Neurology
Part 2
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Movement – Learning Objectives

Ataxia
• Know the common causes of acute ataxia
• Know how to evaluate a child with ataxia
• Know the prognosis of childhood acute cerebellar ataxia

Vertigo
• Know the common causes of vertigo

Chorea
• Know the common causes of chorea
• Know the etiology, clinical features, prognosis, and treatment of Sydenham chorea
Movement – Learning Objectives

Tics
• Distinguish between tics and other involuntary movements
• Differentiate between tic disorder and Tourette syndrome
• Know that tics and Tourette syndrome can be worsened by certain drugs
• Know the drugs useful in the treatment of Tourette syndrome
• Know that Tourette syndrome is associated with behavioral difficulties, learning disabilities, and attention deficit disorder

Drugs
• Know which drugs reverse the symptoms of drug-induced movement disorders
• Know which drugs can cause movement disorders
Movement Disorders

You have to see it ... to know what it is

• Classification:
  • Ataxia*
  • Chorea
  • Athetosis
  • Dystonia
  • Stereotopy
  • Tic
  • Myoclonus
  • Tremor

Each has a very precise definition which determines the differential
Ataxia

- Impaired ability to coordinate muscle activity in the execution of voluntary movement

- Clinical examination:
  - Gait:
    - Broad based, staggering, lurching
  - Dysmetria:
    - Dysdiadochokinesia, titubation, loss of checking response
  - Speech:
    - Slow, slurred, monotonous, irregular
  - Oculomotor:
    - Gaze-evoked nystagmus, abnormal saccades
Ataxia

**Etiology**
- Tumor
- Hemorrhage/Stroke
- Infection
- Guillain-Barre syndrome
- Labyrinthitis
- Toxic
- Migraine
- Trauma
- Acute cerebellar ataxia

**Evaluation...directed by history & PE**
- Toxicology screen
- Consider metabolic screening
- Consider LP
- Neuroimaging if:
  - Altered mental status
  - Focal neurologic exam
  - Asymmetry
  - Concern for increased ICP
  - Trauma

**Acute cerebellar ataxia**
- Acute-subacute onset
- 2-5 years of age
- Usually post-infectious
  - 4 days – 3 weeks post infection
  - ¼ associated with varicella
- Self-limited
  - Resolves by 2-3 weeks
  - Median duration 10-12 days
- Diagnosis of exclusion
Chorea

• Greek for “dance”

• **Clinical exam:**
  • Flowing, continuous, involuntary, irregular movements
  • Most often proximal extremities, neck, trunk and face
  • Flow from one joint to another
  • Exacerbated by voluntary action, stress and emotion

• **Etiology:**
  • Structural/Static injury
    • Cerebral palsy
    • Infarction/Vascular
    • Kernicterus
  • Simple metabolic
    • Electrolytes
    • Thyroid
  • Infection/Autoimmune
    • Sydenham
    • Lupus
    • Antiphospholipid Ab
  • Drugs/Toxins
  • Pregnancy
Sydenham Chorea

• Most common form of acquired chorea in childhood

• Acute rheumatic fever
  • Age 5-15 years
  • Female predominance
  • 1-8 months after infection

• Generalized (20-30% hemichorea)

• Most have psychological dysfunction
  • Personality changes, OCD, irritability, ADHD, regressed behaviors
  • May precede movement disorder by months

• No specific diagnostic test
  • 80% serological evidence of streptococcal infection

• Prognosis
  • Most resolve in 1-6 months
  • 20-40% recurrence

• Treatment:
  • Penicillin 10 days → prophylaxis
  • Valproic acid, haloperidol, pimozide, diazepam, carbamazepine
Stereotypy

• **Clinical:**
  - Onset <3 years
  - Involuntary, repetitive, rhythmic
  - Fixed pattern
  - Last minutes-hours
  - Suppressible with distraction
  - Increased with excitement, stress, anxiety and when engrossed
  - Hand waving, arm flapping, head nodding, body rocking

• 25% positive family history

• **Prognosis:**
  - Complex stereotypies can persist
  - 18% develop tics*

Harris, KM., Pediatric Neurology, 2008;38:267-272
Tics

• **Differentiate from:**
  • Stereotypy
  • Myoclonus

• **Provisional Tic Disorder**
  • Motor or vocal tics
  • <12 months
  • Most common tic disorder

