

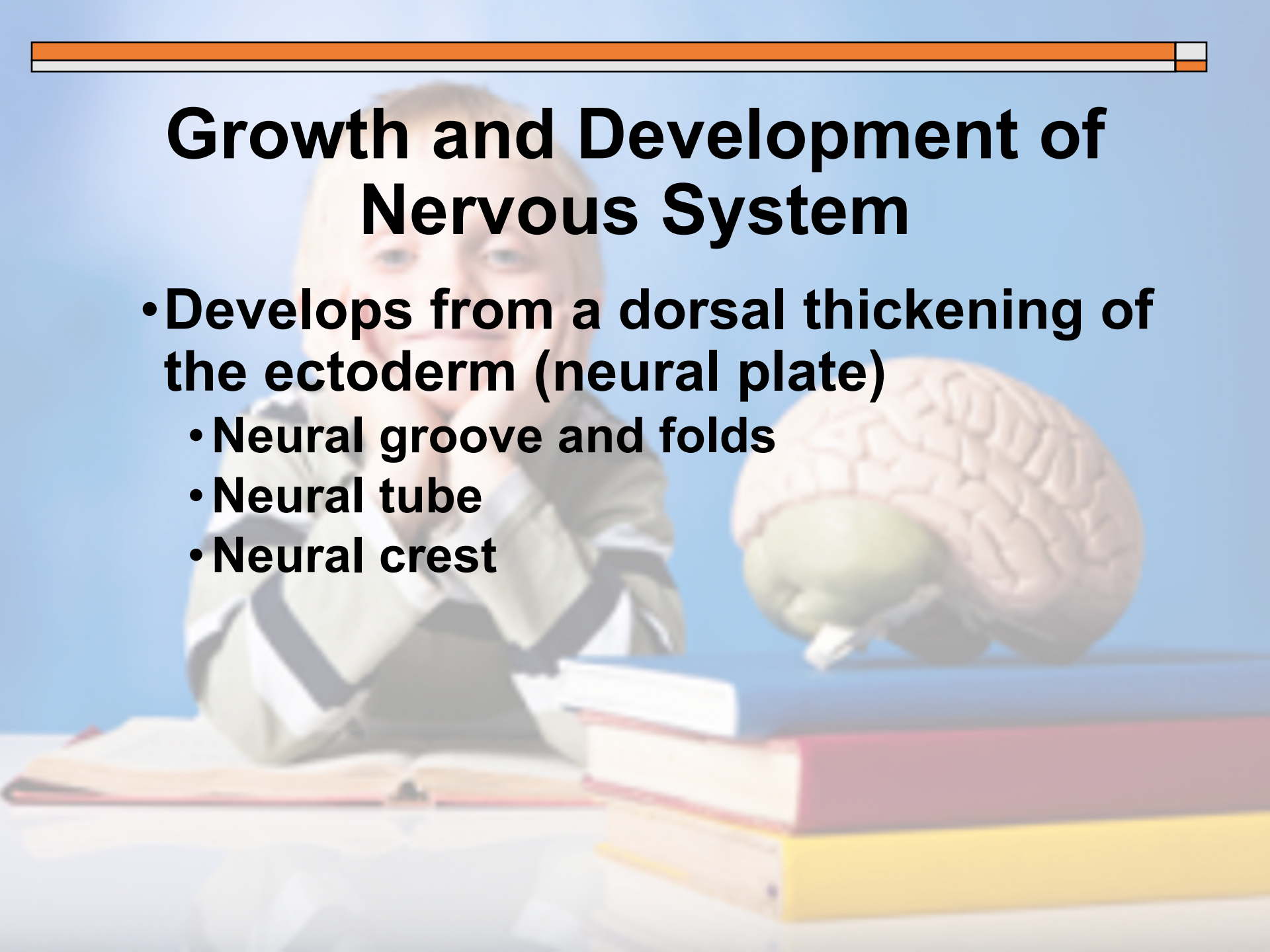
Pediatric Neurological Disorders

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
- 
- The background of the slide features a young girl with blonde hair, wearing a light-colored sweater, sitting at a desk with several books. To her right is a 3D anatomical model of a human brain, showing the cerebral cortex and brainstem. The overall scene is set against a light blue background.

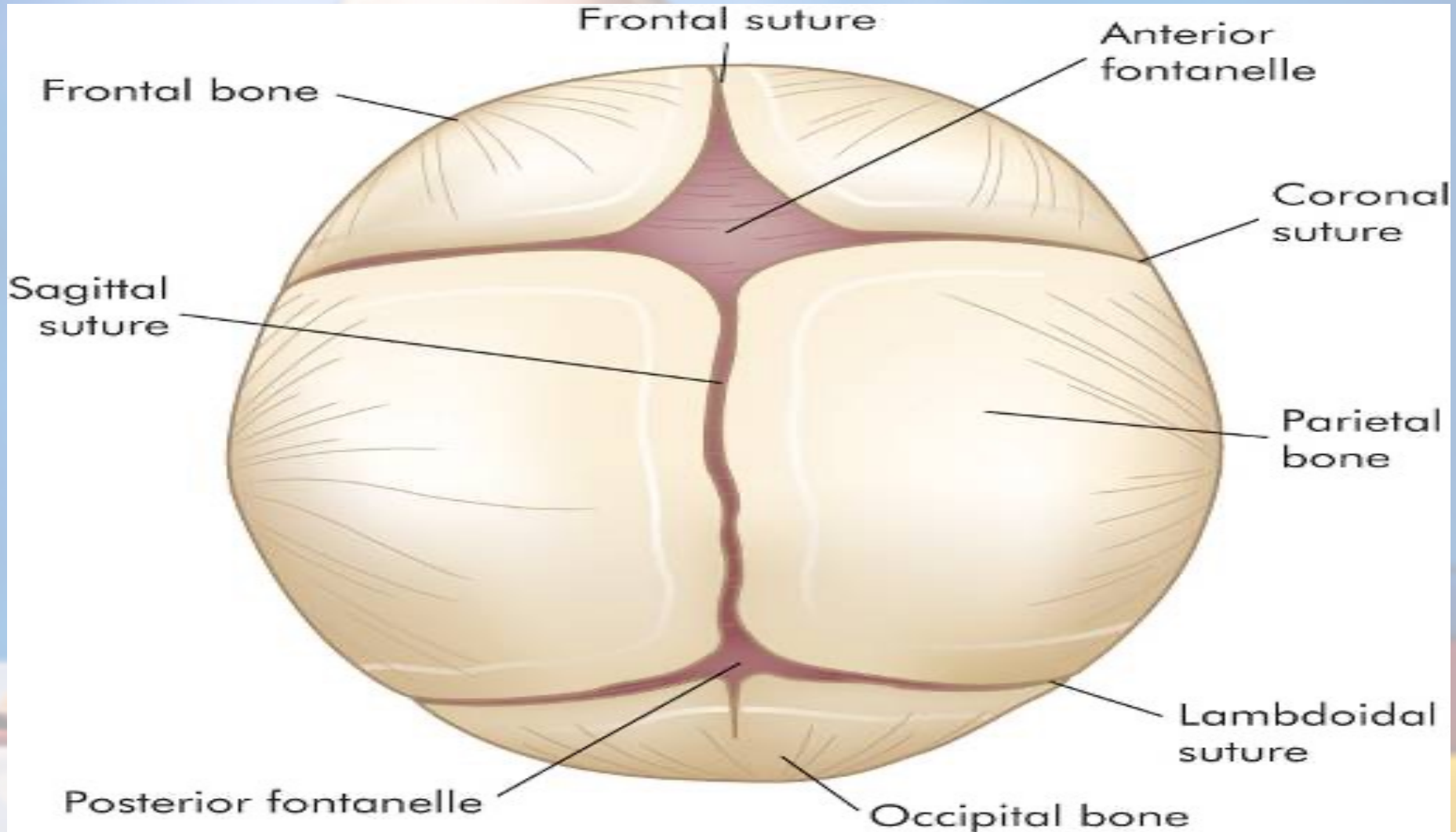


Growth and Development of Nervous System

- **Mesoderm**

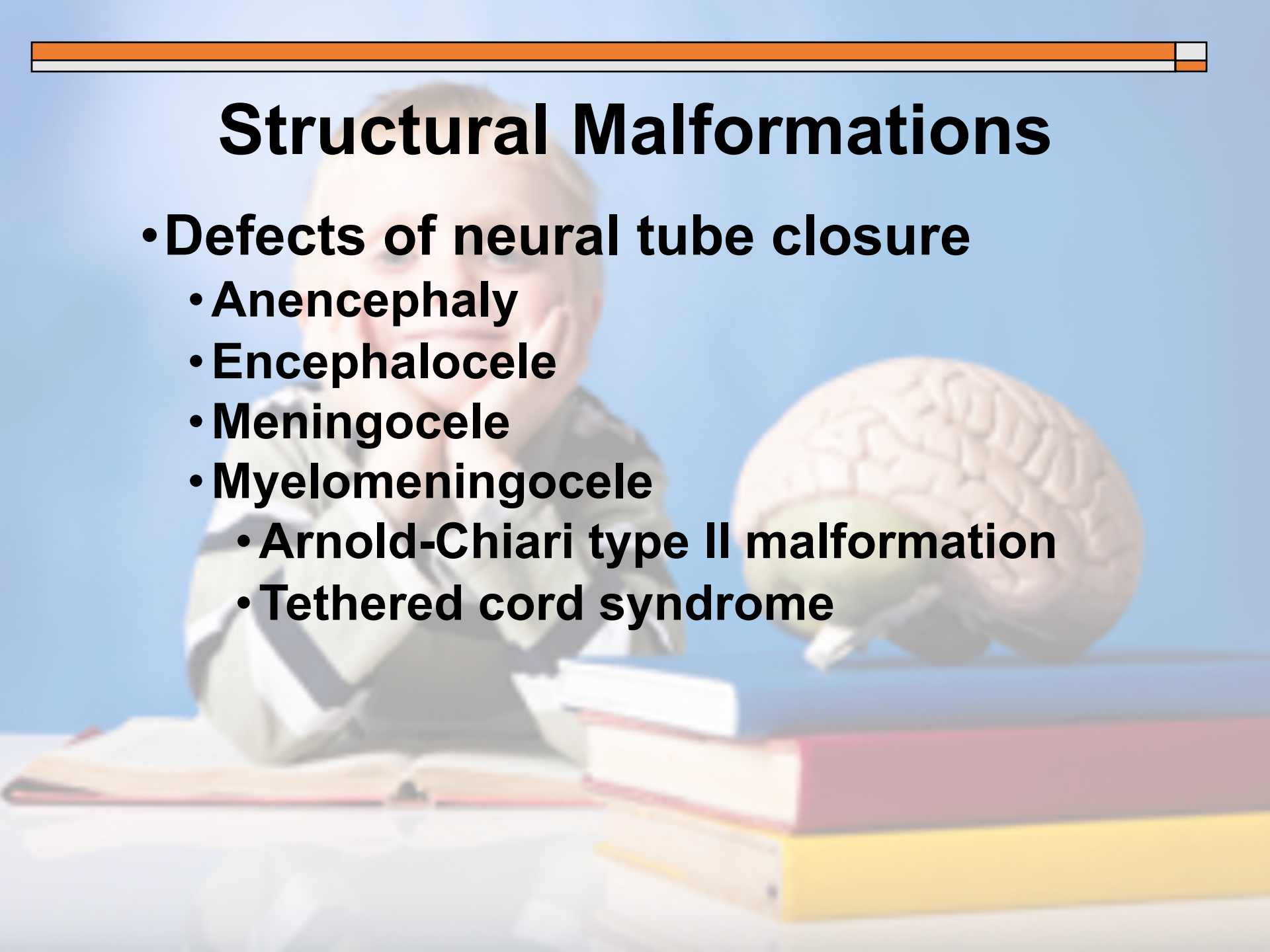
- **Blood vessels, microglial cells, dural and arachnoid layers of the meninges, the capsule of some peripheral nerve endings, and nerve coverings**
- 
- The background of the slide features a soft-focus image of a young child with blonde hair, wearing a light-colored sweater, sitting at a desk with books. To the right of the child, a human brain is visible, rendered in a semi-transparent, light brown color. The overall background is a light blue gradient.

