Pediatric Cases

Kentucky Coalition of Nurse Practitioners and Nurse Midwives 31st Annual Coalition Conference
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PRIMARY CARE
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Objectives

- Distinguish atypical presentations of pediatric diagnoses and early recognition.
- Integrate lessons learned from interesting cases into practice.

Case Presentation

- 6y 2m Hispanic male
- CC: neck pain for 4 days
- Not turning head to the right
- c/o not feeling well at school
- Mild headache, no blurry vision
- No trouble swallowing or breathing, throat is not sore
- No fever, N/V/D
- Attends daycare; exposure to strep
- VS: T 37.1 (98.7), HR 112, RR 22
- 14 point ROS otherwise negative except as noted in HPI

Physical Exam

- General: NAD, Alert and active, Non-toxic
- HEENT: Head normocephalic and atraumatic. PERRL, EOMI. TM's pearly grey with LR and LM visible. FROM of neck; no meningeal signs. Oropharynx without erythema or exudates.
- CV: RRR; no murmurs, rubs or gallops.
- Lung: CTA bilaterally with good air movement. No wheezes, rhonchi, or rales.
- Lymph: Palpable, tender cervical nodes, no erythema or warmth
- Abdomen: Soft, non-tender, non-dilated, active bowel sounds, no masses or hepatosplenomegaly.
- Extremities: No clubbing, cyanosis, or edema. No obvious deformity or bony tenderness. 2+ peripheral pulses.
- Skin: WDI. No rashes or lesions. Cap refill < 2 sec.
- Neuro: Age appropriate; moving UE and LE bilaterally.

Differential Diagnoses

- Viral illness
- Strep Throat
- Lymphadenitis

Plan of Care

- Rapid Strep Test: Negative
- Strep Culture sent
- Tylenol or ibuprofen for pain
- Encourage fluids
- RTIC: fever, increased lymphadenopathy (increase in size, erythema, warmth), no improvement in 3-5 days.
RTC next day

- **HPI:** Sore throat for 5 days with fever. Spiked temp to 103 last night. PO intake 30% of normal. Continues with left-sided neck pain.
- **PE:** No change from previous day.
- **Differential DX:** Mononucleosis, strep, viral illness.
- **POC:** Mono spot (negative), CBC (WBC 26.8). Strep culture negative. Continue supportive care.

RTC 2 days later

- **HPI:** Continues with fever up to 103 (now 7 days). Swelling around right eye developed last night. Mom thinks the eye looked a little red yesterday. No cough or congestion, no diaphoresis. Child's primary complaints are slight sore throat, headache and fever.
- **PE:** Right-sided periorbital edema and mild conjunctivitis. Left-sided cervical lymphadenopathy. Dried, erythematous cracked lips. No swelling of hands or feet. No rash.
- **Differential DX:** Kawasaki Disease, pneumonia, viral URI
- **POC:** CBC (WBC 14), CMP (normal), CRP (4.0), ESR (118)
  - Consulted with Peds ID—sent to KCH ED for further evaluation
  - Admitted to KCH: DX Atypical Kawasaki Disease

**KAWASAKI DISEASE**

- Acute self-limiting inflammatory disorder.
- Associated with vasculitis, affecting predominantly medium-sized arteries, particularly the coronary arteries, causing coronary artery aneurysms (CAA) in 15–25% of untreated patients while 2–3% of untreated cases die as a result of coronary vasculitis.
- The second commonest vasculitic illness of childhood after Henoch Schönlein purpura. In developed countries KD is the commonest cause of acquired heart disease in childhood.
- Etiology remains unknown. Currently believed that one or more as yet unidentified infectious agents induce an intense inflammatory host response in genetically susceptible individuals.
  - World-wide distribution with a male preponderance, an ethnic bias towards Asian children (particularly East Asian), seasonality and occasional epidemics.

