Neurological and Neuromuscular Disorders

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Neuromuscular Birth Injuries: Overview

- Nerve damage caused by trauma during delivery
  - Prolonged labor
  - LGA
  - CPD
  - Abnormal presentation
  - Instrument-assisted delivery
- Nerves most commonly implicated
  - Cervical nerves 5, 6, 7, and 8
  - Thoracic nerve 1
  - Cranial nerve VII
  - Phrenic nerve
Neuromuscular Birth Injuries: Brachial Plexus Injuries

- **Presentation**
  - **Erb’s palsy:**
    - No spontaneous abduction or external rotation of affected arm (absent Moro)
    - Hand function is often preserved (grasp reflex present)
  - **Global plexus palsy (Erb-Duchenne-Klumpke):**
    - Flaccidity of affected arm and hand
    - Absent Moro and grasp reflexes
  - **Klumpke palsy:**
    - Flaccidity of hand and fingers of affected arm (present Moro, absent grasp)

- **Cause**
  - **Erb’s palsy:**
    - Most common, injury to nerve roots C5 and C6
  - **Global plexus palsy (Erb-Duchenne-Klumpke):**
    - Second most common, injury to nerve roots C5 through T1
  - **Klumpke palsy:**
    - Injury to nerve roots C8 and T1 only
Neuromuscular Birth Injuries: Brachial Plexus Injuries

- **Management**
  - Physical examination to assess extent of neurological involvement
  - X-ray if concern for fracture or shoulder dislocation
  - Neurology, orthopedic, and PT consultation
  - Passive ROM exercises when post-injury neuritis has resolved (7-10 days)
  - Use of wrist and/or finger splints, if indicated
  - Caregiver education regarding importance of passive exercise to maintain joint function

- **Complications**
  - Contractures may develop without passive exercise
  - Decreased sensation may lead to developmental deficits in affected arm

- **Outcome**
  - Spontaneous resolution generally occurs within 12 months
  - Best predictor of recovery is return of biceps function by 3 months of age
Neuromuscular Birth Injuries: Phrenic Nerve Injury

- **Presentation**
  - Typically associated with brachial plexus injury, but can occur alone
  - Respiratory distress often requiring oxygen and supportive ventilation

- **Cause**
  - Damage to phrenic nerve impairs nervous system stimulation of ipsilateral half of diaphragm

- **Management**
  - Supportive therapies including respiratory support
  - Surgical plication of diaphragm, if indicated

- **Complications**
  - Respiratory failure, pulmonary infection, growth failure, death

- **Outcome**
  - Mortality rate is 10 – 15%
  - Surviving infants generally recover within a year
Neuromuscular Birth Injuries: Facial Nerve Palsy

• **Presentation**
  – Persistent open eye on affected side
  – Suck with drooling on affected side
  – Mouth drawn to normal side when crying

• **Cause**
  – Trauma to nerve sheath (CN VII) during birth
  – Associated with instrument-assisted deliveries (forceps)

• **Management**
  – Provide artificial tears to open eye, a patch may be needed
  – Family support

• **Complications**
  – Feeding impairment

• **Outcome**
  – Spontaneous resolution is common (> 90% recover without intervention)
Hypoxic Ischemic Encephalopathy (HIE): Overview

- Cerebral injury associated with hypoxia and ischemia
- Incidence: 1-2 cases per 1000 term births with a mortality rate of 10 – 20%
- Hypoxemia: decrease in amount of oxygen circulating in the blood
- Ischemia: decrease in blood flow to brain (decreased perfusion)
  - Decreased glucose available
- Asphyxia:
  - Impairment of oxygen and carbon dioxide exchange
  - Initially causes increase in cerebral blood flow
  - Increasing levels of carbon dioxide contribute to acidosis
- Associated with widespread systemic injury secondary to hypoxic-ischemic insult
Hypoxic Ischemic Encephalopathy (HIE): Overview

