracheomalacia was necessary in 6 (50%, 6/12) patients. Median follow-up was 8 years (mean, 8.6 ± 6.4 years; range, 5 months to 20.6 years), and all survivors (100%, 17/17) remain asymptomatic.

Conclusions: Children with PA sling who do not require tracheal surgery have excellent outcomes. Mortality is determined by the need for tracheal surgery. However, with the advent of the slide tracheoplasty technique, mortality can be reduced. Survival beyond 1 year after surgery offers excellent prognosis. (J Thorac Cardiovasc Surg 2013;145:1033-9)

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Pulmonary artery (PA) sling is a rare congenital vascular anomaly whereby the left PA originates from the posterior aspect of the right PA. The left PA passes between the trachea and esophagus toward the left lung, compressing the lower trachea. The anomaly is often referred to as a ''ring–sling complex'' owing to high prevalence of complete tracheal rings (50%-65%).[1,2] External compression and intrinsic stenosis of the trachea lead to respiratory symptoms. The natural history of PA sling is poor with death resulting from airway obstruction.[3] The first successful surgical repair of PA sling was performed in 1953 by Potts, Holinger, and Rosenblum.[4] Current surgical management often involves concomitant repair of tracheal stenosis and coexisting cardiac lesions. Various techniques of tracheal repair have been described.[5] Reported outcomes in this rare group of patients are limited to case reports and small studies. Herein we reviewed our experience with PA sling repair performed at a single institution over a period of 27 years.

PATIENTS AND METHODS

Patients

The institutional research ethics board approved this study. Patients who underwent surgery for PA sling at the Royal Children's Hospital from 1984 to 2011 were identified (n = 21). Data were obtained by review of medical records from admission until the last follow-up. The patients were followed up regularly by cardiologists or respiratory physicians postoperatively with echocardiograms or bronchoscopy or both.

Definitions

Operative mortality was defined as death occurring within 30 days of surgery or before hospital discharge. All other deaths were considered late. Tracheal stenosis was associated with compression by the PA sling with or without complete cartilage rings. This was determined at preoperative bronchoscopy, bronchography, computerized tomography (CT), or magnetic resonance imaging (MRI).

Surgical Technique

Surgery was performed via median sternotomy. The ductus or ligamentum was ligated and divided. Standard hypothermic cardiopulmonary bypass (CPB) was then achieved. The left PA reimplantation technique was as follows. The left PA was dissected to the left hilar branches. Care was taken to avoid phrenic nerve and recurrent laryngeal nerve injury. The left PA was clamped and detached from its origin leaving a small cuff. The left PA was mobilized, brought anterior to the trachea, and

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Abbreviations and Acronyms

CPB = cardiopulmonary bypass
CT = computed tomography
ECMO = extracorporeal membrane oxygenation
MRI = magnetic resonance imaging
PA = pulmonary artery

anastomosed to the main PA trunk (Figure 1). Great care was taken to have a larger anastomosis than the actual diameter of the left PA and to perform the anastomosis at the posterior left aspect of the main PA (Figure 1, *C*).

The tracheal repair techniques performed at the Royal Children's Hospital included end-to-end anastomosis, patch tracheoplasty (pericardium, tracheal autograft, aortic homograft), and slide tracheoplasty. Patch tracheoplasty was performed with an anterior midline incision through the stenotic segment. The tracheal lumen was enlarged with a patch of autologous pericardium. If the stenotic segment was short, it was resected and trachea was anastomosed end-to-end. The slide tracheoplasty technique was performed by tracheal division at the midpoint of stenosis, longitudinal incisions on the opposite side of proximal and distal tracheal segments, and sliding oblique anastomosis of the segments.[6,7] Carinal stabilization and interposition pericardial flap were used as previously described.[8,9] After completion of tracheal repair, the mediastinum was filled with saline and the lungs were ventilated to test for air leak. Bronchoscopy was occasionally performed intraoperatively to assess the tracheal repair.

