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Pulmonary Endarterectomy and Balloon Angioplasty in Children and Adolescents with Chronic Thromboembolic Pulmonary Hypertension.

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ABSTRACT

Introduction: Chronic thromboembolic pulmonary hypertension (CTEPH) is a rare form of pulmonary hypertension (PH) that can lead to progressive right heart failure and death. Pulmonary thromboendarterectomy (PTE) is the treatment of choice resulting in significant improvements in functional status, cardiopulmonary hemodynamics, and survival [1] [2]. Pulmonary emboli and associated CTEPH is unusual in the pediatric population and mostly reveal an underlying thrombophilic state [3].

Methods: Bakulev National Medical Research Center (BNMRC) for Cardiovascular Surgery, CTEPH database identified patients 18 years or younger at the time of PTE surgery or balloon pulmonary angioplasty (BPA) performance. Medical charts were reviewed for hemodynamics, thromboembolic risk factors, postoperative complications and outcomes.

Results: PTE in pediatric patients resulted in improved functional status and significantly improved cardiopulmonary hemodynamics: mean pulmonary arterial pressure (mPAP) decreased from 44.0 mm Hg (37.8-47.5) to 19.0 mm Hg (17.8-29.0) ($P = 0.008$), pulmonary vascular resistance (PVR) decreased from 560 dynes*s*cm⁻⁵ (393.8-785.8) to 147 dynes*s*cm⁻⁵ (129.5-245.0) ($P = 0.008$), brain natriuretic peptide (NT-proBNP) also decreased from 315.0 pg/ml (163.2-432.5) to 41.0 pg/ml (23.0-69.8) ($P = 0.008$). BPA also improve cardiopulmonary hemodynamics. There were no deaths during PTE or 30 days after surgery, as and after BPA. Long-term survival (5+years) was achieved in 100%. In the long-term we did not observe rethrombosis in our patients.

Conclusions: This study...

Index Terms: Balloon Pulmonary Angioplasty • Chronic thromboembolic pulmonary hypertension • Pediatric population • Pulmonary thromboembolism

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FULL ABSTRACT

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Conclusions: This study demonstrates that PTE and BPA in pediatric patients with CTEPH is well tolerated with improved postoperative hemodynamics, functional status, minimal postoperative complications, and without perioperative mortality.

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Pulmonary Endarterectomy and Balloon Angioplasty in Children and Adolescents with Chronic Thromboembolic Pulmonary Hypertension.

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Abstract

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Keywords: Balloon Pulmonary Angioplasty, Chronic thromboembolic pulmonary hypertension, Pediatric population, Pulmonary thromboembolism

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1 Introduction

The main causes of the development of pulmonary embolism (PE) in children can be a number of pathological conditions or iatrogenic changes in the structure of blood vessels or chambers of the heart. The most common are various types of coagulopathies, such as thrombophilia and antiphospholipid syndrome [4, 5, 6]. In the pediatric population, cases of pulmonary embolism are very rare and, according to some data, can be 0.9 cases per 100,000 children per year [7], and 6 per 10,000 children hospitalized with this nosology [8]. Compared to adults, these pathological conditions in children become more frequent causes of the development of rethrombosis and CTEPH

CTEPH, as well as a combined approach including balloon dilatation of thrombotic masses and PTE [19, 20].

In this paper, we describe successful surgical and endovascular treatment of CTEPH in children and adolescents at one institution.

2 Methods

2.1. Study Design

Retrospective and prospective review of BNMRC CTEPH database identified patients who were 18 years of age or younger at the time of PTE or BPA surgery. Results of PTE in 8 patients and BPA in 4 patients operated on from July 2016 to April 2026 were conducted. PTE

and BPA were performed at the Department of Heart Diseases with Progressive Pulmonary Hypertension Surgical Treatment in the BNMRC using previously described standard techniques [21, 22]. Evaluation of pre- and postoperative clinical examinations and invasive pulmonary artery pressure (PAP) measurement in all patients involved were systematically analyzed. This study was conducted according to the principles expressed in the Declaration of Helsinki and was approved by the Institutional Review Board. Written informed consent was obtained from each patient before interventions.

