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CRITICAL CARE RESUSCITATION

MULTIDISCIPLINARY TEAM DISCUSSION

Complications of Pulmonary Embolism in a Pediatric Patient

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ABSTRACT

A 16-year-old boy presents with a massive acute pulmonary embolism requiring emergent surgical embolectomy and extracorporeal membrane oxygenation for right ventricular failure. Subsequently he was diagnosed with catastrophic antiphospholipid syndrome requiring immunosuppression, and then pneumatoceles causing tension pneumothoraxes. The rarity of presentation in a child required collaboration across pediatric and adult disciplines. (JACC Case Rep. 2025;30:103221) © 2025 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

CASE PRESENTATION

A 16-year-old boy with obesity and depression presented to the emergency department (ED) with tachycardia, tachypnea, and desaturation. He presented to his pediatrician a week prior with an upper respiratory infection with increased work of breathing and was prescribed azithromycin, steroids, and then remdesivir (COVID subsequently negative). He re-presented a third time to his pediatrician with worsening dyspnea, tachycardia, and hypoxemia, and was referred to the ED.

On presentation to the ED, blood pressure was 114/73 mm Hg, pulse was 177, temperature was 36.8 °C, respiratory rate was 48, and SpO2 was 88% on 15L non-rebreather oxygen. The patient appeared ill, was

TAKE-HOME MESSAGES

- This patient's care required unique multidisciplinary decision making at each step including collaboration between adult and pediatric medicine and surgical colleagues, surgical embolectomy, and activation of the Pediatric Pulmonary Embolism Response Team (**Figure 8**).
- We attribute this patient's excellent outcome despite very severe disease with multiple complications to aggressive, early embolectomy, supported by a multidisciplinary team whose pooled experience enabled better decision making in multiple rare diseases.

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ABBREVIATIONS AND ACRONYMS

APS = antiphospholipid syndrome

BNP = brain natriuretic peptide

CAPS = catastrophic antiphospholipid syndrome

CT = computed tomography

ED = emergency department

ECMO = extracorporeal membrane oxygen

LV = left ventricle

PE = pulmonary embolism

RV = right ventricle

unable to lie flat and could not speak in full sentences. Although rare in healthy children, pulmonary embolism (PE) was suspected because of the patient's tachypnea and tachycardia; labs showed extremely elevated D-Dimer, brain natriuretic peptide (BNP), and troponins (**Table 1**) as well as abnormal arterial blood gases (**Table 2**). Computed tomography (CT) pulmonary angiography results confirmed PE with bilateral main pulmonary artery thrombi with extension into the interlobar and segmental pulmonary arteries bilaterally (**Figures 1A and 1C**). Point-of-care ultrasound showed elevated right ventricular (RV) pressures and strain including RV dilation, flattened interventricular septum, qualitatively diminished RV systolic function, and dilated interior vena cava. Although there is no consensus definition of "RV strain" in pediatric PE, this constellation of RV dysfunction and dilation with increased afterload is consistent with the RV dysfunction seen in high-risk PE[1,2] and accepted definitions of RV strain.[2] The patient became hypotensive, requiring initiation of norepinephrine and epinephrine infusions for vasoactive support, and he was started on heparin for PE. The Pediatric Pulmonary Embolism Response Team was activated.

QUESTION 1: WHAT IS THE BEST INITIAL TREATMENT FOR ACUTE HIGH-RISK PE IN THIS PEDIATRIC PATIENT?

Acute PE is stratified into 3 groups: high-risk, intermediate-risk, and low-risk. The 2019 European Society of Cardiology PE-risk categorization is not validated in pediatrics but applied widely to adolescents in clinical practice.[1] High-risk PE causes cardiopulmonary arrest, sustained hypotension (a late feature of pediatric PE due to compensatory increase in systemic vascular resistance), or normotension with signs or symptoms of shock. Treatment of high-risk PE in adults includes reperfusion therapies such as systemic thrombolysis, and catheter-based therapies such as catheter-based thrombolysis and suction thrombectomy. Adult guidelines recommend surgical embolectomy for intermediate-high and high-risk PE when there is a contraindication to systemic thrombolysis, there is rapid deterioration of the patient, there is thrombus in transit in the heart, or catheter-directed therapies have failed.[3] The optimal approach for high-risk PE in the pediatric population is unknown and the superiority of reperfusion is also not established in pediatrics. In a pediatric cohort, Ross et al[2] proposed primary reperfusion, namely thrombolysis or surgical embolectomy, citing the lack of data and limited experience with catheter-based interventions in children as well as availability of expertise and equipment for these therapies in pediatric hospitals. In the absence of pediatric guidelines, high-risk pediatric PE management is influenced by hospital capabilities including availability of a pediatric cardiothoracic surgeon, experience with catheter-directed therapies including immediate availability of specialty equipment, patient profile, time to each therapy, availability of extracorporeal membrane oxygen (ECMO), and physician judgment.

