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Textbook of

Pediatric Nursing

As per the Revised Indian Nursing Council Syllabus (2021-22)

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Neurological Disorders

Chapter Outline

- Introduction
- Meningitis
- Seizure Disorders
- Cerebral Palsy
- Reye's Syndrome
- Encephalitis
- Traumatic Brain Injury
- Hydrocephalus
- Spina Bifida
- Neuroblastoma
- Brain Tumors

INTRODUCTION

Disorders of central nervous system (CNS) are common in children, accounts for 20–30% of childhood illnesses. Early diagnosis of the CNS disease conditions helps in reducing the morbidity and mortality among the children. This chapter explores various nervous system disorders like Meningitis, Convulsive disorders, Cerebral palsy, Reye syndrome, Encephalitis, and Head Injury.

MENINGITIS

Meningitis is a common CNS infection causing inflammation of meninges. Various microorganisms like bacteria, virus, fungus and parasites can cause meningitis. The disease is influenced by the age of the child, his immune status and the epidemiology of the pathogens. Regardless of the causative organisms, most of the clinical manifestations of meningitis are same.

Related Anatomy and Physiology

Brain and the spinal cord are surrounded by meninges. There are three layers of meninges namely dura mater, arachnoid mater and pia mater from outside to inside (Fig. 20.1).

The cerebrospinal fluid (CSF) is located in a space between arachnoid and pia mater called the subarachnoid space. The meninges protect the CNS.

The dura mater is the outermost covering which is a thick, durable membrane closest to the vertebrae and skull. The space

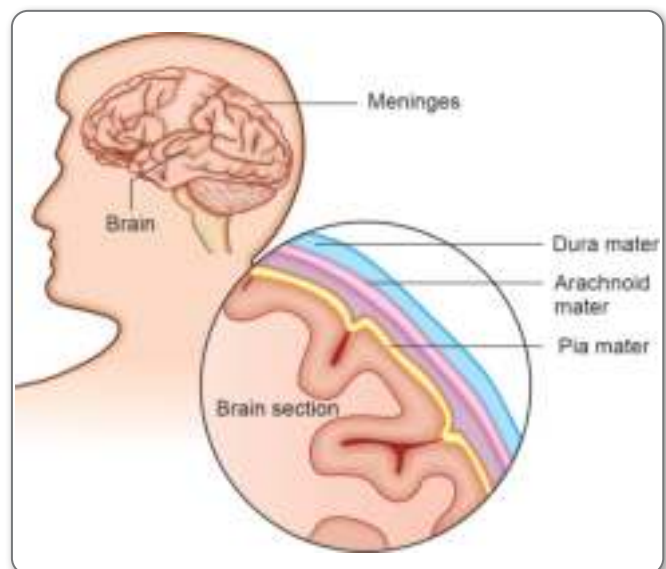


Figure 20.1: Meninges covering the brain and spinal cord

between the skull and dura mater is called epidural space that contains blood vessels and fat. The space between the dura mater and arachnoid space is subdural space, while the space between the arachnoid and pia mater is called subarachnoid space.

Types of Meningitis

Meningitis, depending upon the causative organism, could be classified as:

- Bacterial,
- Viral,
- Fungal and
- Parasitic meningitis.

The bacterial meningitis is the most common one. The bacterial meningitis along with causative organisms are described in Table 20.1. Children with compromised immune status like malignancies, on immunosuppressant drugs are susceptible to meningitis due to fungi, *Listeria* and *Mycoplasma*. Tubercular meningitis (TBM) is the extension of childhood tuberculosis, spreads through hematogenous route.

Pathogenesis

Meningitis spreads mainly through hematogenous route to the meninges either by sepsis or distant foci, for example, through empyema, pneumonia, pyoderma, osteomyelitis, etc. Head injury can also lead to meningitis.

In meningitis, *leptomeninges* (arachnoid and pia mater together) get infiltrated with inflammatory cells. As a result of which the cortex of the brain shows edema, exudate and proliferation of microglia. The ependymal cells are destroyed and the purulent exudate collects at the base of the brain. The subarachnoid space as a result of this inflammatory change gets filled with opaque or cloudy fluid. The exudates may further block the foramina of Luschka and Magendie causing blockage in CSF circulation, leading to the development of hydrocephalus. Children with meningitis may develop neurological sequelae due to infarcts developed as a result of thrombophlebitis of cerebral vessels.

Table 20.1: Causes of bacterial meningitis

Age of onset	Microorganisms
Up to 3 months	<i>Escherichia coli</i> , <i>streptococcus pneumoniae</i> , <i>salmonella species</i> , <i>pseudomonas aeruginosa</i> , <i>Streptococcus faecalis</i> , <i>Staphylococcus aureus</i>
3 months to 3 years	<i>Hemophilus influenzae</i> , <i>S. pneumoniae</i> and <i>neisseria meningitidis</i>
Beyond 3 years	<i>S. pneumoniae</i> and <i>neisseria meningitidis</i>

Table 20.2: Symptoms of bacterial meningitis in neonates, infants and children

Symptoms in neonates	Symptoms in infants and children
<ul style="list-style-type: none"> • Poor feeding, lethargy • Irritability, apnea • Listlessness, apathy • Fever, hypothermia • Seizures • Jaundice • Bulging fontanel • Pallor, shock • Hypotonia • Shriill cry • Hypoglycemia, intractable metabolic acidosis 	<ul style="list-style-type: none"> • Nuchal rigidity (inability to flex the neck forward due to rigidity of the neck muscles) • Opisthotonus (spasm of the muscles causing backward arching of the head, neck, and spine) • Bulging fontanel • Convulsions • Photophobia • Headache • Alterations of the sensorium • Irritability, lethargy • Anorexia, nausea, vomiting • Coma, fever (generally present, although some severely ill children present with hypothermia)

In meningococcal meningitis, the disease course may become fulminating leading to death due to development of endotoxic shock. TBM may occur as part of generalized miliary TB with tubercles in the choroid plexus directly infecting the meninges.

Clinical Features

The three classic symptoms are fever, headache and meningeal signs. Symptoms in neonates and children are summarized in Table 20.2.

The onset of meningitis is acute with fever. The child may appear irritable, cry excessively with nausea and projectile vomiting. Older child may complain of severe headache, resent light and has photophobia. Seizure, a common symptom may be seen at the onset of illness or during the illness. Child may also have altered sensorium. On examination hypertonia, marked neck rigidity is observed. Flexion of the neck is very painful with limited movements. Kernig's and Brudzinski's signs are present. In Kernig's sign, extension of the knee is limited to less than 135° (Fig. 20.2) when child lies flat on back, while in Brudzinski sign, as the neck of the child is flexed, the knees of the child get flexed. There may be hyperarching of back, i.e., opisthotonos (Fig. 20.3).

Meningitis in a neonate and in first 4–6 months of age can present with atypical features such as vacant stare, sepsis, irritability and drowsiness, feed refusal, poor tone, shock and fever.

In meningococcal meningitis along with typical features, child can have petechial hemorrhage. It may be associated with acute fulminant illness, **adrenal insufficiency and death.**

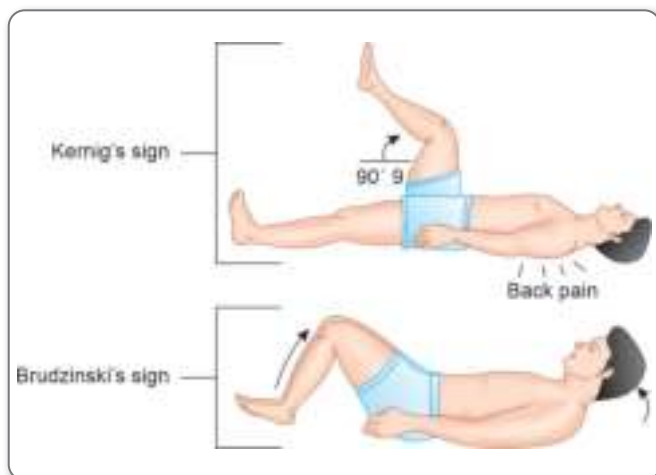


Figure 20.2: Kernig's and Brudzinkski's sign



Figure 20.3: Opisthotonus

The clinical course of TBM is described in three stages namely prodromal stage, stage of meningitis and stage of coma (Table 20.3).

Diagnosis

There is history of fever, irritability, photophobia, headache, convulsions and altered level of consciousness. Diagnosis is confirmed by CSF obtained by lumbar puncture (LP). It shows turbid CSF, with increased cell count ($>1000/\text{cu mm}$) with mostly polymorphonuclear cells, raised protein count ($>100 \text{ mg/dL}$) and sugar (below 50% of the blood sugar level). Gram staining of CSF helps in identifying the microorganisms.

CT scan is not required generally, but can be helpful to rule out subdural effusion, brain abscess, hydrocephalus and tubercular meningitis.

Table 20.3: Clinical stages of tubercular meningitis

Prodromal stage	<ul style="list-style-type: none"> Onset of disease is gradual with low grade fever, loss of weight and appetite. Child may appear irritable, lethargic and restless Vomiting, photophobia, headache
Meningitis stage	<ul style="list-style-type: none"> Appearance of typical features of meningitis Neck rigidity Kernig's sign Rise in body temperature to 39°C Slow heart rate Increased muscle tone
Coma stage	Loss of consciousness Altered respiratory pattern Ptosis, dilated unequal pupils

Serological tests such as ELISA, PCR and HIV may be helpful in suspected cases of TBM.

Treatment

Treatment of meningitis includes mainly antibiotics, corticosteroids and supportive therapy.