All the patients were examined before and immediately after PTE or BPA and at 3–4 months after. Before and after the operation all the patients were evaluated with clinical assessment, transthoracic echocardiogram (TEE), computed tomography angiogram (CTA), right heart catheterization (RHC) with invasive PAP measurement, and dual-energy CT scan (d-e CTs), and the data received were collected. All the patients with acute PE did not fit PTE undergoing. Demographic variables, PAP (systolic [PAPs]), mPAP with subsequent PVR calculation, right ventricle (RV) dysfunction measured by tricuspid annular peak systolic excursion (TAPSE) on 2D echo, d-e CTs, central venous pressure (CVP), oxygen saturation (SatO₂), and any post-invasive and postoperative complications were studied before and after treatment.

2.2. Surgical Protocol

Two of the 8 patients were operated on by using the classical approach proposed by the San Diego group as the preferable method to extract pulmonary thrombi [23]. In these two cases, organized thromboembolic material was classified according to the Jamieson scoring system as type 3 (distal segmental disease with weakly visible thrombus). The other 6 patients were operated on without circulatory arrest [21]. PTE involved cardiopulmonary bypass (CPB), cardioplegia with moderate hypothermia, followed by consecutive endarterectomy of the right and/or left pulmonary arteries. Type 1 disease (organized and chronic thrombus in main or lobar pulmonary arteries) was obtained in 2 patients, type 2 (in lobar or proximal segmental arteries) in 4 patients, and type 3 (distal segmental disease) in 2 patients. These 2 patients were operated on using circulatory arrest. No patient revealed type 4 (inoperable disease/distal arteriolar vasculopathy). Patients' immediate postoperative care took place in the intensive care unit (ICU), where mechanical ventilation, inotropic support, and aggressive diuresis were provided as long as necessary for recovery. For those patients believed to be at a higher risk for rethrombosis after endarterectomy, such as those with thrombophilia [10], intravenous heparin was started within 6 to 8 hours after the operation. Postoperative thrombosis prophylaxis continued until the resumption of warfarin anticoagulation resulted in a therapeutic international normalized ratio (INR). Postoperative cardiopulmonary hemodynamics were measured by thermodilution catheterization techniques before transfer out of the ICU with the patients off both mechanical ventilation and inotropic agents. All other postoperative studies, including TEE, d-e CTs, rest and exercise oxygen saturation testing, and chest radiography occurred while the patients were convalescing on the general hospital ward. All 4 patients who received BPA had type 3–4 vascular changes according to Jamieson's classification.

2.3. Balloon Pulmonary Angioplasty Protocol

Local anesthesia (e.g., 1–2% lidocaine) is administered at the puncture site, followed by percutaneous access to the common femoral vein using the Seldinger technique under ultrasound guidance. An introducer sheath (6–8 Fr) is placed to secure vascular access.

A hydrophilic guidewire is advanced through the introducer sheath, navigating sequentially through the inferior vena cava, right atrium, and RV into the PA. Selective pulmonary angiography is performed by

injecting iodinated contrast medium through a pigtail or multipurpose catheter to delineate stenotic or occluded segments. A 0.014–0.035" guidewire is navigated across the target lesion and positioned distally for stability. A balloon catheter, sized at 0.8–0.9× the reference vessel diameter, is advanced over the wire and inflated with a contrast-saline mixture (1:1 dilution). Inflation pressure is gradually titrated (4–12 atm) under fluoroscopic guidance to avoid complications such as rupture or dissection. Repeat angiography confirms procedural success; residual stenosis (> 30%) may necessitate upsizing the balloon or staged interventions.