Postoperatively, patients were supported by mechanical ventilation and managed by a multidisciplinary team consisting of surgeons, cardiologists, and thoracic and intensive care physicians. The PA reimplantation site was assessed by postoperative echocardiograms. Frequent bronchoscopies were performed in patients who had tracheal repair.

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Data Analysis

All data were analyzed with Stata version 10 (Stata Corp, College Station, Tex). Descriptive statistics for continuous variables were expressed as means ± standard deviations (range), and skewed continuous data were presented by medians and interquartile ranges. Variables were summarized as frequencies and percentages.

RESULTS

The individual patient characteristics, operative procedures, and outcome are summarized in Table 1. There were 12 (57.1%, 12/21) male and 9 (42.9%, 9/21) female patients. Six patients (28.6%, 6/21) had coexisting cardiac anomalies including one or more of the following: atrial septal defect (n = 3; 14.3%, 3/21), ventricular septal defect (n = 3; 14.3%, 3/21), tetralogy of Fallot (n = 2; 9.5%, 2/21), right PA stenosis (n = 1; 4.8%, 1/21), pulmonary atresia with hypoplastic left ventricle (n = 1; 4.8%, 1/21), and partial anomalous pulmonary venous drainage (n = 1; 4.8%; 1/21). In addition, there were 2 (9.5%, 2/21) patients with a tracheal bronchus, 1 (4.8%, 1/21) with agenesis of the right lung, and 2 (9.5%, 2/21) with a hypoplastic right lung. All patients had symptoms of tracheal stenosis ranging from mild stridor to severe respiratory distress. Twelve (57.1%, 12/21) patients required preoperative intubation with 2 (9.5%, 2/21) of them receiving extracorporeal membrane oxygenation (ECMO). PA sling was diagnosed with an echocardiogram (n = 21; 100%, 21/21) along with cardiac catheterization in 7 patients (33.3%, 7/21). Cardiac catheterization has not been performed since 1996. MRI (n = 3; 14.3%, 3/21), CT (n = 10; 47.6%, 10/21), bronchoscopy (n = 13; 61.9%, 13/21), or bronchography (n = 9; 42.9%, 9/21) were used to diagnose and evaluate the degree of tracheal stenosis. There were 3 (14.3%, 3/21) patients who had prior cardiac intervention before PA sling surgery. Two patients had Blalock-Taussig shunts and 1 had balloon dilatation of the pulmonary valve.

Distribution of patients is outlined in Figure 2. The median age at surgery was 5.9 months (2.3-16.4 months). The median weight at surgery was 6 kg (4.2-9.7 kg). The median length was 66 cm (55-85 cm). The chest was opened through midline sternotomy in 20 (95.2%, 20/21) patients and bilateral thoracotomy in the first patient of the series (4.8%, 1/21). Standard CPB was used for surgery with exception of 1 patient who was supported by ECMO. Mean CPB time was 118 ± 50 minutes (47-228 minutes). The mean minimum temperature was 32°C ± 3.2°C (22°-34°). Four (19.1%, 4/21; range, 40-66 minutes) patients required aortic crossclamping. Circulatory arrest was required in 1 patient for 10 minutes. Technique of left PA reimplantation was standard for all patients.

The median hospital stay was 24 days (18-76 days) and the median postoperative ventilation time was 9 days (4-24 days).