- Associated antepartum conditions (20% of cases):
  - Maternal hypotension, placental vasculopathy
  - Contribute to decreased fetal reserves
- Intrapartum events (35% of cases):
  - Prolapsed cord, abruption, traumatic birth
- Combination of antepartum and intrapartum (35% of cases)
- Neonatal conditions (10% of cases):
  - Severe pulmonary disease, recurrent apnea
  - Congenital heart disease
- Preterm infant is at greater risk of HIE than term infant
HIE: Presentation

- **Stage I (mild encephalopathy)**
  - Hyperalert, normal muscle tone, active suck, strong Moro reflex, (+) myoclonus, hyper-responsive to stimuli

- **Stage II (moderate)**
  - Lethargy and hypotonic, (+) myoclonus, seizures common, weak reflexes with overall increased tendon reflexes

- **Stage III (severe)**
  - Comatose, apnea and bradycardia, seizures typical within 12 hours of birth, severe hypotonia and flaccidity, absent reflexes, pupils often unequal, variable reactivity, poor light reflex
HIE: Management

• **Diagnostic testing:**
  – Neurologic examination (Sarnat criteria)
  – Conventional EEG (cEEG)
  – Amplitude-integrated EEG (aEEG)
  – Neuroimaging
    • Head ultrasound
    • CT scan
    • MRI

• **Interventions:**
  – Resuscitation and stabilization
  – Therapeutic hypothermia
  – Family support and education
  – Palliative care
HIE: Complications

- Multisystem disorders are common with stage II and III HIE
  - Renal and cardiac abnormalities
  - Pulmonary hypertension
  - Liver function abnormalities
  - Thrombocytopenia
  - Disseminated intravascular coagulation (DIC)
HIE: Outcome

• Mild encephalopathy:
  – Recovery expected
  – Good outcome with very small risk of long-term disability

• Moderate encephalopathy (in absence of therapeutic hypothermia):
  – 6% death
  – 30% disability

• Severe encephalopathy (in absence of therapeutic hypothermia):
  – 60% death
  – 100% disability
Intraventricular Hemorrhage (IVH): Overview

- **Significant injury in the preterm brain**
- **Germinal matrix hemorrhage:**
  - Germinal matrix is immature and highly vascularized area of preterm infant brain
  - Site of neuron and glia development
  - Poorly supported and fragile blood vessels, sensitive to blood pressure fluctuation and reperfusion injury
    - Hypotension/hypertension, perinatal asphyxia, rapid volume infusions, myocardial failure, hypothermia, hyperosmolarity, etc.
  - Involution of germinal matrix occurs with advancing gestational age, germinal matrix disappears by 36 weeks, GM hemorrhage less common in infants > 32 weeks
- **Germinal matrix hemorrhage may extend to fill lateral ventricles and worsening IVH**
Intraventricular Hemorrhage (IVH): Overview

- **Incidence:**
  - 30 – 40% of infants <1500 grams or <30 weeks PMA
  - <228 weeks PMA have a 3-fold higher risk than 28 – 31 weeks PMA
  - 2 – 3% in term infants

- **Timing of onset:**
  - 50% by 24 hours
  - 80% by 48 hours
  - 90% by 72 hours
IVH: Presentation

- Sudden deterioration: oxygen desaturation, bradycardia, metabolic acidosis, falling hematocrit, hypotonia, shock, hyperglycemia
- Symptoms of worsening hemorrhage: full or tense fontanelle, increased ventilator support, seizures, apnea, decreased activity, decreased level of consciousness
- Rapid and profound clinical decline associated with increased severity of IVH
- Grading of IVH
IVH: Management

• **Neuroimaging**
  – Routine head ultrasound screening of infants born at < 30 weeks PMA
  – Serial head ultrasounds to monitor progression
  – MRI if parenchymal injury is suspected

• **Supportive Care**
  – Minimize stimulation
  – Avoid wide swings in blood pressure
  – Closely monitor respiratory support
  – Avoid acidosis, hypercarbia, fluid overload
IVH: Complications

- Neurodevelopmental disabilities
- Progressive hydrocephalus
- Seizures
- Death
IVH: Outcome

- **Mild/small IVH**
  - Neurodevelopmental disabilities (NDD) similar to premature infants without hemorrhage, major NDD 10%

