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ОРИГІНАЛЬНІ ДОСЛІДЖЕННЯ

Pulmonary artery sling: diagnosis and surgical treatment

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The aim — to study the experience in surgical treatment of isolated and associated pulmonary artery sling including tracheal, cardiac and aortic arch anomalies, to analyze the treatment strategy, complications and their correction.

Materials and methods. Esophagography, digital subtraction angiography, contrast chest CT, tracheobronchoscopy, echocardiography were used for diagnosis. Surgery was used in all cases despite patient's age, severity and associated lesions.

Results and discussion. Over the 35-year period (1982—2017), 93 children were diagnosed and operated with different types of vascular rings in one center. Most of patients were under 3 year of age (n = 75; 80.6 %). Total of 8 (8.6 %) patients having pulmonary artery sling were included to the study. One of them had double vascular ring which included pulmonary artery sling and double aortic arch. Patients' age at operation ranged from 21 days to 24 months (mean 12.9 ± 2.3, median 12.5), and weight ranged from 3.7 kg to 12.5 kg (mean 9.6 ± 0.9 kg, median 9.3 kg). Four of them (group I) (50 %) had isolated pulmonary artery sling and other 4 had associated airway or cardiac malformations (group II). Associated anomalies were as following: complete rings tracheal stenosis (n = 4), ventricular septal defect (n = 1), double vascular ring and total anomalous venous drainage (n = 1). Isolated pulmonary artery sling was corrected through left thoracotomy without cardiopulmonary bypass. Vascular anastomosis was performed using «fish mouth» technique to prevent postoperative stenosis. Median sternotomy and bypass were used in case of associated tracheal stenosis and ventricular septal defect. Tracheal resection or slide tracheoplasty were used for tracheal stenosis. In patient with double vascular ring and anomalous venous drainage surgery was staged: Rashkind procedure; then division of double aortic arch and left pulmonary artery reimplantation. Survival rate in the group I was 100 %, whereas in the group II 3 patients died, including that with double vascular ring. Overall mortality was 37.5 %. Stenosis of vascular anastomosis was corrected by balloon dilation in one case.

Conclusions. Isolated pulmonary artery sling could be corrected through left thoracotomy without cardiopulmonary bypass, whereas associated anomalies should be managed using sternotomy approach and bypass. «Fish mouth» vascular anastomosis is effective in prevention of its stenosis. Balloon dilation of stenotic anastomosis in late period allows restoring an adequate blood flow in the left lung. Associated tracheal stenosis and cardiac defects has significant influence to the increase of mortality in patients with pulmonary artery sling. In case of associated heart defects simultaneous repair of cardiac hemodynamics and decompression of respiratory tract is expedient.

Key words: vascular ring, pulmonary sling, tracheal compression, tracheal stenosis, double vascular ring.

Vascular rings are congenital anomalies of great vessels that encircle and compress the trachea and esophagus or both [7]. Although pulmonary artery (PA) sling is not complete anatomic «ring» it is classified with the classic vascular rings because of the similarities in patient presentation, diagnosis, and surgical treatment [5]. PA sling is a very rare life-threatening vascular ring anomaly with complex anatomy, which is characterized by abnormal development of pulmonary trunk, aberrant origin, hypoplasia and pathological course of the left PA, and compression or anatomic stenosis of the trachea. Abnormal left PA originates from the right PA, turns over the right main bronchus to the top and back, circumflexes the distal trachea and then enters the left lung hilum under the aortic arch and behind the ductal ligament [7, 9, 11, 22]. This vascular sling causes compression of the trachea and the right main bronchus. Decreasing of the volume blood flow and severe respiratory insufficiency are typical pathophysiologic criteria of PA sling. About 58—83 % of patients have associated anomalies, in particular, heart disease are found in 50 % of cases [26, 29] and up to 65 % have tracheal stenosis (complete cartilaginous

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rings) [7, 9, 14]. Those comorbidities significantly deteriorate the course and prognosis, even with timely and adequate surgical treatment [9, 17, 20]. Mortality rate reaches 90 % in the case of a natural course of the defect by the age of 6 months [10], and even with the surgical treatment the mortality in the last two decades was 5—45 % [4, 6, 8, 18, 26, 28, 29] and it is associated with tracheal surgery in most cases [29]. Current controversies in the treatment of PA sling are related to surgical access, the use or non-use of cardiopulmonary bypass (CPB), the method of reconstruction of the pulmonary artery and the methods of tracheal reconstruction [4]. Postoperative complications may include tracheal obstruction due to granulation tissue, restenosis or tracheomalacia following tracheoplasty, stenosis or obstruction of vascular anastomosis and diaphragm relaxation, resulting in disorders of ventilation or perfusion, and worsen prognosis [9, 14, 19, 25].

The aim — to study the experience in surgical treatment of isolated and associated pulmonary artery sling including tracheal, cardiac and aortic arch anomalies, to analyze the treatment strategy, complications and their correction.

Materials and methods

Over the 35-year period (1982—2013), 93 children with different types of vascular rings were diagnosed and operated in one center. Distribution of types of vascular ring and age of patients is demonstrated in Table. Most of patients were aged under 3 years (n = 75; 80.6 %).

Eight (8.6 %) patients with PA sling were included to the study. One of them had double vascular ring (DVR) since PA sling was one of the component of the DVR along with double aortic arch. Their age at operation ranged from 20 days to 24 months (mean 12.9 ± 2.3, median 12.5), and patients' weight ranged from 3.7 kg to 12.5 kg (mean 9.6 ± 0.9 kg, median 9.3 kg). There were 5 boys and 3 girls, ratio 1.7 : 1. Four of them (group I) (50 %) had isolated PA sling and other 4 had associated airway or cardiac malformations (group II). Two patients had association of PA sling with complete tracheal rings stenosis only, one additionally had ventricular septal defect (VSD) and another one newborn baby had a unique association consisted of PA sling and double aortic arch forming double vascular ring along with intracardial total anomalous pulmonary venous drainage (TAPVD) and complete tracheal rings. Long segment complete ring tracheal stenosis was observed in 2 of these patients.

Plain chest X-ray, esophagography, digital subtraction angiography (DSA), contrast chest computed tomography (CT), tracheobronchoscopy, echocardiography were used for diagnosis. If PA sling was diagnosed the surgery was indicated despite patient's age, severity and associated lesions.