Tracheal Repair

A total of 12 (57.1%, 12/21) patients had severe tracheal stenosis necessitating tracheal surgery. Difficulties with ventilation and/or inability to wean from mechanical ventilation were the indications for tracheal repair. Nine (42.9%, 9/21) patients underwent tracheal repair at the time of PA sling repair, whereas 3 (14.3%, 3/21) further patients had tracheal reconstruction after PA sling repair. The techniques performed included: autologous pericardial patch tracheoplasty (n = 5; 41.7%, 5/12), aortic homograft patch repair (n = 1; 8.3%, 1/12), slide tracheoplasty (n = 4; 33.3%, 4/12), tracheal autograft repair facilitated with a small patch of aortic homograft (n = 1; 8.3%, 1/12), and end-to-end anastomosis (n = 1; 8.3%, 1/12). In comparing patients requiring tracheal repair with PA sling repair only, the mean CPB time was 148 ± 39 minutes (95-228 minutes) and 84 ± 39 minutes (47-175 minutes), respectively. Median hospital stay was 35 days (20-131 days) and 22 days (8-32 days) for patients requiring tracheal repair and PA sling repair only, respectively. Median intubation time for tracheal repair was 14 days (6-55 days) compared with 1 day (<1-11 days) for the patients with PA sling repair only.

Concomitant Repair

At the time of PA sling repair, 6 (28.6%, 6/21) patients had repair of associated cardiac anomalies. This included

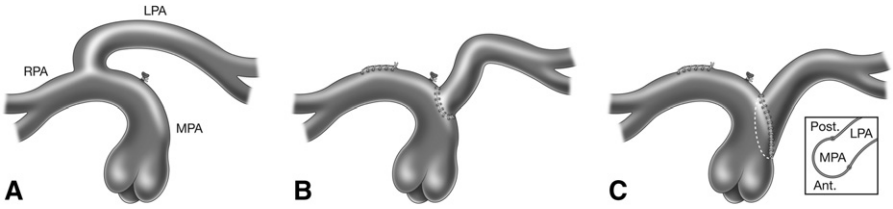

FIGURE 1. Pulmonary artery sling reimplantation technique. A, After ligation and division of the patent ductus arteriosus, the RPA is to be reimplanted to the MPA. B, Reimplantation of the LPA into the distal part of the MPA may cause kinking and result in restenosis. C, The LPA is opened widely and reimplanted to the posterior wall of the proximal MPA immediately above the pulmonary valve. *Ant*., Anterior; *Post*., posterior; *LPA*, left pulmonary artery; *MPA*, main pulmonary artery; *RPA*, right pulmonary artery.

one or more of the following: ventricular septal defect closure (n = 2; 9.5%, 2/21), atrial septal defect closure (n = 3; 14.3%, 3/21), shunt insertion (n = 2; 9.5%, 2/21), correction of tetralogy of Fallot (n = 1; 4.8%, 1/21), right PA enlargement with a polytetrafluoroethylene patch (W. L. Gore & Associates, Inc, Flagstaff, Ariz) (n = 1; 4.8%, 1/21), and partial anomalous pulmonary venous drainage repair (n = 1; 4.8%, 1/21).

Mortality

Overall operative mortality was 14.3% (n = 3; 3/21). Mortality by era is displayed in Table 2. There were no deaths (n = 0; 0%, 0/9) in patients who had PA sling repair only. In patients requiring tracheal surgery, there were 3 early deaths (25%, 3/12). No deaths have occurred in 5 patients since 2004, although all have required tracheal repair during the same admission. There were no deaths among patients who had concomitant cardiac repair (n = 0; 0%, 0/6). Detailed description of the patients who died during hospital admission is as follows:

A 6-month-old girl (Table 1, patient 6) was transferred with ventilatory support from interstate. PA sling, agenesis of the right lung, and severe tracheobronchial stenosis were diagnosed. The patient underwent repair of PA sling and pericardial patch tracheoplasty. Postoperatively, the patient was ventilator dependent and cardiac tamponade developed, necessitating reoperation. She then had episodes of respiratory distress with bradycardia, and eventually her condition deteriorated with increasing airway obstruction owing to patch collapse. Treatment was withdrawn on parental request. The patient died of respiratory failure 19 days after the operation.