**Kawasaki Disease Diagnosis**

- **COMPLETE KD**
  - Fever
  - 4 of 5 symptoms
- **INCOMPLETE KD**
  - <3 symptoms
  - Symptoms may present successively not simultaneously

**Kawasaki Disease Diagnosis**

- persistent fever (> 5 days) in combination with
  - polymorphous exanthema,
  - cervical lymphadenopathy,
  - non-purulent conjunctival injection,
  - changes of the lips and oral cavity (including strawberry tongue, cracked lips, redness of the mucosa), and
  - changes in extremities (swelling and redness of the palms, desquamation in the subacute phase)
Clinical Course of KD

(1) ACUTE—the period lasting 1-2 weeks if untreated, when the child has a spiking, often remittent 40°C Celsius fever and principal symptomatic features and may present with cardiac manifestations including valvitis, pericarditis, and myocarditis

(2) SUBACUTE—the approximately 2 week period following the abatement of fever when the child is at the greatest risk of sudden death due to myocardial infarction

(3) CONVALESCENT—the clinically invisible period following the cessation of symptoms and continuing until acute-phase reactants return to normal serum levels

(4) CHRONIC—which describes patients who require follow-up management due to coronary artery involvement

 PRIMARY CARE

Immunization with live viral vaccines should be deferred for at least 11 months following an episode of KD treated with IVIG

Due to the potential lack of effectiveness of live vaccines following IVIG and the potential for any vaccine to induce potentially detrimental immune activation during the convalescent phase of KD.

Thereafter, all vaccines should be administered as recommended by national schedules.

Inactive immunizations are unaffected by serum IVIG

Check cholesterol levels every 5 years

Kawasaki Disease

What is known:

- Kawasaki disease (KD) is a pediatric vasculitis with coronary artery damage as its main complication.
- CAA in 25% of untreated cases
- Although KD approaches its 50th birthday since its first description, many aspects of the disease remain poorly understood.

What is new:

- In recent years, multiple genetic candidate pathways involved in KD have been identified, with recently promising information about the RPRC pathway.
- As increasing numbers of KD patients are reaching adulthood, increasing information is available about the long-term consequences of coronary artery damage and broader cardiovascular risk.

References


Interesting Pediatric Cases

Kentucky Coalition of Nurse Practitioners and Nurse-Midwives
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Vicki Stringfellow, MSN, APRN

Breathless in Kentucky
Case #1
16 year old female with 2 day history of shortness of breath and palpitations
PMH: vasovagal syncope, exercise-induced asthma, obesity, polycystic ovarian syndrome
Home medications:
- Combined oral contraceptives, started 2 months ago for PCOS
- PRN albuterol for EIA, last used today
Social: sedentary lifestyle; denies tobacco use, no smokers in home
Family: maternal uncle and grandfather died before age 50 years with "heart disease"

Physical Exam
- Afebrile; HR 99; RR 16
- BP 124/82
- Wt 83.3kg (> 95th percentile; < 97th percentile)
- BMI 29.5 (> 95th percentile; < 97th percentile)
- SpO2 in room air: 91% (placed on nasal cannula at 2LPM)
Obese adolescent female, anxious appearing, mild dyspnea. Skin color pale pink. Breath sounds clear, HR regular without murmur. Distal pulses normal, hands clammy and cool to touch. No edema or discoloration of extremities. Abdominal exam unremarkable and GU exam deferred.

