Pediatric Neurology Referral Guidelines

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Pediatric Neurology Referral Guidelines

The CHOC Children's Specialists Neurology Division at CHOC Children's and UCI Medical Center is available for consultation 24 hours a day, seven days a week and provides diagnostic services, medical treatment and follow-up care to infants, children, and adolescents who have suspected or confirmed neurological disorders. Our pediatric neurologists manage and treat a variety of patients including those with epilepsy disorders, neuromuscular disorders and spasticity.

A pediatric neurologist has completed a residency in pediatrics and had additional training in adult and child neurology. All our pediatric neurologists have certification or are eligible for the American Board of Psychiatry and Neurology (with special qualifications in child neurology). Our group also has pediatric neurologists with board certification in clinical neurophysiology (epilepsy and neuromuscular diseases), palliative care and neurodevelopmental disabilities.

Pediatric neurologists combine the expertise in diagnosing and treating disorders of the nervous system (brain, spinal cord, muscles, nerves) with an understanding of medical disorders in childhood and the special needs of the child and their family. In many cases, pediatric neurologists work as a team with pediatricians or other primary care doctors. In addition, pediatric neurologists may work with other pediatric specialists to care for children with more complex or serious medical issues, such as epilepsy, birth defects, or developmental delay.

The following conditions may be best treated by a pediatric neurologist:

- Epilepsy and seizures — including intensive long term video EEG monitoring for epilepsy and related disorders
- Motor system disorders — including tics, Tourette's Syndrome; neuromuscular diseases, including congenital myopathies, muscular dystrophy, hypotonia and other genetic muscular disorders; and cerebral palsy
- Headaches with neurological findings or patients who have failed first line medications
- Developmental and behavioral disorders — including learning disabilities, ADHD, developmental disorders and autism
- Neurogenetic and neurometabolic disorders — including neurodegenerative diseases such as mitochondrial disorders, neurofibromatosis, tuberous sclerosis, Rett's Syndrome and Down Syndrome
- Neuroimmunological disorders — including dermatomyositis and post-infectious encephalopathy
- Neurological aspects of head injuries, brain tumors, brain malformations, and hydrocephalus
- Complications of central nervous system infection

Clinical services are complemented by the multidisciplinary care provided by other CHOC Children's Specialists, nurses, pharmacists, EEG lab technicians, therapists, clinical dietitians, child psychologists and social workers at CHOC Children's and UCI Medical Center.

CHOC Children's Specialists

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A. Recurrent Unprovoked Seizures (Epilepsy) [ICD-9 Code: 345.00] [ICD-10 Code: G40.*]

Suggested Workup & Initial Management

- Sleep deprived EEG (prefer EEG obtained at CHOC Children's) is recommended to determine seizure type.
- Neuroimaging is generally indicated after a first unprovoked seizure. **MRI of the brain is the preferred neuroimaging study** (prefer MRI obtained at CHOC Children's).
  - Emergent if suspicion of a serious structural lesion.
  - Non-urgent if there is no clear cause for seizure
  - CT scan of brain unnecessarily exposes patient to radiation and does not adequately evaluate intraparenchymal structures.

When to Refer

- ► Consultation to general neurology. If patient has already failed 2 or more anti-epileptic medications, then consultation should be directed to one of our epileptologists.

B. Febrile Seizures [ICD-9 Codes: 780.31 (simple) 780.32 (complex)][ICD-10 Codes: R56.00 (simple) R56.01 (complex)]

Suggested Workup & Initial Management

- Infants and toddlers 6 months and 5 years of age with simple (benign) febrile seizures do not require brain imaging, EEG, or neurological consultation

When to Refer

- ► Children with multiple recurrences of simple febrile seizures may benefit from consultation on a case-by-case basis.
- ► Consultation is recommended for children with atypical (complex) febrile seizures defined as lasting >15 minutes, a febrile seizure with partial onset, focal features during or after the seizure, recurrent febrile seizures within a 24 hour period.

