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SHORT-TERM AND LONG-TERM OUTCOMES OF ENDOVASCULAR VERSUS SURGICAL PALLIATION PROCEDURES FOR PULMONARY ARTERY ATRESIA WITH INTACT VENTRICULAR SEPTUM

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With the purpose to compare in-hospital and late outcomes of endovascular palliation, and surgical interventions in patients with pulmonary artery atresia 138 consecutive patients with the same pathology were analyzed since 2006 (115 patients underwent surgery – 160 surgical procedures, and 23 – underwent 32 endovascular procedures). Long-term outcomes studied on 12 years. The necessity for re-interventions following endovascular, and surgical procedures was analyzed. In surgical group mean length of hospital stay was 26±18.6 days, in endovascular group–22±5.7 days. Rate of surgical and endovascular procedures for pulmonary artery atresia Type I was 53.9 % and 56.5 %, respectively, whereas patients with pulmonary artery atresia Type II: 41.7 % vs.17.4 %, respectively ($p<0.001$). Patients with Types III and IV predominantly underwent endovascular procedures in 8.7 % and 17.4 %. Endovascular palliation follows with three-fold lower rate of fatal complications vs. surgical interventions: 4.3 % vs. 13 %. Endovascular interventions are choice of option for first stage palliation for infants with pulmonary artery atresia.

Key words: pulmonary artery atresia, endovascular interventions, surgical procedures.

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КОРОТКОСТРОКОВІ ТА ВІДДАЛЕНІ РЕЗУЛЬТАТИ ЕНДОВАСКУЛЯРНИХ І ХІРУРГІЧНИХ ПАЛІАТИВНИХ ПРОЦЕДУР ПРИ АТРЕЗІЇ ЛЕГЕНЕВОЇ АРТЕРІЇ З ІНТАКТНОЮ МІЖШЛУНОЧКОВОЮ ПЕРЕГОРОДКОЮ

З метою порівняння госпітальних та віддалених результатів ендоваскулярної паліативної допомоги та хірургічних втручань у пацієнтів з атрезією легеневої артерії було проаналізовано 138 пацієнтів з тією ж патологією з 2006 року (115 пацієнтів перенесли операцію – 160 хірургічних процедур, а 23–32 ендоваскулярні процедури). Віддалені результати вивчалися протягом 12 років. Було проаналізовано необхідність повторних втручань після ендоваскулярних та хірургічних процедур. У хірургічній групі середня тривалість перебування у стаціонарі становила 26±18,6 днів, в ендоваскулярній групі – 22±5,7 днів. Частота хірургічних та ендоваскулярних процедур при атрезії легеневої артерії I типу становила 53,9 % та 56,5 % відповідно, тоді як у пацієнтів з атрезією легеневої артерії II типу: 41,7 % проти 17,4 % відповідно ($p<0,001$). Ендоваскулярна паліативна терапія характеризується втричі нижчим рівнем фатальних ускладнень порівняно з хірургічними втручаннями: 4,3 % проти 13 %. Ендоваскулярні втручання є вибором варіанта першої стадії паліативної допомоги для немовлят з атрезією легеневої артерії.

Ключові слова: атрезія легеневої артерії, ендоваскулярні втручання, хірургічні процедури.

Pulmonary artery atresia (PAA) is a pathology which has various forms of association: with or intact interventricular septum [2, 8]. PAA with intact interventricular septum is ductus-dependent complex congenital heart disease associated with right-heart hypoplasia, considerable morphologic heterogeneity and poor outlook without in-time treatment. The size of the right ventricular cavity is proportional to the Z value of the diameter of the tricuspid valve.

Pulmonary atresia with intact interventricular septum accounts for 1–3 % in the structure of all critical congenital heart defects. PAA with intact interventricular septum has a high mortality rate in the natural course of the malformation. About 50 % of newborns die within the first two weeks of life; up to 85 % die before 6 months of life. The mortality in infants with ductus-dependent hemodynamics up to 12 months is 90 %. The mean survival rate of untreated patients with PAA ranges from 6 months to 2 years [2].

Patients with multiple sources of pulmonary blood flow, large aortopulmonary collaterals and moderate cyanosis, could survive to the third decade of life. There are few observations of patients with pulmonary atresia and intact interventricular septum surviving to 21 years of age with natural course of the malformation. In one such patient, reaching the age of 21 years of life was due to the provision of pulmonary circulation by a fistula of the right coronary artery with the pulmonary artery, lived to a similar age due to aorto-pulmonary window [6].