Method of surgical correction depended on the variant of the malformation. In the cases of isolated PA reimplantation of left PA and division of ductal ligament were performed through left thoracotomy without CPB. However, CPB machine should be on standby since partial clamping of pulmonary trunk for anastomosis may result in cardiac arrest. Patients with associated congenital tracheal stenosis had reimplantation of the left PA and tracheal reconstruction surgery through median sternotomy with use of CPB. In the case of associated VSD the procedure was supplemented with transatrial VSD closure at the time of cardioplegia. The unique newborn with DVR+TAPDV initially on the 1st day of life underwent balloon atrioseptostomy (Rashkind procedure) to stabilize hemodynamics and then, on day 21 double ring correction (division of patent ductus arteriosus, division of left patent aortic arch and reimplantation of left PA) was performed through the left lateral thoracotomy.

Surgery for isolated PA sling includes a dissection of abnormal left PA and cutting it off the abnormal origin site, moving the divided left PA from behind the trachea and anastomosing it with main PA inside the pericardium at the probable place of normal bifurcation. Deep tracheal intubation is necessary to prevent tracheal kinking and obstruction during the left PA dissection. The access is left lateral thoracotomy through 4th interspace and it should be wide enough to ensure adequate access to anterior surface of the

Table

Type of vascular ring and age of the patients

Type of vascular ring	Patients age, years				Total
	< 1	1—3	4—7	8—17	
Double aortic arch	20	15	1	3	39 (41.9 %)
Right aortic arch with left ductal ligament/ductus	4	4	2	4	14 (15.1 %)
Aberrant right subclavian artery	3	4	2	4	13 (14.0 %)
Brachiocephalic trunk anomalies	13	4	–	2	19 (20.4 %)
Pulmonary artery sling	2	5	–	–	7 (7.5 %)
Double vascular ring	1	–	–	–	1 (1.1 %)
Total	43 (46.2 %)	32 (34.4 %)	5 (5.4 %)	13 (14.0 %)	93 (100 %)

heart and posterior mediastinum. Mediastinal pleura is opened between *n. vagus* and *n. phrenicus* and descending aorta is dissected. Ductal ligament or patent ductus arteriosus is divided between two purse-string sutures placed on its aortal and pulmonary ends. The recurrent laryngeal nerve is identified and preserved. The division of the ductal ligament facilitates the exposure of PA sling, decompresses PA and improves the conditions for reimplantation. Left PA is dissected thoroughly within the left lung hilum and then, after moving down of aortic arch, it is dissected away from the aorta, posterior trachea and anterior esophagus and identified originated from the superior aspect of the right PA. The pericardium is opened widely in front of the phrenic nerve. Test of partial main PA occlusion helps to assess the possibility of performing the procedure off-pump. Satinsky or Cooley vascular clamp is placed on the left anterior aspect of main PA by half of its diameter between valve's ring and ductal ligament stump. Stable heart rate and arterial pressure allows performing anastomosis without CPB. The dissected left PA is ligated deeply in the mediastinum at the level of the right tracheal aspect and cut off from the site of its origin on right PA to form a small proximal stump. Decompression of trachea should be carried out as soon as possible to prevent the hypoxia and bradycardia. Divided left PA is irrigated with heparin solution. Left anterior aspect of main PA is clamped again as mentioned above. If heart functions remain stable left PA is anastomosed into the pulmonary trunk in end-to-side manner utilizing running 7—0 polypropylene suture. The left PA is often lesser in diameter than the right one; hence the anastomosis is configured in end to side «fish mouth» manner to enlarge its diameter [1]. The pericardium closes with interrupted sutures. Patient heparinization during the operation and after it is not mandatory. The tube is placed into the pleural space only.

If concomitant tracheal and cardiac anomalies have to be repaired simultaneously median sternotomy is performed. The thymus is removed, and pericardium is opened. The aorta and pulmonary artery are separated from each other and ductal ligament is ligated and divided. CPB is instituted using bicaval and aortic canulation. The child is cooled slowly to 32 °C. In case of associated VSD the defect was sutured through right atriotomy after custadiol cardiac arrest. After resumption of heart beating the left PA is dissected away from aorta, trachea and esophagus and transected at its origin on superior aspect of right PA using a partial occluding clamp. The opening on right PA is closed with running 7—0 polypropylene sutures. The transected left PA is moved from behind the trachea, trimmed to prevent kinking and sewn to the main PA using partial occluding clamp.

If the tracheal repair is necessary the area of tracheal stenosis has to be fully exposed. Care should be taken to avoid damaging the lateral blood supply, especially along the distal half of the trachea. Considering that outer size of trachea does not correspond exactly with the inner diameter of stenosis intraoperative tracheoscopy is necessary to define the limits of stenotic area. Slide tracheoplasty should be performed if the length of stenosis is 1/2 or more of trachea [12, 23]. Short segment tracheal stenosis should be corrected with segmental tracheal resection. Tracheal anastomosis is performed with interrupted 5—0 polydioxanone sutures. Postoperative care includes antibiotics and, prolonged tracheal intubation.

All survived patients had follow-up investigation in late postoperative period from 2 to 25 years. Physical investigation, chest X-ray, echocardiography were routinely used for follow-up in all patients. Tracheobronchoscopy was used for assessment of operated airway. Late postoperative DSA was used to control the patency of vascular anastomosis in one patient along with the control of the effectiveness of its balloon dilation. To estimate the influence of associated congenital anomalies on survival rate for patients with PA sling we compared survival rates in patients with isolated PA sling (group I, n = 4) and in patients having associated tracheal or cardiac anomalies (group II, n = 4).

Results

All patients with PA sling and associations had respiratory symptoms since first days of life. Those included stridor, respiratory distress, and recurrent airway infection. Associated cardiac anomalies also presented with cyanosis. The patient with associated congenital tracheal stenosis had several episodes of respiratory arrest requiring tracheal intubation. The DVR + TAPVD patient had severe respiratory and hemodynamic disorders since 20 hours after birth and required tracheal intubation and ventilation.

Chest radiographs showed obstructive emphysema (n = 2), atelectasis of the left lung (n = 1), signs of inflammation (n = 4).