TABLE 1 Elevated Markers Suggestive of Acute Pulmonary Embolism

Troponin	564.2 ng/L
BNP	419.8 pg/mL
D-Dimer	>20.0 μg/mL

BNP = brain natriuretic peptide.

For this patient, our local Pulmonary Embolism Response Team, consisting of hematology, intensive care unit, ED, interventional cardiology, cardiac anesthesia, and interventional radiology teams,[4] used rapid multidisciplinary decision making to mobilize resources including ECMO and cardiothoracic surgery. Because of signs of impending cardiovascular collapse, including hypotension, tachycardia, dyspnea, RV dysfunction, tachypnea, and lactic acidosis (**Table 2**), the patient was taken emergently for surgical embolectomy. Catheter-based thrombolysis was considered but surgical embolectomy was chosen because of worsening hemodynamic instability, concern for failure of catheter-directed therapy given the size of the PE, and immediate availability of cardiothoracic surgery.[3] Systemic thrombolysis was deferred in favor of rapid transfer to the operating room. Surgeons removed massive central clots with extension into the hilar branches (**Figure 2**) and noted a mottled RV with very diminished function. Postoperative transesophageal echocardiogram showed severe RV dysfunction and dilation with normal left ventricular (LV) function (**Figure 3**).

QUESTION 2: WHAT ARE THE PRIMARY CAUSES OF PE IN CHILDREN?

Despite similar presentations, pediatric and adult PE have different etiologies. Risk factors for adolescent PE closely mirror risk factors in the adult population. This patient's unprovoked high-risk PE occurred in the setting of previously undiagnosed thrombophilia (antiphospholipid syndrome [APS]), obesity, and

TABLE 2 Arterial Blood Gases and Lactate on Arrival to the Emergency Department

pH arterial	6.86
pCO_2 arterial	86 mm Hg
PO_2 arterial	141 mm Hg
HCO3 arterial	16 mEq/L
Base excess arterial	−18 mmol/L
Lactate	12.8 mmol/L

The patient was critically acidotic and hypercarbic with lactemia indicating cardiogenic shock.
pCO_2 = partial pressure of carbon dioxide; PO_2 = partial pressure of oxygen.

infection. Additional risk factors for pediatric PE include sickle cell disease, neoplastic disorders, congenital cardiac or pulmonary defects, and oral contraceptives.[5]

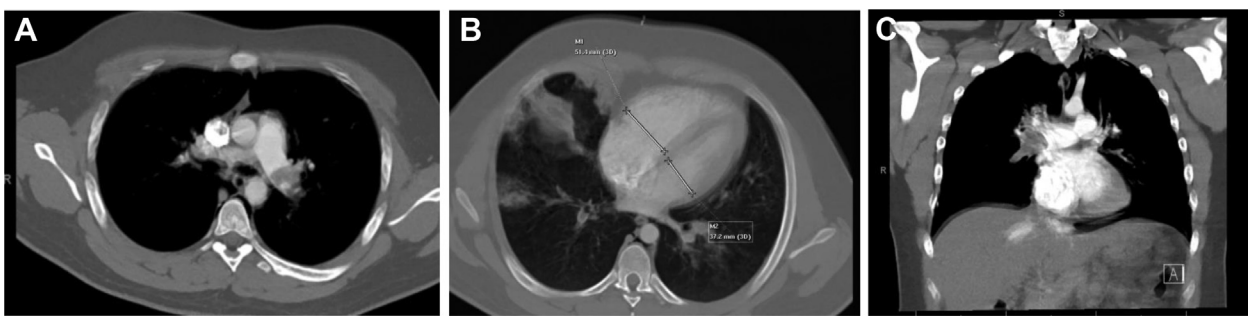
Evaluation for the cause of PE in this patient included testing for both inherited and acquired thrombophilia. This includes the antiphospholipid profile, lupus anticoagulant, anticardiolipin antibodies, and anti-beta2 glycoprotein antibodies. Inherited thrombophilia included genetic testing for Factor V Leiden and prothrombin gene mutations, and plasma-based activities of antithrombin and proteins C and S. The patient was found to have a positive lupus anticoagulant and positive cardiolipin antibody immunoglobulin M (206 units/mL). Although APS requires persistent presence of positive antiphospholipid antibody profile recorded at least 12 weeks apart, this patient was diagnosed with presumed APS in the context of acute thrombosis and positive antiphospholipid antibodies in addition to anemia with positive direct antiglobulin test and hypocomplementemia.

