Pediatric referral guidelines

Corewell Health™
Helen DeVos
Children's Hospital

Pediatric specialties

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital Direct phone: **616.391.2345**

Helen DeVos Children's Hospital wants to make referrals to easy, fast and efficient for our primary care providers. We've developed these guidelines to help maximize productive office visits for you and your patient.

Each guideline includes appointment priority guide, common conditions treated, information about each service offered and how to refer. For most specialties, we've included three sections for referral considerations: suggested workup and initial management, when to refer and information needed for referral. Please note, suggested work-ups may not apply to all patients, but these are studies we generally consider during office visits and may help us when initially evaluating your patient.

In some cases, we will 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.

These referral guidelines were developed as a general reference to assist referring providers. Pediatric medical needs are complex and these guidelines may not apply in every case. HDVCH 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.

We view this as a "living" document and welcome your feedback to further refine the guidelines.

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Pediatric allergy and clinical immunology

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. NE | Suite 3003

About pediatric allergy and clinical immunology

We care for patients from birth to age 18.

Most common referrals

- Food allergy.
- Anaphylaxis.
- Asthma.
- Recurrent viral wheeze.
- Allergic rhinosinusitis.
- Allergic conjunctivitis.
- Chronic sinusitis.
- Nasal polyps.
- Primary immunodeficiency (frequent/recurrent, unusual infection, periodic fever).
- Positive newborn TREC screen.
- Chronic and acute urticarial/angioedema.
- Hereditary angioedema.
- Bee sting allergy.
- Atopic dermatitis/eczema.
- Drug or vaccine allergy.
- Eosinophilic disorders (especially hypereosinophilia and eosinophilic esophagitis).
- Mast cell disorders.

Notes

With the exception of some drug and bee allergy testing, we do not use needles for any skin testing.

Allergy and clinical immunology appointment priority guide

Immediate (e.g., a positive TREC newborn screen for severe combined immunodeficiency)	Call Corewell Health Helen DeVos Children's Hospital Direct at **616.391.2345** and ask to speak with on-call allergist/immunologist or send to the closest emergency department.
Urgent (e.g., severe eczema or history of food allergy <1 year of age; allergic reaction to medication that is needed/critical for continued care)	Likely to receive an appointment 48 hours. Call our department at **616.267.8150**.
Routine	Will receive first available appointment. Fax completed referral form and records to **616.267.2851** or send through Holon.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Food allergy and food oral immunotherapy	No testing needed prior to visit. Prescribe/instruct on use of epinephrine autoinjector for patient to carry with them at all times. *Note: we do not recommend IgE food allergy "panels" that test a broad range of food allergens in one test. These have a high false positive rate and can lead to false diagnosis and potential harm to the patient. If testing is pursued, specific IgE to single food groups based on history is preferred. IgG to food has been shown to be of no clinical value in food allergy and should not be ordered.	• Any question of food allergy. • History of anaphylaxis. • We recommend all patients with food allergy have care established with an allergist. • Urgent referral: For patient <1 year of age and history of severe eczema/food allergy as literature shows we may have the opportunity to prevent food allergy in these patients.	• Request for consult. • Summary of all previous reactions. • Summary from any ED visits. • Summary of any previous allergy testing.
Anaphylaxis	Could consider baseline tryptase.	Any cases of anaphylaxis, especially unexplained, should be referred to an allergist.	• Request for consult and brief history of anaphylactic event. • Any labs obtained, especially tryptase if this is obtained during ED visit for anaphylaxis.
Asthma	None	• Has been hospitalized. • Intubated/ICU admission. • Frequent ED visits. • Frequent need for oral steroid bursts. • Unresponsive to usual therapy with increasing medication use. • Complicating conditions such as allergic rhinitis, sinusitis, GERD and/ or pneumonia. • Abnormal spirometry or needs frequent monitoring with spirometry.	• Request for consult. • Chief concern. • Summary of previous treatments and response • Respiratory history since birth. • All lab results. • All chest films (must have chest X-ray).* • If sweat chloride test was obtained, must be from CF Center accredited lab.**

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Recurrent cough or wheeze **Recurrent bronchiolitis or bronchitis**	Chest X-ray: PA and lateral.* Sweat chloride at an accredited CF Center.** Trial of bronchodilators at any age. Trial of oral and/or inhaled corticosteroids. or Singular® (if age appropriate) if bronchodilators non- responsive. Oral prednisone is typically dosed ~2 mg/kg/day x five days minimum.	• Has been hospitalized. • Intubated/ICU admission. • Frequent ED visits. • Frequent need for oral steroid bursts. • Unresponsive to usual therapy with increasing medication use. • Complicating conditions such as allergic rhinitis, sinusitis, GERD and/ or pneumonia. • Abnormal spirometry or needs frequent monitoring with spirometry.	• Request for consult. • Chief concern. • Summary of previous treatments and response. • Respiratory history since birth. • All lab results. • All chest films (must have chest X-ray).* • If sweat chloride test was obtained, must be from CF Center accredited lab.**
Allergic rhinitis **Chronic rhinitis** **Allergic conjunctivitis** **Chronic sinusitis** **Nasal polyps**	Trial of second generation H-1 antihistamines (i.e., Zyrtec [cetirizine] or Allegra [fexofenadine]) at any age. Trial nasal steroid if tolerated.	• Symptoms refractory to antihistamine and nasal steroid. • Need to clarify diagnosis of allergy vs. nonallergic. • Need to identify specific allergens for environmental management. • Need for evaluation for allergy shots.	• Request for consult. • History of symptoms. • Therapies to this point.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Immunodeficiency/ concern for frequent infections	CBC with differential IgG, IgA, IgM HIV.	• Four or more ear infections in one year; two or more serious sinus infections in one year. • Two or more months on antibiotics with little effect. • Two or more CXR proven pneumonias in one year. • Failure of an infant to gain weight or grow normally. • Recurrent, deep skin or organ abscesses. • Persistent thrush in mouth or fungal infection of skin. • Need for IV antibiotics to clear infections. • Two or more deep seated infections. including septicemia Family history of primary immunodeficiency. • Infection with rare or low virulent organisms. • Unexplained bronchiectasis. • Urgent referral: For concern for serious immunodeficiency.	• Request for consult. • Brief summary of infections and hospitalizations. • All previous radiology results (including CD of film if not done in our system). • All culture results. • All lab results.
Atopic dermatitis/ eczema **Allergic contact dermatitis**	Topical corticosteroids (cream/ointment not lotion) to effected area. Frequent emollients. Oral H-1 antihistamine at night.	• Continued flares of atopic dermatitis despite current treatment. Urgent referral for all patients <one year of age with severe eczema to evaluate for early introduction/prevention of food allergy in accordance with LEAP study, EAT study and 2016 food allergy practice parameters.	• Request for consult. • Brief history of treatments to this point.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Urticaria/ angioedema	**Acute urticaria (<six weeks)** Investigate viral or allergic causes of urticaria. Oral second generation H1 antihistamine for acute control. **Chronic Urticaria (present most days >six weeks)** Empiric trial of Zyrtec (cetirizine) or Allergra (fexofenadine) twice daily and Pepcid (famotidine) twice daily. Consider trial of daily Singulair (montelukast), if age appropriate. In general laboratory testing is not needed/ indicated. If there are concerning systemic symptoms (fever, weight loss, night sweats, joint pain, etc.), you can consider limited laboratory testing (CBC with differential, ESR and/ or CRP, LFTs and TSH). Angioedema/swelling without urticaria or family history of hereditary angioedema C4.	• Unexplained acute urticaria. • Symptoms that are refractory and continues despite BID H-1 and H-2 antihistamine. • Angioedema without urticaria accompanied by low C4 (concern for hereditary angioedema). • Family history of hereditary angioedema	• Request for consult. • History of previous treatment. • Any labs that were obtained.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Drug or vaccine allergy	Due to high rate of false negatives, unless needed for urgent/ emergent reasons (i.e., chemotherapy) we cannot test to drugs until 6 weeks after reaction.	• History of allergy/reaction to a medication that is medically indicated for the patient to take in the future. • History of allergy/reaction to multiple medications that make prescribing future medications difficult. • Any history of penicillin allergy in children >10 years old.	• Request for consult. • History of reaction to all medications.
Hypereosinophilia	CBC with diff. Toxocara canis antibody and strongyloides Note: There is risk of death if prednisone is given to patient with strongyloides. Test for scabies.	• Absolute eosinophil count >1000 with negative toxocara canis antibody and strongyloides antibody.	• Request for consult. • All laboratory results (including all CBCs that have been obtained).
Venom Allergy	Prescribe injectable epinephrine.	• All patients with history of reaction to stinging insect that is more than a large reaction at the site of the sting/bite.	• Request for consult. • Brief history of reaction.

Notes

*We prefer to look at all X-rays/CT scans ourselves during the visit. If your patient has not obtained their X-rays/CT scans at Corewell Health, we ask that the patient obtain a CD that includes all their X-rays/CT scans and bring it to our office visit.

**Accredited CF care centers include: Helen DeVos Children's Hospital (Grand Rapids), Sparrow Hospital (Lansing), Bronson Hospital (Kalamazoo), Children's Hospital of Michigan (Detroit) and University of Michigan (Ann Arbor).

Pediatric behavioral health

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. NE | Suite 5201

About pediatric behavioral health

We accept referrals for children and teens from birth to age 18 who also have medical comorbidity.

We strive to care for the whole child which includes attending to their mental health, physical health, personal, family, school and community needs. Our experts utilize a multidisciplinary and comprehensive approach to diagnose and determine the root of the child's problem using a biological, psychological and social approach to create a personalized treatment plan for each child.

Most common referrals

- Autism spectrum disorders.
- Developmental disabilities.
- Neuropsychology.
- Child psychiatry.
- Pediatric psychology.
- Hospital-based consultation.

Pediatric behavioral health appointment priority guide

Immediate/ urgent	If patient is experiencing a psychiatric crisis, contact: • Medically clear patient, with private insurance coverage – Pine Rest or Forest View. • Medically clear patient, with Medicaid – Network180 (Kent County) or regional community mental health. • Not medically clear, with private insurance coverage – emergency department.
Routine	Send referral via EPIC care link, fax completed referral form to **616.267.2850,** or send referral through Holon. Detailed clinical information with referrals helps to place the child with the best behavioral health specialist. If making the referral in EPIC, within the system enter ref 81. Then indicate if the child is referred for psychiatry, psychology (therapy), testing (general educational testing), neuropsychological testing (psychological testing in a patient who has a medical history that affects brain development) or autism testing. For consultation on mild-to-moderate mental health concerns, consider: MC3 (**mc3.depressioncenter.org**) or Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345**.

Clinic	About the clinic	Other information
Autism spectrum disorders/ developmental disabilities (Certified BCBS Center of Excellence)	We currently provide comprehensive evaluation services and recommendations for educational and treatment planning. We do not see patients for autism testing who have are covered under Medicaid, (other than McLaren) or United Healthcare insurance plans. Once assessment is complete, we assist with providing a resource list of clinical care organization for treatment.	Symptoms of ASD include: • Deficits in social-emotional reciprocity. • Deficits in nonverbal communicative behaviors used for social interaction. • Deficits in developing and maintaining relationships, appropriate to developmental level (beyond those with caregivers). • Stereotyped or repetitive speech, motor movements or use of objects. • Excessive adherence to routines, ritualized patterns of verbal or nonverbal behavior, or excessive resistance to change. • Highly restricted, fixated interests that are abnormal in intensity or focus. • Hyper- or hypo-reactivity to sensory input or unusual interest in sensory aspects of environment.
Neuropsychology	We help place a patient for psychological and neuropsych testing. Detailed background information regarding the child assists with differentiation in testing: • Neuropsych – medical illness which may be impacting brain development or learning. • Psychological – concerns regarding development or dyslexia, without medical illness.	We provide evaluation and treatment for: • Prenatal alcohol or drug exposure. • Prematurity and/or neonatal complications. • Developmental delay and intellectual disability, transition to adulthood. • Genetic disorders and other rare illnesses. • Hypoxic/anoxic events (i.e., low or lack of oxygen). • Cerebral palsy. • Neuro-immunology and neuro-infectious diseases (e.g., meningitis, HIV, etc.). • Concussion and traumatic brain injury. • Epilepsy. • Pre-surgical and post-surgical evaluations. • Neurovascular disorders. • Neurodegenerative and demyelinating disorders. • Leukemia, brain tumor and treatment with chemotherapy and/or radiation. • Bone marrow transplant. • Solid organ transplant. • Congenital heart disease. • Chronic kidney disease.
Child psychology	We provide short-term consultative care for stabilization and treatment.	We see patients with medical illness that affects mental illness or mental illness which affects physical illness (e.g., patients with diabetes and depression that are not adherent to treatment, have conversion disorder or somatic symptom disorder. We will also see patients <6 years who are failing first line treatments for behavioral health concerns.

Clinic	About the clinic	Other information
Pediatric psychology	We specialize in the evaluation, diagnosis and treatment of mental health disorders in children and teens. Our diagnostic consultations generally consist of one to three visits and are designed to provide comprehensive diagnostic services as well as identify the best avenue of care for each child and family. As part of the evaluation, we may conduct cognitive, academic, social, emotional and/or personality testing.	We provide evaluation and treatment for: • Adjustment to chronic illness. • Adherence to medical treatment regimens. • Anxiety and depression in the context of physical illness. • Autism spectrum disorders. • Behavioral and school problems in children <six years. • Chronic pain. • Enuresis and encopresis. • Gender care. • Neurodevelopmental disorders. • Somatoform disorders including conversion disorder. • Parenting behavioral advice. • Dyslexia and other learning disabilities.
Hospital-based services	The pediatric behavioral health consultation liaison team provides psychiatric and psychological consultation, evaluation and treatment for children and families with acute illnesses. Our psychiatrists and psychologists work closely with social work, psychiatric nurses, child life specialists and behavioral health technicians during a medical hospitalization to provide well-rounded and holistic care.	

Pediatric pain and palliative medicine

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. NE | Suite 5301

About pediatric pain and palliative medicine

The pediatric pain and palliative medicine program offers personalized care to help alleviate pain and address troublesome symptoms in infants, children, and teens facing serious medical illness with a particular focus on improving the overall quality of life for our patients and families. Our care is coordinated with a patient's other pediatric specialists, as well as with the maternal-fetal medicine team for prenatal visits. Our palliative medicine team members include fellow-trained pediatric hospice and palliative medicine physicians, nurse practitioners, nurses, and social workers.

The chronic pain program uses evidence-based approaches to evaluate and treat children and adolescents with a variety of chronic pain conditions. This multidisciplinary team includes a medical provider, pediatric pain psychologist, physical therapist and social worker who work together to address the unique needs of children and adolescents with chronic pain conditions with the goal of reduction of pain and increase in overall functioning.

Services provided – our comprehensive care includes, but is not limited to:

- Working to improve quality of life for their child with serious medical illness.
- Reducing burden of physical symptoms including pain.
- Coordinating care for complex patients.
- Integrating medical advice from numerous specialties.
- Assisting families with medical decision-making.
- Addressing barriers to care.
- Evaluation and treatment of children and adolescents with chronic pain conditions.

Pediatric pain and palliative medicine appointment priority guide

Immediate/ urgent	• Palliative Medicine: Contact Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** and ask to speak to the on-call palliative care provider. We will see the patient within 24 hours. • Chronic Pain: Contact the clinic directly at **616.391.8842** and ask to speak to the chronic pain nurse. We will see the patient within seven business days.
Routine	Palliative Medicine: Contact Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** and ask to speak to the on-call palliative care provider. Chronic Pain: Send referral via Epic or fax to **616.391.2978**. We will see the patient within two to three weeks.

Clinic	About the program	Other information
Chronic pain	The chronic pain clinic provides a comprehensive evaluation to determine an accurate diagnosis and develop a personalized treatment plan. We evaluate each patient's symptoms and functioning from three perspectives- medical, psychological, and physical therapy. Our team uses a whole-person perspective to care for pediatric patients with a range of chronic pain conditions.	We provide evaluation and treatment for: • Outpatient pain medicine management. • Individual and family pain psychology intervention (cognitive behavioral and bio behavioral treatments, including biofeedback). • Psychical therapy. • Social work services. • Care coordination with other medical specialties.

Clinic	About the team	Other information
Palliative medicine	The goal of palliative medicine is to improve quality of life for both the child and the family, by relieving or reducing suffering, whether it is physical, psychological, or emotional, even in our most medically complex patients. The team also assists families with medical decision-making concerns. The team works to integrate medical advice from numerous specialties to help support parents in determining the plan of care specific to their child, with a focus on treating the patient, rather than treating the specific conditions.	We seek to provide relief from the symptoms, pain and stress of a serious illness-whatever the diagnosis. This may include, but is not limited to: • Genetic disorders. • Cancer. • Prematurity. • Neurologic disorders. • Heart and lung conditions.

Pediatric dermatology

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. NE | Suite 5201

About pediatric dermatology

We care for children and teens from birth to age 16, with a referral.
*Exception for severe/scarring acne that may require isotretinoin for ages 16-18.

Most common referrals

- Acne
- Warts
- Atopic dermatitis/eczema
- Infantile hemangioma
- Capillary malformations/ port wine stains
- Vascular malformations including lymphatic, venous and mixed malformations
- Pyogenic granulomas
- Psoriasis
- Fungal infections of skin and nails not responding to treatment
- Café-au-lait macules
- Moles, Spitz nevi
- Congenital nevi
- Nevus sebaceous
- Vitiligo
- Rash/dermatitis, skin lesions, cysts
- Hidradenitis suppurativa (HS)

Notes

- To ensure appropriate triage, please upload photos to Epic or send photos with the referral documentation.

Pediatric dermatology appointment priority guide

Urgent	Likely to receive an appointment within two weeks. We welcome phone calls from the referring office in circumstances where a patient should be seen more urgently. Send referral marked URGENT.
Routine	Some diagnoses may have a six+ month scheduling timeline. Send referral via EPIC care link, fax completed referral form to **616.267-2401**, or send referral through Holon.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Atopic dermatitis/ seborrheic dermatitis Refer to American Academy of Pediatrics Patient Care page about AD.	Prior to visit, educate about emollients, sensitive skin care, and use of class six or seven topical steroid, or class four or five topical steroid in older children. Consider wound culture if suspected infection.	Infants <six months, usually scheduled within two weeks. **If severe, or actively infected, please call for urgent appointment.**	• Send growth chart with patient referral, if possible.
Psoriasis	Prior to visit, trial of topical steroid of appropriate class. If acute onset, check for concurrent strep infection (pharynx or perianal).	**If >30% BSA involvement, consider urgent referral.**	
Acne Refer to the American Academy of Pediatrics journal article on acne.	**Mild** Use BPO +/- topical antibiotic, +/- topical adapalene 0.1% gel. **Moderate** Add oral antibiotic (Doxycycline or Minocycline, 100mg), po BID. **Severe** Oral antibiotics + retinoid + BPO. Do not promise isotretinoin if no treatment has been tried; most health plans require three to six months of oral antibiotics + retinoid for coverage of isotretinoin.	Most can be seen via telemed.	• All previous prescriptions for acne.
Warts	Prior to visit, use OTC salicylic acid +/- in-office cryotherapy if appropriate.	Facial or multiple warts	Document treatment(s) tried and locations/ number of warts.
Infantile hemangioma Refer to American Academy of Pediatrics Clinical Practice Guideline for the Management of Infantile Hemangiomas.	For five or more, schedule a liver ultrasound if under two months of age.	• No improvement following timolol gel treatment for small superficial focal hemangiomas. • Refer early if in cosmetically sensitive area, or ulcerated; better response to propranolol if started at two months of age. • For large segmental lesions on face, refer immediately to the pediatric hematology/oncology vascular anomalies clinic for PHACE syndrome evaluation.	

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Capillary malformations on face in forehead, hemifacial, or median facial regions are highest risk for Sturge-Weber Syndrome Refer to Consensus statement for Port Wine Birthmarks	Recommend referral to neurology and referral to ophthalmology. Consider MRI of brain before 3 months of age.	• Patients will be seen urgently if no workup has been completed in high-risk lesions. • Pulsed dye laser treatments begin at one month of age to maximize results without repeated anesthesia.	
Venous and lymphatic malformations	Ultrasound if unclear diagnosis.	• Send referral to pediatric hematology/oncology vascular anomalies clinic.	
Pyogenic granuloma	Please note if bleeding excessively, or not. Can start topical timolol gel forming solution and cold Vaseline BID – this treatment has been shown to shrink pyogenic granulomas. Treatment can take two to four months, re-check patients at one month.	• Patients are usually seen within one to two weeks. • Referral may be redirected to pediatric plastic surgery depending on location/ scheduling availability.	
Moles (nevi)	We do not perform routine skin checks for patients with no risk factors. Risk factors include family history of melanoma, personal or family history of dysplastic/ atypical moles, long-term immunosuppression.	• Rapidly changing • Bleeding on its own • Strong family history of melanoma and/or dysplastic/ atypical moles • Patients on long-term immunosuppressants	• Note if mole is changing, bleeding, if there is family history of melanoma in 1st degree relative, and if known genetic susceptibility to melanoma.
Congenital nevi Refer to journal article on Care of CMN		• Refer urgently for baby with medium-sized or greater CMN.	• Note size and location in referral
Cysts			• Note location in referral • Facial lesions will be deferred to plastic surgery

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Vitiligo	If vitiligo is extensive and rapidly progressing, consider screening/testing for thyroid disease, vitamin D and celiac disease.		Note if rapidly progressive and if strong patient or family history of autoimmune disease.
Alopecia areata	Consider screening/testing for Vitamin D, ferritin, CBC, TSH. If area is scaly and (+) occipital lymphadenopathy, perform fungal culture with Eswab. Include pulled hair and skin scales in container. If no infection, consider initial treatment with Class I or II steroids QD x 6 weeks.		Note if rapidly progressive.
Rashes	Trial of topical therapy based on working diagnosis.	• If worsening or not improving with one month of trial of topical therapy, then refer.	• Please include a differential diagnosis. • Include description of skin findings or photos with referral as this will help to triage more appropriately.
Urticaria	Consider trial of non-sedating antihistamine QD.	• Please send referral to pediatric allergy.	
Hidradenitis suppurativa (HS) Refer to HS algorithm from Children's National		• Multiple flares per month or one persistent active lesion.	
Lichen sclerosus	Start hydrocortisone 2.5% ointment BID.	• Refer after infectious work-up for UTI and vaginitis has been completed.	• Please send urgently. Physical exam of genital area w/description of findings is necessary for triage. Include information if (+) family history of Lichen sclerosus.
Hyperhidrosis	Trial of dry sol nightly. Consider glycopyrrolate 1-2 mg BID if age appropriate.		

Pediatric endocrinology

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. NE | Suite 1800

Outreach locations:
Lansing, Muskegon, St. Joseph, Traverse City

Healthy weight center at Corewell Health Helen DeVos Children's Hospital
616.391.7999 Phone
616.391.8750 Fax
devoschildrens.org/healthyweightcenter

About pediatric endocrinology

We care for children and teens from birth to age 18.

Most common referrals

- Diabetes
- Short stature or failure to thrive
- Tall stature
- Obesity
- Precocious puberty
- Early childhood breast development in girls
- Delayed puberty
- Premature menses.
- Congenital hypothyroidism
- Acquired hypothyroidism
- Acquired hyperthyroidism (Grave's Disease)
- Goiter/thyromegaly
- Calcium disorders
- Hypoglycemia
- Adrenal insufficiency

Resources

Fit Kids 360 | **fitkids360.org**
A comprehensive, healthy lifestyle program developed to fight childhood obesity, combining basic education about nutrition, behavior and exercise with a wide range of physical activities.

Nutrition counseling | Corewell Health: **616.391.1875**
Trinity Health: **800.639.6366** | University of Michigan Metro Health: **616.252.4461**

Services are offered in locations throughout West Michigan. A physician referral is required.
Insurance coverage varies.

