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Transcatheter treatment of pediatric patients with pulmonary atresia

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Keypoints

To evaluate the safety and effectiveness of the method in infants with pulmonary atresia, to determine the factors influencing the immediate and long-term results of the intervention.

Abstract

Introduction

Treatment of patients with pulmonary atresia still remains a pressing and unresolved problem. The aim of the study is to evaluate the safety and effectiveness of the method in infants with pulmonary atresia, to determine the factors influencing the immediate and long-term results of the intervention.

Materials and methods

The paper presents an analysis of the results of endovascular interventions in 23 children with pulmonary atresia. Balloon valvuloplasty of the pulmonary artery was performed on 23 infants.

Results

In patients with pulmonary atresia, a step-by-step treatment was used, which included step-by-step endovascular correction of the elements of the defect, thereby reducing the trauma of the operation.

Conclusion

1. Endovascular surgical interventions in newborns and infants of the first year of life of patients with pulmonary atresia are the operations of choice, contributing to the life support of newborns and infants of the first year of life, allowing to prepare the patient for the next stage of surgical treatment.

2. Endovascular interventions are minimally invasive, which increases the chances of survival for patients with complicated anatomy, despite the severity of their condition upon admission to hospital.

Keywords

pulmonary atresia, balloon valvuloplasty, patent ductus arteriosus.

Introduction

Treatment of patients with pulmonary atresia still remains a pressing and unresolved problem. Considering the large variability of anatomical forms of pulmonary artery atresia, low reliability of precursors of prognostic criteria for the effectiveness of intervention, the need for re-intervention in the early and late postoperative period, the lack of clear recommendations for postoperative treatment and cardiac surgical tactics in the early and late periods after interventions, a small number of similar studies assessing changes in anatomical and hemodynamic parameters in the late period, there remain a large number of open questions in planning cardiac surgical care for such patients.

The aim of the study: to evaluate the safety and effectiveness of the method in infants with pulmonary atresia, to determine the factors influencing the immediate and long-term results of the intervention.

Materials and methods

Treatment of patients with pulmonary atresia using endovascular methods began in 2006. The paper presents an analysis of the results of endovascular interventions in 23 children with pulmonary artery atresia. In 25% of patients, who are related to more modern observations, the defect was diagnosed perinatally and confirmed by echocardiography immediately after birth. In the remaining cases, the pathology was identified at birth, which allowed the newborn to be immediately taken to the clinic for highly qualified medical care. Among children with pulmonary atresia, male patients predominated: 18 (77%) boys and five (23%) girls. The average age of children on the day of surgery was 220.5+650.2 days (from 1 to 698) days; body weight was 5.0+4.4 kg (from 1.851 to 6.055 kg). The length of hospital stay was 22+5.7 days. According to the classification of J. Somerville (1970) [9], 56.5% of our patients were diagnosed with type I pulmonary artery atresia, which was characterized by atresia of the pulmonary artery valve with preservation of the pulmonary trunk, right and left branches of the pulmonary artery. In four observations of type II defect, atresia of the pulmonary artery valve and pulmonary trunk were detected without changes in the right and left branches of the pulmonary artery. In four observations of type IV defect, atresia of the valve, trunk, and both branches of the pulmonary artery was verified, while the lungs were supplied with blood by large aortopulmonary collateral arteries. And only in two observations was type III defect diagnosed with atresia of the valve, trunk, and one branch of the pulmonary artery (Table 1).

Type of defect according to J. Somerville	Number of interventions n (%)
I	13 (56,5%)
II	4 (17,4%)
III	2 (8,7%)
IV	4 (%17,4)

Table 1. Frequency of endovascular interventions in patients with pulmonary atresia

In all patients in this group, the diagnosis of pulmonary artery atresia was confirmed by echocardiography data with a comprehensive assessment of the anatomical variants of the defect. At the preoperative stage, according to the echocardiography examination, the absolute dimensions of the interatrial junction ranged from 3.1 to 9.2 mm, with an average of 5.54±1.5 mm. Five patients showed signs of restriction of the atrial septal defect, accompanied by the presence of a pressure gradient between the left and right atria of more than 5 mm Hg, and a defect size of less than 5 mm (Table 2).