A 4-month-old boy (Table 1, patient 9), born prematurely, was admitted with worsening stridor since birth and required intubation. PA sling with severe tracheal stenosis extending from the thoracic inlet to the carina was diagnosed. Left PA reimplantation and pericardial patch tracheoplasty were performed. The patient had postoperative difficulties with ventilation owing to patch collapse. Reoperation to place a Dacron tube to stent the collapsing portion of trachea was performed. Subsequently, a tracheostomy tube was inserted. He had difficulties being weaned from the ventilator and had a prolonged stay in the intensive care unit (>4 months). The patient required 2 balloon dilatations and an intratracheal Palmaz stent insertion (Cordis Europa NV, Roden, The Netherlands). A subsequent bronchoscopy revealed swelling between the base of the tracheal tube and stent, believed to be due to trauma. The tracheal tube was removed and the patient placed on nasal continuous positive airway pressure. The patient died of respiratory failure 5 months after the operation.

An 18-month-old girl (Table 1, patient 15) was diagnosed with PA sling during infancy and had increased respiratory distress and stridor necessitating preoperative intubation and ECMO. The following day, the patient underwent repair of PA sling and slide tracheoplasty. The postoperative course was complicated by a cerebral vascular accident and rupture of the slide tracheoplasty anastomosis requiring reconstruction of the trachea using an aortic homograft. In addition, the patient required a tracheostomy and postoperative ECMO. This was complicated by homograft collapse and difficulty with ventilation. She required 4 balloon dilatations and insertion of a Y-shaped silicone rubber Dumon stent (Novatech Technologies, Bouches Du Rhone, France). Subsequently, the stent dislodged; thus a Palmaz stent (Cordis Europa NV) and a Polyflex stent (Rüsch AG, Kemen, Germany) were inserted into the bronchus and lower trachea, respectively. The airway obstruction worsened and an airway could not be secured. Treatment was withdrawn on parental request and the patient died of respiratory failure 4.4 months after the operation.

Among 18 operative survivors, there was 1 late death (5.6%, 1/18). A 7-month-old boy (Table 1, patient 5) had respiratory distress requiring admission to the intensive care unit and continuous positive airway pressure support. Surgery to correct PA sling and pericardial patch tracheoplasty was performed. Postoperative bronchoscopy revealed stenosis of the trachea. Reoperation to further repair the trachea was planned, but dense adhesions prevented any surgical repair. The patient had 5 balloon dilatations of the trachea, eventually was extubated, and was discharged home. He died suddenly at home 5 months after discharge (8 months after the operation). Autopsy could not determine the cause of death.