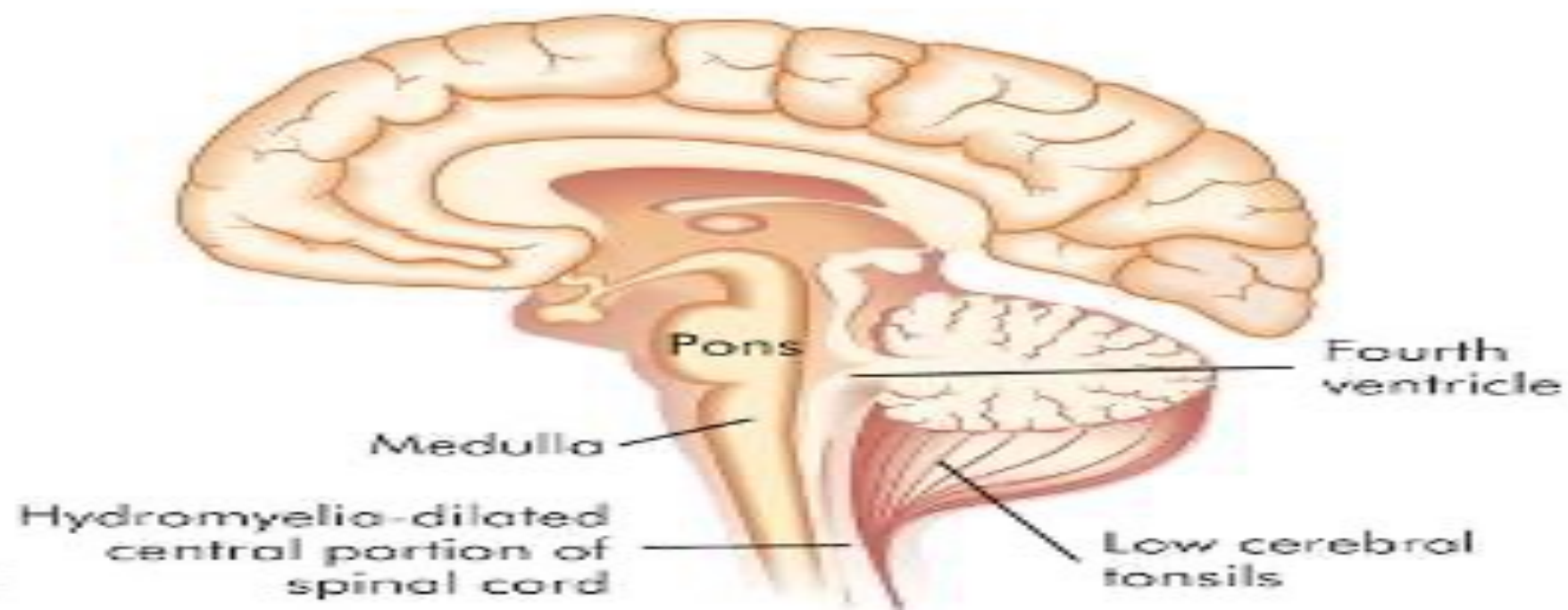
Cranial Sutures and Fontanelles





Structural Malformations

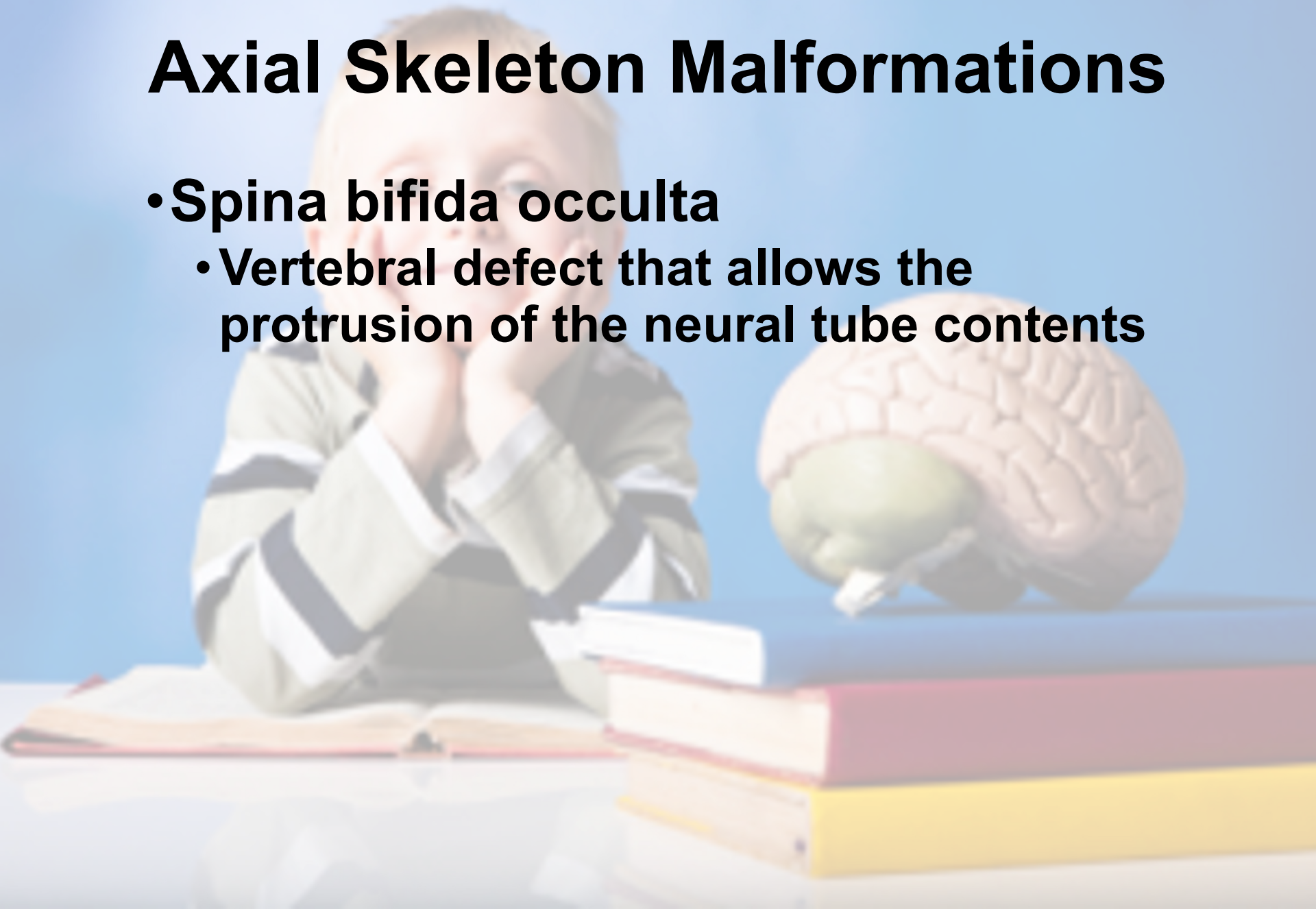
- **Defects of neural tube closure**
 - **Anencephaly**
 - **Encephalocele**
 - **Meningocele**
 - **Myelomeningocele**
 - **Arnold-Chiari type II malformation**
 - **Tethered cord syndrome**
- 






Axial Skeleton Malformations

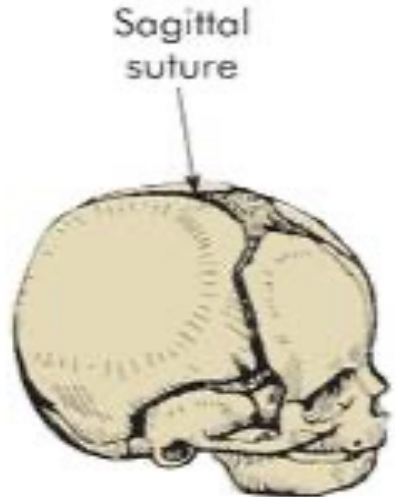
- **Spina bifida occulta**
 - **Vertebral defect that allows the protrusion of the neural tube contents**





Axial Skeleton Malformations

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



NORMAL SKULL



BRACHYCEPHALY



MICROCEPHALY AND CRANIOSTENOSIS



OXYCEPHALY OR ACROCEPHALY



SCAPHOCEPHALY OR DOLICHOCEPHALY



PLAGIOCEPHALY

Childhood Brain Tumors

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



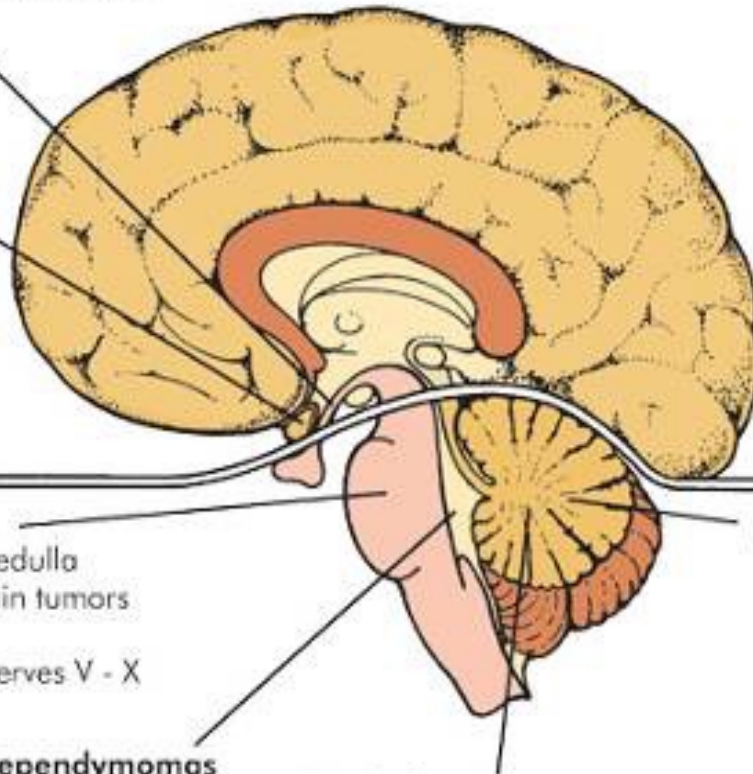
Craniopharyngiomas

- 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

See Notes Page 95

Optic nerve gliomas

- Most often a low-grade astrocytoma



Cerebral tumors

- Astrocytomas invade surrounding structures but grow slowly

- Ependymomas arise from lining tissue of lateral ventricle

Supratentorial

Brain stem gliomas

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

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

Infratentorial

Infratentorial ependymomas

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

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

Other Childhood Tumors

- **Embryonal tumors**
 - **Neuroblastoma**
 - **Retinoblastoma**
 - **Inherited**
 - **Acquired**



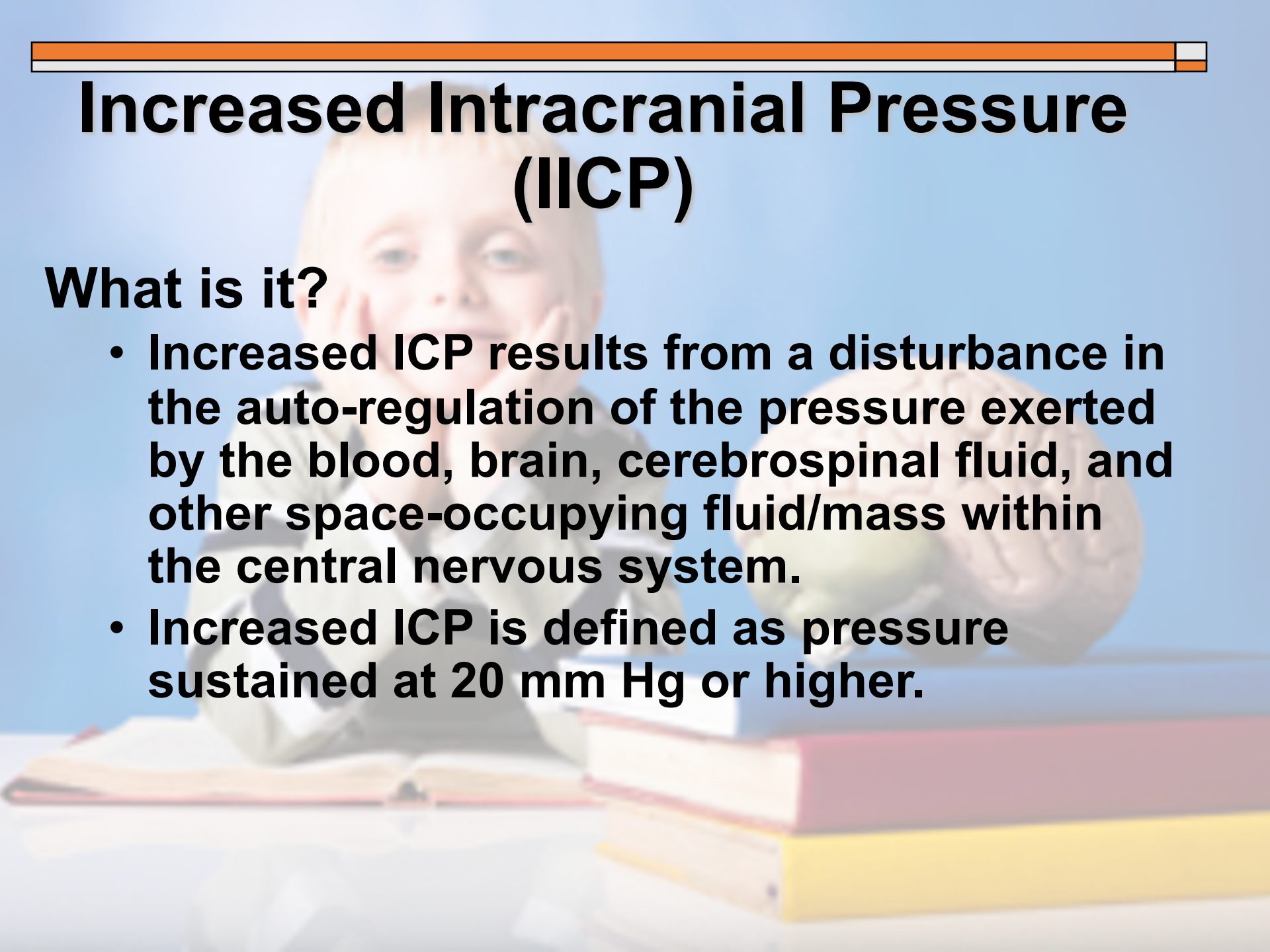
A young girl with blonde hair is sitting at a desk, resting her chin on her hand. She is wearing a green and white striped shirt. In front of her are several books. To her right, a model of a human brain is visible. The background is a light blue gradient. The text "Increased Intracranial Pressure" is overlaid in the center in a large, bold, black font with a drop shadow.

