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Case Report

Pulmonary Endarterectomy for Chronic Thromboembolic Pulmonary Hypertension in a 5-year-old Girl with No Risk Factors - A Case Report

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Abstract: Chronic thromboembolic pulmonary hypertension (CTEPH) in pediatric population is rare. We present a case of five-year-old female child, diagnosed as CTEPH on warfarin and home oxygen for 6 months. Imaging showed extensive thrombus in distal right pulmonary artery (RPA) than left pulmonary artery (LPA). She clinically deterioration inspite of maximal medical management and was referred for surgical option. Considering lack of literature evidence on surgical outcomes in pediatric CTEPH, decision to perform surgery was made after multi-disciplinary team discussion between adult and paediatric pulmonary hypertension and cardiothoracic surgical teams across two units in UK. We performed pulmonary thromboendarterectomy (PEA) using cardiopulmonary bypass under deep hypothermic circulatory arrest (DHCA). The surgical procedure was uneventful with good clinical recovery. Our report highlights the need for earlier and prompt diagnosis with a need for referral to a specialized surgical center at an earlier stage before intractable vascular resistance ensues. Surgery can be safely performed for CTEPH in pediatric populations.

Keywords: Pulmonary Endarterectomy, Pulmonary Hypertension, Chronic Thromboembolism

1. Introduction

Chronic thromboembolic pulmonary hypertension (CTEPH) is defined as mean pulmonary arterial pressure (mPAP) above 20 mm Hg and pulmonary capillary wedge pressure below 15 mm Hg in the presence of multiple chronic thrombi in the elastic pulmonary arteries after at least three months of effective anticoagulation [1].

CTEPH may represent manifestation of unresolved pulmonary embolus with an underlying hypercoagulable state or result from pulmonary arteriopathy. It is classified as Group 4 pulmonary hypertension as per Paediatric Task Force of the 6th World Symposium on Pulmonary Hypertension (2018) [2]. Risk factors include indwelling vascular catheter, ventriculo-atrial shunts, hypercoagulable state, history of malignancy, splenectomy, thyroid replacement therapy, chronic inflammatory conditions, etc.

Progressive dyspnoea and exercise intolerance are early symptoms. Flow murmurs or bruit over the lung fields have been described and are accentuated during inspiration indicating turbulent flow in partially obstructed pulmonary arteries [3]. Chest radiograph may be normal or show area of hypoperfusion.

Suspected cases undergo ventilation/perfusion (V/Q) lung scan (sensitivity of 96 to 97%, specificity of 90 to 95%) followed by computed tomographic pulmonary angiography (CTPA) to rule out non-thromboembolic causes such as sarcoma, veno-occlusive disease, fibrosing mediastinitis,

extrinsic pulmonary vascular compression, etc [4]. CTPA is limited in its ability to differentiate to an acute intraluminal thrombus from a well endothelialized chronic thrombus.

Digital subtraction pulmonary angiography is the gold standard for diagnosis [5]. Characteristic findings include, abrupt narrowing of the major pulmonary arteries, obstruction of lobar or segmental vessels at their origin, pulmonary artery webs or bands, pouch defects or intimal irregularities [6].

The decision about operability is based on the clot burden, absence of extensive distal thrombus, risk/benefit for the patient, comorbidities, presence and severity of any hemodynamic impairment, findings on pulmonary angiogram supplemented by a ventilation/perfusion scan and most importantly the experience of surgeon and team [7-9].

We present a case of a 5-year-old girl with symptomatic CTEPH, successfully managed by PEA at Great Ormond Street Hospital (GOSH), London, United Kingdom.

2. Case Report

A 5 year 5-month-old girl, weighing 16.3 kg, was accepted for PEA surgery at our institution after experiencing pulmonary hypertensive crisis. She was found to have a occlusive thrombus in right pulmonary artery (RPA) with decreased perfusion of the right middle and lower lobes on computer tomography (CT Scan: Figure 1). The V/Q scan suggested that the right lung received 26% perfusion as compared to 74% by left lung (as shown in the figure 2) and was started on warfarin. She underwent right heart catheterisation (RHC) which demonstrated a cardiac index (CI) OF 2.66L/min/m^2, supra-systemic mPAP of 68mmHg and pulmonary vascular resistance index (PVRi) of 21.1 WU.m^2. The decision to operate on her was based on complex multi-disciplinary team discussion between adult and paediatric pulmonary hypertension and cardiothoracic surgical teams across two units, especially considering the lack of data supporting such a procedure in paediatric age group.