Esophagography was the first diagnostic study in 3 early patients having isolated PA sling. The lateral view showed semioval indentation of anterior wall of esophagus (Fig. 1) — Wittenborg sign [10]. The diagnosis was confirmed then by DSA or chest CT.

Selective pulmonary DSA was performed in 4 of 8 our patients. It was 100 % diagnostic for PA sling. It shows the origin and course of left PA (Fig. 2), allows assessing perfusion of the lungs and the patency of anastomosis, and to compare the lung perfusion in late postoperative period.

Contrast chest CT with 3D reconstruction was performed in 3 patients. It allows delineate vascular and tracheal anatomy and confirm the diagnosis of PA sling and associated anomalies (Fig. 3).

Tracheobronchoscopy was the first diagnostic study for «ring-sling» complex and performed preoperatively to rule out the type, degree and extent of

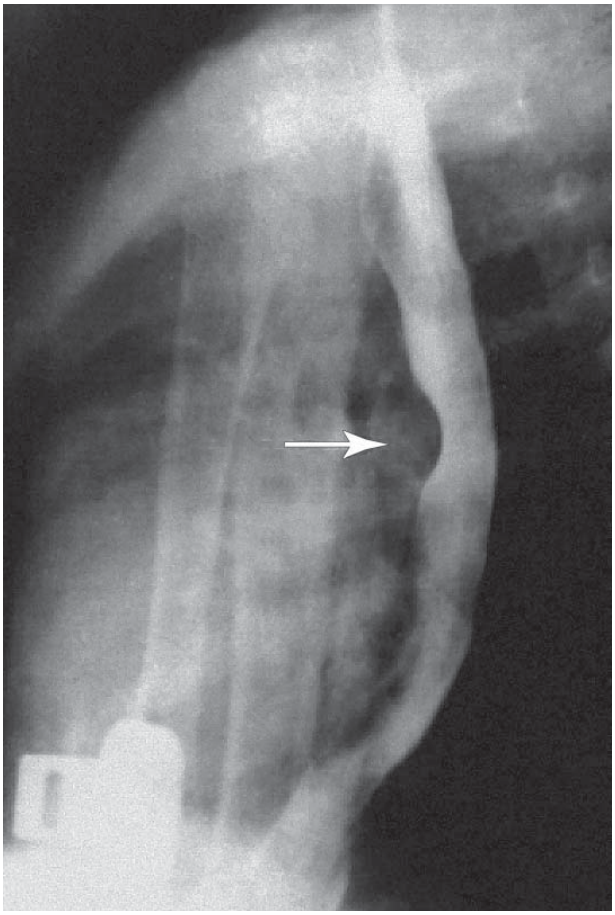

Fig. 1. Esophagography in lateral view in patient L. with pulmonary artery sling, age 2 years. Compression deformity of the anterior oesophageal wall (arrow)

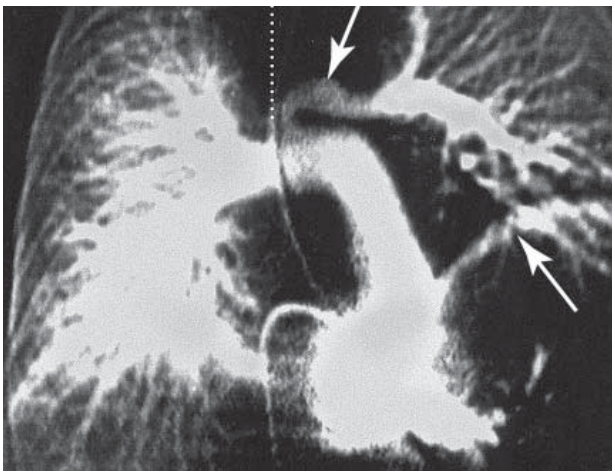

Fig. 2. Digital subtraction angiography in the same patient. The left pulmonary artery (arrow) branches from the right one, has a lesser diameter, and forms the sling on its way to the left lung hilum

