

Pulmonary Embolism in Children

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Abstract

Pediatric pulmonary embolism (PE) has become increasingly more common. It occurs most frequently in adolescent girls because of the effect of estrogen on blood coagulation. Prompt diagnosis is essential for lowering long-term complications and the mortality risk. However, this is challenging due to the varied and often asymptomatic clinical presentation of PE in children. On imaging, a filling defect within a pulmonary artery is the diagnostic finding. Imaging may also show signs of right ventricular strain, which helps inform treatment options. PE is treated based on risk and burden of disease. Potential treatments include anticoagulation, low-dose systemic thrombolysis, surgical embolectomy, or catheter-directed embolectomy.

Keywords: Vascular, Arterial, Pulmonary

Case Summary

An adolescent girl with a family history of blood clots presented to the emergency room with 1 week of dyspnea on exertion and pleuritic chest pain located in the center of her chest. On the morning of admission, she experienced sweating, jaw pain, and dizziness. The patient started taking an oral contraceptive pill 8 months prior to experiencing symptoms.

Imaging Findings

CT pulmonary angiogram (Figure 1) showed large filling defects within the right and left pulmonary artery extending into the lobar and segmental pulmonary artery branches. There was evidence of right heart strain, with enlargement of the right ventricle and straightening of the intraventricular septum.

Diagnosis

Pulmonary embolism (PE)

Discussion

PE is relatively rare in children but has increased in incidence by approximately 200% in the last decade,¹ with an estimated 8.6-57 cases per 100,000 hospitalized children.² Because PE in children is rare, often asymptomatic, and more variable in clinical presentation compared with PE in adults, diagnosis is often delayed or initially missed in children.³ Presenting symptoms are nonspecific and may include chest pain, dyspnea, hemoptysis, cough, syncope, as well as signs of deep vein thrombosis (DVT) and right heart failure.² Delays in treatment significantly increase the risk of complications, including

cardiac arrest, pleural effusion, recurrent venous thromboembolism, pulmonary hypertension, post-thrombotic syndrome, and death.^{4,5}

Among pediatric patients, PE most commonly affects adolescents, specifically female teenagers, due to the effects of estrogen on blood homeostasis.⁵ Additionally, pregnancy and the use of combined hormonal contraceptives are estrogen-related factors that contribute to the increased risk of venous thromboembolism and PE in the female adolescent population.⁵ Other risk factors for pediatric PE include hypercoagulability, presence of a central venous line, immobilization, and prior PE, or DVT.³

DVT is one of the most common risk factors for PE. DVT and PE are present concurrently in an estimated 54-72% of pediatric PE cases.⁶ In children with PE, additional symptoms caused by DVT may include tachycardia, tachypnea, and extremity edema.² Thrombi can be detected with Doppler ultrasonography and may be found in the popliteal vein, femoral vein, and iliac vein.⁶

Laboratory metrics used in the evaluation of PE in adult patients include elevated D-dimer, troponin I, and B-type natriuretic peptide levels.⁵

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Figure 1. (A, B) Axial standard images and (C) coronal max maximum intensity projection image from CT pulmonary angiogram showing filling defects (arrow) within the right and left pulmonary arteries. The filling defects extend to lobar and segmental branches of the pulmonary arteries. (D) Axial image at the level of the heart showing enlargement of the right ventricle and straightening of the intraventricular septum (arrowhead).

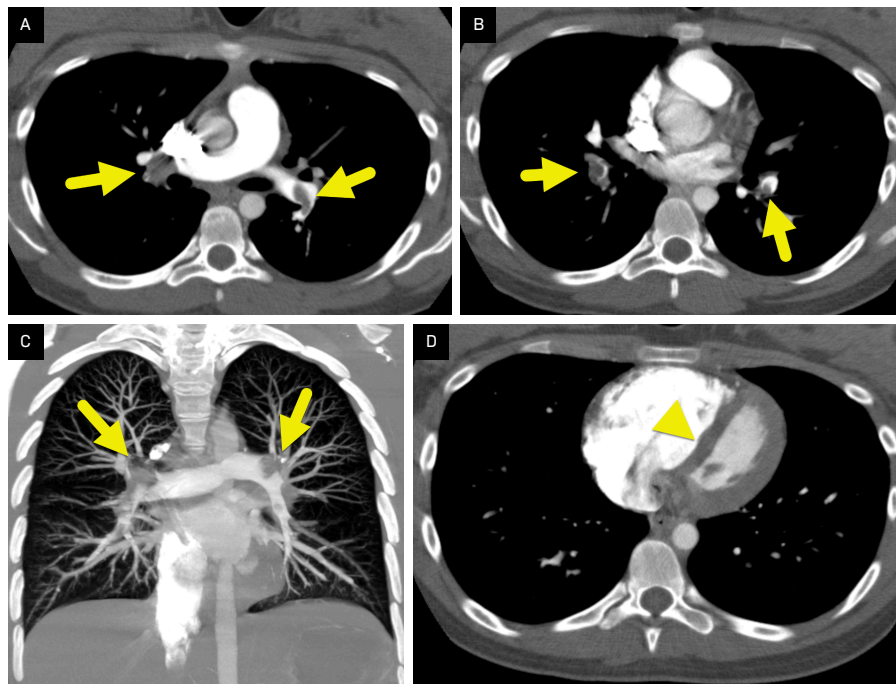


Table 1. Summary of Treatment Recommendations for High-Risk and Intermediate-Risk Pulmonary Embolism in Children

	Intermediate-Risk/Sub-Massive Pulmonary Embolism	High-Risk/Massive Pulmonary Embolism
Criteria	Acute pulmonary embolism without hypotension or compensated shock, but with evidence of right ventricular strain by imaging and/or of myocardial necrosis by elevated cardiac troponin levels	Acute pulmonary embolism causing cardiopulmonary arrest, sustained hypotension, or normotensive shock
Treatment options	• Parenteral anticoagulation • Low-dose systemic thrombolysis • Catheter-directed thrombolysis	• Parenteral anticoagulation • Primary reperfusion with systemic thrombolysis or surgical embolectomy • Catheter-directed thrombolysis • Catheter-directed embolectomy • Mechanical thrombolysis

However, these metrics do not consistently differentiate pediatric patients with PE from those without the disease and are therefore infrequently used in diagnosis.^{3,5} Further, the Wells Criteria and Pulmonary Embolism Rule-Out Criteria are used in diagnostic risk prediction of PE in adult populations, but lack both sensitivity and specificity in pediatric patients.⁵

Imaging is the most reliable diagnostic tool for detecting PE in children, and CT pulmonary angiography (CTPA) is the preferred imaging modality. An abnormal CTPA demonstrates a filling defect in one or more pulmonary arteries. In addition to the vascular filling defect, there may also be decreased or absent contrast enhancement of lung at the level of

the embolism.³ Peripheral wedge-shaped consolidation, characteristic of parenchymal ischemic injury, is often seen and is a more significant finding supporting the diagnosis of PE in children than in adults.⁷ Other features of PE on CTPA include pulmonary consolidation with surrounding ground-glass opacity.³ Right ventricular enlargement relative to the left ventricle may also be seen, indicating right ventricular strain.⁸ Additionally, flattening or bowing of the interventricular septum toward the left ventricle indicates right ventricular strain. Dual-energy CT is an emerging technology that may be more sensitive in detecting parenchymal enhancement and perfusion defects.³

Nuclear medicine has historically been used in PE diagnosis, and ventilation/perfusion (V/Q) scanning remains the supplemental modality of choice when CTPA is contraindicated or nondiagnostic. If PE is present, V/Q data show at least one wedge-shaped perfusion defect without a matched ventilation abnormality.⁹ However, difficulties in active inhalation of radiolabeled gas decrease the utility of this modality in young children.^{3,9}

Current treatment guidelines for pediatric PE are largely extrapolated from adult studies and include anticoagulation and thrombolytic therapy.⁴ Therapeutic recommendations are associated with PE risk stratification, which is summarized in Table 1.^{2,4,8} Patients with right ventricular strain on CTPA are categorized into the intermediate-risk/sub-massive PE category.⁸ Current guidelines do not recommend systemic thrombolysis for adults with intermediate-risk PE due to increased risk of bleeding and stroke.⁸ By contrast, a more aggressive approach may be warranted in children with intermediate-risk PE to mitigate the long-term consequences on chronic PE.⁸ Therefore, children in the intermediate-risk category may benefit from low-dose systemic thrombolysis with close monitoring to prevent complications like bleeding.

Patients who present with cardiopulmonary arrest, sustained hypotension,

or normotensive shock are categorized into the high-risk/massive PE category.⁸ Rapid primary reperfusion, with systemic thrombolysis or surgical embolectomy, is indicated for children with high-risk/massive PE. Catheter-directed thrombolysis or mechanical thrombectomy are minimally invasive interventional therapies used to lyse, aspirate, or fragment clot to relieve obstruction and re-establish blood flow.² These minimally invasive approaches are favored in high-risk patients and patients with failed pharmacologic therapies. Catheter-directed approaches have demonstrated improved outcomes in pediatric patient groups with both intermediate-risk and high-risk PE.^{2,4,8} In children with high-risk/massive PE, preliminary data have shown the efficacy of catheter-directed embolectomy in the removal of nearly all occlusive clots.⁴

Conclusion

Pediatric PE has become increasingly more common. It occurs most frequently

in adolescent girls because of the effect of estrogen on blood coagulation. Prompt diagnosis is essential for lowering long-term complications and the mortality risk. However, this is challenging due to the varied and often asymptomatic clinical presentation of PE in children. On imaging, a filling defect within a pulmonary artery is the diagnostic finding. Imaging may also show signs of right ventricular strain, which helps inform treatment options. PE is treated based on risk and burden of disease. Potential treatments include anticoagulation, low-dose systemic thrombolysis, surgical embolectomy, or catheter-directed embolectomy.

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