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HEMATOLOGY/ONCOLOGY

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Disclosures

Albrey Berber DNP, CPNP-PC

- Has no financial relationship with commercial interests
- This presentation contains no reference to unlabeled/unapproved uses of drugs or products

Learning Objectives

Upon completion of this review, the course attendee should be able to:

- Describe the process of history taking and physical assessment of hem/onc concerns
- Summarize common diagnostic tests (laboratory and radiology) utilized when evaluating a hem/onc
- Compare and contrast the pathophysiology, clinical presentation, management, and follow-up of the most common hem/onc diagnoses seen in primary care
- Describe education needs related to the most common hem/onc

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Presentation Outline

I. History and Physical Assessment

II. Common Diagnostic Tests

III. Pathophysiology, Clinical Presentation, Management and Follow-up

IV. Education Needs

Hematologic Disorders

- **Erythrocyte Disorders**
  - Anemia
    - Iron-deficiency
    - Thalassemia
  - Sickle Cell Disease
  - Hereditary spherocytosis
  - Lead poisoning

- **WBC Disorders**
  - Leukemia
  - Lymphoma (Non-Hodgkin, Hodgkin)

- **Platelet/Coagulation Disorders**
  - Idiopathic Thrombocytopenic Purpura (ITP)
  - Hemophilia A, B, & von Willebrand Disease
  - Henoch-Schonlein purpura (HSP)

Physical Examination

- Head to toe examination
- Growth parameters, including BMI
- Pallor
- Jaundice
- Petechiae
- Bleeding
- Lymphadenopathy
- Joint or extremity pain
- Heart murmurs
- Hepatomegaly/splenomegaly

Common Diagnostic Tests

- CBC
- PT, PTT, INR
- Reticulocyte count
- Peripheral blood smear
- Ferritin, Fe, TIBC
- Hbg electrophoresis
- Bilirubin, LDH

Iron-Deficiency Anemia

- **Key Characteristics:**
  - Most common anemia in childhood
  - RBC or Hemoglobin concentration > 2 standard deviations below mean in healthy children
  - Routine screening between 9-12 months
  - Common between 1-3 years of age due to inadequate dietary iron due to cow's milk as major staple in diet

- **S/S:**
  - Mild anemia (Hgb 9.5-11) may be asymptomatic
  - Sometimes only minimal S/S w/severe anemia (<8-9.5)
  - Hx of fatigue, irritability, excessive milk intake, headaches
  - Mild (normal PE)
  - More severe
    - Poor weight gain
    - Sclera or palmar pallor
    - Splenomegaly (15%)
    - Tachycardia
    - Systolic flow murmurs w/progression

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Evaluation of Iron-Deficiency Anemia

IDA confirmed by the following:

- CBC:
  - RBC: microcytic & hypochromic
  - MCV (decreased)
  - Ratio of MCV/RBC > 13 (Mentzer Index)
    - >13 = IDA
    - <13 = Thalassemia trait
  - Serum ferritin (decreased)
  - Serum iron (decreased)
  - Total Iron binding capacity (increased)
  - Iron saturation (decreased)

Management of Iron-Deficiency Anemia

- **Nutritional strategies:**
  - Breast and/or formula until 1 year of age
  - Reduce milk to no more than 16-24 oz/day at 1 year of age
  - Increase intake of high-iron foods (dark green veggies, beans, whole cereals, pork, beef
- **Ferrous Sulfate: Iron therapy for infants & children:**
  - **May start as early as 2 months of age in premature infants & 4-6 months for term infants**
  - **Mild to moderate IDA:** Elemental iron 3/mg/kg/day in 1-2 divided doses
  - **Severe IDA:** Elemental iron 4-6 mg/kg/day in 2-3 divided doses

- Recheck Hemoglobin in 1 month
- Treat until H & H reaches normal ranges, then give at least 1 month additional to replenish iron stores
- If response is not adequate in 1-2 months consider further diagnostic testing for GI bleeding or other anemias

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Thalassemia

- **Key Characteristics:**
  - Inherited anemia that affects M/F and can be mild/severe
  - Alpha or Beta
  - Included on the NBS
  - Most often in Italian, Greek, Middle Eastern, Asian, & African descent

- **S/S:**
  - No symptoms if Alpha/silent carrier
  - May/may not have S/S w/mild anemia (most often fatigue)
  - S/S of severe anemia due to Thalassemia occur the first 2 years of life & include other health issues
  - Mild (Normal PE)
  - Severe: Pale, poor appetite, dark urine, jaundice, liver, spleen, & heart enlargement, bone problems (facial), slow growth

Thalassemia

- **Evaluation:**
  - CBC w/diff
  - Iron tests: To r/o IDA
  - Hemoglobin electrophoresis testing
  - Genetic studies

- **Management:**
  - **Mild:** usually needs no treatment, but education & genetic counseling for family to prevent need for iron supplements throughout lifespan
  - **Mod/Severe: Referral to Hematologist,** regular blood transfusions, Iron chelation therapy (transfusions can lead to build up of iron), Folic acid for retic > 3%: B vitamin that helps build healthy RBCs
  - Bone marrow/stem cell is only cure