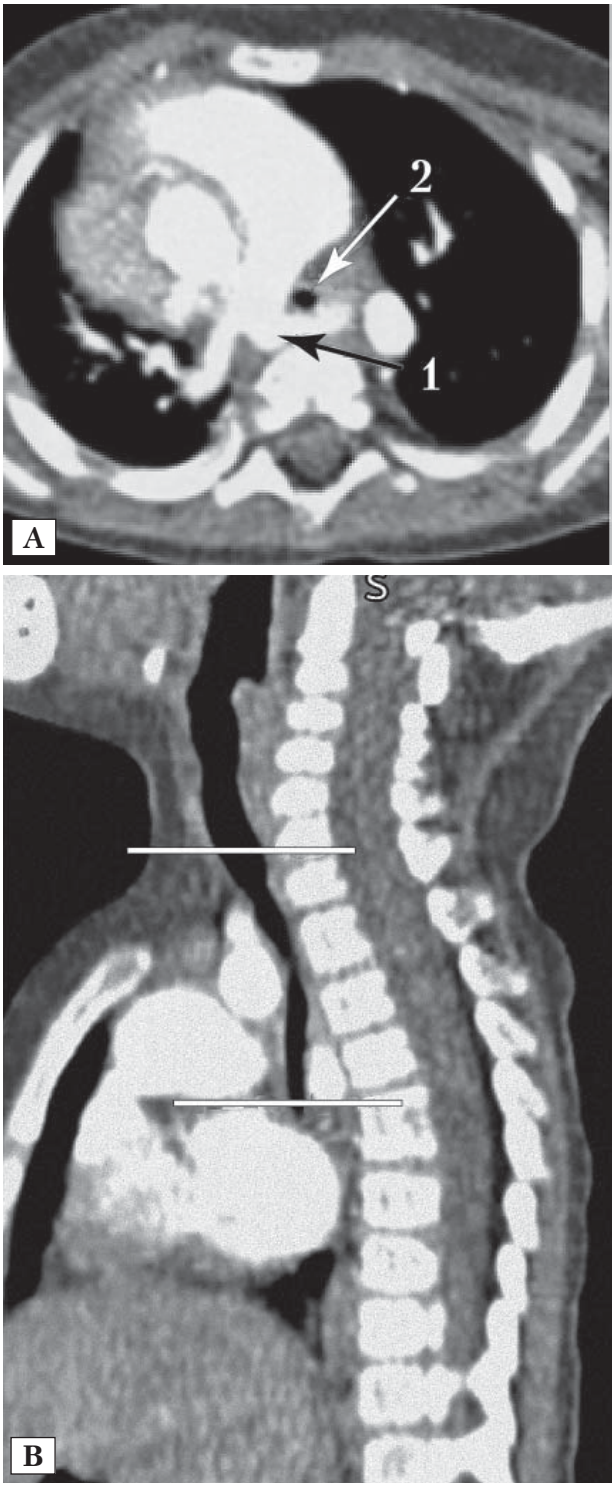

Fig. 3. Chest CT with contrast enhancement in 8 months old patient with pulmonary artery sling associated with complete ring long segment tracheal stenosis. Axial scan (A). Pulmonary artery sling (1), congenital stenosis of the thoracic segment of the trachea (2). Chest CT, sagittal reconstruction (B). Long segment congenital stenosis of the thoracic trachea (length 4 cm (83 %), the degree of narrowing 60 %)

stenosis. In isolated PA sling patients the asymmetric and slightly pulsatile elastic narrowing of the tracheal lumen 60—70 % just above bifurcation or at the right main stem bronchus orifice was registered. The zone of compression stenosis can easily be passed by the rigid bronchoscope. In complete cartilaginous ring cases tracheoscopy reveals rigid circular stenosis (up to 65 %) and absent membranaceous wall (Fig. 4). Rigid mucosal edema could be seen in case of DVR + TAPVD.

Echocardiography is considered to be optimal non-invasive diagnostic study at present time [4, 8, 14]. PA has no bifurcation and takes up incorrect position in all patients with PA sling. However, the visualization can be difficult due to lung emphysema. The intracardiac anatomy and hemodynamic was assessed in patients with associated cardiac anomalies, such as VSD or TAPVD. Doppler ECHO is useful and non-invasive method in follow-up examination to define the hemodynamic parameters in the zone of reimplanted PA and to assess the diameter and patency of anastomosis.

Operative mortality and intraoperative complications were not observed in both groups I and II. All patients with isolated PA sling demonstrated disappearance of respiratory symptoms in a few days after surgery; there were no postoperative complications in near period. All isolated left PA sling patients were extubated on the 1st day after surgery. Mean postoperative hospital stay in those patients was 12.0 ± 0.9 days. One late complication in the group I was the stenosis of vascular anastomosis which was revealed at DSA 2 months after surgery. Balloon dilation was performed resulting in normalization of anastomotic patency with left lung perfusion up to 99 % and 85 % confirmed by DSA in 2 and 10 years postoperatively. In patients of group II with associated malformations only one patient having PA sling associated with tracheal stenosis and VSD survived the simultaneous repair (left PA reimplantation, transatrial VSD closure and slide tracheoplasty) through median sternotomy and with CPB. She was extubated 10 days after surgery and discharged on 26th postoperative day with no respiratory or hemodynamic symptoms. Follow-up tracheobronchoscopy showed normal tracheal patency in this patient in 6 and 18 month postoperatively. All patients were under follow-up observation over a period 2—25 years. All of them have normal physical condition with respiration's compensation and good life quality. 3 (37.5 %) early postoperative deaths were registered. One of them was patient with DVR + TAPVD. The child died due to respiratory, hemodynamic and renal failure on 11th day postoperatively before the surgical correction of TAPDV could be undertaken. Another 2 patients with PA sling associated with complete ring tracheal stenosis died in early postoperative period due to necrosis of tracheal mucosa and acute airway obstruction. Comparing the survival rate in both groups of patients (group I — 100 %, n = 4; group II — 25 %, n = 1; p = 0.003) we confirmed that associated tracheal and cardiac malformations have significant influence on postoperative course and survival rate.

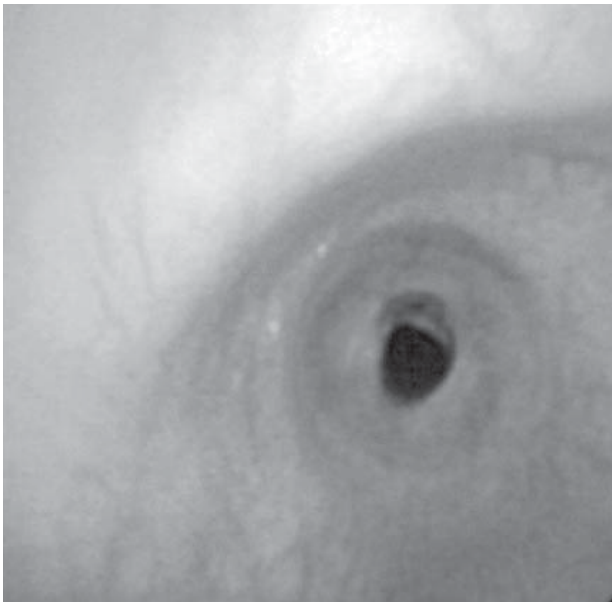

Fig. 4. Endophotograph in a female infant N., aged 15 months having congenital tracheal stenosis associated with pulmonary artery sling («ring-sling» complex). Zone of stenosis: complete rings, absent membrane

Case report 1

Male neonate was born at 40 weeks by normal labor with body mass 3700 g and length 52 cm from prima gravida. Apgar score at delivery was 8/9. Deterioration occurred in 20 hours after birth with an appearance of respiratory failure and weak heart sounds. Oxygen test was negative. Congenital heart disease had been suspected; newborn was intubated. Echocardiography on the 1st day revealed the collector of pulmonary veins behind left atrium, intracardial TAPVD, dilation of coronary sinus, obstruction of interatrial communication, decrease of left ventricle and atrium, dilation of the right atrium and ventricle and hypertrophy of right ventricular wall. DSA on 2nd day showed DVR consisted of double aortic arch with right arch dominated and PA sling, patent ductus arteriosus (PDA), and intracardial type of TAPVD. Rashkind balloon atrioseptostomy was performed to compensate intracardiac hemodynamics. Tracheobronchoscopy revealed tracheal stenosis 1.5 cm below glottis with minimal diameter of 2 mm. There was hard mucosal edema and hyperemia and the stenosis could not be passed by bronchoscope. Chest multispiral contrast enhanced CT confirmed the diagnosis of DVR consisted of double aortic arch with both patent aortic arches, left PA sling and PDA. Left PA was two times lesser than the right one (Fig. 5). All pulmonary veins merged to form common venous collector behind

