Abstract:

Infants and children are commonly brought to the emergency department (ED) with a complaint of acute weakness. A good history and physical are paramount in determining etiology. Once a diagnosis is suspected, tests such as a lumbar puncture and magnetic resonance imaging of the spine will help to further define the etiology and guide therapy. Many causes of acute weakness have the potential for respiratory compromise, and ED practitioners must continually and closely evaluate patients and may choose to electively intubate. This article explores the 2 most common diagnoses made in children presenting with weakness, namely, Guillain-Barré syndrome and transverse myelitis, and briefly discusses other less common etiologies. Effective treatment of these disorders is not possible without prompt recognition of these patterns of weakness and initiation of diagnostic testing in the ED.

Keywords:

Guillain-Barré syndrome; transverse myelitis; spinal cord ischemia; tic paralysis; ischemic myelopathy; compressive myelopathy; pediatric acute weakness

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Conflict of interest: The author has no conflicts of interest to disclose.

1522-8401
Published by Elsevier Inc.

The Child With Acute Weakness

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Acute weakness is a fairly common presenting symptom of children in the emergency department (ED). The differential diagnosis is broad, but key pieces of the history and physical examination will help to localize the disease process and aid in making the diagnosis. The 2 most frequently encountered disorders in the ED causing acute weakness are Guillain-Barré syndrome (GBS) and acute transverse myelitis, which will be the focus of this article. Other less common disorders are summarized in Table 1. Despite the range of potential etiologies (inflammatory, vascular, traumatic, compression of the spinal cord [intradural or extradural], and infectious), an understanding of basic neuroanatomy and common disease features can help focus the diagnostic approach for the ED clinician.

Infants and preschool age children may have nonspecific symptoms of irritability, restlessness, waking repeatedly from sleep, refusal to walk and frequently wanting to be held, or loss of previously attained milestones.[2] Assessing a sensory level can be challenging in young children, but recognizing consistent landmarks such as the clavicle (C4), nipple (T4), umbilicus (T10), and inguinal ligament (T12) can help. Bedside examination of rectal tone, bladder retention by use of bladder ultrasound, respiratory status in older cooperative patients with use of negative inspiratory force, and close observation of vital signs are paramount for patients with acute weakness in the ED.

If the patient's examination demonstrates a sensory level, one must be suspicious for a spinal cord lesion, and urgent magnetic resonance imaging (MRI) is the imaging modality of choice. Table 2 illustrates examination findings in upper motor neuron vs lower motor neuron disorders. Acute transverse myelopathy (neurologic dysfunction of the spinal cord, in contrast to radiculopathy, dysfunction of nerve roots, and neuropathy, peripheral nerve dysfunction) is a medical emergency in children and adults.

GUILLAIN-BARRÉ SYNDROME

Background and Presentation

Guillain-Barre is an acute monophasic inflammatory demyelinating polyneuropathy characterized by rapidly progressive,

TABLE 1. Localization and characteristics of weakness along the neuroaxis.

Location of Disease	Category	Key History and Exam Findings	Key Laboratory and Electrodiagnostic Findings	Examples
Spinal cord	Myelopathy	Acute flaccid paralysis Loss of sphincter tone		Myelitis Infarction
	Motor neuron disease	Initial areflexia below the lesion		Trauma Tumor
Muscle	Myopathy	Proximal weakness Normal sensation May have pain, rash	Increased creatine kinase Diminished duration of motor potentials, reduced recruitment on EMG	Muscular dystrophy Postviral myositis Juvenile dermatomyositis
Peripheral nerve	Neuropathy	Distal weakness	Increased CSF protein, normal CSF WBC	GBS
		Diminished reflexes	Tick	Tick paralysis
		Ophthalmoplegia, ataxia	Decreased amplitude on NCS with axonal variants	MF variant of GBS
Neuromuscular junction	Neuropathy	Fatiguable weakness +Bulbar symptoms, ptosis	Fatigue with stimulation on EMG Fatigue with stimulation on EMG	Botulism Myasthenia gravis

NCS indicates nerve conduction studies; MF, Miller-Fisher; EMG, electromyelogram; CSF, cerebrospinal fluid; WBC, white blood cell; GBS; Guillain-Barre Syndrome.
Data from Cirillo.[1]

ascending, essentially symmetric weakness and areflexia in a previously well child,[3] which peaks in less than 4 weeks, is followed by a plateau phase, and then has a slow recovery.[4] The disease is thought to be caused by the autoimmune-mediated destruction of the peripheral myelin sheath and inflammation of the nerve roots. It is triggered by a preceding respiratory or gastrointestinal infection in up to two thirds of cases.[5–8] *Campylobacter jejuni* is the most frequently found gastrointestinal pathogen in patients with GBS. Incidence rates vary between 0.38 and 2.53 per 100 000 based on region of the world.[9] The overall rate for GBS in children is 0.34 to 1.34 per 100 000.[5,9] There are several variants based on examination and electromyography and nerve conduction velocity studies. The Miller-Fisher variant presents as a triad of ataxia, ophthalmoplegia, and areflexia. Overall, acute demyelinating inflammatory polyneuropathy is the most common variant occurring in 72 to 95% of cases.[7,8,10]

TABLE 2. Upper motor neuron versus lower motor neuron signs.

Characteristics	Upper Motor Neuron	Lower Motor Neuron
Lesion	Interrupt a neural pathway at a level above the anterior horn cell: motor pathways in cerebral cortex, internal capsule, cerebral peduncle, brainstem, spinal cord	Interrupt the spinal reflex arc. Clinical features result from lesion of spinal motor neurons, motor root, or peripheral nerve
Reflexes	Increased	Decreased
Babinski	Yes	No/mute
Muscle tone	Increased; spasticity	Decreased; flaccid
Weakness	Yes	Yes
Fasciculations	No	Yes
Etiology	Spinal cord lesions, stroke, multiple sclerosis, other acquired brain injuries	Guillain-Barré, early spinal cord shock, amyotrophic lateral sclerosis, botulism, polio, cauda equina syndrome

There is male predominance in GBS, and the mean age of onset is 4.9 (range, 5-7.5) years.[4–7,10,11] Presenting signs include weakness and areflexia or hyporeflexia in nearly all patients as well as neuropathy, with myalgias or leg pain and paraesthesias.[3,4,6–8,10] Children with GBS experience pain to a much higher extent than adults. Ataxia may also be present due to weakness or associated with the Miller-Fisher variant.[3] Sensory symptoms and cranial neuropathies including facial palsy, ophthalmoplegia, or bulbar involvement occur in up to half of patients.[3–6] Patients may be nonambulatory or have bladder or bowel sphincter disturbance at presentation.[4,8]

Guillain-Barré syndrome must be differentiated from transverse myelitis (TM) and anterior spinal