Pediatric Neurological Disorders

Dr. Gary Mumaugh

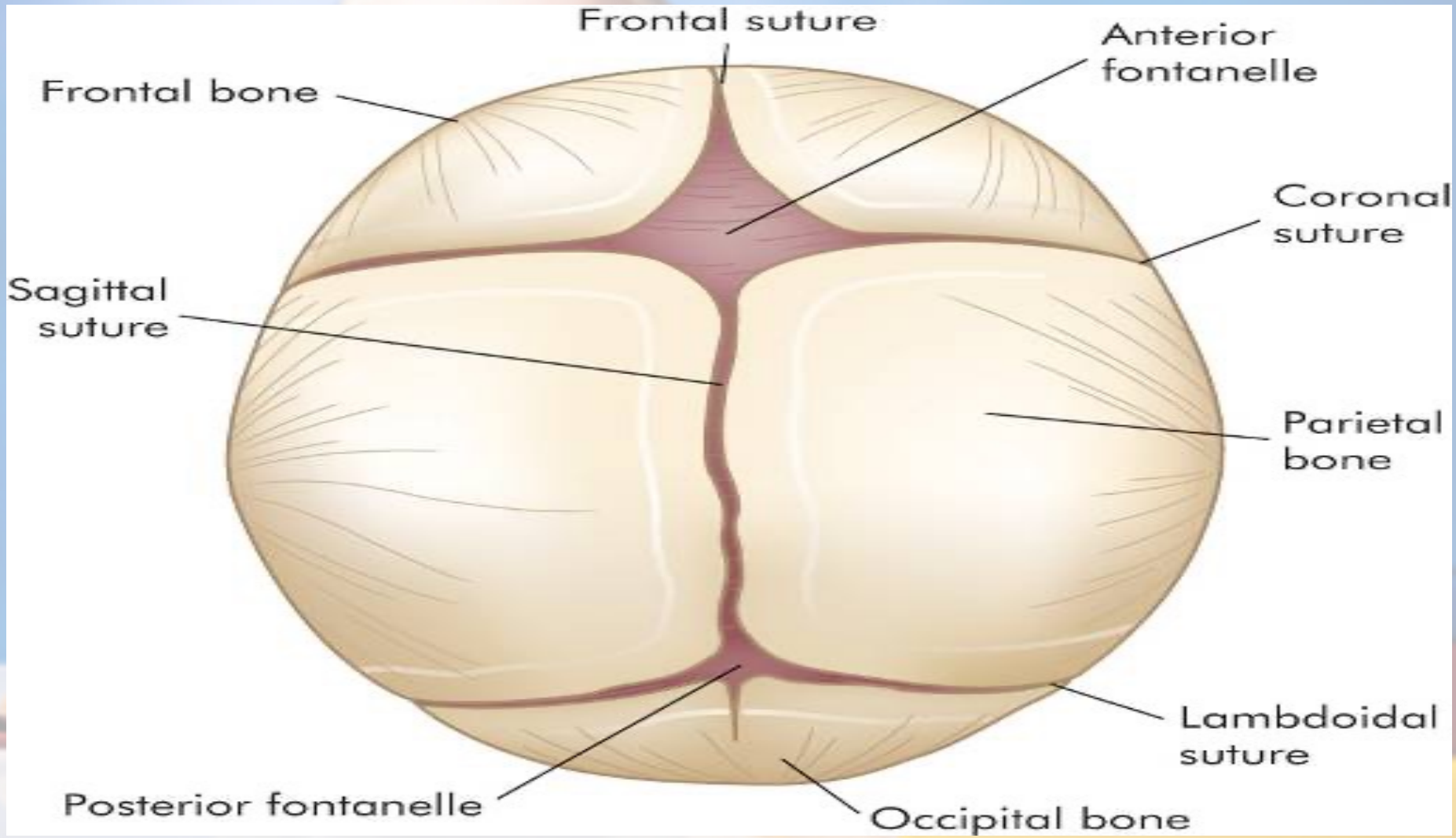
Growth and Development of Nervous System

- Develops from a dorsal thickening of the ectoderm (neural plate)
  - Neural groove and folds
  - Neural tube
  - Neural crest

Growth and Development of Nervous System

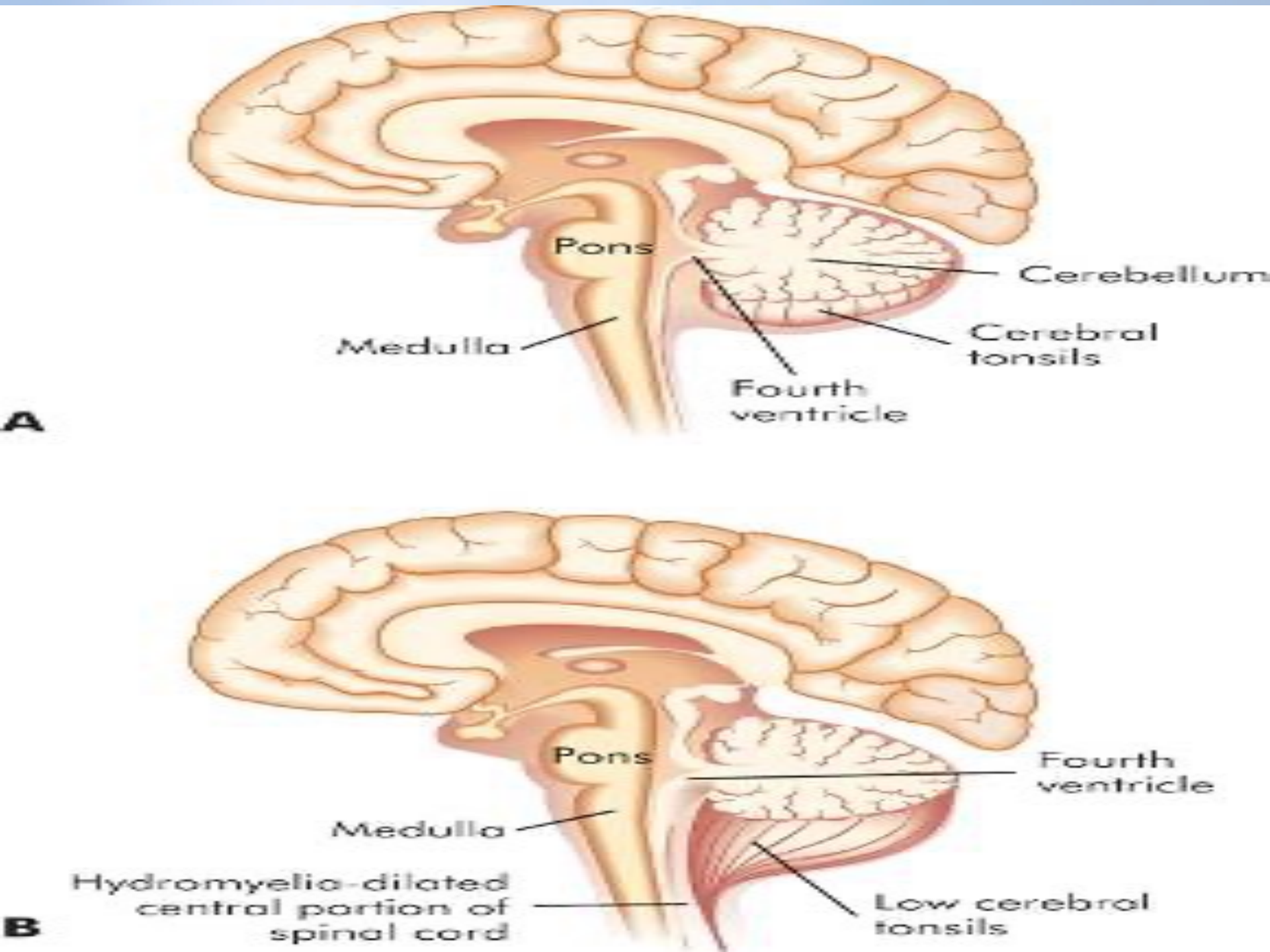
- Mesoderm
  - Blood vessels, microglial cells, dural and arachnoid layers of the meninges, the capsule of some peripheral nerve endings, and nerve coverings

Cranial Sutures and Fontanelles



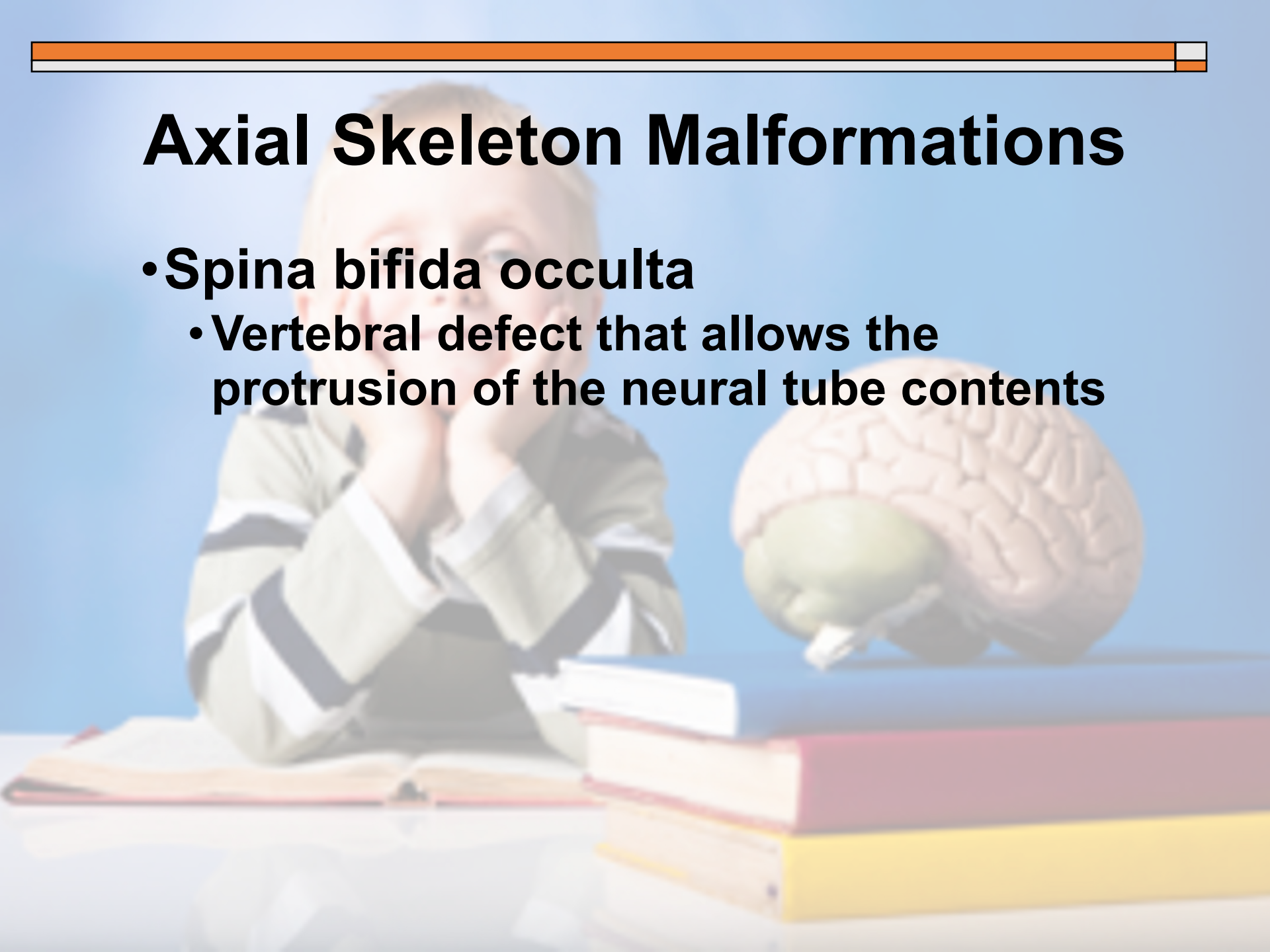
Structural Malformations

- Defects of neural tube closure
  - Anencephaly
  - Encephalocele
  - Meningocele
  - Myelomeningocele
    - Arnold-Chiari type II malformation
    - Tethered cord syndrome