QUESTION 3: WHAT ARE ACUTE CARDIOPULMONARY COMPLICATIONS SEEN AFTER HIGH-RISK PE?

Immediate concerns after acute PE are RV failure, cardiogenic shock, and reperfusion injury with a 25% to 65% mortality rate for high-risk PE in adults with limited data in children.[6] The primary cause of death is RV failure due to massive and sudden increase in RV afterload. Guidelines define RV dysfunction based on an RV:left atrium diameter (apical 4-chamber) >0.9, RV systolic dysfunction, elevation of BNP or ProBNP, new right bundle branch block or anteroseptal ST-segment elevation/depression, or anteroseptal T-wave inversions. They further describe myocardial necrosis based on elevation of troponin I or troponin T.[1,7] In a study evaluating outcomes after surgical embolectomy, patients with high-risk PE were more likely to require ECMO before embolectomy, and more likely to require postoperative ECMO. Nonetheless, RV function normalized after embolectomy, with normalization of central venous pressure, pulmonary artery systolic pressure, and improvement in RV fractional area change.[6]

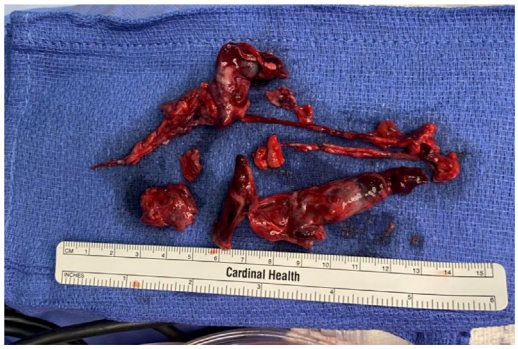
This patient had evidence of RV dysfunction at presentation with an RV:LV ratio by CT of 1.38 (**Figure 1B**) and by echocardiogram immediately after embolectomy with an RV:LV ratio of 1.32 (**Figure 3A**). RV:LV ratios represent an easy-to-implement tool to measure RV dysfunction and can be measured by

FIGURE 1 Axial and Coronal Computed Tomography Angiography Showing Massive Pulmonary Embolism (Right Ventricular:Left Ventricular Ratio = 1.38)



(A, C) Bilateral main pulmonary artery thrombi with extension into bilateral interlobar and segmental pulmonary arteries and extensive pneumomediastinum extending into the upper thoracic inlet and lower cervical epidural space. (B) right ventricle enlargement suggestive of right heart strain.