2.4. Statistical Analysis

Statistical analysis was performed using Statistica software, version 13.1 (TIBCO Software Inc., Palo Alto, CA, USA). Continuous variables are expressed as median with interquartile range (IQR); categorical variables are expressed as absolute numbers and percentages. In the PTE group ($n = 8$), pre- and postoperative hemodynamic and biochemical parameters (mPAP, cardiac output [CO], PVR, and NT-proBNP) were compared using the Wilcoxon signed-rank test for paired non-parametric data, given the small sample size and the inability to reliably confirm normality of distribution. A two-tailed p -value < 0.05 was considered statistically significant. In the BPA group ($n = 4$), the sample size was insufficient to yield statistically meaningful results with formal hypothesis testing; therefore, pre- and post-procedural values are reported descriptively as individual patient-level data and as group median with range, together with the percentage change from baseline for each patient. No formal significance testing was performed for this subgroup, and results were interpreted on the basis of the consistency and magnitude of individual clinical change rather than statistical inference. A p -value < 0.05 was considered statistically significant throughout the analysis, where applicable.

3 Results

3.1. Demographics

Baseline patient data and clinical characteristics are presented in **Table 1**.

The inclusion criteria suggested by the American College of Chest Physicians were the following: 1) New York Heart Association (NYHA) class symptomatology; 2) preoperative PVR > 300 dynes · s · cm⁻⁵; 3) surgical accessibility of thrombi in main, lobar, segmental pulmonary arteries (PA) as seen in CTA; 4) no debilitating comorbidities. All the patients with acute pulmonary embolism did not fit CPB undergoing. The age range of the patients was 29 months to 18 years. The number of men and women was identical (50%). The most common presenting symptom was exertional dyspnea and/or exercise intolerance (80%). Syncope or presyncope were observed in 2 children, both were under 3 years of age. Other presenting symptoms included chest pain (25%), cough (10 %), oedema (25%). Cyanosis was observed in 1 patient and none of them had hemoptysis. At the time of hospitalization initial diagnosis was PE in 3 patients, but after a thorough examination, it was replaced by CTEPH. Finally, all patients had known CTEPH at the time of surgery or intervention and in one of them the embolism was recurrent. NYHA functional class (FC) I was in 1 patient (8%), class II in 4 patients (33%) and class III in 7 patients (69%). No one patient was in class IV.

3.2. Thromboembolic Risk Factors

Six (50%) of the 12 patients had 1 or 2 risk factors for thromboembolism as depicted in **Table 2**. The most common thrombophilic state was antiphospholipid syndrome ($n = 2$).

Table 1. Pediatric patient baseline demographics.

Patient	Age (y)	Gender	WHO score	Initial diagnosis
1	2.5	M	3	PE
2	6	M	3	CTEPH
3	17	F	2	PE
4	16	M	3	CTEPH
5	12	F	3	CTEPH, Recurrent PE
6	18	F	1	CTEPH
7	17	F	3	CTEPH
8	17	F	2	CTEPH
9	17	M	3	CTEPH
10	5	M	2	PE
11	14	M	3	CTEPH
12	16	F	2	CTEPH

Table 2. Triggers for the development of CTEPH.

Patients / age (years)	Thromboembolic risk factors	Comorbidity	Surgical treatment / management	CVC / VAS	Deep venous thrombosis	Intracardiac thrombi
1 / 2.5	No	Agenesis of the corpus callosum. Giant arachnoid cyst.	2 neurosurgical operations.	+/+	No	RA, SVC
2 / 6	No	Nephrotic kidney disease.	No	+/-	No	RA, SVC
3 / 17	No	Post-COVID pneumonia	No	+/-	+	No
4 / 16	No	Post-COVID pneumonia	No	+/-	+	No
5 / 12	No	Injury of the lower limb. Prolonged immobility.	No	-/+	+	RA
6 / 18	No	Fracture of the lower limb. Prolonged immobility.	Trauma surgery	No	+	No
7 / 17	Prothrombin gene mutation (G20210A). Hyperhomocysteinemia	No	No	No	+	No
8 / 17	Homozygous thrombophilia (TGA 2807 > T-TT).	No	No	No	No	No
9 / 17	Antiphospholipid syndrome. Lupus anticoagulant.	CHD. PAPVC. PE LPA (14 years ago).	PAPVC correction + PTE	No	No	No
10 / 5	Elevated factor VIII.	CHD. AS. RA thrombus (3 years ago).	AS correction + RA thrombectomy	No	No	RA
11 / 14	Protein C and S deficiency.	Pneumonia.	No	+/-	+	RV
12 / 16	Antiphospholipid syndrome.	No	No	No	+	No