- **Moderate IVH**
  - Major NDD in 40%
  - Mortality rate 10%
  - Progressive hydrocephalus in 20%

- **Severe IVH**
  - Major NDD in 80%
  - Mortality rate 50 – 60%
  - Progressive hydrocephalus common
Periventricular Leukomalacia (PVL): Overview

- **Severe white matter injury** highly associated with preterm birth
- **Focal injury: cystic necrotic lesion** found bilaterally
  - Nonhemorrhagic and symmetric
  - Caused by ischemia from fluctuations in arterial circulation
- **Diffuse white matter injury**
  - Noncystic lesions associated with disturbances in myelinization
  - Often associated with germinal matrix hemorrhages or IVH
- **Leukomalacia**: ”softening” of white matter
PVL: Presentation

- **Acute phase:**
  - Subtle
  - Altered muscle tone in lower extremities, hypotension, lethargy

- **6 – 10 weeks after white matter injury**
  - Irritable, hypertonic, increased flexion of arms and extension of legs, frequent tremors and startles
  - Moro reflex abnormalities
PVL: Management

• **Diagnostic evaluation:**
  – Head ultrasound
  – CT scan or MRI

• **Interventions:**
  – Treat primary insult
  – Supportive care to prevent further hypoxic-ischemic damage
  – Treatment of hydrocephalus and associated neurological sequalea
  – Family support and anticipatory guidance
  – Developmental care, PT/OT, feeding support
PVL: Complications

- Spastic diplegia
- Intellectual deficits, learning disorders
- Hyperactivity disorders
- Visual impairment
- Lower limb weakness
PVL: Outcome

- Determined by location and extent of injury
- Spastic diplegia reported in as many as 50% of infants with PVL
- Neurodevelopmental follow-up and developmental support improve outcomes related to learning and behavioral disorders
Seizures: Overview

- Sign of malfunctioning neuronal system
- Excessive simultaneous electrical discharge

Causes include:
- Metabolic encephalopathies
- Structural abnormalities
- Meningitis
- Drug withdrawal
- Genetic etiology
Seizures: Overview

- **Metabolic encephalopathies:**
  - Hypoglycemia
  - Ischemia
  - Hypoxemia
  - Hypo- or hypernatremia,
  - Hypocalcemia
  - Hypomagnesemia
  - Inborn error of metabolism
  - Pyridoxine deficiency
  - Hyperammononemia
Seizures: Overview

- **Structural abnormalities:**
  - HIE
  - IVH
  - Intrapartum trauma
  - Perinatal stroke
  - Cerebral dysgenesis
Seizures: Overview

- **Other causes:**
  - Meningitis
    - Group B streptococcus
    - Listeria monocytogenes
    - TORCH etiology
  - Drug withdrawal
    - Prenatal or postnatal exposure to opiates
  - Genetic (familial)
    - Self-limiting
Seizures: 
*Presentation*

- **Subtle (motor automatisms)**
  - Rowing, stepping, pedaling movements, eye blinking/fluttering, staring, lacrimation, smacking of lips, salivation, sucking
- **Clonic**
  - Rhythmic movements of muscle groups in a focal distribution
  - Rapid phase followed by a slow return to movement
  - Not stopped with flexion
- **Tonic (postural)**
  - Sustained generalized tonic extension of all extremities or flexion of the upper limbs with extension of the lower extremities
  - Characteristic of preterm infants with severe IVH
  - May closely mimic decerebrate or decorticate posturing
- **Multifocal clonic (generalized)**
  - Clonic movements that migrate from one limb to another without a specific pattern
  - Associated with significant morbidity and mortality
Seizures: Management

• Diagnostic evaluation:
  – Review perinatal/neonatal clinical course and family history
  – Blood glucose immediately to rule out hypoglycemia
  – Physical examination
  – Lab studies (blood gas, electrolytes, CBC with differential)
  – Septic workup if infectious etiology suspected
    • Blood, urine, CSF cultures
    • Nasal and rectal swabs if HSV suspected
  – Metabolic studies
  – Head ultrasound, CT, MRI
  – EEG
Seizures: Management