FIGURE 2 Surgical Embolectomy Pulmonary Embolism Specimen



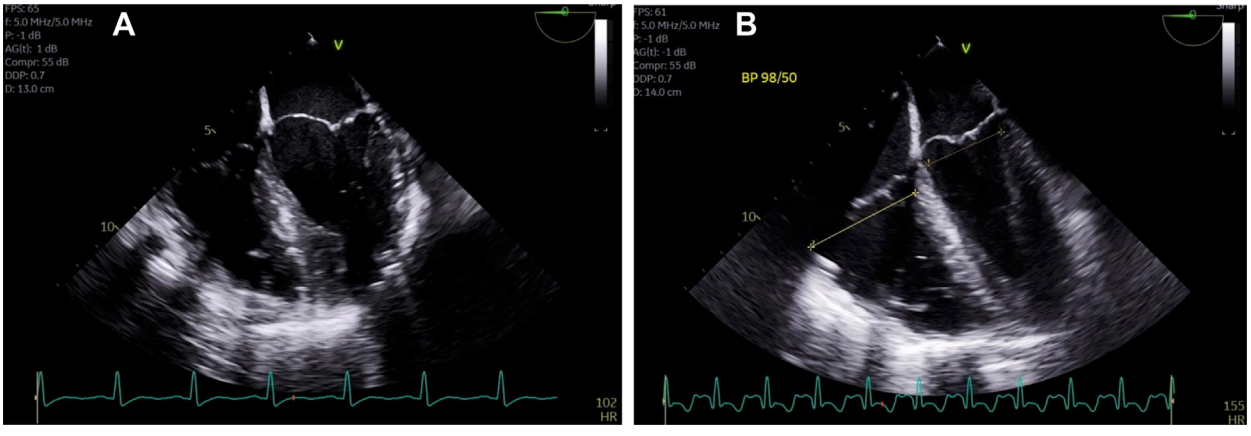
Surgeons removed massive central clots with extension into hilar branches bilaterally.

echocardiogram, CT, or cardiac magnetic resonance imaging; one study found that an RV:LV ratio >1 was 88% sensitive and 88% specific for diagnosing RV dysfunction compared with qualitative function evaluated on echocardiography.[8] The patient's RV likely experienced a period of ischemia due to extremely increased afterload coupled with poor coronary perfusion and myocardial necrosis (reflected by elevated troponins). There was likely pulmonary vasculature reperfusion injury in the setting of acute embolectomy, with severe hypercarbia and respiratory acidosis. The patient was placed on central venous-arterial ECMO due to RV failure and inability to ventilate, decannulating 72 hours later with normalization of RV function and improvement in RV:LV ratio to 0.74 at decannulation (Figure 3B).

QUESTION 4: WHAT ARE POTENTIAL COMPLICATIONS OF ANTIPHOSPHOLIPID SYNDROME AFTER MASSIVE PE AND SURGICAL EMBOLECTOMY?

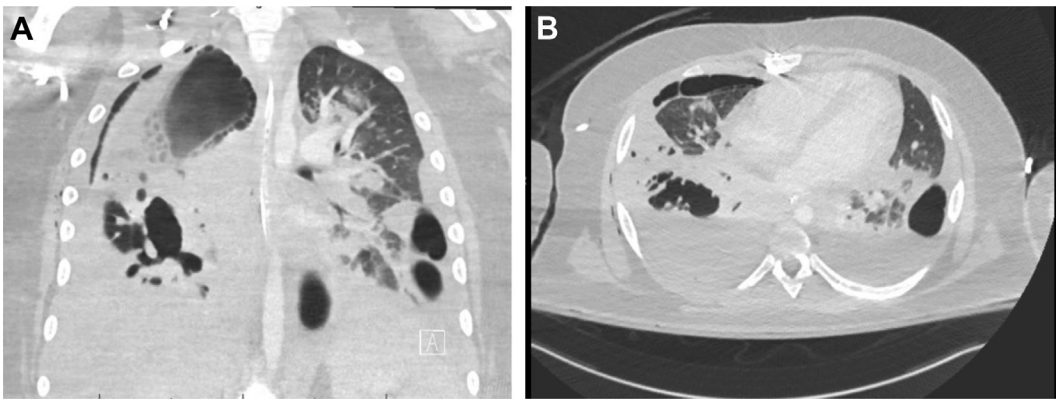
Antiphospholipid syndrome (APS) is a systemic autoimmune disease that can cause venous, arterial, or microvascular thrombosis. Catastrophic antiphospholipid syndrome (CAPS) is a rare but serious complication of APS characterized by diffuse microvascular thrombosis; infection and surgery are typical precipitating events. CAPS diagnosis is considered definite if the following criteria are met: 1) involvement of at least 3 organs, systems, or tissues; 2) symptoms that development over <1 week; 3) histologic confirmation of small vessel occlusion, and 4) the presence of antiphospholipid antibodies (anticardiolipin, anti-beta2-glycoprotein I, lupus anticoagulant) documented twice at least 12 weeks apart. Probable CAPS does not require histologic confirmation or laboratory evaluation. Pediatric CAPS is even more rare, with only 10% of patients in the international CAPS registry diagnosed before age 18. Pediatric CAPS differs from adult CAPS in that there is no female predominance and infection is a more

