

# Disorders Commonly Seen in CAP/C Children

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- Hydrocephalus
- Seizure Disorders
- Neural Tube Defects/Spina Bifida
- Duchenne's Muscular Dystrophy
- Cerebral Palsy
- Cortical Visual Impairment
- Airway Obstructions
- Gastroesophageal Reflux
- Pervasive Developmental Disorders

For nurses, nurse aides, and case managers

# Caution

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The information in this module is general in nature. Each child will present in their own unique way. When the information in this module contradicts the information given to you by your agency or by the physician, defer to the agency or physician.

# Target Audience

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This module is mandatory for all nurses, nurse aides, and case managers with less than one year of experience in pediatrics who will be caring for a child on the CAP/C program.

This module is recommended for other nurses, nurse aides, and case managers.

# Objectives

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- Recognize the signs and symptoms of shunt malfunction and shunt infection in children with hydrocephalus.
- Describe the different types of seizures. List the possible treatments for seizure disorders.
- Describe the differences between the different types of spina bifida. Know how to implement latex precautions in all patients with spina bifida.
- Describe the typical course of Duchenne's Muscular Dystrophy.

# Objectives

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- ❑ Understand what the different classifications of cerebral palsy mean in terms of the patient's movement. Describe the typical signs and symptoms of cerebral palsy. Describe the typical management of a child with cerebral palsy.
- ❑ Describe cortical visual impairment and ways in which to accommodate this visual impairment.
- ❑ Define three types of anatomical airway obstruction.

# Objectives

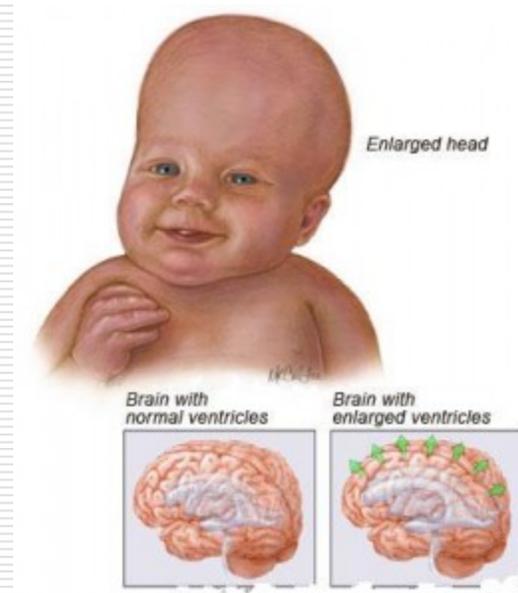
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- ❑ Describe typical symptoms of gastroesophageal reflux, potential complications, and typical treatments.
- ❑ List the five pervasive developmental disorders and briefly describe the differences among them.

# Hydrocephalus

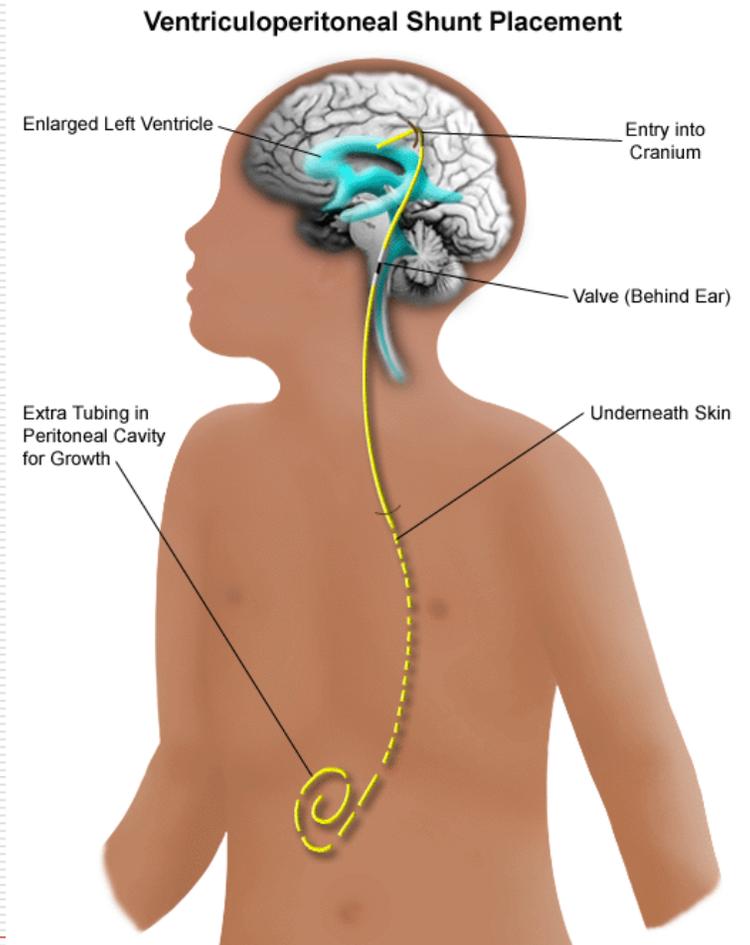
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- Hydrocephalus is a condition in which cerebrospinal fluid (CSF) — the clear, water-like fluid that surrounds and cushions the brain and spinal cord — is unable to drain normally from the brain. It builds up in the ventricles of the brain, causing increased intracranial pressure.
- In children whose skull bones have not yet fused, the skull will expand to accommodate the extra fluid and prevent the increase in pressure.



