Acute Flaccid Paralysis Approach in children

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Acute flaccid paralysis

- Acute flaccid paralysis (AFP) is a clinical syndrome characterized by rapid onset of weakness, that frequently includes respiratory and bulbar weakness, progressing to maximum severity within several days to weeks.
- The term "flaccid" indicates the absence of spasticity or other signs of disordered central nervous system motor tracts such as hyperreflexia, clonus, or extensor plantar responses

- AFP is a complex clinical syndrome with a broad potential etiologies.
- Accurate diagnosis of the cause of AFP has profound implications for therapy and prognosis.
- If not managed appropriately, paralysis can progress to respiratory failure and death

- AFP, a syndrome that encompasses all cases of paralytic poliomyelitis, also is of great public health importance because of its use in surveillance for poliomyelitis in the context of the global polio eradication initiative.
- Any case meeting the AFP definition undergoes a thorough investigation to determine if the paralysis is caused by polio

- Each case of AFP is to be reported and 2 stool samples (>24 h apart, each of 10g) are collected within 14 days of paralysis onset and sent to WHO accredited laboratory.
- In the Global Polio Eradication Initiative, acute flaccid paralysis is defined as:

Any case of AFP in a child aged <15y, or any case of paralytic illness in a person of any age when polio is suspected

Objectives

The objective of this presentation will be:

- To provide practical approach to diagnosis in an individual patient
- - To provide an approach to rational use of diagnostic tests
- To discuss the common causes of AFP in children

- Each case of AFP is a clinical emergency and requires systemic evaluation and management .
- The immediate priorities in a child who presents with acute progressive weakness are:

- To detect and manage respiratory muscle weakness
- To detect and manage bulbar weakness
- To evaluate for cardiovascular instability
- To detect and manage dyselectrolytemia and toxemia
- To detect and manage spinal compression

Detect and manage respiratory muscle weakness

- Younger children with respiratory muscle weakness may present with nonspecific irritability, sweating, shallow or paradoxical breathing, poor feeding

-Older children may complain of respiratory difficulties, air hunger, agitation and poor respiratory effort

- Careful serial examination is critical to pick up the weakness early
- Early elective intubation and respiratory support may be critical to save these affected patients

Detect and manage bulbar weakness

- Symptoms of voice change, poor cry, pooling of secretions, gurgling sound in throat, difficulty in swallowing and choking on feeds may be markers of bulbar dysfunction
- Avoid oral feeding
- Provide regular suction

Evaluate for cardiovascular instability

- Some conditions that can lead to AFP eg. GBS, spinal trauma can result in cardiac rhythm abnormalities and cardiovascular insufficiency.
- ECG/cardiac monitor is an essential early step in the management

Detect and manage dyselectrolytemia and toxemia

- Hypokalemia/hyperkalemia and snake envenomation should be excluded in all children with AFP by history and examination early in management course
- Rapid assessment of electrolyte and ECG

Detect and manage spinal compression

- Patients with possible spinal injury should be identified by hx and examination
- Immediate spinal stabilization and administration of corticosteroid in patients with trauma would be a priority
- early neurosurgical intervention might be required to relief spinal cord compression and prevent long term disability

- For all cases, a detailed clinical description of the symptoms should be obtained, including fever, myalgia, distribution, timing, and progression of paralysis.
- The symptoms of paralysis may include gait disturbance, weakness, or troubled coordination in one or several extremities
- Many children with weakness present with nonspecific symptoms of irritability, lethargy and clumsy walk or refusal to walk
- Pseudoparalysis due to limb pain may result from trauma, arthritis, myositis or joint bleeding

- Comprehensive neurologic examination, including assessment of muscle strength and tone, pattern of weakness, deep tendon reflexes, cranial nerve function, and sensation
- Look for meningismus, ataxia, or autonomic nervous system abnormalities (bowel and bladder dysfunction, sphincter tonus, neurogenic reflex bladder)

Neuroanatomical approach

- It is useful to remember the possible causes of AFP in children using a neuroanatomical approach