- Antibiotics:** Antibiotics are administered for 10–14 days period. Third generation cephalosporin drugs like ceftriaxone ($150\text{--}200 \text{ mg/kg/day}$, or cefotaxime ($100\text{--}150 \text{ mg/kg/day}$) is prescribed. A combination of ampicillin (200 mg/kg/day) and chloramphenicol (100 mg/kg/day) for 14 days is the second empiric therapy. If fever or the meningeal signs (neck rigidity, Kernig's or Brudzinkski's signs) are still present, CSF needs to be re-examined along with review of antibiotics. There are some specific antibiotics depending upon the type of meningitis described in Table 20.4.
- Corticosteroids:** Dexamethasone in a dose of 0.15 mg/kg IV every 6 hourly for first four days is highly beneficial. Corticosteroids are administered 15 minutes prior to antibiotics, they help in reducing the incidence of residual neurological sequelae such as sensorineural deafness, hydrocephalus and behavioral changes.
- Supportive therapy:** Children with meningitis require supportive therapy in the form of osmotic diuretics, anticonvulsant drugs, fluid and electrolyte balance and management of hypotension.
 - Osmotic diuretics:** One of the clinical presentations of the child with meningitis is related to rise in intracranial pressure (ICP). Injection mannitol (20%) 0.5 mg/kg is administered in 6 doses, every 4–6 hours apart.
 - Anti-convulsant drugs:** Injection phenytoin $10\text{--}15 \text{ mg/kg}$ as loading dose followed by maintenance dose (5 mg/kg) is given.
 - Fluid and electrolyte balance:** Fluids are given as $2/3\text{rd}$ of maintenance dose to reduce the cerebral edema and manage syndrome of inappropriate antidiuretic hormone (SIADH).

Table 20.4: Specific antibiotic therapy

Sl. no.	Type of meningitis	Antibiotics
1.	Meningococcal or pneumococcal	Penicillin G (4–5 lacs/kg/day), Cefotaxime (150–200 mg/kg/day) Ceftriaxone 100–150 mg/kg/day
2.	H. influenzae	Cefotaxime (150–200 mg/kg/day) Ceftriaxone 100–150 mg/kg/day Ampicillin (200 mg/kg/day) and Chloramphenicol (100 mg/kg/day)
3.	Staphylococcal	Vancomycin, if methicillin or penicillin resistance is suspected. Additional rifampicin to the regimen to increase the CSF penetration
4.	Listeria	Ampicillin (300 mg/kg/day) along with aminoglycosides (gentamycin, amikacin or netilmicin)
5.	Gram negative rods	Cefotaxime (150–200 mg/kg/day) Ceftriaxone 100–150 mg/kg/day Ampicillin (300 mg/kg/day) along with aminoglycosides (gentamycin, amikacin or netilmicin)
6.	Pseudomonas	Ceftazidime with aminoglycosides

- **Treatment of hypotension:** If child develops hypotension, it is corrected using fluid therapy and vasopressors such as dopamine and dobutamine.

In tubercular meningitis, anti-tubercular therapy (ATT) is given for a period of at least 12 months. A combination of the following 4 drugs INH (5 mg/kg, maximum 300 mg), rifampicin 10 mg/kg (maximum 600 mg) orally early morning empty stomach, ethambutol (15–20 mg/kg/day), pyrazinamide (30 mg/kg/day). Streptomycin in a dose of 30–40 mg/kg/day may be given for a period of 2–3 weeks. Other drugs given to treat TBM are steroids (Dexamethasone 0.15 mg/kg every 6 hours IV followed by oral prednisolone).

Complications

Post treatment complications are subdural empyema and hydrocephalus due to the collection of exudates, vascular insults, and release of endotoxins. The subdural empyema is treated with antibiotics and hydrocephalus may require placement of ventriculo-peritoneal shunt.

Follow-up

Children with bacterial meningitis are regularly followed-up for early detection management and rehabilitation of residual neurological handicaps.

Nursing Management

- **Ineffective cerebral tissue perfusion related to increased intracranial pressure (ICP), cerebral edema as evidenced by delirium, hallucinations, drowsiness, hypercapnia**
 - Monitor vital signs and neurological status. Increasing systolic blood pressure accompanied by decreasing diastolic blood pressure is a sign of increased ICP.
 - Observe for any signs of increased ICP, headache, drowsiness, decreased alertness, vomiting, bulging fontanel (infants).
 - Assess for signs of meningeal irritation, which may happen because of infection, i.e., nuchal rigidity, twitching, increased restlessness, and irritability.
 - Observe for increasing restlessness, moaning, and guarding behaviors which may indicate increased ICP or pain.
 - Monitor ABGs and oxygen saturation.
 - Maintain head or neck in midline position, provide small pillow for support. Turning head to one side compresses the jugular veins and inhibits venous drainage, thereby increasing ICP.
 - Provide comfort measures and decrease external stimuli. Provide quiet environment, use soft voice, and gentle touch.
 - Administer oxygen as needed.
 - Administer medications: Osmotic diuretic: Mannitol to treat cerebral edema by promoting cerebral blood flow, Anticonvulsants: Diazepam or phenytoin.
- **Hyperthermia related to infection as evidenced by increased body temperature, hot, flushed skin, increased heart rate.**
 - Assess the child's vital signs closely.
 - Assess for signs of dehydration such as dry mouth, sunken eyes, sunken fontanel, and low concentrated urine output.
 - Reduce temperature by performing tepid sponge, it decreases temperature by liberating heat by conduction and convection. Elevated body temperature increases the metabolic rate, hence, increases the insensible fluid loss. Administer antipyretics as indicated.
 - Maintain adequate fluid intake as tolerated.
 - Administer antibiotics as indicated
- **Acute pain related to increased ICP, meningeal irritation as evidenced by neck stiffness, headache, irritability, nuchal rigidity**
 - Assess for headache and photophobia.
 - Assess for Kernig's sign and Brudzinski's sign.
 - Maintain a quiet environment and keep child's room darkened.
 - Prevent stimulation and restrict visitors.
 - Assist range of motion (ROM) exercises to prevent joint stiffness and neck pain.
 - Administer antibiotics and corticosteroids to reduce the inflammation and pain.



- Administer analgesics such as acetaminophen or NSAIDs.
- **Disturbed sensory perception r/t decreased LOC, cerebral edema as evidenced by altered sensorium**
 - Assess level of consciousness using pediatric Glasgow Coma Scale.
 - Observe and notify physician for persistent deterioration in LOC.
 - Change in mentation, seizures, increased BP, bradycardia, or respiratory abnormalities may indicate increasing ICP with decreased cerebral perfusion pressure.
 - Assess for signs of cerebral edema such as dizziness, headache, irregular breathing, neck pain, nausea or vomiting.
 - Assess cognitive function by checking ability to follow simple or complex commands.
 - Evaluate presence or absence of protective reflexes: Swallow, gag, blink, and cough. Absence of reflexes is a late sign indicative of increasing ICP.
 - Assess for signs of meningeal irritation: Headache, photophobia, nuchal rigidity, opisthotonic position, Kernig's sign, Brudzink's sign.
 - Elevate head of bed up to 30°–45° with the client's head in neutral position.
 - Reorient the client to the environment, as needed.
 - Assist with diagnostic testing: EEG, Lumbar puncture for CSF, MRI, CT Scan, or ventriculogram
 - Initiate seizure precautions: Observe and provide care during seizure. Maintain a quiet environment and keep the lights dim. Observe and document pattern and frequency of seizure. Notify physician of seizure activity. Administer and monitor anticonvulsants drug levels.
 - Assess pupil size every 3 hours during the first 24 hours and consequently every 6 hours. Increased ICP will result in uneven pupil sizes, fixed dilated pupil.
- **Anxiety r/t disease process and outcome**
 - Teach about disease process and behaviors, physical effects and symptoms of disease.
 - Clarify any misinformation and answer questions in lay terms
 - Encourage the parent to stay with the child
- **Deficient knowledge r/t treatment**
 - Provide information and explanations in clear language that is understandable
 - Teach about administration of medications
 - Assist to plan feedings
 - Teach to isolate other children in the family for 24 hours if respiratory infection present or until the culture is negative.
- **Risk for injury r/t seizure**
 - Assess neurologic status, changes in consciousness, behavior patterns and pupillary/ocular responses appropriate for age

- Measure head circumference in infant.
- Attach cardiac and respiratory monitor to assess for bradycardia and hypoxia.
- Note any seizure activity including onset, frequency, duration and type of movements before, during, or after seizure; pad bed and remove objects/toys from bed and administer any ordered anticonvulsants.
- Keep head in neutral position
- Administer antibiotics
- Administer stool softeners

SEIZURE DISORDERS

Definition

A seizure occurs when one or more parts of the brain has a burst of abnormal electrical signals that interrupt normal brain signals. There is uncontrolled firing of nerves in the brain.

Epilepsy is recurring seizures. **Convulsion** is muscle contraction and relaxation during seizure.

Convulsive disorders can be seen in different age groups. About 5% of convulsions are seen in first 5 years of life. The incidence of neonatal seizures accounts for 1–2% to 20% in preterm neonates. The reason for neonatal seizures is mainly the poor myelination and incomplete arborization of dendrites. The seizures in neonates can be subtle, focal clonic, multifocal clonic, generalized tonic or myoclonic. Subtle seizures are manifested as up-rolling of eyeballs, sucking movements, twitching of limb, and conjugate deviation of eyes. The main causes of neonatal seizures are listed in Table 20.5.

There are many disorders in different age groups that may mimic seizures in neonatal period like jitteriness, benign neonatal sleep myoclonus, apnea; in infants breath holding spells; childhood and adolescents can have syncopal attack, night terrors, cardiac arrhythmias, complicated migraine, hyperventilation attack, pseudo-seizures and tics.