Sickle Cell Disease

- **Key Characteristics:**
  - Autosomal-recessive heme disorder (single gene from both parents)
  - Sickle shaped hemoglobin
  - Predominantly in AA (also found in Mediterranean & Arabic)

- **Evaluation:**
  - Included on Newborn Screening
  - Electrophoresis
  - Repeat resting w/abnormal hemoglobin pattern on NBS
  - CBC w/hemoglobin MCV

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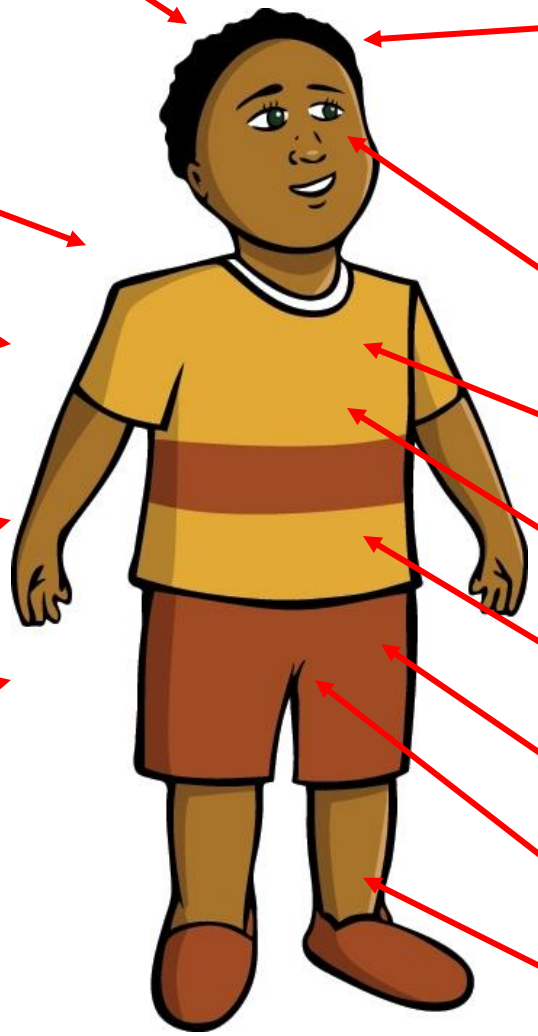
Sickle Cell Disease

- **S/S:**
  - Chronic hemolytic anemia: Pallor, jaundice, tachycardia, fatigue
  - Vaso-occlusion results in ischemia to tissues
    - Painful crisis: Infarcts of muscle, bone, bone marrow, lung, intestines
  - Cerebrovascular accident
  - Acute Chest Syndrome
  - Priapism
  - Ocular retinopathy
  - Gallbladder disease
  - Splenomegaly
  - Cardiomyopathy
  - Growth failure
  - Dactylitis (swelling of the hand/feet)
  - Psychological problems (due to chronic illness)

Complications of Sickle Cell Disease



Psychological problems

Hematology (anemia, leukocytosis)

Growth and maturation

Pain

Infections

Stroke, silent infarcts, and decreased neuro-cognitive function

Retinopathy

Acute chest syndrome

Pulmonary hypertension

Splenic function

Avascular necrosis in bone

Priapism

Leg ulcers

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Sickle Cell Disease

- **Management:**
  - **Children with SCD are co-managed w/PCP & Hematology**
  - Education to families, Adequate fluid intake, immediate care for fevers **(101 fever is an emergency & sepsis until proven otherwise)** importance of prophylactic treatment
  - Penicillin Prophylaxis:
    - 2 mo-3 yrs: PCN VK 125 mg PO BID
    - 3-5+ yrs: PCN VK 250 mg PO BID
  - Hydroxyurea (HU) therapy: Recommended to start HU at 9 months of age to prevent painful crisis
  - Pain medications: Tylenol/NSAIDS (mild) Tylenol w/codeine (mod) Morphine (severe)

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Sickle Cell Disease

- **Emergency admission/referral is necessary in the presence of the following:**
  - Fever (to r/o **Sepsis**) greater than 101 (38.3)
  - Pneumonia, chest pain, or other pulmonary symptoms (**Acute Chest Syndrome**)
  - **Sequestration crisis** (Splenomegaly w/decreased Hgb or Hct)
    - Sudden trapping of blood in the spleen
    - Left sided abdominal pain (Abdominal distention)
    - May be associated w/ fever, pain, vomiting, dyspnea
    - Shock
    - Circulatory collapse & death can occur in less than 30 min
  - **Aplastic crisis** (Decreased Hct & retic count)
  - **Severe painful crisis**
  - **Unusual headache, visual disturbance**
  - **Priapism**

