Pediatric neurology

Neuroanatomy

- The neurologic system is composed of;
- Central nervous system (CNS)
  - Comprises the brain and spinal cord.
  - The brain and the spinal cord are covered by a protective three-layer membrane called the meninges.
  - CSF contains water, glucose, protein, and minerals. CSF surrounds the brain helping to absorb shock
  - The brain is divided into three areas. These are the cerebrum, cerebellum, and brainstem.
  - The spinal cord transmits impulses to and from the brain
- Peripheral nervous system
  - Divided into cranial and spinal nerves
  - Responsible for voluntary function and for reflex actions, and conscious and subconscious mental processes.
- Autonomic nervous system (ANS).
  - Divided into sympathetic and parasympathetic nervous systems
  - Responsible for involuntary function.

N**euroanatomical differences in children and adults**

- Small head size, 2/3 brain weight of adult but large in relation to body
- Cranium not well developed, prone to fracture
- Very vascular brain, prone to hemorrhage
- Weak neck muscles not able to adequately support head
- CSF produces at 100ml per day (500ml per day in adults)
- Open fontanelles
- Fragile neurons, nerve cells not completely myelinated
- Excessive spinal mobility
- Developmental; poor judgment of danger, unsteady gait, immature immune system

Neurological examination

Evaluation of function involves;

- Cerebral function; mental status and behavior.
  - Includes appearance, judgment, memory, thought process, language and speech, mood and affect and orientation.
  - Level of consciousness involves assessment of ability to cry, level of activity, positioning and general appearance
- Cranial nerves; possible to test all in school age onwards
- Cerebellar function; posture, balance and coordination
- Motor system; musculoskeletal system
- Sensory system
- Reflex status
  - Deep tendon reflexes-intactness of reflex arc at specific spinal levels- biceps, triceps, brachioradialis, patellar, achilles. Evaluate strngth and symmetry on R&L side
  - Superficial reflexes- receptors in skin- abdominal, cremasteric, gluteal, planter
  - Newborn reflexes- must be lost before motor development proceeds

Neurological alterations

Meningitis

Inflammation of the meninges.

- Causes include
  - Microbial (age dependent)
    - Meningococcus.
    - Streptococcus.
    - Staphylococcus.
    - Haemophilus type B
    - Pneumococcus.
  - Contaminated head injury.
  - Infected shunt.
  - Contaminated lumbar puncture.
- The severity of the disease is dependent upon the specific microorganism involved, the presence of other neurological disorders, the general health of the patient, the speed of diagnosis, and the initiation of treatment

Pathophysiology

- Infectious microorganisms travel to the meninges via the bloodstream or through direct extension from an infected area (such as the middle ear or paranasal sinuses)
- Gain access to CSF and spread to subarachnoid space
- Inflammatory response occurs and WBC's accumulate over surface of brain with thick purulent exudate
- Brain becomes hyperemic and edematous resulting in increased intracranial pressure
- Hydrocephalus may occur if CSF flow is impeded due to infection and obstruction of the ventricles

DX

- Lumbar puncture to identify the causative organism in the cerebrospinal fluid.
- Blood cultures.
- Physical examination.

Clinical presentation

- Infants and toddlers; irritability, lethargy, bulging fontanelle , elevated temperature, alterations in feed/sleep pattern, vomiting and diarrhea, diminished level of consciousness with depressed respiratory rate
- Older child;
  - GI upset
  - Flu like symptoms
  - Chills.
  - Headache (often severe).
  - Nausea, vomiting or diarrhea
  - Cortical involvement; irritability, agitation,confused, delirious, lethargic, projectile vomiting
  - Nuchal rigidity (stiffness of the neck) and Opisthotonos (extreme hyperextension of the head and arching of the back due to irritation of the meninges), myalgias and headache-spinal nerves involvement
  - Photophobia, diplopia, tinnitus-cranial nerve involvement
  - Altered level of consciousness.
  - Signs of shock
  - Rash with multiple petechiae on the body- meningococcal, urgent need of medical attention
  - Hyper reflexes with positive kerning and brudzinski signs

Nursing management

1. Administer intravenous fluids and medications, as ordered by the physician.
    1. a. Antibiotics should be started immediately.
    2. b. Corticosteroids may be used for the critically ill patient.
    3. c. Drug therapy may be continued after the acute phase of the illness is over to prevent recurrence.
    4. d. Record intake and output carefully and observe patient closely for signs of dehydration due to insensible fluid loss.