Fatal outcomes in the natural course of the malformation are due to severe systemic hypoxemia and metabolic acidosis, coinciding, as a rule, with spontaneous closure of the patent ductus arteriosus. One of the leading predictors of lethal outcomes are small size (Z-score) of the tricuspid valve annulus diameter and dependence of the right ventricle on coronary circulation. Some authors mark that the presence of

coronary artery fistulae, right ventricular dependence, or the tricuspid valvar Z-score did not prove to be risk factors for death [4, 11].

The treatment of such patients is multistage. To date, the tactics of surgical intervention in the first and subsequent stages of treatment of newborns and infants with pulmonary atresia and intact interventricular septum remain controversial. As a rule, indications for single or hybrid surgical interventions are based on the peculiarities of the malformation morphological structure in each individual case [7, 11].

Number of questions remains in the planning of cardiac surgery care for these patients caused by variety of anatomical features of PAA with intact ventricular septum; right ventricular chamber and tricuspid valve annulus dimensions; right ventricular dependence on coronary circulation; low reliability of predictive criteria for the effectiveness of intervention; necessity for early, and late re-interventions; absence of clear recommendations for choice of option following first intervention; poor evaluation of late changes in anatomical and hemodynamic parameters [1, 5, 10].

The purpose of the study was to evaluate the safety, and preferences of endovascular hemodynamic palliation for pulmonary artery atresia and intact ventricular septum in newborns and infants and to analyze predictors of early and late outcomes following endovascular interventions.

Materials and methods. The study was performed at the basis of Azerbaijan Medical University, (Baku, Azerbaijan), Amosov National Institute of Cardiovascular Surgery (Kyiv, Ukraine), National Pedagogical Dragomanov University (Kyiv, Ukraine) and Topchibashev Research Center of Surgery (Baku, Azerbaijan). 138 consecutive newborns and infants with PAA and intact ventricular septum underwent surgical (n=115), and endovascular intervention (n=23) since 2006 to 2022. Enrolled group included 81 (58.6 %) boys and 57 (41.3 %) girls. In 25 % of cases PAA was revealed prenatally; in 75 % – detected at birth.

Surgical group (n=115) included 62 (54 %) boys and 53 (46 %) girls with mean age at surgery 607.8±1023.7 days, and mean weight 8.8±8.1 kg. The length of hospital stay comprised 26±18.6 days. Endovascular group (n=23) included 18 (77 %) boys and 5 (23 %) girls with mean age at the first stage of treatment 220.5±650.2 days, and mean weight 5.0±4.4 kg. The duration of hospital stay after endovascular interventions was 22±5.7 days.

75 (54.34 %) patients presented with PAA Type I: atresia of the pulmionary artery (PA) valve with preservation of the pulmonary trunk, right and left branches of the PA; 52 (37.7 %) –with PAA Type II: atresia of the PA valve and the pulmonary trunk without changes in the right and left branches of the PA; 7(5.1 %) of patients presented with PAA Type III: atresia of the PA valve, pulmonary trunk, and both branches of the PA with ensuring pulmonary circulation by major aorto-pulmonary collateral arteries (MAPCA). PAA type IV revealed in 4 (2.9 %) patients: atresia of the PA valve, pulmonary trunk and one branch of the PA. Types of PAA were classified by J.Somerville.

All patients were separated depending of surgical, and endovascular approach. 115 (83.33 %) patients underwent surgery: 23 (12.6 %) – endovascular interventions.

Among patients who underwent surgery the distribution was as follows: Type I–62 (53.91 %); Type II–48 (41.73 %); Type III–5 (4.34 %). The same distribution for endovascular approach was as follows: Type I–13 (56.52 %); Type II–4 (17.39 %); Type III–2 (8.69 %) and Type IV–4 (17.39 %).

All patients received preoperative medical support from birth to the time of surgery (1–10 days) to correct systemic and pulmonary vascular resistance for adequate systemic perfusion (SO_2 up to 80 %). Prostaglandin E infusion was performed to prevent closure of the patent ductus arteriosus (PDA) and further development of cardiogenic shock.