Nutrition websites: eatright.org | **kidshealth.org** | **nutrition.gov** | **choosemyplate.gov**

Pediatric endocrinology appointment priority guide

Immediate	Contact Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** and ask to speak to the on-call endocrinologist and/or send to closest emergency department.
Urgent	Likely to receive an appointment within two days. Call Corewell Health Helen DeVos Children's Hospital Direct and ask to speak to the on-call endocrinologist regarding an urgent referral.
Routine	Likely to receive an appointment within 14 days. Send referral via EPIC care link, fax completed referral form to **616.267-2401**, or send referral through Holon.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Diabetes: new onset referral Immediate referral recommended. New diagnosis education is offered seven days a week. Not all patients are admitted; we will assist with inpatient or outpatient management.	History and exam: • Height, weight, BMI. • Symptoms: history of excessive thirst or urination, weight loss, vomiting, abdominal pain, fatigue or other significant history. HbA1c, urine and/or serum ketones, blood glucose (fasting, random).	• HbA1c ≥6.5%. • Positive urine or blood ketones. • In this case will often need lab work (HC03). • Fasting blood sugar ≥126. • Random blood sugar ≥200 with symptoms of diabetes.	• Growth chart. • Relevant lab studies. • Previous physician notes.
Diabetes: transfer referral Patients transferring diabetes care to Corewell Health Helen DeVos Children's Hospital.	History and exam: • Height, weight, BMI. • Last known insulin regimen. HbA1c, ketones, blood sugar (fasting, random).	• Signs of insulin resistance or conditions associated with insulin resistance (acanthosis nigricans, hypertension, dyslipidemia, polycystic ovarian syndrome). • Previous DX T1/T2DM.	• Growth chart. • Relevant lab studies. • Previous physician notes.
Short stature or failure to thrive Please consider a referral to Nutritional Services or Intensive Feeding Program in a child with poor weight gain in the face of normal linear growth (exceptions are infants with midline abnormalities or males with hypospandias or cryptorchidism).	History and exam Note: Linear growth is better evaluated after age two. TSH, Free T4, CMP, CBC, ESR, IGF-1, IGFBP3, Karyotype for Turners, 30 cell count (in all girls, Transglutaminase IgA, IgA level. Radiology: bone age.	• Strongly recommend referral if child is >two years and growth velocity <four cm a year for more than a year. • If after age three, crossing centile downward. • Child is growing more than two centile lines below mid-parental height*, with a delayed bone age. • Child is less than 3rd percentile in height. *Boy mid-parental height in inches = (mother's height + father's height)/2 + 2.5. Girl mid-parental height in inches = (mother's height + father's height)/2 – 2.5	• Prior growth data/ charts. • Relevant lab studies. • Ask patient's family to bring bone age X-ray to clinic, if completed. • Pertinent medical records. • Results of any additional tests.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Tall stature	History and exam. TSH, Free T4, CMP, CBC, IGF-1, Karyotype. Radiology: bone age.	• Child is >two years and is greater than 97th percentile for height and greater than two centile lines above mid-parental height.* • Child is >two years and progressively crossing centiles for height. *See previous entry for mid-parental height calculations.	• Prior growth data/ charts. • Relevant lab studies. • Ask patient's family to bring bone age X-ray to clinic, if completed. • Pertinent medical records. • Results of any additional tests.
Obesity We recommend a referral to endocrinology for children with BMI greater than 99th percentile and <three years For children 3-17 years, consider a referral to Corewell Health Helen DeVos Children's Hospital ooptimization center (**616.391.7999**) Before referral, please follow American Academy of Pediatrics guidelines for stage I and stage II obesity treatment.	History and physical Fasting CMP, HbA1c, UA, fasting lipid panel or non-fasting total and HDL cholesterol. • See co-management guidelines for lipids, screening of T2DM and PCOS. • Not recommended: fasting insulin Formal nutritional consultation: • Three to five day diet diary evaluation and calorie count. • Ongoing continuity of care and follow-up with a nutritionist. Establishment of a regular exercise regimen.	• Highly suspected endocrine disorder. • Secondary complications of endocrine disorder. • Clear evidence of insulin resistance: HbA1c, acanthosis nigricans. • Secondary causes of obesity (genetic syndromes such as Prader-Willi) are evident or strongly suspected. • Poor linear growth or short stature in comparison with excessive weight gain. • Short history (<12 months) of marked weight gain. • History of brain injury, brain tumor, CNS disease. • Suggestive phenotypic features: developmental delay, significant obesity beginning before three years. • When an obesity-related complication is confirmed.	• Prior growth data/ chart. • Relevant lab studies. • Pertinent medical records. • Results of any additional tests.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Precocious puberty	History and exam (please include Tanner staging). FSH, testosterone (males and virilized females), estradiol, TSH, Free T4, DHEAS, 17 OH progesterone. Radiology: bone age.	• Breast development or pubic hair in girls <eight years. • Testicular enlargement (3 cc or >2.5 cm), increased penile size or pubic hair in boys <nine years. • Linear growth increasing, with advanced bone age.	• Prior growth data/ charts. • Relevant lab studies. • Ask patient's family to bring bone age X-ray to clinic, if completed. • Pertinent medical records. • Results of any additional tests.
Early childhood breast development in girls Palpable breast buds in girls less than 24 months is not uncommon and usually not of concern.	History and exam. FSH, estradiol, TSH, Free T4, LH.	• Progressing over time. • Accelerated growth, linear velocity. • Vaginal bleeding. • Café au lait spots on physical exam (possible McCune-Albright syndrome).	• Prior growth data/ charts. • Relevant lab studies. • Pertinent medical records. • Results of any additional tests.
Delayed puberty Chronic illness should be considered.	History and physical exam. CBC, ESR, CMP, TSH, Free T4 or T4 total, prolactin, LH, FSH, estradiol, testosterone: morning read (male), celiac screen. Radiology: bone age.	• For boys: no testicular enlargement by 14 years (4 ccs, 2.5 cms). • For girls: no breast development by 13 years, or no menses by 16 years, or no menses ≥four years after onset of breast development. • More than six months without a menstrual cycle.	• Prior growth data/ charts. • Relevant lab studies. • Ask patient's family to bring bone age X-ray to clinic, if completed. • Pertinent medical records. • Results of any additional tests.
Premature menses Consider vaginal foreign body or trauma.	History and exam. FSH, prolactin, estradiol, TSH, Free T4 Radiology: pelvic ultrasound, bone age.	• Vaginal bleeding in girls <ten years. • Vaginal bleeding in any girls without signs of puberty.	• Prior growth data/ charts. • Relevant lab studies. • Ask patient's family to bring bone age X-ray to clinic, if completed. • Pertinent medical records. • Results of any additional tests.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Congenital hypothyroidism Urgent referrals recommended. Appointments within 24 hours.	History and exam. Thyroid function (TSH and Free T4).	• Abnormal newborn screen. • Please follow instructions of the State of Michigan newborn screening program. For questions, please call Corewell Health Helen DeVos Children's Hospital Direct phone: **616.391.2345** to be connected to on-call endocrinologist.	• Thyroid function tests, including results from State of Michigan newborn screening program and any other labs obtained. • Birth history, gestational age, weight and height.
Acquired hypothyroidism If thyromegaly, please see referral guidelines for goiter	History and exam. TSH, if elevated TSH, TPO will provide autoimmune study, Free T4. Please see co-management guidelines for details regarding lab level decision-making.	• If TSH is elevated and free T4 is normal, please see co-management guidelines. • Refer if Free T4 is low. No referral is necessary: • If TSH and Free T4 are normal – even if thyroid antibodies are positive – but, consider repeating labs in three to six months. • If normal TSH and elevated TPO.	• Prior growth data/ charts. • Pertinent medical records. • Relevant lab studies, including thyroid peroxidase antibody, if obtained. • Thyroid scan and ultrasound is not needed, but please provide if obtained. • Results of any additional tests.
Acquired hyperthyroidism (Grave's Disease) Goiter is not always present. Appointments available within 24 hours.	History and exam TSH, Free T4, Total T3, thyroid stimulating immunoglobulin, thyroid binding inhibitory. Radiology: thyroid scan, ultrasound.	• Suppressed TSH. • Elevated T4: Total or free. • Elevated T3: Total or free.	• Prior growth data/ charts. • Pertinent medical records. • Relevant lab studies. • Results of any additional tests.
Goiter/ thyromegaly	History and exam. Thyroid function (include TSH and Free T4; Total T3 may be helpful if TSH is suppressed and Free T4 is normal), thyroid peroxidase antibody.	• Abnormal thyroid function tests. • Palpable nodules or asymmetry. • Increasing in size. • Causing discomfort.	• Prior growth data/ charts. • Pertinent medical records. • Relevant lab studies. • Results of any additional tests.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Calcium disorders Consider urgent referral for symptomatic hypocalcemia, hypercalcemia, total calcium. <7mg/dl or >12 mg/dl, ionized calcium <0.9 mmo/L or >1.6 mmo/L	History and exam. CMP, ionized calcium, phosphorus, magnesium, PTH, 25-OH Vitamin D, 1,25 OH Vitamin D, urine Ca/Cr, skeletal survey for rickets.	• Low or elevated calcium. • Elevated phosphorus. • Evidence of Rickets with a normal or elevated 25 OH Vitamin D. Note: Nutritional rickets is a common disorder that can be managed by the primary care provider. No referral or DEXA scan is required. We are available to assist with questions or concerns.	• Prior growth data/ charts. • Relevant lab studies. • Ask patient's family to bring bone age X-ray to clinic, if completed. • Pertinent medical records. • Results of any additional tests.
Hypoglycemia Note: The definition of hypoglycemia in infants and children continues to be controversial.	History and exam. Serum glucose; if possible, obtain the following critical sample at the time of hypoglycemia: venous serum glucose (not POC), insulin level, c-peptide, beta hydroxybutyrate, cortisol, growth hormone, free fatty acids, lactate, urine ketones.	• Documented hypoglycemia (plasma glucose <50 mg/dl).	• Prior growth data/ charts. • Relevant lab studies. • Pertinent medical records. • Results of any additional tests.
Adrenal insufficiency Urgent appointments available for new diagnosis and positive newborn screen.	History and exam. CMP, glucose, morning cortisol and ACTH (before 9am); if primary adrenal disease is suspected, consider also obtaining renin and aldosterone.	• Low morning cortisol level.	• Prior growth data/ charts. • Relevant lab studies. • Pertinent medical records. • Results of any additional tests.

Pediatric gastroenterology

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. NE | Suite 4150

Outreach locations:
Lansing, St. Joseph, Traverse City

About pediatric gastroenterology

We accept referrals for children up to age 18.

Most common referrals

- Abdominal pain.
- Constipation/encopresis.
- Diarrhea.
- Vomiting.
- GERD.
- Suspected inflammatory bowel disease, celiac disease or eosinophilic esophagitis.
- Elevated liver enzymes or cholestasis.
- Failure to thrive.
- Dysphagia/feeding problems.
- Short bowel syndrome/intestinal failure.

Pediatric gastroenterology appointment priority guide

Immediate	Contact Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** and ask to speak to the on-call gastroenterologist and/or send to the closest emergency department.
Urgent	Likely to receive an appointment within two days. Send referral via EPIC care link, fax completed referral form to **616.267.2401**, or send referral through Holon
Routine	Likely to receive an appointment within 10 days. Send referral via EPIC care link, fax completed referral form to **616.267.2401**, or send referral through Holon

Diagnosis/ symptom	Suggested workup/initial management	When to refer	Information needed
Abdominal pain	Diet modification: eliminate carbonated beverages, caffeine, gum chewing, and decrease intake of greasy or gas-producing foods, consider trial of dairy-free diet. Consider: • Counseling to address potential stress/anxiety issues and to learn relaxation techniques. • Trial of a probiotic. • Trial of an antispasmodic (hyoscyamine or dicyclomine). • Trial of a stool softener (PEG 3350). If not improving with the above recommendations, consider CBC/differential, CRP, ESR, CMP, lipase, total IgA, transglutaminase antibody, include deamidated gliadin antibody if patient <three years of age, urinalysis, fecal hemoccult x3. Would not recommend imaging unless lab abnormalities or symptoms suggest a more specific diagnosis.	For patients zero to four years: • If persistent for more than two weeks, or if accompanied by persistent fever, diarrhea, vomiting, weight loss/growth failure or GI bleeding. For patients >five years: • If pain is persistent for more than six weeks and no improvement with conservative IBS management techniques, or if accompanied by persistent fever, diarrhea, vomiting, weight loss/growth failure or GI bleeding.	• Growth chart. • All lab and radiology reports. • List of treatments tried.

Diagnosis/ symptom	Suggested workup/initial management	When to refer	Information needed
Constipation/ encopresis	Diet modification: decrease intake of dairy, increase intake of water/high fiber foods. Colonic clean out*, if indicated, and then daily use of stool softener. Behavioral techniques (regular toilet time/sticker chart system). If not improving with the above recommendations: • Consider KUB if needed to assess fecal load or if obstruction suspected. • Consider barium enema if Hirschsprung's Disease or neurogenic bowel suspected. – MRI if concerned about tethered cord. • Consider CBC/differential, CMP, TSH, total IgA, transglutaminase antibody, deamidated gliadin antibody if patient <three years of age. • Consider sweat chloride.	• If not responding to standard bowel regimen, or accompanied by obstructive symptoms or urinary incontinence.	• Growth chart. • All lab and radiology reports . • List of treatments tried.

Constipation regimen guidelines

Colonic clean out*:

- PEG 3350 one capful (17 gms) per year of age daily (maximum dose 14 capfuls/day).
- Mix in Gatorade or other clear liquid, can mix 17 gms per four ounces of liquid for the duration of the clean out.
- Give daily for three consecutive days.
- Can stop clean out early if passing clear stools.
- Maintain a primarily clear liquid diet during clean out to obtain best results.

Maintenance:

- PEG 3350 one capful (17 gms) daily mixed in eight ounces clear liquid.
- Dose can be titrated by ½ capfuls as needed to achieve soft daily stools.

*Use caution to avoid dehydration during clean out in patients <two years of age, with fixed fluid intake, or with renal disease.

Diagnosis/ symptom	Suggested workup/initial management	When to refer	Information needed
Diarrhea	Trial of two-week dairy free diet and/or decrease clear liquids, caffeinated beverages and simple sugars. If blood in stool, or if patient fails to respond to dietary management: • Fecal hemoccult x3, fecal lactoferrin or calprotectin, bacterial culture or enteric pathogen PCR, O&P (complete if patient is immunocompromised or has history of recent foreign travel), C. diff screen if patient >two years of age, CBC/differential, CMP, CRP, ESR, total IgA, transglutaminase antibody, deamidated gliadin antibody if patient <three years of age. • Consider sweat chloride. • Consider fecal pancreatic elastase if there are growth concerns (weight or height).	• After infectious etiologies have been ruled out and appropriate dietary management has been initiated. • And, if persistent for more than two weeks, or accompanied by blood in stool or associated weight loss/growth failure.	• Growth chart. • All lab and radiology reports. • List of treatments tried.
Vomiting	Consider trial H2 antagonist or proton pump inhibitor Consider CBC/differential, CRP or ESR, CMP, lipase, total IgA, transglutaminase antibody, deamidated, gliadin antibody if patient <three years of age, urinalysis. Although not routinely recommended, if you feel helicobacter pylori testing is necessary, obtain fecal h. pylori antigen or urease breath test not h. pylori serology. Would **not** recommend helicobacter pylori testing in patients <one year of age. Consider KUB or UGI if anatomic etiology suspected.	• If persistent for more than two weeks. • If experiencing recurrent episodes more than four times per year. • If accompanied by bilious emesis or hematemesis may need immediate referral to emergency department.	• Growth chart. • All lab and radiology reports. • List of treatments tried.

Diagnosis/ symptom	Suggested workup/initial management	When to refer	Information needed
GERD	Conservative GERD measures (see NASPGHAN guidelines). Consider trial H2 antagonist or proton pump inhibitor if H2 antagonist not effective. Consider UGI if dysphagia present or anatomic etiology suspected. **naspghan.org/professional-resources/medical-professional-resources/reflux-gerd/**	• If accompanied by weight loss or failure to thrive, respiratory symptoms, severe irritability in an infant or nonverbal patient, dysphagia, or pain despite observing conservative anti-reflux measures and using appropriate acid suppressive therapy. • If dependent on acid suppression for control of symptoms (has failed two or more attempts to wean acid suppression). • If accompanied by bilious emesis or hematemesis may need immediate referral to emergency department.	• Growth chart. • All lab and radiology reports. • List of treatments tried.
Inflammatory bowel disease	Fecal hemoccult x 3, lactoferrin or calprotectin (calprotectin preferred, if a covered benefit), bacterial culture or enteric pathogen PCR (bacterial culture preferred if a covered benefit), O&P (complete if patient is immunocompromised or has history of recent foreign travel), C. diff screen if patient >two years of age), CBC/differential, CMP, CRP, ESR, total IgA, transglutaminase antibody, deamidated gliadin antibody if patient <three years of age. Please do not initiate corticosteroid therapy for IBD before consulting with pediatric gastroenterology.	• Immediate referral: If inflammatory bowel disease is strongly suspected.	• Growth chart. • All lab and radiology reports. • List of treatments tried.
Elevated liver enzymes	Provided on a case-by-case basis. In patients with BMI ≥95% or acute significant weight gain with mild elevation of transaminases (less than twice the upper limit of normal), initiate lifestyle modification strategies (most importantly elimination of sugar-sweetened beverages) and re-check in one to six months.	• Elevated liver enzymes (ALT greater than 44 for girls, 52 for boys) for over one month.	• Growth chart. • All lab and radiology reports. • List of treatments tried.

Diagnosis/ symptom	Suggested workup/initial management	When to refer	Information needed
Cholestasis	Provided on a case-by-case basis. Initial ultrasound of the liver with doppler may be helpful if it can be performed promptly. Initial lab tests include: CMP **with direct bilirubin,** GGT, CBC, PT/INR.	• Urgent referral: Any infant or child with cholestasis (elevated direct bilirubin, >20% of total bilirubin). Do not delay referral if labs are unable to be obtained. • Contact HDVCH Direct (616.391.2345) and ask for on-call gastroenterologist regarding any cholestatic infant.	• All prior lab testing including imaging studies. • Growth chart. • Previously obtained laboratory studies.
Failure to thrive	For infants, fortify calories in formula or supplement breast feeding with bottle feeding. For toddlers and older children, supplement with Pediasure or equivalent formula. Consult with a dietician. Consider CBC/differential, CMP, CRP, ESR, TSH, total IgA, transglutaminase antibody, deamidated gliadin antibody if patient <three years of age, sweat chloride, fecal pancreatic elastase, urinalysis.	• If patient fails to respond to dietary modification. Consider pediatric endocrinology referral.	• Growth chart. • All lab and radiology reports. • List of treatments tried.
Celiac disease	Consider CBC/differential, CMP, CRP, ESR, total IgA, transglutaminase IgA antibody, include deamidated gliadin antibody if patient <three years of age. First degree relatives: Screen asymptomatic patients >three years of age, or symptomatic patients <3 years of age. total IgA, transglutaminase IgA antibody, include deamidated gliadin antibody if patient <three years of age. Please do not initiate gluten free diet before consulting with pediatric gastroenterology.	• If celiac antibody testing is positive.	• Growth chart. • All lab and radiology reports. • List of treatments tried.
Feeding problems or dysphagia	Obtain outpatient feeding evaluation and swallow study. Consider esophagram. Consider trial of H2 antagonist or PPI.	• If patient fails to improve with feeding therapy and/or acid suppression. • If esophagram demonstrates stricture or other abnormality.	• Growth chart. • All lab and radiology reports. • List of treatments tried.

Pediatric hematology oncology and vascular anomalies/ malformations

Consult and referral guidelines

Pediatric hematology oncology clinic referring provider line: **616.267.1908** (business hours)

Corewell Health Helen DeVos Children's Hospital
100 Michigan St. NE

Traverse City
217 S. Madison St.

About pediatric hematology oncology

We care for children and teens from birth to age 21.

Most common referrals

- 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, hemihypertrophy, Li-Fraumeni, neurofibromatosis and others.
- Lymphadenopathy.
- Abnormal coagulation tests.
- Bleeding disorders including hemophilia, other factor deficiencies, von Willebrand disease.
- Hereditary thrombophilia including factor V Leiden mutation.
- Hemangiomas: infantile and congenital.
- Vascular anomalies and malformations.
- Lymphedema.

Pediatric hematology oncology appointment priority guide

Immediate	During business hours, call referring provider line at **616.267.1908**. After hours and on weekends, contact Corewell Health Helen DeVos Children's Hospital Direct phone: **616.391.2345** and ask to speak to the on-call pediatric hematology 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 referring provider line at **616.267.1908**. After hours and on weekends, call Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** and ask to speak to the on-call pediatric hematology 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 Holon.

Diagnosis/symptom	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 and 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 (not satellite lab): CBC with manual differential, CMP, LDH, phosphorus, uric acid. Obtain chest X-ray.	• Urgent referral: Patient with large lymph nodes (>2 cm) without known infectious etiology, or firm, non-tender or matted lymph nodes. 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).
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 and 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): CBC with manual differential, CMP, LDH, phosphorus, uric acid. Consider chest X-ray.	• Immediate referral: If there is concern for acute leukemia based on physical exam or laboratory findings. 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/symptom	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 and 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 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 and 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). • Pediatric patients with an extremity mass are often referred to Corewell Health Orthopedic Oncology 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/symptom	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 and history of NF-1. Physical exam paying attention to weight/growth curve, complete neurologic exam and 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 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 ED. This helps us to make quick schedule changes in those challenging situations where we are asked to meet a patient in the ED 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 Weidemann, 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 corticocarcinoma. Consider ordering abdominal ultrasound prior to consultation visit.	• Routine referral: We will generally follow these children until eight years of age.	• Office notes. • Any prior workup.
Familial cancer syndromes Including Li-Fraumeni, von Hippel Lindau, Lynch syndrome, Familial Adenomatous Polyposis syndrome.	Referral to medical 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.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Conditions associated with bone marrow failure	Detailed history. Physical exam paying particular attention to microcephaly, features of VACTERL association, thumb anomalies and short stature. Obtain CBC with manual differential, reticulocyte count.	• Immediate or urgent referral: Those with significant pancytopenia 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: retic 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.
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.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Microcytic anemia (Low hemoglobin with low MCV.)	Detailed history, especially diet history (quantitation of cow's milk), menstrual history and any GI symptoms. Labs: 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. • Immediate or urgent referral: Patients with hemoglobin less than 7g/dL, depending on patient factors. • Routine referral: Patients with lack of response to oral iron supplementation after two weeks, ensuring medication adherence. • Males and non-menstruating females above two years with iron deficiency anemia without excessive milk intake but with normal iron absorption will likely need referral to Pediatric Gastroenterology for possible inflammatory bowel disease.	• Office notes. • Any prior workup including all laboratory studies.
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 <three years. • 250 mg twice daily for child >three years.	• All patients should be referred and will be seen for routine consultation.	• Office notes. • Newborn screening results.
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.	• One-time routine consultation to discuss inheritance, etc., is offered.	• Office notes. • Newborn screening results and/or hemoglobin fractionation.
Alpha thalassemia trait or beta thalassemia trait	Patients with thalassemia trait do not need ongoing care from a hematologist.	• One-time routine consultation to discuss laboratory findings, inheritance and potential confusion with iron deficiency anemia is offered.	• Office notes. • Newborn screening results and/or hemoglobin fractionation.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Newborn with Rh sensitization	No additional work up needed beyond usual CBC/retic and bilirubin.	• We like to see these patients prior to hospital discharge; inpatient consult should be requested. • Urgent referral: If born outside of Corewell Health Helen DeVos Children's Hospital, refer so that monitoring plan can be put in place.	• Office notes. • Birth records, including laboratory studies.
Newborn with ABO incompatibility	No additional work up needed beyond usual CBC/retic 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.
Newborn with family history of hereditary spherocytosis (HS)	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 >20,000 but <50,000 without other cytopenias will be seen on an urgent or routine basis depending on patient factors. • Platelets >50,000 will be seen on a routine basis.	• Office notes. • Any prior workup.

Diagnosis/symptom	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 sent immediately to the ED, and will be hospitalized.** Contact us to assist with fast-tracking these patients in the ED. • Patients with neutrophils >500/uL but < 1000/uL with fever **should be sent immediately to the ED for treatment of fever and neutropenia, but will not necessarily be hospitalized.** Contact us to assist with fast-tracking these patients in the ED. • Routine referral: Patients with persistent neutropenia with ANC <1000/uL over at least three to six weeks may need referral. Phone consultation should be first step. • Patients with persistent neutropenia with ANC >1000/uL but <1500/uL may not necessarily require referral. Phone consultation should be first step.	• Office notes including growth charts. • Any prior workup.
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 eight 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 work up.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
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.
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 aPTT 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 and aPTT 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.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
History of familial thrombophilia <u>without</u> 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.
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 and 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: 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: 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/symptom	Suggested workup/initial management	When to refer	Information needed
Capillary malformation on face in V1, V2 distribution, high risk for Sturge-Webber syndrome	MRI brain should be ordered.	• We do not generally care for these patients 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, pediatric ophthalmology and potentially pediatric neurology. If there is uncertainty in terms of diagnosis, we are happy to see the patient and make recommendations for referral.	
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 infectious diseases

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. NE | Suite 4150

Outreach location:
Lansing

About pediatric infectious diseases

We care for children and teens from birth to age 18.

Most common referrals

- Chronic or recurrent infections.
- Chronic or recurrent fevers.
- Recurrent sinopulmonary or otitis infections.
- Immunodeficiency evaluation and care.
- Chronic hepatitis B management.
- Hepatitis C (chronic infection management and evaluation of infants born to hepatitis C-positive mothers).
- HIV management and care (including care for infected children, perinatal evaluation and non-occupational postexposure prophylaxis [nPEP]).
- Histoplasmosis.
- Lyme Disease.
- Recurrent MRSA infections.
- Travel medicine clinic.
- Adoption counseling for adoptees with HIV, hepatitis B or hepatitis C.