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C. First Unprovoked Seizure [ICD-9 Codes: 780.39] [ICD-10 Codes: R56.9]

Suggested Workup & Initial Management

- Labs tests should be individualized to historical and clinical findings such as vomiting, diarrhea, dehydration, or failure to return to mental alertness. Toxicology screens should be done if there is a suspicion of ingestion.
- Lumbar puncture is of limited value and should only be done if meningitis or encephalitis is suspected.
- Sleep deprived EEG (prefer EEG obtained at CHOC Children's) is recommended to determine seizure type and risk for recurrence. (Caution should be advised in interpretation of EEG, as some abnormalities such as postictal slowing or central sharp waves are transient or may not be clinically significant. Clinical correlation is required. Normal EEG does not exclude the diagnosis of epilepsy.)
- Neuroimaging is generally indicated after a first unprovoked seizure. MRI of the Brain is the preferred neuroimaging study (prefer MRI obtained at CHOC Children's).
  - Emergent if suspicion of a serious structural lesion.
  - Non-urgent if there is no clear cause for seizure
  - CT scan of brain unnecessarily exposes patient to radiation and does not adequately evaluate intraparenchymal structures.

When to Refer

- ► Any seizure with partial onset or focal features during or after the seizure.
- ► After 2nd event (withholding treatment until after the second seizure does not alter the long-term prognosis of epilepsy and long-term mortality is low after a single unprovoked seizure).
- ► If initial EEG is abnormal (Caution should be advised in interpretation of EEG, as some abnormalities such as postictal slowing or central sharp waves are transient or may not be clinically significant. Clinical correlation is required.)
- ► Abnormal neuroimaging (Clinical correlation is required.)

Resources used in development of these Referral Guidelines:

Recurrent Unprovoked Seizures, Febrile Seizures, First Unprovoked Seizure:

- Clinical Practice Guideline—Febrile Seizures: Guideline for the Neurodiagnostic Evaluation of the Child with a Simple Febrile Seizure – Pediatrics, Volume 127, Number 2, February 2011
- Steering Committee on Quality Improvement and Management, Subcommittee on Febrile Seizures: American Academy of Pediatrics. Febrile seizures: clinical practice guideline for the long-term management of the child with simple febrile seizures. Pediatrics. 2008;121:1281-1286.
- G. Fenichel – Clinical Pediatric Neurology, Sixth Edition, 2009
- Shinnar S, Glauser TA. Febrile Seizures. J Child Neurol 2002;17:S44-S52
- Hirtz, D, Ashwal, S, Berg, A, et al. Practice parameter: evaluating a first nonfebrile seizure in children. Neurology 2000; 55:616-623.
- Hirtz,D, Ashwal,S,Berg,A, et al . Practice parameter: treatment of the child with a first unprovoked seizure: Report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. Neurology 2003; 60; 166.
- Shinnar, S, O'Dell, C, Berg, AT. Mortality following a first unprovoked seizure in children: a prospective study. Neurology 2005; 64:880.

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D. Developmental Delay [ICD-9 Codes: 783.40] [ICD-10 Codes: R62.50]

Suggested Workup & Initial Management

- **First Line:**
  - Karyotype (Chromosomal microarray may replace karyotype as test of choice)
  - DNA for Fragile X syndrome
  - Lead level
  - Thyroid function tests
  - CBC (screening for iron deficiency)
  - Comprehensive metabolic panel
  - Uric acid (screen for purine disorders, which can cause isolated developmental delay; more stable than lactate and ammonia)
  - Biotinidase (if not born in the USA)
  - Evaluations of vision and hearing
- **Second Line:**
  - Neuroimaging (preferably MRI) if abnormal head size, seizures, focal neurological findings.
  - EEG(sleep deprived) if speech regression, seizures, history suggestive of neurodegenerative disorder.
  - Consider video EEG telemetry if frequent paroxysmal events, or speech regression.
  - Genetics referral if dysmorphic features, family history.
  - Metabolic workup if family history of metabolic disorders, consanguinity, regression, organomegaly, coarse facial features or the combination of epilepsy and developmental delays. Blood: Lactate, amino acids, ammonia, very long chain fatty acids, carnitine, isoforms of transferrin, acylcarnitine profile. Urine: organic acids, oligosaccharides, glycosaminoglycans.