Patients manifested with shock underwent correction of metabolic acidosis. The correction was carried out with extreme caution to avoid the threat of a decrease in pulmonary vascular resistance and, as a result, an increase in Qp/Qs. This reduces systemic circulation, and the “vicious circle” closes and exacerbates acidosis. Very slow infusion of alkaline solutions also helps to avoid brain haemorrhage.

A microbiological study was carried out, performing cultures for the existing microflora and determining its sensitivity to antibiotics. Antibiotic therapy was prescribed to prevent the development of infections before the results of the bacterial culture were obtained. Individual parenteral nutrition was prescribed to avoid peripheral circulatory disorders, including ischaemic and necrotic changes in the gastrointestinal tract.

The indications for perforation and balloon valvuloplasty (BV) of the PA were the presence of a mebranous form of atresia, hypoplasia of the right heart (Z-score of the tricuspid valve <–4; the presence of 2–3 anatomical parts of the right ventricle), and absence of right ventricular–dependent coronary blood flow. In patients with concomitant tricuspidal valve organic lesion (Ebstein-like anatomy or shortening one

of the leaflets), preference was given to surgical repair including plastic surgery of tricuspidal valve, and reduction of the dilated right ventricle.

Due to the unfavorable anatomy of the defect, it was not possible to restore antegrade blood flow in the PA by perforation of valve in 6 patients, and they underwent surgical valvuloplasty of the PA. In 23 cases (16.7 %) of 138 patients with PAA, balloon valvuloplasty was performed. The mean age at the time of endovascular treatment comprised 2±6 days (5 hours to 15 days); the mean body weight –3.4±0.52 (2.2–4.7) kg.

All patients required prostaglandin infusion at the preoperative stage. The level of SaO2 at the preoperative stage after prostaglandin infusion reached 78.1±10.4 (42–94 %). Nine patients (41.5 %) were hospitalized in severe condition with signs of decompensation of the defect, cardiac and respiratory failure. Seven (29.3 %) patients required ventilatory support due to respiratory failure and decompensated status. One patient had clinical and laboratory signs of pneumonia at the time of admission. Sympathomimetics were prescribed to 10 patients at the preoperative stage. Due to the impossibility of stabilizing the condition with medication, 3 patients underwent urgent surgery for vital reasons.

Statistical processing of all obtained data was carried out using descriptive statistics. For each group, the mean numerical value (M), standard deviation (σ2) of the mean numerical value and its standard error (m), as well as the minimum (min) and maximum (max) values of the series were determined. Differences were considered statistically significant at $p<0.05$. Statistical processing was carried out using the Statgraphics Centurion 18 (USA) program.

Results of the study and their discussion. In the treatment of PAA, a staged approach is used, correcting a part of the defect at each stage and thus reducing the trauma of the operation. In our case, the defect was corrected using traditional surgical methods and endovascular treatment.

The number of endovascular procedures was 32. In 14 (43.75 %) of patients pulmonary artery (PA) valve perforation 14 (43.75 %) was performed, in 5 (15.62 %)–major aorto-pulmonary collateral (MAPCA) Stenting, in 4 (12.5 %)–patent ductus arteriosus (PDA) Stenting. 9 (39.1 %) of 23 patients, required a concomitant Rashkind procedure. In more complex cases, it was advisable to perform surgical interventions.

Subsequently, 45 patients underwent the next stage of treatment, and the remaining 70 patients were discharged from the clinic to await the next stage. The following procedures were performed: Blalock-Taussig shunt–115 (71.9 %); Valve-containing conduit (PAD–ASD)–25 (15.6 %); Bidirectional Glenn–17 (10.6 %) and Fontan operation–3 (1.9 %). In one case, after surgical complications on the open heart, stenting of the PA branches was performed. The complications after endovascular and surgical interventions presented in Table 1.

Table 1

Complications in patients with PAA underwent surgical and endovascular interventions

Complications	Surgical group (n=115)	Endovascular group (n=23)	Total
Acute heart failure	9 (7.8 %)	3 (13.0 %)	12 (8.7 %)
Acute respiratory failure	5 (4.3 %)	2 (8.7 %)	7 (5.1 %)
Sepsis	5 (4.3 %)	0	5 (3.6 %)
Wound complications	6 (5.2 %)	0	6 (4.3 %)
Chylothorax	5 (4.3 %)	0	5 (3.6 %)
Femoral vein thrombosis	0	2 (8.7 %)	2 (1.4 %)
RVOT perforation	0	3 (13.0 %)	3 (2.2 %)
Total	30 (26 %)	10 (43.4 %)	40 (28.9 %)

Note: RVOT indicates right ventricular outflow tract.