Increased Intracranial Pressure



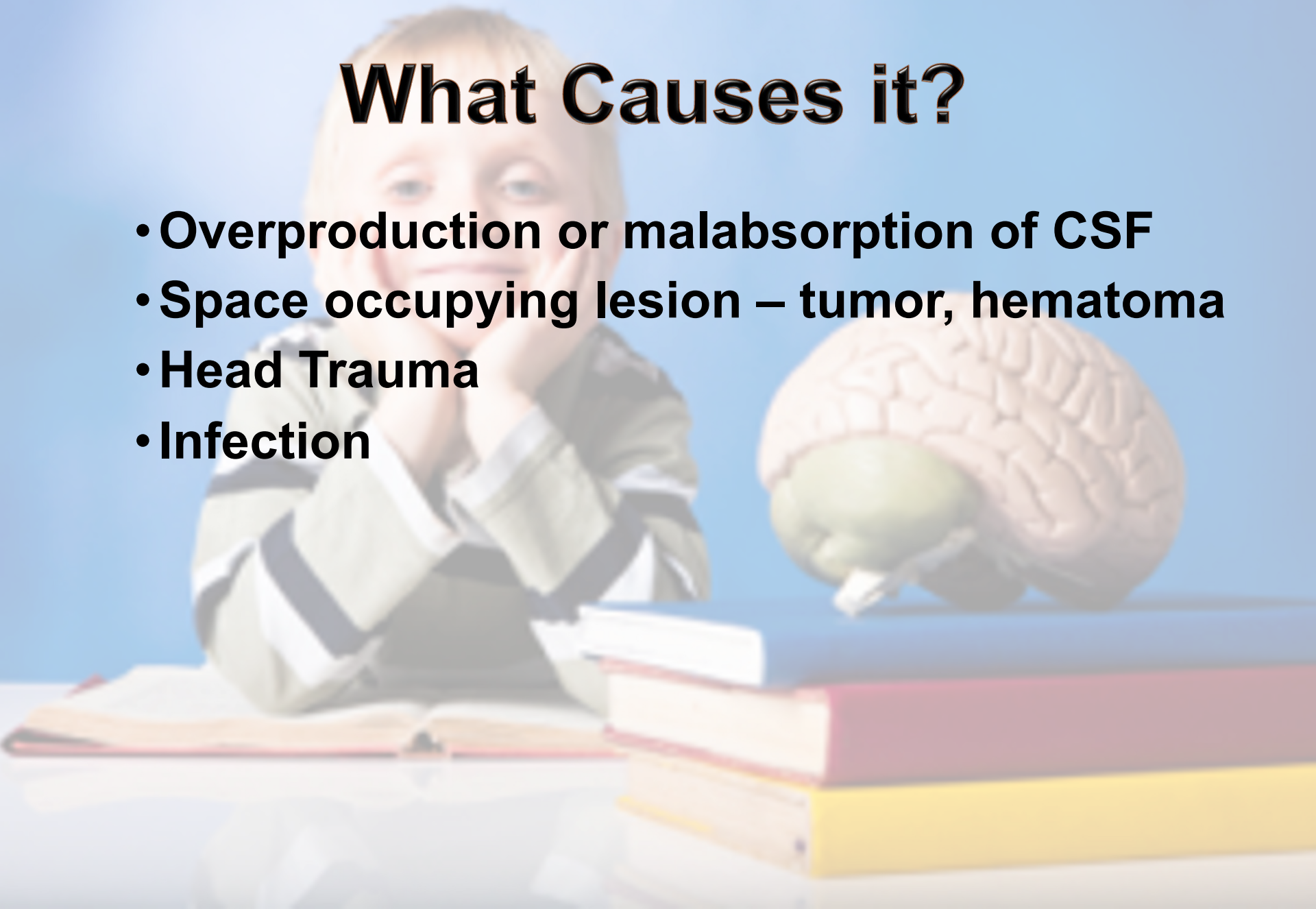
Increased Intracranial Pressure (IICP)

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.**
- 
- The background of the slide features a soft-focus image of a young child with blonde hair, resting their chin on their hands in a thoughtful pose. To the right of the child, a human brain is visible. In the foreground, there is a stack of several books with various colored spines (red, yellow, and white).




What Causes it?

- **Overproduction or malabsorption of CSF**
 - **Space occupying lesion – tumor, hematoma**
 - **Head Trauma**
 - **Infection**
- 
- The background of the slide features a young child with blonde hair, wearing a green and white striped shirt, sitting at a desk with several books. To the right of the child is a 3D anatomical model of a human brain, with a green, rounded mass representing a lesion or tumor on its surface.




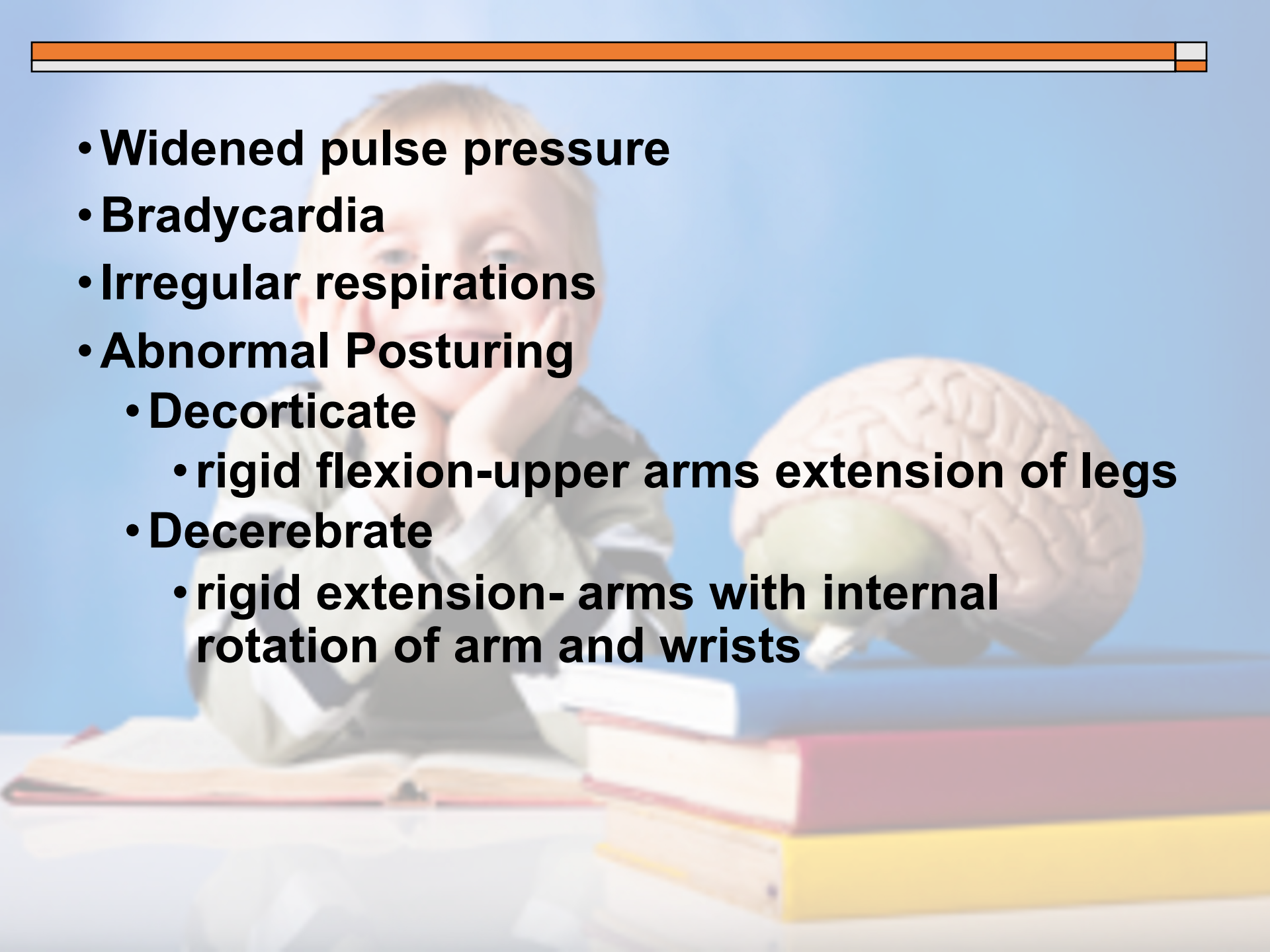
Clinical Manifestations: Infant

- **Irritability and restlessness; high-pitched cry**
 - **Full to bulging fontanelles; 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)**
- 

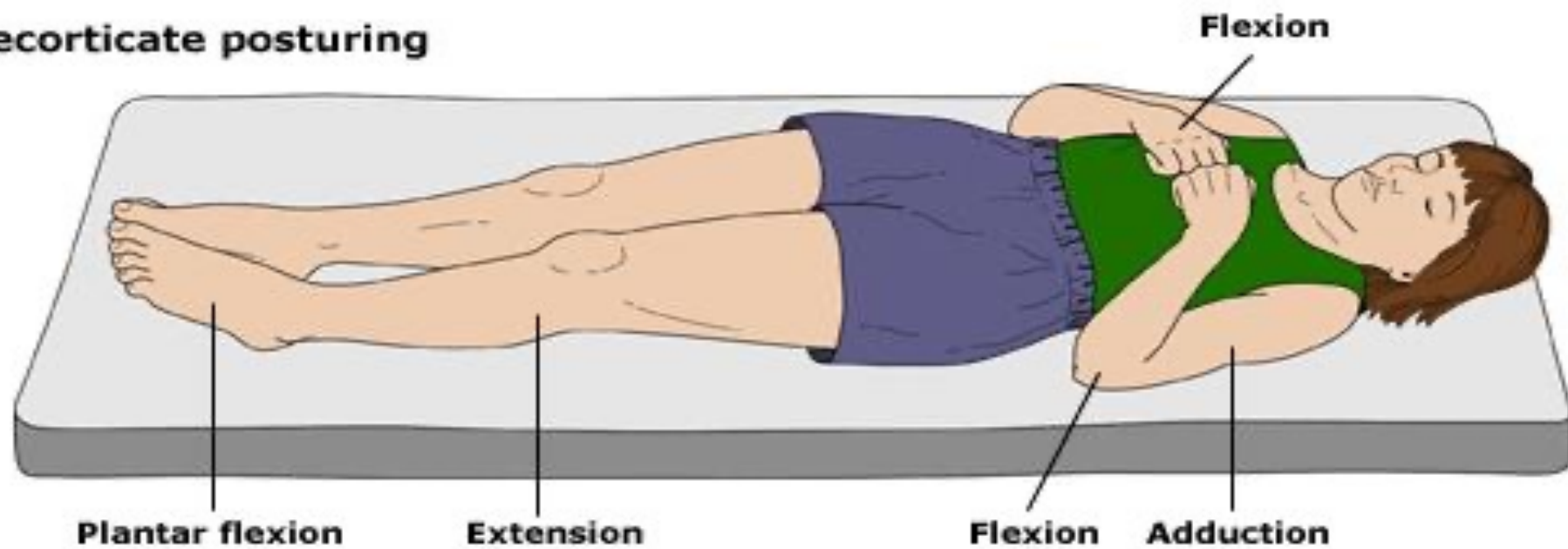


Clinical Manifestations: Child

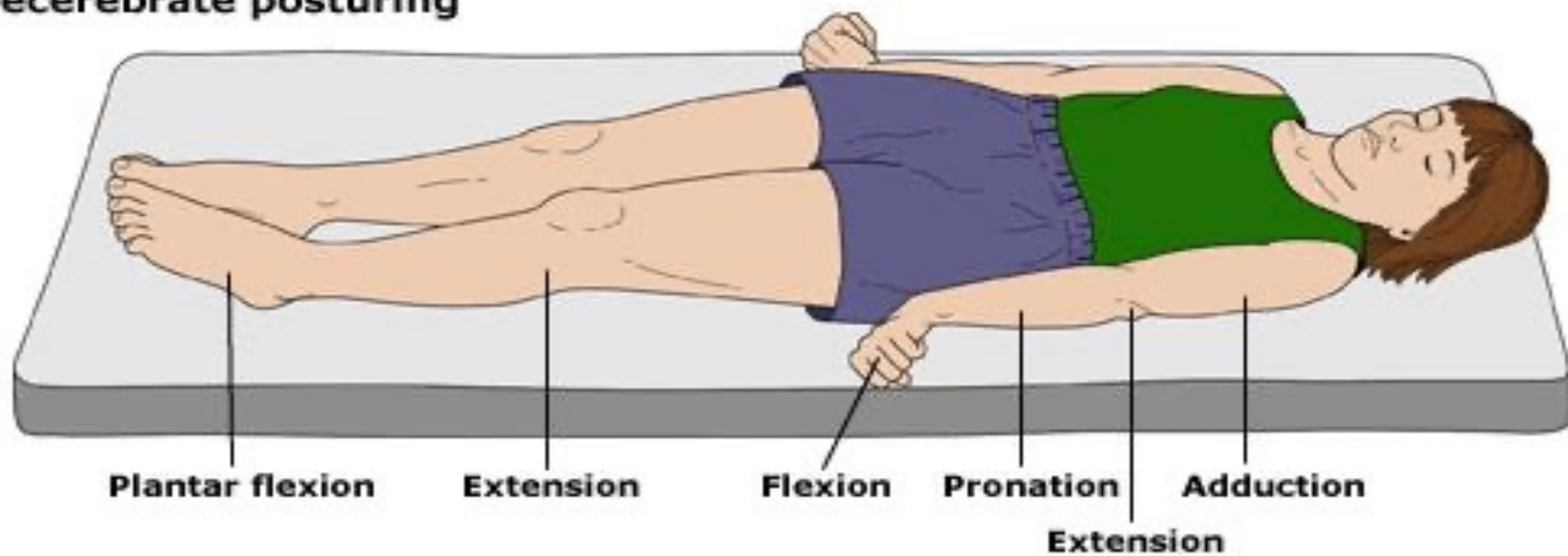
- Headache
 - Visual disturbances - diplopia
 - Nausea and Vomiting
 - Dizziness or vertigo
 - Irritability, lethargy, mood swings
 - Ataxia, lower extremity spasticity
 - Nuchal rigidity
 - Deterioration in school performance, or cognitive ability
- 

- 
- A young child with blonde hair is sitting at a desk, looking towards the camera. The desk is cluttered with several books of various colors (red, yellow, white). To the right of the child, a human brain is visible, and a green apple is placed in front of it. The background is a solid light blue color.
- **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