When to Refer

- ► 0-3 years of age: Refer to regional center for evaluation and early intervention services
- ► Over 3 years of age: Refer to the school system for evaluation for early childhood program
- ► Possibly to Neurology dependent on results of evaluation

Resources used in development of these Referral Guidelines for Developmental Delay:

- Majnemer A, Shevell MI. Diagnostic yield of the neurologic assessment of the developmentally delayed child. J Pediatr 1995; 127:193-9.
- Yeargin-Allsopp M, Murphy CC, Cordero JF et al. Reported biomedical causes and associated medical conditions for mental retardation among 10-year old children, metropolitan Atlanta, 1985 to 1987. Dev Med Child Neurol 1997; 39:142-149.
- Schaefer GB, Bodensteiner JB. Evaluation of the child with idiopathic mental retardation. Pediatr Clin North Am 1992; 39:929-943.
- Battaglia A, Bianchini E, Carey JC. Diagnostic yield of the comprehensive assessment of developmental delay/mental retardation in an institute of child neuropsychiatry. Am J Med Genet 1999; 82: 60-66.
- Shevell M, Ashwal S, Donley D et al. Practice parameter: Evaluation of the child with global developmental delay. Neurology 2003; 60: 367-380.
- McDonald L, Rennie A, Tolmie et al. Investigation o global developmental delay. Arch Dis Child 2006; 91: 701-705.
- Michelson DJ, Shevell MI, Sherr EH, Moeschler JB, Gropman AL, Ashwal S. Evidence report: Genetic and metabolic testing on children with global developmental delay: report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. Neurology. 2011;77(17):1629-35.

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E. Tics [ICD-9 Code: 780.39] [ICD-10 Code: R56.9] and Tourette's [ICD-9 Code: 307.23] [ICD-10 Code: F95.2, F95.9]

Suggested Workup & Initial Management

- When tics are mild and not socially disabling, no treatment is required.
- Overall, simple childhood tic disorder and Tourette's syndrome have a favorable prognosis, with follow-up studies suggesting approximately 1/3 of children are essentially symptom-free as adults and another 1/3 with mild tics that do not require clinical attention.
- In the absence of other neurologic findings (cognitive delays, deterioration, abnormalities other than motor/vocal tics), tics, compulsions, and habits typically require no further diagnostic workup. Symptoms are expected to wax and wane with time. Family history of tics or obsessive-compulsive behavior is helpful in confirming the diagnosis; however, a positive family history is not essential to the diagnosis.
- If akathisia is present, it is important to determine whether it may be caused by other medications that the child is taking or has recently taken. It is also important to be aware of signs of an anxiety disorder and to closely observe to be sure that symptoms do not actually represent a form of chorea.

When to Refer

- If the tics are severe or socially disabling and have failed biofeedback, relaxation methods, or other behavioral techniques helpful in alleviating stress that potentially aggravates tics. In addition, patients with associated behavioral difficulties are often helped by individualized academic, vocational, social, or other supportive services.
- Consider psychiatry referral depending on presence and severity of co-morbidities such as obsessive compulsive disorder or anxiety disorders.

Resources used in development of these Referral Guidelines:

Tics/Tourette's:

- Cath DC, Hedderly T, Ludolph AG, et al. European clinical guidelines for Tourette syndrome and other tic disorders. Part I: assessment. Eur Child Adolesc Psychiatry. 2011;20(4):155-71.
- American Psychiatric Association (Corporate Author). Diagnostic and Statistical Manual of Mental Disorders, Dsm-5. American Psychiatric Publishing, Incorporated; 2013.
- Srour M, Lespérance P, Richer F, Chouinard S. Psychopharmacology of tic disorders. J Can Acad Child Adolesc Psychiatry. 2008;17(3):150-9.
- Irwin RS, Glomb WB, Chang AB. Habit cough, tic cough, and psychogenic cough in adult and pediatric populations: ACCP evidence-based clinical practice guidelines. Chest. 2006;129(1 Suppl):174S-179S.