Sickle Cell Disease

- **Routine Well Child Care:**
  - Birth to 6 months: Visit every 2 months
    - CBC every 2 months
  - 6 mo-2 years: Visit every 3 months
    - CBC every 3-6 m, UA annually
    - Ferritin, TIBC, BUN CREAT, LFTs once at 1-2 years
    - Influenza vaccine annually
    - Start folic acid 1mg/day at 1 year or when retic >3%
  - 2-5 years: Visits every 6 months
    - CBC and UA annually
    - BUN, CREAT, LFTS every 1-2 years
    - 23-Valent pneumo vaccine at 2 year w/booster at 5 years
  - Older than 5 years: Visits every 6-12 months
    - CBC, UA annually
    - BUN, CREAT, LFTS every 2-3 years
    - Continue folic acid
    - May opt to stop PCN V if titers indicate adequate protection

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Hereditary Spherocytosis

- **Key Characteristics:**
  - Hemolytic anemia characterized by a deficiency or abnormality of the RBC membrane protein, reduces the RBC surface area (more spherical in shape)
  - RBCs get sequestered and destroyed by the spleen due to their shape
  - Mainly northern European ancestry

- **S/S:**
  - Jaundice usually in newborn period (hard to distinguish from hyperbili due to ABO incompatibility)
  - After 2 years of age splenomegaly is usually present
  - Chronic fatigue
  - Malaise
  - Abdominal pain

Hereditary Spherocytosis

- **Evaluation:**
  - Chronic anemia: Hgb is 6-10 g/dL
  - Retic count ranges from 5-20%
  - On peripheral smear a small proportion of RBCs are spherocytic and smaller than normal

- **Management:**
  - If severe HS w/multiple transfusions a splenectomy usually produces a clinical cure but should be differed until >6 because of increased risk of encapsulated bacterial infection
  - Pneumococcal & meningococcal vaccines should be given prior to surgery
  - Like in SCD prophylactic PCN & Folic acid
  - Fevers should be vigorously treated
  - Gallstone formation is common & US should be performed annually (before splenectomy)

Lead Poisoning

- **Key Characteristics:**
  - Most common widespread environmental health concern for children < 6
  - Usually exposure occurs in the home

- **S/S:**
  - **Many resemble common complaints:** Headache, stomach ache, irritability tiredness, poor appetite
  - **Subtle:** Poor attention span and memory, sleep disturbances
  - Weight loss
  - Muscle weakness (diminished DTRs)
  - Seizures (signs of anemia)
  - Loss of visual motor coordination

Lead Poisoning

- **Evaluation:**
  - CBC
  - Blood lead level
  - Free erythrocyte protoporphyrin (FEP)

- **Management:**
  - Blood lead level determines treatment
  - Stop unusual exposure to lead
  - Good nutrition
  - **Refer all values > 70**
  - Refer any patient not responding to treatment or continued increased level when source of exposure is removed

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Bleeding & Clotting Disorders

- Bruising & bleeding are a part of active childhood (knees, shins, nosebleeds normal)
- Becomes a health problem when the bleeding is hard to control, or the bruising is in abnormal areas (back, stomach)
- In adolescence heavy menses could be normal or could indicate bleeding problem

- Congenital:
  - Hemophilia A or B
  - von Willebrand
- Acquired
  - ITP
  - HSP

Idiopathic Thrombocytopenic Purpura (ITP)

- **Key Characteristics:**
  - Characterized by thrombocytopenia (less than 150,000/mcL) in the absence of other causes
  - Occurrence in children 2-8 years
  - Peak occurrence between 2-4 years
  - Most common after a febrile, viral illness
  - Easy bruising: Petechiae (usually in mucous membranes & sclera) ecchymoses
- **S/S:**
  - Acute onset of petechiae, purpura, & bleeding in otherwise healthy child
  - Recent viral illness (1-4 weeks)
  - Hemorrhage of mucous membranes (gums, lips)
  - Epistaxis (severe/difficult to control)
  - Menorrhagia
  - Liver, spleen & lymph nodes are not generally enlarged
  - Bone pain & pallor are rare

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Idiopathic Thrombocytopenic Purpura (ITP)

- **Evaluation:**
  - Low platelet count (otherwise normal CBC)
  - Severe thrombocytopenia when platelet count less 20,000 is common
  - Normal PT, aPTT
  - Megathrombocytes on the peripheral smear
  - Normal WBC & RBC
  - Hgb may be decreased if there is severe epistaxis but the MVC remains normal

- **Differential Diagnoses:**
  - If smear slows fragmented RBCs, BUN & CREAT should be measure to rule out HUS
  - If PT & aPTT are elevated w/ thrombocytopenia, DIC is a possibility & cultures should be taken to identify source of infection
  - A prolonged Pt and aPTT w/a normal platelet count suggest coagulation factor deficiency
  - In a sick, febrile child w/isolated thrombocytopenia, petechaiae, or purpura, the diagnosis to consider 1st is menincococcemia (needs hospitalized for presumed sepsis)

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Idiopathic Thrombocytopenic Purpura (ITP)

- **Management:**
  - Prognosis is excellent, w/spontaneous recover in the majority of pediatric cases w/in 6 months therefore most managed OP w/o specific therapy
  - If platelet count is > 20,000 and no bleeding is observed children/parents should be advised to avoid contact sports, aspirine, & NSAIDs, & to notify if signs of bleeding occur
  - Corticosteroids for children w/highest risk for serious bleeding (1st line)
  - Administration of IVIG (active severe bleeding, or in cases where corticosteroids are contraindicated)
  - Splenectomy in select cases (chronic ITP)