# Hydrocephalus

- Treatment is surgical insertion of a shunt that drains the fluid from the ventricles to another part of the body so that it can be reabsorbed by the bloodstream.



# Hydrocephalus

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## □ Types of shunts

- ventriculoperitoneal shunt (VP shunt)
- ventriculo-atrial shunt (VA shunt)
- ventriculo-pleural
- ventriculo-gall bladder

## □ Locations of Shunts

- frontal
- parietal
- occipital

# Hydrocephalus

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- Shunt malfunctions and shunt infections can occur rather commonly.
  - Signs of shunt malfunction: head growth, bulging fontanel, headaches, new or increased seizure activity, unusual irritability, behavior/personality changes, repeated vomiting, crossed eyes or inability to look up, apnea, difficulty swallowing, hoarse or weak cry, or difficulty staying awake

# Hydrocephalus

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- Signs of shunt infection: signs of shunt malfunction, fever, pain/redness/tenderness/drainage from incision site or tract, and abdominal pain

# Hydrocephalus

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**Always maintain a high index of suspicion for shunt malfunction and infection. Report any findings that are different for the child.**

**Shunt complications can also cause any number of symptoms that may not be obviously shunt related.**

**Listen to the parents' gut feelings about shunt problems – they are usually right.**

# Seizure Disorders

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## □ Types of Seizures

- **Partial/Focal** – involves only one part of the brain
  - Simple partial – person remains conscious
  - Complex partial – person loses consciousness
- **Generalized** – involves the entire brain
  - Absence/Petit Mal – lapse of awareness/staring spell
  - Myoclonic – rapid brief muscle contraction/jerky movement
  - Clonic – 2-3 episodes of myoclonus per second
- **Tonic-clonic=Grand Mal** – stiffening of the body (tonic) followed by jerking of limbs and face (clonic). Often accompanied by cyanosis and incontinence
- **Atonic** - drop seizures, person falls down
- **Status epilepticus** – continuous seizure activity with no recovery between seizures; requires emergency medical assistance
- **Infantile spasms** – onset in first year of life, characterized by stiffening of body, arms, and legs, usually occur soon after arousal from sleep, stop between ages 2 and 5 but usually replaced by some other form of seizure activity

# Seizure Disorders

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## □ Treatment of Seizures

### ■ scheduled medications

Usually more than one; for example, Dilantin, phenobarbital, keppra, topomax, valium, ativan, klonopin, depakane, tegretol

### ■ PRN medications

Given during a seizure; for example, Diastat, Ativan

### ■ ketogenic diet

a high fat low carbohydrate diet used to treat difficult to control seizures

# Seizure Disorders

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- Vagal Nerve Stimulator (VNS)  
An implanted device that emits an electrical signal that prevents seizures. When a breakthrough seizure does happen, a magnet is swiped over the device to activate an extra electrical signal
- Surgery  
removal of the portion of the brain causing the seizures, for example, anterior temporal lobectomy, hemispherectomy

# Seizure Disorders

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- Seizures can be very subtle, and will vary among children. You may see nothing more than a small twitch or change of expression. Work with the parents to learn how to identify the child's seizures.