Pediatric Neurology Referral Guidelines

F. Autism / Pervasive Developmental Disorder [ICD-9 Code: 299.00, 299.01] [ICD-10 Code: F84.0, F80.89, F84.9]

Suggested Workup & Initial Management

- Developmental surveillance should be performed at all well-child visits from infancy through school age, and at any age thereafter if concerns are raised about social acceptance, learning, or behavior.
- Recommended developmental screening tools include the Ages and Stages Questionnaire, the BRIGANCE® Screens, the Child Development Inventories, and the Parents' Evaluations of Developmental Status.
- Further developmental evaluation is required whenever a child fails to meet any of the following milestones: babbling or gesturing by 12 months; single words by 16 months; two-word spontaneous (not echolalic) phrases by 24 months; loss of any language or social skills at any age.
- Siblings of children with autism should be monitored carefully for acquisition of social, communication, and play skills, and the occurrence of maladaptive behaviors. Screening should be performed not only for autism-related symptoms but also for language delays, learning difficulties, social problems, and anxiety or depressive symptoms.
- For all children failing routine developmental surveillance procedures, screening specifically for autism should be performed using one of the validated instruments: the Checklist for Autism in Toddlers (CHAT) or the Autism Screening Questionnaire.
- Laboratory investigations, including audiologic assessment and lead screening, are recommended for any child with developmental delay and/or autism. Early referral for a formal audiologic assessment should include behavioral audiometric measures, assessment of middle ear function, and electrophysiologic procedures using experienced pediatric audiologists with current audiologic testing methods and technologies. Lead screening should be performed in any child with developmental delay and pica. Additional periodic screening should be considered if the pica persists.
- Genetic testing in children with autism, specifically high-resolution chromosome studies (karyotype) and DNA analysis for Fragile X, should be performed in the presence of intellectual disability (or if intellectual disability cannot be excluded), if there is a family history of Fragile X or undiagnosed intellectual disability, or if dysmorphic features are present. However, there is little likelihood of positive karyotype or Fragile X testing in the presence of high-functioning autism.

(Autism continued of next page)

When to Refer

- ► 0-3 years of age: Refer to regional center for evaluation and early intervention services
- ► Over 3 years of age: Refer to the school system for evaluation for early childhood program
- ► If dysmorphisms, consider genetics referral

Pediatric Neurology Referral Guidelines

F. Autism / Pervasive Developmental Disorder [ICD-9 Code: 299.00, 299.01] [ICD-10 Code: F84.0, F80.89, F84.9] (Continued)

Suggested Workup & Initial Management

- Selective metabolic testing should be initiated by the presence of suggestive clinical and physical findings such as the following: evidence of lethargy, cyclic vomiting, or early seizures; presence of dysmorphic or coarse features; evidence of intellectual disability; or if occurrence or adequacy of newborn screening is questionable.
- There is inadequate evidence to recommend an electroencephalogram study in all individuals with autism. Indications for an adequate sleep-deprived electroencephalogram with appropriate sampling of slow wave sleep include clinical seizures or suspicion of subclinical seizures and a history of regression (clinically significant loss of social and communicative function) at any age, but especially in toddlers and preschoolers.
- There is no clinical evidence to support the role of routine clinical neuroimaging in the diagnostic evaluation of autism, even in the presence of megalencephaly.
- There is inadequate supporting evidence for hair analysis, celiac antibodies, allergy testing (particularly food allergies for gluten, casein, Candida, and other molds), immunologic or neurochemical abnormalities, micronutrients such as vitamin levels, intestinal permeability studies, stool analysis, urinary peptides, mitochondrial disorders (including lactate and pyruvate), thyroid function tests, or erythrocyte glutathione peroxidase studies

When to Refer

- ► 0-3 years of age: Refer to regional center for evaluation and early intervention services
- ► Over 3 years of age: Refer to the school system for evaluation for early childhood program
- ► If dysmorphisms, consider genetics referral

Resources used in development of these Referral Guidelines:

- Asperger Syndrome and High Functioning Autism Tool Kit & the First 100 Days Kit available from www.autismspeaks.org
- Zwaigenbaum L, Bryson S, Lord C, et al. Clinical assessment and management of toddlers with suspected autism spectrum disorder: insights from studies of high-risk infants. Pediatrics. 2009;123(5):1383-91.
- Filipek PA, Accardo PJ, Ashwal S, et al. Practice parameter: screening and diagnosis of autism: report of the Quality Standards Subcommittee of the American Academy of Neurology and the Child Neurology Society. Neurology. 2000;55(4):468-79.