Indicator	Average value	SD	Minimum	Maximum
TV diameter (mm)	11	2,29	6	17
Z-score TV	-1,04	1,3	- 5,1	+1,4
Ratio of TV and MV	88,8	19,7	46	135
Pulmonary valve diameter (mm)	7,45	1,2	6	12
Z-score PV	-0,8	0,9	-2,5	+0,9
PV to AV ratio	87	156	59	129
Size DIS ДМIIII (mm)	5,54	1,5	3,1	9,2
Systolic pressure RV	99,7	19,7	60	125
EDP RV	13,7	4,9	7	26
Mean pressure gradient at the interatrial junction	4,5	0,8	2	8
Diastolic diameter RVOT	6,8	1,9	2,9	11
SpO_2	82	12	45	97

Table 2. Baseline echocardiographic and radiographic characteristics of patients with pulmonary atresia who underwent endovascular intervention.
Legend: TV – tricuspid valve; MV – mitral valve; PA – pulmonary artery valve; EDP – end-dystolic pressure; RV – right ventricle; RVOT – right ventricular outflow tract.

The tricuspid valve size ranged from 6 to 17 mm, averaging 11±2.29 mm; the average tricuspid valve Z-score was - 1.04±1.3 (- 5.1- +1.3). The tricuspid to mitral valve ratio ranged from 46 to 135, averaging 88.8±19.7. The pulmonary artery valve size ranged from 6 to 12 mm, averaging

7.45±1.2 mm, which corresponded to a Z-score of -0.8±0.9 (-2.5- +0.9). Hypoplasia of the right heart chambers in the analyzed patients was expressed to varying degrees: in 19 (82.6%) observations, absence or insignificant hypoplasia of the right heart chambers was verified; moderate hypoplasia of the right heart chambers was detected in 3 (13.03%) patients and pronounced hypoplasia of the right heart chambers was detected in one (4.33%) patient. Ebstein-like dysplasia of the tricuspid valve was observed in three patients.

When conducting selective pulmonary arteriography in axial projection, it was found that in patients with pulmonary atresia types I - II, the pulmonary artery system was developed satisfactorily. In 16 (69.6%) cases, hypoplasia of the pulmonary artery system of varying severity was noted. In two patients with type III defect, only one left pulmonary artery of satisfactory size was detected. In four (2.9%) patients with type IV pulmonary atresia, the pulmonary artery system was not visualized. All patients underwent preoperative drug preparation for 1-10 days in order to correct systemic and pulmonary vascular resistance, as well as to increase arterial oxygen saturation to 80%. From birth until surgery, prostaglandin E1 infusion was administered to maintain adequate systemic perfusion to prevent the development of cardiogenic shock and prevent physiological closure of the patent ductus arteriosus. If necessary, inotropic drugs were prescribed and metabolic acidosis was corrected. In some patients, correction was performed with alkaline solutions with caution in order to prevent the risk of a decrease in pulmonary vascular resistance and, thus, an increase in Qp/Qs, which in turn reduces systemic blood flow with a high probability of forming a "vicious circle" and increasing acidosis.To prevent septic complications, children were tested for microflora and antibiotic sensitivity immediately after birth, and antibiotics were prescribed depending on the results of the tests. Individual parenteral nutrition was prescribed to prevent ischemic and necrotic changes in the gastrointestinal tract. This anatomical form of the defect belongs to the critical category, characterized by the absence of an exit from the right ventricle and communications at the level of the interventricular septum. Indications for perforation and balloon valvuloplasty of the pulmonary valve were: the presence of a membranous form of valve atresia; hypoplasia of the right heart (Z-score of the tricuspid valve was more than 4); the presence of 2-3 anatomical structures of the right ventricle; the absence of coronary blood flow dependent on the right ventricle.