• **Chronic Tic Disorder**
  • >12 months

• **Tourette syndrome**
  • Onset < 21 yrs of age
  • Multiple motor tics
  • One or more vocal tic
  • A waxing and waning course
  • Duration > 1 yr
  • Absence of medical explanation for tics
  • Verification by knowledgeable observer
Tourette Syndrome

Education is key!!!
- Treat tics only if they are disabling

Tic treatment
- 1st tier
  - Clonidine
  - Guanfacine
- 2nd tier
  - Atypical antipsychotics
  - Typical antipsychotics

Psychiatric co-morbidity is common
- ~50% ADHD
- ~50% OCD
- Learning disabilities
- Anxiety disorders

Treat the most distressing symptoms...if any at all
Drug-induced Movements

• Drug induced chorea:
  • Anticonvulsants (phenytoin, lamotrigine)
  • Antiemetics (metoclopramide)
  • Neuroleptics/neuroleptic withdrawal
  • Levadopa
  • Stimulants (amphetamines, cocaine)
  • Tricyclic antidepressants
  • Oral contraceptives
  • Theophylline

• Drug induced tremor:
  • Valproic acid

• Tardive dyskinesia:
  • Antipsychotics (typical>atypical)
  • Metoclopramide

• Treatment
  • Withdrawal/reduction of responsible drug

• Rx of TD
  • Benzodiazepines
  • Tetrabenazine
  • Anticholinergics
Vertigo

Environment is rotating relative to the patient
Localizes to the **vestibular system**
Both peripheral and central nervous system components

**Etiology**

- **Otitis media**
  - Mastoiditis, cholesteatoma
- **Migraine headache**
  - 19% of children have vertigo as part of aura
- **Benign paroxysmal vertigo of childhood**
  - Brief (<1 min) recurrent episodes
  - Occur in clusters over several days
  - Usually remits by 5 years of age
  - Frequently family history of migraine → may evolve to migraine

- **Trauma/Toxic**
- **CNS infection**
Neuromuscular – Learning Objectives

General

• Distinguish among acute and chronic causes of weakness
• Know that a Gowers sign indicates proximal muscle weakness

Testing

• Know the benefits and limitations of ancillary neurodiagnostic tests in the evaluation of weakness (eg, serum creatine kinase activity, electromyography)
• Understand the laboratory studies available to diagnose muscle disease of childhood
Neuromuscular – Learning Objectives

Hypotonia
• Distinguish between central and peripheral nervous system causes of hypotonia
• Know the differential diagnosis of hypotonia in infants
• Know how to evaluate hypotonia in infants

Guillain-Barre Syndrome
• Know the presenting signs and symptoms of Guillain-Barre syndrome
• Know the risk factors that are associated with Guillain-Barre syndrome (eg, recent immunization, varicella infection)
• Know the differential diagnosis of Guillain-Barre syndrome
• Know that cranial nerves may be affected in Guillain-Barre syndrome
• Know that autonomic dysfunction in Guillain-Barre syndrome may be prominent and dangerous
• Know that respiratory compromise may occur rapidly in Guillain-Barre syndrome
• Know the expected results of laboratory procedures such as examination of the cerebrospinal fluid, nerve conduction studies, and electromyography in Guillain-Barre syndrome
• Understand the treatment of Guillain-Barre syndrome (eg, IVIG, plasmapheresis)
Neuromuscular – Learning Objectives

Peripheral Neuropathy
• Recognize the clinical manifestations of childhood peripheral neuropathy
• Know the common causes of peripheral neuropathy in childhood (eg, hereditary sensory and motor neuropathy)
• Recognize the signs and symptoms of and plan treatment for Bell palsy
• Recognize the clinical manifestations of neonatal brachial plexus injuries
• Know the management and prognosis of neonatal brachial plexus injuries

Myasthenia
• Know the signs and symptoms of myasthenia gravis
• Understand the laboratory and electrophysiologic studies to evaluate children with myasthenia gravis
• Understand the appropriate management for a patient with myasthenia gravis
Neuromuscular – Learning Objectives

Muscular Dystrophy

• Formulate a differential diagnosis for a patient who has weakness and an increased serum creatine kinase activity

• Formulate a differential diagnosis for a patient who has an acquired muscle disorder (eg, inflammatory, infectious, toxic)

• Know the clinical features of dystrophinopathy (Duchenne/Becker muscular dystrophy)

• Know the natural history and late complications of the muscular dystrophies

• Know the genetics of dystrophinopathy (Duchenne/Becker muscular dystrophy)
Neuromuscular