Causes of Seizures

There are different causes of seizures according to different age groups described in Table 20.5.

Classification of Seizures

Seizures can be classified depending upon the type of abnormal activity seen in child.

The two main categories of epileptic seizures are focal (partial) seizure and generalized seizure.

Focal seizures take place when abnormal electrical brain function occurs in one or more areas of one side of the brain.

1. Focal Seizures

Focal seizures are also called partial seizures that account for 60% of childhood seizures. Some of the common causes

Table 20.5: Causes of seizures in children in different age groups

Age group	Causes
Neonatal period	Birth asphyxia, intra-ventricular and intra-cerebral hemorrhage, hypoglycemia, hypocalcemia, dysnatremia, inborn error of metabolism (phenylketonuria, maple syrup urine syndrome, galactosemia), pyridoxine dependency, maternal withdrawal of medications, injection of local anesthetic agents on the fetal scalp
One month to 3 years	Febrile convulsions Metabolic causes (hypocalcemia, hypomagnesia, inborn error of metabolism) Neurological infections (bacterial meningitis, tubercular meningitis encephalitis, cerebral malaria) Space occupying lesions (neoplasm, brain abscess, tuberculoma, cysticercosis) Post infection or vaccinal (mumps or measles infection or post pertussis vaccination) Drugs or poisons (phenothiazines, phenytoin, carbon monoxide, lead poisoning)

of seizures are birth asphyxia, vascular insults, head trauma, neoplasm and inflammatory granulomas. They can be simple partial, complex partial and partial seizures with secondary generalization.

- **Simple partial seizures:** The symptoms can be motor or sensory depending upon the focus. Children may complain of pain, tingling sensation, sometimes visual/olfactory/auditory or taste hallucinations. Focal seizures that spread from one part to another part of body are called *Jacksonian march*.
- **Complex partial seizures:** Generally originate from parietal or temporal lobe. They are characterized by automatisms or loss of consciousness. Abnormal movements like lip smacking, chewing, and fidgetiness may be observed. Complex partial seizures can be seen with secondary generalization. Tuberculoma and neurocysticercosis are the important disease conditions in which complex partial seizures are seen.

2. Generalized Seizures

A generalized seizure occurs in both sides of the brain. Types of Generalized seizures:

- **Absent seizures:** Absent seizures are less common than GTC seizures, and are seen in childhood between 6–8 years of age. A typical absent seizure is not preceded by an aura. This seizure lasts for less than 30 seconds, characterized by abrupt lapse of consciousness. Child may have staring spells, eye fluttering or rhythmic movements.
- **Atonic Seizures:** Atonic seizures are also called akinetic seizures or drop attacks. They are brief and child gains consciousness by the time he strikes the floor.

- **Generalized Tonic Clonic Seizures (GTCS):** Generalized tonic clonic seizures are the classic form of childhood epilepsy, having four phases namely (1) aura, (2) tonic (3) clonic (4) postictal phase.

1. Aura is a premonitory symptom that occurs before the onset of seizure. It can be sensory, motor, visceral or autonomic. Only one third of children can describe aura. Aura is followed by tonic phase, in which the skeletal muscles undergo spasm.
2. In tonic phase, muscular rigidity is the typical feature of antigravity muscles (flexors of the arms and extensors of legs). The duration of tonic phase is about 30–35 seconds. The child loses his consciousness and falls on the floor. At the time of seizure, face looks pale, eyes are rolled up, and pupils are dilated. Frothing is seen from the mouth and child may involuntarily pass urine and feces.
3. This is followed by clonic phase, in which rhythmic alternating contractions of muscles are observed for few minutes.
4. In last stage, i.e., postictal phase child looks confused, complains of headache and performs automatic actions.

- **Myoclonic seizures:** Neurological disorders like west syndrome also called infantile spasms and Lennox Gastaut syndrome are characterized by salaam spells and mental retardation (MR).

Febrile Convulsions

Febrile convulsions are seen in approximately 3–5% of children in children below five years of age (6 months – 5 years). They are generally genetically determined, self-limiting, occur during fever, in the absence of CNS infections. They are called simple, atypical, and benign seizures, usually occur as single seizure within 24 hours of the onset of fever, and last for less than 10 minutes. Majority of febrile convulsions are generalized without any postictal stage.

Febrile convulsions are managed by administration of antipyretic drug and/or hydrotherapy. Seizure is controlled by intravenous administration of Diazepam or Midazolam (0.2–0.3 mg/kg/dose). Children with recurrent febrile convulsions can be treated with Sodium Valproate (10–20 mg/kg/day) or Phenobarbitone (3–5 mg/kg/day).

Parents can be taught to abort the febrile convulsions by administering nasal/buccal midazolam (0.05–0.2 mg/kg) or rectal diazepam (0.5 mg/kg) as part of domiciliary treatment of convulsions. Feeding tube of 8F size, lubricated with paraffin or xylocaine jelly is used (approximately 4 cm) for per-rectal administration of diazepam, followed by flush with tap water.

Epilepsy

Epilepsy is a chronic disorder characterized by two or more seizures that are unprovoked occurring more than 24 hours



apart. Epilepsy is episodic, paroxysmal involuntary activity associated with abnormal electrical activity of the neurons. Approximately 5% of children are estimated to be affected with epilepsy; may present with sensory/motor/psychomotor phenomenon with altered sensorium.

Causes of Epilepsy

Main causes of epilepsy are as follows:

- **CNS infections:** Encephalitis, meningitis and neurocysticercosis
- Vascular malformations
- **Injury to the brain:** At the time of birth, or afterwards a traumatic injury
- Metabolic disorders

Clinical Features

Epilepsy, for the purpose of description is described as generalized or partial. The generalized seizures may be tonic/clonic or tonic-clonic, absent (petitmal)/atonic/myoclonic.

Diagnosis

- Diagnosis is established by careful history taking. Ask parents or eye witness to describe the event, progression of clinical condition and the duration of the event including postictal stage.
- Look for evidence of tongue bite, incontinence to confirm the episode of seizure.
- If child is already on anti-epileptic drugs (AED), enquire about compliance of child with prescribed drug therapy.

Investigations

- **Biochemical tests:** The tests include estimation of glucose, calcium, amino acids, blood ammonia, and blood and CSF lactate/pyruvate levels.

- **Electroencephalography (EEG):** This is done to find out the type of seizures, to locate epileptic focus and determine the anatomic basis.
- **Cranial imaging:** Magnetic resonance imaging (MRI) and CT scan are required to diagnose partial seizures. In case of focal EEG, physician may order for CT scan or MRI.
- Lumbar puncture is required to diagnose meningitis

Medical Management

The goals of management of epilepsy is fourfold.

- To ensure adequate vital signs and oxygenation both systemic and cerebral.
- To terminate seizure activity
- To prevent recurrence of seizures
- To establish the diagnosis and treatment of underlying disorder

Domiciliary or prehospital treatment is advised for all children with recurrent prolonged seizures as described in febrile convulsions section.

Hospital treatment is recommended for children who are convulsing actively in the emergency department of the hospital. Intravenous line is established. The routinely recommended drugs to control seizure activity are benzodiazepines (lorazepam/diazepam/midazolam).

Manage airway, breathing and circulation appropriately depending upon the clinical condition of the child.

Monitor vital signs immediately after the seizures.

Protect child from sustaining any injury or accident during seizures.

Anticonvulsant Therapy

Commonly used anticonvulsant drugs are described in Table 20.6.

Table 20.6: Anticonvulsant drugs

Sl. no.	Name of drug	Initial dose (mg/kg)	Route	Rate of infusion
1.	Diazepam	0.1–0.3 0.2–0.5	IV Rectal	1 mg/min, followed by loading dose of phenytoin
2.	Lorazepam	0.05–0.1 0.1–0.4	IV Rectal	1 mg/min (has longer duration of action, less likely to cause respiratory depression in comparison to diazepam)
3.	Midazolam	0.05–0.2 0.1–0.2	IV IM, buccal, nasal	1–18 µg/kg/min
4.	Phenytoin	15–20	IV	0.5–1 mg/kg/min, diluted in normal saline. Monitor child for hypotension and dysrhythmias
5.	Valproic acid	20	IV, rectal	Indicated in status epilepticus, dilute with equal amount of sterile water
6.	Phenobarbitone	10–20	IV	1–2 mg/kg/min, monitor child for respiratory depression, and hypotension
7.	Paraldehyde	0.15 mL/kg 0.3 mL/kg	IM Rectal	Use glass syringe Dilute with olive oil/coconut oil in 1:3 ratio

Table 20.7: First line anti-epileptic drugs

Medication	Indication	Dose (mg/kg/ day)	Side effects
Carbamazepine	Partial, tonic clonic, atonic and akinetic	10–30	Hepatitis, GI symptoms, rashes, bone marrow depression
Phenytoin	Tonic clonic, atonic and akinetic	5–10, 1–2 doses	Gingival hyperplasia, nystagmus, diplopia, hirsutism, megaloblastic anemia
Sodium valproate	Broad spectrum	20–60, 2–3 doses	Nausea, sedation. Weight gain, hair loss
Phenobarbitone	Tonic clonic, febrile and neonatal seizures	5–10, single dose	Drowsiness, drug dependency, hyperkinesia
Ethosuximide	Absence seizures	20–25, 1–2 doses	Nausea, drowsiness, photophobia, leucopenia, rarely blood dyscrasias
Clonazepam	Atonic, akinetic, resistant absence seizures	0.02–0.2, 2–3 doses	Fatigue, excessive secretions, hypotonia, somnolence
Levetiracetam	Partial, generalized seizures, myoclonus, photosensitive epilepsy	10–60, 2 doses	Drowsiness, behavioral changes
Clobazam	Partial generalized epilepsy	0.3–2.0, 1–2 doses	Sedation, ataxia, drooling, hyperactivity

Medications Used for the Treatment of Epilepsy

Epilepsy is a chronic disease that causes emotional and psycho-social disturbances to the child and his family. Anti-epileptic drugs (AED) used for the treatment of epilepsy is very carefully planned by the physician, keeping in mind the associated side effects of AED, due to the prolonged therapy. Mono-therapy is preferred over poly drug therapy due to difficulty in monitoring the drug compliance and side effects. If the monotherapy fails to control seizures, then only addition of second drug is considered.