diminished left atrium, the venous collector drained to dilated coronary sinus. Left lung was in atelectasis. Thoracic part of the trachea was narrowed with minimal diameter 0,1cm at the level of double aortic. Preoperative treatment included mechanical ventilation, antibiotics, dopamine, dobutamine and diuretics. SaO_2 was 65—75 %, blood gases showed hypercapnia (pCO_2 74—88 mm Hg) and hypoxemia (pO_2 32—51 mm Hg). The surgery for DVR correction was performed at the age of 21 days. The operation included division of patent ductus arteriosus, division of the left aortic arch, reimplantation of the left PA into the pulmonary trunk and posterior aortopexy through a left lateral thoracotomy. Decompression of trachea and esophagus was achieved with normalization of left lung ventilation, increase of SpO_2 up to 85—90 % and decrease of pCO_2 from 130 to 50 mm Hg. There were no intraoperative visual sings of congenital tracheal stenosis. The surgery for TADPV correction was postponed regarding the inflammation in the right lung. Postoperative period was complicated by pneumonia, recurrence of left lung atelectasis with respiratory instability and required prolonged mechanical ventilation. Tracheoscopy on the 10th postoperative day showed circular tracheal narrowing due to hard mucosal edema at the site of DVR. Mucus aspiration improved ventilation of the left lung and increased SpO_2 up to 95—100 %. However, hemodynamic and ventilation failures occurred because of redistribution in systemic and pulmonary perfusion, decrease of cardiac output, acute renal failure, and increase of tracheal stenosis due to mucosal edema and hypervolemia of pulmonary circuit. Baby died on 11th day after surgery. Autopsy confirmed the clinical diagnosis. Tracheal lumen was obstructed with edematous and thickened mucosa.

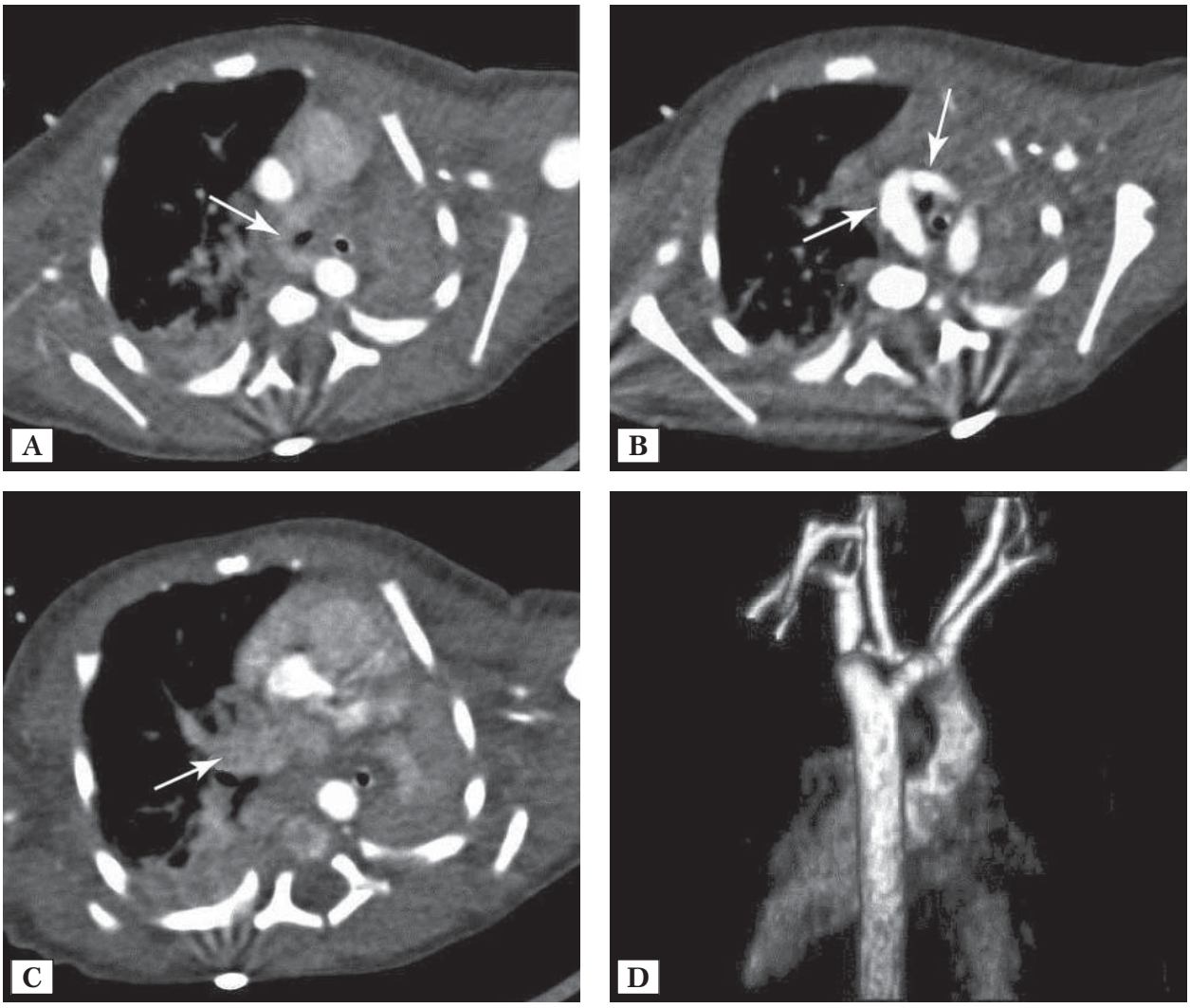

Fig. 5. Multispiral contrast enhanced chest CT in a newborn boy R., with double vascular ring, total anomalous pulmonary venous drainage and tracheal stenosis. A — Axial scan: left PA forms the sling around distal trachea (arrow), atelectasis of the left lung. B — Double aortic arch with both arches patent and dominating right arch (arrows). C — Venous collector that drains into the right atrium (arrow). D — 3D reconstruction demonstrating double aortic arch and separate brachiocephalic vessels (posterior view)