# Neural Tube Defects

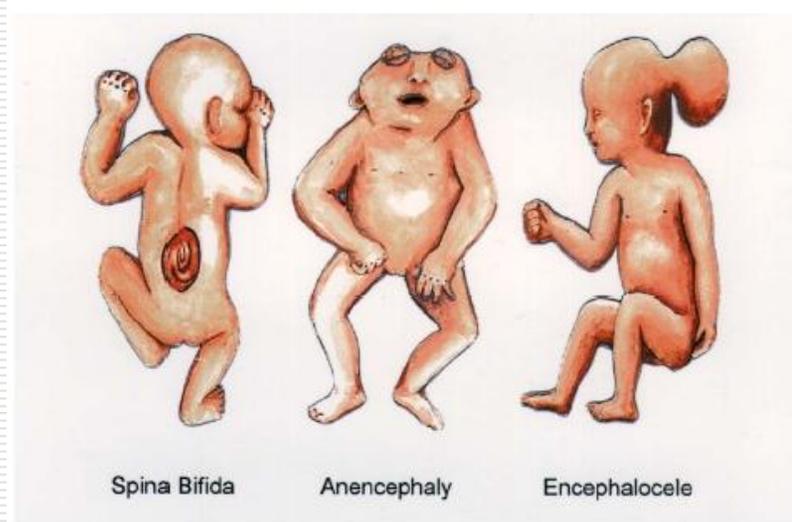
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Neural Tube Defects are malformations that occur during fetal development in the tissue that eventually becomes the brain and spinal cord. Neural tube defects include Spina Bifida, anencephaly, and encephalocele.

# Neural Tube Defects

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- ❑ Spina bifida appears as a localized mass on the back covered by skin or by the meninges, the three-layered membrane that envelops the spinal cord.
- ❑ Anencephaly is a lethal birth defect characterized by absence of all or part of the skull and scalp and malformation of the brain.
- ❑ Encephaloceles are rare and are characterized by protrusion of brain tissue and membranes through the skull.



# Neural Tube Defects - Spina Bifida

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There are three types of Spina Bidfida:

- Spina Bifida occulta
- meningocele
- myelomeningocele

# Neural Tube Defects – Spina Bifida

	Spina Bifida Occulta	Meningocele	Myelomeningocele
severity	mild	moderate	severe
description	The spinal cord and the nerves are usually normal and there is no opening on the back There is a small defect or gap in a few of the small bones(vertebrae) that make up the spine	The protective coatings (meninges) come through the open part of the spine like a sac that is pushed out.	the meninges (protective covering of the spinal cord) and spinal nerves come through the open part of the spine

# Neural Tube Defects – Spina Bifida

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	Spina Bifida Occulta	Meningocele	Myelomeningocele
treatment	usually requires no treatment	repaired surgically	operated on within 24 hours after birth. Surgery is generally performed to minimize the risk of infection and to preserve existing function in the spinal cord.

# Neural Tube Defects – Spina Bifida

	Spina Bifida Occulta	Meningocele	Myelomeningocele
prognosis	Although there may be no motor or sensory impairments evident at birth, subtle, progressive neurologic deterioration often becomes evident in later childhood or adulthood. In many instances, however, spina bifida occulta is so mild that there is no disturbance of spinal function at all.	no nerve damage no paralysis Individuals may suffer minor disabilities. New problems can develop later in life. Although most children with meningocele develop normally, affected children should be evaluated for hydrocephalus and bladder problems in order that they may be treated properly	nerve damage and more severe disabilities, including: <ul style="list-style-type: none"> <li><input type="checkbox"/>hydrocephalus</li> <li><input type="checkbox"/>bowel and bladder complications (need for catheterizations, bowel regimen)</li> <li><input type="checkbox"/>tendonitis</li> <li><input type="checkbox"/>obesity</li> <li><input type="checkbox"/>skin breakdown</li> <li><input type="checkbox"/>gastrointestinal disorders</li> <li><input type="checkbox"/>learning disabilities</li> <li><input type="checkbox"/>depression</li> <li><input type="checkbox"/>social issues</li> </ul>

# Neural Tube Defects – Spina Bifida

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## □ Latex allergy

All individuals with Spina Bifida should be considered at high risk for having an allergic reaction to natural rubber (not synthetic rubber) and should avoid contact with latex products in all settings from birth. Alternative products usually made of silicone, plastic, nitrile or vinyl can usually be safely substituted.