A neuroanatomical differential diagnosis of acute flaccid paralysis in children

Site	Pathophysiology	Disease
Spinal cord	Compressive	Traumatic spinal injury, epidural abscess, hematoma, discitis
	Inflammatory	Transverse myelitis
Anterior horn cell	Viral	Poliomyelitis, vaccine associated poliomyelitis, Enteroviral myelitis, Japanese encephalitis
	Vascular	Anterior spinal artery infarction
Roots/nerves	Immune mediated	Guillain Barre syndrome,
	Toxin	Post diphtheritic, porphyria, arsenic
	Viral	Rabies
	Trauma	Injection related sciatic neuritis
Neuromuscular junction	Immune mediated	Myasthenia Gravis
	Drugs, toxins	Organophosphates, snake venom, drugs (aminoglycosides), Botulism
	Dyselectrolytemia	Hypermagnesemia
Muscle	Infection	Viral myositis
	Inflammation	Inflammatory myopathy (polymyositis)
	Channelopathy	Hypokalemic periodic paralysis
	Dyselectrolytemia	Hypokalemia

Selected clues in history and examination while evaluating a child with acute flaccid paralysis

Points in history and/or examination	Remarks
Fever at onset	Polio or enteroviral myelitis, Transverse myelitis, myositis, epidural abscess, and Koch spine (prolonged history)
Trauma: head/neck	Trivial trauma may lead to spinal compression in patients with cervical vertebral instability (Patients with Downs syndrome, congenital cervicovertebral anomalies or juvenile idiopathic arthritis)
Exposure	Toxins: lead, arsenic
	Snake envenomation
	Dog bite: Rabies
Preceding infectious prodrome/vaccination	Guillain Barre syndrome or transverse myelitis
	Sore throat, neck swelling, diphtheretic polyneuropathy (non/partly immunized)

Selected clues in history and examination while evaluating a child with acute flaccid paralysis (cont.)

Precipitating factors	Diarrhea: Hypokalemia, enteroviral myelitis
	Exertion or post parandial: Hypokalemic periodic paralysis
	Intramuscular injection: Polio, traumatic sciatic neuritis
Sensory loss/level	Compressive myelopathy, transverse myelitis
Early bowel/bladder involvement	Compressive myelopathy, transverse myelitis
Constipation in <1 y	Botulism (H/o honey exposure)
Prominent autonomic signs/symptoms	Guillain Barre syndrome, Rabies, acute myelopathy
Ascending weakness	Guillain Barre syndrome, Rabies, Varicella zoster virus, ascending myelitis

Selected clues in history and examination while evaluating a child with acute flaccid paralysis (cont.)

Ascending weakness	Guillain Barre syndrome, Rabies, Varicella zoster virus, ascending myelitis
Descending weakness	Diphtheria, Botulism
Prominent and early ptosis	Myasthenia Gravis, Botulism
Facial weakness	Guillain Barre syndrome, Myasthenia Gravis, Botulism
Fluctuating symptoms, fatigability	Myasthenia Gravis
Muscle tenderness	Myositis, inflammatory myopathy, (myalgias may be severe in Guillain Barre syndrome)
Muscle stretch reflexes	Absent: Guillain Barre syndrome, Polio, Diphtheria, spinal shock, at level of spinal cord damage
	Preserved : Myasthenia Gravis, periodic paralysis, Botulism
	Exaggerated: Below level of spinal lesion, Upper motor neuron lesion
Spinal tenderness, painful spine movement	Spinal trauma, epidural abscess or other extradural compression
Neck stiffness	Polio, enteroviral myelitis, Guillain Barre syndrome, transverse myelitis

Investigations

MRI Spine is indicated when there is a suspicion of spinal cord compression or transverse myelitis

More specifically, any child with hx of neck or back trauma, rapid onset flaccid quadriparesis, early or persistent bladder or bowel involvement, sensory loss or sensory level on exam., spinal tenderness or appearance of UMN signs on exam. Should get an MRI of the spine

