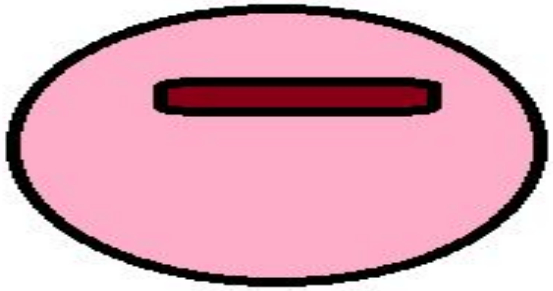
Background of the lecture

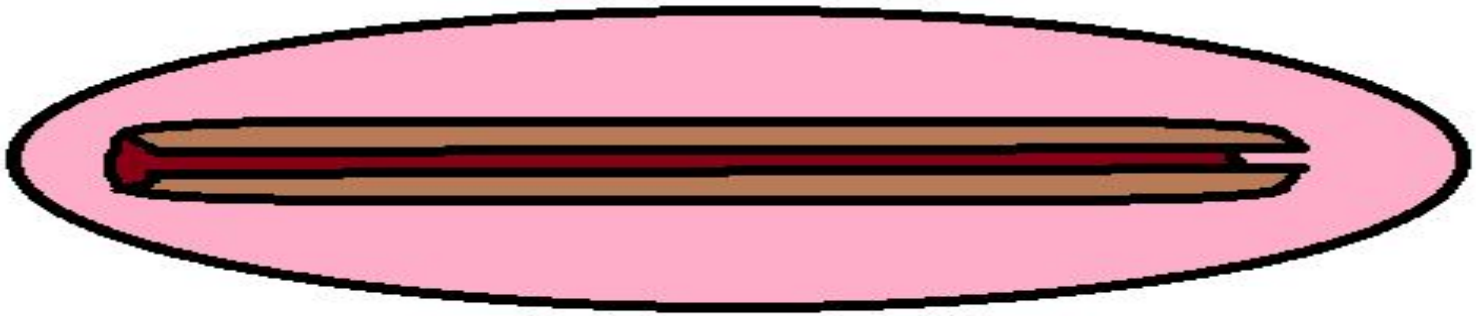
- **Development of CNS in embrio. Clinical evaluation of abnormalities.**
- **Features of CNS in fetus and newborn.**
- **Neurological examination**
- **Complaints&History.**
- **Level of consciousness (LOC)**
- **Mentality**
- **Head examination**
- **Evaluation of motor system**
- **Main semiotics of CNS disorders. Meningitis.**

Anatomo-physiological peculiarities of CNS in children and their clinical importance

The central nervous system appears at the beginning of the 3rd week as a slipper-shaped plate of thickened ectoderm, the neural plate.



Its lateral edges soon become elevated to form the neural folds. With further development, the neural folds become more elevated, approach each other in the midline, and finally fuse, thus forming the neural tube.



Neural tube defects account for the most congenital anomalies of the CNS and result from the failure of the neural tube to close spontaneously between the 3rd and 4th wk of in utero development. Neural tube defects (NTDs) involve the meninges, vertebrae, muscles, and skin.

Neural tube defects (NTDs)

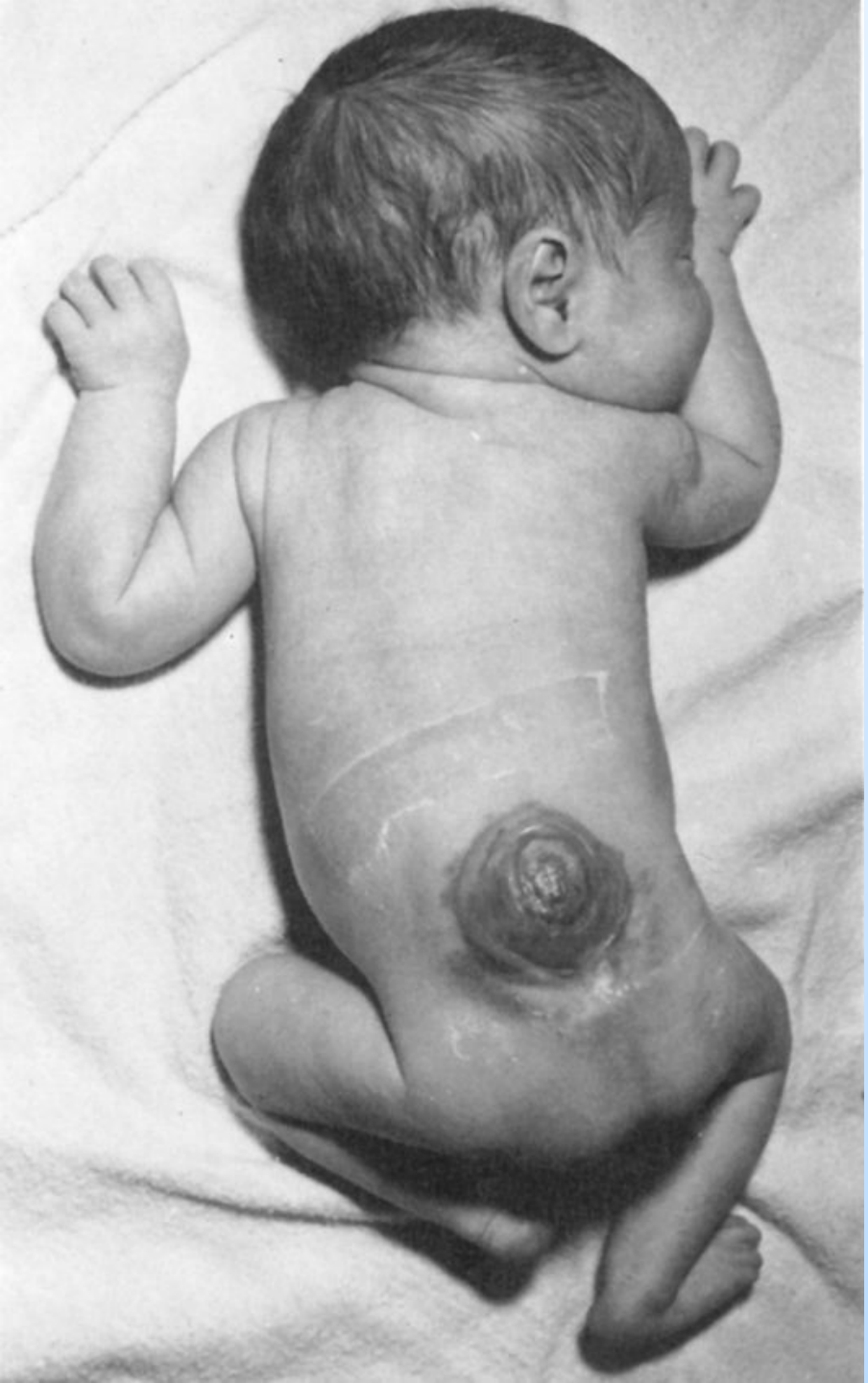
- spina bifida occulta
- meningocele
- myelomeningocele
- encephalocele
- anencephal

Neural tube defects (NTDs)

- can be diagnosed prenatally by ultrasound, and by determination of ά-fetoprotein (AFP) levels in maternal serum and amniotic fluid. The cranium or vertebra can be visualized since 12 weeks of gestation, and defects can be detected.
- Recent evidence indicates that folic acid (folate) reduces the incidence of NTDs in certain populations.

Neural tube defects (NTDs)