# Duchenne Muscular Dystrophy

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- ❑ The most common and the most severe form of Muscular Dystrophy. It affects about 1 out of every 3,500 boys. (Girls can carry the gene that causes the disease, but they usually have no symptoms.) This form occurs because of a problem with the gene that makes dystrophin. Without this protein, the muscles break down and a child becomes weaker.
- ❑ In cases of Duchenne MD, symptoms usually appear around age 5, as the pelvic muscles begin to weaken. Most kids with this form need to use a wheelchair by age 12. Over time, their muscles weaken in the shoulders, back, arms, and legs.
- ❑ Eventually, the respiratory muscles are affected, and a ventilator is required to assist breathing. Kids who have Duchenne MD typically have a life span of about 20 years.
- ❑ Although most kids with Duchenne MD have average intelligence, about a third experience learning disabilities and a small number are intellectually disabled.
- ❑ While the incidence of Duchenne is known, it's unclear how common other forms of MD are because the symptoms can vary so widely between individuals. In fact, in some people the symptoms are so mild that the disease goes undiagnosed.

# Cerebral Palsy

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"Cerebral" refers to the brain and "Palsy" to a disorder of movement or posture. If someone has cerebral palsy it means that because of an injury to their brain (cerebral) they are not able to use some of the muscles in their body in the normal way (palsy).

# Cerebral Palsy

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Cerebral palsy is classified by the type of movement problem:

- ❑ **spastic cerebral palsy** — causes stiffness and movement difficulties, the muscles are unable to relax, rigidity, hypertonia/contractures (Infants may start out floppy/hypotonic, and then develop spasticity.)
- ❑ **athetoid cerebral palsy** — leads to involuntary and uncontrolled movements
- ❑ **ataxic cerebral palsy** — causes a disturbed sense of balance and depth perception
- ❑ Some people also use a classification of **mixed**, which means that the child has components of two or more of the above. Usually, though the component which seems to be causing the most problem is used as the categorizing term. For example, the child with spastic cerebral palsy has mostly spastic muscle problems, but the child may also have a smaller component of athetosis and balance problems.

# Cerebral Palsy

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Cerebral Palsy is also classified by the body parts involved:

- ❑ hemiplegia – involves one side of the body
- ❑ paraplegia – involves the lower part of the body
- ❑ diplegia – involves both legs, infers some arm involvement
- ❑ triplegia – involves three extremities, usually both legs and one arm
- ❑ quadriplegia – involves both legs, both arms, as well as trunk and neck muscles

# Cerebral Palsy

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These classifications are combined into a type of cerebral palsy.

For instance, many children who receive CAP/C have spastic quadriplegia.

# Cerebral Palsy

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Symptoms of Cerebral Palsy include:

- ❑ Abnormal muscle tone: Muscles may be very stiff (spastic) or unusually relaxed and “floppy.” Limbs may be held in unusual or awkward positions. For example, spastic leg muscles may cause legs to cross in a scissor-like position.
- ❑ Abnormal movements: Movements may be unusually jerky or abrupt, or slow and writhing. They may appear uncontrolled or without purpose.
- ❑ Skeletal deformities: People who have cerebral palsy on only one side may have shortened limbs on the affected side. If not corrected by surgery or a device, this can lead to tilting of the pelvic bones and scoliosis (curvature of the spine).

# Cerebral Palsy

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- ❑ Joint contractures: People with spastic cerebral palsy may develop severe stiffening of the joints because of unequal pressures on the joints exerted by muscles of differing tone or strength.
- ❑ Mental retardation: Some, although not all, children with cerebral palsy are affected by mental retardation. Generally, the more severe the retardation, the more severe the disability overall.
- ❑ Seizures: About one third of people with cerebral palsy have seizures. Seizures may appear early in life or years after the brain damage that causes cerebral palsy. The physical signs of a seizure may be partly masked by the abnormal movements of a person with cerebral palsy.

# Cerebral Palsy

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- ❑ Speech problems: Speech is partly controlled by movements of muscles of the tongue, mouth, and throat. Some individuals with cerebral palsy are unable to control these muscles and thus cannot speak normally.
- ❑ Swallowing problems: Swallowing is a very complex function that requires precise interaction of many groups of muscles. People with cerebral palsy who are unable to control these muscles will have problems sucking, eating, drinking, and controlling their saliva. They may drool. An even greater risk is aspiration, the inhalation into the lungs of food or fluids from the mouth or nose. This can cause infection or even suffocation.
- ❑ Hearing loss: Partial hearing loss is not unusual in people with cerebral palsy. The child may not respond to sounds or may have delayed speech.