FIGURE 3 Postoperative 4-Chamber TEE and Post-ECMO Decannulation TEE



(A) Immediately postoperatively, the patient's RV:LV ratio was 1.32, consistent with significant RV dilation. (B) The patient was de-cannulated 72 hours later and repeat ECMO showed normalized RV function and an improved RV:LV ratio of 0.74. ECMO = extracorporeal membrane oxygenation; RV = right ventricular; RV:LV = right ventricular:left ventricular; TEE = transesophageal echocardiogram.

FIGURE 4 Axial and Coronal Computed Tomography Angiography

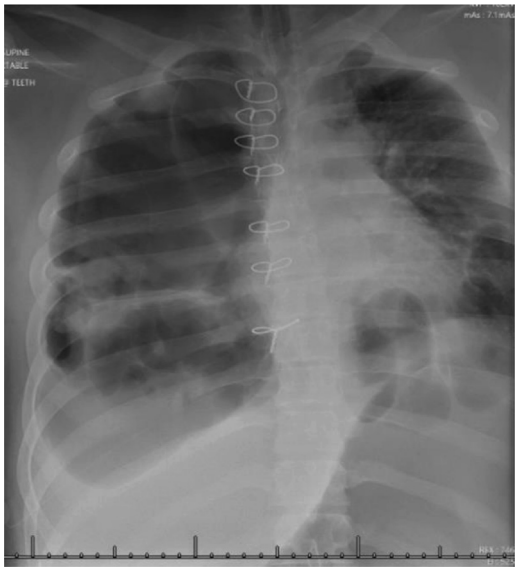


(A and B) Consolidative opacities in the right middle and lower lobes and dependent portion of upper lobe with necrotic changes, some with air-fluid levels. Consolidative opacities in the left upper lobe, lower lobe, and lingula with necrotic changes and air-fluid levels.

common trigger for the disease.[9] Children were also more likely to present with CAPS as a primary presentation of APS.

After decannulation from ECMO, the patient developed persistent fevers, hemolytic anemia, persistent tachycardia, hypertension, persistent bloody sputum, and evidence of renal and liver injury despite cardiac recovery. The multiorgan involvement and signs of end-organ failure raised concerns for CAPS. An interdisciplinary meeting (cardiovascular intensive care unit, hematology, rheumatology, cardiology, adult PE team) agreed on empiric CAPS treatment with pulse steroids, plasma exchange, and a course of rituximab given probable CAPS in a critically ill patient. With treatment, there was dramatic improvement in lung function, delirium, defervescence of fever, normalization of blood counts, and normalization of kidney and liver function.

FIGURE 5 Chest X-Ray



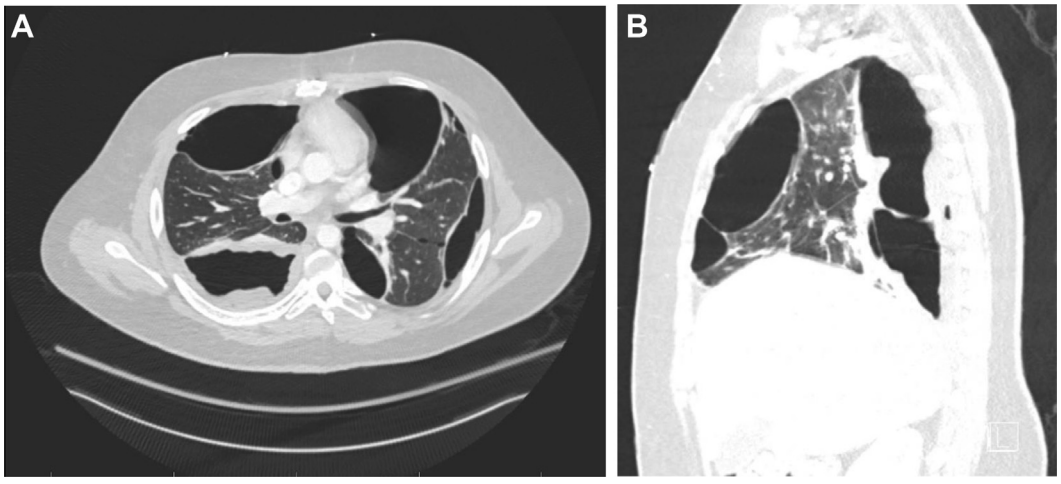
Moderate-large right pneumothorax with unusual lucencies in bilateral lung parenchyma possibly related to pleural air.