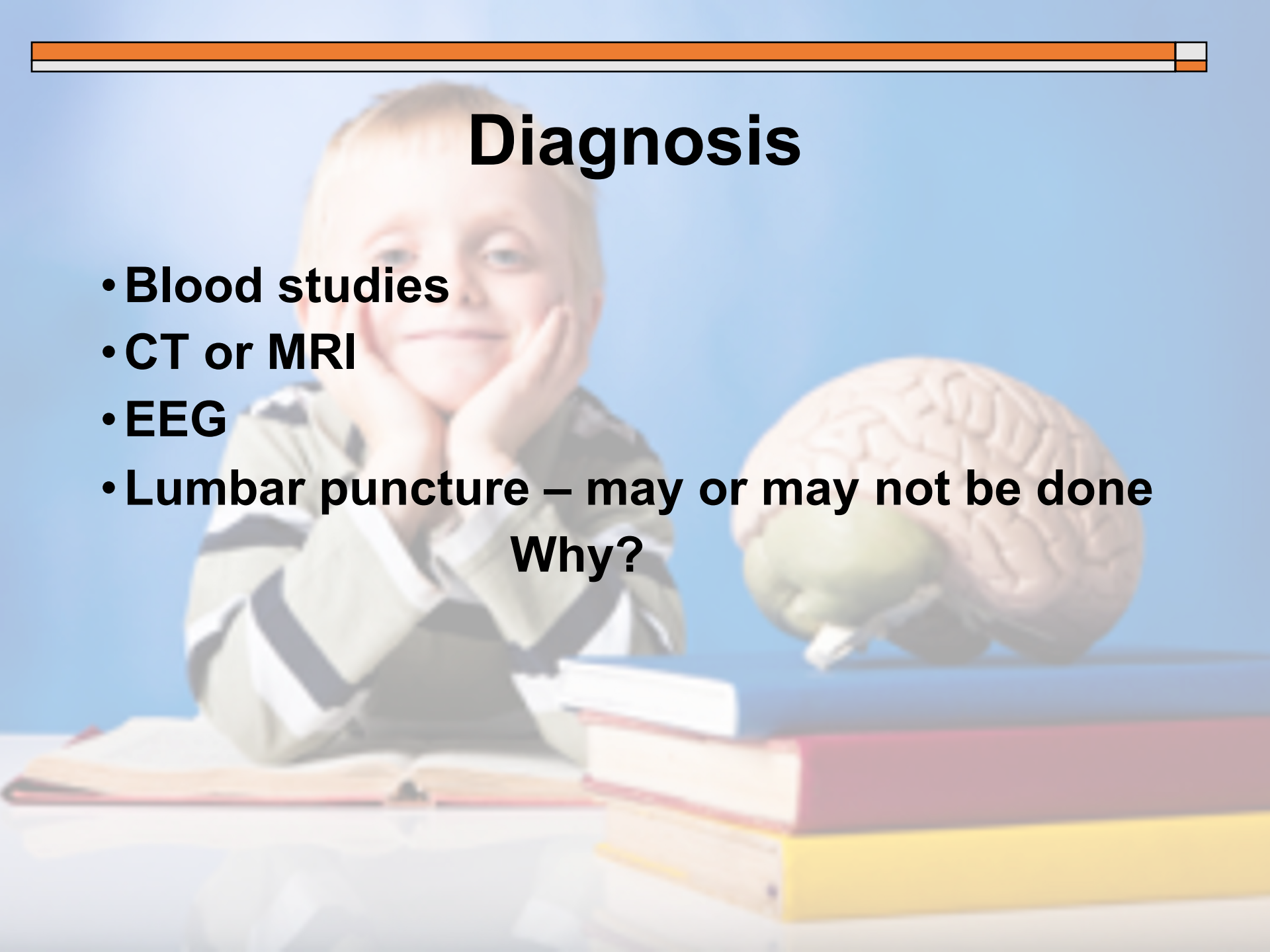




Diagnosis

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

Why?

A young child with blonde hair, wearing a green and white striped shirt, is sitting at a desk. The child is resting their chin on their hands and looking towards the camera. On the desk in front of the child are several books, including one with a red cover and one with a yellow cover. To the right of the child is a large, realistic model of a human brain. The background is a solid light blue color.



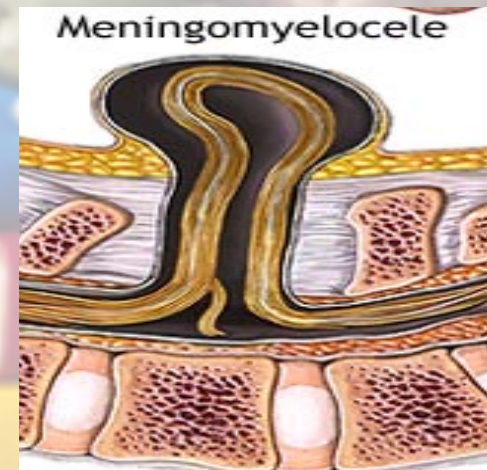
Spina Bifida
Meningocele
Meningomyelocele

What is the difference?

- **Spina Bifida**

- **Meningocele:**

- **Myelomeningocele:**



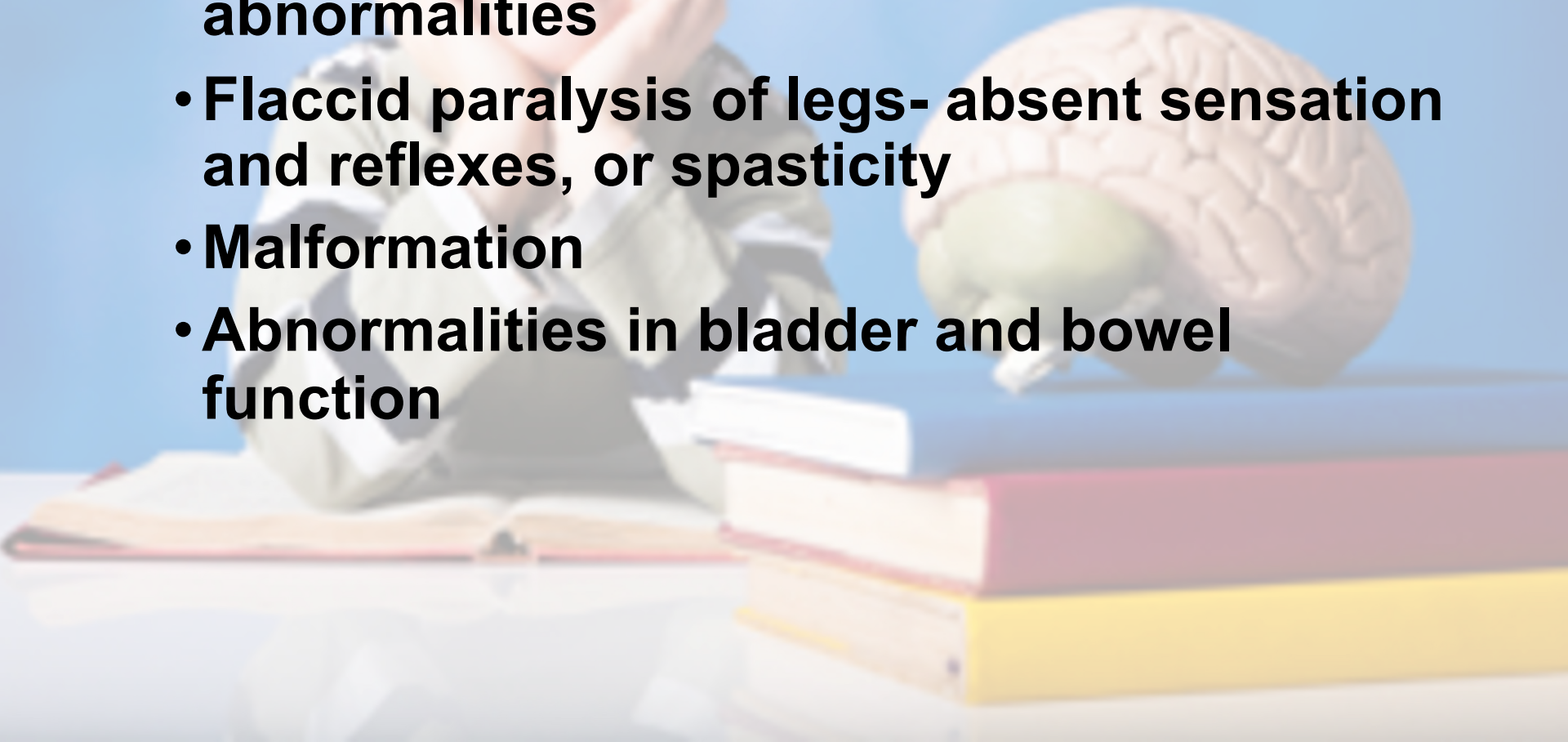
See Notes Page 96

A young child with blonde hair is sitting at a desk, resting their chin on their hands. In front of them are several books and a model of a human brain. The background is a light blue gradient. At the top of the image, there is a horizontal bar with an orange segment on the left and a white segment on the right.

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



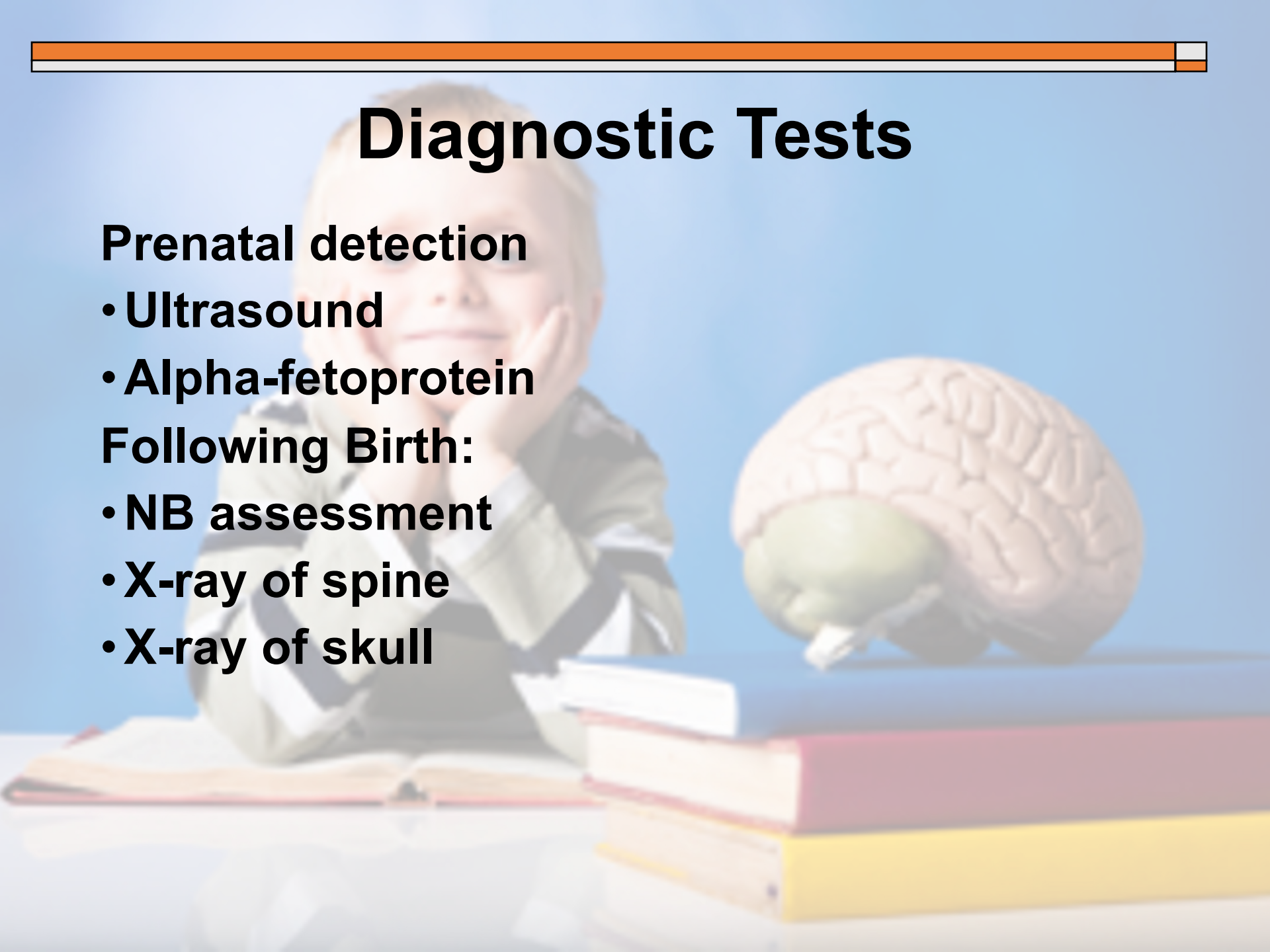


Diagnostic Tests

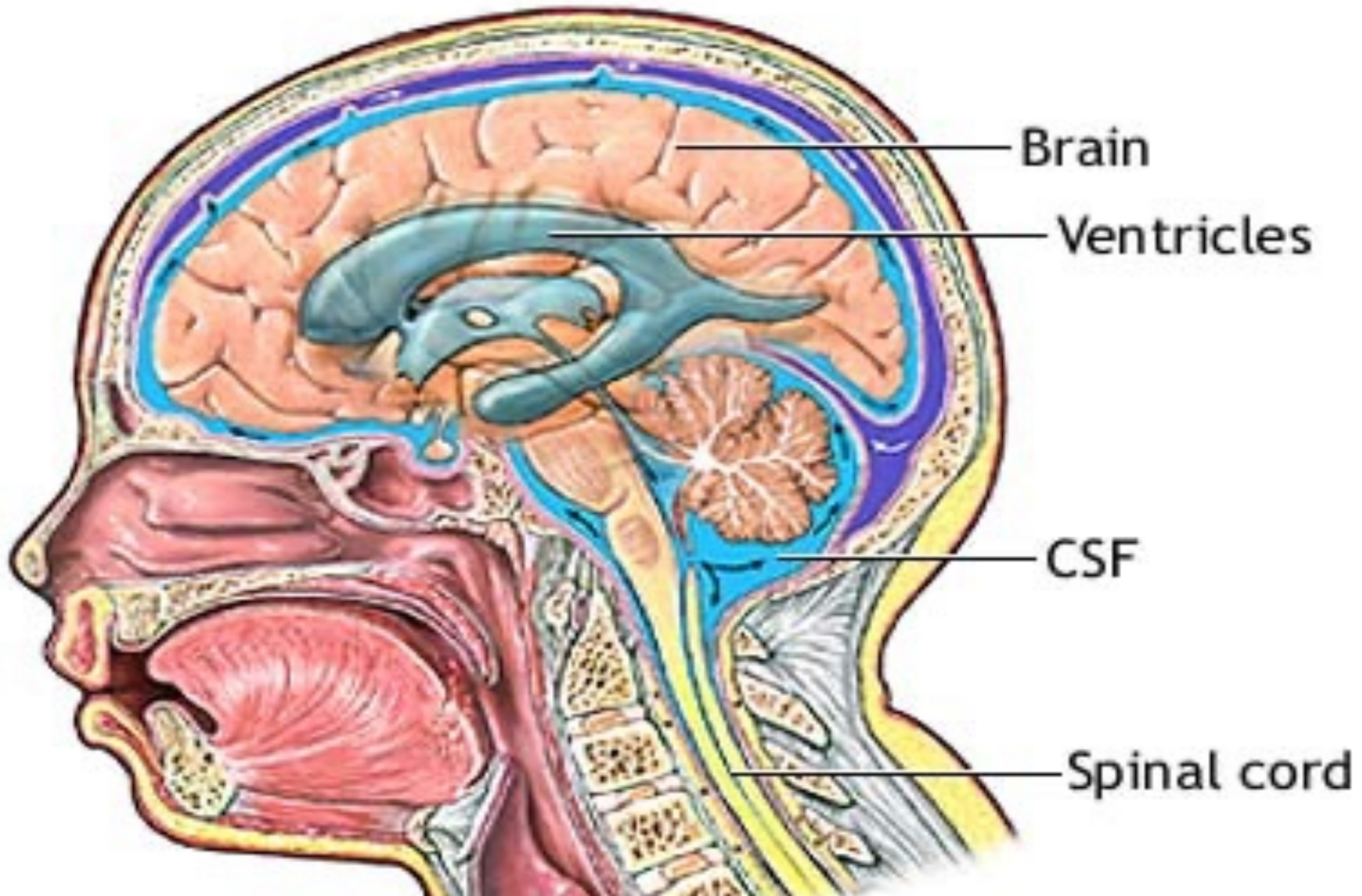
Prenatal detection

- Ultrasound
- Alpha-fetoprotein

Following Birth:

- NB assessment
 - X-ray of spine
 - X-ray of skull
- 
- A young child with blonde hair is sitting at a desk, resting their chin on their hand. On the desk are several books, a model of a human brain, and a green apple. The background is a light blue gradient.