Pediatric infectious diseases appointment priority guide

Immediate	Contact Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** and ask to speak to the on-call infectious diseases physician and/or send to the closest emergency department.
Urgent	Likely to receive an appointment within two days. Call Corewell Health Helen DeVos Children's Hospital Direct and ask to speak to the on-call infectious diseases physician regarding an urgent referral.
Routine	Likely to receive an appointment within seven days. Send referral via EPIC care link, fax completed referral form to **616.267.2301**, or send referral through Holon.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
General referrals			• Records including growth chart, immunizations, previous cultures with sensitivities, CBC and radiographic studies.
Chronic or recurrent infections	Detailed history of infectious history, IgG (subclasses note recommended), IgA, IgM levels, CBC, culture results.		See general referrals. • Antibiotic courses given. • Any positive family history for immune deficiencies.
Chronic or recurrent fevers	Detailed history of fever episodes with associated symptoms/signs, fever log, CRP, ESR and culture results (while febrile and afebrile).	• Ongoing fevers for more than three episodes, or concerning associated symptoms.	See general referrals. • Antibiotic courses given. • Any positive family history for periodic fevers.
Recurrent sinopulmonary or otitis infections	Evaluation for atopy, cultures and sensitivities. IgG, IgA and IgM levels.	• Consider ENT evaluation before referral or concurrently.	See general referrals. • Antibiotic courses given. • ENT, pulmonology and/or allergy notes.
Immunodeficiency, evaluation and care	IgG, IgM and IgA levels, CBC with differentials. Documentation of infections with unusual or opportunistic organisms (pneumocystis jiroveci pneumonia, mycobacterium, candida infections in older children).	• If there is recurrent or persistent infections, an unusual organism causing infection, severe course of a typically mild infection, or family history of immunodeficiency. • If the newborn screen for SCIDS is positive, immediately call Corewell Health Helen DeVos Children's Hospital Direct phone: **616.391.2345** and ask for the on-call allergist/ immunologist. If they cannot be reached, ask for the on-call infectious diseases physician.	See general referrals. • Immunoglobulin levels. • FISH 22q11 if DiGeorge. • Any flow cytometry results (if performed).

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Chronic hepatitis B management	Hepatitis B viral load, hepatitis B antigen/antibody, complete metabolic profile, alpha fetoprotein level, CBC, hepatitis C testing, HIV testing.	• When a pediatric patient is identified as having positive hepatitis B.	See general referrals. • Previous or current antiviral therapy. • Adoption or refugee papers (if an international immigrant). • Any liver ultrasound studies.
Mom with diagnosis of hepatitis C	Hepatitis C viral load and HIV testing of mother		
Chronic hepatitis C management	After the child has been identified as having hepatitis C: hepatitis C viral load, hepatitis C antibody, complete metabolic profile, alpha fetoprotein level, CBC, hepatitis B testing, HIV testing. Nucleic acid viral load if child is <18 months.	• When a pediatric patient is identified as having positive hepatitis C, or was born to a hepatitis C-positive mother.	See general referrals. • Any liver ultrasound studies.
HIV management **Care for infected children**	Labs: HIV antibody, HIV viral load, CD4 count, CBC with differential, complete metabolic profile.	• When a pediatric patient is identified as having HIV, including international adoptees and refugees.	• Initial management labs. • Previous and current antivirals. • Prior or current opportunistic infections. • Developmental status. • Psychiatric comorbidities. • Nutritional status.
Perinatal evaluation	Labs: HIV DNA or RNA, PCR, CBC with differential, complete metabolic profile.	• When an infant is born to a mother with known or suspected HIV infection.	• Maternal HIV testing results. • Maternal treatment history. • History of maternal comorbidities.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Non-occupational post-exposure prophylaxis (nPEP)	Labs: HIV antibody, CBC with differential, complete metabolic profile, Hepatitis C antibody, hepatitis B surface antibody, hepatitis B surface antigen.	• When a child is exposed to blood or body fluids (including sexual assault) that is potentially contagious for HIV, as well as hepatitis B and/or C.	• Prior testing results for HIV, hepatitis B, hepatitis C, hepatitis B vaccine receipt. • Time of exposure.
Pre-exposure prophylaxis (PrEP)	Labs: HIV antibody, CBC with differential, complete metabolic profile.	• When an HIV-negative adolescent or teenage has increased risk of HIV infection, and desires preventative medication.	• Prior testing results for HIV and sexually transmitted infections.
Histoplasmosis	Histoplasma serologies, histoplasma urine antigen, complete metabolic profile, chest X-ray and/or thoracic CT scan.	• If symptomatic for more than one month or has pulmonary nodules.	See general referrals. • Chest radiographic studies. • Histoplasma labs.
Lyme disease	First, lyme disease serology screen. Second, confirmatory IgG and IgM Western Blot results (HDVCH currently sends to Mayo Clinic). If patient has erythema migrans bullseye rash, and reasonable exposure history, testing does not need to be performed and treatment should be given immediately.	• Treated patients without symptoms do not need to be referred. • Refer to AAP redbook for recommended antibiotic treatment. • Patients with ongoing or recurrent symptoms after initial treatment should be referred.	See general referrals. • Lyme testing results from a laboratory that uses FDA-approved assays. • Previous treatment courses.
Recurrent MRSA infections	Culture of abscess material with sensitivities, treatment with Bactrim or clindamycin. Refer to AAP website for bleach bath protocol.	• When patient has multiple infections in a short period of time or if multiple family members are having infections.	See general referrals. • Culture results with sensitivities.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Travel medicine clinic	None.	• When children, adolescents, teens and even parents will be traveling abroad. • We care for the whole family and can accommodate travelers with special needs and chronic health conditions.	• List of prior vaccinations, including routine and travel vaccines. • Anticipated travel destinations and dates of travel; it is preferable to be seen six to eight weeks before arrival in country for travel immunizations to be effective. • The cost of visits is not covered by insurance and will need to be paid out of pocket. Many vaccines will need to be paid out of pocket.
Adoption counseling For adoptees with HIV, hepatitis B or hepatitis C.	Parents considering adoption of a child with one of these infections can have a meeting with a pediatric infectious diseases physician to review available medical records. We will also discuss treatment options, prognosis and long-term care issues.	• When a potential adoptee with one of these conditions is being considered.	• Any medical records that were provided by the adoption agency. • The cost of these visits is not covered by insurance and will need to be paid out of pocket.

Pediatric medical genetics

Consult and referral guidelines

25 Michigan St. NE | Suite 2000

About pediatric medical genetics

We see both pediatric and adult patients. In many cases, our initial evaluation of the patient will result in testing/evaluation of multiple family members, as genetic testing results often have a wide-reaching impact.

Most common referrals

- Known/suspected genetic conditions such as Down syndrome, Noonan syndrome, Turner syndrome, etc.
- Single/multiple congenital anomalies and dysmorphic features.
- Counseling for previous genetic testing results.
- Family history of a genetic condition.
- Neurodevelopmental disorders, such as autism spectrum disorders, intellectual disabilities/cognitive disabilities and developmental delays.
- Neurologic conditions, including cerebral palsy, hyper/hypertonia, muscular dystrophies, ataxias and seizure disorders where individuals appear syndromic or have additional health/developmental concerns.
- Cardiovascular disease, including congenital heart defects, cardiomyopathy, long QT syndrome, arrhythmias.
- Abnormal growth, including short stature/growth restriction, overgrowth and hemihypertrophy/hemihyperplasia.
- Fetal alcohol spectrum disorders.
- Preconception counseling.

Pediatric medical genetics appointment priority guide

Immediate **urgent** **routine**	A genetics physician is on call 24/7 and can be reached by PerfectServe, EPIC in basket, email or by calling our main medical genetics clinic phone number **616.391.2700** during business hours or by calling Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** after hours or weekends. A genetic counselor is on call during business hours and can be reached by calling our main medical genetics clinic phone number.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
General	We typically do not recommend ordering genetic or other testing prior to referral. We will order or recommend studies pre- or post-evaluation if appropriate. If referring providers wish to begin studies or discuss this prior to the initial appointment, please contact our team for assistance.	• Family member with confirmed genetic disorder. • Suspicion of a genetic disorder in patient without previously diagnosed family member.	• All previous genetic testing results (for patient, or for family member if referral is for family history of genetic condition). • Family history, including specific name of the condition of concern and which family member(s) is affected. • Specific condition of concern. • Clinic notes from other subspecialty evaluations not viewable in EPIC. • Relevant lab and/or imaging results. • Relevant neuropsychological evaluation reports. • Growth charts.

Indications we do not routinely see in our office

- Personal or family history of cancer.
- Testing for an asymptomatic pediatric patient for adult-onset conditions or carrier status. Examples of this include certain types of muscular dystrophy, Huntington's disease, etc. Find details here (will have LINK).
- Ehlers-Danlos Syndrome (EDS), hypermobile type. Find details here (will have LINK).
- Personal or family history of Alzheimer's disease, when age of onset is greater than 50 years of age. Find details here (will have LINK).
- MTHFR testing or counseling of previous abnormal results with the exception of homocystinuria caused by MTHFR mutations (typically presenting in infancy). Find details here (will have LINK).
- Testing for personal or family history of autoimmune conditions, such as multiple sclerosis, HLA-B27 testing, lupus, arthritis, etc. Find details here (will have LINK).

Corewell Health Helen DeVos Children's Hospital have other specialty clinics for some genetic disorders and certain patient types and indications. If there is any uncertainty about where to refer a patient, please contact our main medical genetics clinic for assistance at 616.391.2700.

For pregnancy-related genetic concerns (e.g., family history of genetic disorder, abnormal prenatal screening) refer to Corewell Health Maternal Fetal Medicine: **616.391.3681.**

For pediatric patients **affected** with cancer or other features of a potential hereditary cancer condition (e.g., familial adenomatous polyposis, Cowden syndrome, Gorlin syndrome, Li-Fraumeni syndrome, juvenile polyposis syndrome, retinoblastoma, Peutz-Jeghers syndrome, MEN1, MEN2, hereditary paraganglioma-pheochromocytoma syndrome), refer to Corewell Health Cancer Genetics: **616-486-6218**.

For **unaffected** pediatric patients with a known family history of an adult-onset hereditary cancer condition (e.g., BRCA-related hereditary breast and ovarian cancer syndrome, Lynch syndrome), genetic testing is often deferred until the patient is 18 or older, as cancer screenings would not begin during childhood. However, if there are uncharacteristically young cancers in the family (diagnosed at age 28 or younger), referral to Corewell Health Cancer Genetics clinic and consideration of genetic testing for these (usually) adult-onset conditions is appropriate.

For **unaffected** pediatric patients with a family history of cancer, referral of parent(s) to Corewell Health Cancer Genetics is recommended to identify or rule out hereditary cancer risk to their child.

For metabolic, biochemical and mitochondrial genetic conditions or inborn errors of metabolism refer to Corewell Health Helen DeVos Children's Hospital Biochemical Genetics: **616.486.9830**.

For abnormal newborn screen results, the patient's results report will indicate the appropriate contact specific to the abnormality. For questions pertaining to newborn screening, please contact the Corewell Health Helen DeVos Children's Hospital Biochemical Genetics: **616.486.9830**.

Other specialty clinics

- For adult patients with hypertrophic cardiomyopathy (HCM) to consider genetic testing or for first degree relatives of someone with HCM for cardiac surveillance and/or genetic testing recommendations, refer to Corewell Health Cardiovascular Medicine HCM: **616.885.5192**.
- For patients with suspected or confirmed in utero alcohol exposure or suspected fetal alcohol spectrum disorder (FASD), refer to Corewell Health Helen DeVos Children's Hospital Medical Genetics FASD: **616.391.2700**.
- For patients with suspected or confirmed spinal muscular atrophy, Charcot Marie Tooth Disease or Duchenne muscular dystrophy, refer to the Corewell Health Helen DeVos Children's Hospital Neuromuscular: **616.267.2500**.
- For patients with suspected or confirmed genetic epilepsy not requiring a dysmorphology exam, refer to Corewell Health Helen DeVos Children's Hospital Epilepsy Genetics: **616.267.2500**.
- For patients with suspected or confirmed cystic fibrosis, refer to Corewell Health Helen DeVos Children's Hospital Pulmonology and cystic fibrosis care center: **616.267.2200**.
- For pediatric patients with suspected or confirmed cardiomyopathy or arrhythmia or a family history of cardiomyopathy or arrhythmia, refer to Corewell Health Helen DeVos Children's Hospital Cardiogenetics: **616.267.9150**.
- For patients with oral clefts, refer to the Corewell Health Helen DeVos Children's Hospital Oral Cleft coordinated by pediatric plastic surgery: **616.486.5885**.
- For patients with suspected or confirmed Huntington's disease, refer to medical genetics clinic: **616.391.9007**.
- For adult patients with suspected or confirmed neurofibromatosis type 1, refer to Corewell Health Helen DeVos Children's Hospital Neurofibromatosis: **616.391.2414**.
- For pediatric patients with suspected or confirmed neurofibromatosis type 1, refer to Corewell Health Helen DeVos Children's Hospital Pediatric Neurology: **616.267.2500**.
- For patients with suspected or confirmed tuberous sclerosis, refer to Corewell Health Helen DeVos Children's Hospital Pediatric Neurology: **616.267.2500**.

Pediatric nephrology

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. NE | Suite 5201

Outreach locations:
Kalamazoo, Lansing, Reed City, St. Joseph, Traverse City

About pediatric nephrology

We care for children and teens from birth to age 21.

Most common referrals

- End-stage renal disease/dialysis/transplant care.
- Congenital renal disease (i.e., dysplasia, obstructive uropathy, hydronephrosis, including abnormal prenatal imaging/prenatal consult).
- Glomerular Disorders (glomerulonephritis, proteinuria).
- Nephrotic syndrome.
- Recurrent UTIs/reflux nephropathy.
- Enuresis/voiding dysfunction.
- Polyuria/polydipsia.
- Electrolyte Imbalance/metabolic acidosis/RTA.
- Hypertension.
- Nephrolithiasis.
- Genetic renal disease (i.e., cystinosis, Lowe syndrome, etc.).

Pediatric nephrology appointment priority guide

Immediate	Contact Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** and ask to speak to the on-call nephrologist and/or send to the closest emergency department.
Urgent	Likely to receive an appointment within two days. Call Corewell Health Helen DeVos Children's Hospital Direct and ask to speak to the on-call nephrologist regarding an urgent referral.
Routine	Likely to receive an appointment within 10 days. Send referral via EPIC care link, fax completed referral form to **616.267-2401**, or send referral through Holon.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
ESRD/dialysis/ transplant		• Immediate referral: Call **616.391.2345** and ask to speak to on-call nephrologist	• Comprehensive records.
Electrolyte imbalance or abnormalities		• Any abnormalities, call **616.391.2345** with questions or concerns. • We will be glad to provide consultations and interpretation of tests and management guidance .	• Imaging and laboratory data, growth charts.
Enuresis	R/o constipation, consider polyuria or OSA. UA, behavioral modifications, consider bedwetting alarm or DDAVP trial.	• After six months of failed behavioral modifications. • Patients with non-psychogenic polydipsia and polyuria, especially if water deprivation test may need to be considered. • Immediate referral: Any secondary without a psychosocial trigger.	• Laboratory data including all urine results. • Any prior ultrasound images – please send CD if not in PACS.
Glomerular disorders Microscopic hematuria – UA with 5RBC/HPF. AND/OR protein/ creatinine >0.2 mg/mg on random (ideally first AM) analysis.	(Ideally first morning) void for protein/creatinine ratio – no need to order 24-hour urine collection. Renal panel, C3, C4, CBC, random urinary calcium/ creatinine ratio.	• Would encourage referral with any signs of glomerulonephritis and urgent with concurrent hypertension/edema and or renal dysfunction.	• All laboratory data.
Henoch Schonlein Purpura	See co-management guidelines.		• All laboratory data.
Hydronephrosis Congenital by prenatal ultrasound or found on any postnatal renal ultrasound.	See co-management guidelines.		• Any prior workup including renal ultrasounds and maternal prenatal imaging.
Hypertension	See co-management guidelines.	• Immediate referral: If symptomatic, call **616.391.2345** and ask to speak to on-call nephrologist.	• Any imaging and laboratory data.
Kidney stones and hypercalciuria As defined by renal ultrasound or CT/ suggestive history.	Renal ultrasound. Strongly discourage use of CT scan as follow-up. If stone is retrieved, pursue analysis.	• Immediate referral: If symptomatic, strongly consider Urology evaluation initially and Nephrology follow up for metabolic workup and chronic management.	• Any prior workup including renal ultrasounds if done (please send CD) and urine studies.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Gross hematuria	Renal ultrasound, UA, urine protein/creatinine and calcium/creatinine ratios. CMP, complete blood count, phosphorus, C3, C4, strep screen if appropriate.	• Immediate referral: Call **616.391.2345** and ask to speak to on-call nephrologist.	• Any prior workup including renal ultrasounds if done (please send CD) and urine studies.
Recurrent UTIs	Renal ultrasound	• Any time with recurrent urinary tract infections.	• Any prior workup including renal ultrasounds if done (please send CD). • Any prior culture results with sensitivities; urinalysis with method in which urine was obtained.

Pediatric neurodevelopmental

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. NE | Suite 3003

About pediatric neurodevelopmental

We care for children and teens from birth to age 18.

Most common referrals

- Dysphagia gia, feeding problems.
- Syndromic children with developmental delays.
- High-Risk NICU follow-up.
- Infant and early childhood development.
- Cerebral palsy.
- Gastrostomy or other tube feeding management.
- Feeding (calories, tips for formula changes, etc.).
- Sleep and other day-to-day care issues in children with delays or disabilities.

Pediatric neurodevelopmental appointment priority guide

Immediate	Contact Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** and ask to speak to the on-call neurodevelopmental provider and/or send to the closest emergency department.
Urgent	Likely to receive an appointment within two days. Send referral via EPIC care link, fax completed referral form to **616.267-2401,** or send referral through Holon.
Routine	Likely to receive an appointment within one to four weeks. Send referral via EPIC care link, fax completed referral form to **616.267-2401,** or send referral through Holon.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Dysphagia, feeding problems Including need for tube feeding.	Refer for oromotor assessment (speech or occupational therapy; varies in different centers). Consider videofluoroscopic swallowing study.	• Concerns not address by prior assessment. • Coordinated management with other Corewell Health Helen DeVos Children's Hospital specialists.	• Therapy and swallow study reports.
Syndromic children with developmental delays	Genetic testing. Metabolic labs. Refer to early on.		
High risk NICU follow-up Babies born at less than 31 weeks gestational age. Hypoxic ischemic encephalopathy or other brain abnormality. Feeding problems.	Refer to early on.	• First visit at three months adjusted age.	• NICU discharge summary for babies outside of Corewell Health.
Infant and early childhood developmental delays	Early on/Intermediate school district referral. No other pre-evaluation is recommended.	• For consultation.	
Language delay	Refer to early on. Refer to speech therapy.	• If no improvement with therapy If has associated problems (dysmorphic features, delay in other areas).	
Cerebral palsy	As appropriate to child's picture.	• Assistance with diagnosis, counseling, early therapy management or later management. • Early and later management to include feeds, therapies and associated conditions.	

Other referral recommendations

- **Autism**
  Good first referral sources:
  - Community mental health if covered by Medicaid.
  - Autism assessment clinic if covered by commercial insurance.

  Insurance often requires a diagnosis of autism for coverage of related care. Community mental health and the autism assessment clinic can provide a diagnosis. These centers also assist with coordinating referrals. Our team can serve as a resource after the initial visit to community mental health or the autism assessment clinic.
- **Significant behavioral concerns**
  Refer to pediatric behavioral health, depending on nature and severity of the concern.
- **ADHD and/or learning disabilities**
  Refer to pediatric behavioral health if unresolved with school testing.
- **Tics** - Refer to pediatric neurology if consultation desired.
- **Advanced Spastic Cerebral Palsy**
  Consider cerebral palsy clinic.

Pediatic neurology

Consult and Referral Guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. NE

Outreach locations:
Marquette, Traverse City

About pediatric neurology

We see children and teenagers from birth to age 18s.

Most common referrals

- Seizures (first-time seizures, epilepsy and further evaluation of undiagnosed spells).
- Migraine and tension headaches.
- Nerve and muscle disorders such as muscular dystrophies, inherited neuropathies, myasthenia gravis, hyperCKemia.
- Movement disorders (tics, tourette syndrome, tremors and chorea).

Notes

- Please ensure the patient has been seen in your office for the complaint in question prior to referring to neurology so that an accurate description and confirmation of the concern is available..
- We prefer to look at all EEGs ourselves during the visit. If your patient has an EEG from a non-Corewell Health facility, we ask that the patient obtain a CD that includes all their EEGs and bring them to our office visit. If no EEG has been conducted, we can often schedule an EEG on the same day as an appointment.

Pediatric neurology appointment priority guide

Immediate	Contact Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** and ask to speak to the on-call neurologist and/or send to the nearest emergency department.
Urgent	Likely to receive an appointment within seven days. Call Corewell Health Helen DeVos Children's Hospital Direct, the practice, or use Perfect Serve to request an urgent appointment.
Routine	Some diagnoses may have a scheduling timeline. Send referral via EPIC care link, fax completed referral form to **616.267-2401**, or send referral through Holon.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Spells/seizure	Detailed history of event. EEG. Consider MRI brain without contrast if developmentally delayed (preferably at Corewell Health). Have parents video events.	• Unprovoked seizures. • Recurrent events or parental education needed.	• Detailed description of event or reason for referral. • Any prior workup. • Head circumference. • Growth charts.
Breath holding spells Episodes of crying followed by color change, loss of tone/ consciousness and occasionally seizure-l ike movements.	Consider EKG if atypical breath holding spells. CBC and ferritin.	• If episodes do not follow typical sequence - three months to three years - and no antecedent crying.	• Detailed history of spells. • Head circumference. • Growth charts.
Febrile Generalized tonic-clonic convulsion associated with fever (>101° F) in an otherwise neurologically normal child (six mo. – six yrs.) with no prior afebrile seizures.	If simple febrile, no focal features, <15 mins, then no additional workup required. Parental reassurance. Education regarding diagnosis.	• Complex febrile, focal features, recurrent, >15 mins. • Developmental delay, neurological abnormalities or development of non-febrile seizures, positive family medical history for epilepsy.	• All previous workup results. • Head circumference. • Growth charts.
Tics/tourette spectrum Movement is repetitive, quick, brief and typically worsens with stress, anxiety or excitement. Vocal component is similarly repetitive and may include cough, snort, bark, s niff, throat clearing (among others). Tourettes: Tics are common, with motor and vocal components appearing for a year or more.	None required. Parental education regarding diagnosis and reassurance. ASO titer is not indicated. Monitor for common co-morbidities: anxiety, OCD and depression. As a general rule, stimulants may be used in epilepsy and tics and do not exacerbate these diagnoses.	• Characteristics of seizure, refractory, symptoms interfere with ADLs.	• Description of tics. • Evaluation of psychiatric to morbidities and prior/ current treatments. • Head circumference. • Growth charts.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Neuro muscular disorders Chronic muscular weakness, slowly progressive muscular weakness, distal limb atrophy, cramping with exercise, identification of muscle hypertrophy.	CK, repeat if abnormal. Physical therapy.	• Elevated CK, loss or regression of motor skills, +Gowers sign, multisystem involvement (cardiopulmonary).	• Description of progression of symptoms. • All prior labs and imaging (on disk). • Muscle biopsy (if done). • EMG (if done). • +FMHx. • Head circumference. • Growth charts.
Headache	Evaluation and appropriate fundoscopic exam for papilledema. Imaging is optional, usually not necessary. However, if there are any red flags in the history or exam then MRI is the preferred study. Educate about failure of headache hygiene. Avoidance of rebound headache by judicious use of preventative medicine (Tylenol or Motrin two to three times per week), journal of symptoms to review potential triggers, review of psychiatric comorbidities and management by appropriate personnel.	• Failure of prophylactic medications. Options to try include: Periactin/cyproheptadine (if under eight years old), Elavil/amitriptyline, Pamelor/nortriptyline, or Topamax/topiramate. • Worrisome, focal new onset are urgent or inpatient evaluation.	• Description of headache(s). • Evaluation of psychiatric co-morbidities and treatments. • Current and previous headache treatments. • Imaging (if completed), labs. • BP records. • Head circumference. • Growth charts.
Hypotonia/ developmental delay Floppy infant.	Appropriate developmental surveillance according to AAP guidelines with early detection and monitoring of those at risk. MRI brain without contrast. Newborn screen. Chromosomal microarray. Refer to early on.	• Global developmental delay. • Loss or regression of skills or developmental milestones. • Isolated language delay, learning disorders/ school difficulty or apraxia should be referred to speech pathology. • Urgent referral: Infants with severe weakness (will try for appt. within 48 hours).	• Description of progression. • All prior labs and imaging (on disk). • Muscle biopsy (if done). • +FMHx. • Brief description of pre- and post-natal course. • Head circumference. • Growth charts.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Numbness/ tingling	Examination focused on: reflexes strength, delineation of areas of abnormal sensation. CBC, TSH, folate, lead level. Consider screening for anxiety.	• Areflexia or demyelination on MRI. • Abnormal neurologic exam, areflexia, or focal abnormalities on exam. • If associated with hyperventilation or anxiety, consider pediatric behavioral health referral.	• Description of onset and progression. • MRI/LP results (if available). • All prior labs and imaging (on disk). • Current and prior treatment. • Head circumference. • Growth charts.
Syncope History should include classic symptoms of light headedness, tunnel vision, nausea, feeling flushed, occurs most frequently with position change/ standing.	Consider EKG and EEG if atypical. Try conservative measures such as salt and fluid intake. If persistent following conservative treatment, refer to pediatric cardiology or neurology based on history of symptoms.	• If classic history, refer to pediatric cardiology. • If non-classical history, focal seizure or fall preceding spells refer to pediatric neurology. • Note: Post syncopal seizure is a reactive seizure, not a sign of underlying epilepsy and therefore does not require ongoing treatment.	• Description of spell. • EEG. • EKG. • Imaging (if completed). • Labs (CMP). • Head circumference. • Growth charts.

EEG only request guidelines

- You can order a routine EEG to be performed at Corewell Health Pediatric Neurology (35 Michigan, Suite 3003); call **616.267.2500** and ask to schedule an EEG.
- EEGs will be read by one of our pediatric neurologists. You will receive a result note within one to two weeks (patients and families should contact your office for EEG results).