2. Monitor patient's vital signs and neurological status and record.

a. Level of consciousness. Utilize GCS for accuracy and consistency.

b. Monitor rectal temperature at least every 4 hours and, if elevated, provide for cooling measures such as cooling sponge baths, and administration of ordered antipyretics

3. If isolation measures are required, inform family members and ensure staff compliance of isolation procedures in accordance with standard operating procedures (SOP).
4. Provide basic patient care needs.

a. The patient's level of consciousness will dictate whether the patient requires only assistance with activities of daily living or total care.

b. Maintain dim lighting in the patient's room to reduce photophobic discomfort.

5. Provide discharge planning information to the patient and family.

a. Follow up appointments with the physician.

b. Discharge medication instruction.

Spina bifida

- Neural tube defect where there is incomplete closure of the vertebrae and neural tube.
- Develops during the first 28 days of gestation
- It may be as a result of failure of the neural tube to close completely during the 4^{th} week of gestation or as a result of a fissure resulting from increased CSF pressure.
- Exposure to chemicals and medication during pregnancy and mother's low intake of folic acid are associated factors.
- Spina bifida may occur any where along the spinal cord. The most common defects include:

Level of the brain

Anencephaly

- Anencephaly is the most severe form of neural tube defect.
- Neonates with anencephaly rarely survive more than a few hours or days unless in presence of intact midbrain
- These children have been the subject of myths, folklore, and superstitions, and have been referred to as monsters due to frightening appearance.
- It is the most common CNS malformations in the West.
- There is absence of the calvaria with cerebral hemispheres either missing or greatly reduced

Encephalocele

- Protrusion of brain and meninges into a fluid filled sac through a skull defect
- Cranium bifidum, such as an encephalocele, is much more serious.
- Encephaloceles occur when the anterior neuropore fails to close at 26-28 wks gestation.

Cranioschisis

- Skull defect through which neural tissue protrudes.

Exancephaly

- Brain is totally exposed or herniated through a skull defect

Level of spinal cord

Spina bifida cystica

Defect in closure of posterior vertebral arch resulting in meningocele or meningomyelocele

Meningocele

- Herniation of the meninges through the bony defect spina bifida without spinal cord and nerve roots in the dural sac.
- The sac covering may be thin and transluscent or membranous

Myelomeningocele

- Herniation of the meninges through the bony defect spina bifida with spinal cord and nerve roots in the dural sac.
- The lesion is poorly covered so there may be leakage of CSF.
- Most commonly occurs at lumbar or lumbosacral areas of the vertebrae

myelomeningocele

spina bifida occulta

- spina bifida occulta (L5 – S1)-
- midline defect of the vertebral bodies without protrusion of the spinal cord or meninges

signs & symptoms

- +/-vertebral dimple ,dermal sinus tract
- Dimples below the gluteal -benign, nonneurologic.
- A pit, tract) below the gluteal crease is often a pilonidal sinus and need no further evaluation. Above- investigate.
- Small tuft of hair/ lipoma / hemangioma, – lower lumber-sacral region
- neurologic abnormality in the thoracic, lumbar, or sacral region

may be present

Spina bifida occulta

- Orthopedic findings
  - Extremity asymmetry
  - Foot deformities
- Neurological problems
  - Weakness of leg or legs
  - Leg atrophy or asymmetry
  - Loss of sensation, painless sores
  - Hyperreflexia
  - Unusual back pain
  - Abnormal gait
  - Radiculopathy
- Urologic problems
  - Neurogenic bladder
  - Incontinence

Hydrocephalus

- Hydrocephalus is a hydrodynamic disorder of CSF caused by increased production, impaired absorption or blockage of flow of CSF resulting in excessive amount of CSF

Types

Communicating hydrocephalus

- Occurs when full communication occurs between the ventricles and subarachnoid space.
- The obstruction is outside the ventricular system.
- It is caused by overproduction of CSF (rarely), defective absorption of CSF (most often), or venous drainage insufficiency (occasionally).

Non communicating hydrocephalus (99% of cases)

- Occurs when CSF flow is obstructed within the ventricular system or in its outlets to the arachnoid space.
- Most often a result of congenital anomaly.