Case report 2

The 11 months old boy was admitted in the pediatric surgery department with stridor, dyspnea. He had history of recurrent bronchitis and pneumonia required four hospital admissions. Stridor appeared since birth. DSA revealed left PA sling. Left lung perfusion was 95 % comparing to the perfusion of the right lung. Tracheobronchoscopy revealed a 1.0 cm long extrinsic compression narrowing of the trachea by 50—60 % of its lumen localized just above the bifurcation. The mucosa was inflammated with purulent secretions. Esophagography showed the anterior indentation of the esophagus. The off-pump left PA reimplantation surgery through left lateral thoracotomy was performed on April 1989. Postoperative course was uneventful. Follow-up examination in 2 months after surgery showed that the left lung perfusion was decreased to 85 % comparing to the perfusion of the right lung, the diameter of anastomosis was significantly diminished up to 2 mm. Diameters of the main PA, right PA and left PA were 1.23 cm, 1.1 cm and 0.65 cm, respectively. Balloon dilatation of anastomosis was performed. DSA in 2 years after dilation showed the left lung perfusion about 99 % and anastomosis diameter 8 mm. Follow-up examination in 10 years after surgery revealed the 85 % of left lung perfusion compared to the right lung. Diameter of anastomosis was 8 mm, diameters of PA trunk, the right PA and the left PA were 1.9 cm, 1.3 cm and 1.0 cm, respectively (Fig. 6). Patient was doing well.

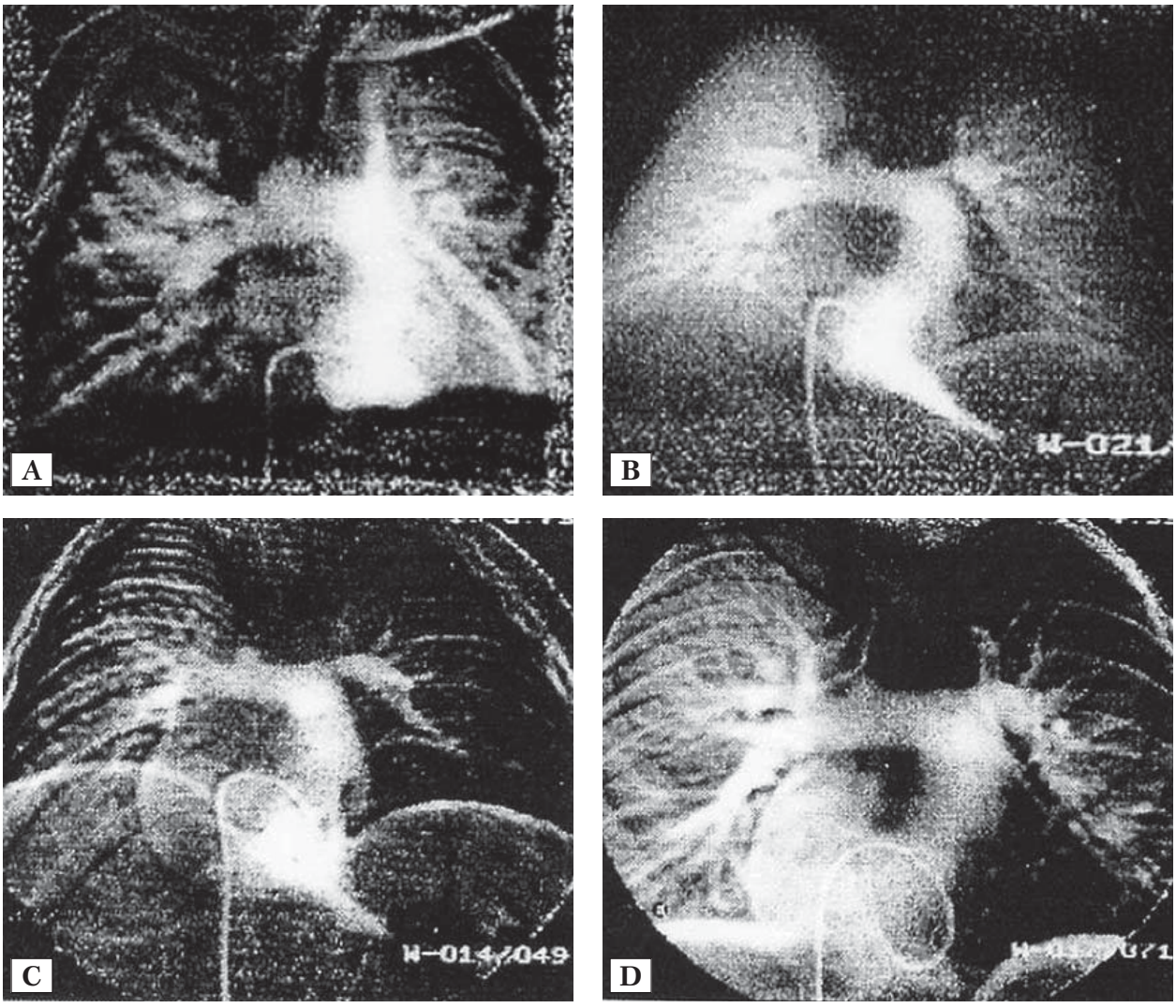

Fig. 6. Treatment of postoperative complication after reimplantation of the left pulmonary artery in patient L.
A — angiopulmonograhy in a 8-month-old patient before operation (the volume blood-flow ratio between the left and right lungs is 95 %). B — 3 months after the operation: stenosis of vascular anastomosis (the volume blood flow ratio is 85 %).
C — 3 years after the operation. Balloon dilatation of the stenosis was performed (the volume blood flow ratio is 99 %).
D — 10 years after the initial operation. Diameters of the vessels: pulmonary trunk 1.9 cm, right pulmonary artery 1.3 cm, left pulmonary artery 1.0 cm

Discussion

PA sling is known since 1897 when it has been described by Glaevecke and Doehle as an autopsy finding in 7-month-old child who died due to severe respiratory distress [11]. The first surgical correction was performed by W.J. Potts in Chicago in 1953. The operation consisted of left PA transection, translocation into ante-tracheal position and reanastomosis resulting in resolving of respiratory symptoms. However, late follow-up in clinically asymptomatic patient revealed occluded left PA [2, 22]. Since that prominent diagnosis was established and surgery was performed the era of intensive investigation of this anomaly had begun.