Investigation

CSF examination

Raised CSF cell count would be seen in transverse myelitis, polio or enterovirus myelitis, rabies, other viruses myelitis

Raised CSF protein with normal cell count suggests GBS, post diphteretic polyneuropathy, rarely may be seen in transverse myelitis

CSF can be normal early in the course of these illnesses

Nerve Conduction studies and Electro Myography

Confirm the involvement of nerve and help in diagnosis of anterior horn cell diseases

Particularly helpful to confirm GBS

Repetitive nerve stimulation test to diagnose myasthenia gravis and botulism

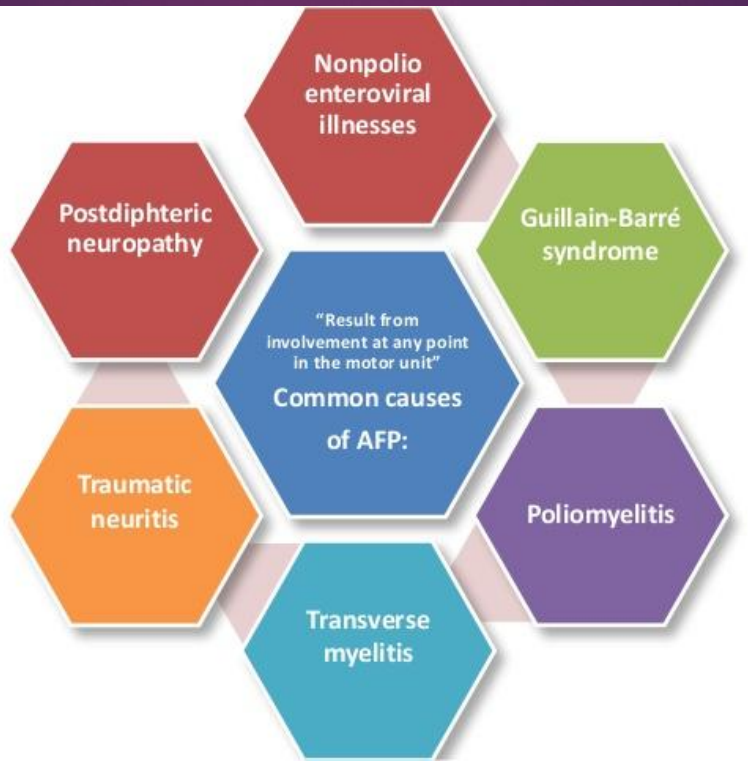
Rarely, may aid the diagnosis of an inflammatory myopathy

Creatine kinase

Raised level of CK reflects acute muscle injury and may points toward a muscle disease

In the setting of AFP this may be seen in children with viral myositis or inflammatory myopathy

Differential diagnosis of AFP



Guillain-Barre Syndrome

- The most common cause of AFP in healthy infants and children
- Occurred world-wide with an annual incidence of 0.34 to 1.34 cases per 100,000 persons aged 18 years or less
- Commonly occurs after an infection triggered immune mediated attack on the nerve axon or myelin
- Approximately two-thirds of patients give a hx of antecedent respiraratory or gastrointestinal infections. Campylobacter infection most commonly identified precipitant of GBS in about 30% of cases
- Other precipitants CMV,EBV, Mycoplasma pneumonia and influenza like illnesses

GBS (cont..)

- A small percentage of patients develop GBS after another triggering event such as immunization, surgery, trauma or bone marrow transplantation
- The available evidence suggests that no increased risk of GBS associated with the H1N1 influenza vaccine in children
- Historically GBS was considered a single disorder. It is now known to be a heterogeneous syndrome with several variant forms.
- The most common underlying subtype of the syndrome is Acute Inflammatory Demyelinating Polyneuropathy (AIDP), Acute Motor Axonal Neuropathy (AMAN)

GBS variants

- Atypical variants of GBS present with local or regional involvement of particular muscle groups or nerves, several have prominent cranial nerves involvement:

*Miller Fisher syndrome

*Bickerstaff brainstem encephalitis

*Polyneuritis cranialis

*Pharyngeal-cervical-brachial weakness

GBS clinical features

- The classic presentation of GBS begins with paresthesia in toes and fingertips, followed by lower extremity symmetric or modestly asymmetric weakness that may ascend to involve the arms and, in sever cases, the muscles of respiration
- Weakness progresses rapidly, about 50% of patients will reach nadir in 2wk, 80%in 3 wk, and the rest by 4 wk
- The predominant symptom of GBS at presentation in children are pain and gait difficulty
- In preschool-aged children, most common symptoms are refusal to walk and pain in legs
- Cranial neuropathy, mostly facial nerves, bilateral
- Pain typically involves the back and the legs

GBS.. Autonomic dysfunction

- Autonomic dysfunction occurs in about one half of children with GBS may include the following:
- A variety of cardiac dysrhythmias (asystole, bradycardia, persistent sinus tachycardia, and atrial and ventricular tachyarrhythmias)
- Orthostatic hypotension
- Transient or persistent hypertension
- Paralytic ileus
- Bladder dysfunction
- Abnormal sweating

GBS (cont..)

- Physical examination:

- Typically reveals symmetric weakness with absent reflexes and gait abnormalities

- Sensory symptoms are usually positive(eg. pain, paresthesia, reflecting nerve irritability) rather than negative(eg. sensory loss)
- Some cases present with initial proximal weakness, or less common findings such as sphincter disturbances, raising concerns about a possible spinal cord lesion

GBS (cont..)

- Risk factors for respiratory failure in GBS
- Cranial nerve involvement
- Short period between antecedent illness and onset of symptoms
- Rapid progression over less than 7 days
- Elevated CSF protein in the first week

Diagnosis of GBS

- Initial diagnosis of GBS based on the clinical presentation
- CSF analysis shows albuminocytologic dissociation, usually after the first week of onset of symptoms. In the acute phase the differentiation from polio or enteroviral myelitis based on CSF
- Nerve conduction studies
- Spinal MRI
- Antibodies against GQ1b, a ganglioside component of nerve, are present in the majority of patients with Miller Fisher syndrome

Management of GBS

- Observation of reparatory and bulbar muscle weakness
- Monitoring autonomic instability
- IVIG, 2g/kg spread over 2-5 days
- Plasmaphoresis

Prognosis of GBS in children

- Children have a shorter clinical course than adults
- Severity of the illness does not correlate with long term outcome, 85% of children have excellent recovery
- 50% are ambulatory by 6 months, 70% walk within a year of onset of the disease

Transverse Myelitis

- Acute demyelinating disorder of the spinal cord that evolves over days usually but may have a hyperacute presentation within hours
- May be associated with demyelination in other parts of the central nervous system, *Acute Disseminated Encephalomyelitis, Neuromyelitis optica*
- Commonly preceded by a viral infection or immunization
- Mean age of onset is 9 years

Transverse Myelitis (cont..)

- The common presentation includes an acute phase of spinal shock with flaccid paraparesis or quadreparesis urinary retention or incontinence, absent reflexes, sensory loss/level is frequently present, commonly thoracic sensory level. After few weeks signs of UMN dysfunction appear
- Visual acuity should be carefully checked to rule out Devic disease
- Back pain is common at the onset
- Urgent spinal MRI is needed to establish diagnosis, and to exclude spinal cord compression and other cause of myelopathy eg. Epidural spinal abscess or hematoma

Transverse Myelitis (cont..)

- CSF often shows increased WBC and protein, rarely positive for oligoclonal band

Management:

High doses of IV steroids (methylprednisolone) followed by tapering doses of prednisone

Poliomyelitis

- Both wild polio and vaccine associated polio virus can cause anterior horn cell damage to result in flaccid paralysis
- Children under 5 years are most frequently affected, however older individual and adults can also develop poliomyelitis
- Initial symptoms of poilo are non-specific and include fever, headache malaise, aseptic meningitis occurs in sever cases. Paralysis follow or accompanies these symptoms
- Rapid progression of paralysis within 1-2 days, pattern of muscle weakness varies but usually asymmetric, proximal more than distal limb paralysis

Poliomyelitis (cont..)