Causes

Congenital causes in infants and children

- Brainstem malformation causing stenosis of the aqueduct of Sylvius -10% of all cases of hydrocephalus in newborns.
- Dandy-Walker malformation -2-4% newborn hydrocephalus.
- Arnold-Chiari malformation type I & II
- Agenesis of the foramen of Monro
- Congenital toxoplasmosis
- Bickers-Adams syndrome: X-linked hydrocephalus accounting for 7% of cases in males. It is characterized by stenosis of the aqueduct of Sylvius, severe mental retardation, and in 50% by an adduction-flexion deformity of the thumb

Acquired causes in infants and children

- Mass lesions -20% of all cases of hydrocephalus in children. eg, medulloblastoma, astrocytoma, cysts, abscesses, or hematoma also can be the cause.
- Hemorrhage: Intraventricular hemorrhage can be related to prematurity, head injury, or rupture of a vascular malformation.
- Infections: Meningitis (especially bacterial) and, in some geographic areas, cysticercosis can cause hydrocephalus.
- Increased venous sinus pressure related to achondroplasia, some craniostenoses, or venous thrombosis.
- Iatrogenic: Hypervitaminosis A, by increasing secretion of CSF or by increasing permeability of the blood-brain barrier, can lead to hydrocephalus. As a caveat, hypervitaminosis A is a more common cause of idiopathic intracranial hypertension, a disorder with increased CSF pressure but small rather than large ventricles.
- Idiopathic

Treatment of hydrocephalus is mainly VP shunting

Clinical features of hydrocephalus are influenced by the following:

- Patient's age
- Cause
- Location of obstruction
- Duration
- Rapidity of onset

Symptoms in infants

history

- Poor feeding
- Irritability
- Reduced activity
- Vomiting

Physical

- Head enlargement:
- Dysjunction of sutures
- Dilated scalp veins
- Tense fontanelle
- Setting-sun eyes
- Positive macewen’s sign
- Increased limb tone -Spasticity caused by stretching of the periventricular pyramidal tract fibers

Symptoms in children

History

- Slowing of mental capacity
- Headaches initially in the morning because of skull rigidity
- Neck pain suggesting tonsillar herniation
- Vomiting, more significant in the morning
- Blurred vision due to papilledema and later of optic atrophy
- Double vision related to unilateral or bilateral sixth nerve palsy
- Stunted growth and sexual maturation leading to obesity precocious puberty or delayed onset of puberty.
- Difficulty in walking due to spasticity secondary to stretched periventricular pyramidal tract. Drowsiness

physical

- Papilledema: failure to treat raised IC- optic atrophy vision loss.
- Failure of upward gaze due to pressure on the tectal plate through the suprapineal recess. The limitation of upward gaze is of supranuclear origin. When the pressure is severe, other elements of the dorsal midbrain syndrome (ie, Parinaud syndrome) may be observed, such as light-near dissociation, convergence-retraction nystagmus, and eyelid retraction (Collier sign).
- Macewen sign: A "cracked pot" sound is noted on percussion of the head.
- Unsteady gait: This is related to spasticity in the lower extremities.
- Large head: Sutures are closed, but chronic increased ICP will lead to progressive macrocephaly.
- Unilateral or bilateral sixth nerve palsy is secondary to increased ICP.

management

Seizures

- Sudden, transient disturbance of brain function, manifested by involuntary motor, sensory, autonomic, or psychic phenomena, alone or in any combination, often accompanied by alteration or loss of consciousness
- It occurs as a result of spontaneous electrical discharge of hyper excited brain cells in epileptogenic focus.
- This may be triggered by multiple factors that cause disturbance to brain function.
- Recurrent seizures without evident cause is referred to as epilepsy.
- Causes of seizures include;

Epileptic seizures

- Idiopathic (70-80%)
- Secondary
  - Cerebral dysgenesis/malformation
  - Cerebral vascular occlusion
  - Cerebral damage, e.g. congenital infection, hypoxic-ischaemic encephalopathy, intraventricular haemorrhage/ischaemia
- Cerebral tumor
- Neurodegenerative disorders
- Neurocutaneous syndromes

- Non-epileptic seizures
  - Febrile convulsions
  - Metabolic
    - Hypoglycaemia
    - Hypocalcaemia/hypomagnesaemia
    - Hypo/hypernatraemia
  - Head trauma
  - Meningitis/encephalitis
  - Poisons/toxins
- The location of epileptogenic foci and number involved determines the nature of the seizure.
- If a small area of the brain is involved, a focal seizure may occur.
- If electrical discharge continues, a generalized seizure will occur. Generalized seizures also occur when epileptogenic foci is located in the brain stem, mid brain or reticular formation