Pons
Cerebellum
Medulla
Cerebral tonsils
Fourth ventricle
A
Pons
Fourth ventricle
Medulla
Hydromyelia-dilated central portion of spinal cord
Low cerebral tonsils
B

Axial Skeleton Malformations

- Spina bifida occulta
  - Vertebral defect that allows the protrusion of the neural tube contents



Axial Skeleton Malformations

- Cranial deformities
  - Acrania
  - Craniosynostosis
  - Microcephaly
  - Congenital hydrocephalus
    - Dandy-Walker deformity
    - Macewen sign (“cracked-pot” sign)

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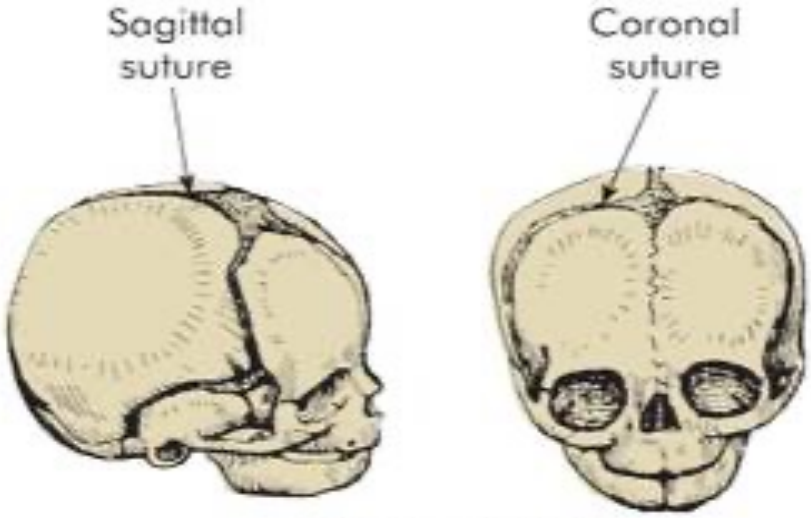
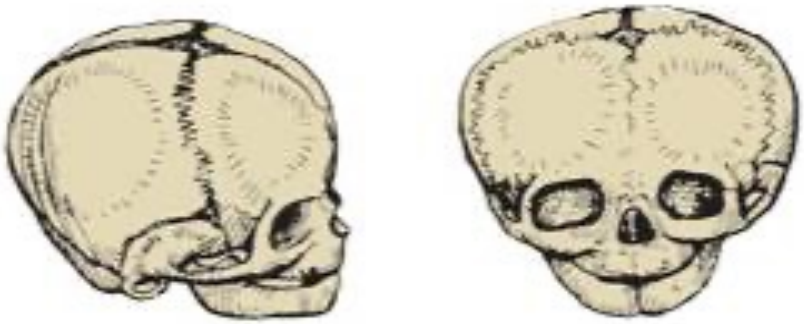


NORMAL SKULL



BRACHYCEPHALY

MICROCEPHALY AND CRANIOSTENOSIS

OXYCEPHALY OR ACROCEPHALY

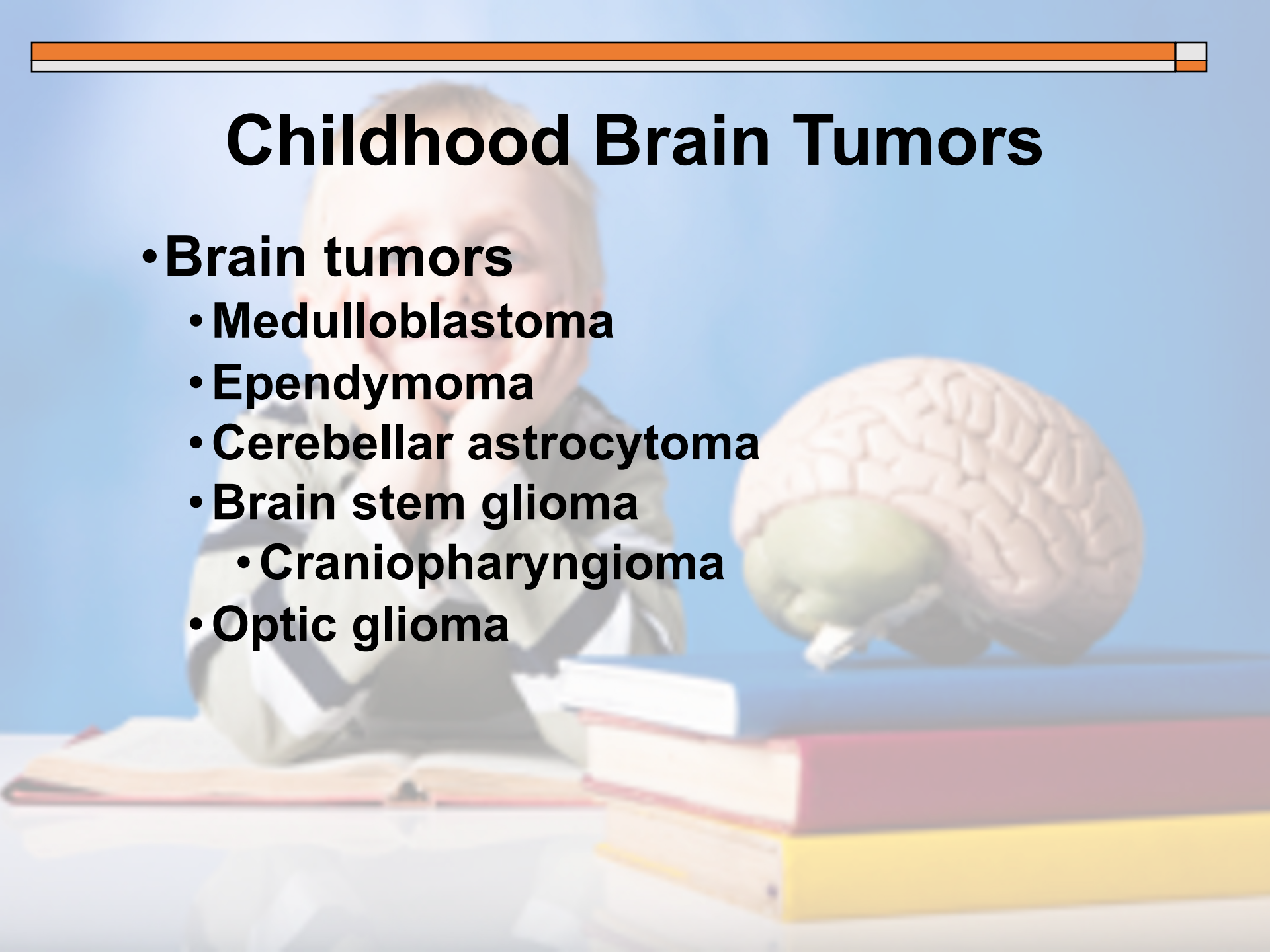
SCAPHOCEPHALY OR DOLICHOCEPHALY

PLAGIOCEPHALY

From Hockenberry MJ: *Wong's nursing care of infants and children*, ed 7, St Louis, 2003, Mosby.

Childhood Brain Tumors

- Brain tumors
  - Medulloblastoma
  - Ependymoma
  - Cerebellar astrocytoma
  - Brain stem glioma
    - Craniopharyngioma
  - Optic glioma



Craniopharyngiomas

- Located adjacent to the sella turcica (structure containing the pituitary gland), often considered to lie supratentorial
- Considered to have benign properties but is life threatening because of its location near vital structures
- 4.9% of brain tumors in children
  5%

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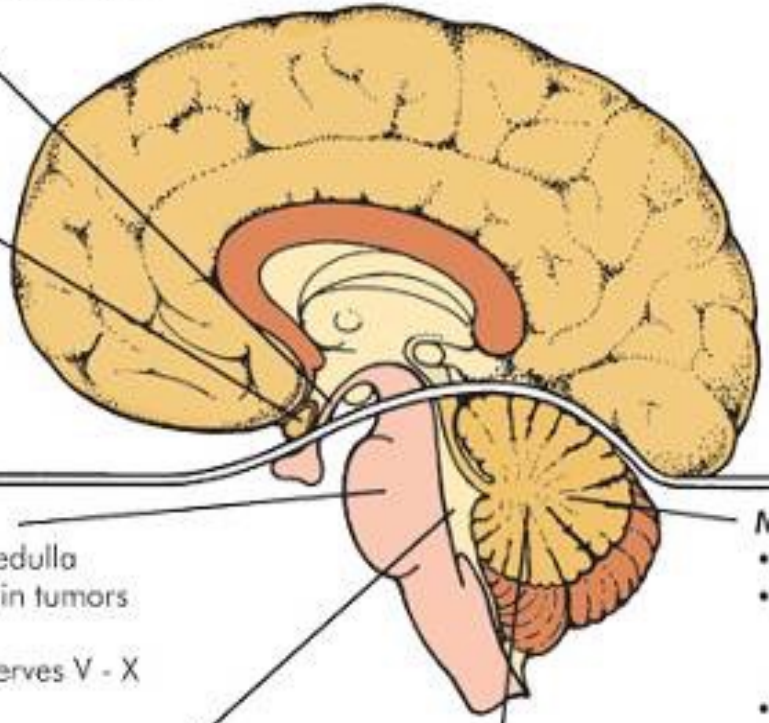


Optic nerve gliomas
- Most often a low-grade astrocytoma
  6%

Cerebral tumors
- Astrocytomas invade surrounding structures but grow slowly
  8%
- Ependymomas arise from lining tissue of lateral ventricle
  6%

Supratentorial

Brain stem gliomas
- Arise from pons or medulla
- 10% of childhood brain tumors
- Slow growing
- May involve cranial nerves V - X
  10%

Infratentorial ependymomas
- Arise from lining tissue of fourth ventricle
- Comprise 13% of childhood brain tumors together with supratentorial ependymomas
  13%

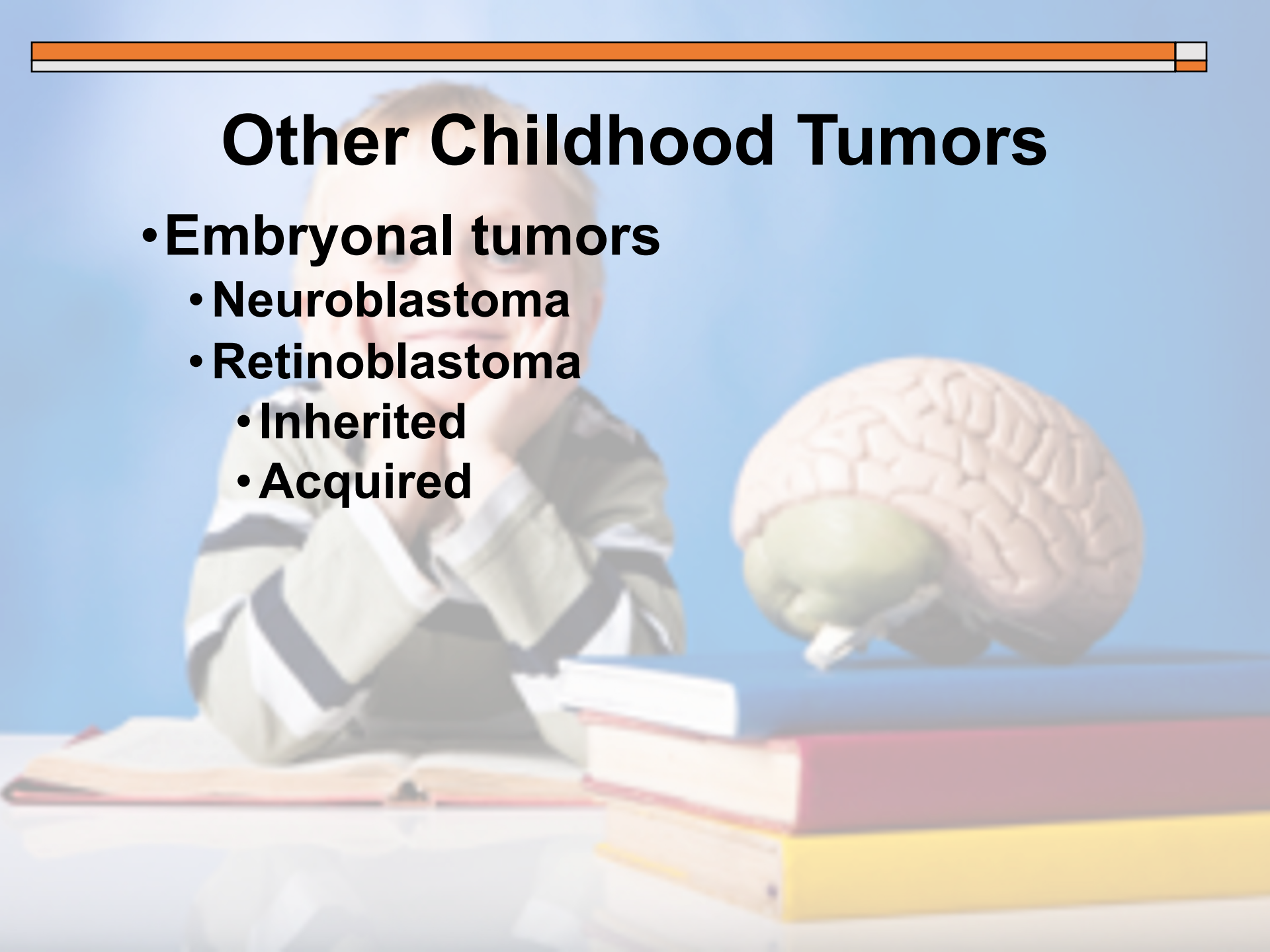
Cerebellar astrocytomas
- Most common brain tumor of childhood (20%)
- Slow growing
- Grading system I to IV with I and II less malignant than III and IV
  20%