Henoch-Schonlein Purpura (HSP)

- **Key Characteristics:**
  - Most common form of systemic vasculitis in children
  - Results from autoimmune reaction where the body attacks its own tissues
  - Usually develops after respiratory infection but can occur after immunization, insect bite, or allergic reactions
  - Mean age of onset is between 6-7 years old
  - Rates of development/duration vary but is self-limited in most cases

- **S/S:**
  - Purpura is the presenting symptom in approximately 3/4 of patients
  - Arthralgia/arthritis –Joint symptoms were the 2nd most common manifestation, occurring in slightly over 1/2
  - Abdominal pain – Colicky pain occurred in approximately 1/2 and GI bleeding 20-30%
  - Renal disease – The frequency of renal involvement ranges from 21-54%

Henoch-Schonlein Purpura (HSP)

- **Evaluation:**
  - No confirmatory lab tests
  - Clinical diagnosis
    - Palpable purpura of the lower extremities/buttocks
    - Arthritis/arthralgia
    - Abdominal pain
    - Renal involvement
  - UA should be performed in all patients with HSP to screen for renal involvement

- **Management:**
  - The vast majority recover spontaneously
  - Therapy is primarily supportive & includes adequate hydration, rest, & relief of pain
  - Edema of the lower extremities, buttocks, & genital area is improved w/rest and/or elevating the affected area

- **Hospitalization is warranted for the following:**
  - Inability to maintain adequate hydration w/oral intake
  - Severe abdominal pain
  - Significant GI bleeding
  - Changes in mental status
  - Severe joint involvement limiting ambulation
  - Renal insufficiency (elevated creatinine), HTN, and/or nephrotic syndrome

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Hemophilia A and B

- **Key Characteristics:**
  - X-linked recessive hereditary disease
  - Disease occurs almost exclusively in males
  - One of the plasma proteins needed to form a clot is missing/reduced
  - Most common is factor VIII deficiency Hemophilia A (85%)
  - 2nd most common is factor IX deficiency Hemophilia B (15%)
- **S/S:**
  - Excessive bruising
  - Prolonged bleeding from mucous membranes after minor lacerations, immunizations, circumcision, or during menstruation
  - Hemarthrosis characterized by pain and swelling in the elbows, knee, ankles

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Hemophilia A and B

- **Evaluation:**
  - + FMH in most cases
  - Greatly prolonged aPTT
  - A specific assay for factor VIII or IX confirms diagnosis

- **Management:**
  - Prevention of trauma
  - Replacement therapy to increase factor VIII or IX
  - Hemarthrosis is the leading type of significant bleeding-apply ice & pressure to affected joints
  - Avoid aspirin & NSAIDs
  - Immunizations should be given SQ w/26 g or IM with 23 g, hold pressure & ice for several minutes
  - Iron therapy may be necessary in children w/severe bleeding disorders

von Willebrand Disease

- **Key Characteristics:**
  - Most common inherited bleeding disorder (*1%*)
  - Occurs in both sexes
  - Autosomal dominant (severe form- known as type 3 is autosomal recessive)
- **S/S:**
  - Mucous membrane bleeding (epistaxis, menorrhagia)
  - Easy bruising
  - Excessive posttraumatic or postop bleeding
  - History of ecchymosis of trunk, upper arms, thighs

von Willebrand Disease

- **Evaluation:**
  - CBC w/diff (usually normal platelet count)
  - PT
  - aPTT
  - Von Willebrand panel (vW antigen, vW ristocetin cofactor, and Factor VII activity)

- **Management:**
  - Referral to Hematology
  - Depends on the type & severity of the bleeding
  - Desmopression DDAVP & Factor VIII-vWF concentrates
  - Local measure to control bleeding
  - Adjunctive therapy (estrogen) depends on the type
  - The use of antifibronolytic agents are sometimes recommended for dental extraction/nosebleeds

Splenomegaly

Initial evaluation of the child with unexplained splenomegaly includes:

- History & physical examination
- CBC w/diff
- Reticulocyte count
- Periphal blood smear
- LFTs
- EBV & CMV Chest radiograph
- Abdominal US

Splenomegaly

- The most common causes of splenomegaly include:
  - Infection (EBV CMV)
  - Malignancy
  - Disorders of immune regulation and hemolytic anemias (portal HTN, storage diseases, & space-occupying lesions)
- A history of recent febrile illness, pharyngitis, & fatigue suggests a viral etiology
- More pronounced symptoms suggest systemic diseases (leukemia, lymphoma, SLE, malaria, or TB)
  - Persistent fevers
  - Night sweats,
  - Weight loss,
  - Decreased activity level
- Other PE findings that are important include:
  - Lymphadenopathy
  - Jaundice
  - Hepatomegaly
  - Rashes
  - Joint swelling
  - Petechiae
  - Ecchymosis

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Question

The pediatric nurse practitioner provides primary care for a 30-month-old child who has sickle cell anemia who has had one dose of 23-valent pneumococcal vaccine. Which is an appropriate action for health maintenance in this child?

