Pediatric Hematology/Oncology and Vascular Anomalies/Malformations

Consult and referral guidelines from Spectrum Health Helen DeVos Children's Hospital

Introduction

We care for children and teens from birth to 21 years. The most common reasons patients are referred are as follows:

- Abnormal CBC results, such as neutropenia, thrombocytopenia or anemia
- Abnormal WBC differential
- Refractory iron deficiency
- Hemolytic anemia, such as congenital spherocytosis
- Hemoglobinopathies, including sickle cell disease and thalassemia
- Bone marrow failure conditions
- Cancers of all types
- Conditions predisposing to cancer, including Fanconi anemia, von Hippel-Lindau syndrome, hemihypertrophy, Li-Fraumeni syndrome, neurofibromatosis and others
- Lymphadenopathy
- Abnormal coagulation tests
- Bleeding disorders, including hemophilia, other factor deficiencies and von Willebrand disease
- Hereditary thrombophilia, including Factor V Leiden mutation
- Hemangiomas: infantile and congenital
- Vascular anomalies and malformations
- Lymphedema

We want to make referrals easy, fast and efficient for primary care providers. This tool was developed to help create productive visits for your patients. In almost all cases, it is our practice to contact you by phone to discuss your patient prior to scheduling a consultation. We do this to gather additional information, triage/assess the urgency of referral and facilitate additional workup prior to consultation. There are times when a telephone discussion is all that is needed, saving time and resources for your patient and his/her family.

Each guideline includes three sections: suggested workup and initial management; when to refer; and information needed. Suggested workups may not apply to all patients, but these are studies we generally consider during office visits.

Feedback regarding these guidelines is encouraged. Please contact Bonnie Doyle at 616.267.1908 to share feedback.
For access to all pediatric guidelines, visit helendevoschildrens.org and type "guidelines" in the search field.

Appointment priority guide

Immediate	During business hours, call the referring provider line (Bonnie Doyle) at 616.267.1908. After hours and on weekends, contact HDVCH Direct at 616.391.2345 and ask to speak to the on-call pediatric hematologist/oncologist. We will help decide if your patient should go to the emergency department or be seen immediately in our clinic.
Urgent	Likely to receive an appointment within two days. During business hours, call the referring provider line (Bonnie Doyle) at 616.267.1908. After hours and on weekends, call HDVCH Direct at 616.391.2345 and ask to speak to the on-call pediatric hematologist/oncologist regarding an urgent referral.
Routine	Likely to receive an appointment within 10 days to six weeks. Send referral via Epic Care Link, fax completed referral form to 616.267.1005 or send referral through Great Lakes Health Connect.

Diagnosis/symptoms	Suggested workup/initial management	When to refer	Information needed
Enlarged lymph node	DO NOT, for any reason, give systemic steroids. Detailed history paying attention to constitutional symptoms, weight loss/failure to thrive, musculoskeletal pain, exposure to cats Physical exam paying attention to weight/growth curve, all lymph node regions of neck, axilla and inguinal areas, abdominal exam for hepatosplenomegaly and bruising on skin exam If suspicion for malignancy is high, send to a hospital laboratory (preferably not satellite lab where specimens must be couriered): CBC with manual differential, CMP, LDH, phosphorus, uric acid. Obtain chest X-ray.	Patient with large lymph nodes (>2 cm) without known infectious etiology, or firm, non-tender or matted lymph nodes should be referred urgently. Those with abnormal labs may need to be seen immediately.	Office notes Any prior workup, including all laboratory studies and chest X-ray if performed

Diagnosis/symptoms	Suggested workup/initial management	When to refer	Information needed
Concern for acute leukemia	DO NOT, for any reason, give systemic steroids. Detailed history paying attention to constitutional symptoms, weight loss/failure to thrive, musculoskeletal pain, complaints of enlarged lymph nodes. Physical exam paying attention to weight/growth curve, all lymph node regions of neck, axilla and inguinal areas, abdominal exam for hepatosplenomegaly, and bruising or pallor on skin exam. If suspicion is high, send to a hospital laboratory (not satellite lab where specimens must be couriered): CBC with manual differential, CMP, LDH, phosphorus, uric acid. Consider chest X-ray.	Anytime there is concern for acute leukemia based on physical exam or laboratory findings, call right away (see Immediate Referral). We will be glad to provide consultation and interpretation of tests and management guidance.	Office notes Any prior workup, including all laboratory studies and chest X-ray, if performed