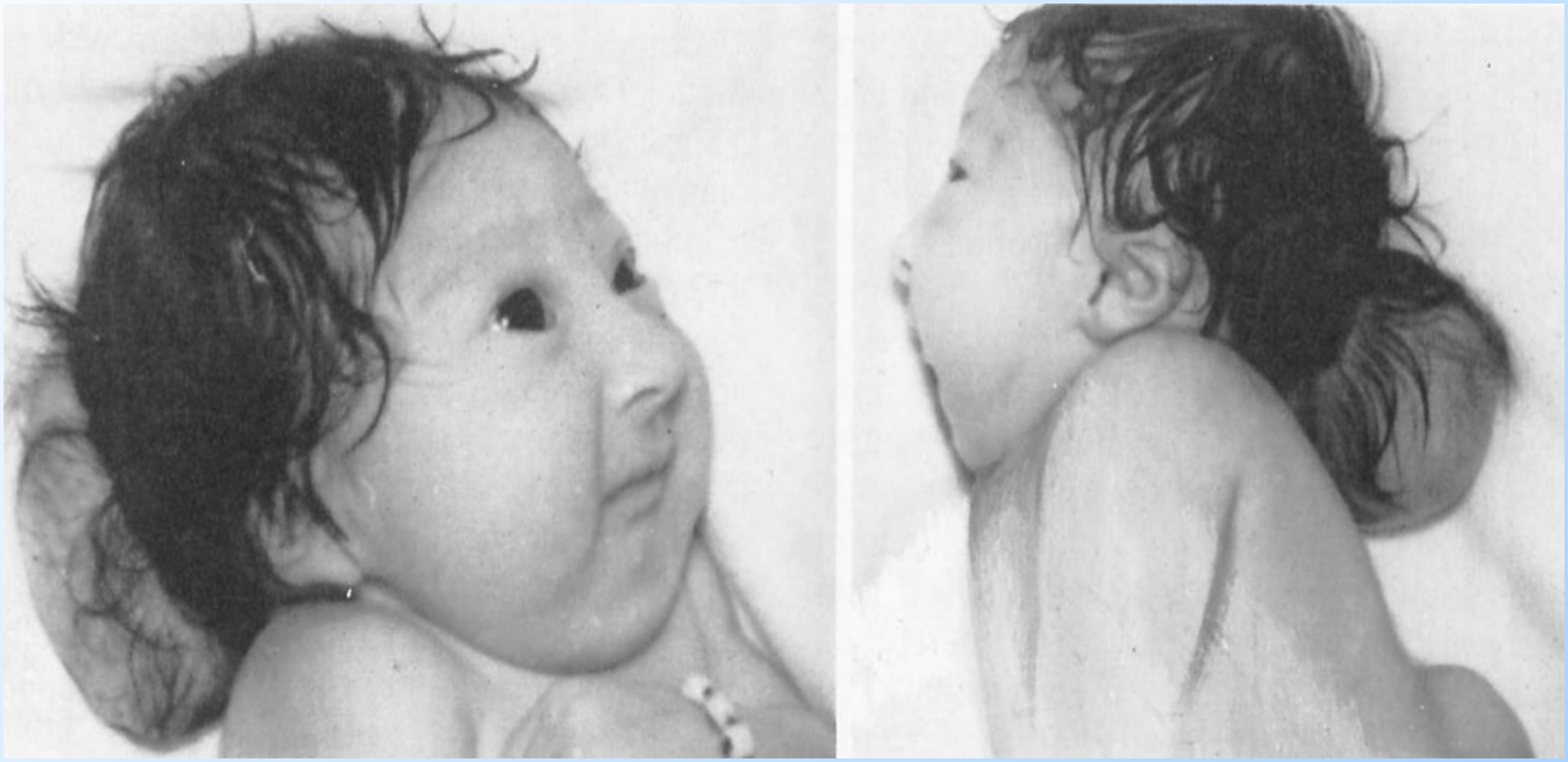
- **Meningocele (Meningoencephalocele)** is herniation of meninges and brain(medulla) through a defect in the skull or vertebra split producing a fluid-filled sac in the occipital or lumbar region.



Neural tube defects (NTDs)

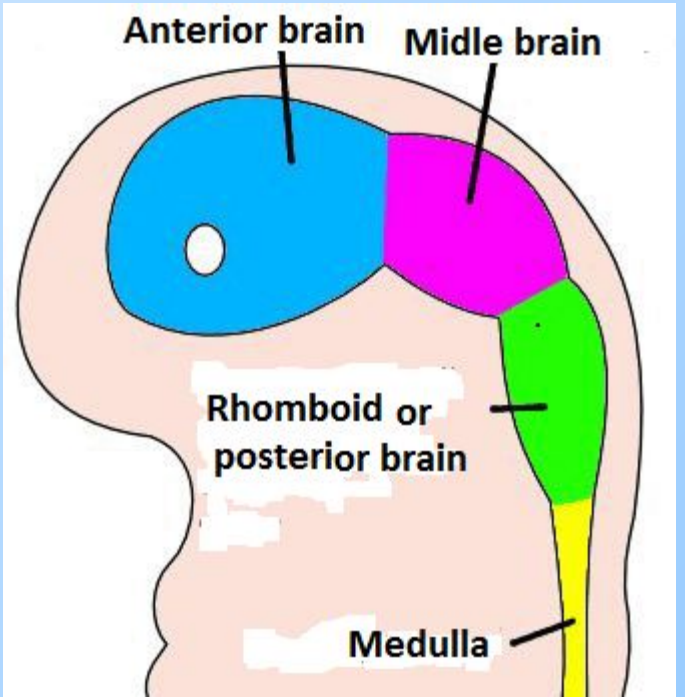
lumbar meningomyelocele in a 3-day-infant

Occipital meningoencephalocele

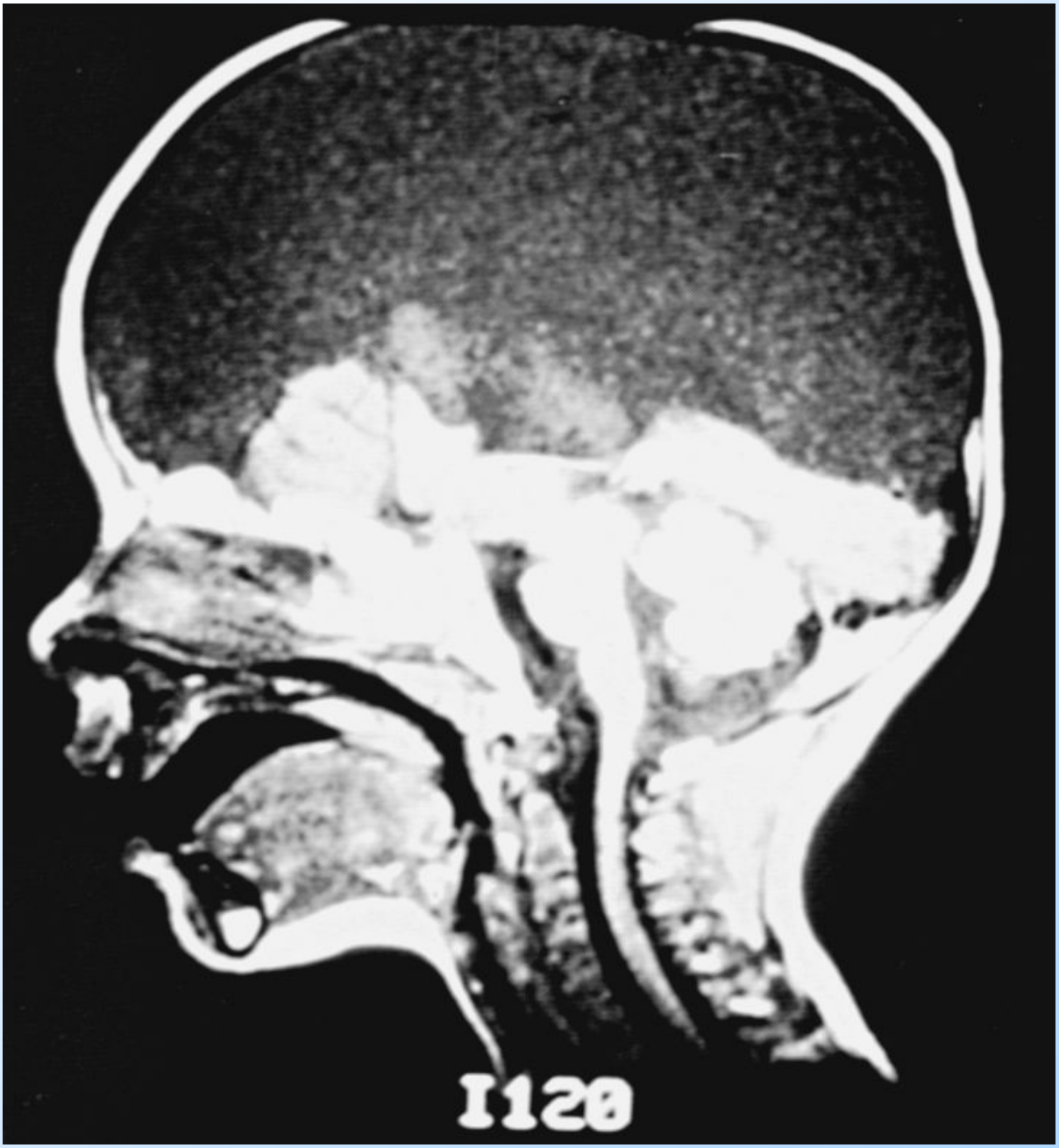


The sloping forehead and small head circumference are evident, although progressive ventricular enlargement often subsequently occurs in such children.

In embryo at its cephalic end of the neural tube the brain bladders are forming from which all parts of the brain are originated within approximately 2-3 months of in utero development, including neural parts of ear, eye and sense of smell.



Hemispheres of the brain are developed from the first brain bladder. Errors of embryogenesis, connected with an action of a teratogen (the factor inducing abnormalities) can lead to severe pathology of the fetus and newborn, for example, *microcephaly and anencephaly.* The cerebral hemispheres and cerebellum are usually absent, and only a residue of the brain stem can be identified when anencephaly presents.



Hydranencephaly

Magnetic resonance imaging (MRI) shows the brain stem and spinal cord with some remnants of the cerebellum and the cerebral cortex.