Medulloblastomas
- Arise from cerebellum
- Can invade fourth ventricle, subarachnoid space, and cerebrospinal fluid pathways
- 18% of brain tumors in children
- Fast growing
- Arise from embryonic cerebellum
  18%

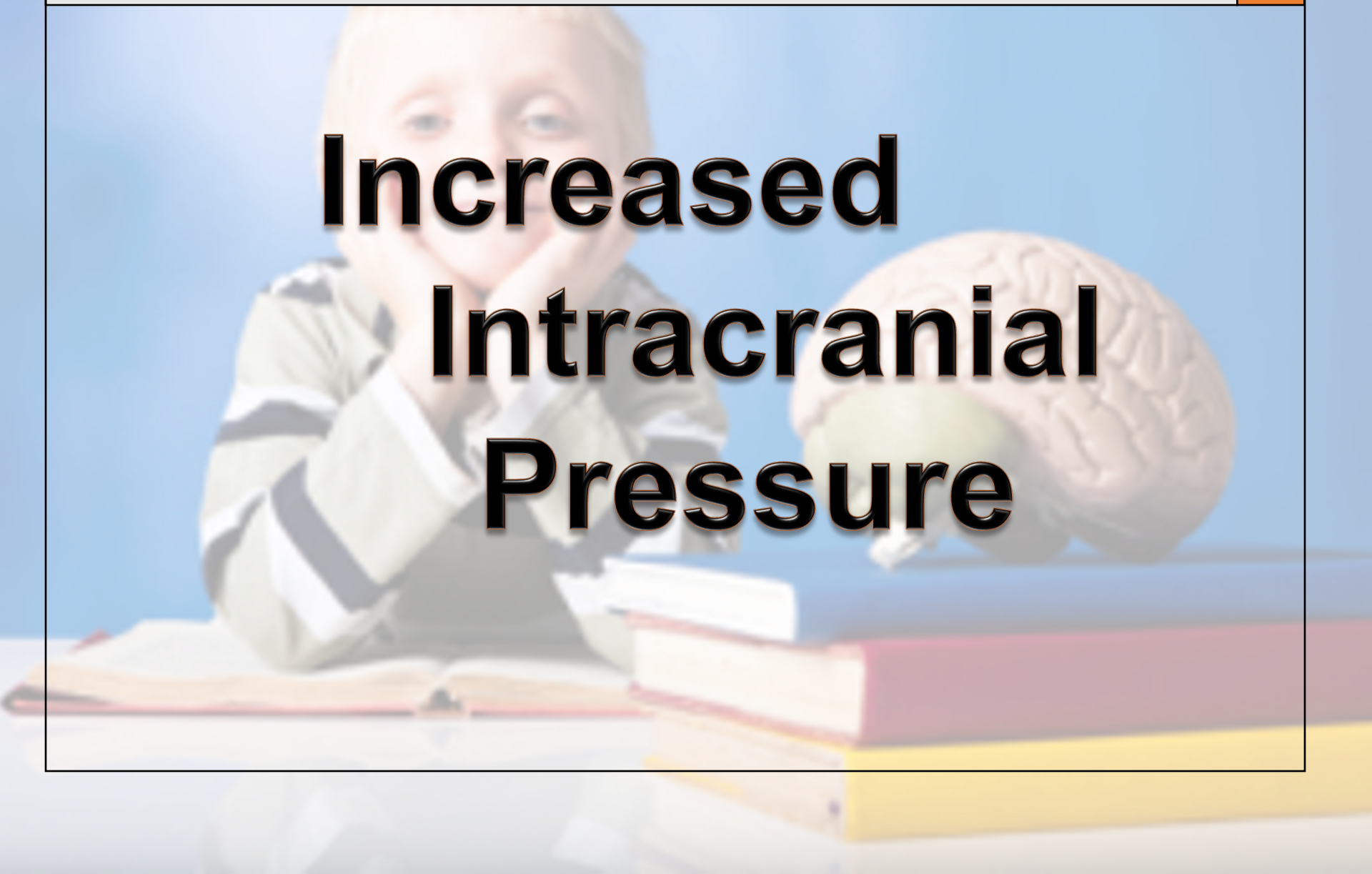
Infratentorial

Other Childhood Tumors

- Embryonal tumors
  - Neuroblastoma
  - Retinoblastoma
    - Inherited
    - Acquired



Increased Intracranial Pressure



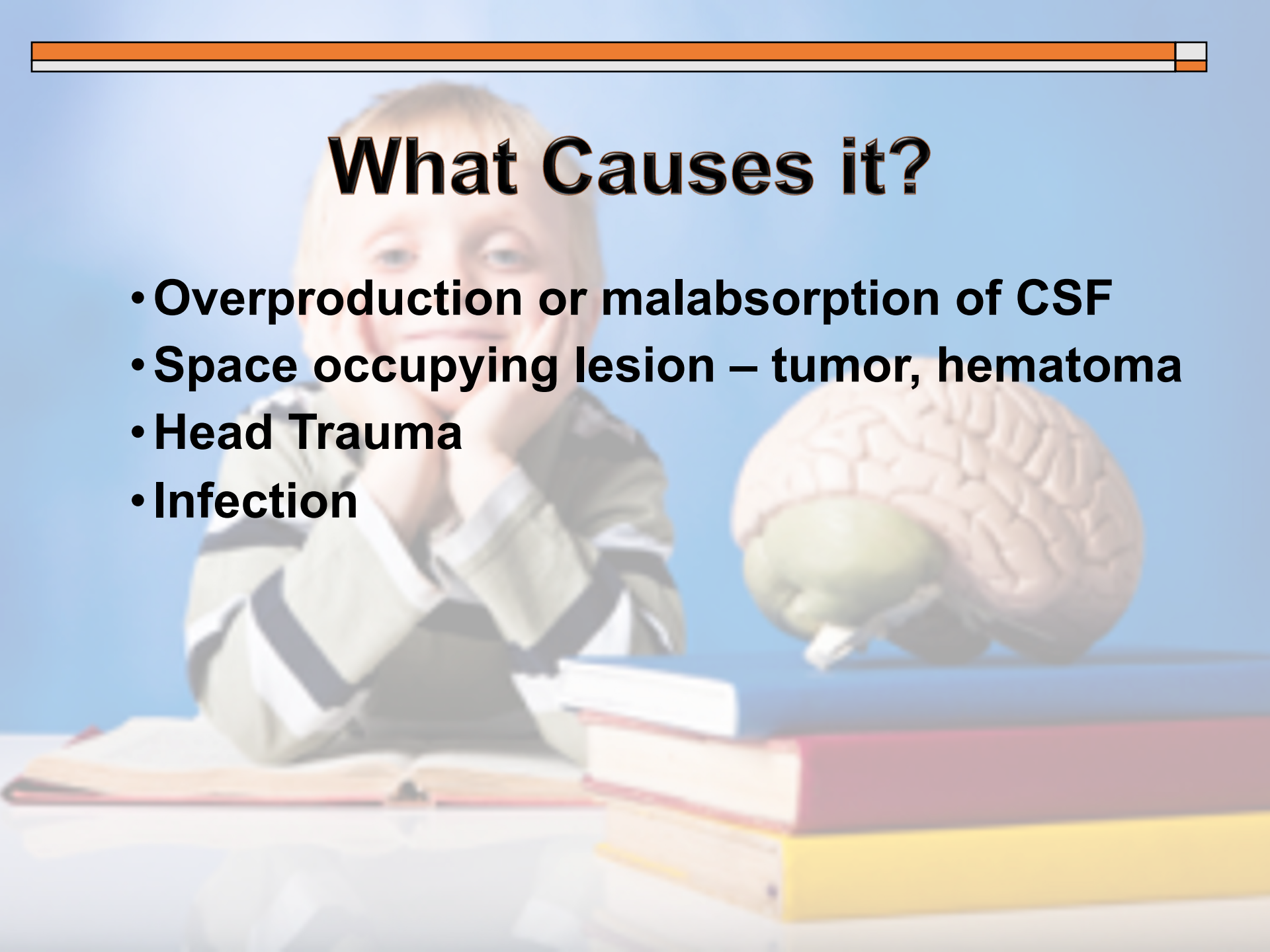
Increased Intracranial Pressure (IICP)

What is it?

- Increased ICP results from a disturbance in the auto-regulation of the pressure exerted by the blood, brain, cerebrospinal fluid, and other space-occupying fluid/mass within the central nervous system.
- Increased ICP is defined as pressure sustained at 20 mm Hg or higher.

What Causes it?