Classification of seizures

- Generalized seizures of nonfocal origin
  - Tonic
  - Clonic
  - Tonic-clonic
  - Absence
  - Atonic
  - Akinetic
  - Bilateral epileptic myoclonus
- Partial (focal) seizures
  - Simple partial seizures with elementary symptomatology (consciousness is not impaired)
    - With motor symptoms (including Jacksonian, adversive, and postural)
    - With sensory symptoms (including visual, somatosensory, auditory, olfactory, gustatory, and vertiginous)
    - With autonomic symptoms
    - With psychic symptoms (including dysphasia and affective changes)
    - Compound (i.e., mixed) forms
  - Complex partial seizures with complex symptomatology (consciousness is impaired)
    - Simple partial seizure followed by loss of consciousness
    - With automatisms
- Unclassified seizures
    - Febrile seizures
    - Metabolic seizures
    - Seizures resulting from poison and toxins

Types of seizures in children

Generalized seizures; due to diffuse electrical activity throughout cortex and brain stem

- Tonic seizure: Unconsciousness, continuous muscle contraction, and sustained stiffness.
- Clonic seizure: Alternating muscle contract in a rhythmic repetitive jerking motion.
- Tonic-clonic (grand mal): Abnormal electrical activity occurs in both hemispheres of the brain and then moves from the cortex to the brainstem. It begins with the tonic phase followed by the clonic phase.
- Absence seizure (petit mal): Brief loss of consciousness lasting <30 seconds with no postseizure confusion and minimal or no loss of postural tone.
- Atonic seizure: Loss of posture tone causing a drop-and-fall action.
- Myoclonic seizure: Involuntary jerking and loss of body tone resulting in falling forward, often occurring when falling asleep or awakening. There is no loss of consciousness or postseizure confusion.
- Akinetic seizure: Brief loss of muscle tone and brief loss of consciousness.

Partial seizures; characterized by local motor, sensory, psychic and somatic manifestations

- Simple partial seizure: Abnormal electrical activity occurs in one hemisphere of the brain or from an area of the cerebral cortex. There is no loss of consciousness and lasts <30 seconds. Examles include jacksonian seizures and rolandic of sylvian seizures
- Complex partial seizures: Abnormal electrical activity occurs in one hemisphere of the brain or from an area of the cerebral cortex resulting in impaired consciousness lasting up to 5 minutes and postseizure confusion.

Others

- Febrile seizure: Rapid rise in body temperature of >102.2° F (39°C) in children 6 months to 6 years of age might result in tonic/clonic seizures lasting <15 minutes.
- Status epilepticus: Continuous seizure or a series of seizures lasting for >30 minutes during which there is a loss of consciousness. Postseizure period can last for up to 2 hours.
- Infantile spasms: Delays in neurologic development or neurologic abnormalities result in abrupt jerking and contracting of the head and neck that begin at 2 months of age and resolve by 2 years of age.

Management

- Dx goals
  - Ascertain child has a seizure disorder
  - Determine cause of seizure
  - Classify type of seizure
- Hx
  - Comprehensive medical history including past illness and hospitalization, medication history, toxic exposures and description of previous seizure episode. Family hx of seizure disorder is also important.
- PE; level of consciousness, reflexes, sensory and motor responses.
- Complete blood count; presence of infections such as meningitis, encephalitis, analysis of serum electrolytes to rule out metabolic disturbances.
- Radiological imaging; CT scan, MRI, PET; visualize structural abnormalities.
- EEG; highlights areas of abnormal brain electrical activity.
- Treatment
  - Pharmacological therapy will be guided by the classification of the seizure

Nursing management in seizure disorders

Objectives of care:

- Manage acute episode and prevent injury
- Determine and treat underlying cause of seizures if possible.
- Prevent recurrence of seizures and therefore allow patient to live a normal life.

- Place oxygen apparatus near and ensure it is in good working condition.
- Take the seizure prone patient's temperature with a rectal thermometer; prevents possibility of patient biting an oral thermometer if a seizure should occur.
- Set up suction equipment at the patient's bedside.
  - Check the equipment daily to be sure it is working properly.
  - Use during or after a seizure to clear the patient's airway.
- Nursing care during a seizure.
  - Turn patient on his side to provide for drainage of oral secretions.
  - Do not forcibly restrain patient during seizure.
  - Remove objects that may obstruct breathing or cause injury to patient.
  - Protect patient's head from injury with pillow, blanket, etc.