The remainder volume of cranium is filled with CSF

CSF (cerebral spinal fluid)

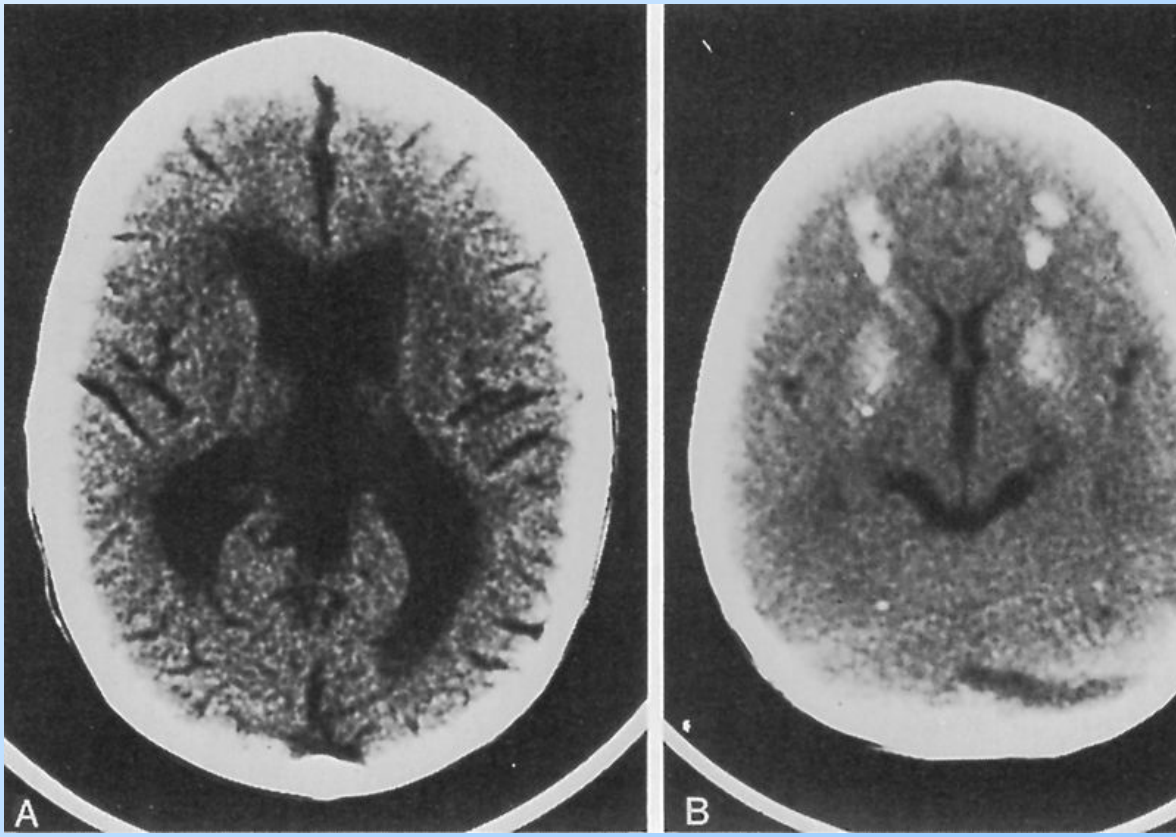
- **CSF flow results from the pressure gradient that exists between the ventricular system and venous channels. The intraventricular pressure is twice higher than the pressure in the superior sagittal sinus.**

CSF

- **Hydrocephalus** resulting from CSF accumulation inside the brain is called internal hydrocephalus.

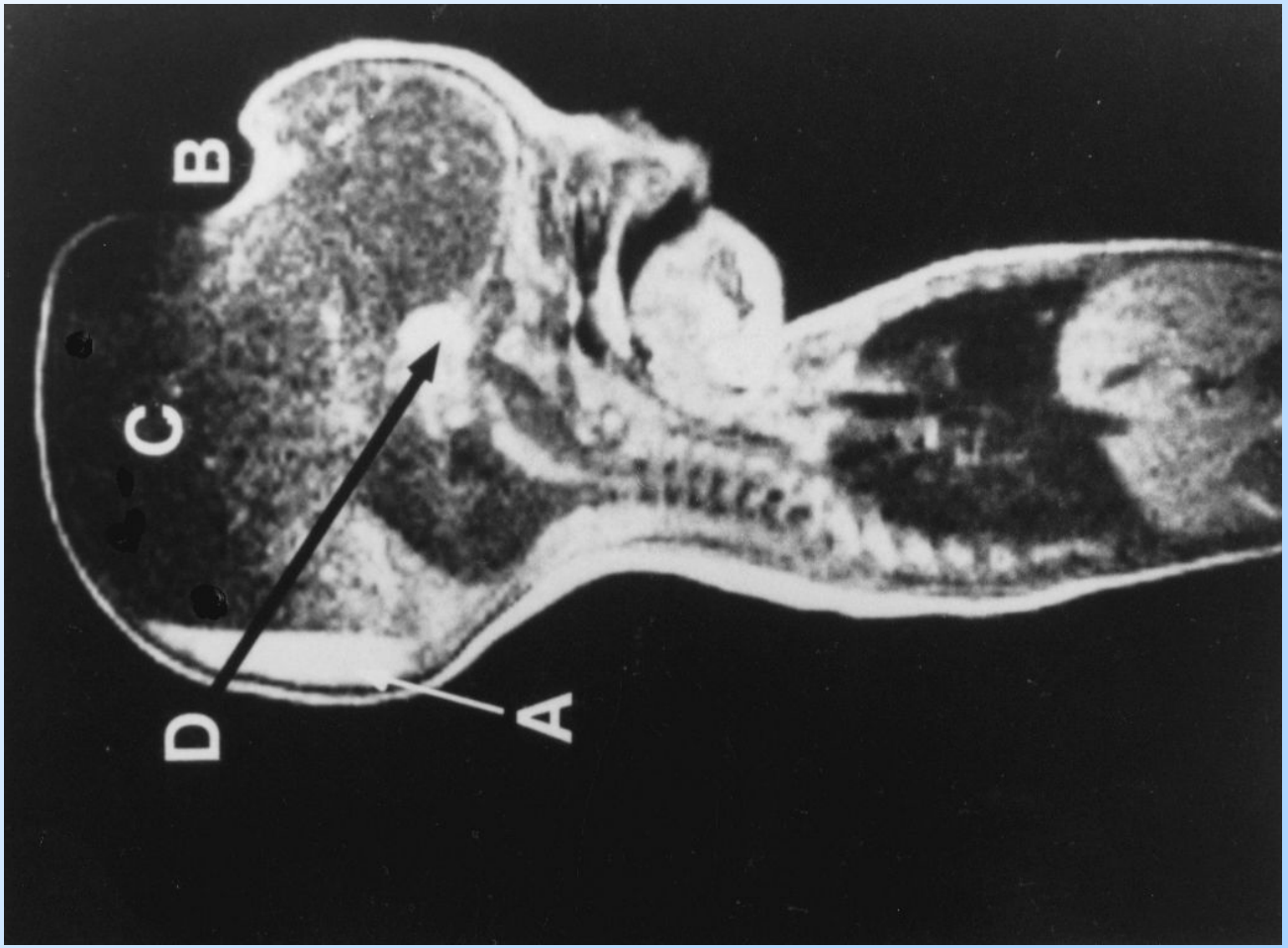
The cranial computerized tomogram (CT) of the infant`s brain with congenital virus-associated encephalopathy

Cerebral atrophy with enlarged ventricles and widened sulsi (internal hydrocephalus).



- CSF is absorbed primarily by the arachnoid villi through tight junctions of their endothelium by the pressure forces.
- **Hydrocephalus** resulting from malfunction of the arachnoid villi is called **nonobstructive or communicating hydrocephalus.**

External hydrocephalus in the newborn with in utero infection of the brain (MRI)



Features of CNS in fetus and newborn

- The brain development is characterizing by gradual formation and maturation of brain structures from ontologically "old" to "young“. Note the line: the spinal cord, brain stem, subcortical formations, cerebellum and at last the cortex are making mature.
- First months of life there is some functional minority of regulating activity of the cortex in favour to the subcortical formations with *domination of thalamopallidal and striopallidal areas.*
- The child’s brain contains more protein than the brain of the adult. Cerebral proteins make the tissues of brain hydrophilic and bent them to cellular edema.

Features of CNS in fetus and newborn (continue)

- There is not clear differentiation of the brain’s layers (grey and white substances are indistinctly differentiated among themselves).
- The gyri and sulci of the cortex are not deep that reduces the absolute and relative area of the child's cortex in comparison to adult.

Features of CNS in fetus and newborn (continue)

The blood-brain barrier (BBB) of the fetus and newborn

- is normally indiscriminately permeable, allowing protein and other large and small molecules to pass freely between the cerebral vessels and the brain.
- becomes mature only to the ending of the neonatal period

Features of CNS in fetus and newborn

- Central and peripheral neurons form myelin coating gradually. Myelinization finally finishes only after the 3-rd year of life.
- Due to undeveloped myelinization in children long time the cortex physiology will be characterizing to be *bent to generalization of irritation and difficulties of neuronal braking*.

Features of CNS in fetus and newborn

- The features of the brain vascular system of fetus when anastomoses develop insufficiently make the brain of premature newborn easily vulnerable to hypoxia, mechanical, and thrombotic damages. This can promote for cerebral ischemia and hypoxia with form of cerebral palsy.