Hydrocephalus



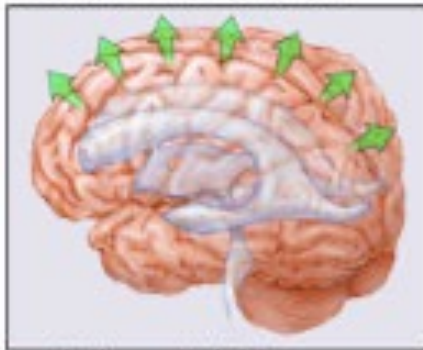
Etiology and Pathophysiology

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

Brain with normal ventricles



Brain with enlarged ventricles



See Notes Page 96

Clinical Manifestations

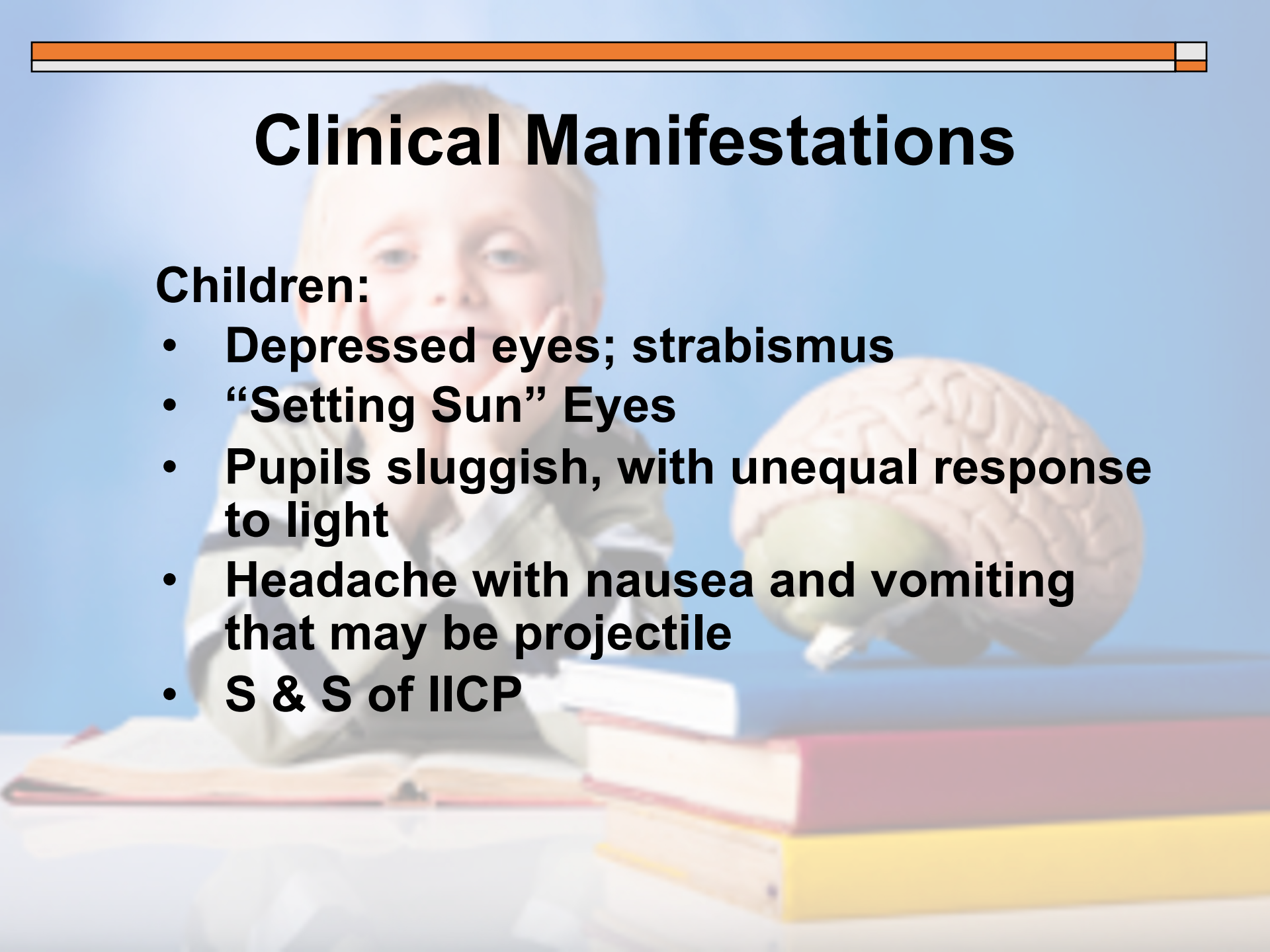
Infants

- Increase in FOC
- Frontal enlargement or bossing
- Head larger than face
- Translucent skin
- Wide palpable suture lines
- Bulging Fontanel
- 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**
- 
- A young child with blonde hair is sitting at a desk, looking towards the camera. The desk is cluttered with several books of various colors (red, yellow, white). To the right of the child, there is a model of a human brain and a glowing lightbulb. The background is a solid light blue color.

Diagnostic Tests

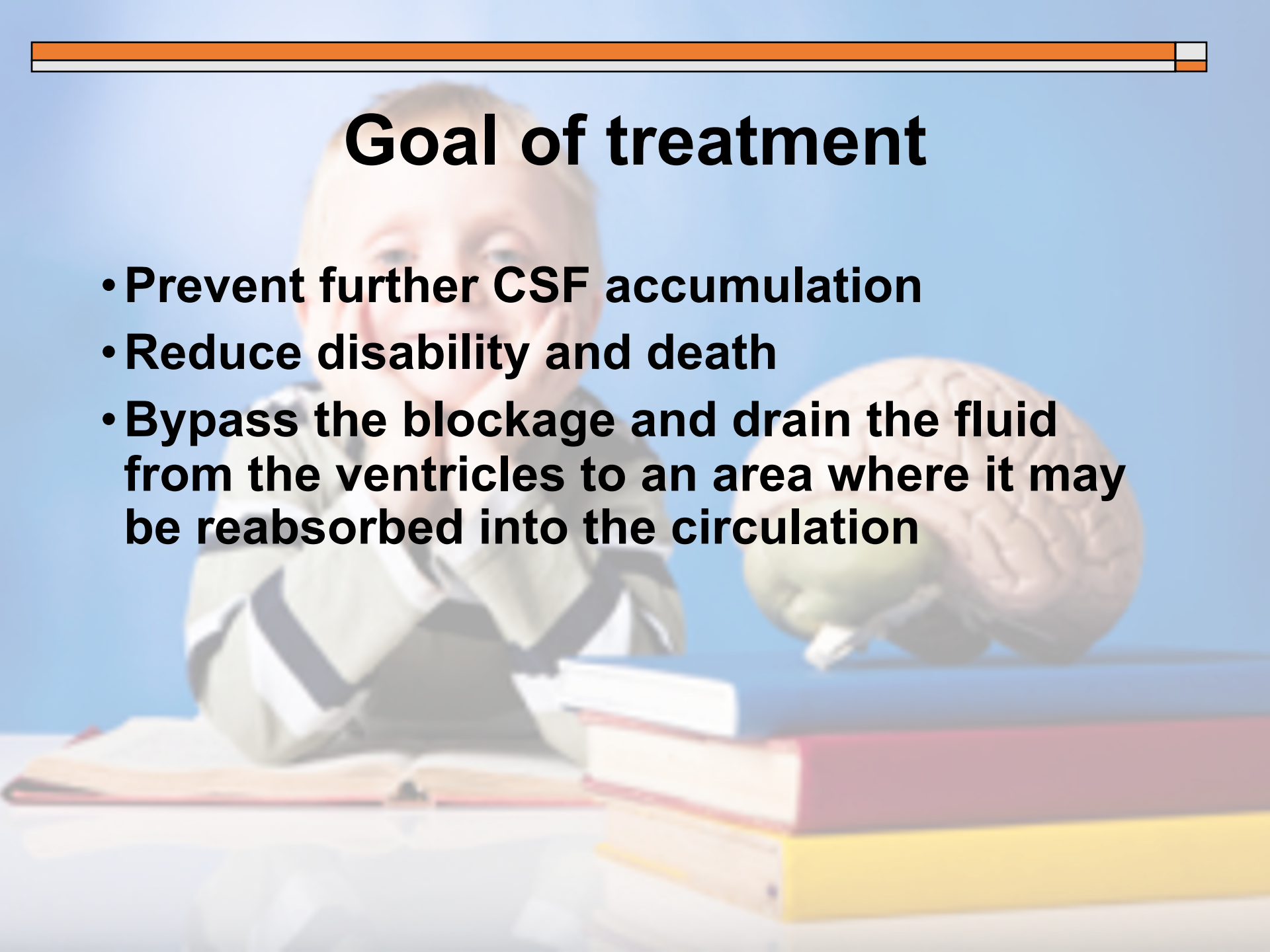
- MRI/ CT scan
- Skull X-ray
- FOC
- Transillumination