Pediatric neurosurgery

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. NE

Outreach locations:
Lansing, St. Joseph, Traverse City

About pediatric neurosurgery

All referrals are reviewed and triaged by a pediatric neurosurgeon. Based on the review, referrals determined to be urgent may be seen by an advanced practice provider in consultation with the pediatric neurosurgeon to facilitate neurosurgical care. All referrals regarding head shape and or size must have all growth charts, particularly head circumference, included with the referral information.

Most common referrals

- Benign extra-axial spaces.
- Chiari.
- Low back pain.
- Sacral dimples.
- Tethered cord.
- Plagiocephaly.

Pediatric Neurosurgery Appointment Priority Guide

Immediate	Contact Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** and ask to speak to the on-call neurosurgeon and/or send to the closest emergency department.
Urgent	Call Corewell Health Helen DeVos Children's Hospital Direct and ask to speak to the on-call neurosurgeon regarding an urgent referral.
Routine	Send referral via EPIC care link, fax completed referral form to **616.267-2401**, or send referral through Holon.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Benign extra-axial spaces/ macrocephaly **Benign extra-cerebral spaces** **Benign extra-hydrocephalus** **Benign extra-axial fluid** **Extra-ventricular hydrocephalus** **Benign subdural effusion**	If performed, MRI for ventricular size or quick brains study. Ultrasound is not recommended.	• If concerning to PCP or parent. • Crossing growth percentiles on a month-to-month basis. • Orbitofrontal head circumference greater than 1 cm over two weeks. • Head circumference crosses second percentile after six months of age. • Neuroimaging reveals increased extra-axial subarachnoid spaces. Note: Increasing orbitofrontal head circumference in children up to approximately 24 months of age, secondary to immature arachnoid granulation preventing the adequate drainage of CSF into the venous system, typically resolves and does not involve neurosurgery intervention.	• Growth chart, including head circumference with notation about large head size.
Chiari	Okay to refer without MRI. MRI, if performed, should be of cervical spine with, or without, brain. The neurosurgery team only requests addition of brain imaging with an MRI if hydrocephalus may be is present.	• If not caused by trauma, headache located in the back of the head. • Valsalva induced (cough, laugh) headache. • Unless headache dominates life, treatment is not recommended.	

Definitions

- Chiari I: Characterized by abnormally shaped cerebellar tonsils that are displaced below the level of the foramen magnum.
- Chiari II: Also known as Arnold-Chiari malformation – characterized by downward displacement of the cerebellar vermis and tonsils, a brainstem malformation with beaked midbrain on neuroimaging, and a spinal myelomeningocele.
- Chiari III: Rare malformation that combines a small posterior fossa with a high cervical or occipital encephalocele, usually with displacement of the brainstem in a spinal canal.
- Chiari IV: Now considered to be an obsolete term that describes cerebellar hypoplasia unrelated to the other Chiari malformations.
- Chiari O (sub-type that is not widely used): Characterized by anatomic aberration of the brainstem (posterior pontine tile, downward displacement of the medulla, low lying obex) but with normally located cerebellar tonsils.
- Chiari 1.5 (sub-type that is not widely used): Chiari II like malformation, but without spina bifida. Both of these sub-types show crowding at the foramen magnum.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Low Back Pain Please also refer to Pediatric Orthopedics guidelines.	MRI imaging is not recommended.	• Mechanical back pain (pain that is completely relieved when a patient lies down and is brought-on when the patient stands up). • Radicular pain (reproducible pain that radiates down the leg in the same place every time and down the same leg every time). • To obtain a second opinion. • Surgery is often not the right treatment option; we will work with patients and families to find alternate care options.	

Important information about low back pain

- In nearly all cases, surgery will not be able to help a patient with back pain only.
- Spine surgery is effective for leg pain (radiculopathy). Differentiating radicular leg pain from non-dermatomal leg pain is a key part of a neurosurgery visit.
- Imaging prior to consultation is discouraged as it will not change management of the condition. Even with radicular pain, conservative management is recommended to most patients.
- We recognize the disabling nature of pain and will always support pediatricians in cases where families are seeking answers. Pediatricians do not think that a patient is a candidate for surgery to send a referral. In addition to helping patients who can benefit from surgery, the neurosurgery team will help families and patients learn why surgery could be harmful.
- Opioids are never recommended, especially for patients with chronic pain. Our office will not prescribe opioids or any other sensorium-altering medications.

Sacral dimples A pit located within the gluteal cleft, often diagnosed in the first year of life.	An ultrasound of the spine may be considered for patients <two months of age. MRI not recommended.	• Only in rare cases do sacral dimples require intervention. • An episode of meningitis requires an expedited workup to determine if the dimple communicates with the intrathecal space. • Refer if with other congenital abnormalities.	• No special information is required.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Tethered cord	MRI of lumbar spine. Note: Some insurance companies only approve this study if the order is written with contrast.	• Progressive or worsening condition. • Progressive orthopedic deformation in a child with other congenital anomalies. • Weakness. • Back pain/radiculopathy. • Leg pain (paresthesia/sensation changes, weakness, reflex changes/spasticity, progressive scoliosis, limb, gait changes). • Bowel/bladder (urinary tract infections, changes in catheterization frequency, loss or change in incontinence, constipation, frequency, loss of bladder function in children who had been potty-trained). • Also consider referrals to primary care, urology, orthopedics, physical therapy.	

Tethered cord definition

- Tethered cord: Conus of the spinal cord is at, or lower than, the superior endplate of L3. This is found through imaging.
- Tethered cord syndrome: Clinical signs and symptoms secondary to the stretch of the spinal cord and/or the nerve roots.
- Simple tethered cord: Fatty filum is greater than 2 mm.
- Complex tethered cord: A tethered cord secondary to etiology of open spina bifida (myelomeningocele) or closed spina bifida which would include lipomyelomeningocele.

Plagiocephaly	Clinical exam including ipsilateral advancement of the occiput, ear and forehead from a “bird’s eye” view. X-rays, CTs and MRI are not recommended and rarely indicated. Parental report with clinical exam is best criteria to diagnose; anthropometric measure and pictures aren’t needed. Consider referral to physical therapy. Also consider referral to plastic surgery. Alter sleep positions.	• Feel a palpable ridge. • Concerns for significant skull malformation. • Surgical correction of this disorder is almost never indicated. • Special care to be given if associated with torticollis.	

Pediatric ophthalmology

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. NE | Suite 5101

About pediatric ophthalmology

We care for children and teens from birth to age 18 and adults with strabismus or diplopia.

Pediatric ophthalmology appointment priority guide

Immediate	Contact Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** and ask to speak to the on-call ophthalmologist.
Urgent	Likely to receive an appointment within two days. Contact the welcome center at **616.267.2605** to schedule same-day or next-day appointment. Or contact Corewell Health Helen DeVos Children's Hospital Direct. Please see list below for conditions that are considered urgent.
Routine	Send referral via EPIC care link, fax completed referral form to **616.267-1408**, or send referral through Holon.

All referrals are placed through a triage process. The following qualify as urgent referrals:

- New diagnosis of concern for cataract.
- Corneal opacity or corneal ulcer.
- Infantile or congenital glaucoma.
- Leukocoria/abnormal or no red reflex.
- Acute or acquired nystagmus.
- Ocular trauma.
- Papilledema.
- Acute or sudden onset ptosis.
- Red eye not responding to treatment or of concern to the PCP.
- Conjunctivitis in infant less than 30-days old.
- Acute or acquired strabismus.
- Sudden vision loss.
- Physician request for emergent or urgent consultation/referral.

All other referrals are triaged based on patient age and diagnosis.

Infants, children and young adults

American Academy of Pediatrics

Visual System Assessment in Infants, Children, and Young Adults by Pediatricians; Committee on Practice and Ambulatory Medicine; Section on Ophthalmology, American Association of Certified Orthoptists, American Association for Pediatric Ophthalmology and Strabismus and American Academy of Ophthalmology; Pediatrics 2016; 137; Published December 7, 2015.

Schedule for visual system assessment					
Assessment	**Newborn to six months**	**Six to 12 months**	**One to three years**	**Four to five years**	**Six years and older**
Ocular history	✓	✓	✓	✓	✓
External inspection of lids and eyes	✓	✓	✓	✓	✓
Red reflex testing	✓	✓	✓	✓	✓
Pupil examination	✓	✓	✓	✓	✓
Ocular motility assessment		✓	✓	✓	✓
Instrument based screening, when available (CPT 99174)		*	✓	✓	✓ Suggested if unable to test visual acuity monocularly with age appropriate optotypes.
Visual acuity fixate and follow response	✓ **	✓	✓		
Visual acuity age-appropriate optotype assessment (CPT 99173)			✓	✓	✓

*The American Academy of Ophthalmology has recommended instrument-based screening at age 6 months. However, the rate of false-positive results is high for this age group, and the likelihood of ophthalmic intervention is low.

**Development of fixating on and following a target should occur by 6 months of age, children who do not meet this milestone should be referred.

Screening examination of premature infants for retinopathy of prematurity (ROP)

American Academy of Pediatrics

Screening Examination of Premature Infants for Retinopathy of Prematurity; American Academy of Pediatrics Section on Ophthalmology, American Academy of Ophthalmology, American Association for Pediatric Ophthalmology and Strabismus, and American Association of Certified Orthoptists; Pediatrics 2013; 131, 189; Published December 31, 2012.

Recommendation for a retinal eye exam with an ophthalmologist at intervals based on gestational age at birth and subsequent disease severity.

Timing of first eye examination based on gestational age at birth		
Gestational age at birth, in weeks	**Age at initial examination, in weeks: postmenstrual**	**Age at initial examination, in weeks: chronologic**
22*	31	9
23*	31	8
24	31	7
25	31	6
26	31	5
27	31	4
28	32	4
29	33	4
30	34	4
Older gestational age with high risk factors: consider timing based on severity of comorbidities.		4

*This guideline should be considered tentative rather than evidence-based for infants with a gestational age of 22 or 23 weeks because of the small number of survivors

Learning disability, dyslexia and vision

American Academy of Pediatrics

Joint Technical Report – Learning Disabilities, Dyslexia, and Vision; Section on Ophthalmology and Council on Children with Disabilities, American Academy of Ophthalmology, American Association for Pediatric Ophthalmology and Strabismus, and American Association of Certified Orthoptists; Pediatrics 2011; 127; e818.

Recommend vision screening and referral to ophthalmology.

Pediatric orthopedics

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. NE | Suite 4150

Outreach locations:
Lansing, St. Joseph, Traverse City

About pediatric orthopedics

We treat all orthopedic ailments in children and teens from birth to age 18.

Most common referrals

- Ankle injury: chronic and acute.
- Back pain: chronic and acute.
- Knee pain.
- Knee injury.
- Shoulder pain.
- Shoulder injury.
- Developmental dysplasia of the hip (DDH).
- Idiopathic toewalking.
- Genu varum/valgum.
- In-toeing.
- Limping child.
- Scoliosis.
- Fractures and acute injuries.
- Metatarsus adductus.
- Flatfoot.
- Clubfoot.

Pediatric orthopedics appointment priority guide

Immediate	Contact Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** and ask to speak to the on-call orthopedic surgeon and/or send to the closest emergency department.
Urgent	Likely to receive an appointment within two days. Call Corewell Health Helen DeVos Children's Hospital Direct and ask to speak to the on-call orthopedic surgeon regarding an urgent referral.
Routine	Likely to receive an appointment within 10 days. Send referral via EPIC care link, fax completed referral form to **616.267-2601**, or send referral through Holon.

Diagnosis/ symptom	Suggested workup/initial management	When to refer	Information needed
Chronic ankle injury	History and exam: assess for joint effusion, areas of tenderness and mechanical symptoms. Obtain standing AP, lateral, Mortise views. Physical therapy evaluation and treatment. Lace-up ankle brace for activities. Rest, ice, compression, elevation, NSAIDs for acute symptoms/ exacerbation.	• No improvement in symptoms after completion of physical therapy. • Abnormal imaging findings.	• History of injury. • Therapies attempted. • Imaging and reports if outside of Corewell Health.
Acute ankle injury	History and exam: assess for joint effusion and areas of tenderness including foot. Order AP, lateral and Mortise view if: • Bony tenderness. • Inability to bear weight. If skeletally mature with no abnormality on X-ray or skeletally immature with no tenderness over growth plate, begin physical therapy and offer ankle stirrup brace. Physical therapy evaluation and treatment. Rest, ice, compression, elevation, NSAIDs.	• Tenderness over growth plate in skeletally immature patient (non-displaced physeal fracture). • Bony injury on X-ray. • No improvement in symptoms and/or continued pain after physical therapy.	• History of injury. • Therapies attempted. • Imaging and reports if outside of Corewell Health.
Chronic back pain	PA and lateral spine radiographs. Weight loss for obese patients. Physical therapy evaluation and treatment. CBC with differential, if associated with constitutional symptoms concerning for malignancy.	• Abnormal radiographs. • Children less than 10 years with chronic back pain. • If symptoms persist despite physical therapy. • With associated radiculopathy or other lower extremity symptoms. • Consider referral to physical medicine and rehabilitation if normal imaging and no neurologic symptoms.	• History of injury. • Therapies attempted. • Imaging and reports if outside of Corewell Health.

Diagnosis/ symptom	Suggested workup/initial management	When to refer	Information needed
Acute back pain	Neurological exam: assess for radicular symptoms. Days of rest, if necessary. Gradual increase in activities over one to two weeks. AP and lateral spine radiographs, if symptoms persist beyond two weeks or if severe pain after trauma. Physical therapy for residual symptoms.	• Abnormal X-rays. • Neurological deficits. • Bowel/bladder dysfunction: refer directly to ED. • If symptoms persist, despite physical therapy. • Consider referral to Physical Medicine and Rehabilitation if normal imaging and no neurologic symptoms.	• History of injury. • Therapies attempted. • Imaging and reports if outside of Corewell Health.
Chronic knee pain	History and exam: assess for joint effusion, areas of tenderness, mechanical symptoms, leg rotation profile. X-rays of knee, include AP, lateral, sunrise patella. Physical therapy evaluation and treatment. Neoprene knee sleeve with activities. Consider MRI if mechanical symptoms, or if continued pain after physical therapy is completed. Consider evaluation for inflammatory condition in patients with recurrent effusions. Hip X-rays, especially in obese adolescents (evaluation for slipped capital femoral epiphysis [SCFE]).	• Mechanical symptoms of knee. • Continued pain after physical therapy completed. • Abnormal findings on X-rays or MRI. • For atraumatic recurrent effusions and pain in young children with normal X-rays, consider referral to pediatric rheumatology.	• History of injury. • Therapies attempted. • Imaging and reports if outside of Corewell Health.

Diagnosis/ symptom	Suggested workup/initial management	When to refer	Information needed
Acute knee injury	History and exam: assess hip and knee range of motion and stability. Three views of knee-standing PA/ AP, lateral and sunrise patellar view. If knee effusion within first one to two hours after injury, obtain MRI to rule out ACL/osteochondral injury. If knee effusion develops overnight – and patient has no mechanical symptoms – begin with physical therapy. Use crutches only as needed. Physical therapy may focus on joint motion, gait training, wean from crutches (if needed) and modalities as needed if adolescent. Rest, ice, compression, elevation, NSAIDs.	• Large knee effusion after injury. • Intra-articular injury on MRI. • No improvement after completion of physical therapy. • Mechanical symptoms. • Persistent effusion, beyond two to three weeks.	• History of injury. • Therapies attempted. • Imaging and reports if outside of Corewell Health.
Chronic shoulder pain	History and exam: assess major joints for effusion and generalized joint laxity, focused shoulder examination to localize primary areas of tenderness: anterior shoulder (biceps and acromio-clavicular joint), posterior shoulder and scapula, and/or lateral shoulder (rotator cuff), assess for instability of the bilateral shoulder joints, assess for voluntary shoulder subluxation/ dislocation. MRI (with athrogram) if older than 12 years and history of unilateral dislocation(s) requiring formal reduction and/or unilateral shoulder instability noted on examination.	• Significant instability or history of dislocation. • Intra-articular abnormalities on MRI (labral tear, large rotator cuff tear, chondral lesions).	• History of injury. • Therapies attempted. • Imaging and reports if outside of Corewell Health.

Diagnosis/ symptom	Suggested workup/initial management	When to refer	Information needed
Acute shoulder injury	History and exam: asses for shoulder or elbow joint effusion, localized areas of tenderness (clavicle, shoulder and elbow), instability of the shoulder joint. X-ray AP of the humerus and axillary view of the shoulder if concern for fracture or dislocation. MRI (with arthrogram) if >12 years if history of unilateral dislocation requiring formal reduction and/or unilateral shoulder instability noted on exam. Rest, ice, NSAIDs as needed. If no acute injury or abnormality on imaging studies and symptoms persist for >three weeks, may begin physical therapy. Physical therapy evaluation and treatment.	• Fracture. • Dislocation or history of instability. • Intra-articular abnormalities on MRI (labral tear, large rotator cuff tear, chondral lesions). • No improvement in symptoms after completion of physical therapy.	• History of injury. • Therapies attempted. • Imaging and reports if outside of Corewell Health.
Developmental dysplasia of the hip (DDH)	History and exam: assess for asymmetric hip range of motion, hip abduction, leg length, instability of hips. Indications for imaging include abnormal exam, breech delivery, family history of DDH (obtain ultrasound at six weeks if exam normal). Ultrasound if less than six months old, X-ray after six months.	• Abnormal imaging. • Abnormal exam.	• History of injury. • Therapies attempted • Imaging and reports if outside of Corewell Health. Note: We may order an ultrasound to be scheduled at Corewell Health prior to the patient's appointment.
Idiopathic toe walking	History and exam: assess for abnormal muscle tone or spasticity, hip/knee/ankle range of motion. Family education; most will resolve spontaneously. Assess for decreasing range of motion or contracture. Assess Gower's sign.	• Achilles tendon contracture. • Consider a Pediatric Neurology evaluation if abnormal neuro exam including abnormal muscle tone, spasticity, proximal muscle weakness or decreasing functional level.	• History of injury. • Therapies attempted. • Imaging and reports if outside of Corewell Health.

Diagnosis/ symptom	Suggested workup/initial management	When to refer	Information needed
Genu Varum/ Valgum	History and exam: observe genu varum if patient <24 months. Observe if genu valgum <seven to eight years. If genu varum persists past 24 months of age, obtain standing limb alignment X-ray with patellae pointed forward. If severe genu valgum persists past seven to eight years of age, obtain standing limb alignment X-ray with patellae pointed forward.	• Unilateral or asymmetric genu varum or valgum. • Pain affiliated with genu varum or valgum. • Genu varum persistent after age 24 months. • Severe genu valgum persistent after >seven to eight years. • Progressive severe genu varum or genu valgum.	• History of injury. • Therapies attempted. • Imaging and reports if outside of Corewell Health.
In-toeing	History and exam: assess alignment of legs for increased femoral anteversion, tibial torsion, genu valgum, and forefoot abduction, leg length discrepancy, increased muscle tone or spasticity. Family reassurance. Observation. Activity as tolerated.	• Unilateral in-toeing or significant asymmetry on exam. • Progressive malrotation. • Spasticity or increased muscle tone (consider pediatric neurology evaluation). • Increased tibial torsion persisting >five years. • Increased femoral anteversion persisting after age 10. • Leg length discrepancy >1 cm in a skeletally immature patient.	• History of injury. • Therapies attempted. • Imaging and reports if outside of Corewell Health.
Limping child	History and exam: obtain information regarding any preceding illness or trauma, assess chronicity of symptoms, examine spine, abdomen, hips and knees to help localize symptoms. X-rays of site of localized pain. If recent history of fever, CBC with manual differential, CRP, ESR. If hip or other joint is irritable, suspected joint infection or inflammatory labs are acutely elevated, refer to emergency department for evaluation.	• Abnormal findings on imaging studies. • Fever, or atraumatic limp persistent for more than 48 hours.	• History of injury. • Therapies attempted. • Imaging and reports if outside of Corewell Health.

Diagnosis/ symptom	Suggested workup/initial management	When to refer	Information needed
Scoliosis	History and exam: neurological exam. Scoliometer measurement. PA and lateral scoliosis films for scoliometer reading over seven degrees. Request evaluation of Risser scoring with X-ray order.	• Abnormal neurologic findings. • Unusual pain or symptoms. • Curves >10 degrees in children younger than 10 years. • Skeletally immature children (Risser 0-3): – Scoliometer reading ≥seven degrees in skeletally immature children. – Curves >20 degrees on X-ray • Skeletally mature children (Risser 4-5): – Curves 0-20 degrees on X-ray – no referral or monitoring necessary. – Curves greater than 20 degrees may require periodic monitoring, suggest referral.	• History of injury. • Therapies attempted. • Imaging and reports if outside of Corewell Health.
Fractures and acute injuries	Assess for focal tenderness or deformity, neurovascular function of the injured extremity. X-rays if bony tenderness or deformity. Consider removable brace or splint for comfort if X-rays normal. Rest, ice, elevation, OTC pain meds.	• Abnormal X-rays. • Consider referral to the emergency department if deformity present. • Large joint effusion on exam. • Failure of symptoms to improve with conservative treatment.	
Metatarsus Adductus	Assess flexibility of foot. If flexible, family stretching and observation.	• Rigid deformity. • Severe deformity after age two.	
Flatfoot	Assess flexibility of foot: when standing on toes, does the patient create an arch and the heel invert. Assess ankle and foot range of motion. Pain or focal tenderness. No treatment needed if painless. OTC arch support if painful.	• Rigid flatfoot (does not create an arch when on toes). • Rigid heel valgus. • Activity limiting pain after OTC arch supports.	
Clubfoot	Assess flexibility of foot. Clubfoot: • Cavus (high arch). • Adductus of the forefoot. • Varus of the heel. • Equinus of the ankle.	• Any clubfoot.	

Pediatric pulmonology and sleep

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. NE | Suite 3003

Outreach locations:
Lansing, Ludington, Traverse City

About pediatric pulmonary and sleep

We care for children and teens from birth to age 18.

Most common referrals

- Recurrent cough or wheeze.
- Recurrent bronchiolitis or bronchitis.
- Asthma.
- Bronchopulmonary dysplasia.
- Recurrent pneumonia.
- Noisy breathing or tachypnea.
- Cystic fibrosis (CF) and CF newborn screening.
- Sleep apnea/sleep disorders.

Notes

- We offer multidisciplinary clinics for cystic fibrosis, home ventilation, neuromuscular diseases and aerodigestive disorders. (For these clinics, patients are seen by a specialist at Corewell Health and followed in these clinics). For the Aerodigestive Disorders clinic, a Corewell Health ENT, pulmonologist or gastroenterologist can refer patients at one visit for all three services or if the primary care provider feels that their patient has combined lung, GI along with ear, nose and throat concerns, please send the referral to our pediatric pulmonary group and label "For aerodigestive disorders clinic."
- CF newborn screening started in October of 2007 so any respiratory condition for patients born before that date should also undergo a sweat test at a Cystic Fibrosis Foundation Accredited Lab. If there are any significant concerns for a CF diagnosis for those born after October 2007, it is prudent to order a sweat test. Although very rare, there have been a handful of false-negative newborn screens statewide.

Pediatric pulmonary and sleep appointment priority guide

Immediate	Contact Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** and ask to speak to the on-call pulmonologist and/or send to the closest emergency department.
Urgent	Likely to receive an appointment within two to five business days. Mark the referral as "urgent."
Routine	Likely to receive an appointment within 7-21 days. Send referral via EPIC care link, fax completed referral form to **616.267-2201**, or send referral through Holon.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Recurrent cough or wheeze **Recurrent bronchiolitis or bronchitis**	Chest X-ray: PA and lateral. Consider trial of bronchodilators at any age. If non-responsive to bronchodilators, consider trial of oral and/or inhaled corticosteroids. Oral prednisone is typically dosed ~2mg/kg/day x five days.	• Hospitalization. • Intubated/ICU admission. • ED visits. • Frequent need for oral steroid bursts. • Age <two years. • Unresponsive to usual therapy with increasing medication use. • Complicating conditions such as rhinitis, sinusitis, GE reflux and/or pneumonia. • Abnormal spirometry or needs frequent monitoring with spirometry. • History of chronic lung disease, prematurity.	• Chief concern. • Summary of previous treatments and response. • Respiratory history since birth. • All lab results. • All chest films.
Asthma	Chest X-ray: PA and lateral. Consider upper GI and/or video fluoroscopic swallow study. Consider allergy evaluation if signs of atopy especially for older childhood and adolescent patients.	• Has been hospitalized. • Intubated/ICU admission. • ED visits. • Frequent need for oral steroid bursts. • Age <two years. • Unresponsive to usual therapy with increasing medication use. • Complicating conditions such as rhinitis, sinusitis, GE reflux and/or pneumonia. • Abnormal spirometry or needs frequent monitoring with spirometry. • History of chronic lung disease, prematurity.	• Chief concern. • Summary of previous treatments and response. • Respiratory history since birth. • All lab results. • All chest films. • Any allergy testing and evaluations.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Bronchopulmonary dysplasia, chronic lung disease of infancy	If patient is having recurrent respiratory illnesses or increasing oxygen need, consider: • Chest X-ray: PA, lateral • UGI • Videofluoro swallow study • Cardiology evaluation • Referral to our office	• Unstable respiratory status or is slow to improve • Oxygen requirement • Difficulty growing or feeding • Problem feeding or G-tube • Re-hospitalization after discharge • Inability to wean medications and/or oxygen	• If obtained outside of Corewell Health: SaO2, echocardiograms, growth and development evaluations, all lab results post-discharge, chest films. • Current treatments and response. • Current oxygen requirements. • NICU discharge summary (if outside NICU is not in care everywhere).
Recurrent pneumonia	Chest X-ray: PA and lateral, if ruling out cystic fibrosis Sweat chloride at an accredited CF Center* Consider upper GI and/or Pediatric Cardiology consult	• Recurrent illness despite treatment • Increasing respiratory symptoms • Symptoms that interfere with daily activities • Respiratory symptoms/ infections and problems with growth and/or development	• Brief pre/postnatal history. • Growth history. • List of treatments and response. • Current treatments.
Noisy breathing and tachypnea	Babies <1 year, with stridor should see an ENT first For non-stridorous noisy breathing and tachypnea, consider a chest radiograph and upper GI	• If ENT feels a pulmonary consultation is necessary to add to the patient's care • If the patient is not improving after reflux therapy has been tried.	• Brief pre/postnatal history. • Growth history. • List of treatments and response. • Current treatments and other consultant evaluations.
Positive cystic fibrosis newborn screen From the State of Michigan: Elevated IRT plus one or more identified CF mutations.	None needed. In the rare circumstance of a suspected bowel obstruction or respiratory.	• As soon as the PCP receives a positive screen from the State of Michigan, please fax referral and newborn screen results to **616.267.2201**. Sweat test order not needed. Pulmonary clinic will call family to schedule appointment and sweat test.	• Referral to include request for consultation, pertinent history and physical.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Sleep apnea/ sleep disorders Including snoring, insomnia and hypersomnia.	Consider treatment for allergic rhinitis. Consider ENT referral. Sleep diary.	• Any symptom of sleep difficulties including sleep disordered breathing, daytime or nighttime symptoms. • Growth delay. • Nocturnal enuresis (only if associated with sleep disordered breathing).	• Chief complaint. • Pertinent history and physical, growth grid. • Treatments pursued and responses. • Any lab results. • Prior ENT evaluations. • Sleep evaluations/ studies.
Non-invasive ventilation with CPAP or BiPAP		• Most primary care providers refer to our sleep clinic for CPAP or BiPAP (PAP) management.	• Previous sleep studies. • Pertinent history and physical. • Previous PAP downloads. • Growth chart. • Any pertinent labs.
Technology Dependent with a Tracheotomy Ventilator/CPAP		• Please call Corewell Health Helen DeVos Children's Hospital Direct for provider referral.	