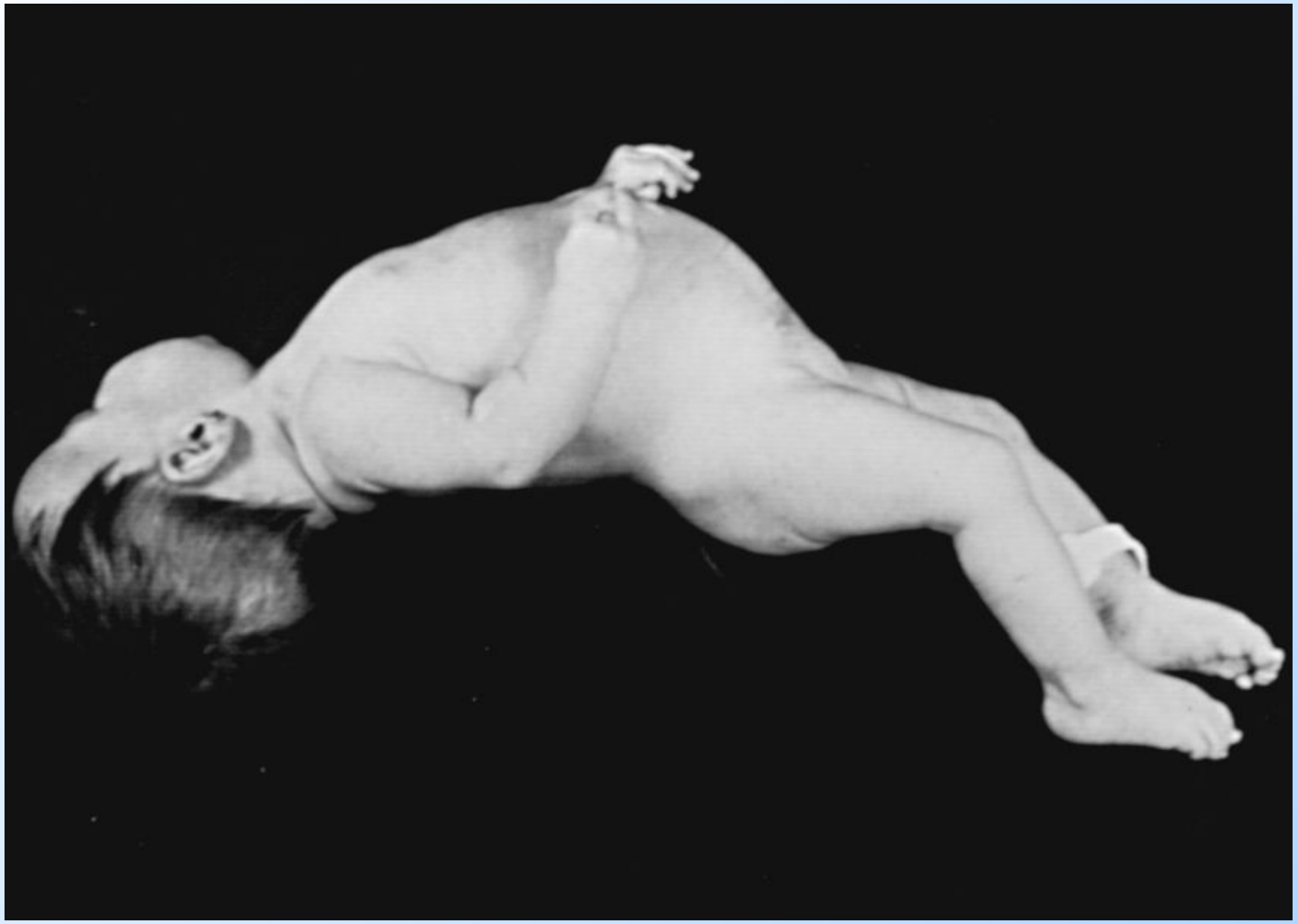
Neurological examination

Neurologic evaluation of the child. Complaints&History.

- **Seizures (convulsion)** are involuntary, violent contraction of muscles. Seizures may be:
- tonic or clonic,
- focal or generalized.

Tonic seizures are characterized by increased tone or rigidity.

Clonic seizures consist of rhythmic muscle contraction and relaxation, when stereotypic, wide movements observe in extremities and other parts of a body.



Opisthotonus in a brain-injured infant.
This is the tonic seizure.

Objective neurological examination

of the child should include 4 main diagnostic aspects:

- 1. Level of consciousness (LOC)
- 2. Mentality
- 3. Head examination
- 4. Evaluation of motor system

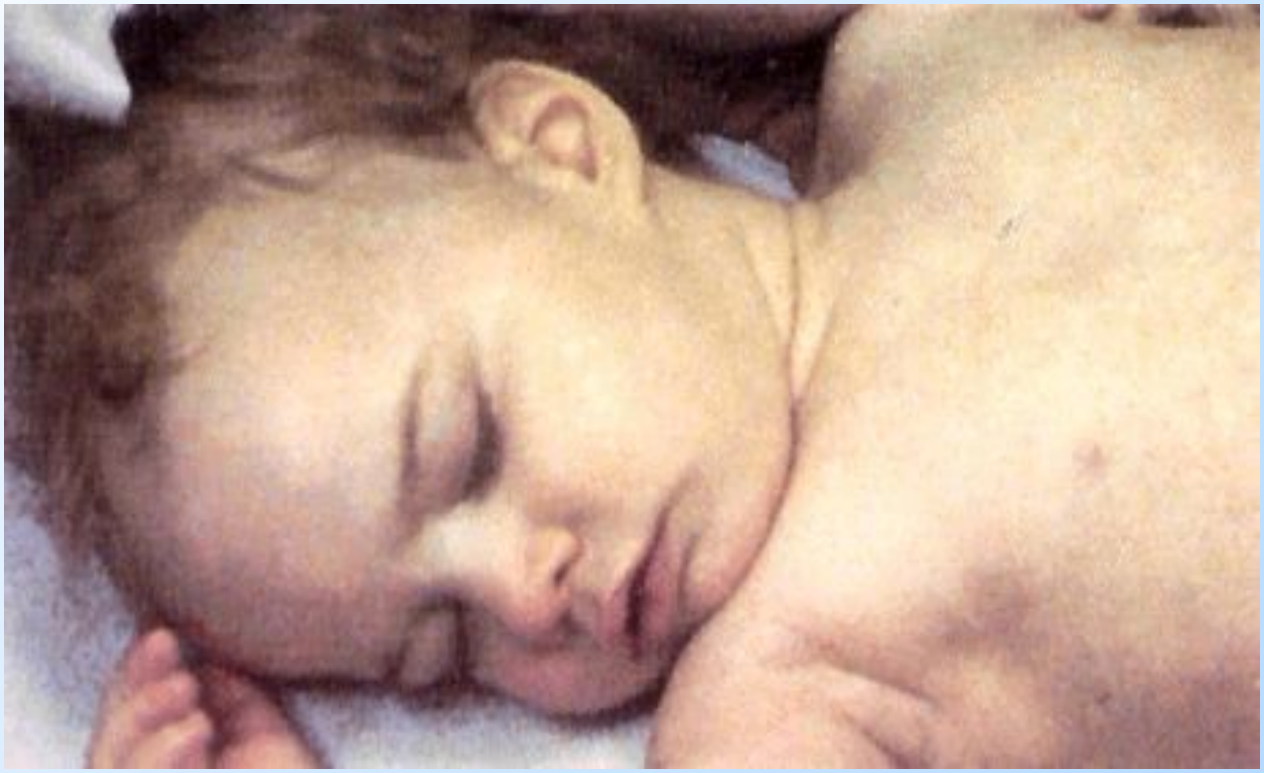
Level of consciousness (LOC)

A well child is conscious, alert and responsive

Level of consciousness (LOC)

- ***Lethargy or pathological sleepy (somnolence)*** is possible to determine as an unusual sleep of the patient.
- ***Confusion.*** The responses of confused patients demonstrate a failure to comprehend their surroundings. The patient is unable to estimate direction or location, is apt to be disoriented in time and may misidentify people.
- ***Coma*** is absence of consciousness.

Level of consciousness (LOC)



- This is a child with meningitis. The child is somnolent and can not arouse. Note the face of a gray color.

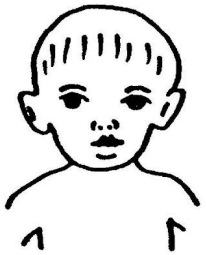
Stages of coma

1. **Stupor**: The stuporous patient arouses from sleep only after painful stimuli. Verbal responses are slow or even absent. The patient lapses into an unresponsive state when the stimulus ceases.
2. **Light coma**: the patient has response to painful stimulus.
3. **Deep coma**: there is no response to painful stimulus.
4. **Terminal coma**: coma with a muscular relaxation and apnea.

Mental development

Mental Development

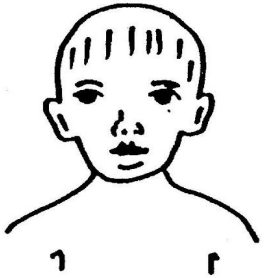
Infancy
(first year)



Social development

- Smiling
- Laughing
- Recognition of mother

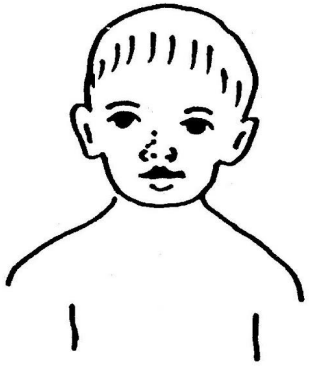
Early Childhood
(1 — 6 years)