The first line of AED includes phenytoin, phenobarbitone, sodium valproate and carbamazepine. Drugs, their doses, indications and side effects are described in Table 20.7.

Carbamazepine is a drug used for the treatment of partial and GTC seizures. One of the advantages of the drug is that it has fewer side effects. Phenobarbitone is a drug used to control seizures in neonates. Phenytoin is used as an initial choice, if there are any financial constraints. There are some newer AEDs such as lamotrigine, topiramate, vigabatrine, gabapentine, zonisamide, oxcarbazine, etc. Epilepsy not responding to medical therapy may be considered for surgical management in which surgical resection of the lesions is done.

Duration of AED

Duration of AED depends upon the child's response to therapy. Drug withdrawal is attempted slowly over a period of 3 months or so, usually 1–2 years in absence attacks and 2 years seizure free period in case of T-C seizures. Partial complex seizures are difficult to manage. Approximately 10–15% of children may show relapse following an adequate course of AED.

Status Epilepticus and Refractory Status Epilepticus

Any child who is brought in a prolonged convulsing state (single or multiple seizures) for a duration of more than

30 minutes, without regain of consciousness is considered to have status epilepticus. Refractory status epilepticus is when the child's seizures are not controlled with two or more antiepileptic drugs (AED).

Management of Status Epilepticus

- Attend to the ABCs before starting any pharmacologic intervention
- Place patient in the lateral decubitus position to avoid aspiration of vomiting
- Make further adjustments of the head and neck if necessary to improve airway patency
- Immobilize the cervical spine if trauma is suspected
- Administer 100% oxygen by facemask
- Assist ventilation and use artificial airways (e.g., endotracheal intubation) as needed
- Suction secretions and decompress the stomach with a nasogastric tube.
- Monitor vital signs, including blood pressure
- Monitor the patient's temperature, as hyperthermia may worsen brain damage
- In the first 5 minutes of seizure activity, before starting any medications, establish IV access and to obtain samples for laboratory tests and treatment.
- Infuse isotonic IV fluids plus glucose at a rate of 20 mL/kg/hr (e.g., 200 mL D5NS over 1 hour for a 10 kg child).
- In children younger than 6 years, use intraosseous (IO) infusion if IV access cannot be established within 5–10 minutes.
- If serum glucose is low or cannot be measured, give children 2 mL/kg of 25% glucose
- If the seizure fails to stop within 4–5 minutes, prompt administration of anticonvulsants may be indicated.

Anticonvulsant selection can be based on seizure duration, as follows:



- **6–15 minutes:** Lorazepam (0.05–0.1 mg/kg IV or IO slowly infused over 2–5 minutes); or diazepam per rectum at 0.5 mg/kg, not to exceed 10 mg
- **16–35 minutes:** Phenytoin (Dilantin) or fosphenytoin, not to exceed infusion rate of 1 mg/kg/min; do not dilute in D5W; if unsuccessful, phenobarbital 10–20 mg/kg IV (not to exceed 700 mg IV); increase infusion rate by 100 mg/min; phenobarbital may be used in infants before phenytoin
- **45–60 minutes:** Pentobarbital anesthesia (patient already intubated); or midazolam, loading dose 0.1–0.3 mg/kg IV followed by continuous IV infusion at a rate of 0.1–0.3 mg/kg/hr

Nursing Management

One of the important nursing responsibilities of a nurse is to monitor a child with seizure disorder during hospitalization. Equally important is to educate the parents about the seizure disorders, drug compliance and the management of seizures at home.

Nursing Diagnosis

- Risk for aspiration
- Risk for injury related to CNS dysfunction
- Knowledge deficit of parents related to disease condition
- Anxiety and fear of parents related to child having life threatening disease condition

Nursing Interventions

At the time of seizures:

- Remain calm and composed
- Note down the time of seizure
- Loosen restrictive clothing
- Remove eye glasses and loose dentures if any
- Clear area of any hazards
- Allow seizures to end without any interference
- If child vomits during seizures, provide side-lying position.
- Avoid restraining the child
- Don't put anything in the mouth of the child
- Don't offer any food or liquid at the time of seizures

After the seizure:

- Check for airway and clear it if required by suctioning or removing visible food particles.
- Assess breathing and provide oxygen if required depending upon the clinical condition of the child
- Provide side lying/recovery position
- Record the period of postictal stage
- Don't offer water or any food at this time
- Remain with the child.
- Make arrangement for transferring the child to the hospital.
- Administer anti-epileptic drugs (AED) as prescribed.
- Ensure adequate safety measures to prevent injury or fall from bed.
- Educate parents about the importance of drug compliance and regular follow-up.

- Provide parental counseling using anticipatory guidance to prevent injury during seizures.
- Encourage family members to involve in daily care of the child
- Hold discussions with parents to address their concerns related to seizures to alleviate their anxiety.

CEREBRAL PALSY

Definition

Cerebral palsy (CP) is a non-progressive neuromuscular disorder having cerebral origin. The severity of CP may range from minor state to total handicap state. Children affected with CP have multiple neurological deficits and varying degree of mental handicap.

Pathogenesis

Factors responsible for the development of CP can be categorized as prenatal, intra-natal and postnatal. Maldevelopment of brain, perinatal hypoxia, birth trauma, chorio-amnionitis, prothrombotic factors, metabolic disturbances, acid base imbalance, hyperbilirubinemia, intra-uterine or acquired infections are the common etiological factors responsible for the development of CP in which pathological lesions such as cerebral atrophy, migration defects, degeneration of basal ganglia and cerebral lesions are observed.

Classification of Cerebral Palsy

Depending upon severity of disease, CP can be classified as mild (20%), moderate (50%) and severe type (remaining 30%), while based on neurological findings CP can be classified as Spastic CP, Hypotonic (atonic) CP, Extrapyramidal CP and Mixed type.

- **Spastic (pyramidal) CP:** The most common form of CP is spastic type. This accounts for 65% of cases. The child can have spastic quadriparesis, diplegia or hemiparesis. There are abnormally persistent neonatal reflexes seen along with feeding difficulties. They also have abnormal stretch tendon reflexes. Affected children with spastic CP may have mental, behavioral or visual handicaps along with seizures.
- **Hypotonic CP:** Children with hypotonic CP are either hypotonic or atonic with hyperreflexia and persistent primitive reflexes.
- **Extrapyramidal or dyskinetic CP:** Extrapyramidal CP accounts for 30% of CP cases. Children with extrapyramidal CP may have the clinical features of dyskinesia such as dystonia (i.e., slow, sometimes rhythmic movements with increased muscle tone and abnormal postures, e.g., in the jaw and upper extremities), athetosis (slow, writhing, involuntary movements, particularly in the distal extremities), chorea (abrupt, irregular, jerky movements),

rigidity and tremors affecting the arms, legs, trunk and neck. It may be suspected when the infant at the age of 6 months may have difficulty in reaching and grasping a dangling ring.

- **Mixed type CP:** A significant number of children with CP can have clinical manifestation from more than one type.

Diagnosis

Diagnosis of CP is suspected in children with history of low birth weight (LBW), perinatal insult, feeding difficulties and does not show normal developmental milestones affecting both gross and fine motor, language and socialization. Urine and plasma tests for diagnosing inborn error of metabolism. CT scan and MRI will help to know the extent of cerebral damage.

EEG in case of seizures, Electromyography and nerve conduction studies are helpful when a muscle or nerve disorder is suspected.

Management

High quality antenatal, intranatal and postnatal care can help in minimizing the occurrence of CP. Managing a child with CP requires a holistic approach. Multidisciplinary approach involving pediatrician, neurologist, orthopedician, physiotherapist, special educator and a nurse are needed to improve the quality of life. Attention is paid to improve the posture, reducing tone, preventing contractures, and early stimulation. Common drugs used are:

- Diazepam is used to control spasticity and athetosis.
- Dantrolene sodium helps in relaxation of muscles.
- Botulinum toxin is a new alternative agent used to reduce the muscle tone.
- AED for seizures.

Surgery

Surgical treatments used in patients with cerebral palsy include the following:

- **Intrathecal baclofen pump insertion:** To treat spasticity and/or dystonia
- **Selective dorsal rhizotomy:** To treat spasticity in the lower extremities
- **Stereotactic basal ganglia surgery:** May improve rigidity, choreoathetosis, and tremor
- **Orthopedic surgical intervention:** To treat scoliosis, joint contractures or dislocation

Nursing Management

- **Impaired physical mobility related to neuromuscular impairment as evidenced by inability to control lower extremities, limited range of motion (ROM) and muscle spasms.**
 - Assess the type of auditory, visual, motor, or intellectual deficit.