*Accredited CF care centers include: Corewell Health Helen DeVos Children's Hospital (Grand Rapids), Sparrow Hospital (Lansing), Bronson Hospital (Kalamazoo), Children's Hospital of Michigan (Detroit) and University of Michigan (Ann Arbor).

Pulmonary function tests (PFTs)

Our services are available for outpatient lung function interpretation at Corewell Health Pediatric Pulmonary Function Laboratory at 35 Michigan in Grand Rapids, plus the Corewell Health Pulmonary Function Laboratories in Big Rapids, Ludington and Greenville.

To request PFTs, please consider the following within your request:

- Baseline spirometry – minimum age five years.
- Spirometry with pre and post bronchodilator – administer bronchodilator only if baseline can be performed.
- Spirometry with lung volumes and airway resistance – minimum age seven years.
- Spirometry with pre- and post-lung volumes and airway resistance – minimum age seven years.

Note: For methacholine challenges and exercise studies, we recommend a pulmonary clinic referral first. For the Methacholine Challenge tests, we must order drug prior to the appointment so if the patient cannot do lung function testing at baseline, the drug is unusable. For exercise testing, there are several types and the visit takes approximately two hours. In addition, we must make sure the patient can do lung function testing and that it is safe to perform the test based on potential underlying diagnoses.

Pediatric rheumatology

Consult and referral guidelines

Corewell Health Helen DeVos Children's Hospital
Outpatient Center
35 Michigan St. N | Suite 4150

Outreach locations:
Lansing, Traverse City

About pediatric rheumatology

We care for children and teens from birth to age 18.

Most common referrals

- Arthralgias.
- Joint swelling, joint contracture, limp joint.
- Weakness.
- Back pain.
- Malar rash.
- Unexplained fevers or weight loss.
- Skin tightening or extremity color changes.
- Iritis.
- Positive (+) ANA.

Pediatric rheumatology appointment priority guide

Immediate	Contact Corewell Health Helen DeVos Children's Hospital Direct **616.391.2345** and ask to speak to the on-call rheumatologist.
Urgent	**Need**
Routine	**Need**

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Arthralgias Possible diagnosis: Juvenile idiopathic arthritis (JIA).	X-ray, if appropriate.	• If patient has persistent joint swelling, limp or joint contracture (four or more weeks).	• Any lab or imaging reports outside of Corewell Health .
Joint swelling, joint contracture, limp child and fever Possible diagnoses: JIA, systemic JIA.	Rule out infection, septic joint If suspicious, refer urgently to orthopedics or emergency department. With fever, CBC, CRP and suggest ferritin within the order.	• If patient has persistent joint swelling, limp or joint contracture that is not attributable to an orthopedic problem. • Urgent referral: With fever and orthopedics ruled out.	• Any lab or imaging reports outside of Corewell Health.
Proximal muscle weakness Possible diagnosis: Juvenile dermatomyositis (JDM).	Check for presence of typical JDM rash (heliotrope rash). Check for proximal muscle weakness. If ordering labs, check muscle enzymes: CK, AST, ALT, LDH, aldolase.	• If weakness persists, and is not attributable to a neurologic condition. • If there is a typical JDM rash.	• Any lab or imaging reports outside of Corewell Health.
Chronic back pain Possible diagnosis: JIA.	Check for sacroiliac joint tenderness, ask about morning stiffness that lasts for more than 30 minutes. Check for ability to flex and extend back. Consider X-ray or MRI (with/without) contrast for LS spine and SI joints.	• If patient shows signs of SI joint tenderness, or X-ray or MRI findings of inflammatory arthritis. • If there is a significant decrease in ROM in the back.	• Any lab or imaging reports outside of Corewell Health. • No need to order HLA B27.
Malar rash Possible diagnoses: Systemic Lupus, mixed connected tissue disease, JDM.	Other symptoms are present. If persistent (for a few weeks), consider screening for ANA (IFA).	• If rash persists or become purpuric or eroded. • If patient has other systemic signs of lupus (joint swelling, oral ulcers, serositis, cytopenias). • If ANA is positive.	• Any lab or imaging reports outside Corewell Health.
Unexplained fevers Possible diagnoses: Systemic JIA, periodic fever syndrome.	Rule out infection (first): Consider a pediatric infectious diseases consult . Rule out malignancy: Consider a pediatric oncology consult. Examine for signs of arthritis.	• If there is no evidence of infection or malignancy. • If there is family history of periodic fever syndrome.	• Any lab or imaging reports outside Corewell Health.

Diagnosis/symptom	Suggested workup/initial management	When to refer	Information needed
Skin tightening or extremity color changes Possible diagnoses: Raynaud's phenomenon, scleroderma, MCTD.	Examine for signs of sclerodactily or skin tightening, esophageal dysmotility, calcinosis, fingertip ulceration and nailfold capillary changes.	• Concern for nail fold capillary changes. • Worsening Raynaud's or concerned about secondary Raynaud's. • If there are signs of systemic disease.	• Any lab or imaging reports outside Corewell Health.
Iritis/uveitis Possible diagnoses: Juvenile idiopathic arthritis, sarcoid, other.	Refer urgently to pediatric ophthalmology. Examine for signs of systemic disease, especially arthritis.	• If ophthalmologist confirms uveitis, systemic symptoms are present and there is not an infectious cause found.	• Any lab or imaging reports outside Corewell Health.
Positive (+) ANA Possible diagnoses: JIA, SLE, Hashimotos (asymptomatic).	Examine for specific autoimmune disease (joint swelling, rash, etc.). Consider C3, C4, CBC, UA, CMP and SED rate. Examine labs for autoimmune, if labs are normal, a referral may not be necessary.	• If patients have specific signs of autoimmune disease, not just a positive ANA.	• Any lab or imaging reports outside Corewell Health.

JAMA Dermatology | Consensus Statement

Consensus Statement for the Management and Treatment of Port-Wine Birthmarks in Sturge-Weber Syndrome

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IMPORTANCE Sturge-Weber syndrome (SWS) is a neurocutaneous syndrome involving the skin, brain, and eyes. Consensus recommendations for management are lacking.

OBJECTIVE To consolidate the current literature with expert opinion to make recommendations that will guide treatment and referral for patients with port-wine birthmarks (PWBs).

EVIDENCE REVIEW In this consensus statement, 12 nationally peer-recognized experts in dermatology with experience treating patients with SWS were assembled. Key topics and questions were formulated for each group and included risk stratification, optimum treatment strategies, and recommendations regarding light-based therapies. A systematic PubMed search was performed of English-language articles published between December 1, 2008, and December 1, 2018, as well as other pertinent studies identified by the expert panel. Clinical practice guidelines were recommended.

FINDINGS Treatment of PWBs is indicated to minimize the psychosocial impact and diminish nodularity and potentially tissue hypertrophy. Better outcomes may be attained if treatments are started at an earlier age. In the US, pulsed dye laser is the standard for all PWBs regardless of the lesion size, location, or color. When performed by experienced physicians, laser treatment can be safe for patients of all ages. The choice of using general anesthesia in young patients is a complex decision that must be considered on a case-by-case basis.

CONCLUSIONS AND RELEVANCE These recommendations are intended to help guide clinical practice and decision-making for patients with SWS and those with isolated PWBs and may improve patient outcomes.

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Supplemental content

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There is a need for a consensus statement regarding an approach to managing Sturge-Weber syndrome (SWS). In 2018, the Sturge-Weber Foundation published a comprehensive review of research needs regarding the pathogenesis, clinical features, and treatment options for SWS.[1] Our consensus aims to provide clinical practice guidelines for the care of the major dermatologic feature of SWS: the port-wine birthmark (PWB).

Sturge-Weber syndrome is a sporadic, congenital, neurocutaneous syndrome involving the skin, brain, and eyes, with an estimated prevalence of 1 in 20 000 to 1 in 50 000 live births.[2] It is caused by a somatic mosaic mutation in the *GNAQ* gene located on chromosome 9q21, affecting neural crest cells emanating from the forebrain region and resulting in vascular abnormalities of the cutaneous forehead, cerebral cortex, and eye.[3,4] Patients with SWS typically have at least 2 of the following 3 components: facial PWB, vascular malformation in the brain, and vascular malformation in the eye. However, clinical manifestations vary, and workup and treatment are guided by the extent of these manifestations.

The goals of this consensus statement are to review the literature and provide an approach to risk stratification and evaluation of PWBs, offer guidance on diagnostic workup for patients with suspected or newly diagnosed SWS, and assess current treatment options for PWBs in light of the patient's age and condition severity. The treatment recommendations are currently applicable to all patients with a PWB.

Methods

Twelve national experts in dermatology were consulted to develop a consensus statement on the management and treatment of cutaneous manifestations of SWS as part of a larger consensus statement. The panel was created from a list of experts provided by the Sturge-Weber Foundation who had significant experience in treating patients with SWS and patients with PWBs who agreed to participate. Three key needs were identified: (1) risk stratification and evaluation of PWBs, (2) optimum treatment strategies for PWBs, and

Box. Key Topics for Dermatologic Management and Treatment in SWS

1. The characteristic skin manifestation of Sturge-Weber syndrome (SWS) is a port-wine birthmark (PWB), a congenital vascular malformation composed of malformed capillary-like vessels that is present at birth as a typically unilateral, bilateral, or centrally located, well-demarcated, pink to red patch on the face.
2. The best timing for evaluation of a facial PWB is at birth.
3. There are a number of factors that should be considered regarding treatment, including minimizing the psychosocial impact, diminishing nodularity and, potentially, tissue hypertrophy, and financial considerations for the family.
4. In the US, light-based devices are the standard of care for PWB treatments, and pulsed dye laser is considered first line.
5. Light-based devices are still first-line treatment for PWBs in patients with skin of color; however, higher rates of adverse effects may be seen than in lighter-skinned patients, mainly dyspigmentation and atrophic scarring. Moderate energy densities, less pulse overlap, and increased cooling are recommended in the treatment of patients with darker skin types to minimize risks.
6. There are a number of alternative therapies that have been investigated for PWBs that do not respond to traditional laser and light-based treatments.
7. The interval between laser treatments is dependent on a multitude of factors. No optimal interval has been established by scientific evaluation; thus, treatment interval must be tailored to each patient.
8. Greater rates of lightening and possible prevention of future darkening and hypertrophy may be attained if treatments are started at an earlier age. The main goal of treatment is to ensure healthy and adequate psychosocial development and minimize the stigma associated with PWBs.
9. Pulsed dye laser in young patients is a safe treatment option with a low incidence of permanent complications when performed by an experienced laser surgeon.
10. Laser treatments can be associated with significant discomfort. The choice of using general anesthesia is complex, and informed decision-making should be shared with the patients and their parents/guardians.

(3) specific recommendations regarding light-based therapies. The expert group was divided into 4 subgroups that formulated questions to address each topic. An extensive literature review was performed using PubMed for English-language articles published between December 1, 2008, and December 1, 2018, an arbitrarily selected date range, to explore articles within the past 10 years. Articles before 2008 or after 2018 were added by the expert panel based on importance. Search terms included *Sturge-Weber syndrome* plus the following: *clinical presentation*, *pathogenesis*, *risk prediction*, *port-wine birthmark* or *port-wine stain*, *diagnostic workup*, *triage*, *management*, *treatment*, *laser therapy*, *light-based therapy* or *treatment*, *photodynamic therapy*, *infantile hemangioma*, and *nevus simplex*. A total of 112 articles were identified; 76 were relevant to dermatology. This number was narrowed to 41 articles based on abstract or full-text review and supplemented with 10 additional references identified by the expert panel. Publications were assigned to questions for each key topic and distributed to each subgroup to develop responses and key guidelines, which were consolidated into 10 key topics (**Box**). One of us (S.S.) drafted the manuscript and presented it to all 4 subgroups for electronic discussion and modification. The drafts were circulated to the full expert panel and edited multiple times until each participant gave final approval.

Key Points

Question What are clinical practice guidelines for treatment and management of port-wine birthmarks, including those associated with Sturge-Weber syndrome?

Findings In this consensus statement, 10 key recommendations for treatment of port-wine birthmarks were formulated. These recommendations address risk stratification, optimum treatment strategies, and recommendations regarding light-based therapies.

Meaning The recommendations provided in this statement may help guide clinical decision-making for these patients.

Dermatologic Factors

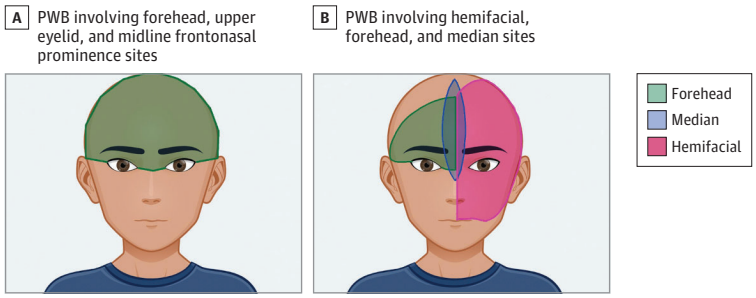
Key topic 1: The characteristic skin manifestation of SWS is a PWB, a congenital vascular malformation composed of malformed capillary-like vessels that is present at birth as a typically unilateral, bilateral, or centrally located, well-demarcated, pink to red patch on the face.

The best predictor for SWS is a facial PWB involving any part of the forehead, including the upper eyelid and the midline frontonasal prominence (**Figure**, A).[3] The distribution appears to follow the patterns of embryologic vasculature, challenging the long-held belief of a trigeminal nerve cause. Not all patients with PWBs will develop SWS; however, certain distributions indicate an increased risk. Studies have reported a 7% to 28% risk for SWS in patients with a PWB in what was previously described as a V1 distribution.[6] More recent research has reported that hemifacial, forehead, and median PWB locations are associated with increased SWS risk (Figure, B).[7] Bilateral PWBs or those that extend from the forehead to include the cheek and skin overlying the mandibles have a higher risk of SWS,[8,9] but the forehead location is the strongest independent predictor of SWS risk.[3,5] Port-wine birthmarks in SWS most often involve the lateral forehead and are less commonly localized to the midline forehead, but there are exceptions, as even a small PWB of the midline forehead can be associated with severe neurologic disease.[9]

Facial PWBs persist throughout life and may become darker red or red-purple over time. Particularly when located over the midface, facial PWBs may develop progressive vascular ectasia/thickening, soft tissue hypertrophy, and proliferative nodules that are prone to bleeding, discomfort, and less commonly, infection.[10] On histologic examination, most such nodules represent vascular ectasias, pyogenic granulomas, or arteriovenous malformations,[11] although other epithelial and mesenchymal hamartomas have been described.[12] Progression may result from both vascular ectasia and specific genetic alterations with PWBs that lead to soft tissue hypertrophy.[13] Progressive changes are uncommon before puberty. Eczematous skin changes (eg, Meyerson phenomenon) have been observed within PWBs earlier in childhood, particularly in children with preexisting atopic dermatitis.[14]

Key topic 2: The best timing for evaluation of a facial PWB is at birth.

Figure. Port-Wine Birthmarks (PWBs) With the Highest Risk of Sturge-Weber Syndrome



A, Forehead PWB location described by Waelchli et al.[3] B, Forehead, median, and hemifacial PWB locations described by Dutkiewicz et al.[5] Illustration by Sara Sabeti, BS.

Identification of an at-risk facial PWB, especially those involving the forehead, should prompt an eye examination for congenital glaucoma and neurology referral. In cases in which the diagnosis is uncertain, referral to an experienced specialist is appropriate (eAppendix in the Supplement). Differential diagnoses include segmental infantile hemangioma that may warrant a PHACE (posterior fossa anomalies, hemangioma, arterial anomalies, cardiac anomalies, and eye anomalies) workup or other capillary malformations, such as nevus simplex. Early diagnosis of a PWB affords the option of maximizing early laser treatments, which may be performed without the need for general anesthesia and may improve treatment outcome.[3]

Determination of the Optimum Treatment

Key topic 3: There are a number of factors that should be considered regarding treatment, including minimizing the psychosocial impact, diminishing nodularity and, potentially, tissue hypertrophy, and financial considerations for the family.

Patients and parents seek treatment for several reasons, including lesion appearance that affects quality of life, confidence, and self-esteem, among other psychosocial issues.[15] Laser treatments may prevent or treat the proliferative nodules that can develop over time.[16] There is no clear evidence that laser treatment definitively prevents tissue hypertrophy. However, superficial overgrowth may be minimized if adequate vessel removal is achieved. Because laser light is unable to reach deep vessels, PWBs may develop hypertrophy despite treatment.

Key topic 4: In the US, light-based devices are the standard of care for PWB treatment, and pulsed dye laser (PDL) is considered first line.

Pulsed dye laser has the longest history of efficacy and safety for treatment of PWBs, and many studies support this device as the standard.[6,17] For infants, PDL is considered the standard of care in the US. Experienced surgeons can safely perform laser surgery in patients of all ages.

Several other wavelength lasers (532, 755, and 1064 nm) and intense pulsed light have been used for PWB treatment. These techniques can be used for all PWBs but are especially useful for those that have demonstrated PDL resistance. The longer wavelengths (755 and 1064 nm) may help target larger or deeper vessels, such as those in patients with nodular and hypertrophic lesions.[6,18] These devices also target hemoglobin but have a higher risk of damage to nontargeted tissue than PDL. To our knowledge, there have been few or no randomized clinical trials with these alternative devices, and children have not been included in most of the reports. However, small studies have shown promising results for recalcitrant PWBs.[19,20] The Alexandrite laser is the most commonly used alternative when PDL is inadequate.[21] Long-pulsed Nd:YAG may also be considered but has a particularly narrow margin of safety.[6]

A variety of fractionated ablative devices designed for facial rejuvenation have also been used for PWB treatment. Many of these devices use infrared laser pulses (carbon dioxide, erbium:YAG, and erbium:glass), while others use bipolar radiofrequency ablation to coagulate skin and blood vessels. In small studies combined with PDL, efficacy has been demonstrated with these devices for recalcitrant PWBs.[22,23]

When discussing the option of laser treatment with families, several factors should be addressed, the first of which is pain control. Topical anesthetics; epidermal cooling methods; injection of local anesthetics; nerve blocks; intramuscular pain medication, such as meperidine; or general anesthesia can minimize discomfort.[16] Multiple factors are involved in choosing the optimal method of pain control for a patient, including but not limited to patient age and state of health, PWB location and extent, availability of methods dependent on practice, surgeon experience, and parent/child preference. Second, adverse effects should be discussed; these are detailed in key topic 10. Third, the family will be faced with financial obligations. Costs may include such factors as professional fees of the laser surgeon, anesthesia fees, and facility fees. The fourth factors are clinical outcomes. Realistic expectations should be set with families as complete PWB clearance is rarely achieved.[24] Pulsed-dye laser can achieve 50% to 90% clearance, and most patients will have more than 50% lightening.[6] Most patients require 8 to 10 treatments or more for optimal results; however, touch-up treatments are frequently needed even after an initial successful series of lightening. Despite considering the following factors, response is difficult to predict.

Several factors have an effect on treatment response. Patients with lighter skin types have a better treatment response.[25] Port-wine birthmarks on the face and neck respond better than those on the extremities,[17] and PWBs on the lateral face respond better than those in the central face.[22,26] Proximal extremity lesions respond better than distal extremity lesions.[17] The eyelids and neck are at higher risk for blistering and scarring, and this possibility should be considered when selecting laser parameters. Based on our experiences, individuals with PWBs associated with SWS may be more resistant to laser. Pink, red, and reticular lesions may respond better than those that are purple and geographic shaped.[17,25] Not unexpectedly, PWBs with overgrowth will show a lesser response than those that are flat, smooth, and not associated with contour change.[6,17]

Fifth, psychosocial consequences should be discussed. It is important that the short- and long-term well-being of the patient from a psychosocial perspective is considered and, above all else, the patient's safety.

Key topic 5: Light-based devices are still first-line treatment for PWBs in patients with skin of color; however, higher rates of adverse effects may be seen than in lighter-skinned patients, mainly dyspigmentation and atrophic scarring. Moderate energy densities, less pulse overlap, and increased cooling are recommended in the treatment of patients with darker skin types to minimize risks.

Port-wine birthmarks occur in patients with all skin types, and this is probably the greatest factor influencing treatment. While limited clinical studies exist for certain races/ethnicities, specifically East Asian and Indian patients, and in certain skin types, particularly Fitzpatrick skin types V and VI, a few studies provide insight for pigmented skin. In one study, PDL was used successfully in Indian patients without permanent adverse effects, although the lightening achieved was modest.[27] This study included 74 flat, nonhypertrophic PWBs with a mean of 7.3 treatment sessions and 24 hypertrophic PWBs with a mean of 8.5 treatment sessions. The mean lightening achieved was 54% in nonhypertrophic lesions and 40% in hypertrophic lesions.[27] In East Asian populations, PDL has also been used safely and successfully. While the percentage of improvement varies, reported results are slightly better than those in the aforementioned Indian study with fewer treatment sessions. In 239 Korean patients, 51.9% showed a good to excellent response, defined as greater than 51% clearance, after a mean of 4.29 sessions.[28] In a study of 848 Chinese patients, a 69.9% response was achieved after a mean of 6.2 sessions.[29] This study also underscored the importance of patient age in treatment response, with a 93.9% response rate reported in children treated during the first year of life and only a 25% response rate in adults treated when they were older than 50 years.

While patients with darker skin types can experience improvement in their PWBs, they are also at higher risk of persistent dyspigmentation, atrophy, and scarring.[17,21] In general, moderate fluences, less pulse overlap, and increased cooling are recommended in treating patients with skin of color, and patients should be counseled that transient hyperpigmentation is common.[27]

Key topic 6: There are a number of alternative therapies that have been investigated for PWBs that do not respond to traditional laser and light-based treatments.

Alternatives to laser or intense pulsed light therapy can be divided into 4 groups: (1) adjuvant medications, (2) photodynamic therapy, (3) surgery, and (4) corrective cover-up. There are currently no adjunctive medical therapies that have demonstrated consistent impressive efficacy for PWBs.[30,31] Several small studies have reported some benefit for PDL plus topical imiquimod vs PDL alone.[30] Similarly, a few small studies and case reports demonstrated some benefit of topical rapamycin as an adjunct with PDL[32,33] but no consistent benefit over PDL alone. Photodynamic therapy involves the intravenous administration of a photosensitizer (various forms of porphyrin) followed by exposure to a light source, producing intravascular singlet oxygen molecules that destroy local tissue. Although photodynamic therapy is not currently performed in the US, studies from China have shown promising results.[34-36] Approximately 20% of patients experience hyperpigmentation and scarring. However, melanin does not influence the efficacy of photodynamic therapy, so this treatment can be performed in patients of all skin types, although patients with darker skin types will still be more susceptible to pigmentary change post treatment.[6,17] Surgery can be used to selectively debulk thick PWBs or lip hypertrophy, remove larger nodules, or completely remove small lesions in which the resulting surgical scar is acceptable to the patient. A variety of corrective cover-up products and concealers are available for dermatologic conditions and can be used in patients with PWBs.

Laser and Light-Based Therapies

Key topic 7: The interval between laser treatments is dependent on a multitude of factors. No optimal interval has been established by scientific evaluation; thus, treatment interval must be tailored to each patient.