Speech development

- Words
- Telegraphic sentences
- Fluent speech

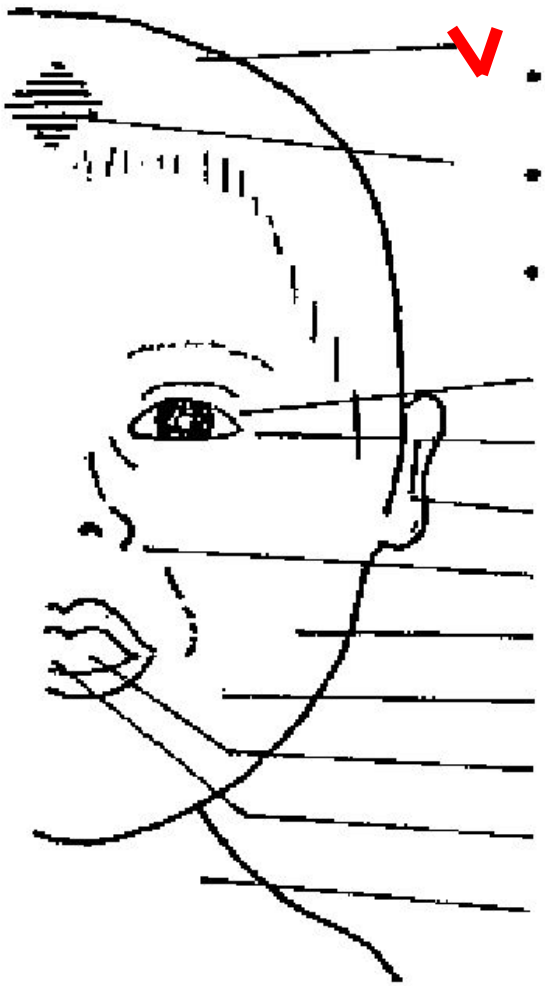
Late Childhood
(6 — 12 years)



Learning ability
(School achievement)

Head size

- **Head examination :**

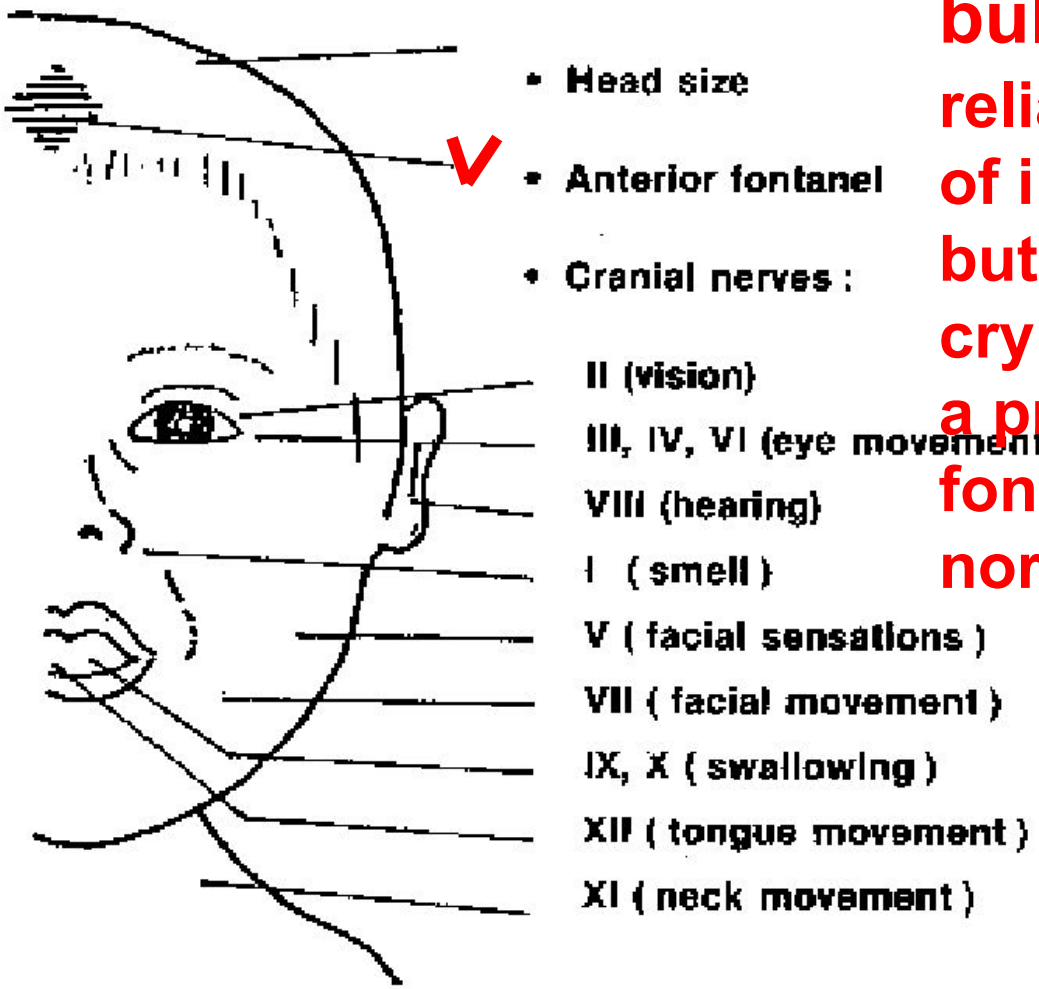


- **Head size**
- **Anterior fontanel**
- **Cranial nerves :**
  - **II (vision)**
  - **III, IV, VI (eye movement)**
  - **VIII (hearing)**
  - **I (smell)**
  - **V (facial sensations)**
  - **VII (facial movement)**
  - **IX, X (swallowing)**
  - **XII (tongue movement)**
  - **XI (neck movement)**

Enlarged head?

AF&PF

- **Head examination :**

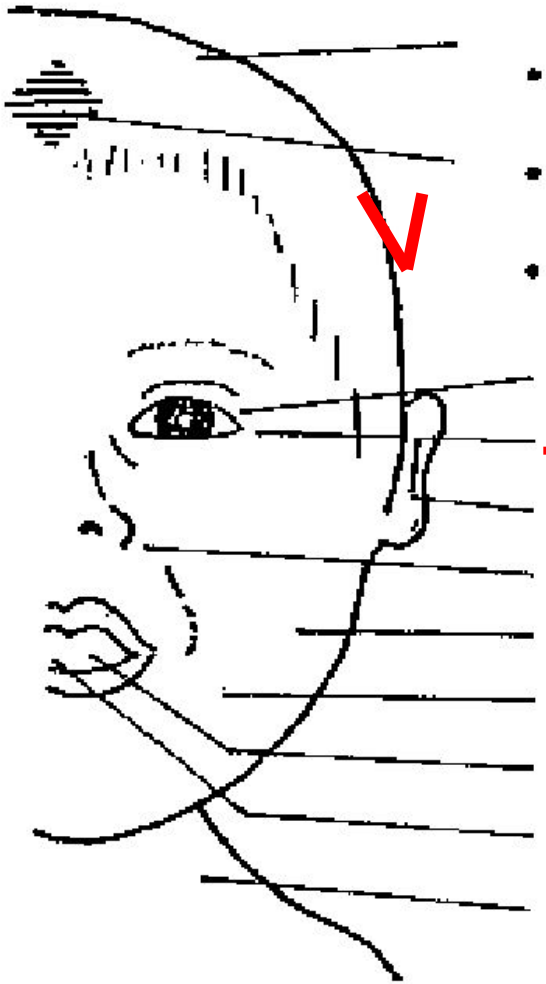


A fontanel bulging is a reliable indicator of increased ICP, but vigorous crying can cause a protuberant fontanel in a normal infant.

ICP-intracranial pressure

Cranial nerves

- Mentality
- Head examination :