• Approach to weakness
  • Time Course
    • Acute: stroke, Guillian-Barre
    • Chronic: muscular dystrophy, CMT
  • Neurologic localization
    • Central nervous system
    • Peripheral nervous system
      = MOTOR UNIT

Gower sign = proximal muscle weakness
Neuromuscular Testing

Uses & Limitations

• Laboratory testing
  • CK
  • Genetic testing

• EMG/Nerve conduction
  • NCS (Large, myelinated fibers)
    • Amplitude (axons)
    • Conduction velocity (myelination)
  • EMG:
    • Myofiber irritability
    • Motor units

• Skin biopsy
  • Small, unmyelinated fibers
Hypotonia

- **Hypotonia:**
  - Decreased resting tone (tension) of the muscle & decreased resistance to passive movement

- **Weakness:**
  - Reduction in maximum power that can be generated

- **Central:**
  - Hypotonia > weakness
  - Other features suggesting CNS involvement:
    - Seizures
    - Altered level of consciousness
    - Micro/macrocephaly
    - Dysmorphism

- **Peripheral:**
  - True weakness
  - +/- sensory loss
  - +/- diminished reflexes
Hypotonia

• **Differential Diagnosis:**
  • **Central:**
    • Brain malformations
    • Chromosomal / Genetic disorders
      (Down syndrome, Prader-Willi)
  • **Anterior horn cell:**
    • Spinal muscular atrophy
  • **Nerve**
    • Charcot-Marie-Tooth
  • **Neuromuscular junction:**
    • Congenital myasthenia
    • Botulism
  • **Muscle:**
    • Muscular dystrophy
    • Congenital/metabolic myopathy

- Brain MRI
- CGH
- Specific gene testing

- SMN gene testing

- EMG/Nerve conduction

- CK
- Gene testing
- Muscle biopsy
Guillain-Barre Syndrome (GBS)

- **Main features:**
  - Acute/subacute symmetric ascending paralysis
    - Progression over days-weeks, peak by 4 weeks
  - Motor > sensory
  - Weakness both proximal and distal
  - Globally depressed/absent reflexes

- **Other features:**
  - Pain (more common in children)
  - Cranial nerve or bulbar involvement
    - Facial nerves often affected
  - Respiratory failure
    - Must monitor FVCs Q4hrs
  - Autonomic involvement
    - Urinary retention, ileus, tachycardia, hyper- or hypotension, arrhythmias

- **Differential:**
  - Brain
    - Brain stem stroke
    - Encephalitis
  - Spinal cord
    - Compressive lesion
    - Transverse myelitis
  - Anterior horn cell
    - Poliomyelitis
    - West Nile virus
  - Peripheral nerve
    - Diphtheria
    - Acute intermittent porphyria
    - Vasculitis
    - Heavy metal intoxication
    - Tick paralysis
  - Neuromuscular junction
    - Myasthenia gravis
    - Botulism
  - Muscle
    - Periodic paralysis
    - Hypokalemia
    - Acute myopathy

GBS is a neurologic emergency

Long ... but narrowed by good neurologic exam
Guillain-Barre Syndrome

• CSF
  • “Cytoalbuminologic dissociation”
  • Elevated protein (80%)
    • May be normal early in disease
  • Normal WBCs

• MRI
  • Enhancement of lumbar nerve roots

• Nerve conduction testing
  • Demyelination
    • Reduced motor conduction velocity
    • Prolonged distal motor latency
    • Conduction block
    • Prolonged F-wave latency

• Treatment:
  • AAN Practice Parameter
    • Plasmapheresis and IVIG effective
    • Steroids are NOT effective (6 trials)
  • What about ambulatory patients:
    • Plasmapheresis helpful in 1 study in adults

• Outcome:
  • 4-15% mortality
  • 20% with residual disability
  • Severity and rapid onset \(\rightarrow\) worse outcome
  • 8-16% relapse
Peripheral Neuropathy

Clinical
- Weakness
- Sensory loss
- +/- autonomic dysfunction
- Depressed/absent reflexes

Pattern:
- Length dependent
- Single nerve territory

Etiology
- Hereditary Motor Sensory Neuropathies (CMT)
- Hereditary Neuropathy with Liability to Pressure Palsies (HNPP)
- Hereditary Sensory Autonomic Neuropathies (HSAN)
- Nutritional
- Toxic
### Specific Peripheral Neuropathies