The interval between laser sessions depends on age, skin type, PWB location, pain tolerance, and presence of hypertrophy, nodules, or blebs. The interval is also influenced by resolution of prior purpura and/or hyperpigmentation, convenience, financial limitations, and potential restriction of activities after treatments. Only a few small studies in infants have analyzed the interval between PDL treatment sessions ranging from 2 weeks to 3 months.[37,38] These studies do not provide a clear recommendation on shorter vs longer intervals, although a subset of patients appears to benefit from shorter intervals. These studies have demonstrated the safety of PDL when performed at short intervals.

In older children, adolescents, and adults, it is uncertain whether there is an optimal timing interval between treatments. A small pilot study in adults suggested that 2-week treatment intervals resulted in greater lightening compared with 6-week intervals.[39] Other studies have failed to show this benefit.[40] While shown to be safe, shorter treatment intervals may result in a higher incidence of undesirable effects, especially in patients with darker skin who often benefit from longer intervals to avoid postinflammatory hyperpigmentation. It is also important to allow purpura to heal before treating again because the increased chromophore can heighten the risk of adverse effects.

Key topic 8: Greater rates of lightening and possible prevention of future darkening and hypertrophy may be attained if treatments are started at an earlier age. The main goal of treatment is to ensure healthy and adequate psychosocial development and minimize the stigma associated with PWBs.

Based on expert observations and limited studies, treatment of PWBs at an earlier age, particularly in the first year of life, results in better outcomes. Factors associated with improved prognosis in young children include proportionately smaller PWBs, more superficial and smaller blood vessels, and less melanin as a competing chromophore for PDL. Liu et al[41] found greater efficacy when treatments were started before age 6 years. Other studies have reported better responses in infants younger than 1 year, particularly with smaller PWBs (<20 cm).[3,42-44] However, these studies are limited by their retrospective design and relatively short follow-up periods. Larger case-control studies are needed to support this observation.

Current laser technology is less successful in reverting progressive PWB changes of darkening, hypertrophy, and nodularity. Thus, performing laser treatment in early childhood may prevent or minimize these changes. Limited retrospective studies support the concept that early treatment inhibits progression; however, longitudinal

studies are needed.[45,46] It has been shown that patients aged 7 to 16 years with facial differences, including PWBs, experience impaired health-related quality of life.[47] The negative impact in the child's psychosocial development and quality of life is one of the main reasons to pursue early treatment. This impact may be diminished when lightening of the PWB is attained at an earlier age.

Key topic 9: Pulsed dye laser in young patients is a safe treatment option with a low incidence of permanent complications when performed by an experienced laser surgeon.

The risks associated with laser treatment of PWBs can be categorized in 2 groups: risks inherent to the procedure and risks associated with the method of analgesia. The latter is discussed with the next key topic. The risks associated with PDL are focused on here because it is the most widely used and safest treatment option in infants and toddlers, particularly when compared with longer-wavelength vascular lasers. The safety and tolerance of PDL was also improved with the addition of cooling technology. Immediate treatment effects include erythema and purpura, which has traditionally been considered the desired clinical end point. The risk of complications has been reported to be less than 10%, and most complications are temporary.[41] Swelling may occur, which in most patients is mild except in the periorbital or lip area. Dyspigmentation may occur in response to direct epidermal and melanocyte damage and is most common in patients with darker skin types, tanned skin, and recent sun exposure. Sun protection is advised before and after treatments. Temporary blistering may occur. Erosions, ulcerations, and secondary infection are rare when appropriate laser settings are used and adequate postprocedure skin care is followed. Permanent scarring, both atrophic and hypertrophic, is one of the most serious potential complications of PDL but has an estimated incidence below 1%.[41,48,49] In addition, PDL treatment over hair-bearing areas may cause hair loss, which is typically temporary but can be permanent in an estimated 1.5% to 2.6% of cases.[50]

The most serious potential complication resulting directly from PDL is ocular damage, especially when the periorbital area is treated. Appropriate use of corneoscleral eye shields is mandatory when treating the skin within the orbital rim. Laser-specific eye shields may be used when treating outside the orbital rim. Special care must be taken when PDL is performed without sedation, as young patients may move in response to the discomfort. Securing the patient's position and ensuring eye protection is crucial when treating infants and toddlers. Parents, nursing staff, and the laser surgeon must also wear adequate protective eyewear.

Key topic 10: Laser treatments can be associated with significant discomfort. The choice of using general anesthesia is complex, and informed decision-making should be shared with the patients and their parents/guardians.

An important factor to consider when treating younger patients is the pain and discomfort associated with laser treatments. While treatment of small and moderate-size lesions is fast and generally well tolerated, patients with larger PWBs may experience significant discomfort, in which case topical anesthesia or sedation may be considered. Topical anesthetics, including lidocaine preparations, can be used safely, but there is a risk of methemoglobinemia, especially in infants.[16] Published guidelines regarding topical anesthesia should be followed if this option is chosen.[51]

General anesthesia requires careful consideration given the potential risks and higher cost. General anesthesia carries a risk of cardiorespiratory complications, which is highest in the neonatal period and decreases with age.[52,53] It is common practice to wait until the infant is at least age 6 months to use general anesthesia for elective procedures, but procedures before this age can be performed without general anesthesia. Anesthesia should be administered by clinicians specialized in pediatric care to reduce the risk of perioperative morbidity. The risk of neurotoxicity with potential long-term negative effects on neurologic development has gained recent attention.[54] The US Food and Drug Administration advises caution in patients younger than 3 years requiring repeated use of general anesthesia and sedation during surgeries or procedures, which is relevant to the management of PWBs as multiple treatments early in life are often performed. The warning was based predominantly on preclinical data, and ongoing trials may help to further clarify this risk.[54] Until more information is available, the decision to use general anesthesia or sedation must be carefully considered.

When general anesthesia or sedation is not used, young infants have an advantage over toddlers. Pulsed dye laser treatment without general anesthesia is more safely and efficiently performed in infants since the area of involvement is proportionally smaller and the patient's position and eye protection can be secured more easily.[55] Nevertheless, the potential impact of painful procedures must be carefully weighed. Noxious stimuli early in life may lead to short-term and possibly long-term effects in behavior, particularly toward medical care.[56] Parental stress and satisfaction must also be considered when making this decision.

Conclusions

This consensus statement provides expert consensus on identification and risk stratification, optimal treatment strategies, and recommendations for light-based therapies for patients with PWBs. These recommendations are intended to help guide clinical practice and decision-making for patients with SWS and those with isolated PWBs and may improve patient outcomes.

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REFERENCES

1. De la Torre AJ, Luat AF, Juhász C, et al. A multidisciplinary consensus for clinical care and research needs for Sturge-Weber syndrome. *Pediatr Neurol*. 2018;84:11-20. doi:10.1016/j.pediatrneurol.2018.04.005

2. Shirley MD, Tang H, Gallione CJ, et al. Sturge-Weber syndrome and port-wine stains caused by somatic mutation in GNAQ. *N Engl J Med*. 2013;368(21):1971-1979. doi:10.1056/NEJMoa1213507

3. Waelchli R, Aylett SE, Robinson K, Chong WK, Martinez AE, Kinsler VA. New vascular classification of port-wine stains: improving prediction of Sturge-Weber risk. *Br J Dermatol*. 2014;171(4):861-867. doi:10.1111/bjd.13203

4. Rosser T. Neurocutaneous disorders. *Continuum (Minneap Minn)*. 2018;24(1, Child Neurology):96-129. doi:10.1212/CON.0000000000000562

5. Dutkiewicz AS, Ezzedine K, Mazereeuw-Hautier J, et al; Groupe de Recherche Clinique en Dermatologie Pédiatrique. A prospective study of risk for Sturge-Weber syndrome in children with upper facial port-wine stain. *J Am Acad Dermatol*. 2015;72(3):473-480. doi:10.1016/j.jaad.2014.11.009

6. Brightman LA, Geronemus RG, Reddy KK. Laser treatment of port-wine stains. *Clin Cosmet Investig Dermatol*. 2015;8:27-33. doi:10.2147/CCID.S53118

7. Zallmann M, Leventer RJ, Mackay MT, Ditchfield M, Bekhor PS, Su JC. Screening for Sturge-Weber syndrome: a state-of-the-art review. *Pediatr Dermatol*. 2018;35(1):30-42. doi:10.1111/pde.13304

8. Kaseka ML, Bitton JY, Décarie JC, Major P. Predictive factors for epilepsy in pediatric patients with Sturge-Weber syndrome. *Pediatr Neurol*. 2016;64:52-58. doi:10.1016/j.pediatrneurol.2016.08.009

9. Dymerska M, Kirkorian AY, Offermann EA, Lin DD, Comi AM, Cohen BA. Size of facial port-wine birthmark may predict neurologic outcome in Sturge-Weber syndrome. *J Pediatr*. 2017;188:205-209.e1. doi:10.1016/j.jpeds.2017.05.053

10. Higueros E, Roe E, Granell E, Baselga E. Sturge-Weber syndrome: a review. *Actas Dermosifiliogr*. 2017;108(5):407-417. doi:10.1016/j.ad.2016.09.022

11. Chen D, Hu XJ, Lin XX, et al. Nodules arising within port-wine stains: a clinicopathologic study of 31 cases. *Am J Dermatopathol*. 2011;33(2):144-151. doi:10.1097/DAD.0b013e3181e169f5

12. Sanchez-Carpintero I, Mihm MC, Mizeracki A, Waner M, North PE. Epithelial and mesenchymal hamartomatous changes in a mature port-wine stain: morphologic evidence for a multiple germ layer field defect. *J Am Acad Dermatol*. 2004;50(4):608-612. doi:10.1016/j.jaad.2003.04.002

13. Nguyen V, Hochman M, Mihm MC Jr, Nelson JS, Tan W. The pathogenesis of port wine stain and Sturge Weber syndrome: complex interactions between genetic alterations and aberrant *MAPK* and *PI3K* activation. *Int J Mol Sci*. 2019;20(9):E2243. doi:10.3390/ijms20092243

14. Fonder MA, Mamelak AJ, Kazin RA, Cohen BA. Port-wine-stain-associated dermatitis: implications for cutaneous vascular laser therapy. *Pediatr Dermatol*. 2007;24(4):376-379. doi:10.1111/j.1525-1470.2007.00453.x

15. Hagen SL, Grey KR, Korta DZ, Kelly KM. Quality of life in adults with facial port-wine stains. *J Am Acad Dermatol*. 2017;76(4):695-702. doi:10.1016/j.jaad.2016.10.039

16. Shahriari M, Makkar H, Finch J. Laser therapy in dermatology: kids are not just little people. *Clin Dermatol*. 2015;33(6):681-686. doi:10.1016/j.clindermatol.2015.09.010

17. Updyke KM, Khachemoune A. Port-wine stains: a focused review on their management. *J Drugs Dermatol*. 2017;16(11):1145-1151.

18. Izikson L, Nelson JS, Anderson RR. Treatment of hypertrophic and resistant port wine stains with a 755 nm laser: a case series of 20 patients. *Lasers Surg Med*. 2009;41(6):427-432. doi:10.1002/lsm.20793

19. McGill DJ, MacLaren W, Mackay IR. A direct comparison of pulsed dye, alexandrite, KTP and Nd:YAG lasers and IPL in patients with previously treated capillary malformations. *Lasers Surg Med*. 2008;40(6):390-398. doi:10.1002/lsm.20638

20. Tierney EP, Hanke CW. Alexandrite laser for the treatment of port wine stains refractory to pulsed dye laser. *Dermatol Surg*. 2011;37(9):1268-1278. doi:10.1111/j.1524-4725.2011.02079.x

21. Carlsen BC, Wenande E, Erlendsson AM, Faurschou A, Dierickx C, Haedersdal M. A randomized side-by-side study comparing alexandrite laser at different pulse durations for port wine stains. *Lasers Surg Med*. 2017;49(1):97-103. doi:10.1002/lsm.22532

22. Bae YC, Alabdulrazzaq H, Brauer JA, Geronemus RG. Treatment of recalcitrant port-wine stains (PWS) using a combined pulsed dye laser (PDL) and radiofrequency (RF) energy device. *J Am Acad Dermatol*. 2017;76(2):321-326. doi:10.1016/j.jaad.2016.03.004

23. Toren KL, Marquart JD. Fractional thermoablation using an erbium-doped yttrium aluminum garnet fractionated laser for the treatment of pulsed dye laser-resistant port wine stain birthmarks. *Dermatol Surg*. 2011;37(12):1791-1794. doi:10.1111/j.1524-4725.2011.02160.x

24. Groopman J. *The Anatomy of Hope: How People Prevail in the Face of Illness*. Random House Publishing Group; 2003.

25. Fitzpatrick RE, Lowe NJ, Goldman MP, Borden H, Behr KL, Ruiz-Esparza J. Flashlamp-pumped pulsed dye laser treatment of port-wine stains. *J Dermatol Surg Oncol*. 1994;20(11):743-748. doi:10.1111/j.1524-4725.1994.tb03197.x

26. Yu W, Ma G, Qiu Y, et al. Why do port-wine stains (PWS) on the lateral face respond better to pulsed dye laser (PDL) than those located on the central face? *J Am Acad Dermatol*. 2016;74(3):527-535. doi:10.1016/j.jaad.2015.08.026

27. Khandpur S, Sharma VK. Assessment of efficacy of the 595-nm pulsed dye laser in the treatment of facial port-wine stains in Indian patients. *Dermatol Surg*. 2016;42(6):717-726. doi:10.1097/DSS.0000000000000723

28. Woo SH, Ahn HH, Kim SN, Kye YC. Treatment of vascular skin lesions with the variable-pulse 595 nm pulsed dye laser. *Dermatol Surg*. 2006;32(1):41-48. doi:10.1097/00042728-200601000-00007

29. Shi W, Wang J, Lin Y, et al. Treatment of port wine stains with pulsed dye laser: a retrospective study of 848 cases in Shandong Province, People's Republic of China. *Drug Des Dev Ther*. 2014;8:2531-2538. doi:10.2147/DDDT.S71710

30. Lipner SR. Topical adjuncts to pulsed dye laser for treatment of port wine stains: review of the literature. *Dermatol Surg*. 2018;44(6):796-802. doi:10.1097/DSS.0000000000001507

31. Passeron T, Maza A, Fontas E, et al. Treatment of port wine stains with pulsed dye laser and topical timolol: a multicenter randomized controlled trial.

Br J Dermatol. 2014;170(6):1350-1353. doi:10.1111/bjd.12772

32. Marqués L, Núñez-Córdoba JM, Aguado L, et al. Topical rapamycin combined with pulsed dye laser in the treatment of capillary vascular malformations in Sturge-Weber syndrome: phase II, randomized, double-blind, intraindividual placebo-controlled clinical trial. *J Am Acad Dermatol*. 2015;72(1):151-158.e1. doi:10.1016/j.jaad.2014.10.011

33. Musalem HM, Alshaikh AA, Tuleimat LM, Alajlan S. Outcome with topical sirolimus for port wine stain malformations after unsatisfactory results with pulse dye laser treatment alone. *Ann Saudi Med*. 2018;38(5):376-380. doi:10.5144/0256-4947.2018.376

34. Li-Qiang G, Hua W, Si-Li N, Chun-Hua T. A clinical study of HMME-PDT therapy in Chinese pediatric patients with port-wine stain. *Photodiagnosis Photodyn Ther*. 2018;23:102-105. doi:10.1016/j.pdpdt.2018.06.006

35. Wu Q, Tu P, Zhou G, et al. A dose-finding study for hemoporfin in photodynamic therapy for port-wine stain: a multicenter randomized double-blind phase IIb trial. *Photodermatol Photoimmunol Photomed*. 2018;34(5):314-321. doi:10.1111/phpp.12384

36. Zhao Y, Tu P, Zhou G, et al. Hemoporfin photodynamic therapy for port-wine stain: a randomized controlled trial. *PLoS One*. 2016;11(5):e0156219. doi:10.1371/journal.pone.0156219

37. Anolik R, Newlove T, Weiss ET, et al. Investigation into optimal treatment intervals of facial port-wine stains using the pulsed dye laser. *J Am Acad Dermatol*. 2012;67(5):985-990. doi:10.1016/j.jaad.2011.11.964

38. Swan BC, Robertson SJ, Tuxen A, et al. Pulsed dye laser treatment of capillary malformations in infants at 2-weekly versus 3-monthly intervals, reducing the need for general anaesthesia. *Australas J Dermatol*. 2017;58(3):214-218. doi:10.1111/ajd.12457

39. Tomson N, Lim SPR, Abdullah A, Lanigan SW. The treatment of port-wine stains with the pulsed-dye laser at 2-week and 6-week intervals: a comparative study. *Br J Dermatol*. 2006;154(4):676-679. doi:10.1111/j.1365-2133.2005.07113.x

40. Yu W, Zhu J, Changc SJ, et al. Shorter treatment intervals of East Asians with port-wine stain with pulsed dye laser are safe and effective—a prospective side-by-side comparison. *Photomed Laser Surg*. 2018;36(1):37-43. doi:10.1089/pho.2017.4315

41. Liu X, Fan Y, Huang J, et al. Can we predict the outcome of 595-nm wavelength pulsed dye laser therapy on capillary vascular malformations from the first beginning: a pilot study of efficacy co-related factors in 686 Chinese patients. *Lasers Med Sci*. 2015;30(3):1041-1046. doi:10.1007/s10103-014-1695-0

42. Morelli JG, Weston WL, Huff JC, Yohn JJ. Initial lesion size as a predictive factor in determining the response of port-wine stains in children treated with the pulsed dye laser. *Arch Pediatr Adolesc Med*. 1995;149(10):1142-1144. doi:10.1001/archpedi.1995.02170230096014

43. Nguyen CM, Yohn JJ, Huff C, Weston WL, Morelli JG. Facial port wine stains in childhood: prediction of the rate of improvement as a function of the age of the patient, size and location of the port wine stain and the number of treatments with the pulsed dye (585 nm) laser. *Br J Dermatol*. 1998;138(5):821-825. doi:10.1046/j.1365-2133.1998.02219.x

44. Ashinoff R, Geronemus RG. Flashlamp-pumped pulsed dye laser for port-wine stains in infancy: earlier versus later treatment. *J Am Acad Dermatol*. 1991;24(3):467-472. doi:10.1016/0190-9622(91)70075-D

45. Lee JW, Chung HY, Cerrati EW, O TM, Waner M. The natural history of soft tissue hypertrophy, bony hypertrophy, and nodule formation in patients with untreated head and neck capillary malformations. *Dermatol Surg*. 2015;41(11):1241-1245. doi:10.1097/DSS.0000000000000525

46. Minkis K, Geronemus RG, Hale EK. Port wine stain progression: a potential consequence of delayed and inadequate treatment? *Lasers Surg Med*. 2009;41(6):423-426. doi:10.1002/lsm.20788

47. Masnari O, Schiestl C, Rössler J, et al. Stigmatization predicts psychological adjustment and quality of life in children and adolescents with a facial difference. *J Pediatr Psychol*. 2013;38(2):162-172. doi:10.1093/jpepsy/jss106

48. Levine VJ, Geronemus RG. Adverse effects associated with the 577- and 585-nanometer pulsed dye laser in the treatment of cutaneous vascular lesions: a study of 500 patients. *J Am Acad Dermatol*. 1995;32(4):613-617. doi:10.1016/0190-9622(95)90346-1

49. Chiu CH, Chan HHL, Ho WS, Yeung CK, Nelson JS, Tsai RY. Prospective study of pulsed dye laser in conjunction with cryogen spray cooling for treatment of port wine stains in Chinese patients. *Dermatol Surg*. 2003;29(9):909-915. doi:10.1046/j.1524-4725.2003.29255.x

50. Feldstein S, Totri CR, Friedlander SF. Can long-term alopecia occur after appropriate pulsed-dye laser therapy in hair-bearing sites? pediatric dermatologists weigh in. *Dermatol Surg*. 2015;41(3):348-351. doi:10.1097/DSS.0000000000000284

51. Kouba DJ, LoPiccolo MC, Alam M, et al. Guidelines for the use of local anesthesia in office-based dermatologic surgery. *J Am Acad Dermatol*. 2016;74(6):1201-1219. doi:10.1016/j.jaad.2016.01.022

52. Cohen MM, Cameron CB, Duncan PG. Pediatric anesthesia morbidity and mortality in the perioperative period. *Anesth Analg*. 1990;70(2):160-167. doi:10.1213/00000539-199002000-00005

53. Habre W, Disma N, Virag K, et al; APRICOT Group of the European Society of Anaesthesiology Clinical Trial Network. Incidence of severe critical events in paediatric anaesthesia (APRICOT): a prospective multicentre observational study in 261 hospitals in Europe. *Lancet Respir Med*. 2017;5(5):412-425. doi:10.1016/S2213-2600(17)30116-9

54. Orser BA, Suresh S, Evers AS. SmartTots update regarding anesthetic neurotoxicity in the developing brain. *Anesth Analg*. 2018;126(4):1393-1396. doi:10.1213/ANE.0000000000002833

55. Jeon H, Bernstein LJ, Belkin DA, Ghalili S, Geronemus RG. Pulsed dye laser treatment of port-wine stains in infancy without the need for general anesthesia. *JAMA Dermatol*. 2019;155(4):435-441. doi:10.1001/jamadermatol.2018.5249

56. Mathes EF, Frieden IJ. Early use of laser for port-wine stains: timing, efficacy, and shared decision making. *JAMA Dermatol*. 2019;155(4):421-423. doi:10.1001/jamadermatol.2018.5189

Care of Congenital Melanocytic Nevi in Newborns and Infants: Review and Management Recommendations

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A pediatric dermatology expert working group performed a narrative review to describe care related to congenital melanocytic nevi (CMN) in neonates and infants. There are no published guidelines for most aspects of care, including routine skin care and visit intervals. Few guidelines exist for surgical management; newer recommendations favor conservative practice. Emerging evidence contributes to recommendations for screening MRI to evaluate for neural melanosis and related central nervous system complications, however, more research is needed. Risk for melanoma is generally low, but those with large, giant, or multiple CMN have a higher risk. Multidisciplinary care, with a focus on family and patient preferences, is of paramount importance. Without standardized screening and management guidelines, questions abound regarding appropriate physical examination intervals, potential treatment including full or partial excision, timing and frequency of imaging, melanoma risk, and assessment for neural melanosis.

This review highlights the current state of knowledge concerning care of patients with CMN, reveals gaps in the literature surrounding skin care, and provides management recommendations. We additionally discuss cutaneous complications of CMN, such as pruritus, hypertrichosis, and wound healing. Resources and references for families and providers can help patients navigate this sometimes challenging diagnosis. Finally, we contribute expert care recommendations to the current body of literature as a foundation for the development of future, more comprehensive care guidelines.

METHODS

A working group of pediatric dermatology experts in the Pediatric Dermatology Research Alliance convened to investigate current best practices and recommendations for the care of CMN in neonates and infants. The Pediatric Dermatology Research Alliance is a collaborative research organization linking pediatric dermatology researchers across North America. A comprehensive review of the literature from January 1, 1998, to December 31, 2017, was performed by using "congenital" and "melanocytic" and "naevus" or "nevus" as keywords in PubMed, Cochrane Database of Systemic Reviews, PyschInfo, Ovid Medline, Embase, and Cumulative Index to Nursing and Allied Health Literature (CINAHL). A second search (which also included Web of Science) of articles published from January 1, 2018, to July 24, 2020, and a hand search of articles identified from reference lists and more recent publications supplemented the initial query. Between these searches, 2594

abstract

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reports were identified, with 1144 eligible for inclusion after removing duplicates, reports in languages other than English, abstracts, and articles not related to CMN. However, the level of evidence was low, with many reviews and case reports. There were no randomized-controlled trials. No reports addressed routine skin care for patients born with CMN, although several addressed surgical interventions, imaging, and melanoma risk. As such, we performed hand search for supplemental articles and commenced a narrative review, including the most relevant, practical, robust, and impactful articles identified in this search as determined by our expert reviewers. We include recommendations (Table 1) for important aspects of care, denoting statements without published guidelines as "authors' recommendation" within the text. Although formal consensus methodology was not used, all of the authors had the opportunity to review and comment on these recommendations, and all authors approved them.

Clinical Presentation and Pathophysiology of CMN

CMN are collections of melanocytes in the skin, with variable extension to adipose tissue, to muscles, and around adnexa. They typically present at birth, although some appear after several months and can be located anywhere on the skin. The reported incidence of CMN of any size varies widely from <1% to 31.7%[1–8]; most studies calculate an incidence of <1% to 3.6%.[3–6,8] Estimates are limited by age at examination, with reports on neonates missing CMN that become visible after birth, and studies of older children potentially including more nevi than are truly congenital on the basis of recall bias and appearance. Additionally, many studies are focused on children in the United States, Europe, and Australia.[1,3,4,7,8]

Initial presentation varies from red to brown to black; they may initially be confused as vascular lesions before the brown color evolves. CMN can be flat (macules and patches), palpable (papules, plaques, and nodules), or both. They often have variations in pigment within a single lesion (Fig 1), or between lesions in patients with >1 CMN. Hypertrichosis may be present at birth or develop later (Fig 2A). Typical phenotypes are described in Table 2. CMN may change over time, making continued monitoring important. Generally, CMN are not identified prenatally.[9]

CMN typically grow in proportion to a child's somatic growth and are categorized by projected adult size (Table 2).[10] Small and medium CMN are more common than large and giant CMN, which are associated with higher morbidity and an increased risk for associated melanoma (see prognosis). Large or giant CMN can occur along with smaller melanocytic nevi, which can be present at birth or develop over time (Fig 2A). These smaller lesions were historically termed "satellite" nevi; the term "satellite," however, is neither spatially nor developmentally precise, and many authors prefer the term "multiple CMN,"[11] with the total number of CMN being important in categorization.[10] Additionally, multiple medium CMN without a large or giant CMN is a distinct presentation. Krengel et al[10] have proposed features to categorize CMN including projected adulthood size, number of "satellite" nevi, site, color heterogeneity, and surface features such as hypertrichosis or rugosity (details in Table 2). This schema can be useful in clinical and research settings.