TABLE 1. Summary of patients

Pt	Year	Age	Sex	Weight (kg)	Associated anomalies	Preoperative ventilation	Tracheal stenosis (% of total length)	Tracheal repair	Reoperation and reintervention	Duration of postoperative intubation (d)	Duration of hospital stay (d)	Outcome
1	1984	1 mo	F	4.4	None	+	None	None	None	8	40	Asymptomatic
2	1986	117 mo	M	40	None	−	33	Pericardial patch	None	4	14	Asymptomatic
3	1988	16 mo	M	9.7	None	−	None	None	None	<1	9	Asymptomatic
4	1990	37 mo	F	13	Pulmonary atresia, VSD, HLV	−	50	Pericardial patch	Bilateral bidirectional Glenn shunt, small Blalock shunt, central shunt, diaphragm plication	4	18	Asymptomatic
5	1991	7 mo	M	6.1	Hypoplastic right lung	−	50	Pericardial patch	Bronchoscopic tracheal balloon (5)	67	126	Late death
6	1991	6 mo	F	5.3	Absent right lung	+	50	Pericardial patch	None	18	20	Early death
7	1992	17 d	M	4.2	None	+	None	None	None	1	22	Asymptomatic
8	1992	2 mo	M	4	None	+	None	None	None	11	15	Asymptomatic
9	1993	4 mo	M	5.4	None	+	100	Pericardial patch	Bronchoscopic tracheal balloon (2), stent, tracheoplasty with Dacron tube	107	164	Early death
10	1995	5 mo	M	8.1	None	−	None	None	None	1	24	Asymptomatic
11	1996	15 mo	F	8	ASD, TOF	+	None	None	TOF repair	24	76	Asymptomatic
12	1996	2 mo	F	4.1	TOF	+	None	None	PA balloon (2); TOF repair	12	23	Asymptomatic
13	1997	56 mo	M	19.5	None	−	None	None	None	<1	7	Asymptomatic
14	1998	8 d	F	3.3	None	−	None	None	None	0	4	Asymptomatic
15	2003	17 mo	F	10	None	+	40	Aortic homograft patch	Bronchoscopic tracheal balloon (4), stent, reoperation for leakage	72	135	Early death
16	2003	22 mo	M	12	None	+	50	Tracheal autograft with aortic homograft	Bronchoscopic tracheal balloon (2), stent, reoperation for leakage	31	84	Asymptomatic
17	2008	10 mo	M	6	Hypoplastic right lung	+	70	Slide	Bronchoscopic tracheal balloon (14), granulation excision, ECMO	42	219	Asymptomatic
18	2009	2 mo	M	4	ASD, VSD	−	70	Slide	Granulation excision	9	35	Asymptomatic
19	2010	2 mo	F	3.4	ASD, VSD	−	60	Slide	None	10	35	Asymptomatic
20	2010	4 mo	M	6	None	+	60	Slide	None	5	31	Asymptomatic
21	2011	9 mo	F	6.5	None	+	20	End to end	None	7	20	Asymptomatic

VSD, Ventricular septal defect; *HLV*, hypoplastic left ventricle; *ASD*, atrial septal defect; *TOF*, tetralogy of Fallot; *ECMO*, extracorporeal membrane oxygenation; *PA*, pulmonary artery.

The overall mortality was 19% (n = 4; 4/21). Fourteen (66.7%, 14/21) patients were operated on under the age of 1 year with 3 deaths (21.4%, 3/14). There was 1 death in those operated on over the age of 1 year (14.3%, 1/7).

Reoperation and Reintervention

Severe left PA stenosis developed in 1 (4.8%, 1/21) patient 9 months after the operation. The patient required 2 balloon dilatations and is currently well with no further stenosis at last echocardiogram. Significant granulation tissue developed in 6 (50%, 6/12) patients after tracheal surgery and necessitated prolonged mechanical ventilation (mean, 55 ± 35 days; range, 9-107). These complications developed within 4 months after the operation. Patients in whom granulation tissue developed required multiple tracheal balloon dilatations (n = 5; 83.3%, 5/6), granulation excisions (n = 2; 33.3%, 2/6), and stent insertions (n = 3; 50%, 3/6). The stent material used included Palmaz

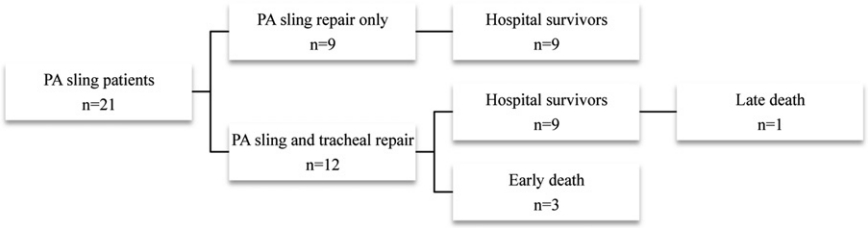

FIGURE 2. Flow chart of pulmonary artery *(PA)* sling patients (n = number of patients).

(Cordis Europa NV), silicone rubber Dumon stent (Novatech), and Polyflex stent (Rüsch AG). Of the 6 patients who encountered this complication, 3 died (50%, 3/6).