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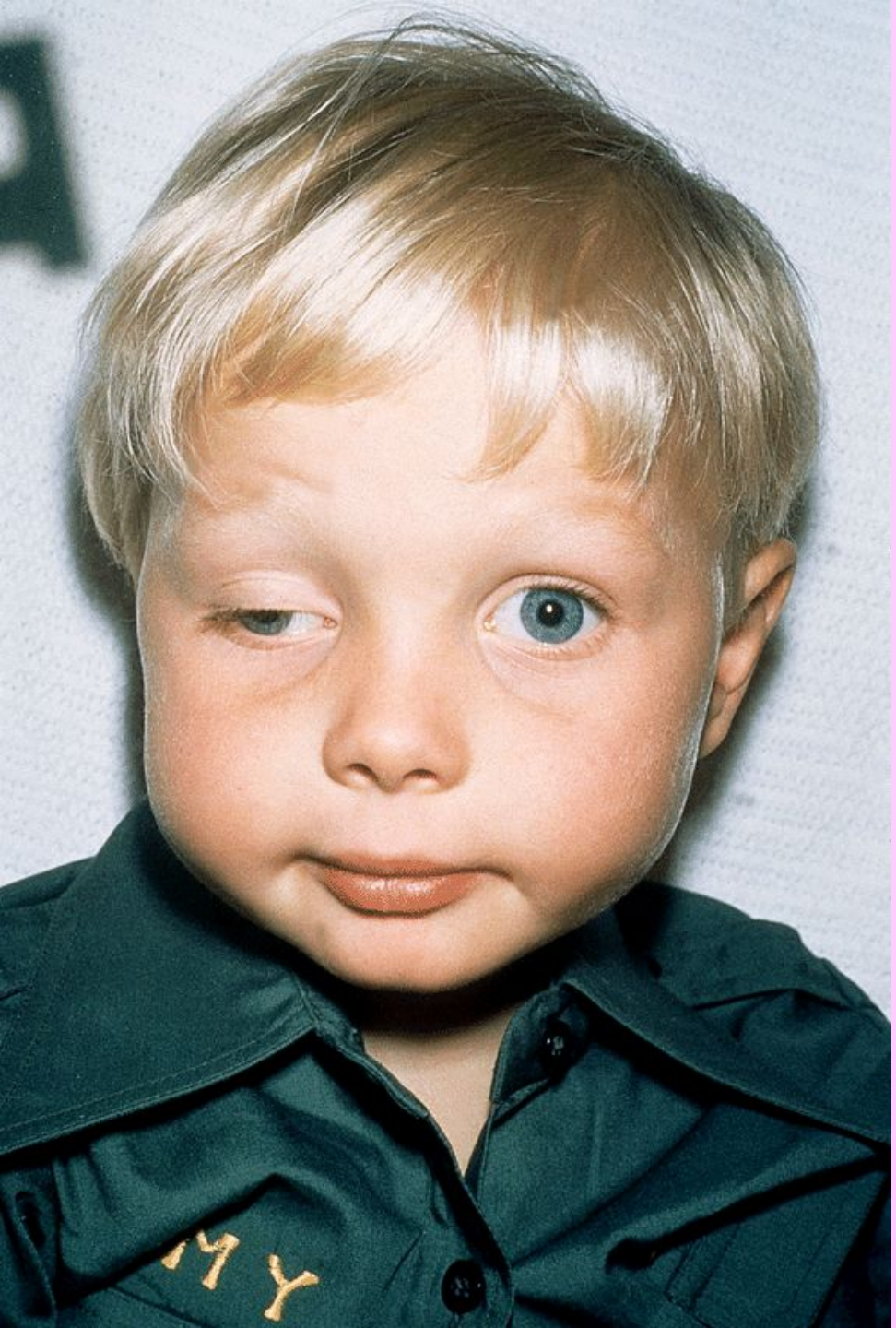
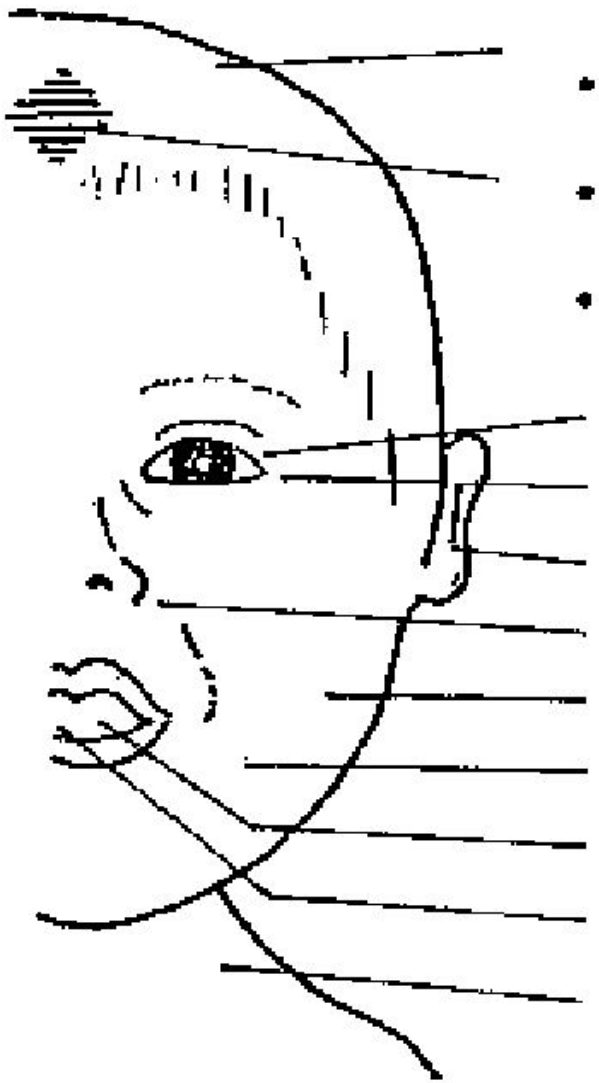


Oculomotor (3-rd) nerve paresis: ptosis (impossibility to lift an upper eyelid) and removal of an eyeball laterally (temporally).