Genetics

CMN are caused by postzygotic somatic mutations and generally not considered to be inherited. *NRAS* variants are the most commonly identified variants in large and giant CMN (see Table 2 for size definitions), but *BRAF, KRAS, APC,* and *MET* variants are also reported as well as rare protein fusions.[12] Other investigations of genotype–phenotype correlations are ongoing.[13]

Melanoma and Neurocutaneous Melanosis

The lifetime risk of melanoma developing in association with a CMN is low.[14–16] This risk varies based on the size of the nevus. Incidence is reported between 0.7% to 2.2% for all lesions but for those with a giant CMN, estimates are higher (3% to 8%).[14–16]

Neural melanosis, characterized by deposits of melanocytic cells along the leptomeninges or cerebral cortex or within the brain parenchyma, can be associated with CMN. Neurocutaneous melanosis (NCM) refers to the cooccurrence of neural melanosis and cutaneous nevi. Estimates of overall NCM incidence vary because different criteria are used in studies and case series examining imaging results, and lower-risk children are not routinely imaged. In children deemed high risk, incidence of neural melanosis ranges from 17% to 41%.[17–19] The presence of neural melanosis is a risk factor for melanoma; in one study, patients who underwent an MRI screening and were found to have neural melanosis had a 12% incidence of melanoma, compared with 2% of those without NCM.[14] Notably, the population screened with an MRI in this study only included patients with multiple CMN, many of which were seen in association with a large or giant CMN. In a small

TABLE 1 Authors' Management Recommendations for Children With CMN

Section	Subsection	Statement
Skin care and specialty management		
General skin care	Bathing	[a]Bathe with water alone or with a nonsoap cleanser[b] at least 2 to 3 times per week followed by the application of a bland emollient.[c]
	Xerosis and pruritus	[a]Apply bland, thick emollients (creams or ointments with minimal or no fragrances or preservatives) for chronic management and low- to mid-potency topical corticosteroids twice daily as needed for acute eczematous flares.
	Skin fragility/wound healing	[a]Cleanse ulcerations or erosions with soap or a nonsoap cleanser and water and apply petroleum jelly or bland ointment and a bandage. [a]Consider hydrocolloid or foam dressings, which are adherent yet easily removable and gentle on skin. Consider wound cultures or biopsy for nonhealing wounds.
	Hypohidrosis/anhidrosis	Counsel on avoidance of overheating and use of cooling techniques.
	Photoprotection	Follow standard recommendations regarding sunscreen, hats, protective clothing, seeking shade, avoiding sun during peak hours.
Specialty care and comorbidities	Dermatology referral: small or medium CMN	[a]Unless there are clinical concerns (color variation, nodules, symptoms, or location), referral can be delayed or deferred to the primary care provider.
	Dermatology referral: large, giant, or multiple CMN	All of these patients should be referred to dermatology.
	Physical examination: CMN palpation	Palpate CMN with elevated melanoma risk at every visit.
	Physical examination: Lymph nodes	Palpate lymph nodes of patients at higher risk for melanoma. [a]Clinical context, imaging, and biopsy, when needed, differentiate benign and malignant lymph node enlargement.
	Frequency and timing of dermatology office visits	[a]Determined by location and characteristics of the nevus, patient's age, parental concerns, and medical comorbidities.
		[a]Follow larger, multiple, and changing nevi closely during infancy or times of expected nevus change, such as puberty, because of increased melanoma risk and need for family counseling; visits every 3 months may be appropriate.
		[a]After the first year of life, in the absence of particular concerns, visit frequency is gradually decreased. Eventually a minimum of a yearly dermatologist evaluation may be appropriate for large, giant, and multiple CMN or smaller CMN with concerning features.
	Changes and growths within CMN	[a]Between visits, patients and/or caregivers should visually inspect and palpate CMN and notify their physician of any concerning changes (rapid growth, bleeding, pain, development of a lump or nodule, or ulceration). These changes require prompt evaluation, preferably by a dermatologist.
	Neural melanosis screening and monitoring	[a]Solitary small, medium, and large CMN are low risk for NCM and MRI screening is not recommended unless signs or symptoms are elicited during examination.
		[a]Patients with multiple medium CMN, ≥10 "satellite" lesions, and giant CMN are at higher risk for NCM and should undergo screening MRI.
		[a]Early MRI screening, with neither contrast nor anesthesia, of the brain and spine in infants with risk for NCM decreases procedure risks for the infant and can provide useful clinical information.
		Children with neurologic symptoms should undergo MRI to evaluate for neural melanosis and other CNS abnormalities.
Surgery and procedures		
Surgery	All nevi	The decision for procedural interventions or removal of a CMN is complicated by numerous factors, including family preference, the size and location of the nevus, patient age, overall health, and prognosis if NCM or melanoma is present. Detailed risk and benefit discussions are required.
Other procedures	Laser	Pigment-specific ablative lasers, curettage, and dermabrasion can be considered, but they may be associated with adverse outcomes, including obscuring clinical evaluation for melanoma and frequent pigment recurrence.
	Hair removal	When hair removal is desired, shaving, waxing, threading, chemical depilation, electrolysis, or trimming are low risk.
Patient and family support	Family support	Provide prompt support and reliable information about CMN and skin care (see Table 3).

[a] Highlighted as "authors' recommendation" within the text.

[b] Nonsoap cleansers are typically liquid with neutral or mildly acidic pH.

[c] Bland emollients are creams or ointments with minimal or no fragrances or preservatives.

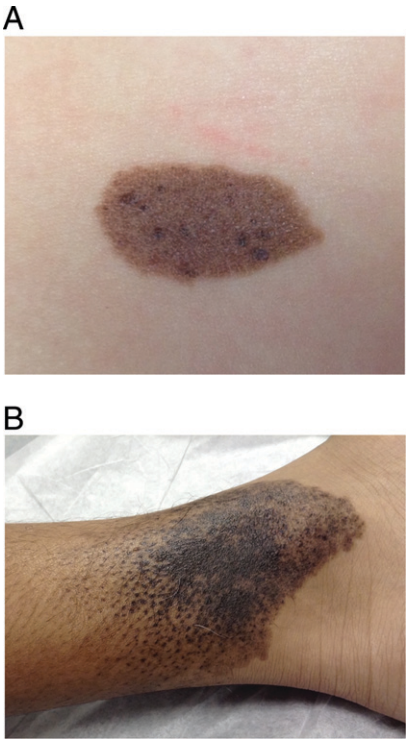

FIGURE 1
Medium-size congenital nevi revealing variety in color and configuration (A) on the abdomen (B) on the lower extremity.

cohort of patients with multiple CMN with giant CMN greater than 60 cm projected adult size or multiple CMN without a largest CMN, the incidence of melanoma was 8% (7/88). The majority of these melanoma cases presented in the CNS.[14] Among fatal cases of pediatric cutaneous melanoma in a separate, large, multisite cohort study, 5 of 6 patients with CMN-associated melanoma had large or giant CMN.[20]

Melanoma and neural melanosis are both more likely in patients with CMN that have a projected adult size of >40 cm in diameter, numerous "satellite" nevi (many CMN), and location on the trunk (which may be a proxy for lesion size) as well as patients with multiple medium CMN.[17,21,22] Patients with multiple CMN are considered at highest risk for NCM. Historically, >20 CMN was considered a strong risk factor.[18,22] A recent proposal

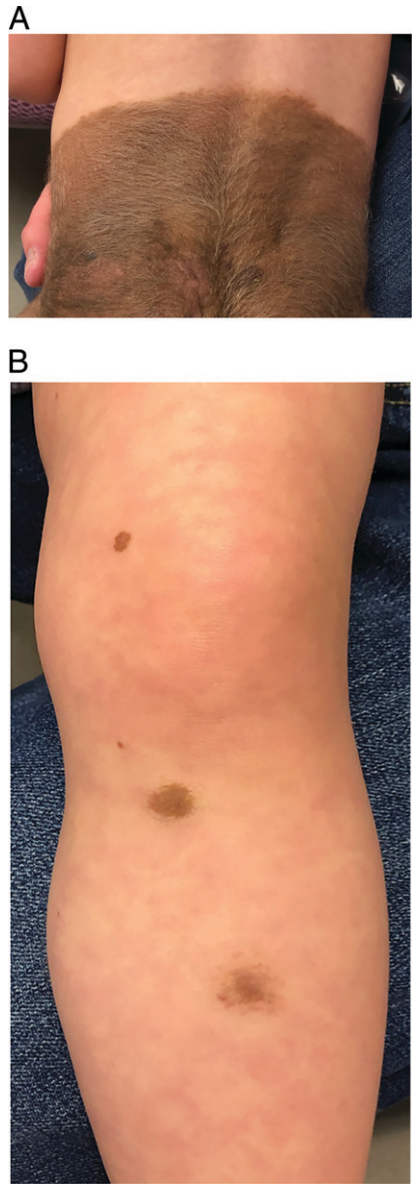

FIGURE 2
(A) Giant congenital nevus of the back that extends to buttocks and legs revealing hypertrichosis and color variation (B) accompanied by several smaller congenital nevi (previously called "satellite nevi").

recommended imaging all patients with >1 CMN; however, this practice has not been accepted by all experts.[19] An MRI of the brain and spine is the preferred screening modality.

Patients with neural melanosis may be symptomatic or asymptomatic. Symptoms may include seizures, headaches, rapidly enlarging head circumference because of hydrocephalus, symptoms of spinal cord compression, and subsequent developmental delays.

Management of CMN

Table 1 summarizes the authors' recommendations for children with CMN.

General Skin Care

There are no evidence-based standards nor guidelines published

TABLE 2 Incidence, Categorization, and Important Subtypes of CMN

Incidence[a]	
CMN	<1 to 3.6%
Melanoma in patients with CMN	0.7% to 1.7%
Categorization of CMN[10]	
Size	Small (<1.5cm)
	Medium (M1: 1.5 to 10 cm; M2: >10 to 20 cm)
	Large (L1: >20 to 30 cm; L2: >30 to 40 cm)
	Giant (G1: >40 to 60 cm; G2: >60 cm)
	Multiple medium (≥3 medium CMN without a principal, largest CMN)
Other characteristics	Localization (body site)
	Number of "satellites"
	Color variation
	Rugosity
	Nodularity
	Hypertrichosis
	Multiplicity
Subtypes of CMN	
"Classic"	**Clinical description:** Shades of brown and black within a macule, papule, patch, or plaque nodules possible. Red-pink ("amelanotic") papules also possible.
	Histopathology: Melanocytes possible within the epidermis and dermis; can involve hair follicles and adnexal structures.
Blue	**Clinical description:** A gray-blue patch or plaque, nodules possible.
	Histopathology: dermal melanocytes (which make the lesions look blue).
Nevus spilus	**Clinical description:** Brown patch with overlying darker brown or black macules, papules, and plaques. May also develop Spitz nevi.
	Histopathology: Similar to a "classic" subtype other than the Spitz nevi.

[a] See text for a detailed discussion.

regarding skin care in the infant with CMN. Recommendations herein were extrapolated from general neonatal skin care and expert consensus.

Bathing

Although studies regarding the barrier function of skin overlying large and giant CMN are lacking, scattered reports and clinical observations of xerosis, pruritus, and skin fragility support the concept of an impaired skin barrier in lesional skin. Bathing recommendations, therefore, include bathing with water alone or with a nonsoap cleanser at least 2 to 3 times per week followed by the application of a bland emollient (authors' recommendation).[23] This can help improve skin hydration and skin barrier function.[24] Nonsoap cleansers are typically liquid with neutral or mildly acidic pH. Bland emollients are creams or ointments with minimal or no fragrances or preservatives. The addition of an emollient after bathing results in less transepidermal water loss without an adverse effect on the pH of the skin.[25]

Xerosis and Pruritus

Pruritus, especially in larger CMN, is common.[26] Pruritus may occur with or without eczematous changes and, fortunately, rarely indicates malignant transformation.[26] Eczematous changes may appear within the nevus or surrounding it (Meyerson phenomenon).[26] Bland, thick emollients (creams or ointments with minimal or no fragrances or preservatives) are recommended for chronic management and low- to mid-potency topical corticosteroids twice daily as needed added for acute eczematous flares (authors' recommendation).

Skin Fragility and Wound Healing

CMN may display increased fragility resulting in ulcerations, erosions, and bleeding with minimal trauma.[27] The neonate, nonetheless, should be handled the same as one would handle an unaffected neonate to encourage parental bonding and minimize parental anxiety. Should ulcerations or erosions occur, wounds should be gently cleansed with soap or a nonsoap cleanser and water, and petroleum jelly or bland ointment and a bandage should be applied (authors' recommendation). Hydrocolloid or foam dressings, which are adherent yet easily removable and gentle on skin, are often useful (authors' recommendation). Topical or oral antibiotics are only indicated if infection occurs. Nonhealing ulcers should be assessed for infection with cultures; additionally, malignancy should be considered when assessing chronic, nonhealing wounds, and biopsies should be performed as appropriate.[27]

Hypohidrosis and Anhidrosis

Anecdotal reports mention lack of sweat glands histologically in some CMN and some patients note hypohidrosis. Although more

research is needed to explore this phenomenon, parents can be counseled on avoidance of overheating and use of cooling techniques, if applicable.

Photoprotection

No studies have specifically examined the effect of UV radiation on CMN. Children with CMN should follow the American Academy of Pediatrics UV radiation protection recommendations.[28] Photoprotective clothing (eg, rash guards and hats) is particularly efficient to safely block the sun.

Specialty Care and Comorbidities

In the neonatal period, patients with large, giant, or multiple CMN of any size should establish care with a pediatric dermatologist or general dermatologist with expertise in CMN. Unless there are clinical concerns (color variation, nodules, symptoms, and/or location), referral for solitary small and medium CMN can be delayed or deferred to the primary care provider (authors' recommendation).

Dermatology Visits

During the initial dermatology visit, a thorough skin examination, education and counseling, discussion of management, and recommendations for potential referrals and follow-up care are reviewed. CMN evaluation involves (1) visual inspection, which can be aided by dermoscopic evaluation, and (2) palpation. Palpation is particularly important because melanoma in CMN can present as deep nodules without overlying color change within the dermis or subcutis, rather than within the epidermis.[14] Serial photographs can be helpful to monitor nevus appearance and changes over time.

Regional lymph node palpation is an important component of the physical examination for patients with CMN at an elevated risk for melanoma; however, both CMN and melanoma can proliferate in draining lymph nodes, and palpation cannot distinguish between benign and malignant proliferation.[29,30] Clinical context, imaging, and biopsy, when needed, inform this determination (authors' recommendation).

There are no evidence-based guidelines specifying visit intervals for CMN of any size. The frequency of dermatologic visits is determined by location and characteristics of the nevus, the age of the patient, parental concerns and needs, and medical comorbidities (authors' recommendation). Benign-appearing, asymptomatic, small or medium CMN (low risk for malignant transformation) often can be followed by a primary care provider at well-child checks, whereas changing or symptomatic small or medium CMN or those with an unusual appearance (eg, variegated color) generally merit referral to dermatology. Larger, multiple, and changing nevi are generally managed closely by a dermatologist during infancy or times of expected nevus change, such as puberty, because of increased risk for melanoma and need for family counseling; visits every 3 months may be appropriate (authors' recommendation). After the first year of life, in the absence of particular concerns, visit frequency is gradually decreased, but a minimum of a yearly evaluation by a dermatologist is generally appropriate for large, giant, and multiple CMN or smaller CMN with concerning features (authors' recommendation). Patients with concurrent medical conditions or who undergo immunosuppression may require special consideration because of a potential increase in melanoma risk.[31,32] Additional factors may play a role, such as the patient's proximity and access to their dermatologist.

Changes and Growths Within CMN

Interestingly, some nevi spontaneously regress. Scalp CMN in particular have a tendency to lighten over time, although continued histologic presence of nevus cells has been reported and thus continued skin examinations should not be foregone.[33,34] The pigmentation of any CMN may evolve to include more mottled or speckled pigment, homogenous or heterogeneous darkening or lightening, and/or a change in the texture.[35] A patient's background skin pigmentation may be the most predictive feature to determine the ultimate nevus color.[35] Surface changes over time include becoming more raised, hypertrichotic, verrucous, cerebriform, mamillated, or papillated.

Between visits, parents, guardians, caregivers and/or patients should monitor nevi visually and with palpation and are advised to notify their physician of any concerning changes, such as rapid growth, bleeding, pain, development of a lump or nodule, or ulceration. These changes within a CMN should be promptly evaluated, preferably by a dermatologist (authors' recommendation). Importantly, melanoma may occur in the skin or the CNS, and some children with large or giant CMN have had metastatic melanoma without a known primary site.[14,16,36]

Proliferative nodules, which are secondary, benign melanocytic growths that can arise over time within larger-sized CMN, have clinical and histologic features overlapping with those of melanoma but lack the genetic instability seen in malignancy. They may present in early infancy or later and are diagnosed clinically or by

pathology.[37,38] Clinically, proliferative nodules can appear as papules, plaques, or nodules, with or without ulceration. Because there can be clinical and histologic uncertainty between proliferative nodules and melanoma, the decision whether to perform a biopsy on a new growth in a CMN is generally on the basis of the clinician's level of concern for malignancy. The size, location, appearance, and feel on palpation (eg, firm versus soft) can help to determine if a biopsy is needed and the type of biopsy performed.

Histopathologic examination of a concerning change or possible proliferative nodule within a CMN should be performed by a dermatopathologist with expertise in pigmented lesions in children because interpretation is often extremely complex. Genetic studies, such as fluorescence in situ hybridization and comparative genomic hybridization, can be complementary diagnostic tools in more ambiguous lesions. Ancillary genomic test results can also drive treatment decisions for melanoma now that targeted treatments exist.

Neural Melanosis Screening and Monitoring

An MRI of the brain and total spine is used to screen for and/or monitor NCM (screening, symptom-directed, and monitoring protocols differ). Solitary small, medium, and large CMN are low risk for NCM, and MRI screening is not recommended unless signs or symptoms are elicited during examination (authors' recommendation). Patients with multiple medium CMN, ≥10 "satellite" lesions, and giant CMN are at high risk for NCM and should undergo MRI screening (authors' recommendation).

Early MR screening, without contrast, can often be performed by using a "feed and swaddle" technique in newborns and until at least 2–3 months of age (although some will attempt this method up to the age of 6 months) to avoid the need for general anesthesia.[39] In this age group, neural melanosis is not obscured by myelination and can be visualized without contrast.[17] This can provide useful information in diagnosing the presence (or absence) of neural melanosis without the need for general anesthesia (authors' recommendation). Telemedicine may be helpful for primary care providers without nearby access to dermatologists in deciding whether early MR screening is warranted. Although institution dependent, some attempt noncontrast screening without anesthesia in older infants and children; however, such imaging may be slightly less definitive because of rapid changes in brain myelination, again emphasizing that early imaging done without contrast is optimal.[17,18,21] In addition, motion artifact must be considered when determining the need for anesthesia. Rapid-sequence ("fast-brain" or "one-bang") MRIs have low resolution.[40] No published data validate rapid-sequence MRI in the investigation for neural melanosis, and this method is not currently recommended for screening nor symptom-directed imaging.

Among patients with ≥2 CMN who underwent screening MRI evaluation, 79% showed normal findings.[19] Intraparenchymal melanosis, the most common abnormality, was seen in 10%.[19] Of note, an MRI can also reveal additional findings in patients with CMN, such as cysts, tumors, malformations, hydrocephalus, and tethered spinal cord.[19,41]

An abnormal MRI in CMN patients is the best predictor of clinical outcomes. Although the large majority of individuals with neural melanosis have good outcomes, its presence, particularly if extensive, signifies an overall increased risk of melanoma. Moreover, early imaging establishes a baseline in case complications or symptoms arise later.[19] Additionally, imaging should be pursued or repeated for new abnormal neurologic findings or developmental deficits. Of note, symptom-directed imaging or follow-up of known lesions is not "screening," and, thus, different guidance for imaging protocols is recommended. An MRI in these cases typically requires contrast and sedation in young children. All imaging should be reviewed by an expert neuroradiologist familiar with NCM.

Patients with proven NCM should be referred to a pediatric neurologist. Further neurodevelopmental assessments, including neuropsychologic testing, may be recommended. Patients with a speech delay or failed newborn hearing screen should undergo audiologic evaluation. Some physicians recommend consultation by ophthalmology, particularly for children with neural melanosis, because melanocytic lesions may be identified on the retina; however, this is controversial. In infants with structural anomalies (eg, hydrocephalus or concern for tethered spinal cord) neurosurgery consultation may be appropriate.

Endocrine Comorbidities

A small number of patients with CMN have been described to have distinct facial features and CNS or endocrinologic anomalies, which are collectively referred to as CMN syndrome.[42] Premature thelarche, undescended testes, insulin insensitivity, and abnormal oral glucose tolerance tests warrant further investigation.[43]

TABLE 3 Patient and Provider Resources

Name	Web Site	Details
Patient associations or research networks		
Caring Matters Now	https://caringmattersnow.co.uk	Group in the United Kingdom supporting patients, funding research, and improving awareness of congenital nevi
Naevus International	https://naevusinternational.com	A collaboration of patient groups, doctors, psychologists, scientists and communications experts focused on improving access to information about nevi for patients and professionals
Nevus Network	https://nevusnetwork.org	A patient support group
Nevus Outreach	https://nevus.org	A nonprofit organization "dedicated to bringing awareness, providing support, and finding cures for people affected by congenital melanocytic nevi and related disorders"
Physician or medical associations (including resources for finding a provider)		
American Academy of Dermatology	https://aad.org	Organization of dermatologists, including pediatric dermatologists, in the United States
European Society of Pediatric Dermatology	https://espd.info	An organization of pediatric, dermatology, and associated providers with goals to "promote clinical care, interdisciplinary research, education and training, and to stimulate international contacts within Europe in the field of Pediatric Dermatology"
Pediatric Dermatology Research Alliance	https://pedraresearch.org	Organization advancing pediatric dermatology research, including a group focused on pediatric nevi
Society for Pediatric Dermatology	https://pedsderm.net	Organization supporting and promoting pediatric dermatology care and research
Sun-protection information and educational programs		
American Academy of Dermatology	https://aad.org/lesson-plans	"Good skin knowledge" lesson plans about sun safety. Also, general sun-protection information at aad.org
American Academy of Pediatrics	https://healthychildren.org/English/safety-prevention/at-play/Pages/Sun-Safety.aspx	Sun safety facts and tips
Centers for Disease Control and Prevention	https://cdc.gov/cancer/skin/basic_info/sun-safety.htm	Sun safety facts
The Community Guide	https://thecommunityguide.org/content/evidence-shows-community-based-skin-cancer-prevention-works	Detailed information on sun-protection educational programs (and whether they work)
Society for Pediatric Dermatology	https://pedsderm.net/for-patients-families/patient-handouts/	Handouts for patients, developed by pediatric dermatologists: "Moles & Melanoma," "Pediatric Skin Cancer," and "Sun Protection"
SunSmart (Australia)	https://sunsmart.com.au/	Skin cancer prevention and early detection program in Australia
SunWise	https://neefusa.org/sunwise	A program for children grades K to 8 to learn about sun safety
Melanoma and skin cancer research organizations		
American Skin Association	https://americanskin.org	Group including patients, families, advocates, physicians and scientists that promotes and funds research as well as provides skin cancer education
Children's Oncology Group	https://childrensoncologygroup.org	Multicenter research group that conducts treatment trials and provides families medical and survivorship information
Melanoma Research Alliance	https://curemelanoma.org	Organization that funds research and promotes knowledge about melanoma
Melanoma Research Foundation	https://melanoma.org	Research group aiming to "eradicate melanoma by accelerating medical research while educating and advocating for the melanoma community"
Skin Cancer Foundation	https://skincancer.org	Organization "devoted to the prevention, early detection, and treatment of skin cancer" through education and research

Psychological Considerations

Referral to psychology can be considered for all affected children to address quality of life and emotional and behavioral health.[44] The impacts of CMN on a child's social relationships and emotional function are not always predictable. In a parental proxy-report study on health-related quality of life and psychological adjustment, parents of children with neurologic sequelae, skin symptoms, and high levels of perceived stigmatization noted increased impairments in these domains.[44] In a study including adolescent self-report, 46% of adolescents reported "no" to a "small" impact on their skin-related quality of life, whereas 54% reported a "moderate" to "extremely large" impact.[45] More research into the relationship of the response with the age of the patient (experience over time) and size and location of the CMN are needed. It is important to support families and children emotionally and with community resources, as appropriate. Many families find support from patient advocacy groups helpful (Table 3).