#### Bell’s Palsy

<table>
<thead>
<tr>
<th><strong>Signs/Symptoms</strong></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Facial weakness—both upper and lower face</td>
<td></td>
</tr>
<tr>
<td>Decreased tearing</td>
<td></td>
</tr>
<tr>
<td>Hyperacusis</td>
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<tr>
<td>Loss of taste on anterior 2/3 of tongue</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Treatment</strong></th>
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</thead>
<tbody>
<tr>
<td>Prednisone x 7 days</td>
<td></td>
</tr>
<tr>
<td>Begin in first 72 hours</td>
<td></td>
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<tr>
<td>+/- valacyclovir</td>
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</tbody>
</table>

#### Brachial Plexus Injuries

<table>
<thead>
<tr>
<th><strong>Clinical manifestations</strong></th>
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<tbody>
<tr>
<td>Erb palsy (C5-C6) 50%</td>
<td></td>
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<tr>
<td>Arm adducted, internally rotated with extended elbow</td>
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<tr>
<td>Hand function intact</td>
<td></td>
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<tr>
<td>Klumpke palsy (C7-T1)</td>
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<tr>
<td>Isolated hand paralysis</td>
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</tr>
<tr>
<td>Horner’s syndrome</td>
<td></td>
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<tr>
<td>~50% other</td>
<td></td>
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</table>

<table>
<thead>
<tr>
<th><strong>Management</strong></th>
<th></th>
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</thead>
<tbody>
<tr>
<td>Conservative</td>
<td></td>
</tr>
<tr>
<td>Surgical repair considered if severe residual weakness at 6 months?</td>
<td></td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Prognosis</strong></th>
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<tbody>
<tr>
<td>Most recover spontaneously in 1-3 months</td>
<td></td>
</tr>
<tr>
<td>10-25% have residual weakness</td>
<td></td>
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</tbody>
</table>
Myasthenia Gravis

- **Clinical:**
  - Fluctuating weakness
  - Extra-ocular
  - Bulbar
  - Ptosis
  - Peripheral
  - Fatigue
  - Respiratory failure

- **Laboratory:**
  - Anti-AChR antibody

- **Nerve Conduction:**
  - Decrement with repetitive nerve stimulation

- **Treatment:**
  - Ach-esterase inhibitors
    - Pyridostigmine (mestinon)
  - Immunomodulating agents
    - Corticosteroids
    - Azathioprine
  - Rapid immunotherapy
    - IVIg
    - Plasmapheresis
  - Thymectomy

Progressive ptosis with sustained upgaze
### Weakness + Elevated CK

<table>
<thead>
<tr>
<th>Chronic</th>
<th>Acquired</th>
</tr>
</thead>
<tbody>
<tr>
<td>Muscular Dystrophy</td>
<td>Inflammatory myopathy</td>
</tr>
<tr>
<td>Duchenne/Becker</td>
<td>Dermatomyositis</td>
</tr>
<tr>
<td>Limb Girdle</td>
<td>Polymyositis</td>
</tr>
<tr>
<td>Emery Dreifuss</td>
<td>Rhabdomyolysis</td>
</tr>
<tr>
<td>Congenital muscular dystrophy</td>
<td>Post-viral</td>
</tr>
<tr>
<td>Metabolic Myopathy</td>
<td>Extreme exercise</td>
</tr>
<tr>
<td>Glycogen storage disorders</td>
<td>Alcohol/toxin/drug</td>
</tr>
<tr>
<td>Pompe</td>
<td>Underlying metabolic muscle disease</td>
</tr>
<tr>
<td>McArdle</td>
<td></td>
</tr>
<tr>
<td>Lipid Storage Disorders</td>
<td></td>
</tr>
<tr>
<td>CPT II deficiency</td>
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</tbody>
</table>
Duchenne/Becker Dystrophy

**Clinical:**
- Onset in early childhood
- Difficulty in running and climbing stairs
- Frequent falls
- Gower’s maneuver (proximal weakness)
- Enlarged calves
- 30% have intellectual impairment
- Loss of ambulation in adolescence
- Death by the third decade
  - Due to respiratory or cardiac

**Inheritance:**
- X-linked (Xp21)
- Dystrophin gene
- 1/3 spontaneous mutations

**Diagnosis**
- CK: 10-20X normal
- Gene Testing
- Dystrophin testing
Other CNS Disorders – Learning Objectives

**Hydrocephalus**
- Recognize the clinical features of hydrocephalus
- Recognize the signs and symptoms of shunt malfunction in hydrocephalus