****lumbar puncture very dangerous and usually NOT done**



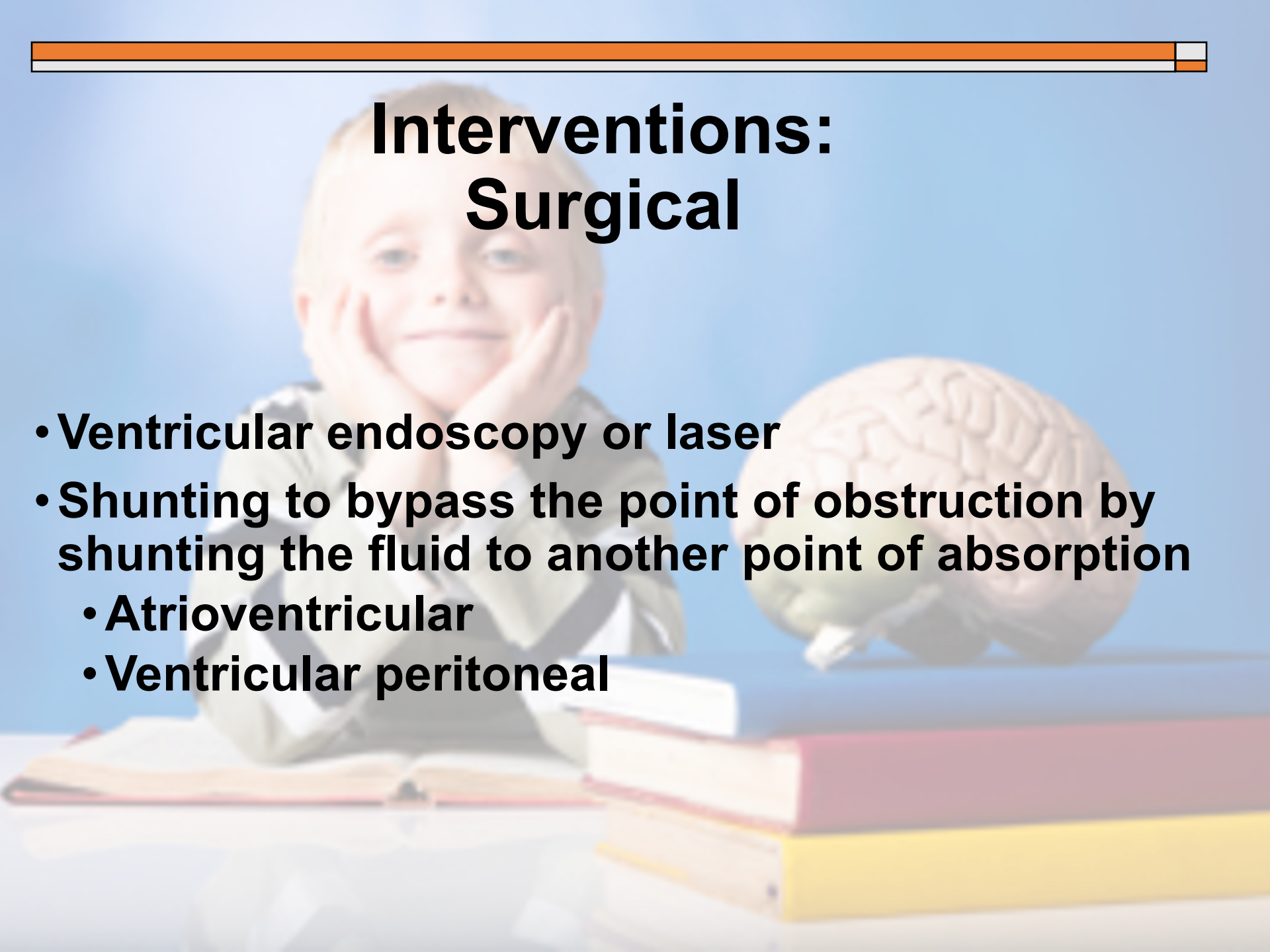


Goal of treatment

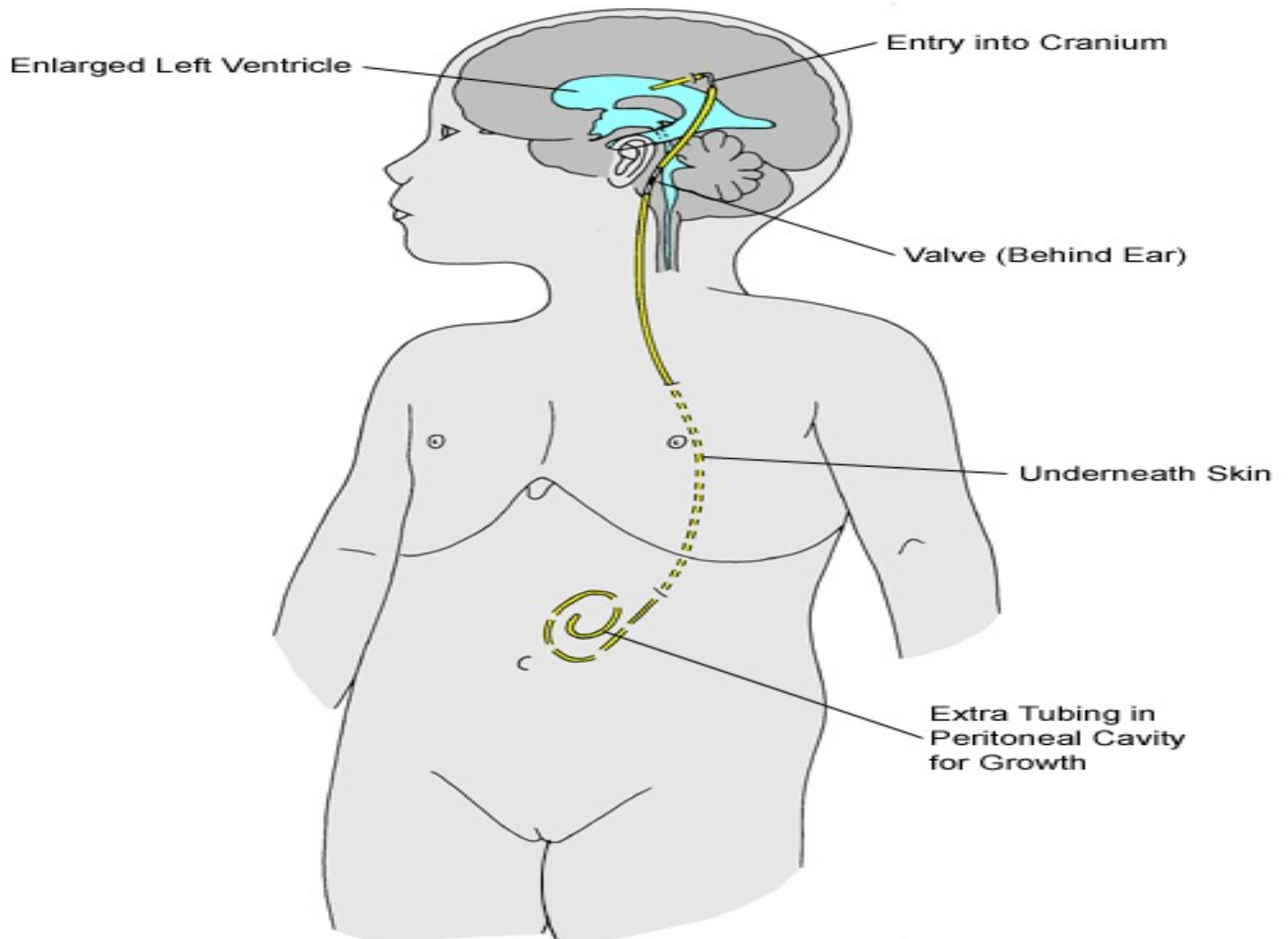
- **Prevent further CSF accumulation**
 - **Reduce disability and death**
 - **Bypass the blockage and drain the fluid from the ventricles to an area where it may be reabsorbed into the circulation**
- 
- The background of the slide features a composite image. On the left, a young child with blonde hair is shown from the chest up, resting their chin on their hand in a thoughtful pose. To the right of the child is a realistic, anatomical model of a human brain. In the foreground, there is a stack of several books with various colored spines (red, yellow, white) and an open book with a red cover. The entire scene is set against a light blue background.



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



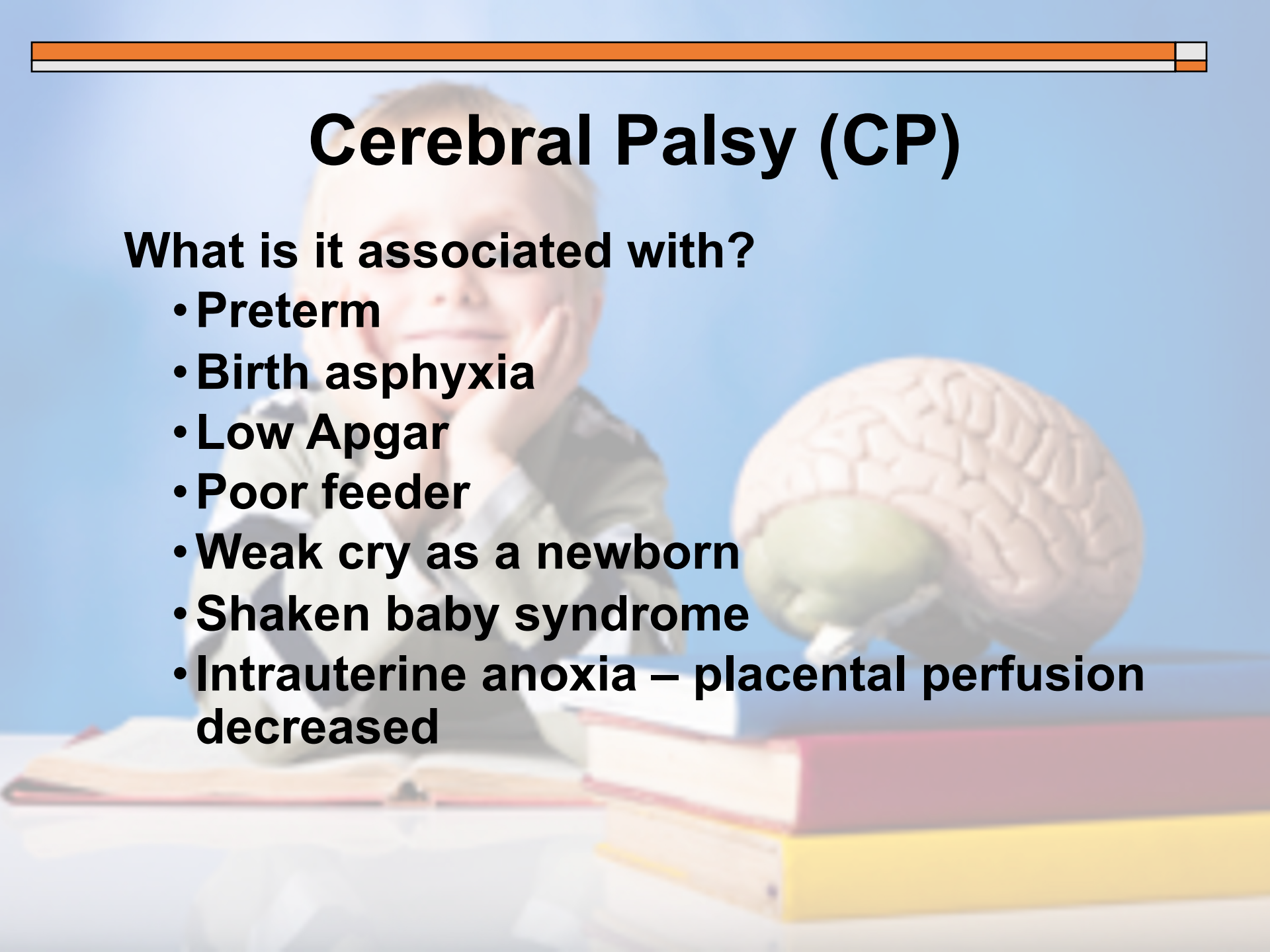
A young child with blonde hair is sitting at a desk, resting their chin on their hands. In front of them are several books stacked on the desk. To the right, a model of a human brain is visible. The background is a solid light blue color. The text 'CEREBRAL PALSY' is overlaid in the center in a bold, black font with a yellow outline, set against a light blue background with a water droplet pattern.