Diagnosis/symptoms	Suggested workup/initial management	When to refer	Information needed
Abdominal mass	Detailed history paying attention to constitutional symptoms, weight loss/failure to thrive, abdominal pain, chronic constipation Physical exam paying attention to blood pressure, weight/growth curve, all lymph node regions of neck, axilla and inguinal areas, abdominal exam for hepatosplenomegaly and abdominal mass Abdominal ultrasound, CBC and CMP are good screening tests.	We will be glad to provide guidance about the most efficient and safest way to work up your patient (prior to referral). If your index of suspicion is high for an abdominal mass, and your patient is being worked up as an outpatient, notify us prior to scheduling diagnostic imaging or triaging to the emergency department (ED) so we are prepared to act upon the results or ED notification. These patients are seen on an immediate or urgent referral basis.	Office notes Any prior workup, including all laboratory studies
Extremity mass	Detailed history paying attention to constitutional symptoms, weight loss/failure to thrive, pain and inability to bear weight Physical exam paying attention to weight/growth curve, all lymph node regions, extremity exam Plain X-ray films of limbs above and below the area of pain are a good initial step. This should be followed up with MRI of the extremity with and without contrast when index of suspicion is high. If sedation is required, consider chest X-ray PA/lateral to evaluate airway and look for lung disease prior to MRI.	We will be glad to provide guidance about the most efficient and safest way to work up your patient (prior to referral). Often, pediatric patients with an extremity mass are referred first to orthopedic oncology at Spectrum Health Cancer Center at Lemmen-Holton Cancer Pavilion. We can help facilitate that referral. These patients are seen on an urgent referral basis.	Office notes Any prior workup, including all laboratory studies

Diagnosis/symptoms	Suggested workup/initial management	When to refer	Information needed
New cranial nerve palsy, onset of weakness, lethargy (concern for hydrocephalus)	Detailed history paying attention to constitutional symptoms, weight loss/failure to thrive, headache, seizures, decline in school performance, history of NF-1 Physical exam paying attention to weight/growth curve, complete neurologic exam, vision	We will be glad to provide guidance about the most efficient and safest way to work up your patient (prior to referral). In many cases, referral to the emergency department is most appropriate. If possible, if your index of suspicion is high for an intracranial mass, and your patient is being worked up as an outpatient, we would be grateful for advance notification prior to scheduled radiology imaging or triage to the emergency department. This helps us to make quick schedule changes in those challenging situations where we are asked to meet a patient in the emergency department or radiology waiting room. These patients are seen on an immediate or urgent referral basis.	Office notes Any prior workup, including all laboratory studies
Overgrowth syndromes, including hemihypertrophy (hemihyperplasia), Beckwith-Wiedemann syndrome, Sotos syndrome, megalencephaly-capillary malformation syndrome, others	These children have a small increased risk for embryonal tumors of childhood, such as Wilms tumor, neuroblastoma, hepatoblastoma and adrenal cortical carcinoma. Consider ordering abdominal ultrasound prior to consultation visit.	Routine referral: We will generally follow these children until 8 years of age.	Office notes Any prior workup

Diagnosis/symptoms	Suggested workup/initial management	When to refer	Information needed
Familial cancer syndromes, including Li-Fraumeni syndrome, von Hippel-Lindau syndrome, Lynch syndrome, familial adenomatous polyposis syndrome	Referral to cancer genetics for appropriate counseling and screening test	Routine referral: After confirmed diagnosis of familial cancer syndrome — even if the child has no personal history of cancer — we will discuss and provide a cancer screening regimen specific to the familial cancer syndrome.	Office notes Any prior workup
Conditions associated with bone marrow failure	Detailed history Physical exam paying particular attention to microcephaly, features of VACTERL association, thumb anomalies, short stature Obtain CBC with manual differential, reticulocyte count	Those with significant pancytopenia may need to be seen immediately or urgently, as leukemia is also in the differential diagnosis.	Office notes, including growth charts Any prior workup
Normocytic anemia (low hemoglobin with normal MCV)	Detailed history Peripheral smear (manual differential acceptable too), reticulocyte count, direct Coombs, CMP. Note: reticulocyte count and direct Coombs can be added to specimen in lab when CBC results show normocytic anemia.	We will be glad to provide consultation and interpretation of tests and management guidance. Immediate referral: patients with positive Coombs test	Office notes Any prior workup, including all laboratory studies