After induction of general anaesthesia, the patient suffered a cardiac arrest, with restoration of spontaneous circulation at 8 minutes on resuscitation.

The surgical procedure was performed via median sternotomy and cardiopulmonary bypass (CPB), this being established semi-urgently due to borderline haemodynamics. Deep hypothermic circulatory arrest (DHCA) was established at 18 degrees C for 25 min, as per protocol. Right pulmonary arteriotomy was performed first and endarterectomy plane identified. There was successful retrieval of an organised thrombus from the distal RPA going into interlobar arteries supplying the middle and inferior lobes (Figure 3). Bypass and perfusion was resumed for 19 mins, while arteriotomy was closed primarily. Using another episode of DHCA, left pulmonary artery was incised for inspection and no thrombus was identified. Similarly, arteriotomy was closed primarily.

Heart regained sinus rhythm on rewarming but initial attempts to come off bypass failed as patient's mPAP was more than systemic pressure along with systemic arterial hypotension. Intraoperative echocardiogram suggested severe right ventricular dysfunction owing to elevated pulmonary vascular resistance. Bypass was re-established in order to optimise the inotrope and vasopressor strategy, including inhaled nitric oxide. The mPAP fell gradually over time and was half systemic when decision was taken to come off bypass again which was uneventful this time.

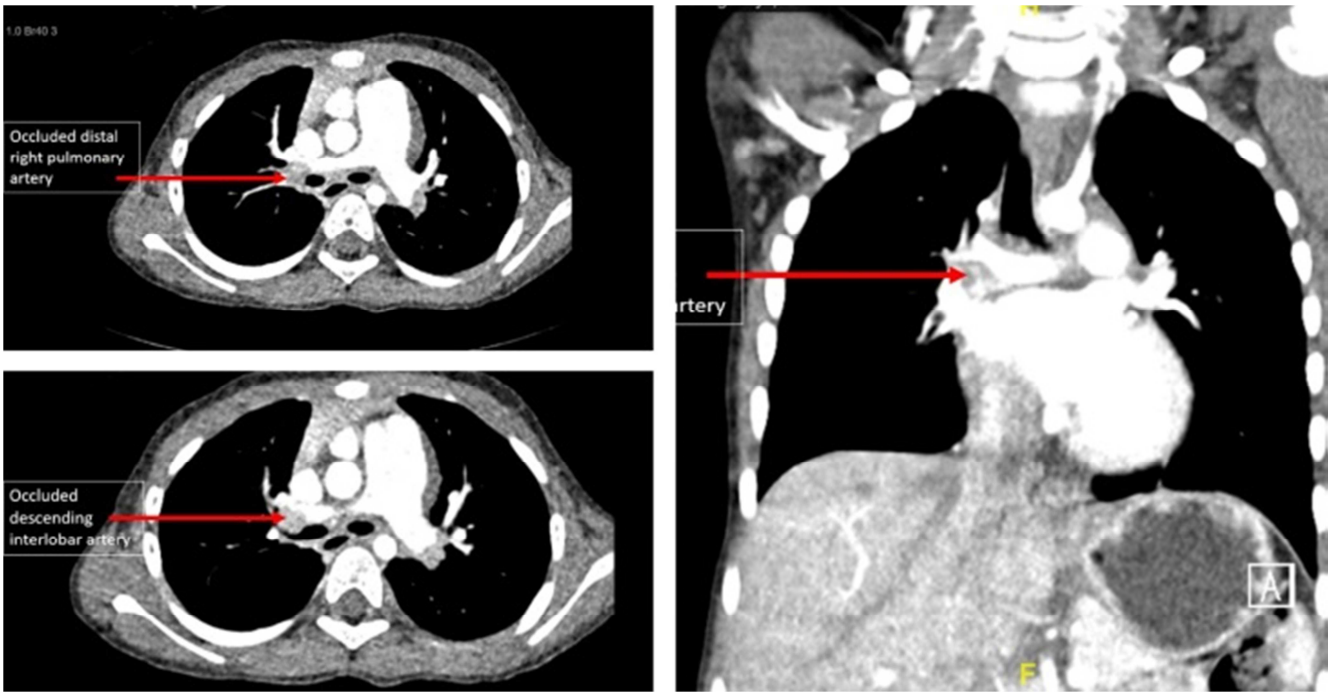
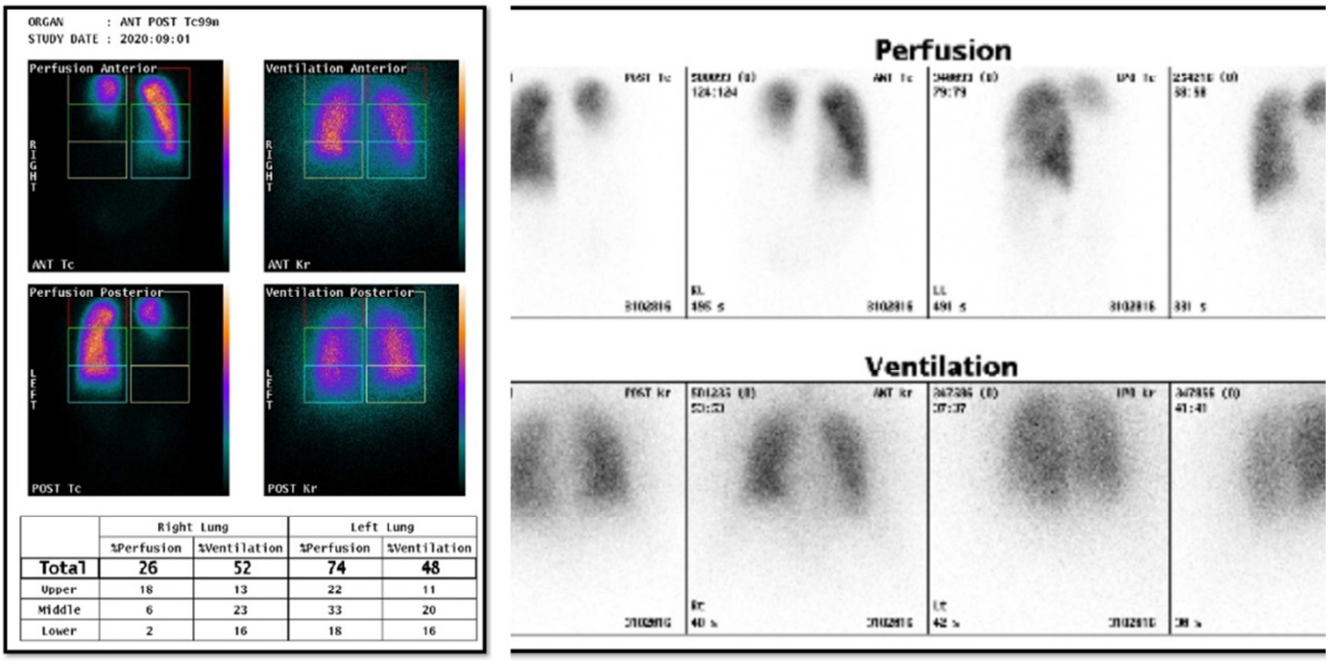


Figure 1. *Cross sectional image of CT scans demonstrating occluded right pulmonary artery and interlobar division on right side due to thrombus within the lumen.*



	Right Lung		Left Lung	
	%Perfusion	%Ventilation	%Perfusion	%Ventilation
Total	26	52	74	48
Upper	18	13	22	11
Middle	6	23	33	20
Lower	2	16	18	16

Figure 2. *Ventilation perfusion scan (V/Q scan) demonstrating poor perfusion on the right side.*

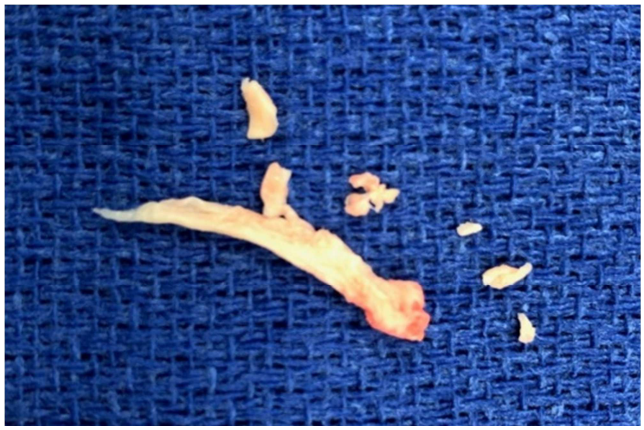


Figure 3. *Surgical specimen showing the thrombi removed from right pulmonary artery.*