Surgical interventions were performed when endovascular treatment was not possible. As a rule, clinical status of these patients was more severe. Open-heart surgery is a serious trauma for the patient, and therefore the postoperative period in such patients was more likely characterized by complications.

The mean length of hospital staying after surgical treatment comprised 26±18.6 days, and for endovascular treatment – 22.0±5.7 days ($p>0.1$). Thus, there was a significant reduction in the length of hospital stay after endovascular treatment, although NS. In 7 cases, the Rashkind procedure was additionally performed after open surgery. As a result of this additional surgical intervention, the patients' status improved and they were discharged in a satisfactory condition. Such complication as perforation of the right ventricular outflow tract by a coronary conduit (3 cases) was eliminated by immediate suturing of the perforation in the open heart.

Transient cardiac rhythm disturbances during the intervention were eliminated with the use of medications in 3 patients. All patients showed angiographic signs of pulmonary antegrade blood flow after

the procedure. During right ventriculography, contrast filling of the PA trunk and main branches was recorded. In all patients the systolic pressure in the RV decreased from initial 99.1±21.5 (58–129) mm Hg to 50.0±9.0 (35–75) mm Hg (p<0.001).

The residual gradient in the outflow tract of the RV after dilatation was 20±8.8 (10–47) mm Hg. No significant changes in end-diastolic pressure in the RV were noted. In addition to the criteria described above, fluoroscopic visualization of the balloon catheter, which was dilated at the level of the atresiated valve, was important. At the moment of balloon inflation, the rupture of the valve membrane could be clearly visualized. Despite the improvement of arterial SaO2 immediately after successful BV, in the postoperative period with the closure of the PDA, the SaO2 significantly decreased in all patients. On average, on the 3rd day after BV with the cessation of prostaglandin infusion and a decrease in blood flow through the PDA, a decrease in the SaO2 level was observed from 91.0±5.09 (70–99 %) immediately after dilatation to 70±18.7 (32–90 %) following procedure (Table 2).

Table 2

Results of pulmonary valve balloon perforation

Data	Before dilatation	After dilatation	P
SaO_2 (%)	75.1±11.5 (40–90)	91.0±5.09 (70–99)	<0.001
RV pressure (mm Hg)	99.1±21.5 (58–129)	50.0±9.0 (35–75)	<0.001
Systolic pressure gradient at RVOT (mm Hg)	–	20±8.8 (10–47)	–
RV EDV (mm Hg)	13.0±4.9 (5–27)	3.4±3.4 (1–10)	<0.001
Δp RA–LA (mm Hg)	4.2±0.8 (0–8)	1.9±0.6 (0–7)	

Note: RV indicates right ventricle; RA – right atrium, LA – left atrium, EDV – end-diastolic volume.

Such dynamics was associated with insufficient pulmonary antegrade blood flow in the setting of closure of the compensatory junction – the PDA.

The insufficient volume of blood flow through the PA valve can be explained by TV hypoplasia and insufficient volume, diastolic dysfunction and low compliance of the severely hypertrophied RV walls, which could not provide ejection into the PA to form sufficient antegrade pulmonary blood flow.

The mean intensive care units (ICU) staying following BV comprised 9±4 days (3 to 50 days). Five patients required sympathomimetic support in the postoperative period for an average of 8.0±4.7 (3–20) days, as well as ventilator support on 4.4±4.1 days (7 hours – 23 days). Fatal complications following endovascular procedures comprised 4.3 %.

Of the 14 patients who underwent BV, only 4 children (28.6 %) did not require re-intervention on 2 days to 12 years, mean 6 years. The remaining 10 patients underwent 16 surgical interventions.

In the early postoperative period, 5 children (37.8 %) required re-interventions. In 4 observations (28.6 %), the operation was aimed at increasing pulmonary blood flow by applying a systemic-pulmonary anastomosis on 5±2 (1–12) days. One patient underwent RVOT repair with concomitant TV repair.