- Overproduction or malabsorption of CSF
- Space occupying lesion – tumor, hematoma
- Head Trauma
- Infection

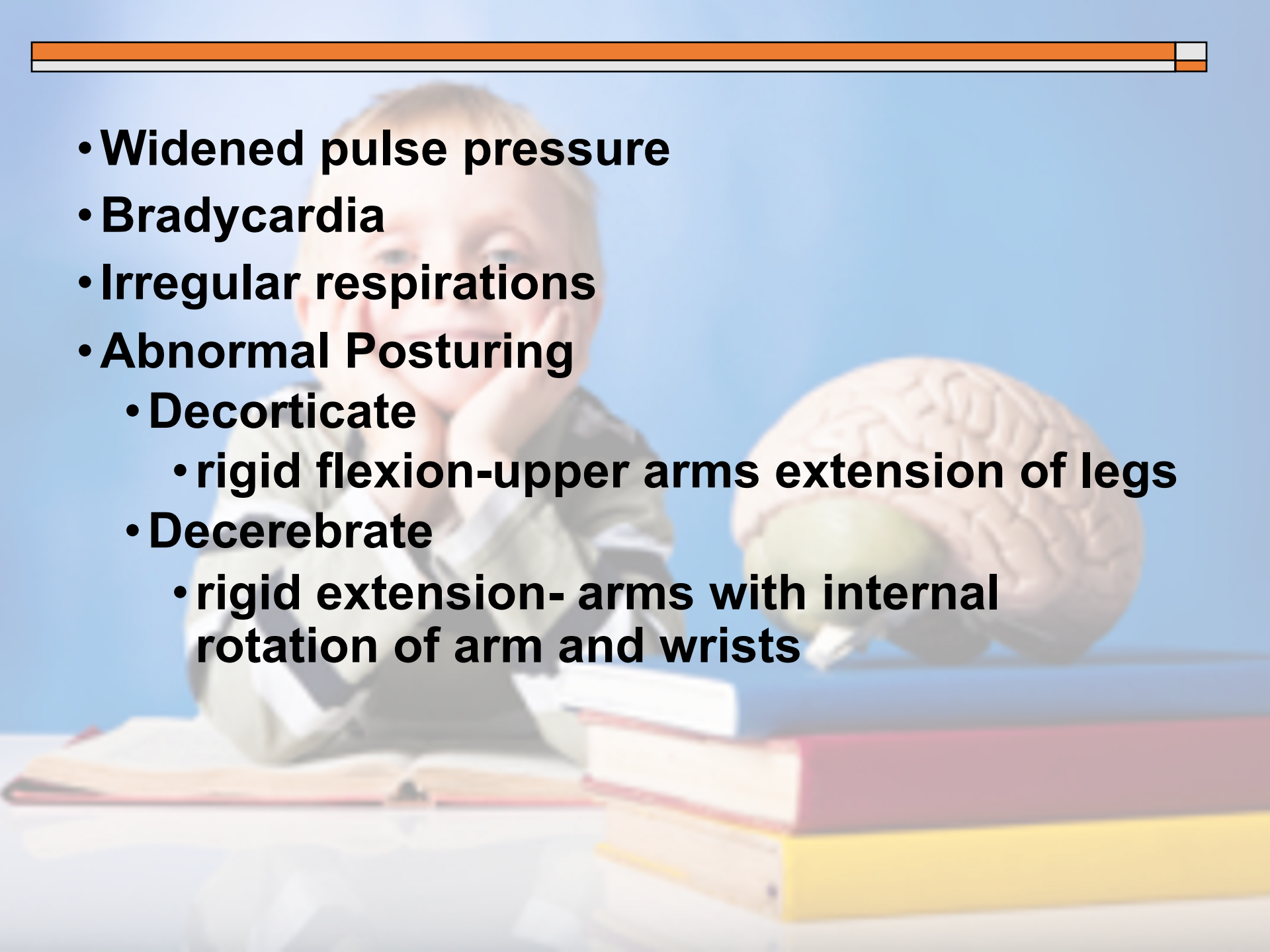


Clinical Manifestations: Infant

- Irritability and restlessness; high-pitched cry
- Full to bulging fontanels; Increase in FOC
- Poor feeding, poor sucking
- Prominence of frontal portion of the skull with distension of superficial scalp veins
- Nuchal rigidity
- Nonreactive; unequal pupils
- Seizures (late sign)

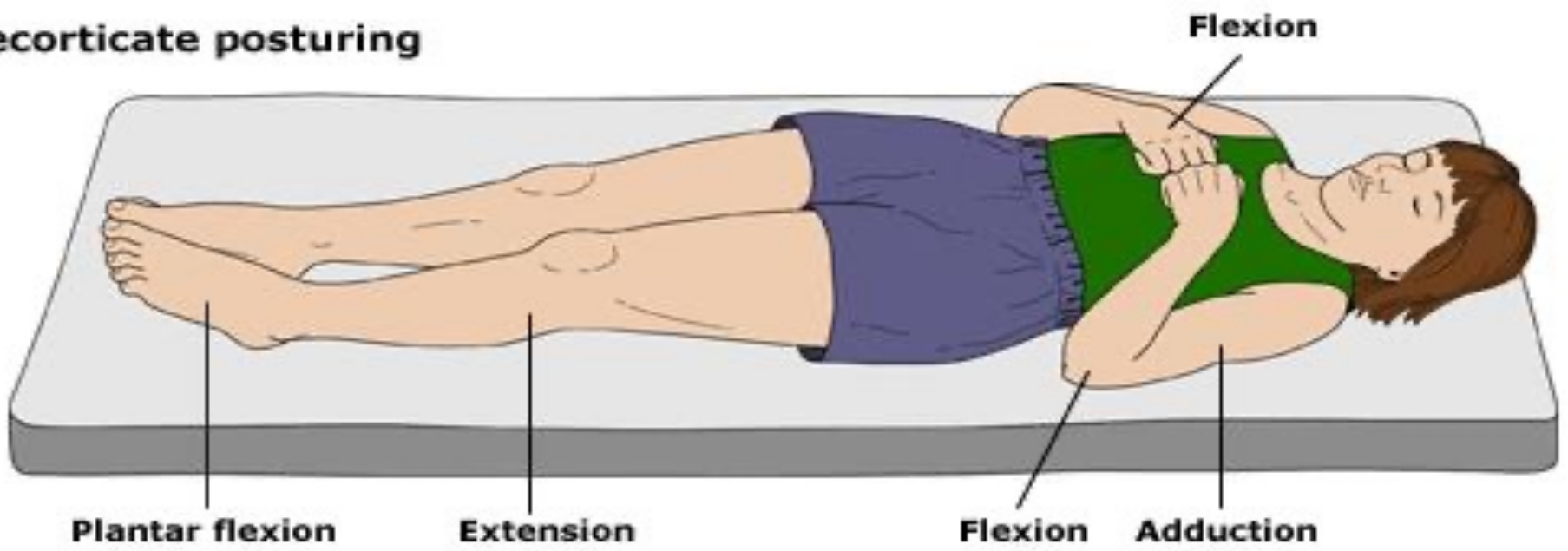
Clinical Manifestations: Child

- Headache
- Visual disturbances - diplopia
- Nausea and Vomiting
- Dizziness or vertigo
- Irritability, lethargy, mood swings
- Ataxia, lower extremity spasticity
- Nuchal rigidity
- Deterioration in school performance, or cognitive ability

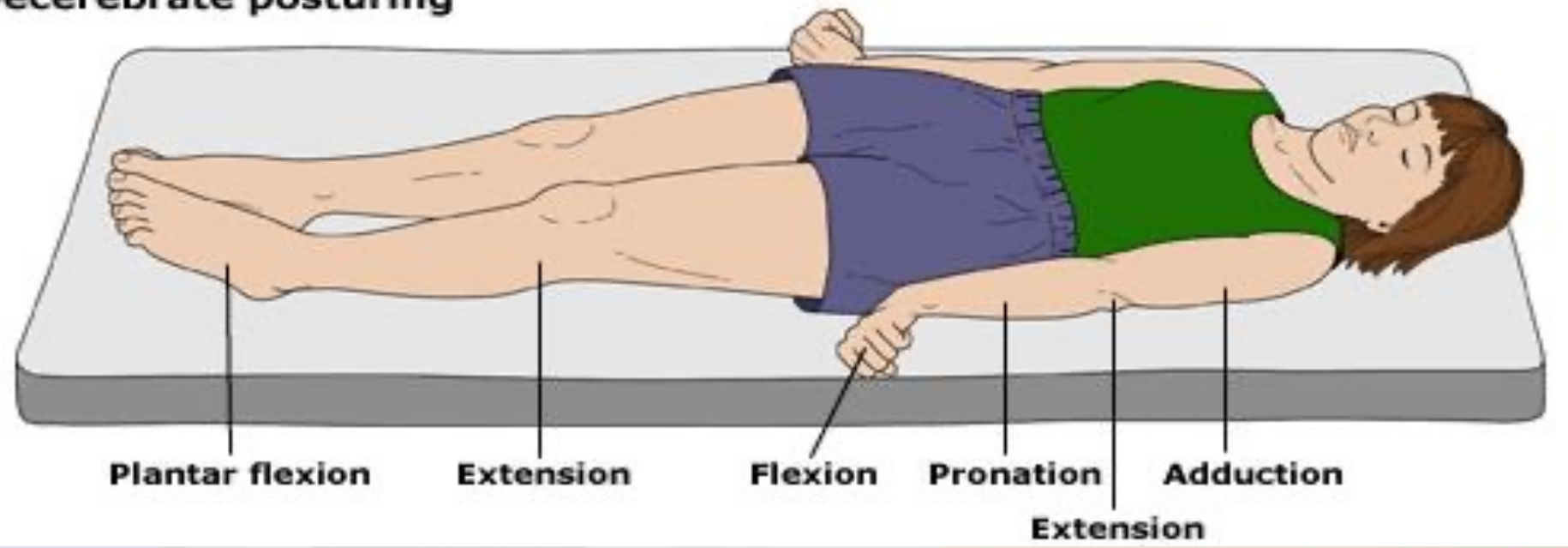


- Widened pulse pressure
- Bradycardia
- Irregular respirations
- Abnormal Posturing
  - Decorticate
    - rigid flexion-upper arms extension of legs
  - Decerebrate
    - rigid extension- arms with internal rotation of arm and wrists

Decorticate posturing



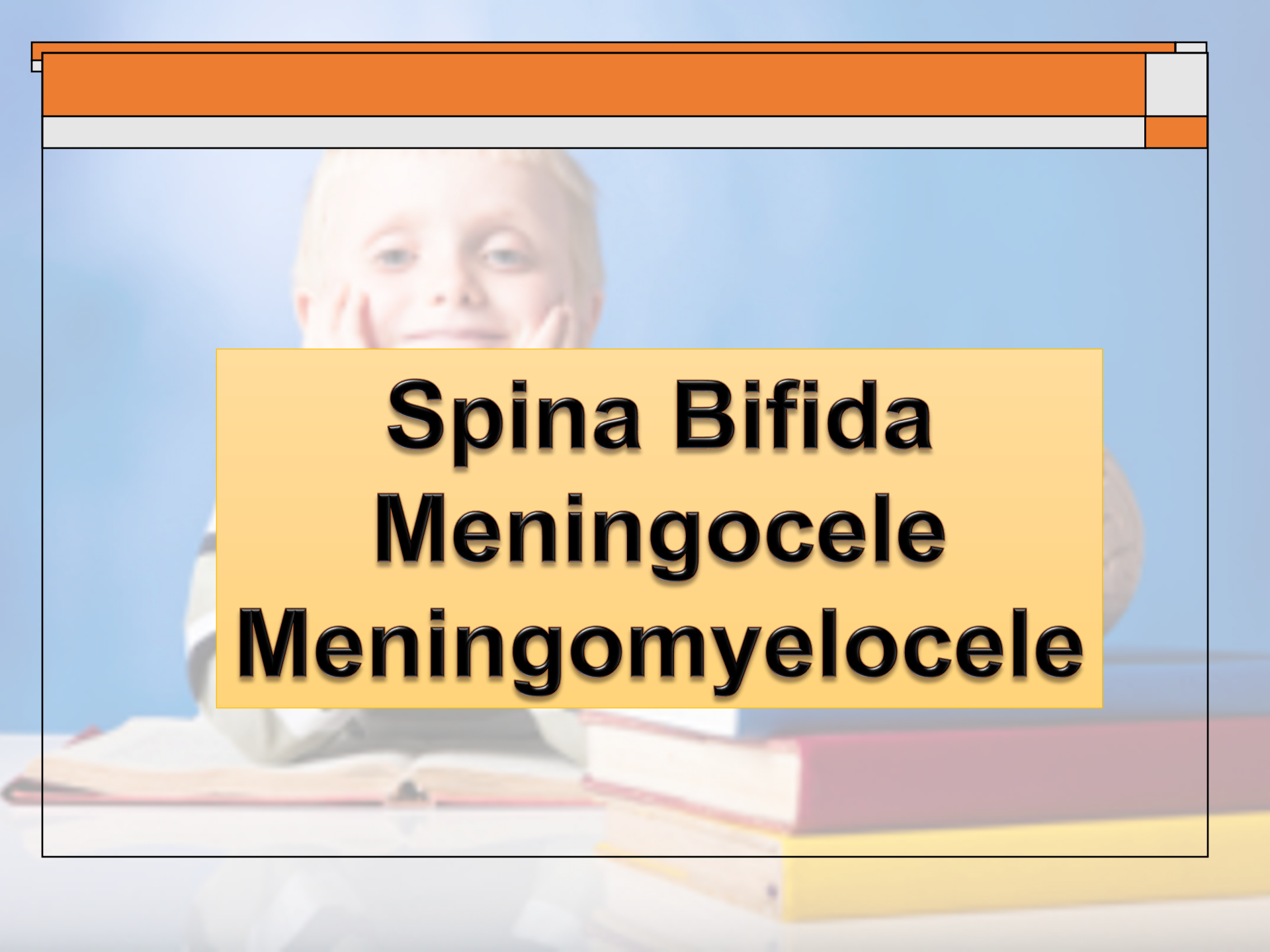
Decerebrate posturing



Diagnosis

- Blood studies
- CT or MRI
- EEG
- Lumbar puncture – may or may not be done

Why?