Differential Diagnosis
- Neuro/CNS/psych: Panic attack
- Chest/ pulmonary: Asthma exacerbation, Pleuritis, Pneumonia/ bronchitis, Pneumothorax, Foreign body aspiration, Malignancy/ mass
- Chest/ cardiac: Pericarditis, Cardiomyopathy, Vascular: Pulmonary embolism, Musculoskeletal: Costochondritis

Narrowed Differential Diagnosis and Initial Diagnostics

Diagnostics Results
- CXR unremarkable; ECG normal; Serum bHCG negative
- Troponin: < 0.01 ng/mL (reference <0.04 ng/mL)
- D-dimer: 300 ng/mL (reference range < 250 ng/mL)
- BNP: 147 pg/mL (reference range 0-178 pg/mL for 15-17 year old female)
- Echo: 1. Qualitatively normal biventricular size and function; normal LV size and wall thickness
2. Trivial tricuspid regurgitation; inadequate to estimate right ventricular systolic pressure.
3. Mild systolic septal flattening noted consistent with elevated right ventricular pressure.
- CTA 1. Bilateral filling defects in the left greater than right lower lobe pulmonary arteries; concerning for pulmonary emboli.
2. Left lower lobe subsegmental atelectasis versus infarcts.
3. Small left pleural effusion

Case #2
17 year old male with chest pain and difficulty breathing; symptoms worse with lying down, became dizzy and nauseated upon sitting up. Rates pain 8/10.
PMH: shoulder injury 1 month ago
PSH: outpatient orthopedic procedure left shoulder 1 week ago
Home medications: none
Social: plays high school baseball; denies tobacco or substance use
Family: negative for cardiac disease, respiratory disease, seizure disorder
Physical Exam

- T 36.6C/ 97.9F; HR 118; RR 22
- BP 100/65
- Wt 71kg (156lbs) (> 50th; < 75th percentile)
- Ht 175 cm (5'9") (50th percentile)
- BMI 23 (> 50th; < 75th percentile)
- SpO2 in room air: 88% (placed on non-rebreather)

Adolescent male; pale, diaphoretic with significant tachypnea and dyspnea. Anxious, able to speak few words at a time. Pupils 3mm, equal and react to light. Breath sounds clear, HR regular without murmur. Central pulses 2+, distal pulses 1+. Extremities without edema or discoloration.

Differential Diagnosis

- Neuro/CNS/psych/behavioral
  - Panic attack
  - Substance use
- Chest/ pulmonary
  - Pleuritis
  - Pneumonia/bronchitis
  - Foreign body aspiration
  - Malignancy/ mass
- Vascular
  - Venous thromboembolism
  - Pulmonary embolism

Narrowed Differential Diagnosis and Initial Diagnostics

<table>
<thead>
<tr>
<th>Potential diagnosis</th>
<th>Initial diagnostic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neuro/psych: Panic attack</td>
<td>None / exclusion</td>
</tr>
<tr>
<td>Chest: Malignancy/ mass</td>
<td>CXR</td>
</tr>
<tr>
<td>Chest/ cardiac: Arrhythmia Myocardial ischemia</td>
<td>Echocardiogram Electrocardiogram</td>
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<tr>
<td>Vascular: Pulmonary embolism</td>
<td>Chest CT angiogram D-dimer</td>
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Diagnostics Results

- CXR: potential enlargement of pulmonary artery
- Troponin: 0.1 ng/mL (reference <0.04 ng/mL)
- D-dimer, BNP: insufficient sample
- Echo: unable to complete prior to intervention
- ECG: 1. Sinus tachycardia
  1. Large bilateral pulmonary emboli with widening of the main pulmonary artery
  2. Left lower lobe subsegmental atelectasis vs infarct
  3. Small left pleural effusion

Risk Factors for Pediatric VTE

- Inherited/congenital
  - Factor V Leiden mutation, protein C or S deficiency, decreased serum homocysteine
  - Congenital heart disease, May-Thurner syndrome
  - Family history
- Acquired
  - Central venous catheters
  - Trauma or major surgery, particularly if orthopedic
  - Malignancy, specific chemotherapy agents
  - Tobacco use
  - Immobility, dehydration, infections
  - High estrogen states: pregnancy and estrogen-containing contraceptives
  - Obesity
  - Nephrotic syndrome
  - Antiphospholipid antibodies
  - Inflammatory conditions (Crohn disease, rheumatoid arthritis, systemic lupus erythematosus)
Pediatric PE