# Cerebral Palsy

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- ❑ Vision problems: Three quarters of people with cerebral palsy have strabismus, which is the turning in or out of one eye. This is due to weakness of the muscles that control eye movement. These people are often nearsighted. If not corrected, strabismus can lead to more severe vision problems over time.
- ❑ Dental problems: People with cerebral palsy tend to have more cavities than usual. This results from both defects in tooth enamel and difficulties brushing the teeth.
- ❑ Bowel and/or bladder control problems: These are caused by lack of muscle control.

# Cerebral Palsy

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Treatment of cerebral palsy:

- ❑ Physical, Occupational, and Speech Therapy
- ❑ Equipment such as AFOs, positioning devices, feeding supplies, and incontinence supplies
- ❑ Symptom specific treatments, for example:
  - Medications for spasticity
  - Seizure medications
  - Feeding tube
  - Special Education
  - Surgical procedures: tendon lengthening, dorsal rhizotomy (nerves are cut to reduce spasticity)

# Cortical Visual Impairment

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Cortical Visual Impairment is a condition in which the eyes work normally, but the person has visual problems because of brain damage.

Cortical Visual Impairment (CVI) is also sometimes referred to as Cortical Blindness, but people with CVI are rarely, if ever, completely blind.

# Cortical Visual Impairment

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## Visual and Behavioral Characteristics of Cortical Visual Impairment

- ❑ The individual prefers to look at old objects, not new, and lacks visual curiosity.
- ❑ Visually attends in near space only
- ❑ Difficulties with visual complexity/crowding (Individual performs best when one sensory input is presented at a time, when the surrounding environment lacks clutter, and the object being presented is simple.)
- ❑ Non-purposeful gaze/light gazing behaviors
- ❑ Distinct color preference (Preferences are predominantly red and yellow, but could be any color.)
- ❑ Visual field deficits (It is not so much the severity of the field loss, but where the field loss is located.)
- ❑ The individual's visual responses are slow, often delayed.
- ❑ Attraction to movement, especially rapid movements.
- ❑ The individual fails to blink at threatening motions.
- ❑ Look and touch occur as separate functions, e.g., child looks, turns head away from item, then reaches for it.
- ❑ Inefficient, highly variable visual sense

# Cortical Visual Impairment

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Children with Cortical Visual Impairment should receive vision therapy, also called Visual Instruction (VI), which helps improve their visual function.