VAS – Ventriculoatrial shunt. CVC – Central venous catheter. AS – Aortic valve stenosis

One patient exhibited testing suggestive of both protein C and S deficiency. Prothrombin gene mutation, the Lupus anticoagulant and Elevated factor VIII were in 1 patient.

Other disorders seen in this cohort that are associated with increased risk of thromboembolism [25, 26] include the presence of a VAS ($n = 1$) or nephrotic syndrome ($n = 1$). Another potential risk factor identified was prolonged immobilization of a limb. Two patients had a prior injury or fracture of their lower limbs. Prolonged immobilization in both cases appeared to be related (temporally or locally) to the subsequent development of a deep venous thrombosis (DVT), as and CVC in two patients with severe post-COVID pneumonia. Three patients had previously undergone surgery. Two for the correction of congenital heart diseases and one more for two neurosurgical operations.

3.3. Severity of Pulmonary Hypertension

Baseline and postoperative (after PTE and BPA) pulmonary hemodynamic measurements are shown in **Tables 3A (A, B), 4A (A, B)** and **Figure 1**. None of the 8 patients who underwent PTE had PVR greater than 1000 dynes $\cdot$ s $\cdot$ cm⁻⁵.

The number of sessions performed and number of dilated segments are presented in **Table 4A**, and the results of BPA are presented in **Table 4B**.

According to NYHA, the clinical conditions have also improved: class I was observed in 6 (50%) patients, class II in 4 (33%) and class III in 2 (17%) patients.

At presentation to BNMRC for surgery 2 (17%) patients (both less than 3 years of age) were on inotropic support (epinephrine -1, dobutamine -1). Ten (83%) patients were receiving pulmonary vasodilator therapy. Preoperative pulmonary hypertension-specific pharmacotherapy included bosentan ($n = 8$), sildenafil ($n = 2$). Therapy was continued in the follow-up.

3.4. Details of Surgical Intervention

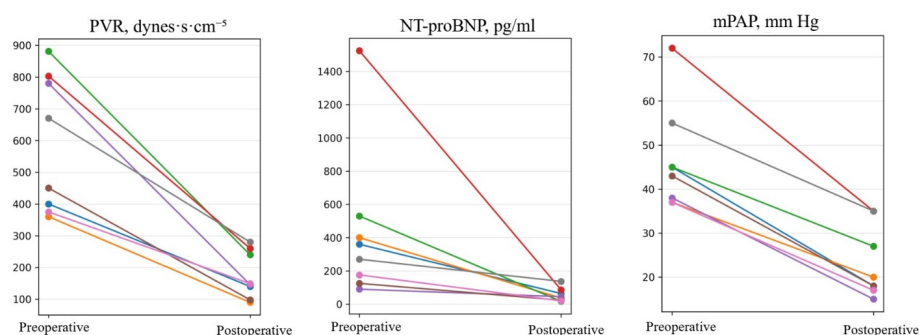
The mean CPB time in 6 patients operated on without deep hypothermic circulatory arrest (DHCA) was 120.7 ± 35.5 min, the mean aortic cross-clamp time was 67.4 ± 24.7 min. Patients were cooled down to $28^\circ\text{C} \pm 3^\circ\text{C}$. In 2 patients operated on with DHCA CPB time was 196 and 217 minutes respectively. Both patients were cooled down to 20°C . Right-sided circulatory arrest time was 17 and 19 minutes, and left-sided circulatory arrest time was 20 minutes in one patient. CPB provided an adequate oxygen delivery ($\text{DO}_2 > 270 \text{ mL/m}^2/\text{min}$) with adequate hemodynamic (average arterial pressure $\geq 50 \text{ mm Hg}$). In all operated patients (100%), the spontaneous restoration of cardiac activity was observed. CPB was completed with moderate doses of inotropic agents (inotropic score— 5.0 ± 4.85). In 2 of the patients observed,

Table 3A. Cardiopulmonary hemodynamics before and after PTE (n = 8).