- Bulbar polio is often life threatening, affected children have prolonged episodes of apnea and require respiratory assistance
- Diagnosis: clinical findings, isolation and viral typing from the stool, CSF shows elevated WBC 50-200/mm
- Management is supportive
- Non polio enteroviruses can cause a polio like paralytic disease, eg coxsackie virus and echovirus

Traumatic neuritis

- Suspected in cases in which there is one limb involvement and definite hx of injection in that limb before the onset of paralysis
- Associated with pain and hypothermia in the affected limb
- Polio is a differential diagnosis; but sensory deficit and lack of CSF pleocytosis favor the diagnosis of traumatic neuritis
- Management is supportive

Periodic paralysis

- Periodic paralysis is a rare neuromuscular disorder, related to a defect in muscle ion channels, characterized by episodes of painless muscle weakness, which may be precipitated by heavy exercise, fasting, or high-carbohydrate meals.
- Periodic paralysis is classified as hypokalemic most common type, hyperkalemic or normokalemic . Most cases of periodic paralysis are hereditary, usually AD.
- Could be secondary to GI or renal loss of potassium
- Acquired cases of hypokalemic PP have been described in association with hyperthyroidism.

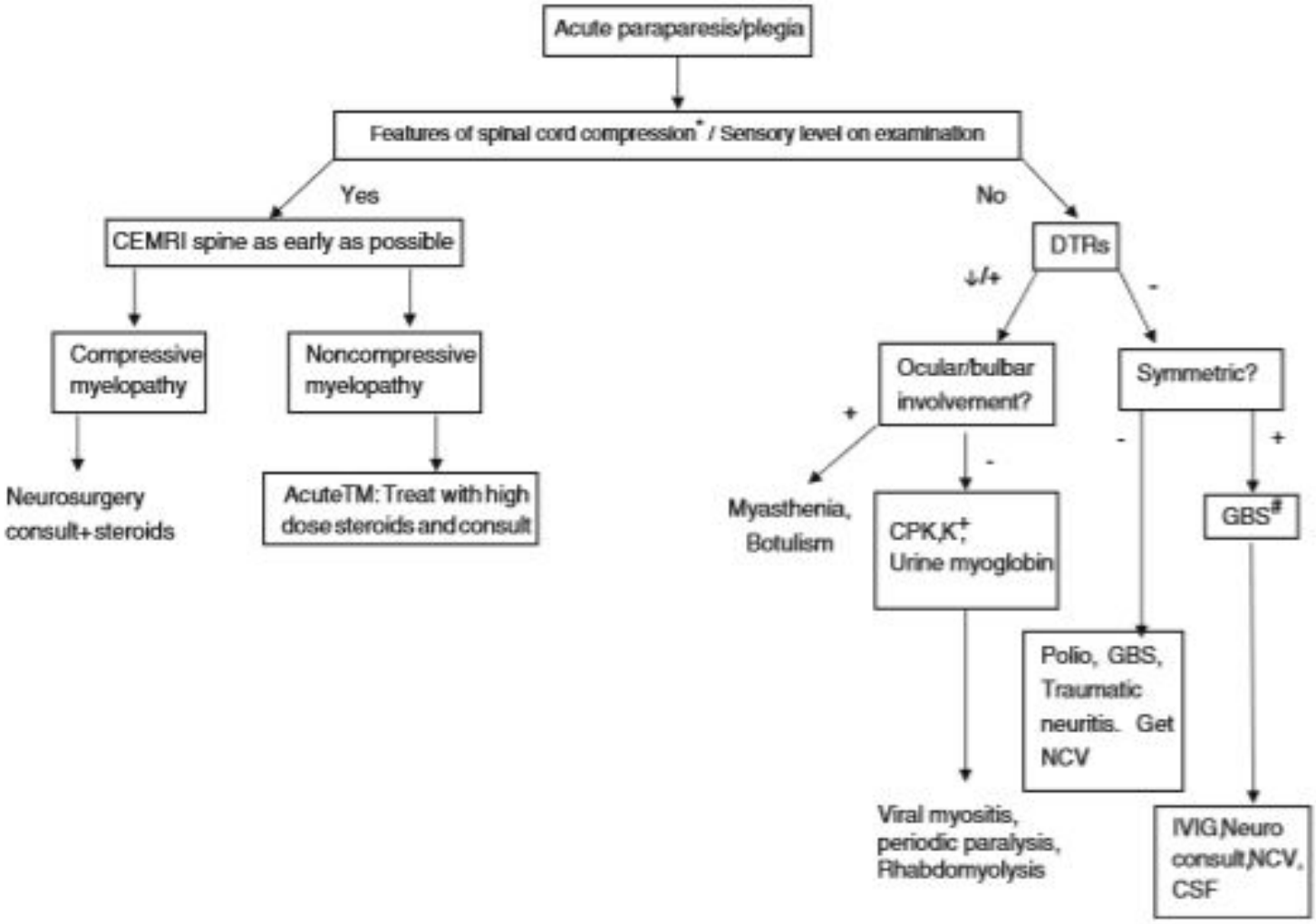
Periodic paralysis (cont..)