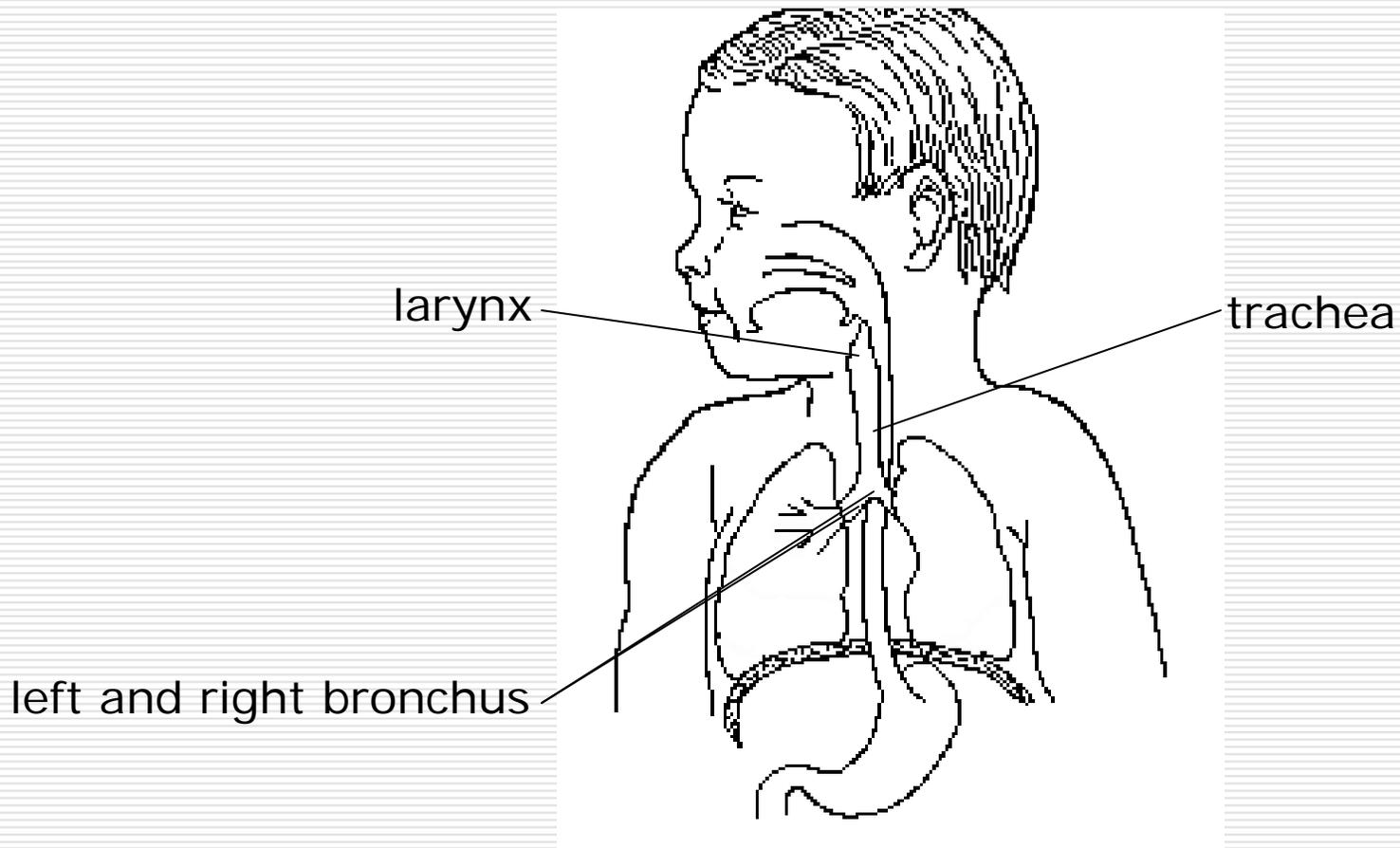
# Airway Obstructions

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- ❑ Laryngomalacia – weakness and floppiness of the walls of the larynx (voice box)
- ❑ Tracheomalacia – weakness and floppiness of the walls of the trachea (windpipe)
- ❑ Bronchomalacia – weakness and floppiness of the walls of the bronchial tubes

# Airway Obstructions

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# Airway Obstructions

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Depending upon the location and severity of the malacia, it can cause anything from stridor or wheezing up to the need for a tracheostomy tube to breathe.

# GERD

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## Gastro-Esophageal Reflux Disease

Children with developmental or neurological conditions, such as prematurity or cerebral palsy, are more at risk for GERD and can have more severe, lasting symptoms.

# GERD

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## □ Symptoms

- fussiness, crying, irritability
- spitting up, vomiting
- poor sleep habits, frequent wakening
- neck and back arching/Sandifer's Syndrome

# GERD

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## Sandfifer's syndrome

- Symptom of reflux in children under 2 years of age and in older children with spasticity
- During and after a feeding, child will turn head and neck to one side and legs to the opposite side, with back arched and body extended. This posture is done repeatedly so that it can sometimes look like a seizure.
- As a result, children may develop torticollis, and because of their lack of eye contact, stiffness, and fussiness after feeding may have problems bonding with caregivers

# GERD

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- Complications/Symptoms of more severe disease
  - Poor weight gain, weight loss, or failure to thrive
  - Oral aversion
  - Respiratory problems – pneumonia, bronchitis, wheezing, aspiration, asthma, night-time cough, apnea
  - Torticollis
  - Difficulty bonding

# GERD

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## □ Treatments

- smaller, more frequent feedings
- thickened feedings
- non-constrictive clothing
- upright for 30 minutes after feeding
- medications
- tube feedings
- Nissen fundoplication
- physical therapy for torticollis/muscle problems related to neck and back arching and stiffness/extension
- speech/feeding therapy for swallowing and for oral aversion

# Pervasive Developmental Disorders

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Pervasive Developmental Disorders are defined as "severe and pervasive impairment in several areas of development."

The five disorders under PDD are:

## Autistic Disorder

Characterized by impaired social interaction and communication skills, and a limited range of activities and interests

## Asperger's Disorder

Children with autistic behaviors but well-developed language skills are often diagnosed with Asperger's Syndrome.

## Childhood Disintegrative Disorder (CDD)

Children who develop normally and then suddenly deteriorate between the ages of 3 to 10 years and show marked autistic behaviors may be diagnosed with childhood disintegrative disorder.

## Rett's Disorder

Girls with autistic symptoms may be suffering from Rett syndrome, a sex-linked genetic disorder characterized by social withdrawal, regressed language skills, and hand wringing.

## PDD-Not Otherwise Specified (PDD-NOS)

Children with some symptoms of autism, but not enough to be diagnosed with classical autism, are often diagnosed with PDD-NOS.