Spina Bifida
Meningocele
Meningomyelocele

What is the difference?

- Spina Bifida
- Meningocele:
- Myelomeningocele:

Spina Bifida
Meningocele
Meningomyelocele

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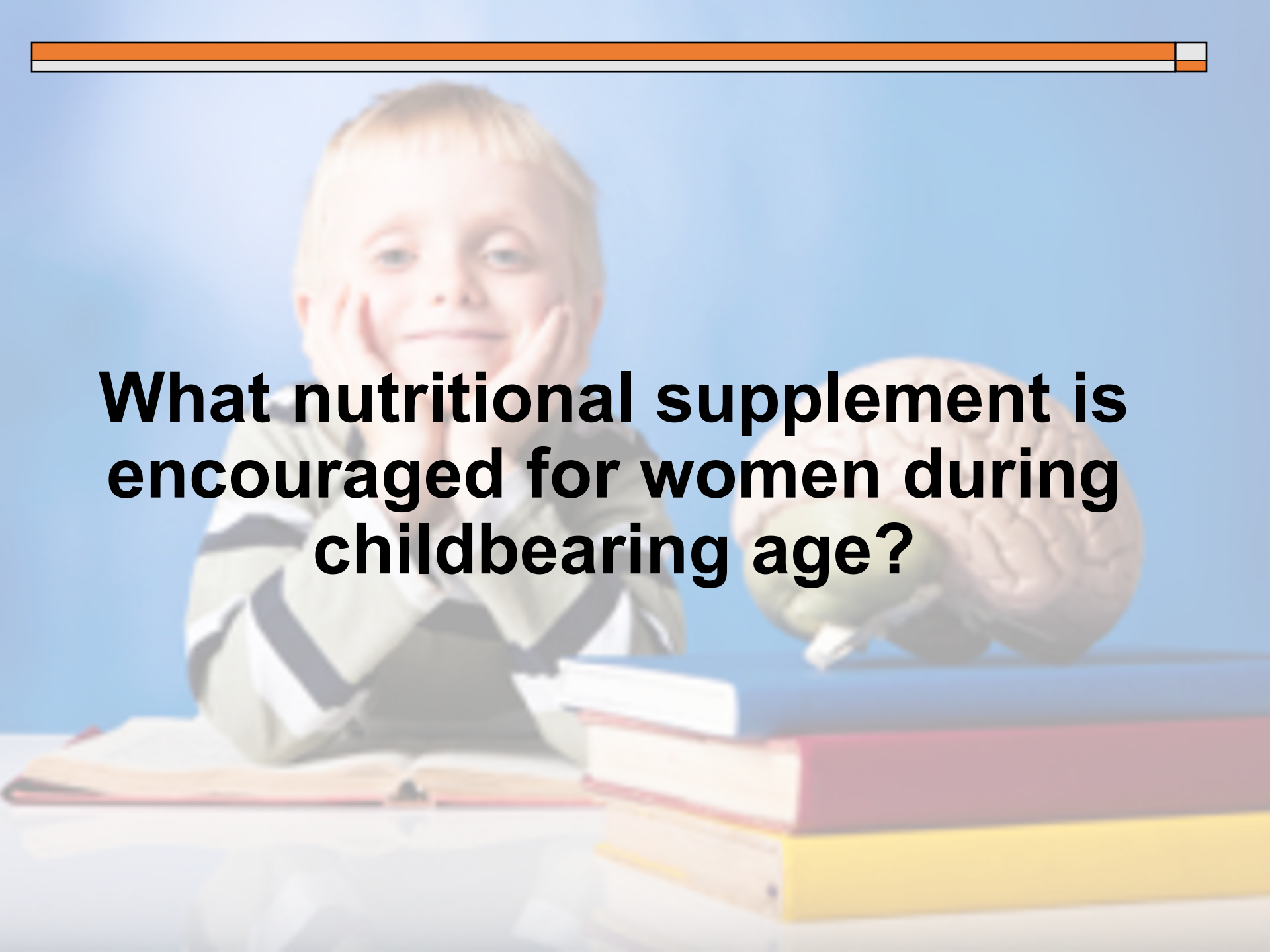


What nutritional supplement is encouraged for women during childbearing age?

Clinical Manifestations

- Visualization of the defect
- Motor sensory, reflex and sphincter abnormalities
- Flaccid paralysis of legs- absent sensation and reflexes, or spasticity
- Malformation
- Abnormalities in bladder and bowel function

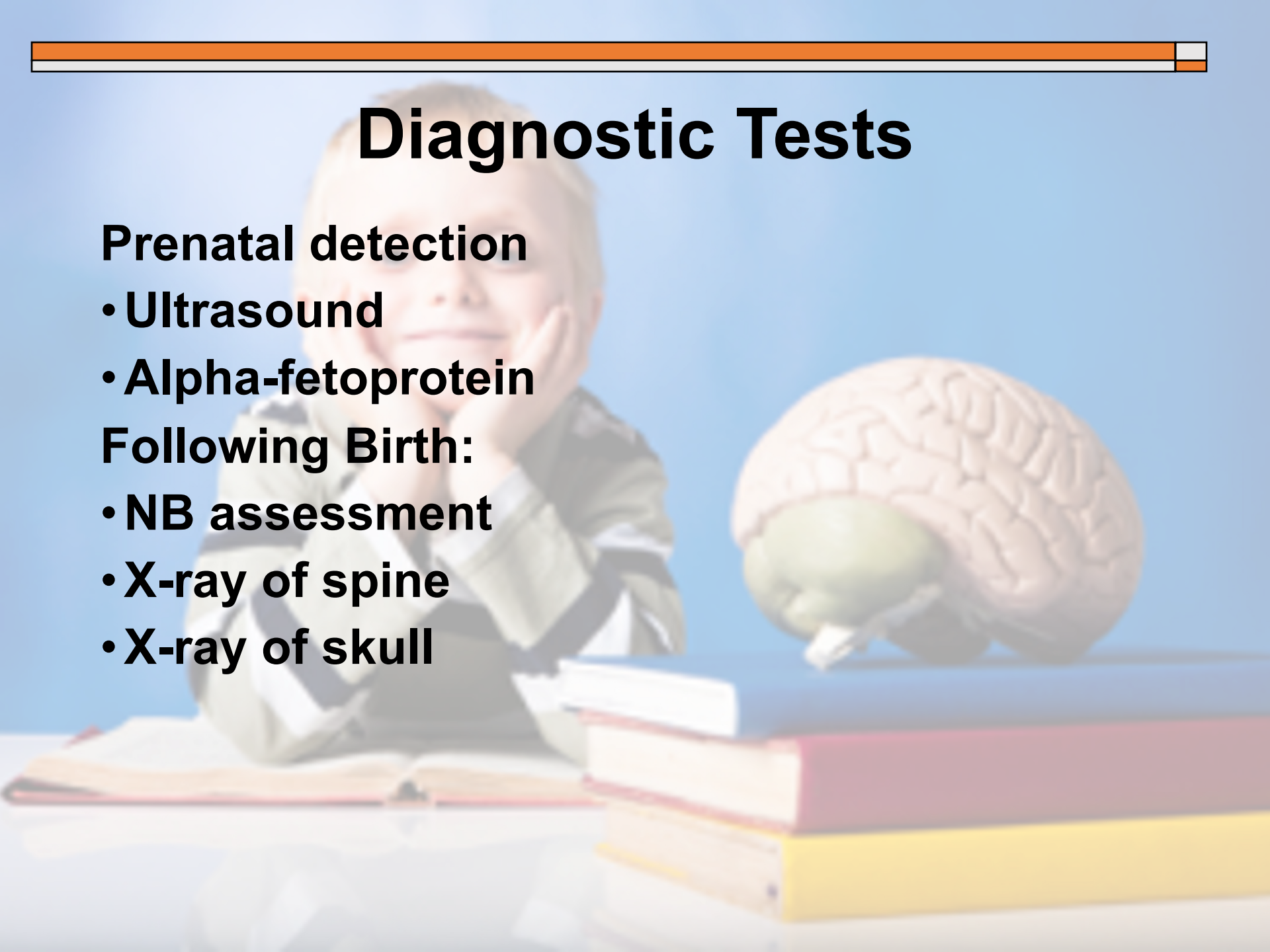
Diagnostic Tests

Prenatal detection

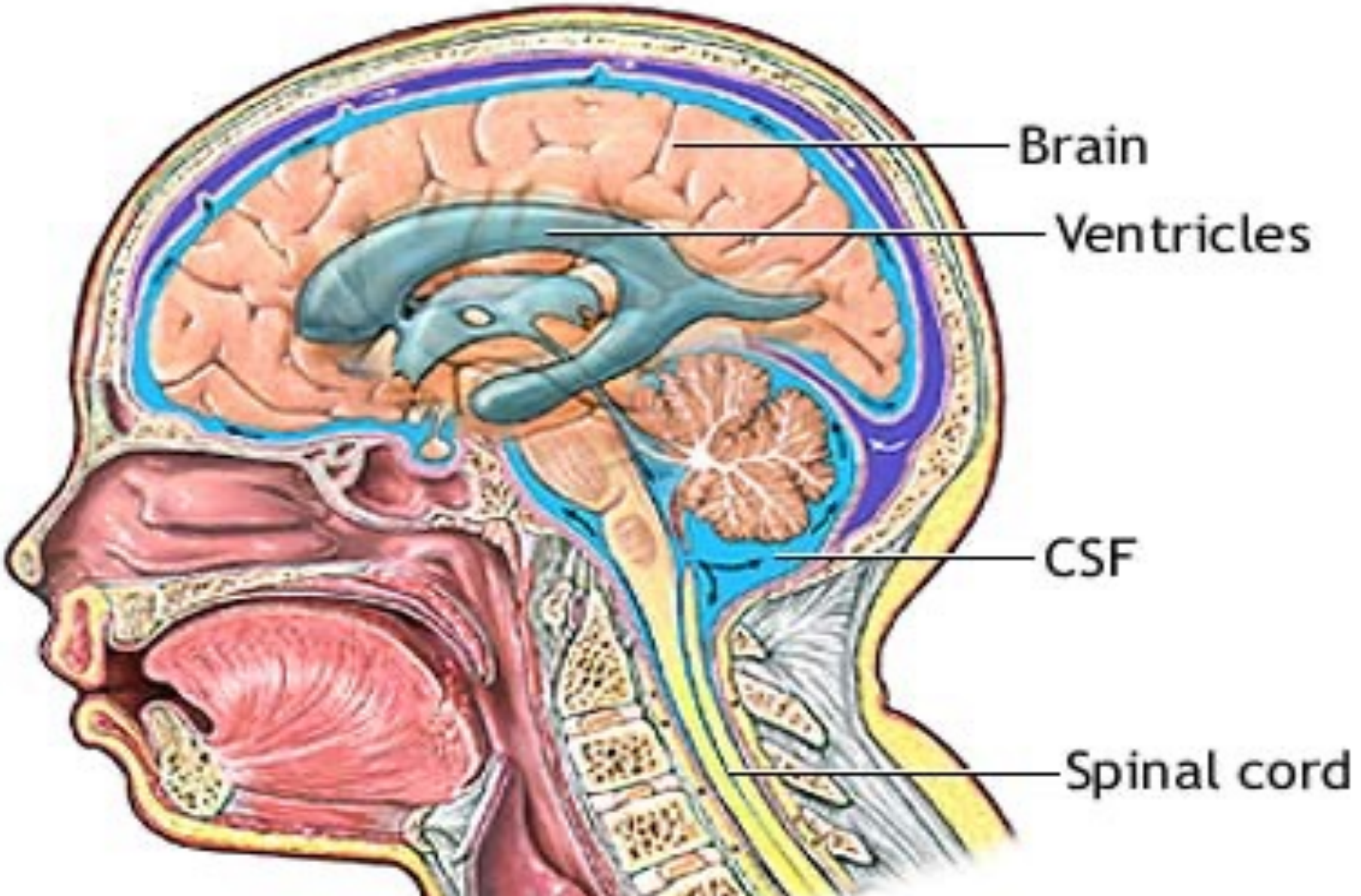
- **Ultrasound**
- **Alpha-fetoprotein**