Of the 4 patients after systemic-pulmonary shunt, 2 – underwent anastomosis closure and atrial septal defect repair; 1 patient required Blalock shunt closure with atrial septal closure in the long-term period at the age of 1 year, and 1 patient underwent bidirectional cavapulmonary anastomosis at the age of 6.8 years.

Five patients (37.8 %) required reintervention in the long-term period after BV. Of these, 4 – underwent repeat BV on 105±42 (40–370) days, and 1 patient underwent Atrial Septal Defect (ASD) repair with TV repair at 5 years.

In 2 patients, after repeated BV, surgical repair of the RVOT with TV plasticity was performed. In the early postoperative period, most of the patients (n=5) required stent implantation of PA, which accounted for 28.6 % of all reoperations in the first month after valvuloplasty. In the long-term period, repeated endovascular valvuloplasty was performed in 5 patients and surgical reconstruction of the RVOT with TV plasticity in 1 patient (Fig. 1).

The balloon valvuloplasty of the pulmonary artery was performed by us in 23 newborn patients. The average age of newborn patients at the time of endovascular intervention ranged from 5 hours to 14 days of life, making up 2 days, on average. The body weight of the operated patients ranged from 2.25–4.7 kg, amounting to 3.37±0.52 kg, on average.

After endovascular interventions, the hospital mortality rate in the group of newborns and infants with pulmonary artery atresia we analyzed was 4.3 %. At the same time, surgical interventions at the first stage of treatment of newborns with pulmonary artery atresia are accompanied by a rather high hospital and long-term mortality, with the need for early re-interventions.

Surgical treatment of patients with PAA is multistage and includes endovascular methods and surgical interventions. It yields good results with satisfactory PA size, adequate peripheral distribution, and

the absence of multiple aorto-pulmonary collateral arteries. Nevertheless, in most patients, there is a fairly pronounced hypoplasia of the PA system, local stenosis of the peripheral branches and significant additional sources of collateral pulmonary circulation, which complicates the treatment process. Therefore, such patients often have to undergo several complex surgical interventions. Recently, endovascular methods have become widespread. At the first stage, as a rule, the application of Rashkind procedure or reconstruction of the RVOT performed. These procedures increase SaO_2 by increasing the minute volume of the small circle of blood circulation and provide a hemodynamic effect (increased pressure in the PA promotes the growth and development of this vascular system).

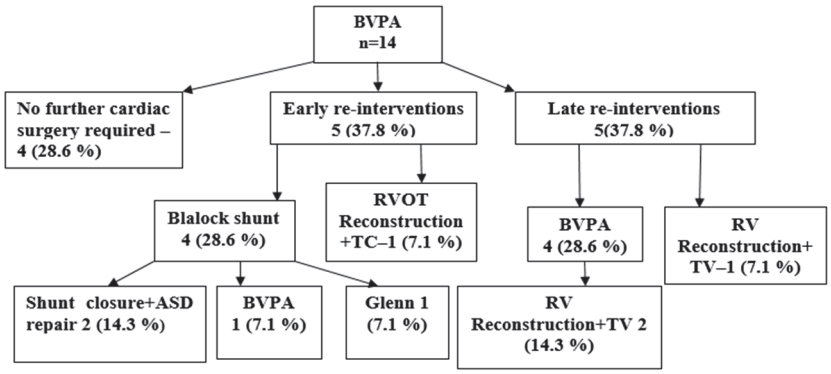

Fig. 1. Necessity for endovascular or surgical re-interventions following balloon valvuloplasty in patients with pulmonary artery atresia. BVPA indicates balloon valvuloplasty of pulmonart artery; ASD – atrial septal defect; RVOT – right ventricular outflow tract; TV – tricuspidal valve.

Vall Camell M, et al noted that patients with critical pulmonary stenosis had better outcomes compared to patients with pulmonary atresia with intact ventricular septum. In accordance with their data, the aggressive strategy of opening the pulmonary valve early on was associated with a good overall survival and correlated to a good functional class [9]. Wolter A, et al, with the purpose to analyze prenatal parameters predicting outcome in pulmonary atresia with intact ventricular septum/critical pulmonary stenosis (PAIVS/CPS) observed 82 foetuses from 01/08 to 10/18 in 3 centres in intervals 1 (<24 weeks), 2 (24–30 weeks) and 3 (>30 weeks). According to their results, 83.0 % survived, and the best single parameter for biventricular outcome was tricuspid/mitral valve (TV/MV) ratio. This ratio as simple parameter has high predictive value. But the study had some limitations, so, the authors emphasized, that the criteria for foetal intervention must further be evaluated [10].