- **Head examination :**



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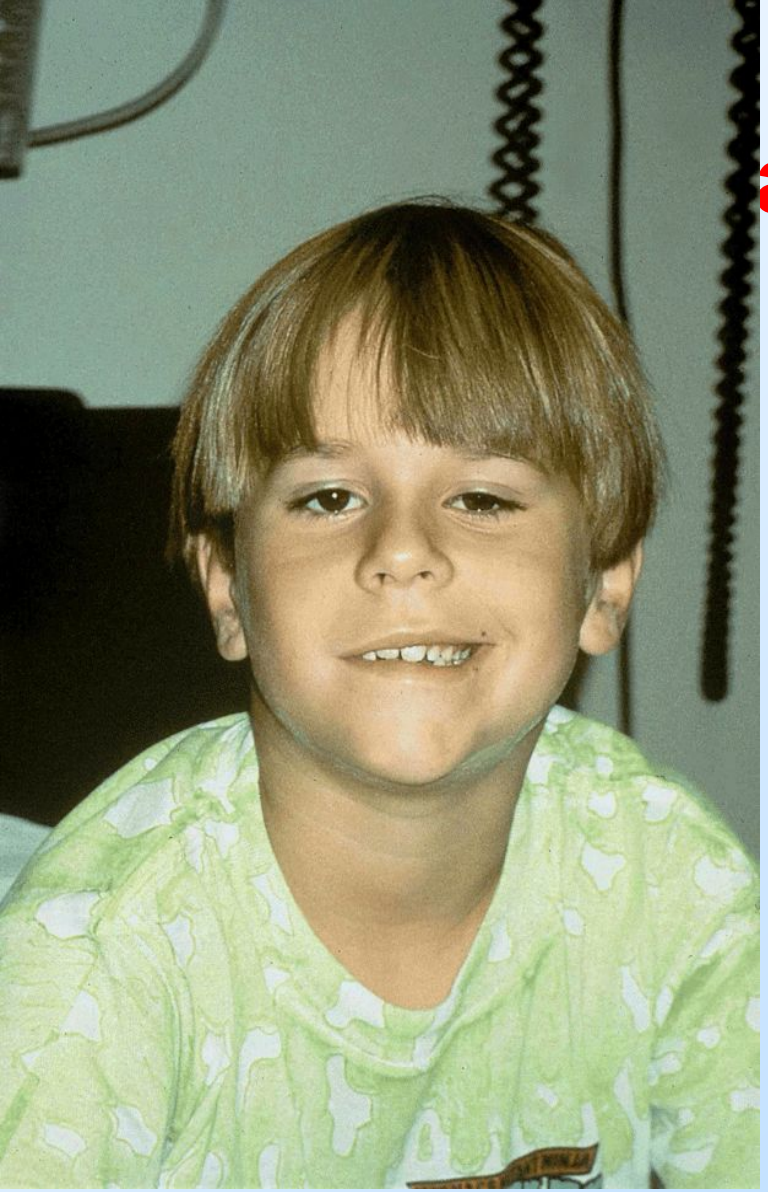
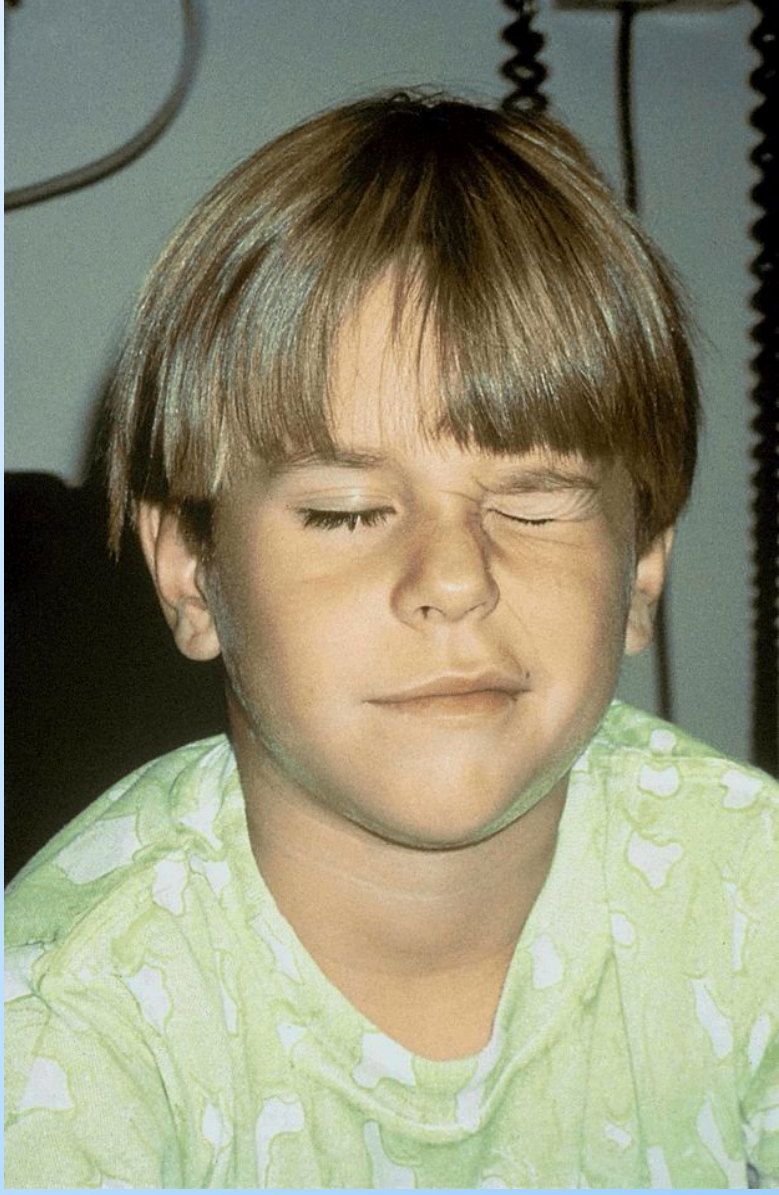
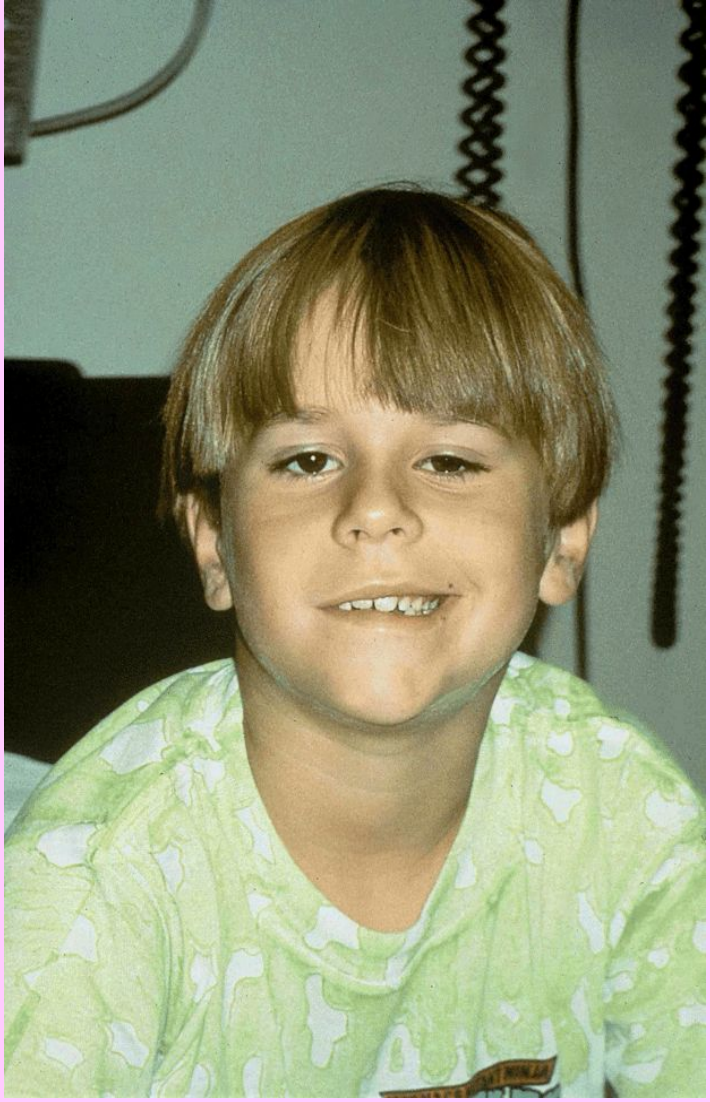
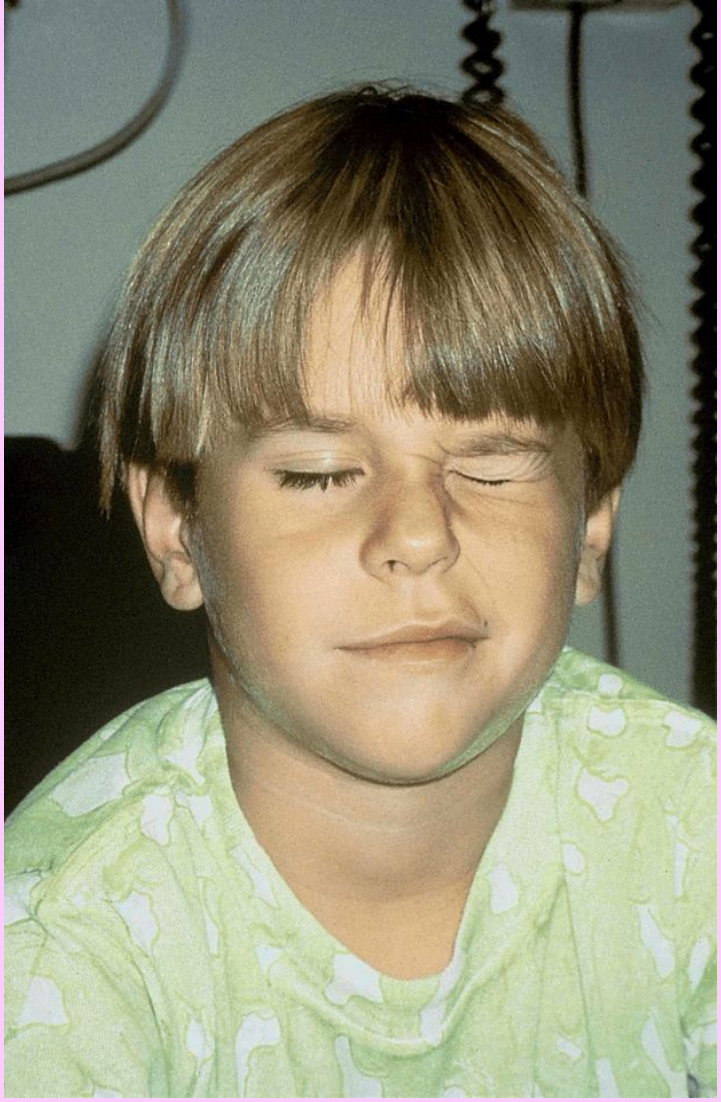