Following Birth:

- **NB assessment**
- **X-ray of spine**
- **X-ray of skull**

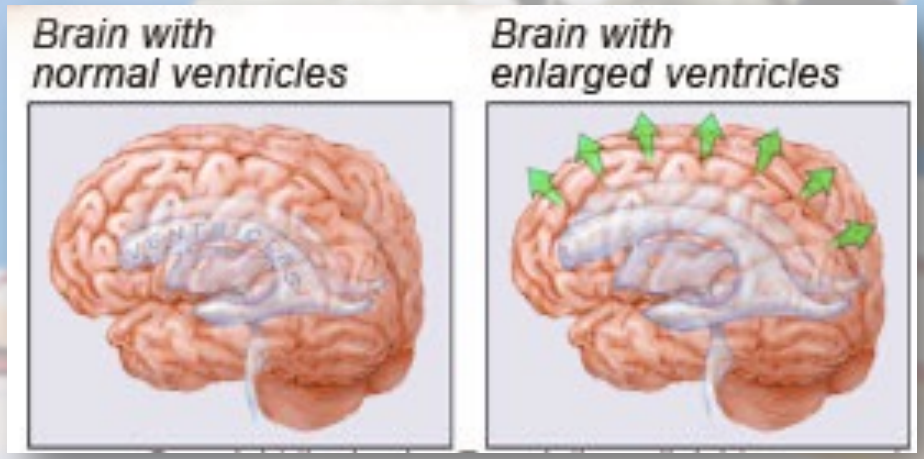


Hydrocephalus



Etiology and Pathophysiology

- Imbalance between the production and absorption of cerebral spinal fluid causing accumulation of fluid in the ventricles



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Clinical Manifestations

Infants

- **Increase in FOC**
- **Frontal enlargement or bossing**
- **Head larger than face**
- **Translucent skin**
- **Wide palpable suture lines**
- **Bulging Fontanels**
- **Eyes -wide bridge between**
- **Behavior changes**

Clinical Manifestations

Children:

- **Depressed eyes; strabismus**
- **“Setting Sun” Eyes**
- **Pupils sluggish, with unequal response to light**
- **Headache with nausea and vomiting that may be projectile**
- **S & S of IICP**

Diagnostic Tests

- MRI/ CT scan
- Skull X-ray
- FOC
- Transillumination

**lumbar puncture very dangerous and usually NOT done

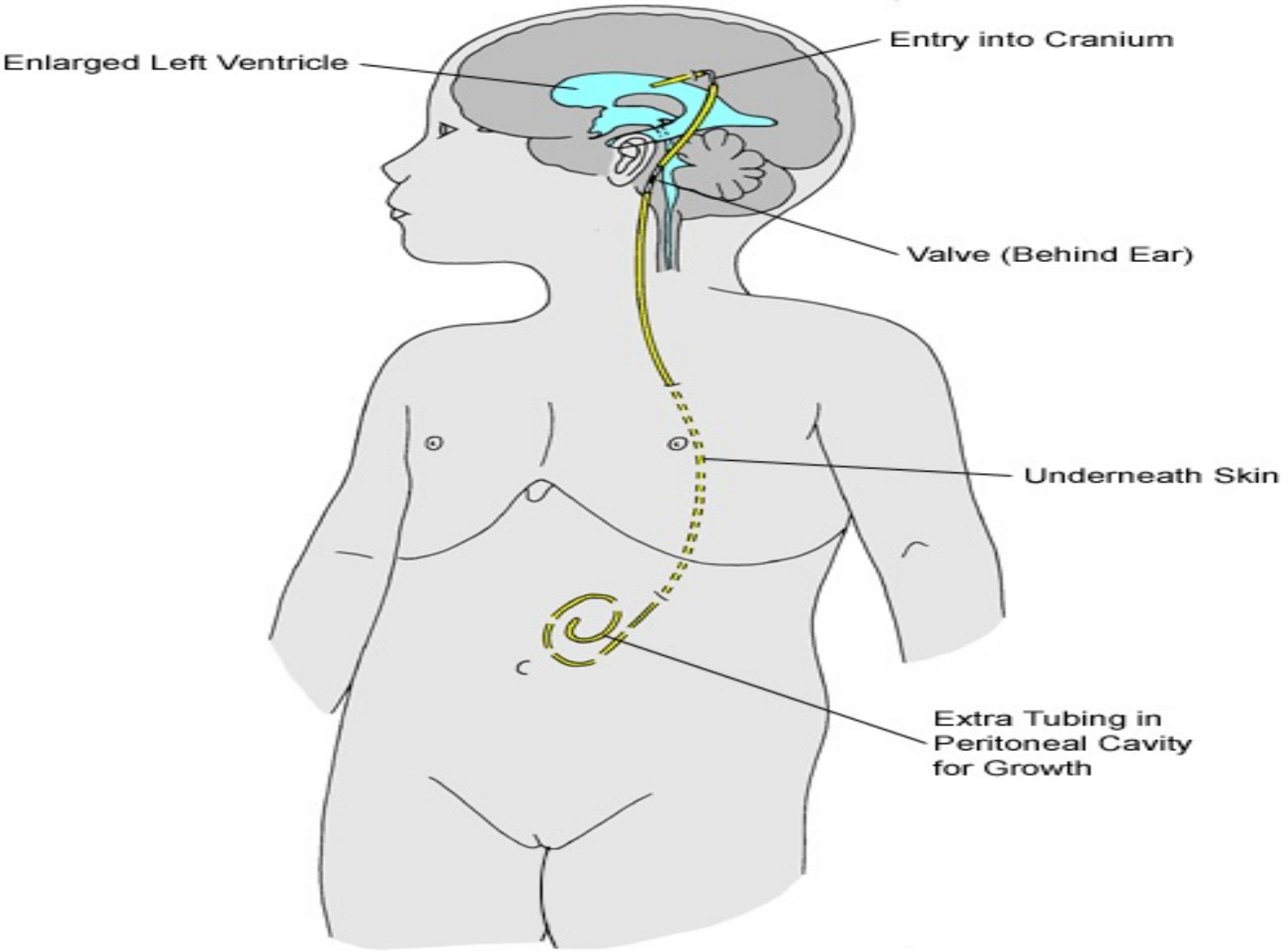
Goal of treatment

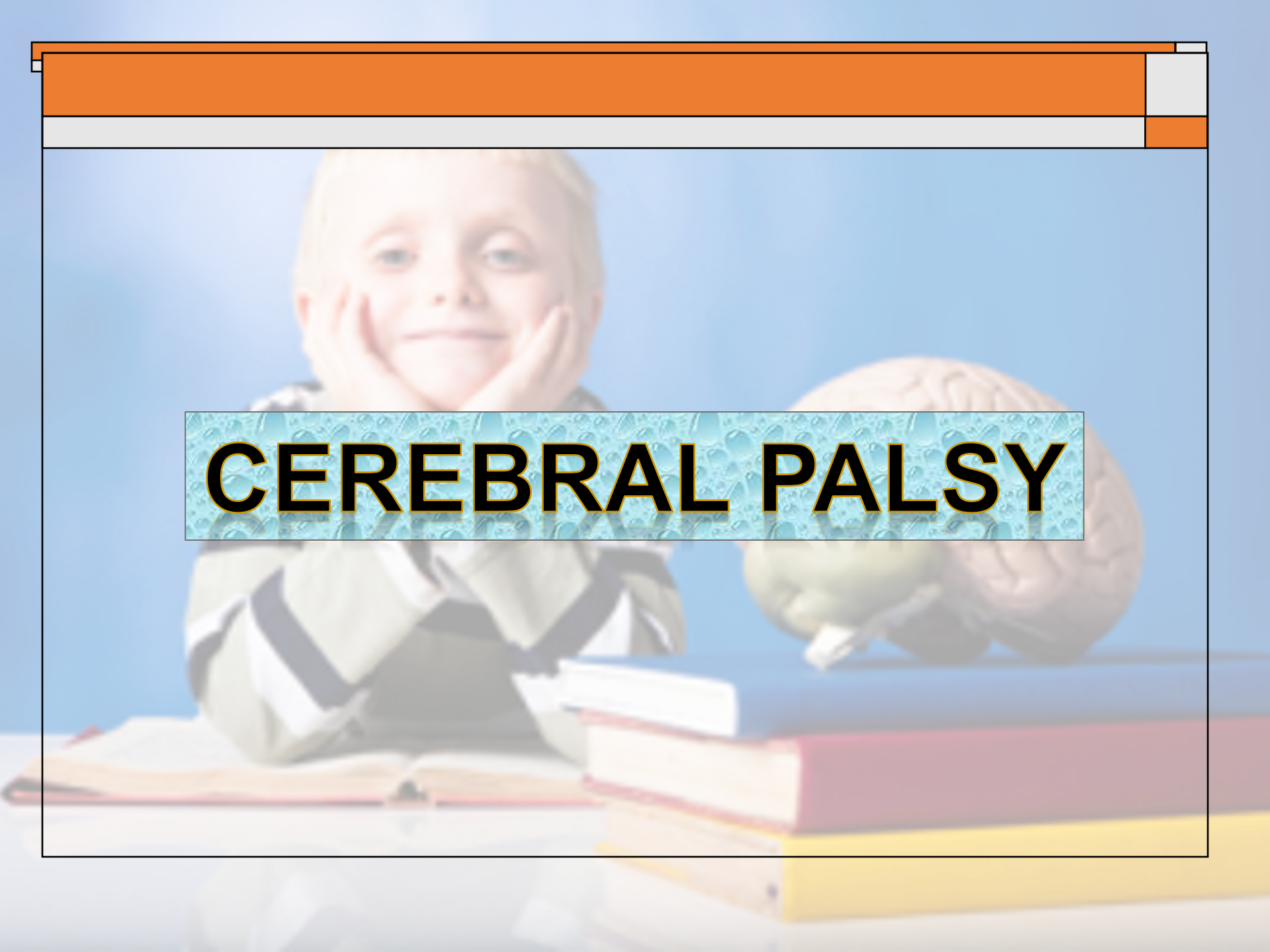
- Prevent further CSF accumulation
- Reduce disability and death
- Bypass the blockage and drain the fluid from the ventricles to an area where it may be reabsorbed into the circulation

Interventions: Surgical

- Ventricular endoscopy or laser
- Shunting to bypass the point of obstruction by shunting the fluid to another point of absorption
  - Atrioventricular
  - Ventricular peritoneal

Ventriculoperitoneal Shunt Placement




CEREBRAL PALSY

Cerebral Palsy (CP)

What is it associated with?