1. Administer an initial meningococcal vaccine.
2. Begin folic acid dietary supplementation.
3. Decrease the dose of penicillin V prophylaxis.
4. Give a second dose of 23-valent pneumococcal vaccine

The pediatric nurse practitioner provides primary care for a 30-month-old child who has sickle cell anemia who has had one dose of 23-valent pneumococcal vaccine. Which is an appropriate action for health maintenance in this child?

Answer: Administer an initial meningococcal vaccine.

Question

The primary care pediatric nurse practitioner reviews hematology reports on a child with beta-thalassemia minor and notes an Hgb level of 8 g/dL. What will the nurse practitioner do?

1. Evaluate serum ferritin.
2. Order Hgb electrophoresis.
3. Prescribe supplemental iron.
4. Refer for RBC transfusions.

The primary care pediatric nurse practitioner reviews hematology reports on a child with beta-thalassemia minor and notes an Hgb level of 8 g/dL. What will the nurse practitioner do?

Answer: Evaluate serum ferritin.

Question

A school-age child comes to the clinic for evaluation of excessive bruising. The primary care PNP notes a history of an URI 2 weeks prior. The physical exam is negative for hepatosplenomegaly and lymphadenopathy. Blood work reveals a platelet count of 60,000/mm3 with normal PT and aPTT. How will the nurse practitioner manage this child's condition?

1. Admit to the hospital for IVIG therapy.
2. Begin a short course of corticosteroid therapy.
3. Refer to a pediatric hematologist.
4. Teach to avoid NSAIDs and contact sports.

A school-age child comes to the clinic for evaluation of excessive bruising. The primary care PNP notes a history of an URI 2 weeks prior. The physical exam is negative for hepatosplenomegaly and lymphadenopathy. Blood work reveals a platelet count of 60,000/mm3 with normal PT and aPTT. How will the nurse practitioner manage this child's condition?

Answer: Teach to avoid NSAIDs and contact sports.

Question

The primary care PNP is managing care for a child diagnosed with iron-deficiency anemia who had an initial hemoglobin of 8.8 g/dL and hematocrit of 32% who has been receiving ferrous sulfate as 3 mg/kg/day of elemental iron for 4 weeks. The child's current lab work reveals elevations in Hgb/Hct and reticulocytes with a hemoglobin of 10.5 g/dL and a hematocrit of 36%. What is the next step in management of this patient?

1. Discontinue the supplemental iron and encourage an iron-enriched diet.
2. Continue the current dose of ferrous sulfate and recheck labs in 1 to 2 months.
3. Increase the ferrous sulfate dose to 4 to 6 mg/kg/day of elemental iron.
4. Refer the child to a pediatric hematologist to further evaluate the anemia

The primary care PNP is managing care for a child diagnosed with iron-deficiency anemia who had an initial hemoglobin of 8.8 g/dL and hematocrit of 32% who has been receiving ferrous sulfate as 3 mg/kg/day of elemental iron for 4 weeks. The child's current lab work reveals elevations in Hgb/Hct and reticulocytes with a hemoglobin of 10.5 g/dL and a hematocrit of 36%. What is the next step in management of this patient?

Answer: Continue the current dose of ferrous sulfate and recheck labs in 1 to 2 months.

Question

A complete blood count on a 12-month-old infant reveals microcytic, hypochromic anemia with a hemoglobin of 9.5 g/dL. The infant has mild pallor with no hepatosplenomegaly. The primary care pediatric nurse practitioner suspects

1. hereditary spherocytosis.
2. iron-deficiency anemia.
3. lead intoxication.
4. sickle-cell anemia

A complete blood count on a 12-month-old infant reveals microcytic, hypochromic anemia with a hemoglobin of 9.5 g/dL. The infant has mild pallor with no hepatosplenomegaly. The primary care pediatric nurse practitioner suspects

Answer: iron-deficiency anemia.

Cervical Lymphadenopathy: History

- Duration & laterality of enlargement & change in size over time
- Associated symptoms: fever, weight loss, fatigue, malaise, conjunctivitis, pharyngitis, nasal, aural, or sinus obstruction w/o discharge, dental problems/mouth sores, cough, arthralgia, skin lesions or trauma
- Immunization status (diphtheria, MMR)
- Ill contacts
- Ingestion of unpasteurized animal milk
- Animal exposures
- Flea/tick bites
- Medications
- Geographic location & travel history

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Cervical Lymphadenopathy: Physical Exam

- Hepatosplenomegaly w/generalized adenitis (systemic infection)
- “Reactive" or shotty nodes- soft & discrete, mobile, minimally tender
- Infected nodes- isolated, asymmetric, warm, & erythematous, they may be fluctuant, less mobile & discrete than reactive lymph nodes (tender)
- Malignant lymph nodes often are hard, fixed, or matted to the underlying structures (nontender)
- Conjunctival injection may indicate Kawasaki disease
- Periodontal disease, ulcers (HSV), gingivostomatitis or pharyngitis
- Generalized rash (non specific viral illness)
- Localized skin lesion (specific etiology)