- Do a developmental assessment and note the development of milestones (such as stand with help, or walk when led).
- Facilitate activities in using fine and gross motor skills (such as giving a ball in hand to encourage throwing, holding a spoon)
- Perform range-of-motion exercises every 4 hours for the child unable to move body parts.
- Educate the family on how to use an orthotic device as indicated.
- Administer medications as prescribed: Anticholinergics (e.g., Benztropine mesylate), Muscle relaxants (e.g., Baclofen), Anticonvulsants (e.g., Gabapentin)
- **Imbalanced Nutrition: Less than body requirements as evidenced by difficulty chewing, swallowing and sucking**
 - Monitor and record height and weight.
 - Assess the infant sucking and swallowing ability.
 - Offer small frequent meals more throughout the day.
 - Position the child upright during feedings to avoid aspiration. Use soft and blended foods.
 - Encourage adequate fluid intake and high fiber foods such as wholegrain cereals, fruit, and vegetables to avoid constipation.
 - Offer high protein
 - Teach the family regarding enteral tube feeding if needed.
- **Impaired verbal communication related to neurologic impairment as evidenced by difficulty vocalizing words**
 - Learn patient needs and pay attention to nonverbal cues.
 - Maintain a calm, unhurried manner. Provide ample time for the child to respond.
 - Keep distractions such as television and radio at a minimum when talking to the child.
 - Provide an alternative means of communication such as flash cards, whiteboards, hand signs or a picture board
 - Involve family and significant others in the plan of care as much as possible.
 - Coordinate the child with a speech therapist as indicated.
- **Risk for Injury related to impaired motor function, seizure activity**
 - Assess level of consciousness
 - Explain to the parents on different stimuli that can trigger a seizure activity. Bright flashing lights, lack of sleep, and lengthy exposure to television or computer games may precipitate a seizure activity.
 - Provide a safe environment for the child by wearing protective gear (helmet, kneepads) if needed.
 - Assist the child in performing ADL
 - Institute seizure precautions such as keeping padded side rails up with the bed in its lowest position.
 - Administer benzodiazepines (e.g., Diazepam [Valium]).
 - Coordinate with a physical therapist for strengthening exercises and gait training to prevent falls.



TRAUMATIC BRAIN INJURY

Traumatic brain injury (TBI) is the leading cause of death and disability in children. The scalp is highly vascularized and a potential cause of lethal blood loss. Even a small loss of blood volume can lead to hemorrhagic shock in a newborn, infant, and toddler, which may occur without apparent external bleeding. Therefore, children are considered to exhibit a specific pathological response to brain injury and accompanying neurological symptoms.

Causes

Head injuries (HI) are the most common types of trauma associated with motor vehicle/cycle injuries, and fall. Child can sustain injury as an occupant, pedestrian or cyclist. Head injury can occur in isolation or in combination with other musculoskeletal and spinal injuries.

HI is a pathological state involving injury to the scalp, skull, meninges or brain due to any mechanical force. HI may occur as skull fracture and/or intracranial lesion.

Classification of Head Injury

Head injury can be classified on the basis of severity and morphology, described in Table 20.9. Pediatric glasgow coma scale (GCS) is shown in Figure 20.5.

HI can also be classified as primary and secondary.

- Primary HI occurs at the time of trauma and may include skull fracture, contusions, intracranial hematoma, and diffuse injury.
- Secondary HI occurs as a result of raised ICP, cerebral edema and hypoxic brain injury.

Pathogenesis

The pathology of brain injury is directly related to the force of impact of injury. The intracranial content like brain, blood or CSF are damaged as a result of the impact of injury. The elastic pliable skull of the child provides protection to the intracranial content due to its ability to absorb the direct energy. Despite the pliable skull, children are more vulnerable due to their large size head and insufficient musculoskeletal support.

Physical forces act through acceleration or deceleration. When the stationary head receives a blow, the sudden acceleration caused deformation of the skull along with mass movement of brain, while deceleration takes place during a fall. This movement causes bruises to the point of contact called **COUP** and on the side injury is called **CONTRECOUP**.

Types of Head Injury

- **Concussion:** Concussion is due to transient and reversible neuronal dysfunction leading to immediate loss of consciousness and persists for a short duration.
- **Contusion and laceration:** Contusion and laceration are the terms used to explain the visible bruising and tearing of cerebral tissues. Occipital, temporal, frontal lobes and irregular surfaces of anterior and middle fossae at the base of skull are susceptible to contusion and laceration. The degree of brain damage in the contused areas depends upon the extent of vascular injuries.
- **Fractures:** Various types of fractures like linear, depressed, comminuted, basilar, open and diastatic fractures can be seen in children. Due to the flexibility of skull bones in children, they are able to sustain a greater degree of deformation. In fact large amount of force is required to produce a fracture in the skull of infants.

Table 20.9: Classification of head injury

Classification			GCS score
Severity	<ul style="list-style-type: none"> • Minor • Moderate • Severe 		13–15 9–12 3–8
Morphology	Skull fracture	Vault	<ul style="list-style-type: none"> • Linear versus stellate • Depressed/nondepressed • Open/closed
		Basilar	<ul style="list-style-type: none"> • With/without CSF leak • With/without seventh nerve palsy (facial nerve)
	Intracranial lesions	Focal	<ul style="list-style-type: none"> • Epidural • Subdural • Intracerebral
		Diffuse	<ul style="list-style-type: none"> • Concussion • Multiple contusions • Hypoxic/ischemic injury • Axonal injury

>1 year			<1 year	
Eye opening	4	Spontaneously	Spontaneously	
	3	To verbal command	To shout	
	2	To pain	To pain	
	1	No response	No response	
>1 year			<1 year	
Best motor response	6	Obeys		
	5	Localizes pain	Localizes pain	
	4	Flexion—withdrawal	Flexion—normal	
	3	Flexion—abnormal (decorticate rigidity)	Flexion—abnormal (decorticate rigidity)	
	2	Extension (decerebrate rigidity)	Extension (decerebrate rigidity)	
	1	No response	No response	
>5 years			2–5 years	0–23 months
Best verbal response	5	Oriented and converses	Appropriate words and phrases	Smiles, coos, cries appropriately
	4	Disoriented and converses	Inappropriate words	Cries
	3	Inappropriate words	Cries and/or screams	Inappropriate crying and/or screaming
	2	Incomprehensible sounds	Grunts	Grunts
	1	No response	No response	No response

Figure 20.5: Pediatric glasgow coma scale

- **Epidural hematoma (EDH):** Blood accumulation between the dura and the skull results in the development of EDH. Bleeding is arterial, rapid in onset, forcing underlying brain contents downward and inward due to expansion of the brain. Parieto-temporal region is the commonest site for EDH. The hemorrhage can be from dural veins or dural sinuses. The typical course of EDH is the momentary loss of consciousness, followed by normal period and then development of lethargy and coma.
- **Subdural hematoma (SDH):** Subdural hematoma results due to bleeding between the dura mater and cerebrum due to rupture of cortical veins that bridge the subdural gap. The SDH tends to develop slowly as compared to EDH. Birth trauma, fall, assault, physical abuse or violent shaking can lead to the development of SDH.

Diagnosis

- A detailed history and physical examination including level of consciousness (LOC), pupillary reactions and witnessed seizures following injury will help in establishing the diagnosis. Physical examination includes primary and secondary survey.
- Radiologic studies like X-ray skull, CT scan and MRI would help in finding out the structural alterations in the brain.

Complications

Major complications seen following HI are hemorrhage, infection, edema and herniation through the tentorium. Some degree of cerebral edema is expected in 24–72 hours following injury after TBI. Cerebral edema can occur due to direct cellular injury, or vascular injury or anoxia.

Management

- Children with mild head injuries usually do not need any treatment other than careful monitoring for 48 hours. For severe TBI:
 - Detailed head to toe examination is performed to find out the other possible associated injuries like abdominal, spine or long bone injuries.
 - Early stabilization of the child in terms of airway, breathing, circulation and maintenance of temperature after primary survey.
 - Intracranial pressure monitoring
 - **Cerebral perfusion pressure (CPP):** Joint management of ICP and CPP is considered a standard practice for management of children with severe TBI
 - Sedatives and analgesics are required for general care of all TBI children to achieve a level of anesthesia needed for invasive procedures, such as airway management, ICP control, to coordinate respiratory efforts with the ventilator, and anxiety relief during diagnostic imaging. Combination of opioids and benzodiazepines for pain control and sedation are used in children with severe TBI.
 - **Hyperosmolar therapy:** IV mannitol and hypertonic saline are routinely used to control intracranial hypertension in children with severe TBI.
 - CSF drainage to reduce the volume of the contents of the intracranial vault for the management of increased ICP.
 - Hyperventilation reduces ICP by lowering CBF by cerebral vasoconstriction of arterioles.
 - Barbiturates to control refractory intracranial hypertension, e.g., Pentobarbital.



- **Temperature control:** Avoid hyperthermia which increases metabolic demands
- **Decompressive craniectomy:** Performed for controlling intracranial hypertension
- **Nutritional management:** It is recommended that full nutritional replacement be instituted by day 7 post-injury.
- **Anticonvulsants:** Immediate prophylactic administration of anticonvulsant is recommended in children with severe TBI.
- Surgical repair (Craniectomy) is indicated in the following cases:
 - CSF leak is clearly recognized,
 - Detection of foreign body,
 - Debridement of the local wound is deemed necessary,
 - Infected wounds,
 - Evacuation of hematoma is required, and
- For cosmetic reasons.

Prevention of Traumatic Brain Injury in Children

- Never leave the baby alone on a raised surface
- Use car safety seats and seat belts
- Make sure that child always wears a properly fitted safety helmet while riding a bicycle or scooter.
- Do not vigorously shake the baby.