- Preterm
- Birth asphyxia
- Low Apgar
- Poor feeder
- Weak cry as a newborn
- Shaken baby syndrome
- Intrauterine anoxia – placental perfusion decreased

Assessment

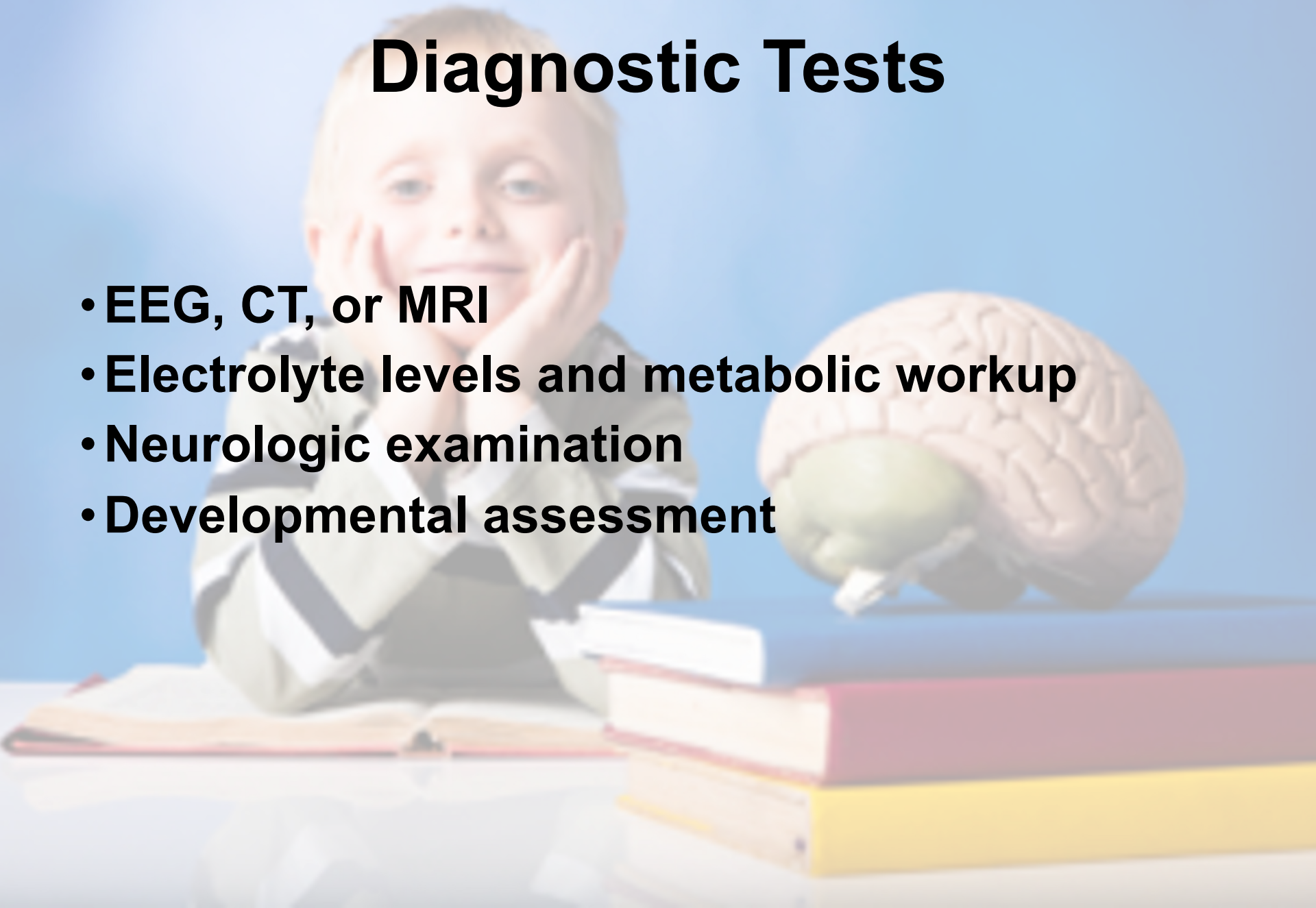
Determining diagnosis or extent of involvement in an infant can be difficult –may be recognizable only when child is older and attempts more complex motor skills, such as walking

- Jittery (easily startled)
- Weak cry (difficult to comfort)
- Experience difficulty with eating (muscle control of tongue and swallow reflex)
- Uncoordinated or involuntary movements (twitching and spasticity)
- Abnormal newborn reflexes – prolonged

Assessment

- Alterations in muscle tone
  - Abnormal resistance
  - Keeps legs extended or crossed
  - Rigid and unbending
- Abnormal posture
  - Do not crawl on knees, scoot on back
  - When try to walk, walk with toes first as in plantar flexion
  - Scissoring and extension (legs feet in plantar flexion)
  - Persistent fetal position (>5 months)

Diagnostic Tests

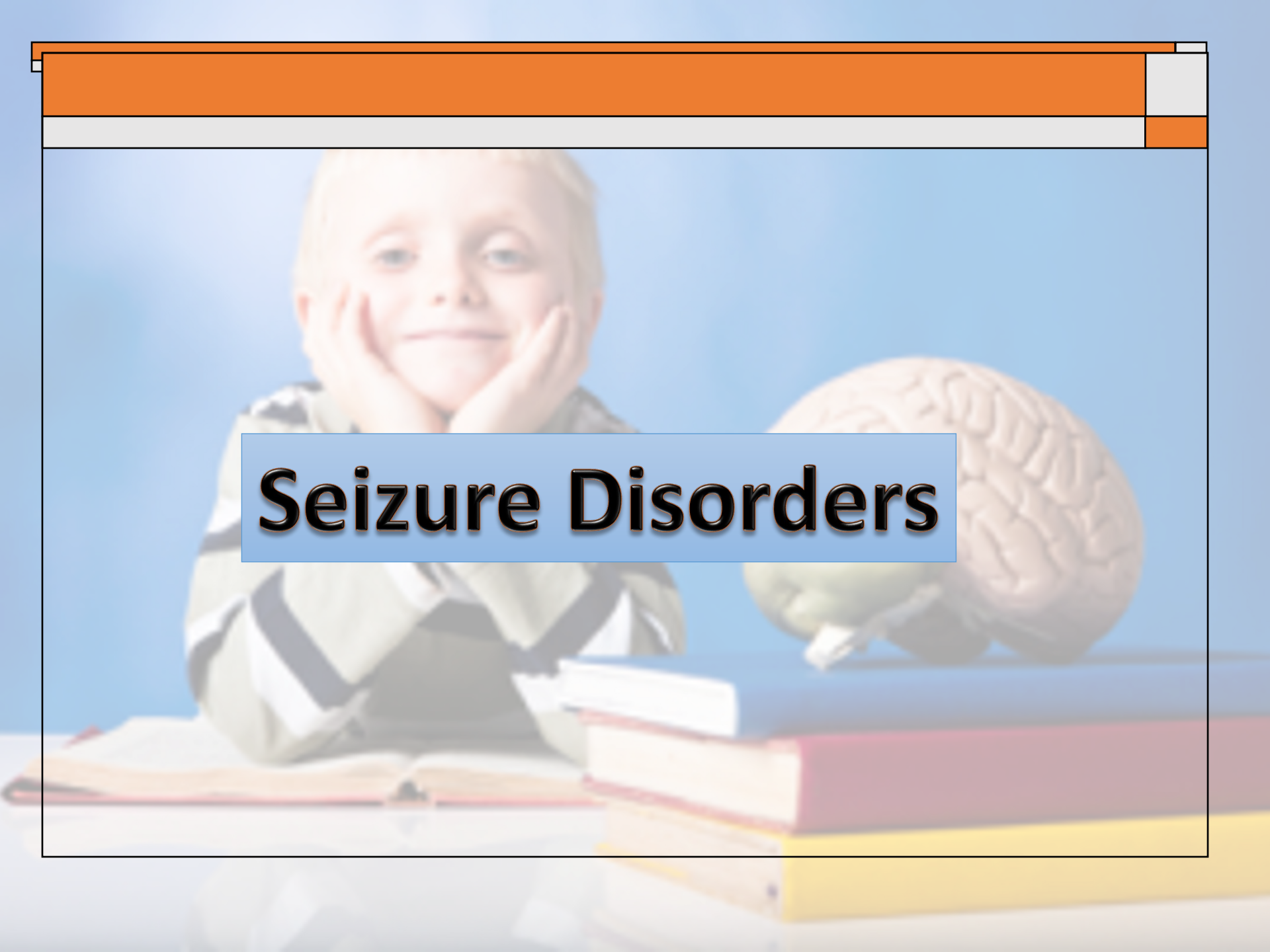


- EEG, CT, or MRI
- Electrolyte levels and metabolic workup
- Neurologic examination
- Developmental assessment

Head Injuries

Shaken Baby Syndrome

- The subdural vessels are torn as the brain moves within the skull, as the brain moves over the skull floor bruising occurs, and the brain stem my become herniated with direct trauma


Seizure Disorders

Seizures

- What are they?
  - Brief convulsive behavior caused by abnormal discharge of neurons.
  - The result of these discharges is involuntary contraction of muscles
  - When numerous nerve cells fire abnormally at the same time, a seizure may result.

Clinical Manifestations of General Seizure/ Tonic - Clonic

- Onset is abrupt. Usually less than 5 minutes duration
- Tonic Phase:
  - Usually lasts 10-20 second
  - Child loses consciousness
  - Jaw clenches shut, abdomen and chest become rigid and may emit a cry or grunt as air is forced through the taut diaphragm.
  - Pale
  - Eyes roll upward or deviate to one side.
  - Arms flexed; legs, head, neck extended
  - increased salivation and loss of swallowing reflex

Clinical Manifestations of General Seizure/ Tonic - Clonic

- Clonic Phase
  - Violent jerky movements as the trunk and extremities undergo rhythmic contraction and relaxation
  - Respirations are irregular and may have stridor
  - May foam at the mouth
  - Incontinent of urine and feces
- Afterwards
  - Drowsy and sleep afterwards

Diagnostic Tests

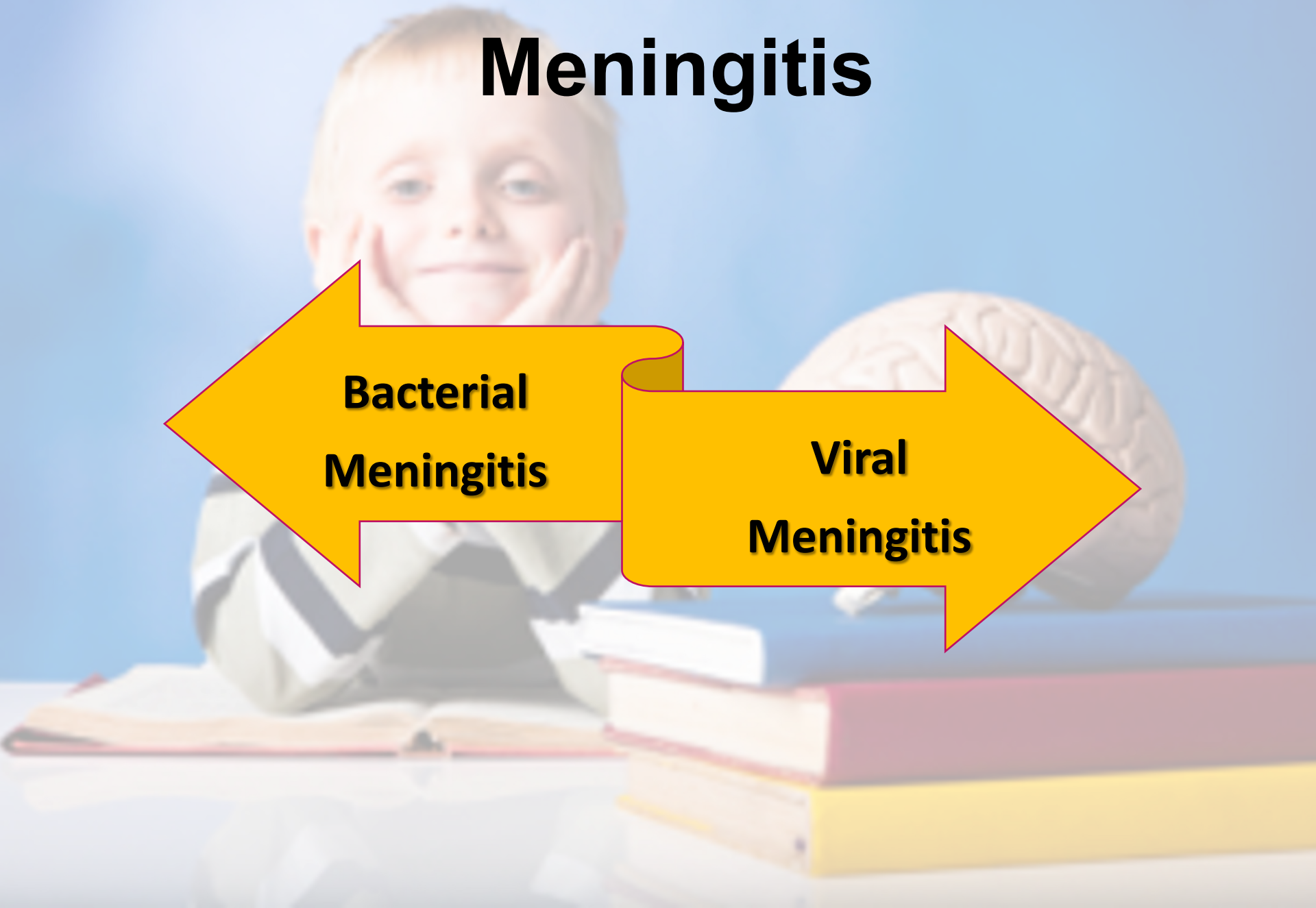
- EEG
- CT, MRI
- Lumbar puncture
- CBC
- Metabolic screen for glucose, phosphorus and lead levels

Seizure Medications

- Phenobarbital
- Carbamazephine – (Tegretol)
- Phenytoin – (Dilantin)
- Diazepam – (Valium) – used mainly for status epilepticus


Meningitis

Meningitis



Bacterial Meningitis

Potentially Fatal

Caused by:

- **Streptococcus**
- **Neisseria meningitides**
- **E coli**

What is it?