Nursing care following a seizure.

- Keep bed flat and patient turned on his side until he is alert.
- Room lighting should be dim and noise kept to a minimum.
- Loosen restrictive clothing (if not done during seizure).
- Check vital signs immediately following seizure and every 30 minutes (or as ordered) until patient is alert.
- Check lips, tongue, and inside of mouth for injuries.
- If patient is incontinent, change clothing and bedding with as little disturbance as possible.
- Document all precautions taken, all activity observed during a seizure, to include the time, location, circumstances, length of seizure activity, and vital signs. Also document any injury sustained during the seizure.

Discharge care

- Institute and reinforce the importance of anticonvulsant drug therapy:
  - Drug therapy is a means of controlling the condition; it is not a cure.
  - Initially, dosage will have to be monitored and altered to provide maximum control with minimum side effects.
- Instruct patient to keep record of events surrounding his/her seizures (number, duration, time, sleep/eating patterns).
  - Use of multidisciplinary approach to cope with social, emotional, and vocational pressures of the person with epilepsy.

Myasthenia gravis

- Chronic disease characterized by rapid fatigability of striated muscle.
- disorder of neuromuscular transmission that results from autoimmune attack on nicotinic postsynaptic receptors for acetylcholine (Ach).
- Release of ACh into the synaptic cleft by the axonal terminal is normal, but the postsynaptic muscle membrane or motor end plate is less responsive than normal.

Clinically divided into

- Neonatal (transient) myasthenia gravis
  - Occurs in 12% of infants born to myasthenic mothers.
  - The condition is due to maternal ACh rcptr antibody transferred across the placenta.
- Congenital (persistent) myasthenia gravis ; mothers of the affected infants rarely have myasthenia gravis, but other relatives may.
  - Congenital myasthenia gravis is not caused by receptor antibodies and often responds poorly to therapy.
  - It may result from a genetic abnormality of the acetylcholine receptor protein, postsynaptic membrane structure, or other myoneural transmission defects.
- Juvenile myasthenia
  - symptoms and signs are similar to those in adults.
  - Receptor antibodies are usually present.
  - Prominent signs are difficulty in chewing,dysphagia, a nasal voice, ptosis, and ophthalmoplegia, pathologic fatigability of limbs, chiefly involving the proximal limb and neck muscles.
  - Associated with autoimmune conditions e.g. thyroid disease.

- Dx
  - Adm of short-acting cholinesterase inhibitor (Neostigmine and Edrophonium); ptosis and ophthalmoplegia improve within a few seconds, and fatigability of other muscles decreases.
  - Serum ACh receptor antibodies are often found in the neonatal and juvenile forms.
  - EMG; Repetitive stimulation of a motor nerve at slow rates over the appropriate muscle reveals a progressive fall in amplitude of the
  - CXR and CT scan may disclose benign thymus enlargement.
- Clinical manifestation
  - Dysphagia and facial weakness in early infancy causing feeding difficulties
  - Ptosis and extraocular muscle weakness
  - Poor head control due to weakness of the neck flexors
  - Diminished tendon stretch reflexes
  - Rapid fatigue of muscles
  - If untreated, may progress to life threatening respiratory muscle involvement with risk of aspiration.

Management

- Myasthenic crisis;
  - Ach inhibitor (Edrophonium) results in dramatic but brief improvement.
  - Suctioning, tracheostomy, respiratory assistance,
  - Fluid and electrolyte maintenance.
- Cholinergic crisis; may result from over dosage of anticholinesterase drugs. The resulting weakness may be similar to that of myasthenia. Muscarinic effects are however absent.
- Immunologic intervention; achieved primarily with prednisone. Plasmapheresis is effective in removing acetylcholine receptor antibody in severely affected patients.
- Surgery; Early thymectomy is beneficial in many patients whose disease is not confined to ocular symptoms

Cerebral palsy

- Non progressive motor dysfunction caused by damage to motor areas of the brain resulting in partial paralysis and uncontrolled movement.
- In addition children with cerebral palsy often have learning difficulties, epilepsy, squints, visual impairment, hearing impairment, speech and language disorders, behavior disorders and feeding problems.
- Causes
  - Prenatal; genetic or chromosomal disorders, ineffective placenta, exposure to teratogens, multiple fetuses, intrauterine infections.
  - Birth; complicated labor and delivery, birth injury, asphyxia due to cord prolapse or strangulation
  - Perinatal; CNS infection, kernicterus
  - Childhood; head trauma, meningitis

Types of cerebral palsy

Spastic / (most common): damage to the upper motor neuron (pyramidal or corticospinal tract) pathway. Limb tone is increased (spasticity) and the child has a scissor-like gait where one foot crosses in front of the other foot.