	Preoperative				Postoperative			
	mPAP (mm Hg)	CO (L/min)	PVR (dynes · s · cm ⁻⁵)	NT-pro BNP (pg/ml)	mPAP (mm Hg)	CO (L/min)	PVR (dynes · s · cm ⁻⁵)	NT-pro BNP (pg/ml)
1.	45	2.2	400	360	18	2.9	140	64
2.	37	2.7	360	400	20	3.6	90	37
3.	45	3.4	881	530	27	3.8	240	17
4.	72	2.2	803	1524	35	3.4	260	87
5.	38	3.2	780	90	15	3.6	146	45
6.	43	3.6	450	125	18	3.6	98	24
11.	37	3.3	375	176	17	3.4	149	20
12.	55	3.8	670	270	35	3.1	280	137

Table 3B. Cardiopulmonary hemodynamics before and after PTE (n = 8). Descriptive and Inferential Statistics.

Parameters	Preoperative Median (IQR)	Postoperative Median (IQR)	Mean relative change	p
mPAP, mm Hg	44.0 (37.8–47.5)	19.0 (17.8–29.0)	−50.8%	<i>p</i> = 0.008
CO, L/min	3.2 (2.6–3.5)	3.5 (3.3–3.6)	+16.1%	<i>p</i> = 0.125
PVR, dynes·s·cm ^{−5}	560 (393.8–785.8)	147.5 (129.5–245.0)	−69.8%	<i>p</i> = 0.008
NT-proBNP, pg/ml	315.0 (163.2–432.5)	41.0 (23.0–69.8)	−79.1%	<i>p</i> = 0.008

**Figure 1.** Hemodynamic changes before and after PTE**Table 4A.** Number of sessions and dilated segments performed under BPA.

N	mPAP (mm Hg)	PVR (dynes · s · cm ⁻⁵)	NT-pro BNP (pg/ml)	Session (n)	Segments (n)
7	68	1391	419	3	10
8	45	640	400	3	8
9	87	1524	803	4	10
10	48	560	460	4	12

Table 4B. Individual patient hemodynamic values before and after BPA (n = 4).

Patient	mPAP pre→post (mm Hg)	%Δ	PVR pre→post (dynes · s · cm ⁻⁵)	%Δ	NT-proBNP pre→post (pg/ml)	%Δ
7	68 → 48	−29.4%	1391 → 870	−37.5%	418 → 173	−58.6%
8	45 → 37	−17.8%	640 → 320	−50.0%	400 → 148	−63.0%
9	87 → 57	−34.5%	1524 → 670	−56.0%	803 → 400	−50.2%
10	48 → 35	−27.1%	560 → 340	−39.3%	460 → 144	−68.7%

%Δ – percentage change

norepinephrine was required to maintain an adequate total peripheral vascular resistance (1 – without and 1 – with DHCA).

3.5. Postoperative Complications

PTE surgery was generally well tolerated by all patients. Residual PH (mPAP = 30 mm Hg) with atrial arrhythmias was observed in one case. Although reperfusion lung injury can occur in up to 40% of adult CTEPH patients [16, 17] there were no cases in our results. In the remaining 7 patients, no significant complications were observed.