Nursing Management

Nursing Diagnoses

- Altered breathing pattern and ineffective airway clearance
- Impaired thermoregulation
- Altered sensorium
- Altered cerebral perfusion
- At risk for fluid and electrolyte imbalance
- At risk for infection due to multiple invasive lines
- Impaired skin integrity
- Knowledge deficit and anxiety of parents related to the disease condition

Nursing Interventions

- Assessment and maintenance of airway. All children with head injury should be suspected to have cervical spine injury unless ruled out.
- Provide C- spine protection using a cervical collar or immobilizing using sand bags.
- Start oxygen at high flow rate (10–12 L/min) using non-rebreathing mask.
- Attach child to the cardiac monitor and observe oxygen saturation.
- Control bleeding using appropriate measures like direct pressure, tourniquet application.

- Keep child NPO. Establish IV lines using two wide bore cannula, draw blood samples for various biochemical parameters.
- Clean the wounds and apply sterile dressings over the wounds.
- Administer mannitol (20%) and phenytoin (AED) as prescribed.
- Explain family members about the child's condition and the type of injuries sustained by him.
- Provide psychological support to the family members to alleviate their anxiety.

HYDROCEPHALUS

Definition

Hydrocephalus is a condition in which abnormal accumulation of CSF in the brain causes increased ICP inside the skull. This is usually due to blockage of CSF outflow in the brain ventricles or in the subarachnoid space at the base of the brain.

Related Anatomy and Physiology

The CSF is secreted at the choroid plexus within the ventricles by ultrafiltration and active secretion. It passes from the lateral ventricle to the third ventricle, fourth ventricle and then to the basal cisterns and then reaches the cerebral and spinal subarachnoid spaces where it is absorbed via the arachnoid villi (granulations) into the venous channels and sinuses (Fig. 20.6). About 20 mL of CSF is secreted in an hour and its turnover is 3–4 times in a day.

Types of Hydrocephalus and their Etiology

Hydrocephalus can be caused by impaired CSF flow, reabsorption, or excessive CSF production. The normal pressure in the skull is a balance between pressure of skull bones and the blood, CSF and brain parenchyma (skull pressure = brain pressure + CSF pressure + blood pressure).

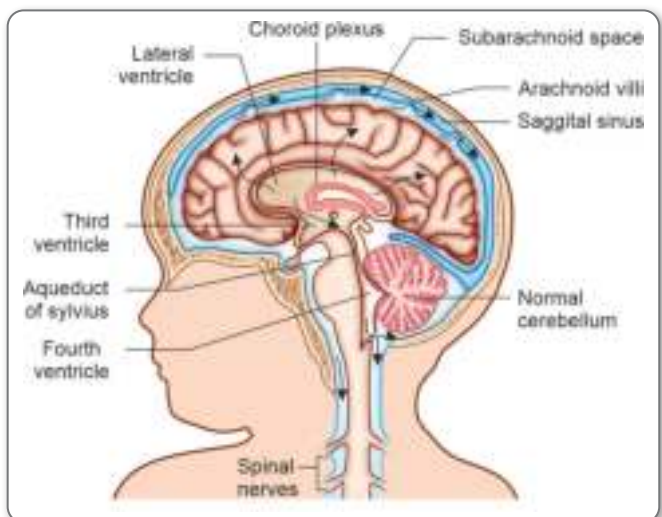


Figure 20.6: Flow of cerebrospinal fluid

Based on its underlying mechanisms, hydrocephalus can be classified into communicating, and non-communicating (obstructive). Both communicating and non-communicating forms can be either congenital, or acquired.

Communicating Hydrocephalus

Communicating hydrocephalus, also known as non-obstructive hydrocephalus, is caused by impaired CSF resorption in the absence of any CSF flow obstruction. It has been hypothesized that this is due to functional impairment of the arachnoid granulations, which are located along the superior sagittal sinus and is the site of cerebrospinal fluid resorption back into the venous system. Various neurologic conditions may result in communicating hydrocephalus, i.e., subarachnoid/intraventricular hemorrhage, meningitis, Chiari malformation, and congenital absence of arachnoidal granulations (Pacchioni's granulations).

Normal pressure hydrocephalus (NPH) is a particular form of communicating hydrocephalus, characterized by enlarged cerebral ventricles, with only intermittently elevated CSF. The diagnosis of NPH can be established with the help of continuous intraventricular pressure recordings (over 24 hours or even longer). Dynamic compliance studies may be also helpful. Altered compliance (elasticity) of the ventricular walls, as well as increased viscosity of the CSF, may play a role in the pathogenesis of normal pressure hydrocephalus.

Noncommunicating Hydrocephalus

Non-communicating hydrocephalus, or obstructive hydrocephalus, is caused by a CSF-flow obstruction (either due to external compression or intraventricular mass lesions).

Foramen of Monro obstruction may lead to dilation of one or, if large enough (e.g., in colloid cyst), both lateral ventricles.

Aqueduct of Sylvius, may be obstructed by a number of genetically or acquired lesions (e.g., atresia, ependymitis, hemorrhage, and tumor) and lead to dilatation of both lateral ventricles as well as the third ventricle.

Fourth ventricle obstruction will lead to dilatation of the aqueduct as well as the lateral and third ventricles.

Foramina of Luschka and Magendie may be obstructed due to congenital failure of opening (e.g., Dandy-Walker malformation).

Subarachnoid space surrounding the brainstem may be obstructed due to inflammatory or hemorrhagic fibrous meningitis, leading to widespread dilatation, including the fourth ventricle.

Congenital Hydrocephalus

As the cranial bones fuse by the end of the third year of life, for head enlargement to occur, hydrocephalus must occur before them.

The causes are usually genetic but can also be acquired. It includes (1) intraventricular matrix hemorrhages in premature infants, (2) intrauterine infections – rubella, CMV (3) congenital malformations- Arnold-Chiari malformation, aqueduct atresia and stenosis, and Dandy-Walker malformation.

In newborns and toddlers with hydrocephalus, the head circumference is enlarged rapidly. Since the skull bones have not firmly joined together by this time, bulging fontanelles may be present.

The infant exhibits restlessness, poor feeding, and frequent vomiting. As the hydrocephalus progresses, the child becomes lethargic, and the infant shows lack of interest in his surroundings. Later on, the upper eyelids become retracted and the eyes are turned downwards (due to hydrocephalic pressure on the mesencephalic tegmentum and paralysis of upward gaze). Movements become weak and the arms may become tremulous. Papilledema is absent but there may be reduction of vision. The head becomes so enlarged that the child may eventually be bedridden.

About 80–90% of fetuses or newborn infants with spina bifida develop hydrocephalus.

Acquired Hydrocephalus

This condition is acquired as a consequence of:

- CNS infections- tuberculosis
- Chronic and pyogenic meningitis
- Posterior fossa tumors—medulloblastoma, astrocytoma, ependymoma,
- Head trauma
- Intracranial hemorrhage (subarachnoid or intraparenchymal) and is usually extremely painful for the patient.

Pathophysiology

The usual cause of accumulation of CSF is a block in the pathway of CSF circulation that hampers absorption of the fluid. This block may be either proximal to the outlet of the fourth ventricle, i.e., within the brain (non-communicating) or there is a block in the subarachnoid space that prevents CSF from reaching the arachnoid villi (communicating).

When CSF collects in the ventricles, it causes the ventricles to distend. In the infants while the fontanelles are open and the sutures are not fused, the increase in ventricular volume can be compensated by increase in the dimension of the vault of the skull. Thus there is no increase in the ICP at first but an increase in the skull circumference is noted.

As the ability of pliable cranium to accommodate increased CSF is exhausted, signs of increased ICP are evident.

Ventricles are dilated. Ependymal lining of ventricles is disrupted resulting in periventricular ooze. Subependymal edema occurs. White matter is compressed. Cortex is generally preserved till late but cortical atrophy may occur. The process may be reversible if the treatment is started early.

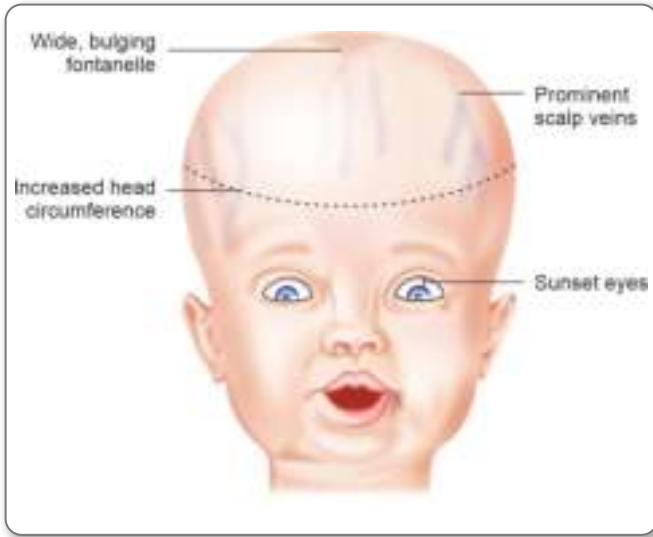


Figure 20.7: Symptoms of hydrocephalus

Signs and Symptoms (Fig. 20.7)

- A full or bulging fontanel
- Increasing head circumference, enlarged head size
- Delayed closure of fontanels and sutures
- Seizures
- Bulging eyes and an inability of the baby to look upward with the head facing forward
- Very noticeable scalp veins
- Increased irritability
- High-pitched cry
- Poor feeding
- Projectile vomiting
- Sleepiness or less alert than usual
- Developmental delays

Associated Symptoms

- Headache, nausea, vomiting
- Personality and behavior disturbances, irritability, head banging, apathy and drowsiness
- Sunset eyes
- Papilledema, pyramidal tract signs, cranial nerve palsies
- Abnormal skull contour, prominent forehead
- Prominent and dilated scalp veins
- Impaired upward gaze
- Limbs become spastic because of stretching of cortical fibers
- Distortion of the brainstem may lead to bradycardia, systemic hypertension and altered respiratory rate
- Toddlers whose sutures have not yet closed also show the signs of head enlargement.
- Older toddlers and children, once their sutures have closed, will show other symptoms of raised ICP caused by their enlarged ventricles.