- There are three main types of spastic cerebral palsy:
  - *Hemiplegia* - unilateral involvement of the arm and leg with the face spared.
  - *Quadriplegia* - all four limbs are affected. The arms may be affected more than the legs.
  - *Diplegia* - legs are affected to a much greater degree than the arms, so that hand function may appear to be relatively normal

Athetoid/ dystonic : The basal ganglia are affected resulting in uncoordinated involuntary motion.

Ataxic: The cerebellum is affected resulting in poor balance and difficult muscle coordination.

Clinical manifestation

- Poor sucking, difficulty feeding
- Facial grimacing and drooling
- Alteration in muscle tone such as abnormal posturing and movements.
- Involuntary muscular contraction and relaxation causing contractures
- Increased deep tendon reflexes
- Persistent primitive reflexes making achievement of milestones difficult
- Fine motor coordination may be affected, interfering with activities of daily living
- Poor vision, strabismus.
- Hearing loss
- Cognitive impairment
- Speech or language delays
- seizures

Dx

- EEG
- CT scan
- MRI
- electrolyte levels; rule out metabolic disorders

Management

- Medication
  - Anticonvulsants for epilepsy
  - If spasticity is severe and causing pain muscle relaxants are used
- Orthopedic surgery
  - Orthopedic deformities may develop as a result of longstanding muscle weakness or spasticity such as dislocation of the hips and fixed equinus deformity of the ankle as a result of muscle spasticity.
  - This is corrected surgically

Nutrition

- Undernutrition commonly occurs in children with cerebral palsy, and can reduce the chances of achieving physical and intellectual potential.
- Food must be given in a form appropriate to the child's ability to chew and swallow.
- Energy-rich supplements in adequate amounts is necessary.

Physiotherapy

- The role of the physiotherapist is crucial in the management of the child with cerebral palsy.
- Advises on handling and mobilization of child in daily activities
- Exercises to prevent the development of deforming contractures.

Occupational therapy

- Advise on special equipment such as wheelchairs and seating, and activities that encourage fine motor function.

GUILLAIN-BARRÉ SYNDROME

- Acute, progressive autoimmune condition that affects the peripheral nerves.
- Symptoms occur as the myelin surrounding the axon on the peripheral nerves is damaged from the autoimmune effect.
- The disease follows a viral infection (Epstein-Barr virus, coxsackievirus, echovirus, cytomegalovirus), surgery or other acute illness.
- Prominent cause of acute paralysis.
- In ascending syndrome, muscle weakness and/or paralysis begins in the distal lower extremities and travels upward. The reverse is true in descending syndrome
- Cranial nerve dysfunction leads to facial weakness, weak nasal speech, and dysphagia.
- Respiratory compromise owing to intercostal muscle weakness poses the major threat, and ventillatory assistance is required

- Patient may experience altered sensory perception in the same areas, such as the sensation of crawling, tingling, burning, or pain.
- The progression of symptoms may take hours or days.

Signs and Symptoms

- Burning or pickling feeling due to demyelination of the nerve axons.
- Symmetric weakness or flaccid paralysis ascending or descending in pattern.
- Absence of deep tendon reflexes due to changes within the nerves, absent babinski reflex
- Recent infection or other acute illness.
- Facial weakness, dysphagia, visual changes in descending disease.
- Labile blood pressure and cardiac dysrrhythmias due to autonomic nervous system response.
- Weak cough and shallow respirations indicate intercostal muscle involvement.
- Urinary incontinence or retention

Dx

- LP; elevated protein in CSF
- Nerve conduction shows slowed velocity.
- Pulmonary function tests show diminished tidal volume and vital capacity

Treatment

- Monitor respirations and support ventilation if necessary.
- Plasmapheresis; remove the antibodies in the circulation.
- Administer immunoglobulin IV after drawing labs for serum immunoglobulin A.
- Nasogastric tube feeding if swallowing is a problem.