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**
- 

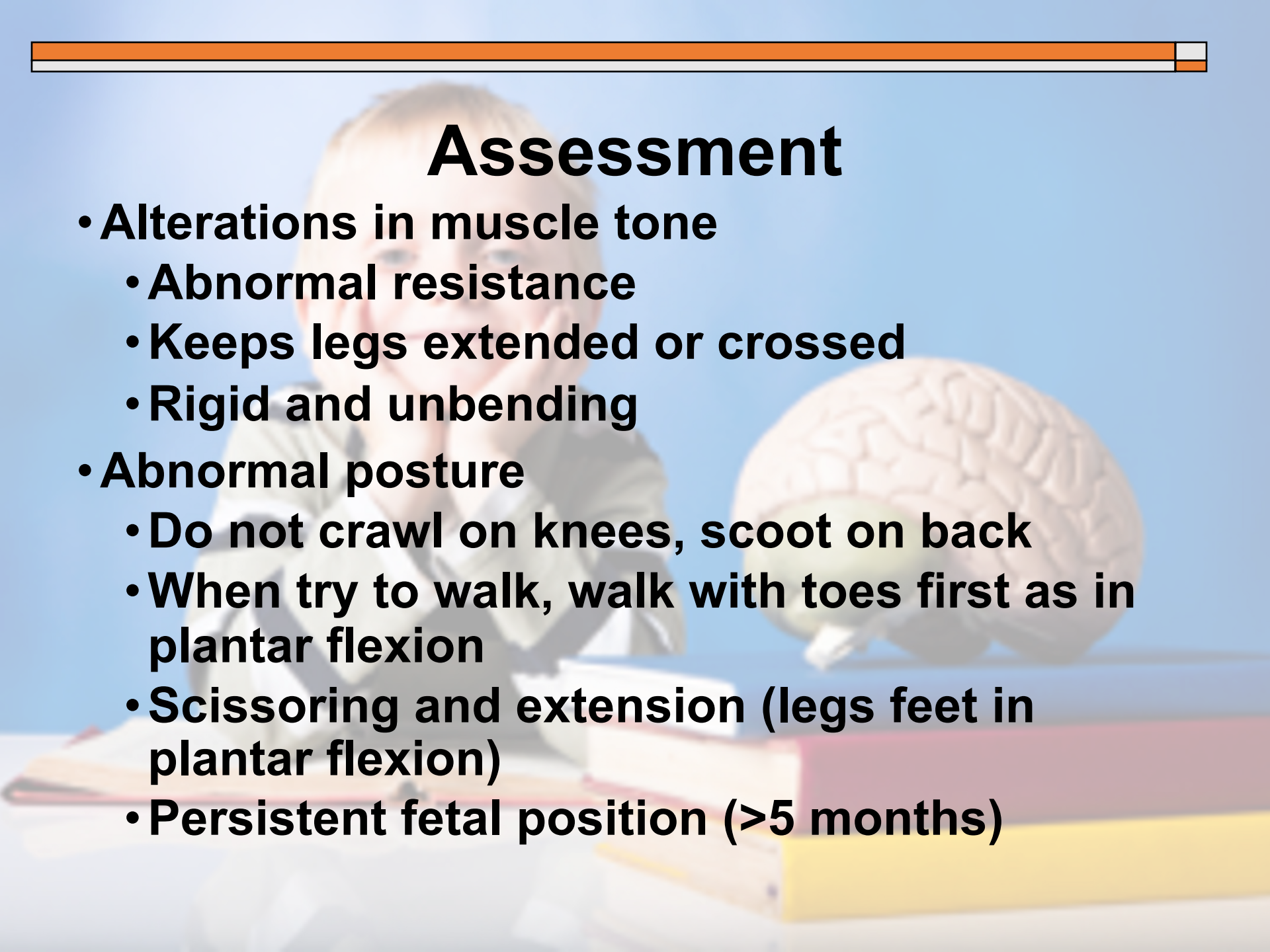
Assessment

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

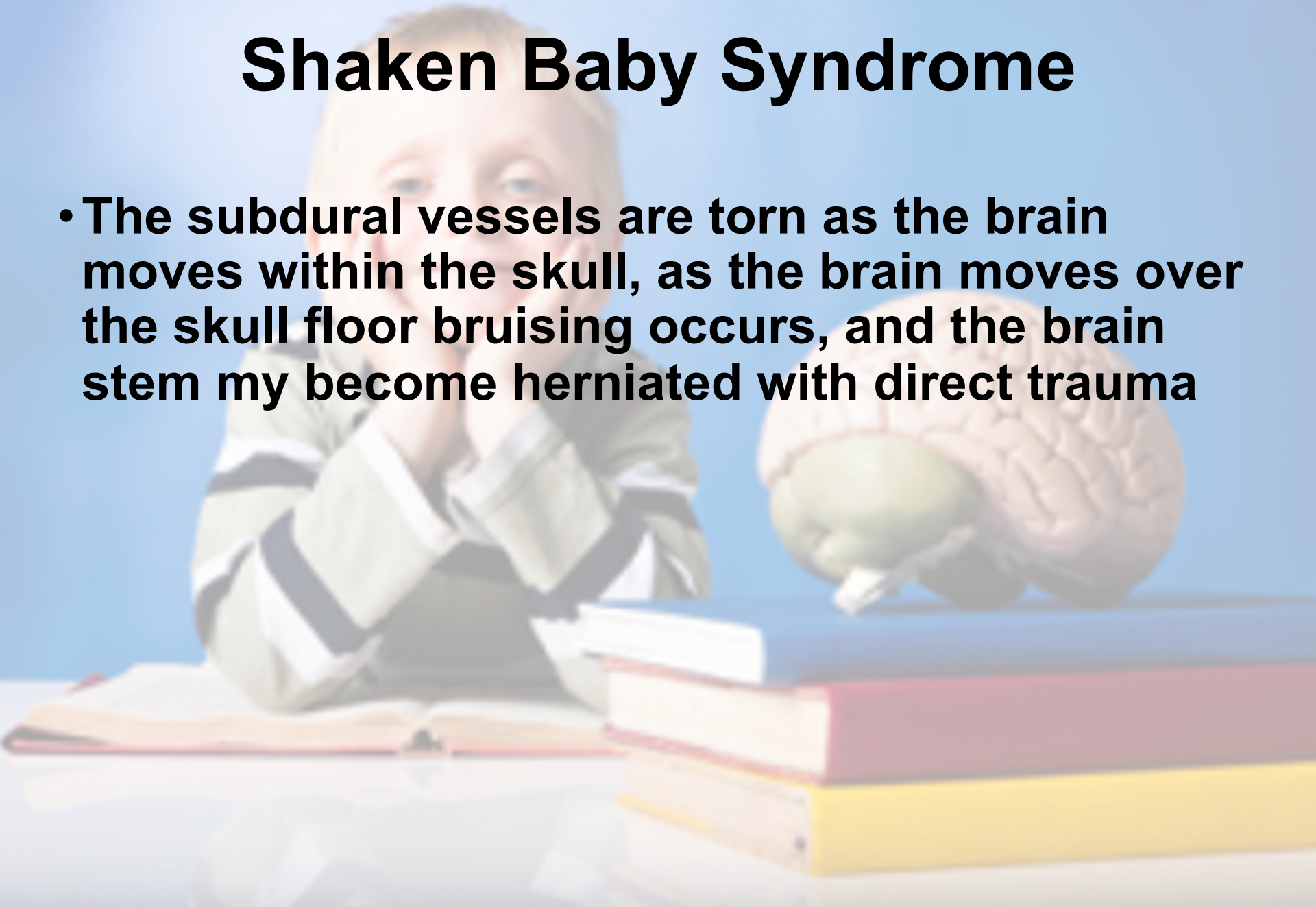
- 
- **EEG, CT, or MRI**
 - **Electrolyte levels and metabolic workup**
 - **Neurologic examination**
 - **Developmental assessment**

A young child with blonde hair is sitting at a desk, resting their chin on their hands. In front of them is a stack of books and a model of a human brain. The scene is set against a light blue background. A semi-transparent blue box with a black border is overlaid on the image, containing the text 'Head Injuries'.

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 may become herniated with direct trauma**

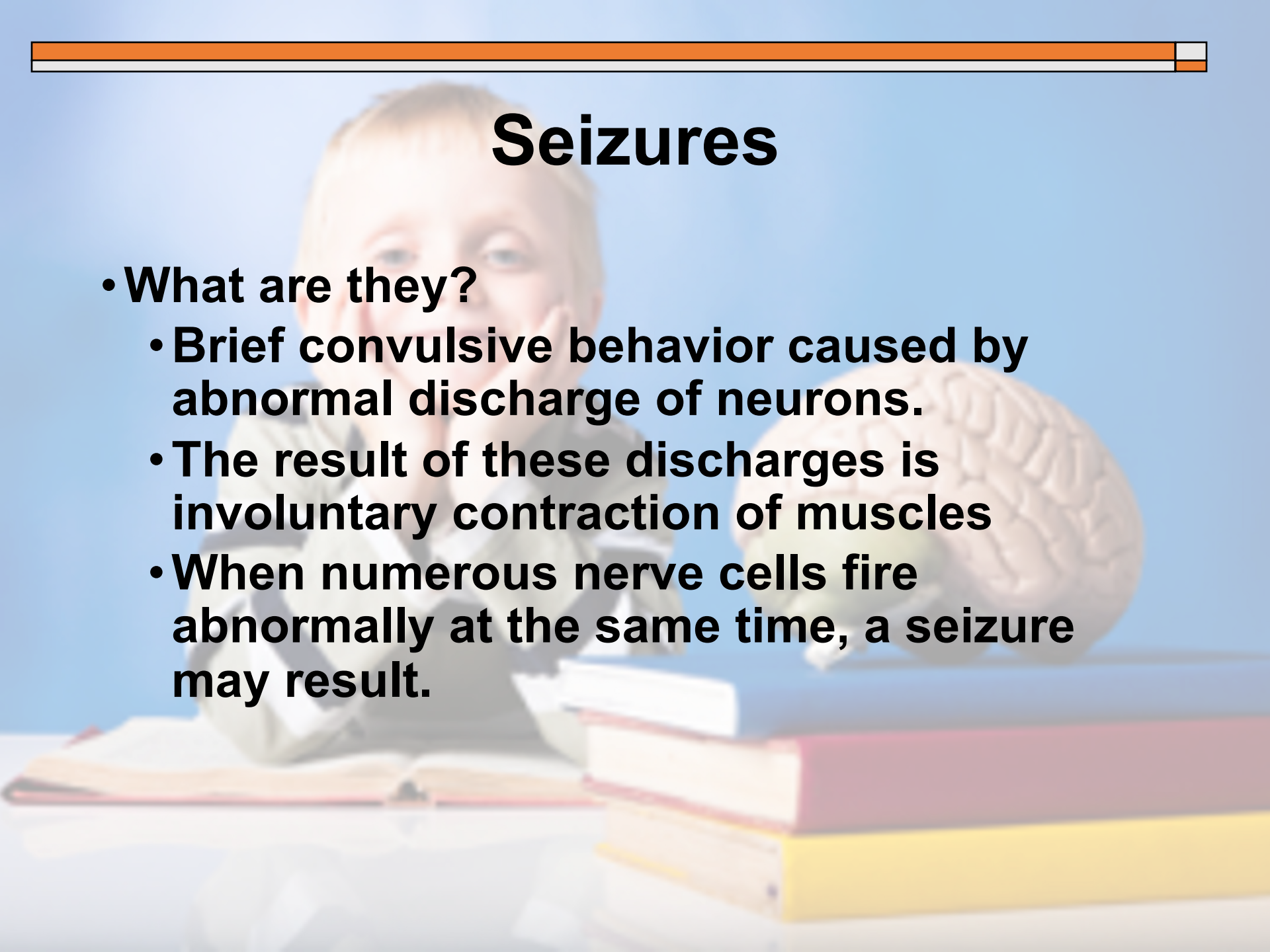



A young child with blonde hair is sitting at a desk, resting their chin on their hands. In front of them are several books, and to the right is a model of a human brain. The background is a light blue gradient. The text 'Seizure Disorders' is overlaid in a blue box with a black border.

Seizure Disorders

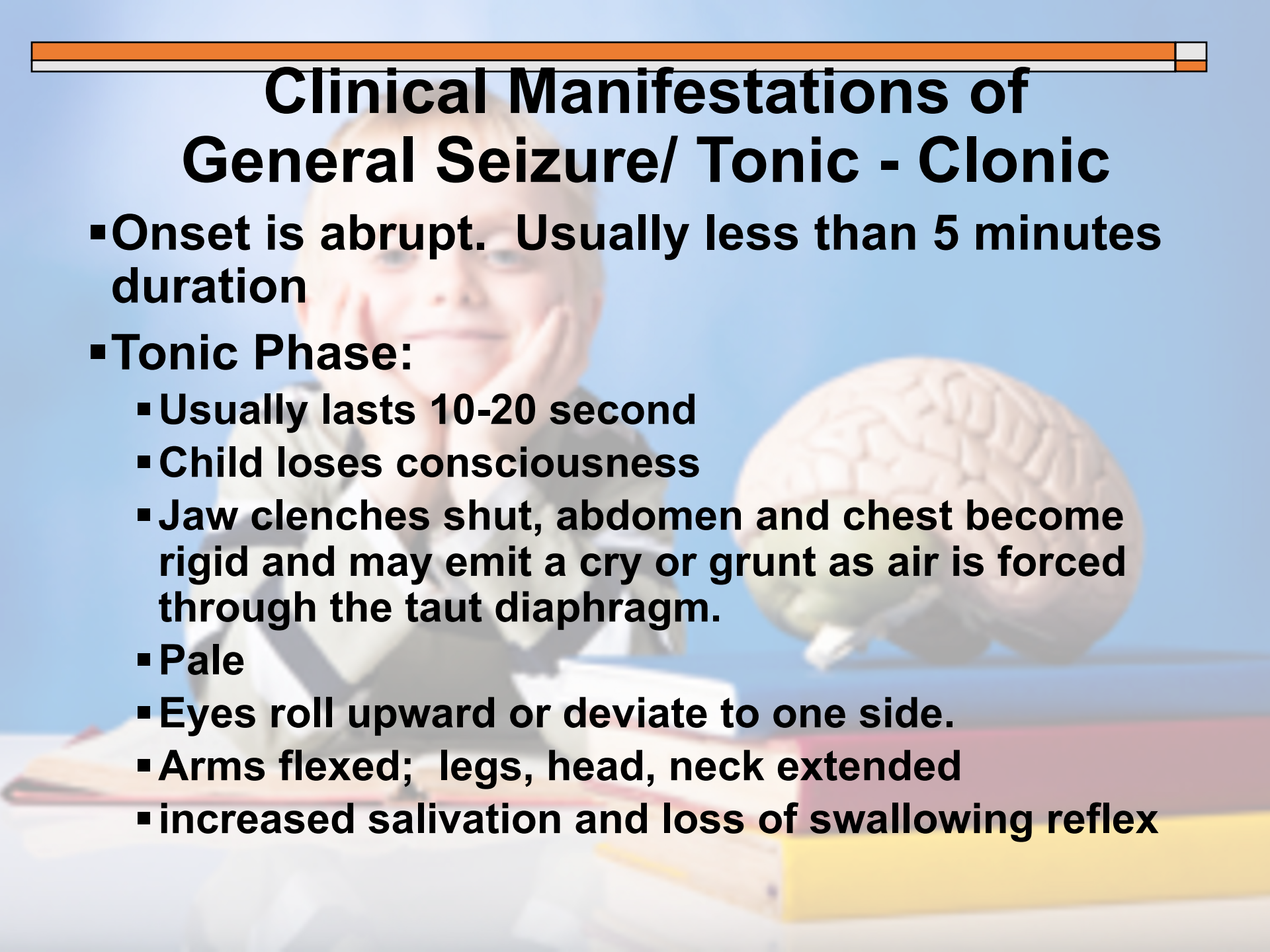


Seizures

- **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.**
- 

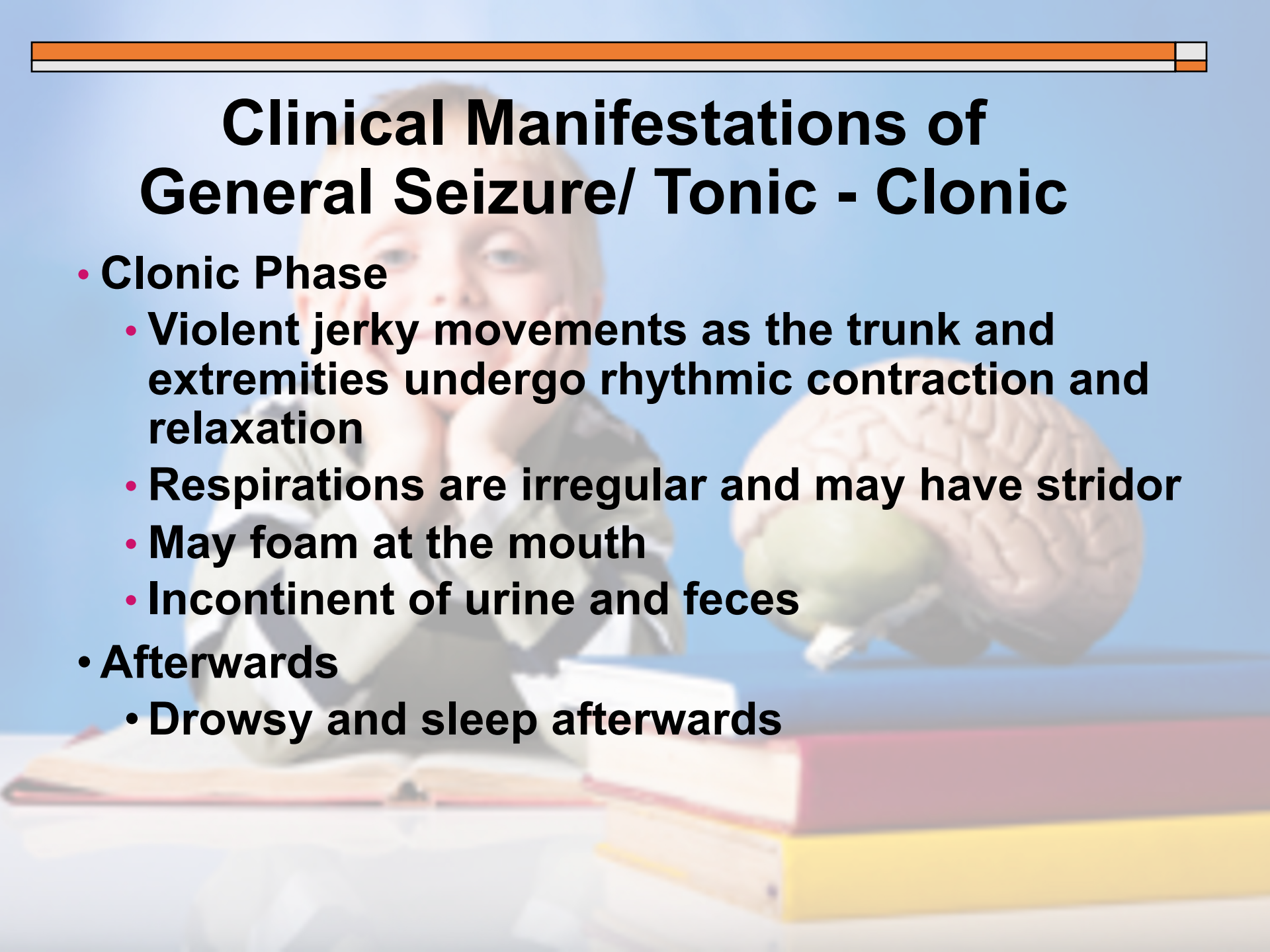


Clinical Manifestations of General Seizure/ Tonic - Clonic

- 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
- 
- A young child with blonde hair is sitting at a desk, looking towards the camera. The child's hands are near their face. In the background, there is a large, realistic model of a human brain. The desk has several books and papers on it. The overall scene is brightly lit with a blue background.