QUESTION 5: WHAT ARE THE RISK FACTORS AND ODDS OF CAVITARY PULMONARY INFARCTION FOLLOWING PE?

An estimated 10% to 50% of PE cases involve pulmonary infarction, despite the lungs being relatively protected against organ ischemia due to dual blood supply from the bronchial and pulmonary circulations.[10] This wide range is due to variable definitions of pulmonary infarction used in studies. Risk factors for pulmonary infarction include infection, surgical iatrogenic vasculitis, and smoking history. Interestingly, younger age (<40 years old) and taller height are associated with an increased likelihood of developing pulmonary infarction secondary to PE; however, obesity is associated with a decreased likelihood.[10]

Three weeks after treatment for CAPS, the patient developed cough and right scapular pain. CT scan demonstrated multifocal necrotizing pneumonia with large pneumatoceles and loculated pleural effusions

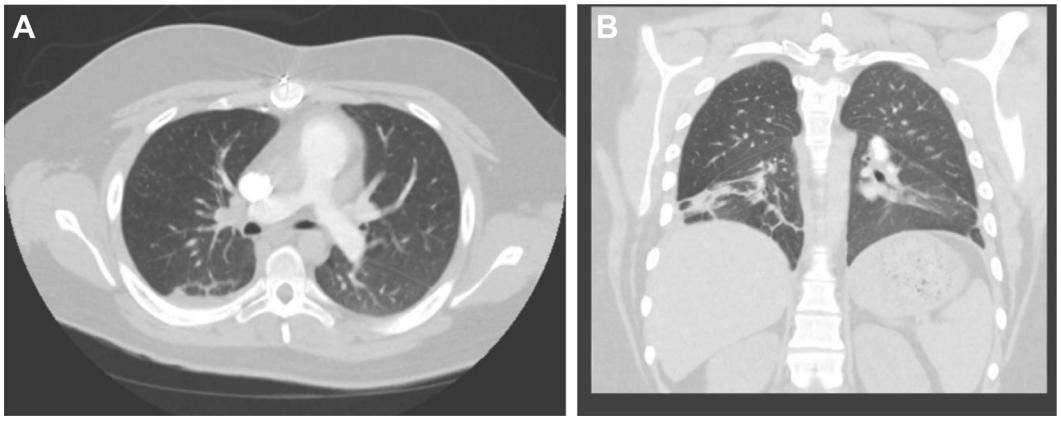
FIGURE 6 Axial and SagittalComputed Tomography Scans



(A and B) Bilateral hydropneumothoraxes with increased lucency overlying left lung base and decreased visualization of lung markings within the right lung base. Bilateral atelectasis and pleural effusions present.