- Diagnosis: molecular testing is available,: but the clinical feature usually establish the diagnosis.. Low serum potassium and ECG changes. If you induce hypokalemia, paralysis follows within 2-3 hrs
- Treatment: correct potassium level
- Other electrolyte disturbances that might cause muscle weakness: hypophosphatemia, hypocalcaemia, hyponatremia and hypernatremia

Characteristics to aid differential diagnosis of acute flaccid paralysis

Feature	Transverse myelitis	Poliomyelitis	Guillain-Barre syndrome	Traumatic neuritis (following injection)
Development of paralysis	From hours to four days	24 to 48 h from onset to full paralysis	From hours to 4 wk	From hours to four days
Fever at onset of weakness	May be present	High, always present at onset of flaccid paralysis	Uncommon	Present, if underlying infection being treated with IM injections
Paralysis	Symmetric	Asymmetric,	Symmetric, mostly ascending	Affects only one limb
Progression of paralysis		Descending•	Ascending	
Muscle tone	Reduced during acute phase	Reduced	Reduced	Reduced
Deep-tendon reflexes	Absent in lower limbs(early); hyperreflexia(late)	Decreased or absent	Absent	Decreased or absent
Sensation	Anesthesia of lower limbs with sensory level	Severe myalgia, backache, no sensory changes	Cramps, tingling, hypoanesthesia of palms and soles	Pain in gluteus
Cranial nerve involvement	Absent	Only when bulbar involvement is present	Often present, affecting nerves VII, IX, X, XI, XII	Absent
Respiratory insufficiency	Sometimes	Only when bulbar involvement is present	Occurs in severe cases	Absent
Autonomic signs and symptoms	Present	Rare	Frequent in severe cases (blood pressure alterations, sweating, blushing, and body temperature fluctuations)	Hypothermia in affected limb
Cerebrospinal fluid	Normal or Pleocytosis	Mild elevation of lymphocytes 10 to 200/mL	Albumin-cytologic dissociation (usually <10 cells/ml, never >50cells/ml)	Normal
Bladder dysfunction	Present- early and persistent	Rare	Occasionally (Transient, at the peak of weakness, 1–3 d (30 %))	Never
Nerve conduction velocity: third wk	Normal	Abnormal: anterior horn cell disease (normal during first 2 wk)	Abnormal: slowed conduction, decreased motor amplitudes	Abnormal: s/o motor-sensory axonal damage
Diagnostic test	MRI–spine	Stool viral detection	Nerve conduction studies	Nerve conduction studies, Electromyography

Approach to child with acute paraplegia



Thank you