Diagnosis/symptoms	Suggested workup/initial management	When to refer	Information needed
Macrocytic anemia (low hemoglobin with high MCV)	Detailed history, including diet Peripheral smear (manual differential acceptable too), reticulocyte count, TSH with reflexive T4, RBC folate, B12 level, CMP	We will be glad to provide consultation and interpretation of tests and management guidance. All patients should be referred for routine consultation. Some patients may require bone marrow aspiration and biopsy to evaluate for marrow failure or myelodysplastic syndromes.	Office notes, including growth charts Any prior workup, including all laboratory studies
Microcytic anemia (low hemoglobin with low MCV)	Detailed history, especially diet history (quantitation of cow's milk), menstrual history and any GI symptoms Ferritin, TIBC and serum iron. Consider stool for hemoccult if appropriate. Trial of oral iron replacement: 3 mg/kg of elemental iron given once daily; discontinue cow's milk	We will be glad to provide consultation and interpretation of tests and management guidance. Patients with lack of response to oral iron supplementation after two weeks, ensuring medication adherence, should be referred and will be seen on a routine basis. Patients with hemoglobin less than 7g/dL will be seen on an urgent or immediate basis, depending on patient factors. Males and non-menstruating females above 2 years with iron deficiency anemia without excessive milk intake but with normal iron absorption will likely need referral to pediatric GI for possible inflammatory bowel disease.	Office notes Any prior workup, including all laboratory studies

Diagnosis/symptoms	Suggested workup/initial management	When to refer	Information needed
Sickle cell disease (hemoglobin SS, hemoglobin SC or sickle beta thalassemia)	Send confirmatory hemoglobin fractionation (not electrophoresis). Immediately start penicillin VK 125 mg twice daily for child under 3 years; 250 mg twice daily for child age 3 and over.	All patients should be referred and will be seen for routine consultation.	Office notes State newborn screen
Hemoglobinopathy trait, including sickle cell trait; isolated hemoglobin C trait or hemoglobin E trait (without concomitant thalassemia or sickle cell)	Patients with hemoglobinopathy trait do not need ongoing care from a hematologist.	A one-time routine consultation to discuss inheritance, etc., is offered.	Office notes State newborn screen and/or hemoglobin fractionation
Alpha thalassemia trait or beta thalassemia trait	Patients with thalassemia trait do not need ongoing care from a hematologist.	A one-time routine consultation to discuss laboratory findings, inheritance and potential confusion with iron deficiency anemia is offered.	Office notes State newborn screen and/or hemoglobin fractionation
Newborn with Rh sensitization	No additional workup needed beyond usual CBC/reticulocyte and bilirubin.	We like to see these patients **prior** to hospital discharge. Inpatient consult should be requested. Urgent referral: If born outside of HDVCH, refer so that monitoring plan can be put in place.	Office notes Birth records, including laboratory studies
Newborn with ABO incompatibility	No additional workup needed beyond usual CBC/reticulocyte and bilirubin.	At birth: Hemoglobin less than 12 g/dL After discharge: Hemoglobin less than 10 g/dL	Office notes Birth records, including laboratory studies