- Reported incidence from 8.6 to 57/100,000 hospitalized children; 0.14 to 0.9/100,000 all children (1,2)
- Bimodal distribution; higher incidence in infants < 1 year of age and in adolescents (2)
  - Central venous catheters (CVC) implicated among infants
  - Pregnancy and hormonal contraceptives implicated in adolescent females
  - Inherited thrombophilias common in adolescents without CVCs
- A majority occur in the context of a serious illness Approximately 2/3 associated with clinically apparent DVT at another location (1,2)

Clinical Presentation of Pediatric PE

- Most common presenting signs and symptoms include shortness of breath, pleuritic chest pain, cough, and hemoptysis
  - Additional signs and symptoms include tachypnea, tachycardia and hypoxemia
  - Diagnostic challenge due to multiple potential etiologies for these

Diagnostic Assessment

- Initial:
  - Point of care: pulse oximetry, ECG
  - Labs: D-dimer, troponin, BNP
  - Imaging: CT pulmonary angiography; less commonly nuclear medicine ventilation/perfusion scan or MR angiography
  - Pregnancy test for post-menarchal females
- Further testing to guide management and investigate for underlying cause
  - CBC, CMP, coagulation profile; ABG (if hypoxemic or significant distress)
  - Thrombophilia screening
  - Imaging: Doppler ultrasound extremities

Treatment

- Treatment is based primarily on evidence from adults; formal guidelines addressing children include
  - 2012 American College of Chest Physicians (ACCP) guidelines
  - 2018 American Society of Hematology (ASH) guidelines

Treatment: Immediate Stabilization

- Tailored to presentation; treat emergent situations by advanced life support guidelines
- Pharmacologic
  - Initial anticoagulation with heparin or low-molecular-weight heparin (LMWH)
  - Transition to vitamin K antagonist; some may continue LMWH
  - Newer anticoagulant therapies; data for pediatrics is sparse but extrapolation to adult-sized older adolescents is reasonable
- Mechanical
  - Thrombolyis, thrombectomy, or embolectomy for massive PE with associated hemodynamic instability

Treatment: Continued Therapy

- Current recommendation for duration of anticoagulation therapy included in the available guidelines
  - 6-12 months for unprovoked PE (1,2)
  - 3 months or until precipitating risk factor is resolved for secondary (provoked) PE (1,2)
Continued Pharmacologic Therapy

- Vitamin K antagonist, warfarin (Coumadin)
  - Oral administration
  - Requires dietary consistency
  - Monitoring: prothrombin time (PT)/ international normalized ratio (INR)
    - typical therapeutic range 2-3
- Low molecular weight heparin, enoxaparin (Lovenox)
  - Subcutaneous injections twice daily
  - Monitoring: anti-Xa levels
    - typical therapeutic range 0.5 - 1 IU/mL

Outcomes and Case Progression

- Overall mortality reported at approximately 10% for pediatric PE\(^{(1)}\)
- Reported recurrence rates range from 7% – 18%\(^{(1)}\)
- Case progression/ outcome
  - Case #1
    - Hormonal contraception stopped. Initially managed with heparin infusion, transitioned to enoxaparin, then to warfarin prior to discharge
    - Outpatient management continued by pediatric hematology: referred to high BMI clinic
  - Case #2
    - Arrest event in ED: resuscitated, emergent thrombectomy, catheter-delivered anticoagulation, ECMO. After decannulation, safety of heparin infusion, transitioned to enoxaparin and then to warfarin.
    - Outpatient treatment continued with pediatric hematology.
    - No inherited thrombophilia identified: attributed to surgery and immobility.

Summary

- VTE and PE are increasing in the pediatric population
- Pertinent modifiable risk factors among adolescents include obesity, tobacco use, and hormonal contraceptive use in females
- Prompt diagnosis improves outcome and requires a high level of suspicion
- Care is multidisciplinary across inpatient and outpatient settings

References