**Micro- / Macrocephaly**
- Know the differential diagnosis of microcephaly
- Know the differential diagnosis of macrocephaly
- Know the causes and management of abnormal head shape
Other CNS Disorders – Learning Objectives

Cerebral Palsy
- Recognize the clinical features of cerebral palsy, including classifications
- Know that birth trauma and obstetric complications are not the leading causes of cerebral palsy
- Know the risk factors associated with cerebral palsy
- Know the disabilities associated with cerebral palsy: cognitive, visual, communication, auditory, motor, seizure activity, behavioral, oral function, nutrition
- Know the principles of management for children with cerebral palsy (eg, feeding, spasticity, mobility, activities of daily living, education)

Stroke
- Identify the clinical features of childhood stroke
- Know the causes of stroke in children
- Identify the clinical features of CNS arteriovenous malformations of childhood
Hydrocephalus

Clinical
• Symptoms
  • Headache
    • Often early morning
  • Vomiting>>nausea
  • Lethargy

• Signs
  • Macrocephaly
  • Dilated scalp veins
  • Setting Sun Sign
  • VI nerve palsy
  • Impaired upward eye gaze
    • Compression of midbrain
  • Spasticity of legs

VP Shunt Malfunction
• Presentation
  • Increased ICP
  • Meningitis

• Evaluation
  • CSF examination
  • HCT/”quick brain” MRI
  • Shunt series

Consult Neurosurgery
Head Shape

Microcephaly
• Etiology
  • Genetic syndromes
  • CNS structural abnormality
  • Metabolic disorders
  • CNS insult
    • HIE
    • TORCH infections
    • Stroke

Macrocephaly
• Etiology
  • Benign familial macrocephaly
  • Hydrocephalus
  • Genetic disorders
    • Neurocutaneous disorders
    • Metabolic disorders
Abnormal Head Shape

- Etiology of abnormal head shape
  - Positional plagiocephaly
    - “Tummy time”
  - Positioning with sleep
  - Helmets
    - Controversial
  - PT (if torticollis present)
  - TIME

- Craniosynostosis
  - Premature fusion of one or more cranial sutures
  - Evaluation with HCT
  - Referral to Neurosurgery
Cerebral Palsy

• Definition of cerebral palsy
  • ...a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain

• Classification
  • Spastic
    • Spastic diplegia
    • Spastic hemiplegia
    • Spastic quadriplegia
  • Dykinetic
  • Ataxic
  • Atonic

Birth trauma and obstetric complications are NOT the leading causes of cerebral palsy

• Preterm
  • Periventricular Leukomalacia (PVL)
  • Intraventricular Hemorrhage (IVH)
  • Ischemic injury
  • Brain malformations

• Term
  • Hypoxic Ischemic Injury
  • Neonatal Stroke
  • Brain Malformations
  • CNS infection
  • TORCH infections
  • Kernicterus

A report: the definition and classification of cerebral palsy April 2006
Dev Med Child Neurol Suppl. 2007 Feb;109:8-14
Cerebral Palsy

• Associated disabilities:
  • “The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, behavior, epilepsy, and by secondary musculoskeletal problems.”
  • Now acknowledged as part of the accepted definition of CP.

• Multidisciplinary team:
  • Pediatrician
  • Developmental pediatrician
  • Neurologist
  • Orthopedist
  • PT/OT/Speech
  • Feeding clinic
  • Psychologist
Stroke

• Clinical
  • Acute onset
  • Focal deficit

• Etiology
  • Congenital heart disease
  • Vasculopathies
    • Arteriopathy
    • Dissection
    • Moya moya
  • Hematologic
    • Sickle cell
  • Metabolic disorders
  • Infection
  • Arteriovenous malformations

• Epidemiology
  • 0.6-7.9/100,000

• Management
  • Supportive
  • ?Anticoagulation
    • Conflicting recommendations
    • Heparin for dissection or identified embolic source
  • Urgent exchange transfusion for SC
  • ASA

• Prognosis
  • Normal 25-37%
  • Mild disability 20-25%
  • Severe disability 16-49%
Spinal Cord – Learning Objectives

**Myelitis**
- Recognize the clinical manifestations of an acute spinal cord lesion
- Recognize the clinical manifestations of myelitis
- Plan the evaluation of a patient in whom post-infectious myelitis is suspected