Comprehensive approach to treatment of PA sling is surgery. However, rare reports of therapeutic approach to PA sling management are also exist [21].

Current areas of controversy in the management of PA sling include surgical approach, use or non-use of CPB, and reimplantation versus translocation with distal tracheal resection [4, 14, 24]. A number of different surgical methods for concomitant tracheal stenosis repair were published: tracheal resection [13], pericardial tracheoplasty [15], chondral tracheoplasty [27], slide tracheoplasty [12, 23], using of tracheal homograft [16] and autograft [3].

Mortality in the case of a natural course of the defect reaches 90 % by the age of 6 months [10]. Associated anomalies significantly deteriorate the course and prognosis, even in cases of appropriate surgical treatment [9, 17, 20] and it is associated with tracheal surgery in most cases [28]. Simultaneous repair is advocated by the most authors [2, 4, 9, 14]. During the last 20 years the results of PA sling surgical treatment improved significantly with the decrease of postoperative mortality from more than 50 % in 1970th years [21] to the range of 5—45 % in the last two decades [4, 6, 8, 18, 26, 28, 29] with absence of mortality in early postoperative period and 5—10 % mortality in late postoperative period in some clinics [2].

C. L. Backer et al suggested surgical correction of PA sling through median sternotomy with CPB in 1985. It was considered that this method has some advantages like a simple administration of bypass, good access to PA and heart, sufficient time limits, and a possibility to perform a tracheoplasty in patients with concomitant congenital stenosis [2, 14].

We consider that it is possible to perform PA reimplantation without CPB in patients with isolated PA sling and a left thoracotomy is suitable for this. This approach allows reaching the left lung hilum, PA trunk and mobilizing the left PA deeply in the mediastinum up to its origin on right PA. In case of stable hemodynamic during a partial clamping of pulmonary trunk anastomosis of the left PA to pulmonary trunk is possible. Also this method is less expensive.

Associations of PA sling with tracheal and cardiac malformations are frequent and run up to 50—65 % [7, 14]. However, we could not find the information about a combination of PA sling, double aortic arch (double vascular ring), and TAPVD in relevant literature.

Our own experience (Prof. D. U. Krivchenya) includes diagnosis and surgical treatment of 93 patients with different types of vascular rings (see Table). But we had met the case of double vascular ring for the first time. Uniqueness of this case consisted of the combination of several malformations of great vessels and heart: double aortic arch and PA sling that formed double vascular ring with tracheoesophageal compression along with intracardiac type of TAPVD, patent ductus arteriosus and complete tracheal rings. Pathophysiology of hemodynamic failure, its severity due to hypervolemia of all thoracic organs with imminent edema of tracheal mucosa and secondary disturbances of respiration is obvious. Tracheal stenosis was formed with compression by double vascular ring and was increased due to tracheal intubation and repeated tracheobronchoscopies. Rashkind procedure was carried out urgently. It improved the baby's condition, decreased respiratory failure and gave the time to perform the diagnosis.

Surgical decompression of trachea by division of the double vascular ring with division of PDA and aortopexy provided the functional compensation during first week. Morphologic features of tracheal stenosis were evaluated during operation. Outer diameter of the trachea after decompression was similar to that of proximal trachea, hence, no visual signs of tracheal stenosis were found. That was confirmed by the immediate improve of respiratory parameters after decompression of the trachea. So, tracheal reconstruction was not considered to be indicated. Correction of TAPVD was necessary for normalizing of hemodynamic. But widening of incision and increase the time of surgery with CPB was considered of extreme risk and inexpedient. The decision was made to postpone this stage of repair. However, we consider that it was our mistake not to perform TAPVD repair on 7th postoperative day. Repeated tracheoscopy with passing the tube through the stenotic trachea should also be considered as ungrounded procedure that did not resulted in improving of tracheal patency but increased tracheal stenosis by posttraumatic mucosal edema. Autopsy showed that tracheal lumen was equal along all its length it was sufficient for a respiration. Tracheal stenosis requiring surgical correction was not found. Only surgery of cardiac malformation with translocation of pulmonary veins into the left atrium could prevent the increase of respiratory tract edema. Atrial opening after Rashkind procedure was insufficient (diameter 0.4 cm) for adequate hemodynamic.

Analysis of this unique case of complex congenital anomalies showed that similar combination of malformations should be corrected more intensively without delay and with simultaneous repair of intracardiac hemodynamics and airway decompression.

Anastomosis for left PA reimplantation should be performed by «fish mouth» type because the usual method may result in stenosis of anastomosis. Follow-up control of anastomosis patency allows performing balloon dilation timely if needed and thus maintain the adequate perfusion of the left lung.

Conclusions

Isolated pulmonary artery sling could be corrected through left thoracotomy without cardiopulmonary bypass, whereas associated anomalies should be managed using sternotomy and cardiopulmonary bypass. «Fish mouth» vascular anastomosis is effective in prevention of its stenosis. Balloon dilation of stenotic anastomosis in late period allows restoring an adequate blood flow in the left lung. Associated tracheal stenosis and cardiac defects have significant influence to the increase of mortality in patients with pulmonary artery sling. Analysis of unique case of total anomalous pulmonary venous drainage and double vascular ring suggested that simultaneous repair of cardiac hemodynamics and decompression of respiratory tract is expedient.

Conflicts of interest: none.

Authorship contributions: conception and design — D. K., E. R; acquisition of data, analysis and interpretation of data, drafting the article — E. R.; critical revision of the article — D. K.

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Петля легеневої артерії: діагностика та хірургічне лікування

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Мета роботи — вивчити досвід хірургічного лікування ізольованої та асоційованої з вадами трахеї і серця петлі легеневої артерії, проаналізувати лікувальну тактику, ускладнення та методи їх корекції.