BPA was less tolerated. In all 4 patients 3 or 4 sessions of BPA with dilatation of 8, 10 or 12 segments were required to normalize the mPAP (< 25 mm Hg) (**Figure 2**). Reperfusion lung injury was observed in 2

patients (**Figure 3**). Initial PVR was more than 1300 dynes · s · cm^{−5} in both cases. Atrial arrhythmias was observed in one case. Sinus rhythm was observed in all patients after PTE and BPA at discharge from the hospital. No bleeding complications and no incidence of postoperative delirium were noted in all our patients.

3.6. Mortality

No pediatric patient in this series died during surgery or within 30 days after surgery or BPA at our institution. Long-term survival (5+ years) was 100%.

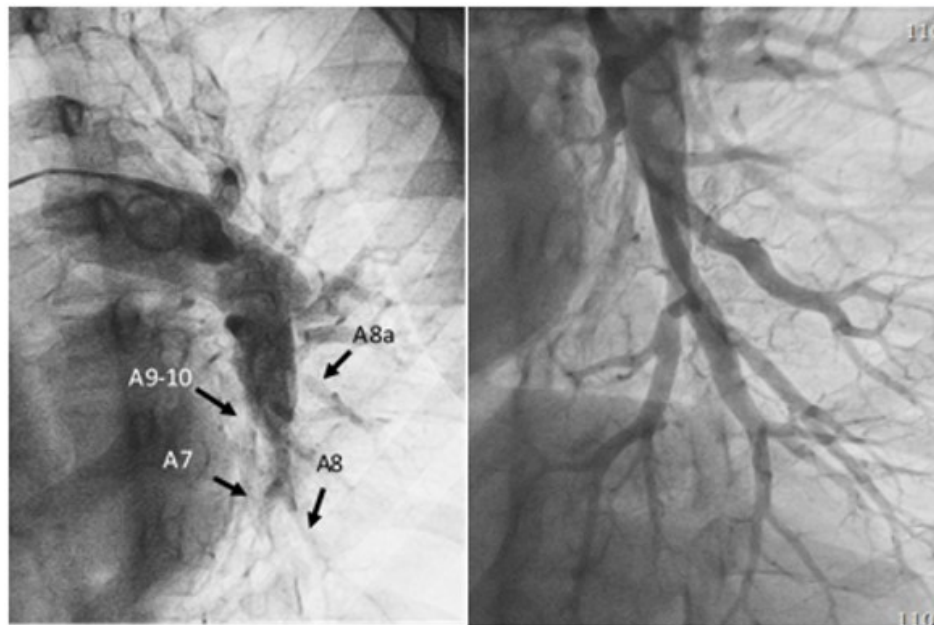


Figure 2. Pulmonary angiography before (A) and after (B) 4 sessions of BPA.



Figure 3. X-ray of the lungs. Left-sided inferior lobe reperfusion pulmonary oedema (area of BPA exposure).

4 Discussion

This report details the case series of pediatric CTEPH patients who have successfully undergone PTE surgery, emphasizing not only that this operation is well tolerated, but also that it is associated with low morbidity, no mortality, and significant improvement in pulmonary hemodynamics and right heart function. This study also serves to highlight important features of pediatric CTEPH patients, including the increased presence of thrombophilic disorders, especially in combination with central venous catheters used in children requiring long-term parenteral nutrition or treatment of pneumonia.

PTE is recommended as a first-line treatment for operable patients [1, 2, 27]. However, despite continued improvement in surgical and interventional techniques, PTE remains a challenging procedure. This is particularly the case in patients with elevated PVR, as several studies have previously shown that high preoperative PVR was associated with an increase in early postoperative mortality [28, 29, 30].

The highest incidence of PE in adult patients is due to the presence of DVT and can reach 51% [31]. In children, DVT can also lead to the development of PE in 56% of cases, but only in a small number of cases is the source of PE thrombi from the veins of the lower limbs [32]. According to the Canadian Registry, this cause of PE was identified in 0.86 per 10,000 patients [33, 34]. Madani et al. described 4 cases of verification of the diagnosis of DVT that caused CTEPH out of 18 operated patients, as well as 3 cases of this complication in children who had severe long-term pneumonia [10].