Some studies related to treatment of patients with pulmonary artery atresia, showed that endovascular balloon valvulotomy of the pulmonary valve in newborns with pulmonary artery atresia is accompanied by an increased risk of re-interventions to increase pulmonary blood flow as compared with the patients who have initially undergone surgery [2, 7].

Loomba RS, et al, performing a retrospective study of patients who were assigned an aortic perfusion score based on the amount of antegrade perfusion to the four main coronary arteries. Various characteristics, including aortic perfusion score, were compared between those who required transplant or died during follow-up vs those who did not. The results showed that aortic perfusion index can be used to predict the composite endpoint of death or transplantation and may also be useful in selecting patients to be included in the transplant list [5].

Assuming that endovascular balloon valvulotomy suffers from limitations similar to performing an “open” valvulotomy or reconstructing the RVOT without concomitant systemic pulmonary anastomosis, some researchers also noted that performing an isolated balloon valvulotomy of the pulmonary valve in newborns with pulmonary artery atresia is fraught with a high risk of repeated interventions than in patients with initially performed surgery [3, 9].

Manhem S, et al conducted a systematic literature review on PA-IVS treatment. All neonates born in Sweden with PA-IVS between 2007 and 2019 were screened for inclusion. The literature review presented heterogeneity in standards for treatment. Their retrospective, population-based, multicenter study showed that both catheter-based intervention and cardiac surgery are safe procedures. The authors' results are comparable or superior to those of a systematic review of the literature. In summary, a systematic review of the literature shows great heterogeneity in study design, with no definitive gold standard for treatment [7].

Patients who initially underwent valvulotomy of the pulmonary valve with or without systemic pulmonary anastomosis often subsequently need reconstruction of the right ventricular outlet tract.

Multistage PAA surgery is designed to ‘rehabilitate’ hypoplastic true lungs by stimulating the growth and development of the true pulmonary bed by increasing blood flow through hypoplastic vessel. In the latter case, stenting of pulmonary branches, and reconstruction of the RVOT without closure of the ASD were used.

The main sources of blood supply to the lungs in PAA are the PDA, bronchial collateral arteries, large aortopulmonary collateral arteries, large mediastinal arteries and coronary pulmonary fistulas. The more alternative sources of blood supply to the lungs and the more pronounced their functional capacity, the lesser the degree of hypoxemia and clinical manifestations of the defect. However, in most patients, blood flow in the lungs is severely depleted due to stenosis of the collateral arteries and small diameter of the ICA.

The variants of the connection between the PDA and PA in PAA vary: in some cases, PA begin independently of the duct and connect to it as usual (Type I–II PAA), in others, the source of PA may be the duct itself (Type III PAA). In 2 % of cases of PAA with ASD, bilateral PDA was found [2].

Thus, pulmonary artery atresia is the serious condition, and any delays in emergency surgery for patients with PA are accompanied by progression of heart failure and increasing failure of other organs and systems, even to death, which was observed in our studies. Namely, the mortality rate is 13 % when using surgical methods of treatment.

Conclusions

1. Rate of surgical and endovascular procedures for pulmonary artery atresia Type I was 53.9 % and 56.5 %, respectively, whereas patients with pulmonary artery atresia Type II: 41.7 % vs.17.4 %, respectively ($p<0.001$). Patients with Types III and IV predominantly underwent endovascular procedures in 8.7 % and 17.4 %.

2. Endovascular palliation follows with three-fold lower rate of fatal complications vs. surgical interventions: 4.3 % vs. 13 %.

Pulmonary artery atresia, in addition to significant hemodynamic disorders, is also characterized by a significant depression of the physiological functions of other organs and systems with their subsequent insufficient blood supply. All this leads to a growing deterioration in the functional state and inevitable death of such patients in the absence of surgical treatment. Thus, endovascular procedures are effective choice of option for first stage palliation in newborns and infants with PAA, contributing to life support like a “bridge” to the next stage of repair.

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