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

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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

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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

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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

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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)

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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**

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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**

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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

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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

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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)

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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

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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**

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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

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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**

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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

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IVH:
Complications

- **Neurodevelopmental disabilities**
- **Progressive hydrocephalus**
- **Seizures**
- **Death**

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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

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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**

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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 abnomalities

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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

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PVL:
Complications

- **Spastic diplegia**
- **Intellectual deficits, learning disorders**
- **Hyperactivity disorders**
- **Visual impairment**
- **Lower limb weakness**

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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**

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Seizures: *Overview*

- **Sign of malfunctioning neuronal system**
- **Excessive simultaneous electrical discharge**
- **Causes include:**
  - Metabolic encephalopathies
  - Structural abnormalities
  - Meningitis
  - Drug withdrawal
  - Genetic etiology

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Seizures: *Overview*

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

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Seizures: *Overview*

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

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Seizures: *Overview*

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

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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

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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

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Seizures:
Management

- **Supportive care**
- **Careful assessment of clinical seizure activity**
- **Medication management:**
  - Phenobarbitol
  - Fosphenytoin
  - Levetiracetam
  - Lorazepam

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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

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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

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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

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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

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Subdural Hemorrhage: *Complications*

- Hydrocephalus
- Seizures
- Neurodevelopmental impairment

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Subdural Hemorrhage: *Outcome*

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

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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)

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Hydrocephalus: *Presentation*

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

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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

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Hydrocephalus: *Complications*

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

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Hydrocephalus: *Outcome*

- **Determined by underlying cause**

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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

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Neural Tube Defects: Anencephaly

- Presentation
- Etiology
- Management
- Complications
- Outcome

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Neural Tube Defects: Encephalocele

- Presentation
- Etiology
- Management
- Complications
- Outcome

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Neural Tube Defects: Myelomeningocele

- Presentation
- Etiology
- Management
- Complications
- Outcome

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Neural Tube Defects: Meningocele

- Presentation
- Etiology
- Management
- Complications
- Outcome

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Neural Tube Defects: Sacral Agenisis/Dysgenisis

- Presentation
- Etiology
- Management
- Complications
- Outcome

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