Diagnosis/symptoms	Suggested workup/initial management	When to refer	Information needed
Newborn with family history of hereditary spherocytosis	CBC with manual differential, reticulocyte count. Consider bilirubin if jaundiced. Osmotic fragility should not be sent in newborn period.	We will be glad to provide consultation and interpretation of tests. Newborn with anemia and hyperbilirubinemia and family history of HS should be referred within two weeks of hospital discharge. Patient will be seen for urgent or routine consultation depending on patient factors.	
Isolated thrombocytopenia	Detailed history, including maternal/gestational history if patient is a newborn, recent medication changes or immunizations Physical exam paying attention to weight/growth curve, oral exam for petechiae, all lymph node regions of neck, axilla and inguinal areas, abdominal exam for hepatosplenomegaly and bruising/petechiae on skin exam If no bruising and incidentally found, consider repeating CBC in case platelets were clumped. If patient has any associated symptoms, obtain CBC with manual differential, CMP, uric acid, LDH, Coombs test, blood type.	Platelets < 20,000 will require immediate referral/probable hospitalization. Platelets $\underline{\geq}$ 20,000 but < 50,000 without other cytopenias will be seen on an urgent or routine basis depending on patient factors. Platelets $\underline{\geq}$ 50,000 will be seen on a routine basis.	Office notes Any prior workup

Diagnosis/symptoms	Suggested workup/initial management	When to refer	Information needed
Isolated neutropenia	Detailed history, including recent illness, history of infections/recurrent fevers, mouth sores, diarrhea, autoimmune disease, race/ethnicity Physical exam paying attention to weight/growth curve, any signs of infection, all lymph node regions of neck, axilla and inguinal areas, abdominal exam for hepatosplenomegaly CBC with manual differential. Consider repeating one to two weeks later to see if low absolute neutrophil count persists.	Patients with neutrophils < 500/uL (severe neutropenia) *with fever* should be triaged immediately to the emergency department, and will be hospitalized. PHO should be contacted to assist with fast-tracking these patients in the ED. Patients with neutrophils ≥ 500/uL but < 1,000/uL *with fever* should be triaged immediately to the emergency department for treatment of fever and neutropenia, but will not necessarily be hospitalized. PHO should be contacted to assist with fast-tracking these patients in the ED. Routine referral: Patients with persistent neutropenia with ANC < 1,000/uL over at least three to six weeks may need referral. Phone consultation should be the first step. Patients with persistent neutropenia with ANC ≥ 1,000/uL but less than 1,500/uL may not necessarily require referral. Phone consultation should be the first step.	Office notes, including growth charts Any prior workup

Diagnosis/symptoms	Suggested workup/initial management	When to refer	Information needed
Excessive bruising or bleeding	Detailed bleeding history (bleeding with surgery, including circumcision, epistaxis, bleeding gums, prolonged bleeding with tooth loss, excessive bruising, heavy menses); family history of bleeding Must assess for non-accidental trauma/need for CPS referral as appropriate. Obtain PT/INR, PTT, fibrinogen, CBC with manual differential, von Willebrand antigen, von Willebrand ristocetin cofactor activity, factor 8 activity level	Routine referral: child with bleeding history and prolonged PT, PTT; low fibrinogen, von Willebrand antigen or von Willebrand ristocetin cofactor activity	Office notes Any prior workup
Isolated prolonged PTT in non-hospitalized patient	Detailed bleeding history (bleeding with surgery, including circumcision, epistaxis, bleeding gums, prolonged bleeding with tooth loss, excessive bruising, heavy menses); family history of bleeding Obtain repeat PTT, as result could be spurious. If abnormal, and no bleeding history, consider repeating in three to four weeks, as transient anti-phospholipid antibodies are common in children.	Note that normal ranges differ for newborns and infants. We will be glad to provide consultation and interpretation of tests and management guidance. Routine referral: child with bleeding history and prolonged PTT confirmed on repeat measurement	Office notes Any prior workup