The child might have problems with vision (blurred or double vision), balance, and delayed development in areas like walking, talking, or poor coordination. As with infants, a child may be more irritable or tired than normal.

The child may show a change in personality or be unable to concentrate or remember things, and their school performance may decline. Older children may have difficulty waking up and staying awake. While at times the symptoms are very noticeable, other times they can be very subtle and progress so slowly that only in retrospect are they appreciated.

Symptoms of increased ICP may include headaches, vomiting, nausea, papilledema, sleepiness, or coma. Elevated ICP may result in uncal and/or cerebellar tonsil herniation, with resulting life-threatening brain stem compression.

The triad of gait instability, urinary incontinence and dementia is a relatively typical manifestation of the distinct entity normal pressure hydrocephalus (NPH). Focal neurological deficits may also occur, such as abducens nerve palsy and vertical gaze palsy.

Complications

Because hydrocephalus injures the brain, thought and behavior may be adversely affected. The child may have learning disabilities, motivation and visual problems, problems with co-ordination and epilepsy.

Diagnosis

- Prenatal ultrasound
- Clinical examination
- Serial recording of head circumference: An increase in the head circumference in the first three months of life >1 cm every fortnight should arouse suspicion of hydrocephalus
- Serial cranial ultrasound: To evaluate ventricular size
- Imaging studies like X-ray, MRI, and CAT scan: To determine the site of obstruction and to identify associated malformation through a combination of large magnets, radiofrequencies, and a computer to produce detailed images of organs and structures within the body.

Treatment

Goal of treatment is to reduce the pressure in the baby's head and to properly drain the CSF. Occasionally, medications or procedures to draw off extra CSF may be used.

Medical Management

Acetazolamide – 50 mg/kg/day. It is a carbonic anhydrase inhibitor which reduces CSF production by the choroid plexus.

Oral glycerol — 1–1.5 g/kg as a single dose, then 500 mg/kg 4–8 hours later. It is an osmotic diuretic which helps in reduction of ICP.

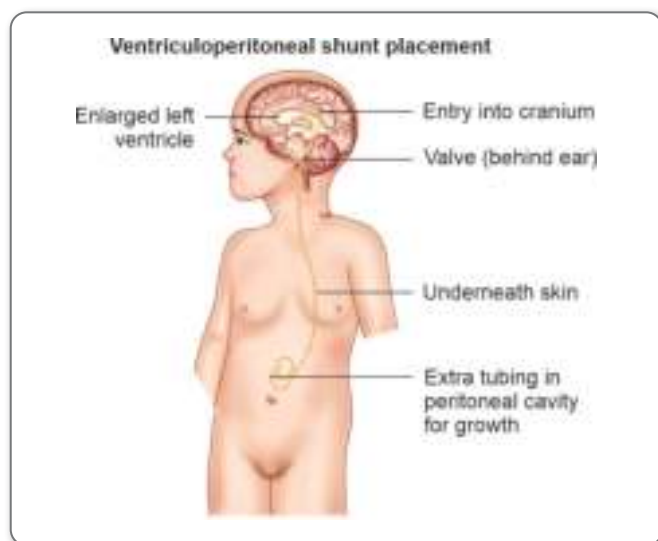


Figure 20.8: Ventriculoperitoneal shunt

Surgical Management

It involves the placement of a ventricular catheter, into the cerebral ventricles to bypass the obstruction and drain the excess fluid into other body cavities, from where it can be resorbed.

Most shunts drain the fluid into the peritoneal cavity (ventriculoperitoneal shunt) (Fig. 20.8). Alternative sites of shunt include the right atrium (ventriculoatrial shunt), pleural cavity (ventriculopleural shunt), and gallbladder.

A shunt system can also be placed in the lumbar space of the spine and have the CSF redirected to the peritoneal cavity (LP Shunt). An alternative treatment for obstructive hydrocephalus in selected patients is the endoscopic third ventriculostomy (ETV). In ETV, an opening is created in the floor of the third ventricle to allow the CSF to flow directly to the basal cisterns, thereby bypassing any obstruction, as in aqueductal stenosis.

The shunt consists of three parts:

1. A tube that is placed inside of the ventricular space
2. A reservoir and valve to control the flow of CSF
3. Tubing that is directed under the skin to the abdomen, or less commonly to the heart or lung area

The shunt redirects the CSF out of the head through the tubing to a location elsewhere in the body where it can be absorbed. The shunt is usually placed behind the ear and the tubing is threaded from behind the ear, under the skin to the area of the abdomen, heart, or lung.

Shunt Complications

Examples of possible complications include shunt malfunction, shunt failure, bleeding and shunt infection. Although a shunt generally works well, it may stop working if it disconnects, becomes blocked, or it is outgrown. If this

happens the cerebrospinal fluid will begin to accumulate again and a number of physical symptoms will develop, some extremely serious, like seizures. The shunt failure rate is also relatively high and it is not uncommon for patients to have multiple shunt revisions within their lifetime.

Ommaya Reservoir

The CSF surrounds the brain and spinal cord. Sometimes, medications need to be given into the CSF because intravenous (IV) medications will not reach that area. Medications can be given into the CSF when an ommaya reservoir is inserted. The ommaya reservoir, or port, is a dome like device that is placed surgically under the scalp.

It may be used to:

- Give medications directly into the CSF
- Measure the pressure of the CSF
- Take samples of CSF to be tested

Prognosis

Affected individuals and their families should be aware that hydrocephalus poses risks to both cognitive and physical development. However, many children diagnosed with the disorder benefit from rehabilitation therapies and educational interventions and go on to lead normal lives with few limitations. Treatment by an interdisciplinary team of medical professionals, rehabilitation specialists, and educational experts is critical to a positive outcome.

Nursing Management

- **Ineffective Cerebral Tissue Perfusion r/t decreased venous or arterial blood flow, increased ICP as evidenced by decreased pulse or respiration, high pitched cry.**
 - Monitoring hourly vital signs to recognize early signs of increased ICP (such as fluctuating blood pressure, tachycardia, and shallow breathing) or Cushing's triad (bradycardia, apnea, and widening pulse pressure).
 - Assess neurological status such as mental status, motor, and balance, reflexes (for newborns and infant), and cranial nerves.
 - Examine the pupils – size, shape, equality, and position of the pupils, and their response to light.
 - Note the quality and tone when children cry. A high pitched cry may indicate increased ICP.
 - Measure the child's head circumference and appearance of anterior fontanel.
 - Provide a non-stimulating environment and adequate rest periods.
 - Elevate the head of the bed gradually about 15°–45° as indicated. Maintain the client's head in neutral position.
 - Provide oxygen therapy as needed.
 - Administer diuretics, carbonic dehydratase inhibitors, corticosteroids as ordered.



- **Anxiety r/t threat to change in health status as evidenced by increased apprehension that condition of infant might worsen**
 - Assess source and level of anxiety and need for information and support about the condition and impending surgery.
 - Communicate therapeutically with parents and answer question calmly and honestly.
 - Allow expressions of concern and opportunity to ask questions about condition and recovery of ill infant/child.
 - Encourage parents to remain involved in care and decision-making regarding infant/child.
 - Encourage parents to stay with infant/child or visit when able if hospitalized, assist in care (hold, feed, diaper) and make suggestions for routines and methods of treatment.
 - Prepare child/parents for diagnostic tests and potential surgical procedures.
 - Clarify any misinformation and answer all questions honestly and in simple understandable language.
 - Teach about shunt placement and reason; possible future revision of shunt placement, signs and symptoms of shunt complication or malfunction.
- **Risk for Injury (Preoperative) r/t sensory, integrative and effector dysfunction preoperatively as evidenced by behavioral changes**
 - Perform neurologic and vital assessment every 4 hours or as needed.
 - **Assess for increased ICP:** a rapidly increased circumference of head, tense, bulging fontanels, widening suture lines, irritability, lethargy, “cracked pot” sound percussion, sunset sign, opisthotonus, spasticity of lower extremities, seizures, high-pitched cry, distended scalp veins, changes in normal feeding patterns.
 - **Assess for early signs:** Headache, nausea, vomiting, diplopia, blurred vision, seizures, irritability, restlessness, decrease in school performance, decreased motor performance, sleep loss, weight loss, memory loss progressing to lethargy and drowsiness.
 - **Late signs:** Decreased level of consciousness, decreased motor response to commands, decreased response to pain, change in pupils, posturing, papilledema.
 - Carry out seizure precautions including padding of crib/bed, remove toys and objects from the bed, and maintain suction and oxygen at bedside, note and report characteristics of seizure, note and report characteristics of seizures.
 - Position with head elevated 30 degrees and support head when handling or changing position; monitor skin integrity with position changes.
- **Risk for injury (postoperative) related to shunt placement and potential complications of shunt functioning caused**

by changing of position of tubing, displacement with growth, increased ICP or kinking or plugging of shunt tubing

- Assess for shunt malfunction – observe signs and symptoms of increased ICP, swelling along shunt tract; note presence or severity of headache and neck pain; behavior changes (lethargy, irritability), physical changes (full fontanel, nausea, vomiting, edematous eyes, tender, swollen abdomen).
 - Note vomiting, drowsiness, irritability, swelling at pump site, redness, exudate, and temperature of the child which indicates shunt blockage.
 - Instruct parent on hydrocephalus and shunt placement. Teach about brain anatomy, hydrocephalus (causes, diagnostic test, treatment, signs of shunt malfunction and infection). Supply written materials. Emphasize the importance of early identification of infection/malfunction and prompt notification.
 - Teach parents about the need for bowel elimination at least every 2 days and steps to take to ensure bowel movement to prevent complications associated with a ventriculoperitoneal shunt.
 - Position carefully on nonoperative side postoperatively; maintain bed position and activity level as ordered depending on shunt dynamics.
 - Instruct activities to be avoided such as rough contact sports.
- **Risk for Infection r/t Invasive procedure of shunt insertion as evidenced by excessive drainage on dressing, elevated temperature, lethargy**
 - Assess site for inflammatory process, temperature elevation, increased WBC, characteristics of drainage on dressings.
 - Monitor temperature every four hours.
 - Teach about signs and symptoms of infection of site and shunt tract and to notify position if noted.
 - Follow principles of asepsis when performing procedures such as dressing changes.
 - Teach parents about wound care and dressing change, emphasize the importance of good hand washing techniques.
 - Administer antibiotics.