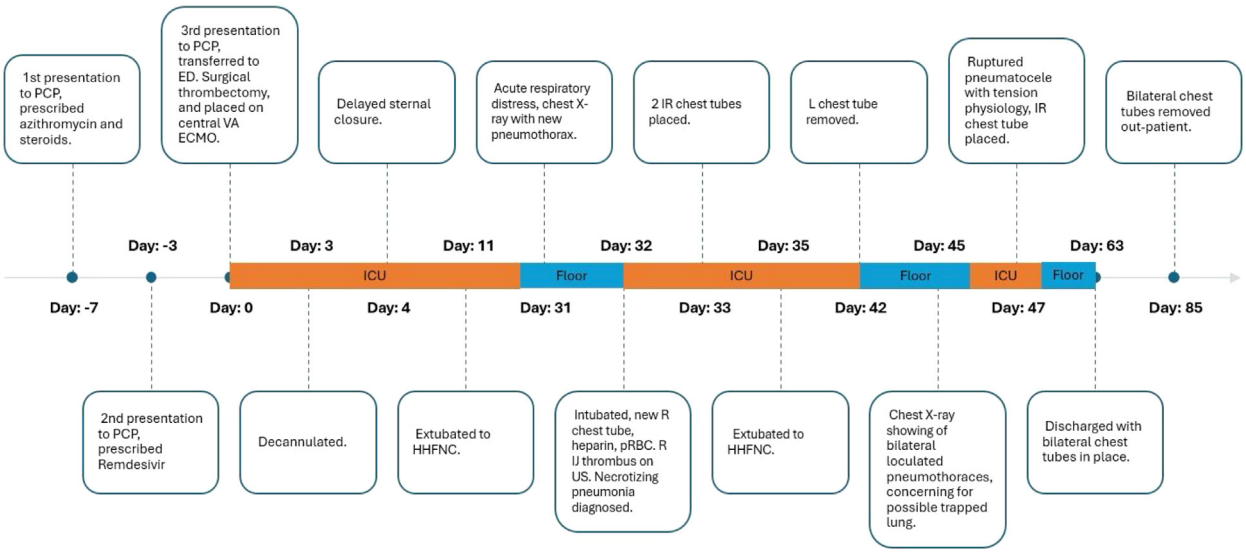
(Figure 4). The patient was re-intubated for respiratory failure in the setting of pneumonia and sepsis requiring vasoactive support. Bilateral chest tubes were placed with pleural fluid cultures positive for coagulase-negative staphylococcus. After recovery and later removal of the left chest tube, the patient developed tachypnea and left-sided scapular pain; X-ray (Figure 5) and noncontrast CT (Figure 6) revealed bilateral loculated pneumothoraxes with possible trapped lung. The pneumothorax rapidly progressed, either due to ruptured pneumatocele or bronchopleural fistula, and an emergent left chest tube was placed for tension pneumothorax with resolution of respiratory symptoms. Surgical

FIGURE 7 Axial and Coronal CT Following Chest Tube Removal



(A and B) CT scans at follow-up outpatient appointment reflect near resolution of bilateral pneumothoraxes with resolving bilateral cavitary lesions. CT angiography showed persistence of clot. CT = computed tomography.

FIGURE 8 The Timeline of This Patient's Complex Hospitalization



ED = emergency department; HHFNC = humidified high-flow nasal canula; IJ = internal jugular; IR = interventional radiology; L = left; PCP = primary care provider; pRBC = packed red blood cells; R = right; US = ultrasound; VA = venoarterial; other abbreviation as in Figure 3.

decortication was considered in consultation with pediatric and adult thoracic surgery. The team opted for conservative treatment with chest tubes, given that he was recovering from infarction and pneumonia, was still on a steroid taper, and not requiring respiratory support. The patient was discharged with chest tubes in place with close outpatient follow-up. Subsequent imaging after chest tube removal showed near complete resolution of pneumothoraxes (Figure 7). The patient has recovered remarkably well and is back to attending school full-time (Figure 8).

QUESTION 6: HOW IS THE PATIENT EXPECTED TO RECOVER FOLLOWING HIGH-RISK PE?

PE rates are increasing among children.[11] By extension, there is increasing emphasis on chronic sequelae of pediatric PE.[12] Despite anticoagulation and resolution of PE, limitations such as exercise intolerance and dyspnea on exertion, also termed the post-PE syndrome, occur commonly, making PE an important cause of disability in children who are otherwise expected to live several decades after the index event. Up to half of adult PE and up to one-third of pediatric PE survivors report persistent dyspnea, exercise intolerance, and/or functional limitations 3 to 6 months after acute PE.[12] Specific long-term outcomes following the 3 risk categories of PE are currently unknown. Despite apparent excellent recovery, our patient's exercise capacity as measured by peak oxygen uptake (VO_2) on maximal cardiopulmonary exercise testing at 4 months post-diagnosis was abnormally low (<80% of predicted) with high dyspnea ratings at submaximal or constant load cardiopulmonary exercise test, highlighting incomplete recovery and the paradox of resting tests that mask post-PE symptoms. Given his extended length of hospital and intensive care unit stay, deconditioning is a likely determinant of his low VO_2 but given persistent clot burden seen on follow-up CT, chronic thromboembolic disease and chronic thromboembolic pulmonary hypertension monitoring will be essential over time.

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