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Cervical Lymphadenopathy: Evaluation/Management

- **Acute bilateral:**
  - URI is the most common cause
  - Ill-appearing, febrile, progressive or persistent symptoms the following evaluation:
  - CBC w/diff
  - GAS
  - ESR
  - CRP
  - Hepatic profile
  - Blood culture
  - EBV, CMV, HIV
  - Chest X-ray

- **Acute unilateral:**
  - Usually causes by S. aureus
  - Children w/poor oral hygiene
  - Blood cultures if ill appearing
  - Throat culture
  - Empiric therapy for children w/moderate symptoms
  - Patients should response to empiric therapy w/in 48-72 hours, if they do not or worsen should be reevaluated promptly

OBSERVE NODES IF:

Bilateral, < 3 cm in size, no erythema & non/minimal tender

EMPIRICALLY TREAT IF:

No systemic symptoms, node > 2-3 cm, unilateral, or erythematous & tender

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Generalized Lymphadenopathy

- **History**:
  - Lymph node enlargement
  - Associated symptoms (local & systemic)
  - Potential exposures
  - Past medical history

- **Physical examination:**
  - Weight loss of >10 percent (malignancy?)
  - Head, eyes, ears, nose, throat
  - Tinea capitis
  - Conjunctival erythema
  - Nasal obstruction/depression of the soft palate
  - Dental problems, pharyngitis, herpangina, HSV gingivostomatitis
  - Adventitious sounds (systemic process)
  - Hepatosplenomegaly (EBV, CMV, brucellosis, HIV, syphilis, neoplastic disease, rheumatologic disease) abdominal mass (neuroblastoma)
  - Localized lesions, generalized rash

Generalized Lymphadenopathy: Evaluation & Management

1. History and examination to look for obvious causes

2. Early biopsy of most abnormal node for children with:
- Supraclavicular nodes
- Massively enlarged nodes (ie, >4 cm [1.6 inches])
- A group of nodes with a total diameter >3 cm (1.2 inches)

3. Initial testing typically includes:
- CBC w/diff, ESR/CRP
- Serology for CMV and EBV & other viral illness as indicated
- TST
- Chest x-ray

4. Provide treatment/additional evaluation for conditions that are identified through initial history, examination, & testing

5. When the cause remains uncertain after the initial evaluation, obtain the following tests:
- Serology for *Bartonella henselae*, toxoplasmosis, histoplasmosis, coccidiomycosis, brucellosis, syphilis, HIV, & other viruses
- ANA

6. Obtain biopsy of the most abnormal node w/in 4 weeks of initial evaluation if:
- Any lymph nodes increase in size
- There is a lymph node ≥2 cm in diameter and **either** of the following:
  - The diagnosis remains uncertain after 4 weeks
  - There is no response to therapy as indicated by the findings of initial or second tests

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Cancer

Uncommon & often present w/symptoms of benign illness

- **S/S are variable/nonspecific & can include:**
  - Unexplained weight loss
  - HA (typically in early morning)
  - Swelling/persistent pain in joints, back, or legs
  - Lumps or masses
  - Excessive bruising/bleeding
  - Rash
  - Recurrent infections
  - Persistent N/V
  - Fatigue
  - Vision changes
  - Recurrent fevers w/o cause

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Leukemia

- **Key Characteristics:**
  - Malignant hematologic diseases in which normal bone marrow elements are are replaced w/poor lymphocytes (blast cells)
  - Most common form of childhood cancers
  - Acute lymphoblastic leukemia (ALL) accounts for 80% of childhood cases & 56% of adolescent (peak incidence between 2-6)
  - Acute myeloid leukemia (AML) is less common

- **S/S:**
  - **All are vague & provider should maintain a high index of suspicion for cancer**
- Child may be anemic, pale, listless, irritable, or chronically tired & have the following:
  - History of repeat infections, fever, weight loss
  - Bleeding episodes (epistaxis, petechiae, & hematomas)
  - Lymphadenopathy
  - Hepatosplenomegaly
  - Bone/Join pain

CNS symptoms, such as HA, vomiting, or lethargy are rare at time of diagnosis but can present due to IC or spinal mass

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Leukemia

- **Evaluation:**
  - CBC w/diff, platelet, and retic count (Thrombocytopenia & anemia are present in most cases)
  - WBC may be elevated, normal, or low w/varying levels of neutropenia
  - Peripheral smear (malignant cells)
  - Bone marrow aspiration

- **Management:**
  - Referral to Oncology
  - Approximately 90% of those diagnosed with ALL can now be cured
  - ALL: Chemo & systemic corticosteroids
  - AML: Chemo, CNS prophylaxis
  - Role of the PCP is crucial to facilitate proper referrals, routine health visits, immunizations when appropriate
  - The child should be monitored by PCP for failed remission or metastasis (CNS, testicles)