SPINA BIFIDA

Spina bifida is a birth defect that involves the incomplete development of the spinal cord or its coverings. It occurs at the end of the first month of pregnancy when the two sides of the embryo's spine fail to join together, leaving an open area. Normally the closure of the neural tube occurs around 28 days after fertilization. However, if something interferes and the tube fails to close properly, a neural tube defect will occur.

Causes/Risk Factors

- Some evidence suggest that genes may play a role, but in most cases there is no familial connection.
- A high fever during pregnancy may increase a woman's chances of having a baby with spina bifida.
- Women with epilepsy who have taken the drug valproic acid to control seizures may have an increased risk of having a baby with spina bifida
- Diabetes, obesity
- Having a relative with spina bifida

Classification

The two types of spina bifida are spina bifida occulta and spina bifida manifesta.

1. **Spina bifida occulta:** It is the mildest form of spina bifida. Occulta means hidden. Most children with this type of defect never have any health problems, and the spinal cord is often unaffected.
2. **Spina bifida manifesta:** It is of two types (Fig. 20.9):
 - i. **Meningocele:** It refers to protrusion of sac from the spinal column containing the meninges and CSF.
 - ii. **Myelomeningocele:** It is the most severe form of spina bifida. There is protrusion of spinal cord along with meninges and CSF. Most babies who are born with this type of spina bifida also have hydrocephalus.

Pathophysiology

Because of the abnormal development of and damage to the spinal cord, a child with myelomeningocele will have some paralysis. The degree of paralysis largely depends on where the opening occurs in the spine. The higher the opening is on the back, the more severe the paralysis tends to be.

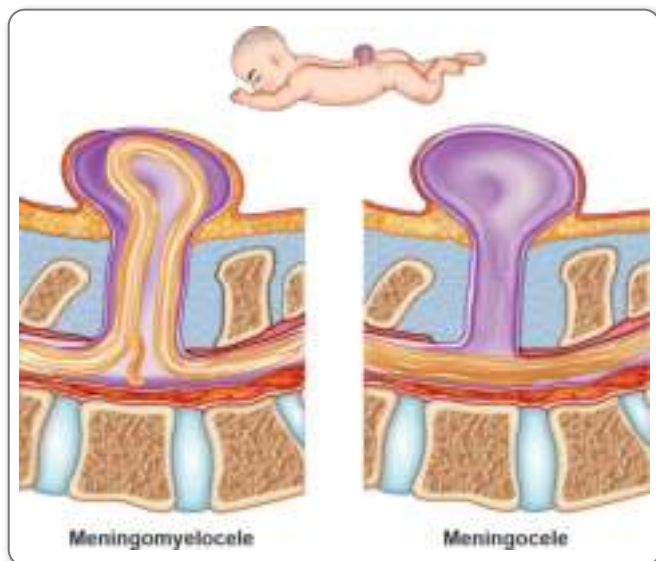


Figure 20.9: Spina bifida manifesta

Children with spina bifida often have problems with bowel and bladder control, and some may have attention deficit hyperactivity disorder (ADHD) or other learning difficulties, such as hand-eye coordination problems.

Symptoms

Spina bifida occulta: Often has no outward signs or symptoms. The spinal cord does not protrude through the skin, although a patch of hair, a birthmark, or a dimple may be present on the skin over the lower spine.

Meningocele: There is a fluid-filled sac visible on the back. The sac is often covered by a thin layer of skin and can be in the shape of a grape or cricket ball.

Myelomeningocele: Also have a sac-like mass that bulges from the back, but a layer of skin may not always cover it. In some cases, the nerves of the spinal cord may be exposed.

Diagnosis

Prenatal Tests

- **The α -fetoprotein (AFP) test:** It is performed between the 16th and 18th week of pregnancy. AFP produced by fetus is measured in the mother's bloodstream. If the amount is high, the test is repeated because in many cases, high AFP readings are false. If the second result is high, other tests is done to double-check and confirm the diagnosis.
- **Ultrasound**
- **Amniocentesis:** Amniotic fluid is tested for AFP.

Prevention

- Maternal folic acid deficiency has been linked to spina bifida, and researchers believe that many cases can be prevented if women of childbearing age consume 0.4 milligrams (400 micrograms) of folic acid every day, from **3 months before conception**, and continue to take it throughout the first trimester.
- It is important that folic acid consumption starts *before* the onset of pregnancy to provide the best protection.
- Sources of folic acid: Eggs, orange juice, and dark green leafy vegetables.
- Many multivitamins contain the recommended dose of folic acid, too.

Treatment of Spina Bifida

- **Spina bifida occulta:** Rarely need treatment.
- **Spina bifida manifesta:** Treatment depends on the type of spina bifida and its severity.
 - **Meningocele:** Surgery is usually done during infancy in which the meninges are pushed back and the hole in the vertebrae is closed. Child will have no other health problems later unless there is nerve tissue involved with the sac.



- **Myelomeningocele:** It needs more immediate attention and often has surgery within the first 1 to 2 days after birth. The spine is pushed back into the vertebrae and the hole is closed to prevent infection and protect the spine.
- A shunt will be needed in case of hydrocephalus.
- Some children need successive surgeries to manage problems with their feet, hips, or spine. The location of the gap in the back often dictates what kind of adaptive aids or equipment a child with myelomeningocele will need. Those with a gap high on the spinal column and more extensive paralysis often need to use a wheelchair, while those with a gap lower on the back may be able to use crutches, leg braces, or walkers.

Nursing Management

Prenatal

- Refer prospective parents to a genetic counselor
- Encourage women of child bearing age to take folic acid supplements (0.4 mg daily)
- Provide psychological support for diagnosis, preoperative and postoperative care

Nursing Diagnosis

- Hypothermia
- Impaired Urinary Elimination
- Bowel Incontinence
- Disturbed Body Image
- Interrupted Family Processes
- Risk for Infection
- Risk for Injury

Nursing Interventions

Preoperative

- Manage hypothermia. Place the child in radiant warmer or incubator.
- Check the leakage, infection at the sac
- Assess for signs and symptoms of CNS infection
- Assess for motor activity below the sac
- Assess bowel and bladder functions
- Teach parents and family measures to prevent contractures, pressure ulcers, UTI and other complications. Keep genital area clean after every elimination episode.
- Practice hand washing.
- Administer antibiotics.
- Maintain the infant in a prone position or side-lying, as permitted, with head lower than buttocks or hips slightly flexed with a pad between the knees; anchor position with sandbags. It reduces pressure on the sac to prevent possible rupture and prevents rolling on side or back.
- Prevent trauma by keeping pressure off the sac
- Prevent the sac from drying, cover it with saline soaked sterile dressing

Postoperative

- Manage hypothermia
- Provide thorough skin care if paralysis occurs
- Teach clean intermittent catheterization to resolve urinary incontinence. Administer anticholinergic, antispasmodic, smooth muscle relaxant to improve bladder storage and continency by increasing bladder action.
- Prevent constipation. Establishes a routine for elimination to empty bowel. Place child on a toilet or potty chair at the same time each day.
- Promote ROM
- Assess motor ability and sensation below the level of the lesion; paralysis is possible
- Assess for latex allergy

NEUROBLASTOMA

Neuroblastoma is a malignant (cancerous) tumor that develops from nerve tissue. It is the most common extracranial solid cancer in childhood and the most common cancer in infancy.

Etiology

Neuroblastoma can occur in many areas of the body. It develops from the tissues that form the sympathetic nervous system.

Most neuroblastomas begin in the abdomen in the adrenal gland, spinal cord, or in the chest. They may also start in other areas. Neuroblastomas can spread to the bones (face, skull, pelvis, shoulders, arms, and legs), bone marrow, liver, lymph nodes, skin, and around the eyes (orbits).

The cause of the tumor is unknown. Neuroblastoma is most commonly diagnosed in children before age 5. The disorder occurs in approximately 1 out of 100,000 children and is slightly more common in boys.

In most patients, the neuroblastoma has already been spread when it is first diagnosed.

Clinical Manifestations

The first symptoms are usually fever, malaise, and pain. There may also be loss of appetite, weight loss, and diarrhea. Other symptoms depend on the site of the tumor, and may include:

- Bone pain or tenderness (if the cancer spreads to the bones)
- Difficulty breathing or a chronic cough (if the cancer spreads to the chest)
- Enlarged abdomen (from a large tumor or excess fluid)
- Flushed, red skin
- Pale skin and bluish color around the eyes
- Profuse sweating, tachycardia

Brain and nervous system problems may include:

- Inability to empty the bladder
- Paralysis of the hips, legs, or feet (lower extremities)



Note



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