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**
- 

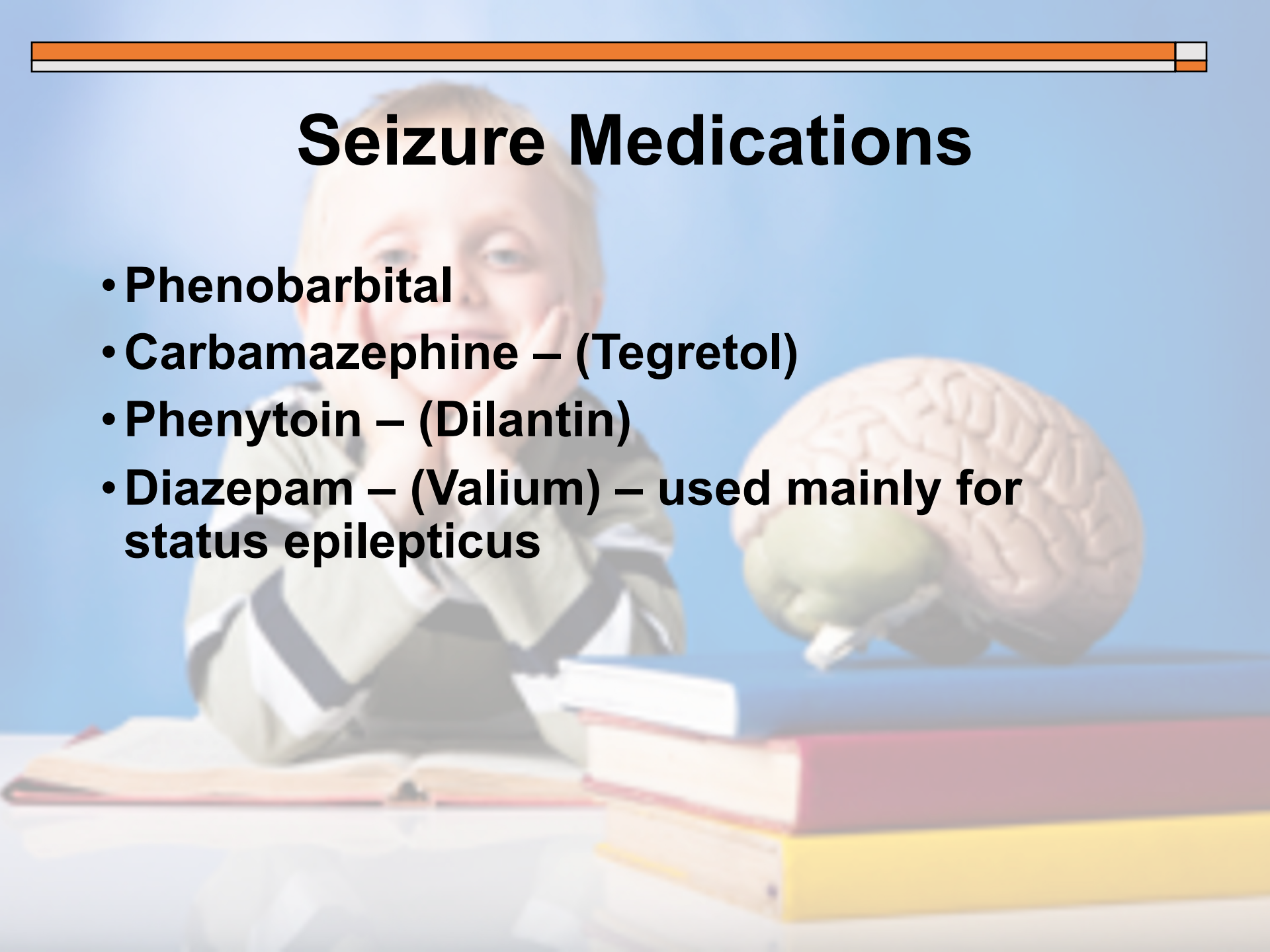
Diagnostic Tests

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





Seizure Medications

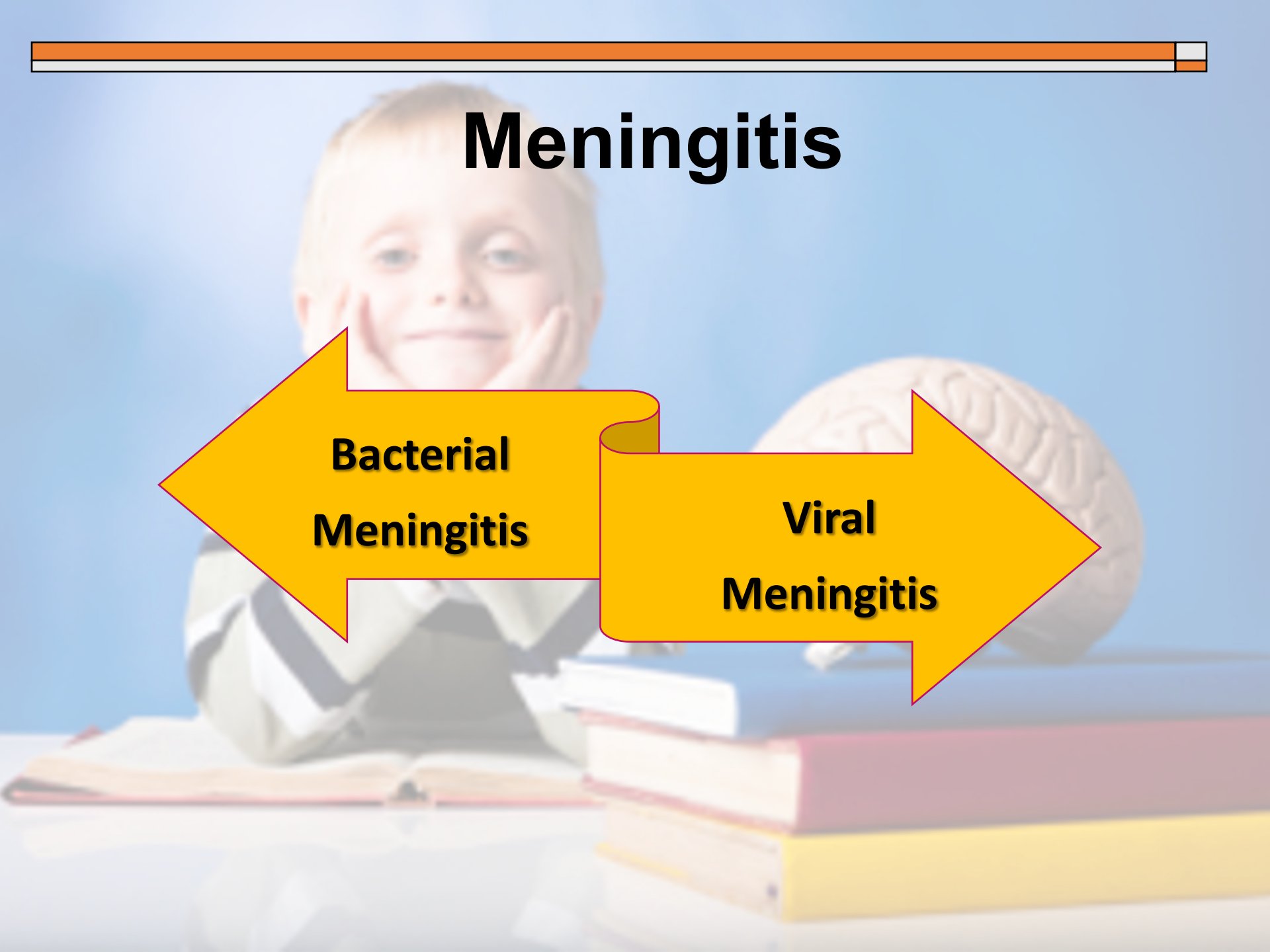
- **Phenobarbital**
 - **Carbamazepine – (Tegretol)**
 - **Phenytoin – (Dilantin)**
 - **Diazepam – (Valium) – used mainly for status epilepticus**
- 
- A young child with blonde hair is sitting at a desk, looking towards the camera. The child is wearing a light-colored shirt with dark stripes on the sleeves. On the desk in front of the child are several books stacked together. To the right of the books is a model of a human brain. The background is a solid light blue color.

A young child with blonde hair is sitting at a desk, resting their chin on their hands. In front of them are several books, including one with a red cover and one with a yellow cover. To the right, a large, realistic model of a human brain is visible. The background is a solid light blue color. The word "Meningitis" is written in a large, bold, black font with a blue shadow effect, centered over the image.

Meningitis



Meningitis



**Bacterial
Meningitis**

**Viral
Meningitis**



Bacterial Meningitis

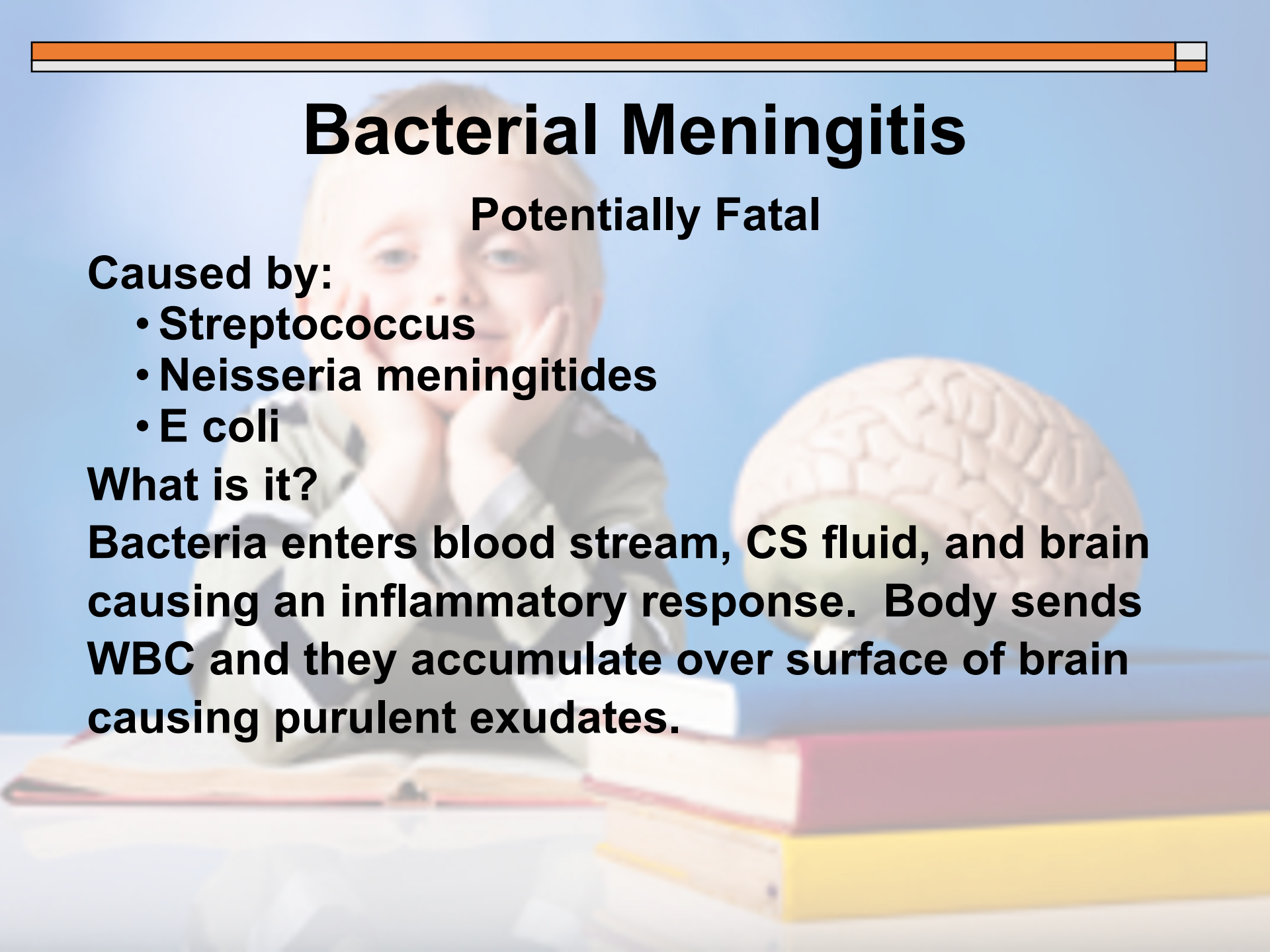
Potentially Fatal

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.

A child with blonde hair is sitting at a desk, resting their chin on their hand. In front of them are several books. To the right, a model of a human brain is visible. The background is a light blue gradient.



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**



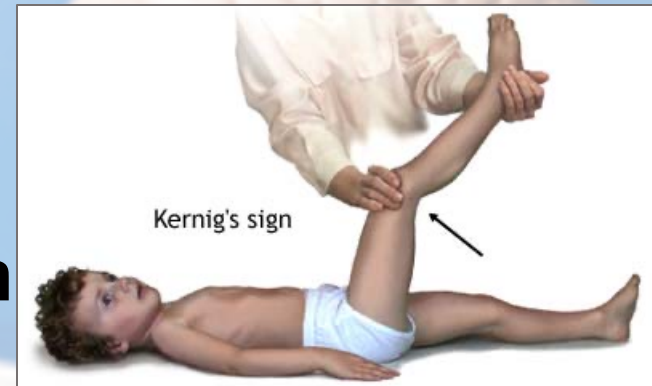
Assessment

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



Signs of Meningeal Irritation

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





Diagnostic Tests

The background features a young child with blonde hair, wearing a green and white striped shirt, resting their chin on their hands. To the right is a realistic model of a human brain. In the foreground, there are several books stacked on a white surface, including one with a red cover and one with a yellow cover. The entire scene is set against a light blue background.

- **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