Diagnosis/symptoms	Suggested workup/initial management	When to refer	Information needed
Isolated prolonged PT or combined prolonged PT & PTT in non-hospitalized patient	Detailed bleeding history (bleeding with surgery, including circumcision, epistaxis, bleeding gums, prolonged bleeding with tooth loss, excessive bruising, heavy menses); family history of bleeding Obtain repeat PT and PTT, as result could be spurious.	Note that normal ranges differ for newborns and infants. We will be glad to provide consultation and interpretation of tests and management guidance. Routine referral: child with bleeding history and prolonged PT or PT & PTT confirmed on repeat measurement	Office notes Any prior workup
Acute thrombosis	Detailed family history of thrombosis, risk factors for thrombosis (modifiable and unmodifiable) If you are considering initiating anticoagulation prior to referral, please obtain the following labs: PT, aPTT, protein C activity, protein S activity and an antithrombin III activity level.	Immediate referral: child with acute thrombosis should be directed to emergency department for further management. We will either consult (on a critically ill child) or admit the patient to our service.	Office notes Any prior workup
History of familial thrombophilia WITHOUT active thrombosis (factor V Leiden mutation, prothrombin G20210A mutation, antithrombin III deficiency, protein S deficiency, protein C deficiency)	Detailed family history of thrombosis, risk factors for thrombosis (modifiable and unmodifiable) Testing for thrombophilia is controversial but could be considered in high-risk patients (obesity, tobacco use, immobilization due to surgery) and those in whom oral contraceptives are being considered. Avoidance of oral contraceptive pills is strongly recommended in patients with family history of thrombosis. Non-estrogen alternative should be considered.	Routine referral: child with familial risk factor for thrombosis can be seen for one-time consultation to discuss laboratory findings, inheritance and risk reduction.	Office notes Any prior workup

Diagnosis/symptoms	Suggested workup/initial management	When to refer	Information needed
Hemangioma	Birth history, time course in terms of initial appearance and growth pattern Physical exam, paying particular attention to size (documenting dimensions), location, potential for organ compromise, ulceration, presence of petechiae or bruising If concern exists for bruises or petechiae, or non-traumatic bleeding from the vascular tumor, immediate evaluation for Kasabach-Merritt syndrome should occur (CBC with manual differential, fibrinogen, PT, PTT), as KMS can be life threatening. If electing to perform ultrasound of area prior to visit, please order ultrasound with Doppler to assess blood flow. For small, flat (< 1 cm), non-ulcerated, superficial hemangiomas that are not near mucus membranes, topical Timolol may be an option. Please call to discuss.	Urgent referral: child with vascular tumor in organ-threatening location (e.g., on face near eye, nose, mouth) or with significant risk or presence of ulceration. Note that infants with “port wine stain” of face may have other associated anomalies that will require additional workup. Routine referral: child with vascular tumor in non-threatening location and without presence of ulceration. Note that infants with segmental hemangioma involving lower body may have other associated anomalies that will require additional workup. Not all children with infantile hemangiomas need to be referred. We will be glad to provide phone guidance to determine if consultation is warranted.	Office notes Any prior workup

Diagnosis/symptoms	Suggested workup/initial management	When to refer	Information needed
Capillary malformation on face in V1, V2 distribution, high risk for Sturge-Weber	MRI brain scan should be ordered.	These patients are generally not cared for by pediatric hematology unless they have a capillary malformation in the context of an overgrowth syndrome. In the case of an overgrowth syndrome, routine referral should be made. Otherwise, these patients should be referred to pediatric dermatology, ophthalmology and potentially pediatric neurology.	
Vascular anomaly, vascular malformation, lymphedema	Birth history, time course in terms of initial appearance and growth pattern Physical exam paying particular attention to size (documenting dimensions), location, potential for organ or airway compromise, pain or acute swelling. Presence of petechiae, bruising, bleeding. Presence of limb length or girth discrepancy. Ultrasound with Doppler of lesion prior to referral is helpful.	Urgent referral: infant or child with vascular malformation or lymphedema causing pain, with acute swelling, or with signs/symptoms of cellulitis Routine referral: infant or child with vascular malformation of limb without pain or acute growth	Office notes Any prior workup

Pediatric Hematology/Oncology Clinic referring provider line: 616.267.1908 (business hours)

HDVCH Direct phone: 616.391.2345

HDVCH developed these referral guidelines as a general reference to assist referring providers. Pediatric medical needs are complex, and these guidelines may not apply in every case. Helen DeVos Children's Hospital relies on its referring providers to exercise their own professional judgment with regard to the appropriate treatment and management of their patients. Referring providers are solely responsible for confirming accuracy, timeliness, completeness, appropriateness and helpfulness of this material and making all medical, diagnostic and prescription decisions.