Post-operatively, she was transferred to the cardiac intensive care unit (CICU) with open chest on high inotropic support and kept on inhaled nitric oxide at 20ppm, which were gradually weaned off and her chest was closed on post-operative day (POD) 2. Her post-surgical cardiac MRI demonstrated dramatic improvement in right ventricular volumes, function, flow to RPA and mPAP of 34mmHg which was improved from 68mmHg preoperatively. She was extubated on POD 8 and discharged to a medical facility, neurologically intact, on POD 26 on sildenafil, warfarin, furosemide, amiloride and nocturnal oxygen of 0.2 l/min via nasal cannula till next follow up.

She remains on NYHA class 2 clinically. She had resumed her school 3 months post-surgery. Repeat echocardiogram on day 30 showed mildly reduced right ventricular function with discrete proximal narrowing of the right pulmonary artery. Cardiac MRI showed significant reduction in mPAP to 25 mmHg from 68 mmHg preoperatively and preserved right ventricular systolic function.

3. Discussion

The incidence of PE is 8.6–57 in 100,000 in hospitalized children, and 0.14–0.9 in 100,000 in the general population of non-hospitalized children [10-14]. Occlusion of the pulmonary arterial bed results in remodeling of right heart and small distal pulmonary arteries in the nonoccluded areas.

Administration of prostacyclin in patients with severe form of CTEPH decreased pulmonary vascular resistance and improved post- operative outcomes [15]. Inferior vena cava filters are no longer placed prior to PEA in view of peri-operative anticoagulation.

Treatment for inoperable or persistent CTEPH in inoperable cases includes anticoagulation, balloon pulmonary angioplasty and medical treatment with drugs (riociguat).

Definitive management of CTEPH has remained surgical, with bilateral pulmonary endarterectomy with mixed results. The intraluminal thrombus composed of fibrous tissue is densely adherent to the intima and is inaccessible to thrombectomy or dilatation. PEA improves cardiopulmonary hemodynamics, gas exchange and survival by removal of the fibrotic remodeled thrombus and the diseased intima. It prevents progressive distal arteriopathy and mitigates further vascular collateralization. PEA is an effective and successful surgical procedure when performed in an experienced high-volume center. Current in-hospital mortality after PEA is between 2% and 5% in experienced centers [16, 17] and the 5-year survival rate is between 76% and 86% [17–19].

Recent evidence points towards similar success in pediatric population with significant reduction in the mean pulmonary artery pressure, pulmonary vascular resistance and cardiac

Output. 5 years survival was achieved in 87.5% in 17 reported cases [20].

There is a paucity of such evidence for paediatric experience. While surgical technique is essentially similar to the reported procedure in adult PEA, the management principle could vary especially around protection of right ventricle. With better bypass strategy and understanding of pulmonary physiology, this surgery has continued to improve with better survival and results.

The postoperative course may be complicated by post-reperfusion pulmonary oedema causing hypoxemia and prolonged mechanical ventilation; right heart failure secondary to persistently high pulmonary pressure; bleeding from arteriotomy rupture; ventilation associated pneumonia, haemoptysis, phrenic nerve palsy and rethrombosis. Long term anti- coagulation therapy is required, and vitamin K antagonists are more effective than direct oral anticoagulants. In spite of intensive management and satisfactory central thromboendarterectomy, 10% of patients have persistent pulmonary hypertension.

Intensive care management post-operatively includes supportive cardiorespiratory management like lung protective ventilation, end organ perfusion, negative to neutral fluid balance and early thromboprophylaxis. Specific complications like pulmonary vascular steal, reperfusion pulmonary edema can occur. Clinical decision is made for therapeutic anticoagulation once drains and wires are removed.

4. Conclusion

CTEPH in pediatric population can also be treated with surgical thromboendartectomy as in adult population, with excellent outcomes. This report adds to the current evidence which suggests that early referral to a CTEPH centre experienced with PEA in cases with suspected or diagnosed CTEPH in children results in improved hemodynamic and functional status. Joint effort of multidisciplinary team including pulmonary hypertension, cardiology, hematology, critical care, anesthesiology and cardiothoracic surgery are paramount for favorable outcomes.

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