In our series, DVT caused thromboembolism and subsequent development of CTEPH in 7 patients (58%), and in 3 cases it was caused by long-term therapy of severe disease (pneumonia) that required the use of a CVC. In the remaining two cases, the cause of venous thrombosis can be considered prolonged immobilization of the lower limb. Our report revealed three children who suffered from severe pneumonia (in two cases viral and in the other bacterial) requiring a wide variety of antibiotic therapy for a long period. In both of these cases, the patients developed DVT, which was the cause of PE. In two cases, the trigger for the development of DVT and subsequent PE was lower limb injury, which required a long period of treatment in a hospital setting with passive mobility. There were no data on coagulation changes in these patients at the time of hospitalization, although this cannot be completely excluded given that they did not undergo specialized blood tests. The most common causes of thrombosis are various types of thrombophilia [4, 5].

Thrombophilia was detected in 12 out of 18 children in the previously mentioned work [10]. Kumbasar et al. described results of 3 cases of PTE in children; thrombophilia was diagnosed in 2 of them [3]. Of the 12 patients in our series, various variants of thrombophilia were observed in 6 (50%) patients, although in 2 cases at the time of hospitalization no specialized studies of the coagulation system were performed. The most common variant was antiphospholipid syndrome (2 cases), in one case in combination with Lupus Anticoagulant. Homozygous thrombophilia (TGA 2807 > T-TT), elevated factor VIII, and protein C and S deficiency were present in one case each. One patient had a combination of genetic predisposition (heterozygous carriage of G20210A mutation in the prothrombin gene) and metabolic disorders (hyperhomocysteinemia) as an etiological factor of PE. A massive subsegmental type of lesion was identified. The patient was found to be inoperable according to the European Guidelines for the Diagnosis and Treatment of PH in Adults [1]. These guidelines recommend alternative therapies for this group of patients, such as BPA, alone or in combination with specific drug therapy. Percutaneous revascularization of pulmonary vascular obstructions with BPA has emerged as a potential treatment option for individuals who are unsuitable candidates for PEA or have

residual obstruction after PEA [35, 36, 37]. Over the past 5 years, BPA has become a guideline-recommended treatment (class IB) for inoperable CTEPH patients or patients with residual PH after PEA [1].

The possibility of damage to the vascular wall during endovascular intervention remains a difficult question, especially in the pediatric population. Does it influence the results of subsequent PEA? Shimahara et al. did not find such danger in their recently published article. No case of technical difficulty caused by a destroyed PA plane after receiving BPA as a first treatment was observed. Endovascular exposure did not cause additional problems when performing open surgery [38]. A number of other authors are also inclined to this opinion [39, 40].

Our results demonstrated that the complication rate after BPA proved to be higher than after CPB. We do not see anything surprising in this due to the fact that in the group of patients who underwent BPA, the severity of PH was significantly higher. That is why the most common complication was reperfusion pulmonary oedema. At the same time, the absence of mortality makes it possible to advocate this approach for the treatment of children with CTEPH and high PH.

5 Conclusion

The efficacy of PTE and BPA for CTEPH treatment in adults has been reported. Historically, data on the results of these interventions in children and adolescents have been limited.

The results of this study suggest that PTE and BPA are effective for CTEPH treatment in children, achieving significant hemodynamic improvement without in-hospital mortality. However, it is necessary to continue refining criteria for patient selection and procedural timing.

LIMITATIONS

The major limitation of this study is the small sample size, which makes it challenging to establish definitive statistical criteria for choosing between surgical and interventional approaches in pediatric CTEPH. Nevertheless, this series provides critical evidence supporting the feasibility and safety of both PTE and BPA in pediatric cohorts.

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KEY TAKEAWAYS

What Is Already Known? PTE is considered the gold standard for CTEPH surgical treatment in adult populations.

What Does This Study Add? PTE and BPA are safe and effective for CTEPH treatment in pediatric and adolescent patients, achieving 100% long-term survival with significant hemodynamic improvement.

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