Bacteria enters blood stream, CS fluid, and brain causing an inflammatory response. Body sends WBC and they accumulate over surface of brain causing purulent exudates.

Viral Meningitis

Same signs and symptoms, may be milder and self-limiting.

Usually lasts a few days

Assessment

Infants:

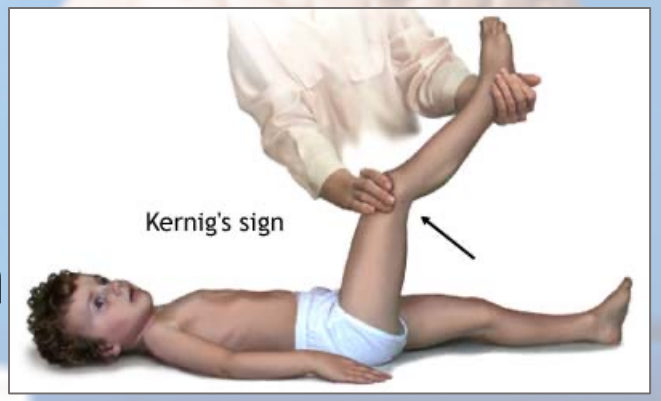
- **Fever (not always present)**
- **Lethargy**
- **Alterations in sleep and feeding habits**
- **Fussy and irritable**
- **Nuchal rigidity (late sign)**
- **Bulging fontanel**
- **High pitched cry**

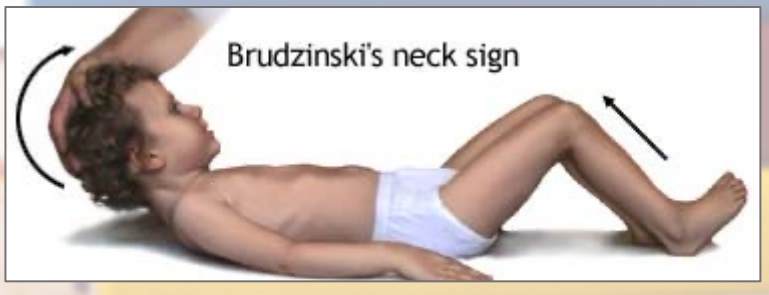
Assessment

- Childhood & Adolescence
  - Hyperthermia
  - S&S of IICP
  - Nausea and vomiting
  - Headache
  - Seizures
  - Photophobia

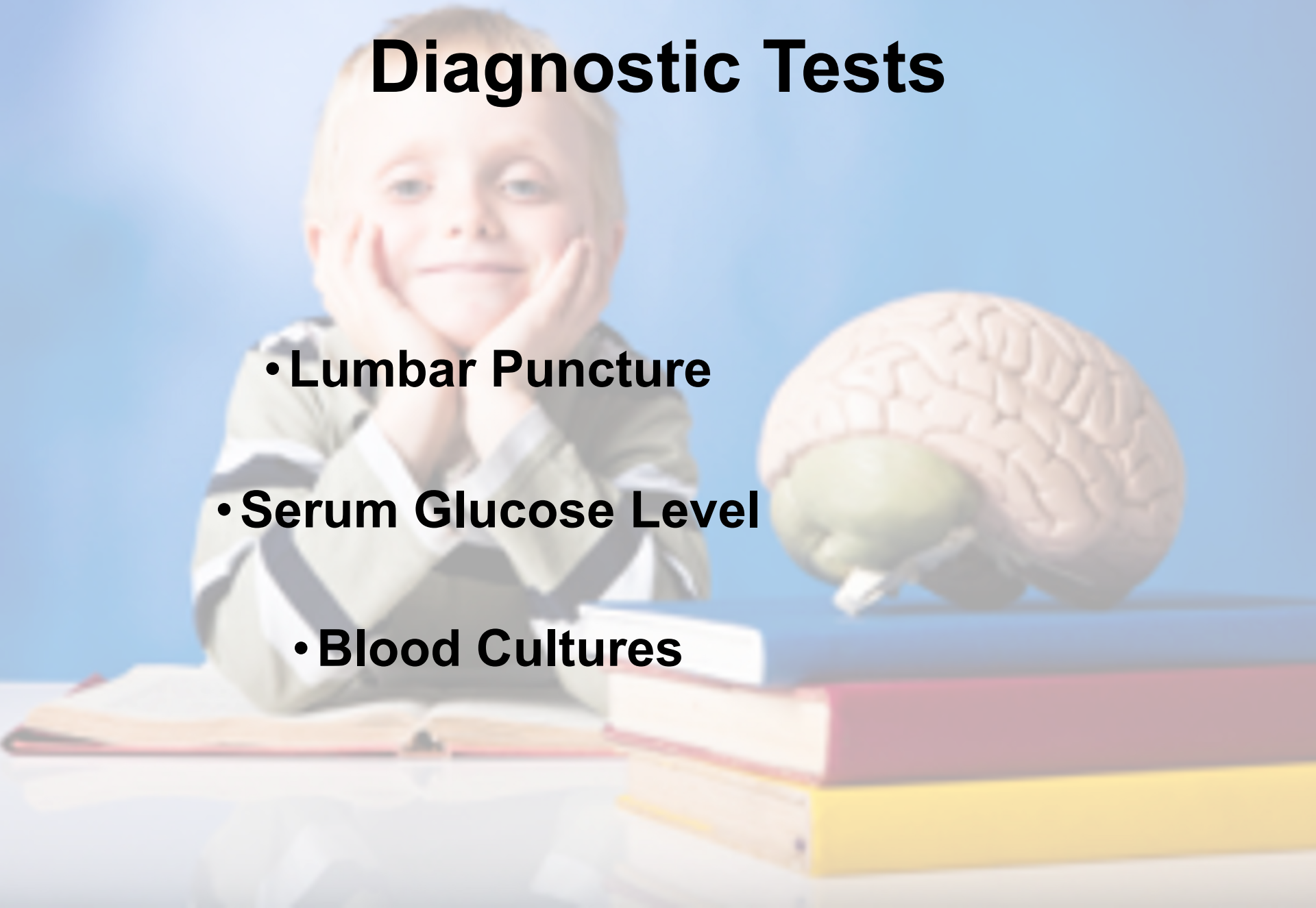
Signs of Meningeal Irritation

- Headache
- Photophobia
- Nuchal Rigidy
- Opisthotonic position
- Positive Kernig's sign
- Postive Brudzinski's sign

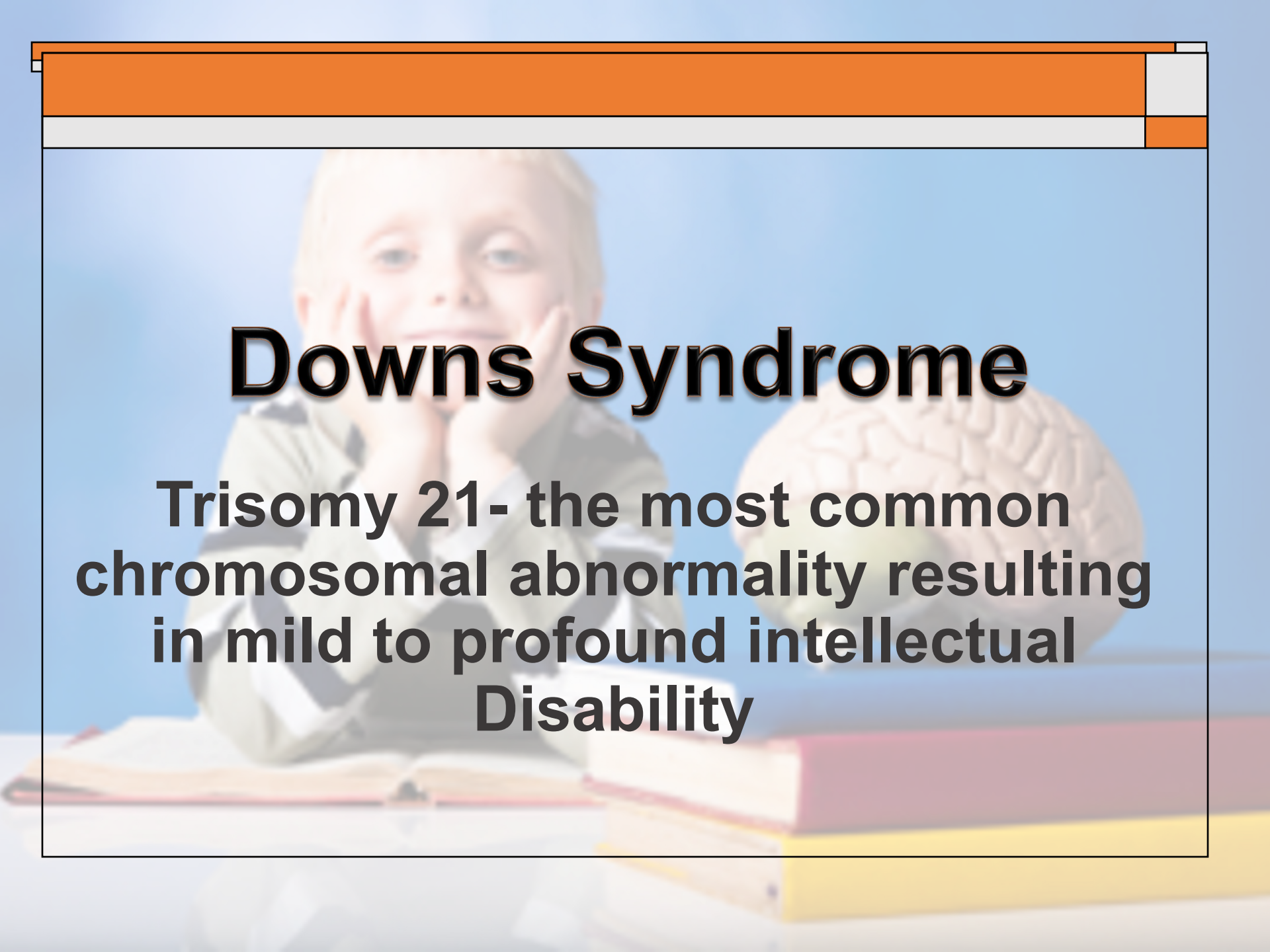




Diagnostic Tests



- Lumbar Puncture
- Serum Glucose Level
- Blood Cultures



Downs Syndrome

Trisomy 21- the most common chromosomal abnormality resulting in mild to profound intellectual Disability

Down Syndrome

Clinical Manifestations:

- Congenital anomalies – cardiac and GI tract
- Flat facial features, nose broad and flat
- Low set ears
- Upward slanting eyes
- Prominent epicanthial folds
- Short hands with simian crease
- Hypotonia
- Neck short with extra fat pad
- Usually sterile