**Myelomeningocele**
- Know that a myelomeningocele is usually associated with hydrocephalus
- Recognize the most common orthopedic problems associated with a myelomeningocele and their relative significance
- Understand the evaluation and fundamental long-term management of neurogenic bladder
- Know the fundamental long-term management of a neurogenic bowel
- Know the clinical and radiographic features and prognosis of spina bifida occulta
- Know the differential diagnosis of acute neurologic deterioration in a child with myelomeningocele
- Identify the clinical manifestations and plan the diagnostic evaluation of spinal
Myelitis

- Acute spinal cord lesion
  - Sudden onset
  - Impaired motor and sensory function ➔ look for a spinal level
  - Bowel/bladder dysfunction

- Neurologic emergency
  - Epidural abscess/hemorrhage
  - Stroke
  - Tumor
  - AVM
  - Transverse myelitis

- Transverse myelitis
  - Inflammation of spinal cord
  - Causes: idiopathic, post-infectious, rheumatologic, neuromyelitis optica

- Evaluation
  - Emergent MRI (with contrast)
  - CSF—MS panel
  - Serum:
    - NMO Ab, infectious, rheumatologic panel

- Treatment
  - Steroid burst
Myelomeningocele

- **Etiology**
  - Failure of primary neurulation (closure of spinal cord) by 28 days

- **Clinical**
  - Spinal level
  - Absent motor/sensory function

- **Diagnostic evaluation**
  - Prenatal: AFP
  - At birth: Abx, closure before 72 hours
  - Follow up: MRI for symptoms

- **Associated conditions:**
  - Chiari II malformation
  - Hydrocephalus
  - Tethered Spinal Cord
    - 25% symptomatic
  - Neurogenic bladder
    - Intermittent catheterization
    - Frequent UTIs
    - Anticholenergics
  - Neurogenic bowel
    - Regular bowel movements
    - Surgical
  - Scoliosis – common
  - Seizures
  - Learning Disabilities

*Spina bifida occulta is the mildest form of spinal dysraphism*
Trauma - Learning Objectives

Head Injury

- Know that the outcome of a head injury is related to the duration and degree of coma
- Know that a linear skull fracture in an infant younger than 1 year of age is a sign of possible child abuse
- Know the long-term neurologic and behavioral consequences of head trauma
- Recognize the neuroendocrine complications of a following head injury
- Recognize that cerebral edema is a consequence of head trauma

Intracranial Hemorrhages

- Recognize the clinical features of epidural hematoma
- Understand the clinical course and management of epidural hematoma
- Recognize the clinical features of subdural hematoma
- Understand the clinical course and management of subdural hematoma
- Recognize that intracranial hematomas can occur in the absence of a skull fracture
- Know the value and limitations of neurodiagnostic techniques such as magnetic resonance imaging, computed tomography, and ultrasonography
Trauma - Learning Objectives

Spinal Cord Trauma

• Recognize the clinical manifestations of an acute spinal cord lesion
• Know the signs and symptoms of spinal trauma
• Recognize the association of cervical cord injury with head trauma
• Know the association between atlantoaxial instability in Down syndrome and potential neurologic complications
• Recognize the significance of bladder and bowel dysfunction in spinal cord disease
• Plan the initial neurodiagnostic evaluation in a patient with acute spinal cord dysfunction
• Know the role of pharmacologic therapy in acute spinal cord or craniocerebral trauma
Head Injury

- **Head injury**
  - Acute issues:
    - Cerebral edema
    - Endocrine
    - Seizures
    - Hemorrhage
  - Long-term
    - Seizures
    - Headache
    - Behavioral

- **Key points:**
  - Outcome of a head injury is related to the duration and degree of coma
  - Linear skull fracture in an infant younger than 1 year of age is a sign of possible child abuse
Intracranial Hemorrhage

**Epidural hematoma**
- "Potential space" between dura and skull

- **Clinical:**
  - "Lucid interval"
  - Irritability, vomiting, lethargy, seizures

- **Management:**
  - HCT: Does not cross sutures → **lens shaped**
  - Trauma eval & Neurosurgical consult
  - Close neurologic monitoring

**Subdural hematoma**
- Between dural and arachoid membranes
- Tearing of bridging veins

- **Clinical**
  - Irritability, vomiting, lethargy, seizures
  - Frequently associated with non-accidental trauma