- Supportive care
- Careful assessment of clinical seizure activity
- Medication management:
  - Phenobarbital
  - Fosphenytoin
  - Levetiracetam
  - Lorazepam
Seizures: 
Complications and Outcome

- Untreated sustained seizures exacerbate underlying pathology
- Outcome varies significantly based upon etiology:
  - Familial seizures: often benign and self-limiting
  - Refractory seizures associated with HIE: severe morbidity and mortality
Subdural Hemorrhage: Overview

- Rupture of draining veins and sinuses that occupy the subdural space
- Due to molding and torsional forces on the head during birth
- Risk factors:
  - Macrosomia, CPD, shoulder dystocia
  - Traumatic birth
  - Vaginal breech presentation
  - Malpresentation
  - Instrument-assisted vaginal birth
Subdural Hemorrhage: Presentation

- Subdural hemorrhage accounts for less than 10% of all intracranial bleeds
- **Large hemorrhage:**
  - Nuchal rigidity, coma, abnormal respiratory pattern, unreactive pupils, signs of increased ICP, seizures, signs of hypovolemia and anemia
- **Small hemorrhage:**
  - Subtle or few signs until hematoma presses on brain tissue, may cause deterioration in mental status, development of hydrocephalus, seizures
Subdural Hemorrhage: Management

- Supportive care and seizure management
  - Volume replacement, respiratory support, pressor support
- Close monitoring of neurologic status
- Subdural tap or subdural shunt in infants with increasing ICP
- Monitor and intervention for progressive hydrocephalus
  - May occur weeks after the hemorrhage
Subdural Hemorrhage: 
*Complications*

- Hydrocephalus
- Seizures
- Neurodevelopmental impairment
Subdural Hemorrhage: *Outcome*

- Outcome dependent upon severity of hemorrhage
- Mortality rate may be as high as 45%
Hydrocephalus: Overview

• Excess of CSF in ventricular system
• Caused by inadequate reabsorption of CSF
  – Aqueductal outflow obstruction (non-communicating hydrocephalus)
    • Dandy-Walker cyst, myelomeningocele with Arnold-Chiari malformation, infection
  – Flow between lateral ventricles and subarachnoid space (communicating, non-obstructive hydrocephalus)
Hydrocephalus: 
*Presentation*

- Increasing head circumference
- Widened sutures
- Full, bulging, or tense fontanelles
- Setting-sun eyes
- Vomiting, lethargy, irritability
Hydrocephalus: Management

- Diagnostic testing: determine underlying cause, identify site of obstruction (if obstructive)
- Supportive care: decreased stimuli, minimal handling, monitor head circumference measurements
- Mechanical CSF drainage:
  - Short term: lumbar puncture or direct ventricular access
  - Long term: ventriculo-peritoneal shunt
  - Procedural and post-op care
Hydrocephalus: Complications

- Neurological deterioration associated with increased ICP
- Infection of VP shunt, infection associated with LP and ventricular access
Hydrocephalus: 

*Outcome*

- Determined by underlying cause
Neural Tube Defects: Overview

• **Primary NTD**
  – Failure of neural tube closure or disruption of closed tube
  – Occurs between 18-25 days of gestation
  – Location of neural tube failure determines presentation
  – Anencephaly, encephalocele, myelomeningocele

• **Secondary NTD**
  – Abnormal development of the lower sacral or coccygeal segments during secondary neurulation
  – Defects present primarily in lumbosacral spinal region
  – Skin typically intact over lesion
  – Meningocele, lipomeningocele, sacral agenesis/dysgenesis
Neural Tube Defects: Anencephaly

- Presentation
- Etiology
- Management
- Complications
- Outcome
Neural Tube Defects: Encephalocele

- Presentation
- Etiology
- Management
- Complications
- Outcome
Neural Tube Defects: Myelomeningocele

- Presentation
- Etiology
- Management
- Complications
- Outcome
Neural Tube Defects: Meningocele

- Presentation
- Etiology
- Management
- Complications
- Outcome
Neural Tube Defects: Sacral Agenesis/Dysgenesis

- Presentation
- Etiology
- Management
- Complications
- Outcome