In cases where, along with pulmonary atresia, there was concomitant tricuspid valve insufficiency caused by its organic pathology, or Ebstein-like anatomy or shortening of one of the valve cusps, preference was given to surgical correction of the defect in combination with tricuspid valve plastic surgery and reduction of the size of the chamber of the dilated right atrium. Due to the unfavorable anatomy of the defect, it was not possible to restore antegrade blood flow into the pulmonary artery by perforation in six patients, so they underwent surgical pulmonary artery valvuloplasty.

Balloon valvuloplasty of the pulmonary artery was performed in 23 infants. The median age at the time of endovascular intervention was 2 [2; 5] days (from 5 hours to 14 days); the average body weight was 3.37±0.52 kg (2.25-4.7 kg). All patients required prostaglandin infusion at the preoperative stage.

Severe cyanosis of the skin at the time of hospitalization was observed in 11 (47.8%) patients. The level of arterial blood saturation at the preoperative stage after prostaglandin infusion fluctuated within 42-94%, amounting to 78.1±10.4%, on average.

Nine (41.5%) patients were hospitalized in severe condition with signs of decompensation of the defect, heart failure and metabolic disorders or respiratory acidosis. Seven (29.3%) patients required mechanical ventilation due to respiratory failure and decompensation. One patient had clinical and laboratory signs of pneumonia at the time of hospitalization. Sympathomimetics were prescribed to 10 patients at the preoperative stage. Due to the impossibility of stabilizing the condition with medication

methods, three patients were operated on for vital indications.

Results

After the endovascular interventions, no fatal outcomes were observed in the analyzed group of newborns and infants in the first year of life. In patients with pulmonary atresia, staged treatment was used, which involved step-by-step endovascular correction of the elements of the defect, thereby reducing the trauma of the operation. Options for performing endovascular interventions in patients with pulmonary atresia are presented in Table 3.

An immediate complication of the endovascular intervention was thrombosis of the femoral vein in the access zone in one patient, which required additional therapy. Such a complication as perforation of the right ventricular outflow tract by a coronary conductor passed without the prescription of additional therapy. Transient cardiac arrhythmias during endovascular intervention were stopped by the use of drugs in three patients.

Option of REV intervention	n. of patients n (%)
Stenting of PA branches	1 (4%)
Perforation and BV of the PA valve	6 (28%)
BALKA stenting	5 (22%)
Stenting of PDA	2 (7%)
Rashkind procedure	9 (39%)

Table 3. Endovascular intervention options in 23 infants with pulmonary atresia
Legend: REV – X-ray endovascular intervention; PA – pulmonary artery: BV – balloon valvuloplasty: BALKA – large aortopulmonary collateral arteries: PDA – patent ductus arteriosus.

All patients showed angiographic signs of opening of antegrade pulmonary blood flow after the procedure. During control ventriculography of the right ventricle, filling of the trunk and branches of the pulmonary artery with contrast agent was recorded. All patients showed a statistically significant decrease in systolic pressure in the right ventricle from 58-129 mmHg to 35-75 mmHg, averaging 99.1±21.5 mmHg and 50.0±9.0 mmHg, respectively ($p<0.001$) (Table 4). The residual pressure gradient in the right ventricular outlet region after dilation fluctuated within 10 - 47 mm Hg, amounting to 20 ± 8.8 mm Hg, on average. We did not observe any significant changes in the right ventricular end-diastolic pressure or the pressure gradient in the interatrial junction.

Indicators	Result	After dilation	P
SpO_2 (%)	75,1±11,5 (40-90)	91,0±5,09 (70-99)	<0,001
LV pressure (mmHg)	99.1±21,5 (58-129)	50,0±9,0 (35-75)	<0,001
Pressure gradient in the RVOT zone (mmHg)	-	20 ±8,8(10-47)	-
RV EDV (mmHg)	13,0±4,9 (5-27)	3,4±3,4(1-10)	<0,001
Δp RA-LA (mmHg)	4,2±0,8(0-8)	3,9±0,6(0-7)	>0,05

Table 4. Pulmonary Valve Perforation Success Rates in Patients with Pulmonary Atresia.
Legend: - arterial blood oxygen saturation; LV – left ventricle; RVOT – right ventricular outflow tract; EDV – end-diastolic volume; RV – right ventricle; RA – right atrium; LA – left atrium.