Reoperation was required in 6 (28.6%, 6/21) patients. Three (14.3%, 3/21) patients had reoperation on the trachea: repair of tracheal rupture with an aortic homograft (n = 2) and stabilization of pericardial patch collapse with Dacron tube graft (n = 1). Three (14.3%, 3/21) patients required further reoperations to correct other cardiac anomalies. The patient with pulmonary atresia, ventricular septal defect, and hypoplastic left ventricle had multiple reoperations including central shunt creation, bilateral bidirectional Glenn shunt, arteriovenous fistula, and left diaphragm plication. Two patients with tetralogy of Fallot had right ventricle outlet tract reconstruction.

Long-Term Status

Median follow-up for the 17 survivors was 8 years (3.1-12.7 years; mean, 8.6 ± 6.4 years; range, 5 months to 20.6 years). All (100%, 17/17) patients were asymptomatic at last follow-up.

DISCUSSION

The literature on the outcomes of PA sling repair is limited owing to the rarity of the condition.[10-13] Thus, we analyzed our experiences with PA sling repair at the Royal Children's Hospital. Case histories of 2 patients from this series have been previously reported.[8,9]

Clinical presentation varies with the spectrum of severity of airway compromise. The majority of patients had stridor. Several patients had severe airway obstruction requiring preoperative ventilatory support and, occasionally, ECMO. Severe obstruction was usually triggered by respiratory tract infection, presumably owing to further airway compromise by mucosal swelling and excessive secretions. As has been the trend with other centers in the world,[11] an echocardiogram became the current investigation of choice at our institution to rule out intracardiac anomalies. Cardiac catheterization has not been performed since 1996. Owing to frequent association of PA sling with tracheal abnormalities,[2,11] others have recommended routine preoperative investigations for tracheal stenosis.[13,14] Similarly, we perform preoperative tracheal evaluation with CT, MRI, bronchoscopy, or bronchogram. Although not all patients with PA sling require tracheal surgery, failure to diagnose significant tracheal stenosis could be detrimental. At our institution, selection of patients requiring tracheal surgery was based on difficulty with ventilation and/or inability to wean off mechanical ventilation.

TABLE 2. Mortality by era

	1984-1993	1994-2003	2004-2011
Operative mortality	22.2%; 2/9	14.3%; 1/7	0%; 0/5
Late mortality	14.3%; 1/7	0%; 0/6	0%; 0/5

Overall operative mortality in this study was 14.3%. However, it was reduced to 0% in the last 6 consecutive patients, despite all of them undergoing tracheoplasty (Table 2). The higher operative mortality between 1984 and 2004 in our patients was similar to that reported by Fiore and associates[11]—operative mortality of 14% (2/14) in 14 patients with PA sling who were operated on between 1983 and 2003 (8 patients had tracheoplasty). Both operative deaths occurred in patients who had patch tracheoplasty. The remaining 6 patients (slide tracheoplasty, n = 2; patch tracheoplasty, n = 4) were discharged from the hospital.[11] Oshima and colleagues[12] reported operative mortality of 6.5% (2/31) in 31 patients with PA sling operated on between 1984 and 2006 (28 patients had tracheoplasty). They concluded that if tracheomalacia involved the carina, the patients had worse outcomes. All patients in their study[12] had slide tracheoplasty, end-to-end anastomosis, or costal cartilage graft and no patch tracheoplasty. Mortality in our patients operated on before 2004 was also determined by the requirement of tracheal surgery. The patients who required tracheal surgery had longer duration of CPB and longer hospital stay.