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G. Concussion [ICD-9 Code: 850.9] [ICD-10 Code: S06.0X*]

Suggested Workup & Initial Management

- Tools and additional resources are available on www.choc.org/concussion
- Anyone who is suspected to have suffered a concussion should be removed from activities that put them at risk for additional concussion including sports participation until he or she is evaluated by a physician with training in the evaluation and management of concussions.
- No athlete should be allowed to participate in sports if he or she is still experiencing symptoms from a concussion.
- Consider MRI scanning in instances where headache or other associated symptoms worsen or persist.
- Complete SCAT3 or Child SCAT3 in clinic.

When to Refer

- For any concussion requiring guidance from a specialist, please call: (714) 509-4054
- Please choose option #1 for Sports Concussion

Resources used in development of these Referral Guidelines:

- Halstead ME, Walter KD. American Academy of Pediatrics. Clinical report--sport-related concussion in children and adolescents. Pediatrics. 2010;126(3):597-615.
- Mccrory P, Meeuwisse WH, Aubry M, et al. Consensus statement on concussion in sport: the 4th International Conference on Concussion in Sport held in Zurich, November 2012. Br J Sports Med. 2013;47(5):250-8.

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H. Headaches [ICD-9 Code: 339.*, 346.*] [ICD-10 Code: G43*, G44*]

Suggested Workup & Initial Management

- Patients with recurrent headache and a normal neurologic exam generally do not require ancillary testing.
- Brain imaging studies (MRI) are suggested for patients who have headaches that awaken them in the middle of the night, patients whose headaches begin shortly after arising from bed in the morning (i.e. positional headache), or for patients whose headaches are associated with transient neurologic deficits (e.g. hemiparesis, ophthalmoparesis, confusion).
- MRI of the Brain is the preferred neuroimaging study (prefer MRI obtained at CHOC Children's). MRA and MRV may also be indicated in certain clinical settings

When to Refer

- ► Patients with a new severe headache of acute onset, headache with a focal neurologic deficit, or headache associated with papilledema should be referred to the Emergency Department
- ► Recurrent headache has been present for at least six months or for patients with headache associated with focal neurologic deficits. Consider referral if patient has severely disabling recurrent headaches that are not well managed with headache hygiene, migraine prophylaxis, and appropriate use of NSAIDs or other first line headache treatment strategies
- ► Consider psychiatry referral depending on presence and severity of co-morbidities such as obsessive compulsive disorder or anxiety disorders.

Resources used in development of these Referral Guidelines:

Headaches:

- Lewis DW, Ashwal S, Dahl G, et al. Practice parameter: evaluation of children and adolescents with recurrent headaches: report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. Neurology. 2002;59(4):490-8.
- Lewis D, Ashwal S, Hershey A, et al. Practice parameter: pharmacological treatment of migraine headache in children and adolescents: report of the American Academy of Neurology Quality Standards Subcommittee and the Practice Committee of the Child Neurology Society. Neurology. 2004;63(12):2215-24.
- Silberstein SD, Holland S, Freitag F, et al. Evidence-based guideline update: pharmacologic treatment for episodic migraine prevention in adults: report of the Quality Standards Subcommittee of the American Academy of Neurology and the American Headache Society. Neurology. 2012;78(17):1337-45.
- Holland S, Silberstein SD, Freitag F, et al. Evidence-based guideline update: NSAIDs and other complementary treatments for episodic migraine prevention in adults: report of the Quality Standards Subcommittee of the American Academy of Neurology and the American Headache Society. Neurology. 2012;78(17):1346-53.