Non-Hodgkin Lymphoma

- **Key Characteristics:**
  - Solid tumors of the lymphatic system
  - Higher incidence in males
  - Usually occur in 2nd decade of life, rare < 3
- **S/S:**
  - Acute abdomen: pain, distention, fullness, and constipation
  - Nontender lymph node enlargement
- **Evaluation:**
  - CBC/diff (may be normal)
  - LFTs
  - Lactate Dehydrogenase (LDH)
  - Uric acid
  - Electrolytes
  - Chest x-ray, US, CT, MRI
- **Management:**
  - Referral to oncology
  - Chemotherapy

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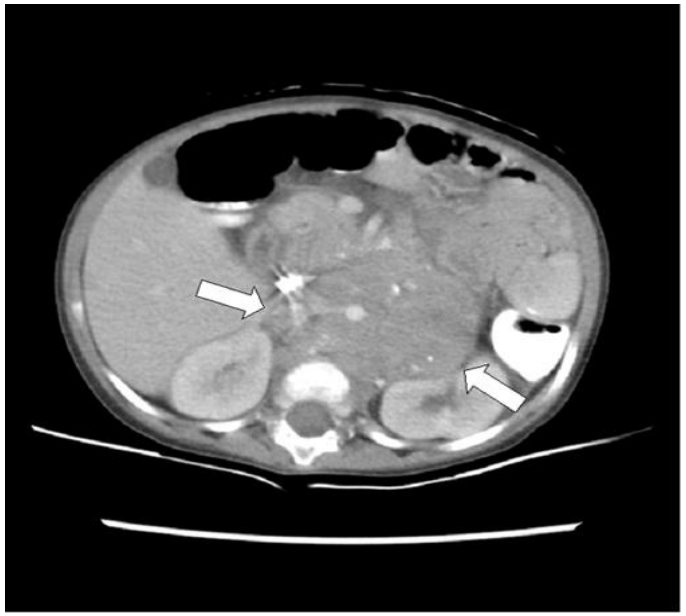
Hodgkin Lymphoma

- **Key Characteristics:**
  - Usually originates in the cervical nodes & spreads to other nodes or other organs if left untreated
  - Rare in children < 15
- **S/S:**
  - Painless enlargement of lymph nodes (usually cervical)
  - Chronic cough (trachea compressed by large mediastinal mass)
  - Fever
  - Decreased appetite
  - Weight loss of 10% or more w/in 6 months
- **Evaluation:**
  - Hematologic findings are often normal but could include:
    - Anemia
    - Elevated ESR, CRP
    - Abnormal LFTs
    - UA may have proteinuria
    - X-ray, US, CT, MRI
- **Management:**
  - Referral to Oncology
  - Optimal results through combination of radiation & chemotherapy

Neuroblastoma

- **Key Characteristics:**
  - Refers to a spectrum of neuroblastic tumors
  - The adrenal gland most common primary site (40%)
  - Abdominal (25%), thoracic (15%), cervical (5%), and pelvic sympathetic ganglia (5%)



- **Children with localized disease can be asymptomatic, children w/advanced disease appear ill at presentation, usually w/ systemic S/S to include the following**:
  - Abdominal mass (retroperitoneal or hepatic)
  - Abdominal pain or constipation
  - Proptosis
  - Periorbital ecchymosis
  - Horner syndrome (miosis, ptosis, anhidrosis)
  - Localized back pain, weakness (from spinal cord compression)
  - Scoliosis, bladder dysfunction
  - Palpable non-tender subcutaneous nodes
  - Fever, weight loss
  - Bone pain
  - Anemia
  - Heterochromia iridis (different colors of the iris or portions of the iris)
  - Hypertension
  - Unilateral nasal obstruction

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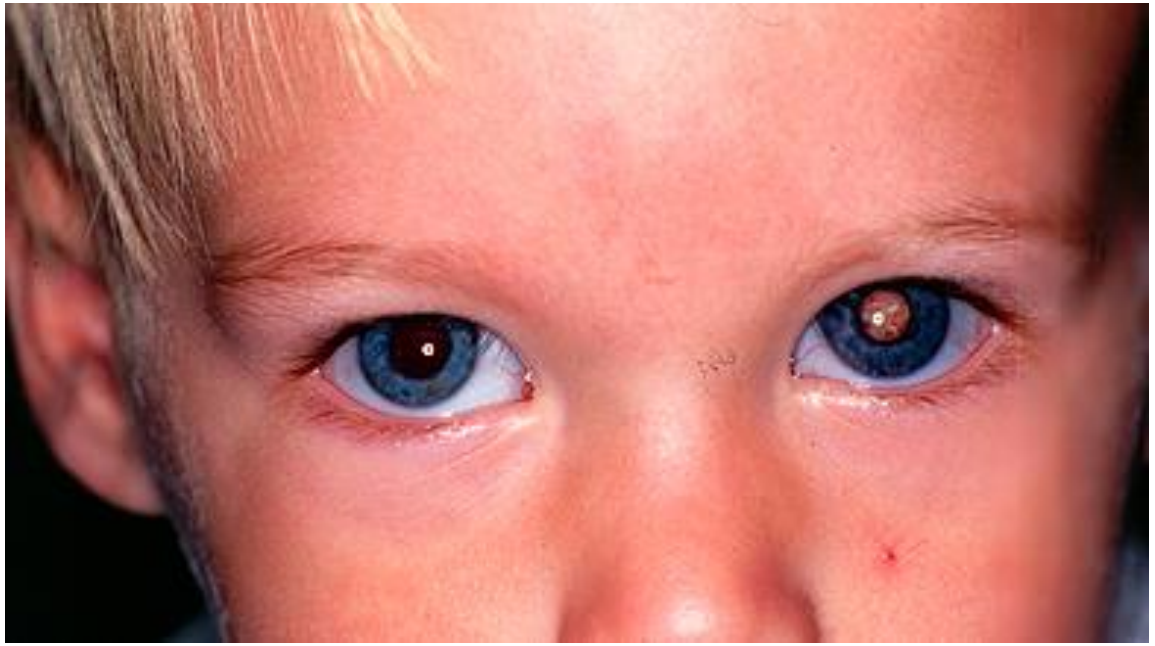
Retinoblastoma