Management of CMN: Surgery and Procedures

The decision for procedural interventions or removal of a CMN is complicated by numerous factors, including family preference, the size and location of the nevus, patient age, overall health, and prognosis if NCM or melanoma is present. Importantly, prophylactic removal does not eliminate the risk of melanoma because residual nevus cells may remain after removal and, especially for larger nevi, it can be almost impossible to surgically remove a CMN entirely.[36] Moreover, CNS melanoma risk is not affected by nevus removal. Prophylactic removal can be associated with several risks, such as multiple procedures, repeated general anesthesia, anxiety associated with treatments, postoperative pain, and scar appearance. Potential complications must be weighed against potential improvement in the patient's appearance, improvement in function if associated impairment exists, and psychosocial benefit (or lack thereof). Shared decision-making can support patients and families in making choices about treatments, focusing the discussion on both patient and family preferences and medical evidence.

Surgery: Small and Medium CMN

In general, uncomplicated small and medium lesions can be observed.[46] Indications for surgery may include functional considerations, symptoms, difficulty managing a lesion clinically, or stigma. Psychosocial impact of the CMN can be variable and should be considered in surgical decision-making.[45]

Surgery: Large and Giant CMN

Surgical approaches to large and giant CMN include serial excision, tissue expansion, local flaps, and grafting with skin or artificial skin substrates. Benefits of serial excision include a single linear scar when size allows and no donor sites nor large flaps. Tissue expansion can allow for a reduced number of surgical procedures and reduced time to complete the excision of a nevus; tissue expansion, however, has an overall high complication rate (18.2%), with 60% of these cases undergoing subsequent successful reconstruction.[47]

Notably, routine excision of large and giant CMN has fallen out of favor, given that surgical interventions present their own risks and have not demonstrated a reduced lifetime melanoma risk.[46,48] Additionally, neither NCM nor risk of CNS melanoma is impacted by surgical approaches.[14] In addition, disfigurement caused by resection and procedures has psychological and functional consequences, and must be considered along with the psychosocial impact of the CMN on the child and family. Conservative management is now frequently encouraged for large and giant CMN, except when worrisome examination findings present.[46]

Other Procedural Treatment Approaches for CMN

Pigment-specific ablative lasers, curettage, and dermabrasion have also been used to treat CMN. These nonexcisional approaches are employed primarily for cosmesis, when surgical excision and reconstruction are not options. Some authors suggest that these interventions may make melanoma more difficult to detect because of resulting fibrosis and scarring.[49] Risks of destructive treatment include scarring, dyspigmentation, need for repeated treatments, poor wound healing, infection, and eventual repigmentation, which is common. Some report a worsening appearance after intervention, including disfiguring scars, keloids, and recurrent nevi.[46,50] Many CMN lighten over time without intervention, which should be considered before any cosmetic intervention.[33,35]

Hair Removal

Hair removal is understudied in children with CMN. Shaving, waxing, threading, chemical depilation, electrolysis, or trimming are low risk. Hypertrichosis often becomes more prominent over time. For infants, simple trimming of hair is generally adequate should caregivers wish to do so. Laser hair reduction[51] and electrolysis are more permanent and require serial treatments. Histologic and dermoscopic changes have been

noted in nevi after laser hair removal,[52,53] but the risk of melanoma from these procedures is thought to be low.

Management of CMN: Medical Care

Medical interventions for CMN are emerging and currently under investigation. They are based on the evolving understanding of the genomics of CMN and target pathways involved in their growth and proliferation. Current approaches are experimental and have infrequently been used for newborns and infants, except in rare cases of compassionate use. Research in this area may validate a role for medical treatment of infants with thick, pruritic, or painful CMN that interfere with development.[54–56]

Patient and Family Support

Families may have little familiarity with CMN before their child is born, necessitating prompt support and provision of reliable information about CMN and skin care. Table 3 includes information about patient support and advocacy organizations, finding a provider, sun protection, and melanoma.

CONCLUSIONS

CMN vary in size, color, location, and associated comorbidities. Classification is based on size, number of lesions, and associated findings at birth, which aids the predicted prognosis and complications. Management of large, giant, and multiple CMN is complex and typically involves multidisciplinary care. Optimal routine skin care recommendations have not been developed specifically for the care of CMN; best practices are extrapolated from general infant care. Screening guidelines are needed to standardize care and optimize outcomes.

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ABBREVIATIONS

CMN: congenital melanocytic nevi
CNS: central nervous system
NCM: neurocutaneous melanosis

acquisition of data, revised the article critically for intellectual connect, provided final approval of the version to be published, and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved; Dr Kruse had substantial contribution to interpretation of data, revised the article critically for important intellectual content, provided final approval of the version to be published, and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved; Drs Jen and Horii had substantial contribution to interpretation of data, drafted the article, provided final approval of the version to be published, and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved; Dr Hawryluk had substantial contribution to conception and design, drafted the article, revised it critically for important intellectual content, provided final approval of the version to be published, and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved; and all authors approved the final manuscript as submitted.

The guidelines/recommendations in this article are not American Academy of Pediatrics policy, and publication herein does not imply endorsement.

In this review with management recommendations, we discuss care, risks, and screening for patients with CMN; we includes resources for providers and families.

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REFERENCES

1. Aguilera P, Puig S, Guilabert A, et al. Prevalence study of nevi in children from Barcelona. Dermoscopy, constitutional and environmental factors. *Dermatology.* 2009;218(3):203–214
2. Alikhan A, Ibrahimi OA, Eisen DB. Congenital melanocytic nevi: where are we now? Part I. Clinical presentation, epidemiology, pathogenesis, histology, malignant transformation, and neurocutaneous melanosis. *J Am Acad Dermatol.* 2012;67(4):495.e1–17
3. Alper JC, Holmes LB. The incidence and significance of birthmarks in a cohort of 4,641 newborns. *Pediatr Dermatol.* 1983;1(1):58–68
4. Castilla EE, da Graça Dutra M, Orioli-Parreiras IM. Epidemiology of congenital

pigmented naevi: I. Incidence rates and relative frequencies. *Br J Dermatol.* 1981;104(3):307–315

5. Fung WK, Lo KK. Prevalence of skin disease among school children and adolescents in a Student Health Service Center in Hong Kong. *Pediatr Dermatol.* 2000;17(6):440–446
6. Ingordo V, Gentile C, Iannazzone SS, Cusano F, Naldi L. Congenital melanocytic nevus: an epidemiologic study in Italy. *Dermatology.* 2007;214(3):227–230
7. Kallas M, Rosdahl I, Fredriksson M, Synnerstad I. Frequency and distribution pattern of melanocytic naevi in Estonian children and the influence of atopic dermatitis. *J Eur Acad Dermatol Venereol.* 2006;20(2):143–148
8. Rivers JK, MacLennan R, Kelly JW, et al. The eastern Australian childhood nevus study: prevalence of atypical nevi, congenital nevus-like nevi, and other pigmented lesions. *J Am Acad Dermatol.* 1995;32(6):957–963
9. Hösli I, Holzgreve W, Danzer E, Tercanli S. Two case reports of rare fetal tumors: an indication for surface rendering? *Ultrasound Obstet Gynecol.* 2001;17(6): 522–526
10. Krengel S, Scope A, Dusza SW, Vonthein R, Marghoob AA. New recommendations for the categorization of cutaneous features of congenital melanocytic nevi. *J Am Acad Dermatol.* 2013;68(3): 441–451
11. Kinsler V. Satellite lesions in congenital melanocytic nevi–time for a change of name. *Pediatr Dermatol.* 2011;28(2): 212–213
12. Martins da Silva V, Martinez-Barrios E, Tell-Martí G, et al. Genetic abnormalities in large to giant congenital nevi: beyond NRAS mutations. *J Invest Dermatol.* 2019;139(4):900–908
13. Polubothu S, McGuire N, Al-Olabi L, et al. Does the gene matter? Genotype-phenotype and genotype-outcome associations in congenital melanocytic naevi. *Br J Dermatol.* 2020;182(2):434–443
14. Kinsler VA, O'Hare P, Bulstrode N, et al. Melanoma in congenital melanocytic naevi. *Br J Dermatol.* 2017;176(5): 1131–1143
15. Kinsler VA, Chong WK, Aylett SE, Atherton DJ. Complications of congenital melanocytic naevi in children: analysis of 16 years' experience and clinical practice. *Br J Dermatol.* 2008; 159(4):907–914
16. Krengel S, Hauschild A, Schäfer T. Melanoma risk in congenital melanocytic naevi: a systematic review. *Br J Dermatol.* 2006;155(1):1–8
17. Jakchairoongruang K, Khakoo Y, Beckwith M, Barkovich AJ. New insights into neurocutaneous melanosis. *Pediatr Radiol.* 2018;48(12):1786–1796
18. Lovett A, Maari C, Decarie JC, et al. Large congenital melanocytic nevi and neurocutaneous melanocytosis: one pediatric center's experience. *J Am Acad Dermatol.* 2009;61(5):766–774
19. Waelchli R, Aylett SE, Atherton D, Thompson DJ, Chong WK, Kinsler VA. Classification of neurological abnormalities in children with congenital melanocytic naevus syndrome identifies magnetic resonance imaging as the best predictor of clinical outcome. *Br J Dermatol.* 2015; 173(3):739–750
20. Hawryluk EB, Moustafa D, Bartenstein D, et al. A retrospective multicenter study of fatal pediatric melanoma. *J Am Acad Dermatol.* 2020;83(5): 1274–1281
21. Price HN, Schaffer JV. Congenital melanocytic nevi-when to worry and how to treat: Facts and controversies. *Clin Dermatol.* 2010;28(3):293–302
22. Marghoob AA, Dusza S, Oliveria S, Halpern AC. Number of satellite nevi as a correlate for neurocutaneous melanocytosis in patients with large congenital melanocytic nevi. *Arch Dermatol.* 2004;140(2):171–175
23. Blume-Peytavi U, Lavender T, Jenerowicz D, et al. Recommendations from a European roundtable meeting on best practice healthy infant skin care. *Pediatr Dermatol.* 2016;33(3):311–321
24. Dyer JA. Newborn skin care. *Semin Perinatol.* 2013;37(1):3–7
25. Garcia Bartels N, Scheufele R, Prosch F, et al. Effect of standardized skin care regimens on neonatal skin barrier function in different body areas. *Pediatr Dermatol.* 2010;27(1):1–8
26. Rolland S, Kokta V, Marcoux D. Meyerson phenomenon in children: observation in five cases of congenital melanocytic nevi. *Pediatr Dermatol.* 2009;26(3):292–297
27. Giam YC, Williams ML, Leboit PE, Orlow SJ, Eichenfield LF, Frieden IJ. Neonatal erosions and ulcerations in giant congenital melanocytic nevi. *Pediatr Dermatol.* 1999;16(5):354–358
28. Balk SJ; Section on Dermatology; Council on Environmental Health. Ultraviolet radiation: a hazard to children and adolescents. *Pediatrics.* 2011;127(3): 588–597
29. Bowen AR, Duffy KL, Clayton FC, Andtbacka RH, Florell SR. Benign melanocytic lymph node deposits in the setting of giant congenital melanocytic nevi: the large congenital nodal nevus. *J Cutan Pathol.* 2015;42(11):832–839
30. Fontaine D, Parkhill W, Greer W, Walsh N. Nevus cells in lymph nodes: an association with congenital cutaneous nevi. *Am J Dermatopathol.* 2002;24(1):1–5
31. Kubica AW, Brewer JD. Melanoma in immunosuppressed patients. *Mayo Clin Proc.* 2012;87(10):991–1003
32. Schaff LR, Marghoob A, Rosenblum MK, Meyer R, Khakoo Y. Malignant transformation of neurocutaneous melanosis (NCM) following immunosuppression. *Pediatr Dermatol.* 2019;36(4):497–500
33. Kinsler V, Bulstrode N. The role of surgery in the management of congenital melanocytic naevi in children: a perspective from Great Ormond Street Hospital. *J Plast Reconstr Aesthet Surg.* 2009; 62(5):595–601
34. Strauss RM, Newton Bishop JA. Spontaneous involution of congenital melanocytic nevi of the scalp. *J Am Acad Dermatol.* 2008;58(3):508–511
35. Polubothu S, Kinsler VA. Final congenital melanocytic naevi colour is determined by normal skin colour and unaltered by superficial removal techniques: a longitudinal study. *Br J Dermatol.* 2020;182(3):721–728
36. Watt AJ, Kotsis SV, Chung KC. Risk of melanoma arising in large congenital melanocytic nevi: a systematic review. *Plast Reconstr Surg.* 2004;113(7):1968–1974
37. Vergier B, Laharanne E, Prochazkova-Carlotti M, et al. Proliferative nodules vs melanoma arising in giant congenital melanocytic nevi during childhood. *JAMA Dermatol.* 2016;152(10):1147–1151

38. Yélamos O, Arva NC, Obregon R, et al. A comparative study of proliferative nodules and lethal melanomas in congenital nevi from children. *Am J Surg Pathol.* 2015;39(3):405–415

39. Bekiesińska-Figatowska M, Sawicka E, Żak K, Szczygielski O. Age related changes in brain MR appearance in the course of neurocutaneous melanosis. *Eur J Radiol.* 2016;85(8):1427–1431

40. Rozovsky K, Ventureyra EC, Miller E. Fast-brain MRI in children is quick, without sedation, and radiation-free, but beware of limitations. *J Clin Neurosci.* 2013;20(3):400–405

41. Foster RD, Williams ML, Barkovich AJ, Hoffman WY, Mathes SJ, Frieden IJ. Giant congenital melanocytic nevi: the significance of neurocutaneous melanosis in neurologically asymptomatic children. *Plast Reconstr Surg.* 2001;107(4):933–941

42. Kinsler V, Shaw AC, Merks JH, Hennekam RC. The face in congenital melanocytic nevus syndrome. *Am J Med Genet A.* 2012;158A(5):1014–1019

43. Waelchli R, Williams J, Cole T, et al. Growth and hormone profiling in children with congenital melanocytic naevi. *Br J Dermatol.* 2015;173(6):1471–1478

44. Masnari O, Neuhaus K, Aegerter T, Reynolds S, Schiestl CM, Landolt MA. Predictors of health-related quality of life and psychological adjustment in children and adolescents with congenital melanocytic nevi: analysis of parent reports. *J Pediatr Psychol.* 2019;44(6):714–725

45. Neuhaus K, Landolt MA, Theiler M, Schiestl C, Masnari O. Skin-related quality of life in children and adolescents with congenital melanocytic naevi - an analysis of self- and parent reports. *J Eur Acad Dermatol Venereol.* 2020;34(5):1105–1111

46. Ott H, Krengel S, Beck O, et al. Multidisciplinary long-term care and modern surgical treatment of congenital melanocytic nevi - recommendations by the CMN surgery network. *J Dtsch Dermatol Ges.* 2019;17(10):1005–1016

47. Adler N, Elia J, Billig A, Margulis A. Complications of nonbreast tissue expansion: 9 Years experience with 44 adult patients and 119 pediatric patients. *J Pediatr Surg.* 2015;50(9):1513–1516

48. Arad E, Zuker RM. The shifting paradigm in the management of giant congenital melanocytic nevi: review and clinical applications. *Plast Reconstr Surg.* 2014;133(2):367–376

49. Ibrahimi OA, Alikhan A, Eisen DB. Congenital melanocytic nevi: Where are we now? Part II. Treatment options and approach to treatment. *J Am Acad Dermatol.* 2012;67(4):515.e1–13

50. Criscito MC, Correa LM, Koshenkov VP, Firoz BF. Recurrent nevi in a skin graft following excision of giant congenital melanocytic nevus. *Dermatol Surg.* 2016;42(9):1113–1116

51. Rajpar SF, Hague JS, Abdullah A, Lanigan SW. Hair removal with the long-pulse alexandrite and long-pulse Nd:YAG lasers is safe and well tolerated in children. *Clin Exp Dermatol.* 2009;34(6):684–687

52. Garrido-Ríos AA, Muñoz-Repeto I, Huerta-Brogeras M, et al. Dermoscopic changes in melanocytic nevi after depilation techniques. *J Cosmet Laser Ther.* 2013;15(2):98–101

53. Soden CE, Smith K, Skelton H. Histologic features seen in changing nevi after therapy with an 810 nm pulsed diode laser for hair removal in patients with dysplastic nevi. *Int J Dermatol.* 2001;40(8):500–504

54. Mir A, Agim NG, Kane AA, Josephs SC, Park JY, Ludwig K. Giant congenital melanocytic nevus treated with trametinib. *Pediatrics.* 2019;143(3):e20182469

55. Rouillé T, Aractingi S, Kadlub N, et al. Local inhibition of MEK/Akt prevents cellular growth in human congenital melanocytic nevi. *J Invest Dermatol.* 2019;139(9):2004–2015.e13

56. Yu Q, Wu M, Sheng L, Li Q, Xie F. Therapeutic effects of targeting RAS-ERK signaling in giant congenital melanocytic nevi. *Am J Transl Res.* 2018;10(4):1184–1194

Algorithm for Hidradenitis Suppurativa (HS)

Children's National

HS has a high rate of co-morbidities that should be screened for at the time of diagnosis[1]

HPI/ROS findings – Ask screening question: *Have you had 2 or more outbreaks of boils in the last 6-12 months in the axillae, groin, inframammary folds or buttocks?*

Treatment goal: Reduce scarring and development of new lesions while preventing progression of disease to improve symptoms and quality of life

Physical exam findings present – Examine intertriginous areas including the axilla, groin, neck, breasts and buttocks

Likely diagnosis of HS

Lifestyle modifications[2]: All treatment modalities should also include include adjunct therapy for pain management, weight loss, appropriate skin care

- No sinus tracts or scarring
- Flares are intermittent and can go weeks to months without any active lesions

Mild

Treatment for flares:
- Topical clindamycin 2x/day[3] **AND**
- Benzoyl peroxide 10% wash[3] **AND**
- Oral doxycycline 100mg 2x/day x 2 weeks

Improvement

Flares are common and may occur at same or different site. Re-treat with previously effective therapy and continue to monitor for progression of disease

- Stop doxycycline
- Start zinc gluconate 50mg 2x/day x 12 weeks AND continue maintenance therapy of choice
- Monitor for disease progression

- Multiple flares of lesions per month, or always with at least one lesion active
- May have no scarring or have signs of spaced out scars or a single sinus tract/ tunnel

Moderate

Treatment:
- Topical clindamycin 2x/day[3] **AND**
- Benzoyl peroxide 10% wash[3] **AND**
- Oral doxycycline 100mg 2x/day x 2 weeks **AND**
- Maintenance therapy of choice

Consider dermatology referral

No improvement

ADD

- Oral doxycycine 100mg 2x/day x 12 weeks[4] **AND**
- Maintenance therapy
  - Metformin 500mg 2x/day for men and women **OR**
  - Spironolactone 50mg 2x.day for women **OR**
  - OCPs[5] for women
- If comorbid menstrual flares and/or irregularities present, screen for PCOS[6] and favor hormonal therapies for women
- Consider dermatology referral for all

Improvement

No improvement

ADD

- Refer to dermatology +/- HS multi-disciplinary clinic
- Consider change or addition of maintenance therapy
- Consider starting clindamycin 300mg 2x/day **AND** rifampin 300mg 2x/day, both x 12 weeks4

- Multiple flared lesions at any given time
- Interconnected sinus tracts and abscesses throughout affected area

Severe

Treatment:
- Topical clindamycin 2x/day[3] **AND**
- Benzoyl peroxide 10% wash[3] **AND**
- Oral doxycycline 100mg 2x/day x 2 weeks **AND**
- Maitenance therapy of choice

Refer directly to HS multi-disciplinary clinic

Additional treatment options for refractory or severe disease:
- Differing combination of topical/oral agents
- Biologic agents
- Laser therapy
- Surgical procedures

Routine incision and drainage **NOT** recommended for acute symptomatic lesions, unless obviously fluctuant with a definite large fluid collection[7]

#	Subject	Description
1	Associated co-morbidities	HS has a very high co-morbidity burden, with newer guidelines recommending that the following systems should be screened for at the time of diagnosis: • **Metabolic (obesity, dyslipidemia, hypertension, metabolic syndrome)** – Exam: BMI, blood pressure – Labs: fasting lipid panel, hemoglobin A1c, fasting blood glucose • **Endocrinologic (diabetes, PCOS, precocious puberty/premature adrenarche)** – History: menstrual irregularities – Exam: PCOS screening (signs of hyperandrogenism), signs of precocious puberty – Labs: PCOS screening labs if appropriate • **Psychiatric (depression, anxiety, substance use disorder)** – History: PHQ-2 and/or PHQ-9, GAD-7, AUDIT-C questionnaire/opioid risk tool • **Inflammatory conditions (inflammatory bowel disease, spondyloarthritis)** – History: arthritis and inflammatory bowel disease screening questions – Labs: IBD screening labs if appropriate • **Dermatologic (acne, pilonidal disease, dissecting cellulitis of scalp, pyoderma gangrenosum)** – Exam: Full skin exam Management and referrals to appropriate specialists if needed should be pursued if signs of these conditions are found.
2	Lifestyle modifications	Lifestyle modifications include weight management counseling with exercise and nutrition recommendations. **Weight loss** of 5-10% is the best supported modification. Other lifestyle modifications include counseling to: • Consider avoiding antiperspirant and using deodorant only, or switching to spray • Wash affected areas gently with fingers; do not scrub with washcloth or brush • Avoid overly tight clothing • Smoking/vaping cessation • NSAIDs or corticosteroids can be considered in short courses to reduce pain and inflammation • Avoid popping/draining new forming lesions[7]
3	Topical therapy	• **Topical clindamycin 1% solution** may help to reduce inflammatory lesions and pustules – Clean involved area with soap and water, dry, and apply the 1% clindamycin solution with fingertip 2x/day in skin areas subject to recurrent flares for **3 months** • **Benzoyl peroxide 10% antiseptic wash** must be used in conjunction with topical clindamycin to prevent Staph aureus resistance • Other antiseptic washes that can be alternated daily with benzoyl peroxide include: – Chlorhexidine gluconate 4% – Shampoo containing zinc pyrithione 1%
4	Long-term oral antibiotics	**Long-term oral antibiotics** have been shown to improve HS, although the mechanism is not definitively known. Patients who achieve satisfactory disease control may stop and then use zinc gluconate 50mg 2x/day for longer disease-free remission. a. **Oral doxycycline** 100mg 2x/day for **3-6 months (at least 3 months** prior to assessing response). **Strongly encourage patients to take this with a full meal to improve tolerability.** Not recommended for pediatric patients under the age of 9. Refer younger patients with HS to dermatology sooner for management. b. **Oral clindamycin** 300mg 2x/day plus rifampin 300mg 2x/day for **12 weeks** as second-line therapy if patients fail to respond to doxycycline.

#	Subject	Description
5	Antiandrogenic agents	Some female patients have noted menstrual variation in their HS, indicating a role of hormones in HS. **Antiandrogenic therapy** in women seem to have a stronger response compared to antibiotic use. **Antiandrogenic agents should NOT be given to pregnant women because of the risk for adverse effects on the fetus.** Always conduct a pregnancy test before considering use of antiandrogenic agents. • **Oral contraceptive pills (OCP)** improve clinical symptoms – **Ethinyl estradiol** 50 mcg (cycled days 5 to 25) and **cyproterone acetate** 50mg (cycle days 5 to 14) for **6 months** – **Ethinyl estradiol** 50mcg and norgestrel 500mcg (cycle days 5 to 25) daily for **6 months** Administration of combined OCPs containing ethinyl estradiol are key. Progesterone-only hormonal therapies can trigger or worsen HS, and it is recommended to switch to a different therapy if an HS patient is already on a progesterone-only agent. • **Spironolactone** for HS associated with improvement in pain, lesions, and disease severity, especially for patients with PCOS. – Start with 25mg/day and go up to 100mg/day for **at least 3 months**
6	Metformin	Insulin-resistance may contribute to HS, and **metformin** has shown benefit in HS along with modest weight loss for patients with obesity, particularly those with metabolic syndrome, PCOS or diabetes. 500mg initial dose 2x/day with food and titrate 500mg 1x/day x 1-2 weeks to minimize side effects, then increase to 500mg 2x/day **for at least 3 months**
7	Acute symptomatic lesions	• When patient has new forming lesion, counsel patient to intermittently apply **warm compress** over area for 10 minutes at a time throughout the day. This can improve symptoms of inflammation. • If **lesion starts to drain** on its own, keep wound clean and wash gently with antiseptic wash. Cover the skin with **petroleum jelly** to avoid dressing from sticking to the wound, and clean/change dressing daily until wound heals. – Adhesive tape should be avoided if possible, and instead an absorbent material should be held in place in a way that minimizes skin trauma, such as an elastic fishnet dressing. – If lesion continues to be painful and inflamed, instruct patient to call doctor to discuss additional methods for treating acute symptomatic lesions. • Additional interventions by dermatology: **intralesional corticosteroid injections** (triamcinolone 10mg/mL), **punch debridement** (partial unroofing) and **topical resorcinol** (topical 15% resorcinol).

Sources:
Children's National Hospital Dermatology Department
American Academy of Dermatology, North American clinical management guidelines for HS, https://doi.org/10.1016/j.jaad.2019.02.068
International Journal of Women's Dermatology, A concise clinician's guide to therapy for hidradenitis suppurativa, Nesbitt et al., doi: 10.1016/j.ijwd.2019.11.004
UpToDate, Hidradenitis Suppurativa: Management, Ingram, https://www.uptodate.com/contents/hidradenitis-suppurativa-management
Liy-Wong C, Kim M, Kirkorian AY, et al. Hidradenitis Suppurativa in the Pediatric Population: An International, Multicenter, Retrospective, Cross-sectional Study of 481 Pediatric Patients. JAMA Dermatol. Published online February 24, 2021. doi:10.1001/jamadermatol.2020.5435