Along with the described criteria, fluoroscopic visualization of the balloon catheter, which was inflated at the level of the atretic pulmonary artery valve, was important. At the moment of inflation, the rupture of the valve membrane on the balloon could be clearly visualized. Despite the increase in arterial blood oxygenation levels immediately after successful balloon valvuloplasty, in the postoperative period, with closure of the patent ductus arteriosus, arterial blood oxygen saturation values significantly decreased in all patients.

On average, on the third day after performing balloon valvuloplasty, with the cessation of prostaglandin infusion and a decrease in blood flow through the patent ductus arteriosus, a decrease in the average arterial blood oxygen saturation level was noted from 91.0±5.09% (70-99%) after dilation to a level of 70±18.7% (32-90%) after performing endovascular intervention.

Such dynamics were associated with insufficient antegrade pulmonary blood flow against the background of the closure of the compensatory communication – the patent ductus arteriosus. Insufficient blood flow through the pulmonary valve can be explained by tricuspid valve hypoplasia and insufficient volume, diastolic dysfunction, and low compliance of severe right ventricular wall hypertrophy. These factors were unable to provide adequate blood ejection into the pulmonary artery to ensure sufficient antegrade pulmonary blood flow. The average period of stay of patients in the intensive care unit after balloon valvuloplasty was 9 [7; 14] days (range 3–50 days). Five patients required sympathomimetic support in the postoperative period for periods ranging from 3 to 20 days, with an average duration of 8.0±4.7 days, as well as artificial ventilation, the duration of which ranged from 7 hours to 23 days, with an average duration of 4.4 [1.4; 8.5] days. Of the 23 patients with pulmonary atresia who underwent balloon valvuloplasty, only nine (39.1%) children did not require repeated interventions during the observation periods from 2 days to 12 years of monitoring. 14 (60.9%) patients underwent 18 cardiac surgeries in the future.

In the early postoperative period after balloon valvuloplasty of the pulmonary valve, repeat interventions were performed in three (13.0%) infants. In two cases (8.7%), repeated intervention was aimed at increasing pulmonary blood flow by creating a systemic-pulmonary anastomosis, on average after 5 [2, 6] (1-12) days. After primary balloon valvuloplasty, two patients underwent systemic-to-pulmonary anastomosis; two underwent closure of Blalock anastomosis and interatrial communication at 1 year and 4 years of age, respectively; one patient underwent cavopulmonary anastomosis as a repeat procedure at 7 years of age. Two patients underwent repeat balloon valvuloplasty at follow-up periods ranging from 41 to 366 days, with an average of 103 [90; 210] days after the primary endovascular intervention. One patient, after repeated balloon valvuloplasty, at the age of 6 years underwent plastic surgery of the right ventricular outflow tract with simultaneous plastic surgery of the tricuspid valve. The absence of the need for repeat cardiac surgery during a mean follow-up period of up to 6 years was 35.9% (95% CI: 20.8-51%).

In the early postoperative period after performing balloon valvuloplasty, in 7 cases there was a need to impose a systemic-pulmonary anastomosis, which accounted for 87.5% of all repeated interventions performed during the first month after the initial valvuloplasty. At long-term follow-up, repeated endovascular valvuloplasties were performed in five patients; surgical reconstruction of the right ventricular outflow tract with simultaneous tricuspid valve plasty was performed in one patient.

Thus, pulmonary atresia is one of the complex ductus-dependent congenital heart defects.

Conclusion

1. Endovascular surgical interventions in newborns and infants of the first year of life of patients with pulmonary atresia are the operations of choice, contributing to the life support of newborns and infants of the first year of life, allowing to prepare the patient for the next stage of surgical treatment.
2. Endovascular interventions are minimally invasive, which increases the chances of survival for patients with complicated anatomy, despite the severity of their condition upon admission to hospital.

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