- **Key Characteristics:**
  - Most common primary intraocular malignancy of childhood
  - Accounts for 10-15% of cancers that occur within the 1st year of life
- **S/S:**
  - Typically presents as leukocoria (white reflex) in a child < 3
  - Other common presenting symptoms include strabismus, nystagmus, and a red inflamed eye



Central Nervous System Tumors

- **Key Characteristics:**
  - Primary malignant CNS tumors are the 2nd most common childhood malignancies, after hematologic malignancies, & are the most common pediatric solid organ tumor
  - 2nd leading cause of death from childhood cancer surpassing ALL

- **S/S:**
  - Headaches (most common symptom)
    - Early morning, relieved by vomiting

- Common symptoms based on tumor location:
  - Posterior fossa tumors: Nausea & vomiting, HA, abnormal gait, & coordination
  - Brainstem tumors: Abnormal gait/coordination, & cranial nerve palsies
  - Spinal cord tumors: Back pain and/or weakness & abnormal gait
  - Supratentorial & central tumors: Symptoms are generally nonspecific, most commonly HA

Diagnosis made by MRI or CT & histological exam

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Question

The primary care pediatric nurse practitioner sees a 12-month-old infant who is being fed goat's milk and a vegetarian diet. The child is pale and has a beefy-red, sore tongue and oral mucous membranes. Which tests will the nurse practitioner order to evaluate this child's condition?

1. Hemoglobin electrophoresis
2. RBC folate, iron, and B_{12} levels
3. Reticulocyte levels
4. Serum lead levels

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Answer: RBC folate, iron, and B12 levels

Question

A 2-year-old child who has SCA comes to the clinic with a cough and a fever of 101.5°C. The child currently takes penicillin V prophylaxis 125 mg orally twice daily. What will the primary care pediatric nurse practitioner do?

1. Give intravenous fluids and antibiotics in clinic.
2. Admit the child to the hospital to evaluate for sepsis.
3. Increase the penicillin V dose to 250 mg.
4. Order a chest radiograph to rule out pneumonia.

A 2-year-old child who has SCA comes to the clinic with a cough and a fever of 101.5°C. The child currently takes penicillin V prophylaxis 125 mg orally twice daily. What will the primary care pediatric nurse practitioner do?

Answer: Admit the child to the hospital to evaluate for sepsis.

Question

The primary care pediatric nurse practitioner evaluates a 5-year-old child who presents with pallor and obtains labs revealing a hemoglobin of 8.5 g/dL and a hematocrit of 31%. How will the nurse practitioner manage this patient?

1. Prescribe elemental iron and recheck labs in 1 month.
2. Reassure the parent that this represents mild anemia.
3. Recommend a diet high in iron-rich foods.
4. Refer to a hematologist for further evaluation.

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Answer: Prescribe elemental iron and recheck labs in 1 month.

Question

The primary care pediatric nurse practitioner is examining a 5-year-old child who has had recurrent fevers, bone pain, and a recent loss of weight. The physical exam reveals scattered petechiae, lymphadenopathy, and bruising. A complete blood count shows thrombocytopenia, anemia, and an elevated white cell blood count. The nurse practitioner will refer this child to a specialist for

1. corticosteroids and IVIG.
2. hemoglobin electrophoresis.
3. bone marrow biopsy.
4. immunoglobulin testing.

The primary care pediatric nurse practitioner is examining a 5-year-old child who has had recurrent fevers, bone pain, and a recent loss of weight. The physical exam reveals scattered petechiae, lymphadenopathy, and bruising. A complete blood count shows thrombocytopenia, anemia, and an elevated white cell blood count. The nurse practitioner will refer this child to a specialist for

Answer: bone marrow biopsy.

Question

The primary care pediatric nurse practitioner reviews a child's complete blood count with differential white blood cell values and recognizes a "left shift" because of

1. a decreased eosinophil count.
2. a decreased lymphocyte count.
3. an elevated monocyte count.
4. an elevated neutrophil count.

The primary care pediatric nurse practitioner reviews a child's complete blood count with differential white blood cell values and recognizes a "left shift" because of

Answer: an elevated neutrophil count.