Матеріали і методи. Для діагностики використано езофагографію, цифрову субтракційну ангіографію, трахеобронхоскопію та ехокардіографію. Хірургічне лікування застосовували в усіх випадках, незважаючи на вік пацієнтів та асоційовані вади.

Результати та обговорення. Протягом 35-річного періоду (1982—2017 рр.) 93 пацієнти з різними варіантами судинного кільця були прооперовані в одному центрі. Більшість пацієнтів були раннього віку (n = 75; 80,6 %). З них 8 (8,6 %), які мали петлю легеневої артерії, залучили в дослідження. Один з них мав подвійне судинне кільце, утворене петлею легеневої артерії та подвійною дугою аорти. Вік пацієнтів на момент операції становив від 21 дня до 24 місяців (у середньому (12,9 ± 2,3) доби, медіана 12,5), маса тіла — від 3,7 до 12,5 кг (у середньому (9,6 ± 0,9) кг, медіана 9,3 кг). Четверо пацієнтів мали ізольовану петлю легеневої артерії (перша група), інші четверо — асоційовані вади трахеї або серця (друга група). Асоційовані вади охоплювали повні хрящові кільця трахеї (n = 4), дефект міжшлуночкової перегородки (n = 1), подвійне судинне кільце й тотальний аномальний дренаж легеневих вен (n = 1). Ізольована петля легеневої артерії була коригована доступом через ліву торакотомію без штучного кровообігу. Судинний анастомоз формували за методикою «риб'ячого рота» для запобігання післяопераційному стенозуванню. У випадках асоційованого стенозу трахеї та септального дефекту серця корекцію виконували через стернотомію в умовах штучного кровообігу. Корекцію стенозу трахеї проводили шляхом резекції трахеї або ковзної трахеопластики. У пацієнта з подвійним судинним кільцем та аномальним дренажем легеневих вен корекцію проводили стадійно: процедура Рашкінда, потім роз'єднання подвійної дуги аорти і реімплантація легеневої артерії. Частота виживання в першій групі — 100 %, тоді як у другій групі троє пацієнтів померли, у тому числі пацієнт із подвійним судинним кільцем. Загальна летальність становила 37,5 %. Стеноз судинного анастомозу в одному випадку був коригований балонною дилатацією.

Висновки. Ізольована петля легеневої артерії може бути коригована доступом через ліву торакотомію без штучного кровообігу, тоді як у разі асоційованих вад корекцію слід виконувати через стернотомію в умовах штучного кровообігу. Судинний анастомоз за методикою «риб'ячого рота» ефективний у запобіганні стенозуванню. Балонна дилатація стенозованого анастомозу легеневої артерії дає змогу відновити кровообіг лівої легені. Асоційовані вади серця і трахеї істотно підвищують летальність у пацієнтів з петлею легеневої артерії. У разі асоційованих вад серця доцільна симультанна корекція внутрішньосерцевої гемодинаміки та декомпресія респіраторного тракту.

Ключові слова: судинне кільце, петля легеневої артерії, компресія трахеї, стеноз трахеї, подвійне судинне кільце.

Петля легочной артерии: диагностика и хирургическое лечение

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Цель работы — изучить опыт хирургического лечения изолированной и ассоциированной с пороками трахеи и сердца петли легочной артерии, проанализировать лечебную тактику, осложнения и методы их коррекции.

Материалы и методы. Для диагностики использованы эзофагография, цифровая субтракционная ангиография, трахеобронхоскопия и эхокардиография. Хирургическое лечение применяли во всех случаях, несмотря на возраст пациентов и сочетанные пороки.

Результаты и обсуждение. На протяжении 35-летнего периода (1982—2017 гг.) 93 пациента с различными вариантами сосудистого кольца были прооперированы в одном центре. Большинство пациентов (n = 75; 80,6 %) раннего возраста. Из них 8 (8,6 %), которые имели петлю легочной артерии, включены в исследование. У одного из них было двойное сосудистое кольцо, образованное петлей легочной артерии и двойной дугой аорты. Возраст пациентов на момент операции составлял от 21 суток до 24 месяцев (в среднем (12,9 ± 2,3) суток), масса тела — от 3,7 до 12,5 кг (в среднем (9,6 ± 0,9) кг). У четырех пациентов была изолированная петля легочной артерии (первая группа), еще четверо имели ассоциированные пороки трахеи или сердца (вторая группа). Ассоциированные пороки включали полные хрящевые кольца трахеи (n = 4), дефект межжелудочковой перегородки (n = 1), двойное сосудистое кольцо и тотальный аномальный дренаж легочных вен (n = 1). Изолированную петлю легочной артерии корригировали через левую торакотомию без искусственного кровообращения. Сосудистый анастомоз формировали по методике «рыбьего рта» для предупреждения послеоперационного стенозирования. В случаях ассоциированного стеноза трахеи и септального дефекта сердца коррекцию выполняли через стернотомию в условиях искусственного кровообращения. Коррекцию стеноза трахеи проводили путем резекции (n = 1) трахеи или скользящей трахеопластики (n = 2). У пациента с двойным сосудистым кольцом и аномальным дренажем легочных вен коррекцию проводили поэтапно: процедура Рашкинда, затем разделение двойной дуги аорты и реимплантация легочной артерии. Частота выживания в первой группе — 100 %, тогда как во второй группе три пациента умерли, включая пациента с двойным сосудистым кольцом; общая летальность составила 37,5 %. Стеноз сосудистого анастомоза в одном случае был корригирован баллонной дилатацией.

Выводы. Изолированная петля легочной артерии может быть корригирована доступом через левую торакотомию без искусственного кровообращения, тогда как в случае ассоциированных пороков коррекцию следует выполнять через стернотомию в условиях искусственного кровообращения. Сосудистый анастомоз по типу «рыбьего рта» является эффективным для предупреждения стенозирования. Баллонная дилатация стенозированного анастомоза легочной артерии позволяет возобновить кровоток левого легкого. Ассоциированные пороки сердца и трахеи существенно повышают летальность у пациентов с петлей легочной артерии. В случае ассоциированных пороков сердца целесообразна симультанная коррекция внутрисердечной гемодинамики и декомпрессия респираторного тракта.

Ключевые слова: сосудистое кольцо, петля легочной артерии, компрессия трахеи, стеноз трахеи, двойное сосудистое кольцо.