- **Management**
  - HCT: Limited by dural attachments → **crescent shaped**
  - Trauma eval & Neurosurgical consult

CT is the imaging modality of choice for blood
Spinal Cord Trauma

- **Clinical**
  - Focal pain
  - Spinal level
    - Flaccid paralysis
    - Absent reflexes
    - Diminished sensation
    - Loss of bowel/bladder function
  - Often associated with head trauma

- **Evaluation**
  - ABCs
  - Neck/spine immobilization
    - Athletic headgear should be left in place
  - Imaging—differs by center

- **Pharmacologic therapy**
  - Meta-analysis showed methylprednisolone within 8 hours → improved motor recovery

- **Atlantoaxial instability in Down syndrome**
  - AAP recommends annual PE to assess for neurologic signs of spinal cord injury
  - Neuroimaging recommended for only those with symptoms
Encephalopathy & Degenerative Disorders
Learning Objectives

Encephalopathy

• Know the common causes of an altered level of consciousness
• Recognize that disorders of metabolism, liver, kidneys, lungs or heart can be manifested as encephalopathy
• Plan the initial phase of evaluation for an altered level of consciousness
• Know to measure ammonia concentration and organic acid concentrations in neonatal coma
• Know which ingestions are likely to result in neurologic toxicity
• In a patient with an altered level of consciousness, know the items of importance in the history of ingestion
• Know which historic and physical findings should lead to consideration of child abuse as a cause of an altered level of consciousness
Encephalopathy & Degenerative Disorders

Learning Objectives

Degenerative Disorders

- Recognize the historical features indicative of a degenerative CNS disorder
- Recognize the signs and symptoms of degenerative CNS disorders
- Know the clinical presentation and course of Rett syndrome
- Understand the initial evaluation of a patient with suspected CNS degenerative disease
Altered Consciousness

Etiology
- Brainstem or Bilateral Cortical Hemisphere Dysfunction
- Toxic - Drugs
- Hypoxic
- Metabolic - HYPOGLYCEMIA
- Infectious/Postinfectious - ADEM
- Neoplastic/Paraneoplastic
- Structural – Hydrocephalus, ICP
- Trauma
- Ictal/Postictal
- Vascular – syncope, stroke, HIE, SAH
- Psychiatric

Evaluation
- History and Physical Examination
- Laboratory testing
  - Glucose
  - Ammonia
  - Lactate
  - Amino/Organic acids
- Urine Toxicology Screen
- Neuroimaging (CT or MRI)
- Lumbar Puncture
- EEG

Especially in neonatal coma

If concern for trauma—including non-accidental
Degenerative Disorders

- Relatively normal early development → progressive loss of neurological function

- Signs/symptoms

- Evaluation:
  - MRI
  - Genetic eval as indicated

- Classification
  - Gray matter
    - Neuronal ceroid lipofuscinosis
  - White matter
    - Adrenoleukodystrophy
  - Spinocerebellar
    - SCAs
  - Metabolic/Mitochondrial
    - Menkes’
Rett Syndrome

- **Clinical progression:**
  - Normal development (0-6 months)
  - Decelerated head growth (3 mo – 4 yrs)
  - Loss of purposeful hand use (9 mo – 2.5 yrs)
  - Stereotypic hand movements (1-3 yrs)

- **Associated manifestations:**
  - Seizures (30-80%)
  - Intellectual impairment
  - Sleep, GI, breathing and psychiatric issues
Neuroimaging

- **Ultrasound**
  - Open fontanelle
  - IVH, PVL

- **CT**
  - Acute blood
  - Calcifications

- **MRI**
  - Anatomy
  - Vascular structures
Neurophysiologic Testing

- EEG
  - Evaluation of seizure or altered consciousness
  - Negative EEG ≠ No seizures
  - Abnormal EEG ≠ Epilepsy

- EMG/nerve conduction testing
  - Peripheral nerves from cord to muscle
  - Only large, myelinated nerves
  - EMG of selected muscle only

- Evoked potentials
  - Electrographic response of brain to external stimulus
  - Auditory: coma, brainstem tumors
  - Visual: multiple sclerosis, neurodegenerative
  - Somatosensory: altered mental status, spinal cord disease
Changes You May Wish to Make in Practice

1. Contrast tics from stereotypies in affected patients
2. Consider treatment options for tics when distressing to the patient / family
3. Be familial with the diagnosis and treatment options for Guillain-Barre Syndrome
References: