



Progressive Shortness of Breath and Hypoxia in an Adolescent Female Athlete: A case report

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History of Presenting Illness

Patient:

Previously healthy 16 year-old female

Chief Complaint:

2 months of progressive shortness of breath

History:

- First noticed symptoms at basketball practice after period of inactivity due to COVID
- Patient reports dyspnea with minimal exertion
- SpO₂ in 80s on home pulse oximeter following exertion
- 40 lb unintentional weight loss over same time period

Physical Exam and Case Details

Physical Exam:

- Vital signs significant for SpO₂ 87-88% with exertion, SpO₂ 90-91% with rest
- Cardiovascular: RRR, no murmurs or rubs, strong radial pulses
- Respiratory: normal work of breathing, clear to auscultation, no wheezes or crackles
- Extremities: warm and well perfused, no edema

Case Details:

- Initial laboratory work up reveals elevated D-dimer. CT scan confirms bilateral pulmonary emboli with right heart strain confirmed on echocardiogram. Ultrasound of extremities reveals a non-occlusive VTE in her left arm consistent with Paget-Schroetter Syndrome.
- Elevated cardiolipin IgG and elevated PTT raise concern for anti-phospholipid syndrome (APS).
- The patient is started on anticoagulation with plan to taper dose at home

6 weeks later:

- Patient presents with worsening dyspnea following reduction of anticoagulant dose
- CT angiogram initially described as bilateral ground glass opacification with improvement in thromboembolic clot burden.
- Over read of CT scan, cardiac catheterization and V/Q scan reveal underlying diagnosis

CTEPH is rare in pediatric patients, although it is frequently associated with an underlying cause such as PSS or APS.

Children with CTEPH and/or PSS should undergo thorough evaluation for hypercoagulable disorders given the high co-incidence in pediatric patients.

Diagnostic Evaluation:

Labs:

Initial Presentation

- D-dimer: 3.8 (ref range <0.5)
- Cardiolipin Antibody IgG: 27 (ref range 0-14)
- PTT: 44.1 (ref range 28-37)
- Histone AB IgG: 2.4 (ref range <0.9)
- Labs within normal limits: Cardiolipin Antibody IgM, Anti thrombin III: 116, Protein C antigen, Protein S antigen, PT/PTT Lupus anticoagulant, ANA, Factor V Leiden, Fibrinogen AG

6 weeks later:

- Cardiolipin AB IgM 14 (ref range <13)
- Labs within normal limits: D-dimer, BNP, CRP, ESR, SARS-COV-2 AB IgG, ANA, ANCA, Von Willebrand antigen, M. Pneumonia AB IgG

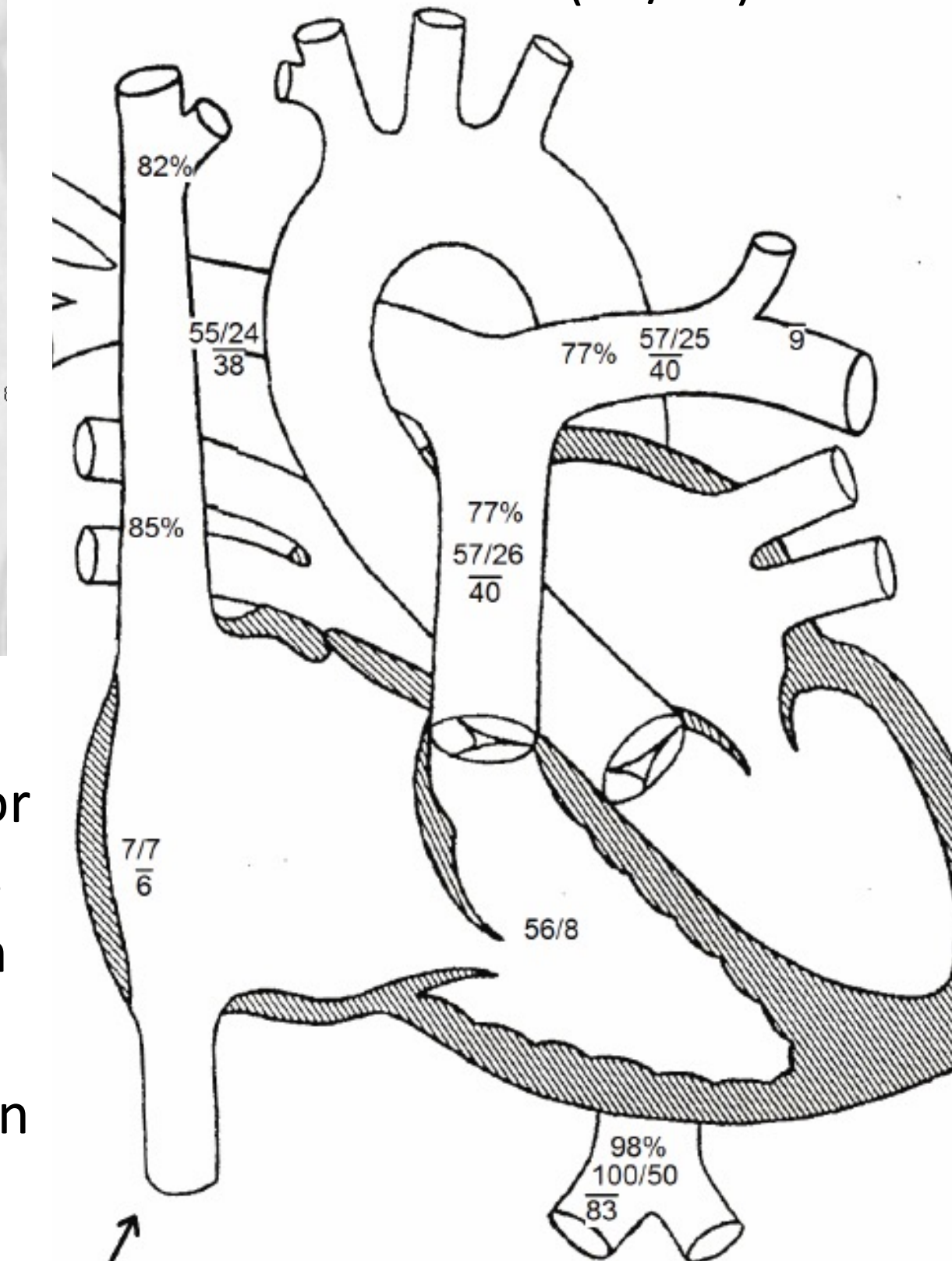
Imaging:



CT Scan (above): Chronic pulmonary embolism with persistent clot in the posterior basal right and lateral basal left lower lobes with multifocal areas of increased perfusion

V/Q Scan: mismatched ventilation/perfusion in posterior and lateral basal segments compatible with areas of known chronic pulmonary embolism

Cardiac Catheterization(below): Moderate pulmonary hypertension (57/26) that responds slightly to nitric oxide (43/20)



CONCLUSIONS

- CTEPH occurs when an acute thromboembolus transforms into a fibrotic mass instead of resolving- this can result in proximal pulmonary vessel obstruction which can lead to pulmonary hypertension, right heart failure and death.
- CTEPH is rare in pediatric patients and is frequently associated with an underlying cause, such as PSS or hypercoagulable state.
- Children with CTEPH should undergo thorough evaluation for hypercoagulable disorders given high co-incidence in pediatric patients.
- Treatment of CTEPH requires life-long anti-coagulation and pulmonary thromboendarterectomy

ADDITIONAL KEY INFORMATION

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