Since the first reports of tracheal stenosis repair in 1982,[15,16] various tracheal repair techniques have been proposed. Over the past decade, slide tracheoplasty, as described by Tsang and coworkers,[7] has emerged as the technique of choice.[6,12,13,17-19] This technique is designed to avoid the use of devascularized graft material and allows immediate stability of the trachea, thus reducing ventilation time and excessive granulation formation.[20] In high-volume centers, survival outcomes after slide tracheoplasty are excellent.[6] Manning and colleagues[6] reported an

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operative mortality of 2.5% (2/80) after slide tracheoplasty in 80 patients operated on between 2001 and 2009. They concluded that slide tracheoplasty with CPB can be performed with low mortality in the pediatric population. In our experience, mortality can be reduced with a multidisciplinary team approach, preferential use of slide tracheoplasty, and the abandonment of patch tracheoplasty. With such an approach we were able to eliminate mortality since 2004. Recently, Backer and coworkers[13] reported excellent results with no operative mortality in 34 patients, 26 of whom underwent tracheoplasty. Thus, low mortality can be achieved in patients with PA sling who require tracheal surgery. With extensive slide tracheoplasty, however, underlying tracheobronchomalacia may result in carinal collapse.[12] Manning and associates[6] described using slide tracheoplasty and temporary stenting to stabilize the carina. To deal with this problem, we have introduced a technique whereby the carina is stabilized by placing external stents.[8]

With tracheal repair using patch material, postoperative patch collapse often leads to increased duration of ventilation and subsequent development of excessive granulation tissue requiring bronchoscopic intervention.[5] Three patients in our study died of respiratory failure owing to persistent patch collapse. The complication of excessive granulations has been described in the literature,[11] and Manning and colleagues[6,20] identified prolonged ventilator support as a risk factor for excessive growth of granulation tissues. Even after introduction of the slide tracheoplasty technique, excessive granulation tissue occurred in 2 of our patients. This, presumably, occurred because of prolonged preoperative ventilation in 1 patient and small pericardial patch used in 1 patient to enlarge the carina.

In 2010, Goldstein and colleagues[21] noted the high incidence of PA stenosis at the reimplantation site in both PA sling and anomalous origin of one PA from the ascending aorta. They reported that 45% of their patients required an intervention for PA stenosis.[21] In contrast, only 1 (4.8%) patient in our series had left PA stenosis. Other reports have similarly described good patency rates.[10,12,13] We implanted the left PA proximal to the main pulmonary trunk, immediately above the sinotubular junction. This allowed adequate anastomostic size and prevented kinking of the left PA.

Late mortality was 5.6%. Our experience compares favorably with the literature. Oshima and colleagues[12] (median follow-up, 4.6 years) and Fiore and associates[11] (mean follow-up, 3.5 years) reported late mortality of 10.3% (3/29; all after tracheoplasty owing to residual tracheal stenosis) and 8.3% (1/12; after patch tracheoplasty), respectively. The recent study by Backer and colleagues[13] reported late mortality of 11.7% (4/34; 2 deaths were after patch tracheoplasty). The only late death in our study occurred 8 months after surgery in a patient with previous patch tracheoplasty. It appears that late mortality may be determined by patch tracheoplasty.

Fiore and coworkers[11] noted that mortality was highest for patients operated on in infancy. Likewise, 75% of deaths in our series occurred in children operated on in infancy. The encouraging observation was that survival beyond 1 year after PA sling and tracheal repair offered good outcomes, at least into early adulthood. At the most recent follow-up, all patients were asymptomatic. It appears that despite extensive airway surgery, there was adequate growth of the trachea and the long-term respiratory complications were rare.

In our study, all patients with PA sling alone, without airway surgery, have had an excellent outcome. Concomitant repair of coexisting cardiac anomalies was performed with no added mortality.

Limitations

The study is a retrospective review. Perioperative techniques have varied during the study period. Statistical analysis was limited owing to small sample size.

CONCLUSIONS

Patients with the diagnosis of PA sling require preoperative evaluation of the trachea. Children with PA sling who do not require tracheal surgery have excellent outcomes. Mortality was determined by the need for tracheal surgery. However, mortality can be reduced with the slide tracheoplasty technique and multidisciplinary team approach. Survival beyond 1 year after surgery offers an excellent prognosis.

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