alsy

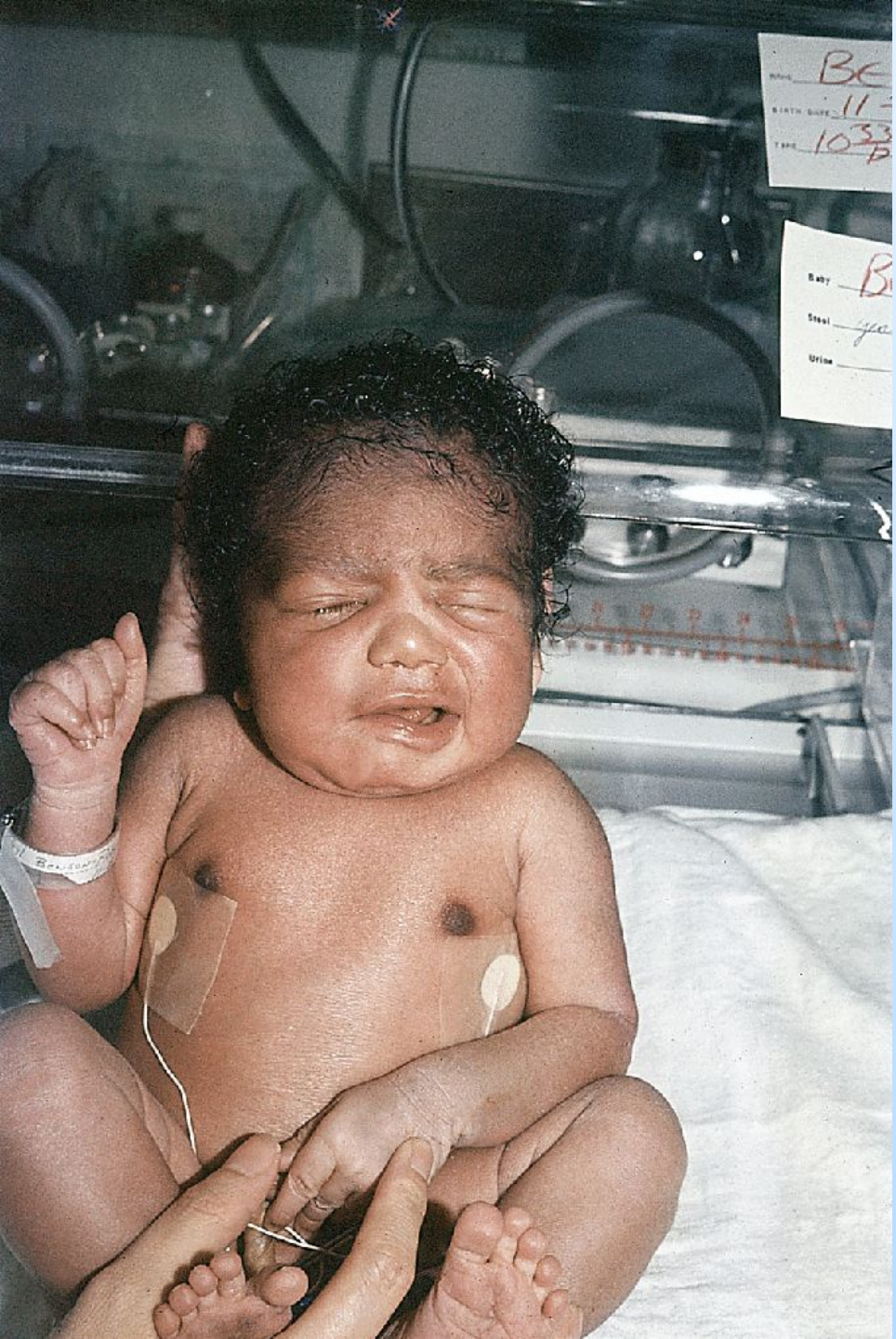


Facial nerve palsy



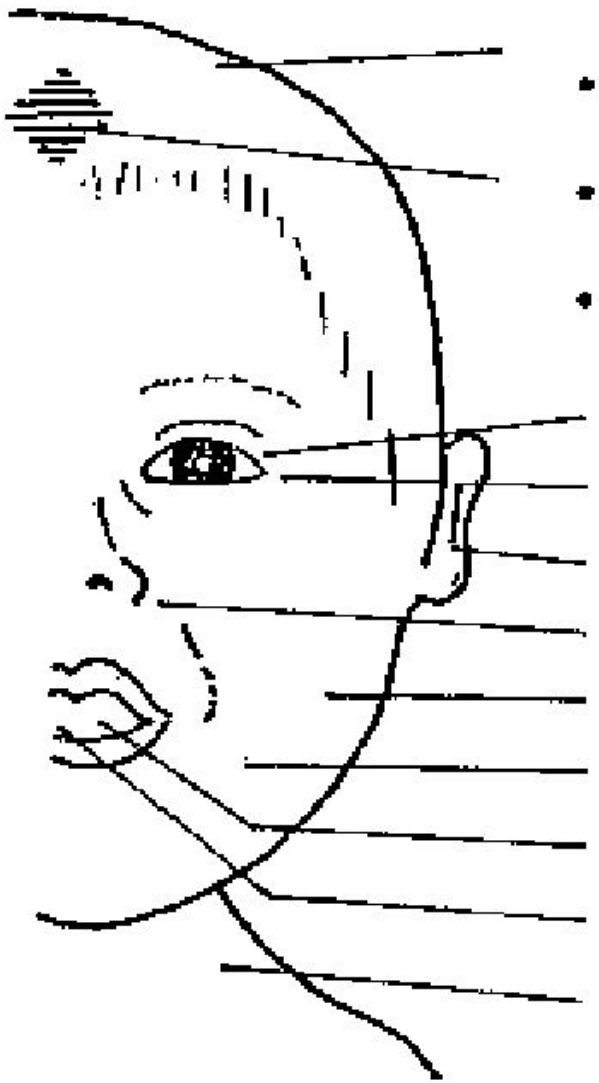


Facial nerve palsy. Notice the loss of the nasolabial fold and the mouth deviated to the left when he smiles.



A newborn with right facial palsy

- **Head examination :**



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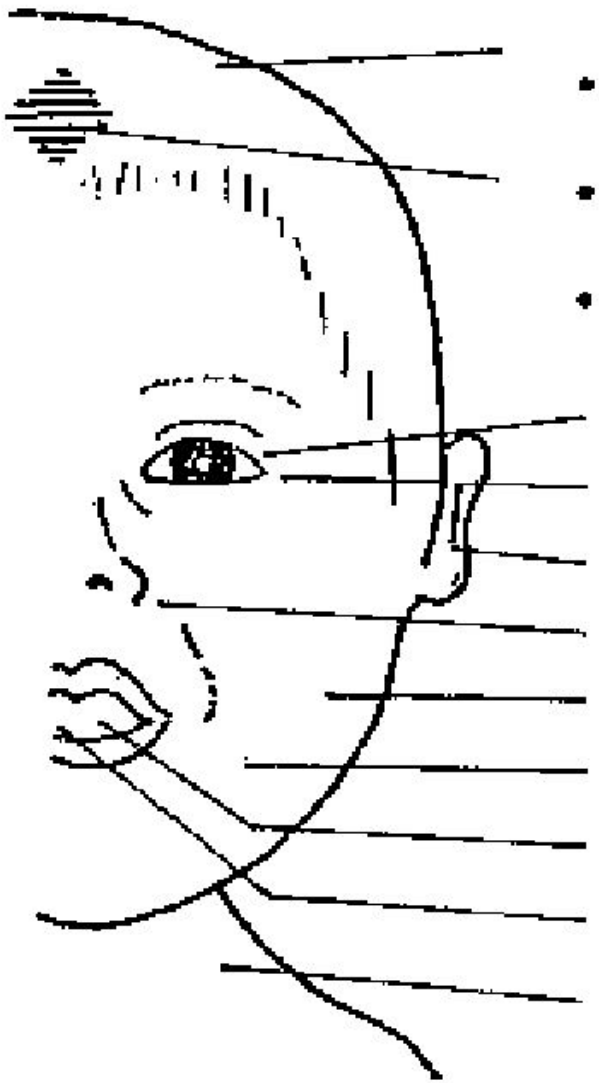
choking

- **Head examination :**



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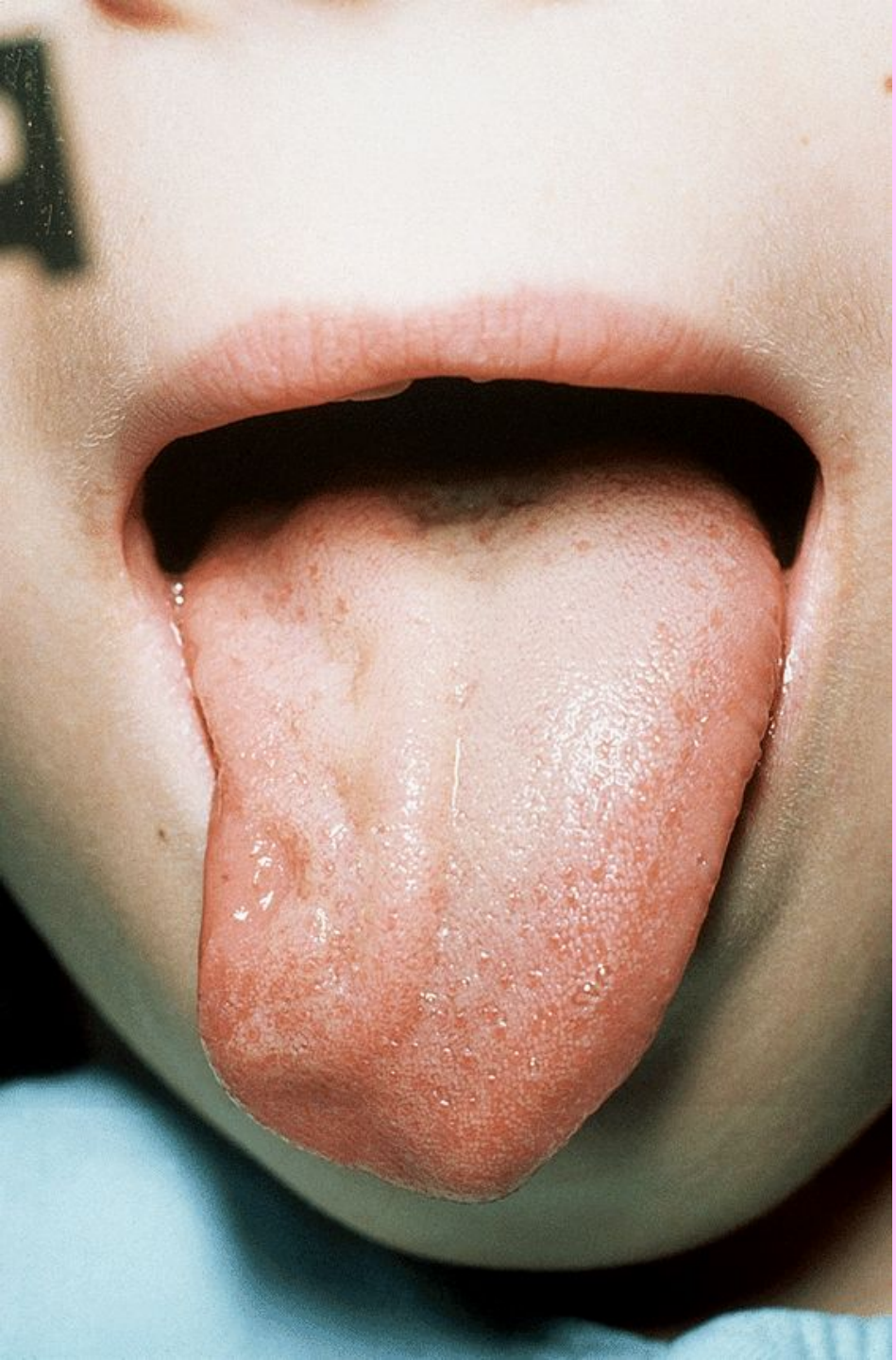
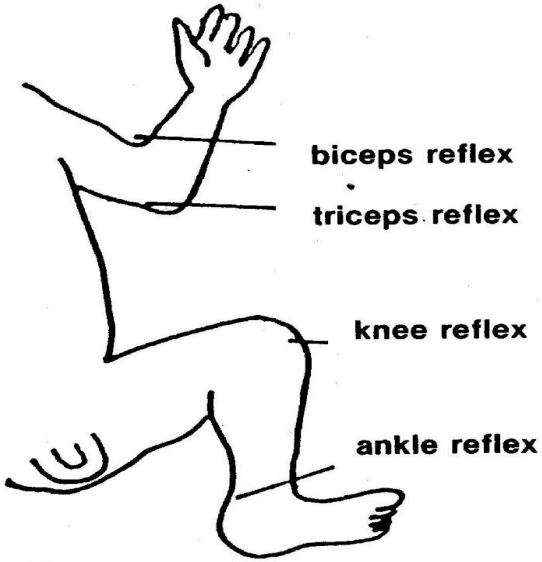


Unilateral (right-side) hypoglossal (12th) nerve paresis. Tongue deviation.

Motor examination

Motor Examination

- Posture or gait
- Movement disorders
- Muscles :
  - Muscle power
  - Muscle tone
  - Muscle bulk
- Reflexes :



- Tendon reflexes

- + ve Babiniski

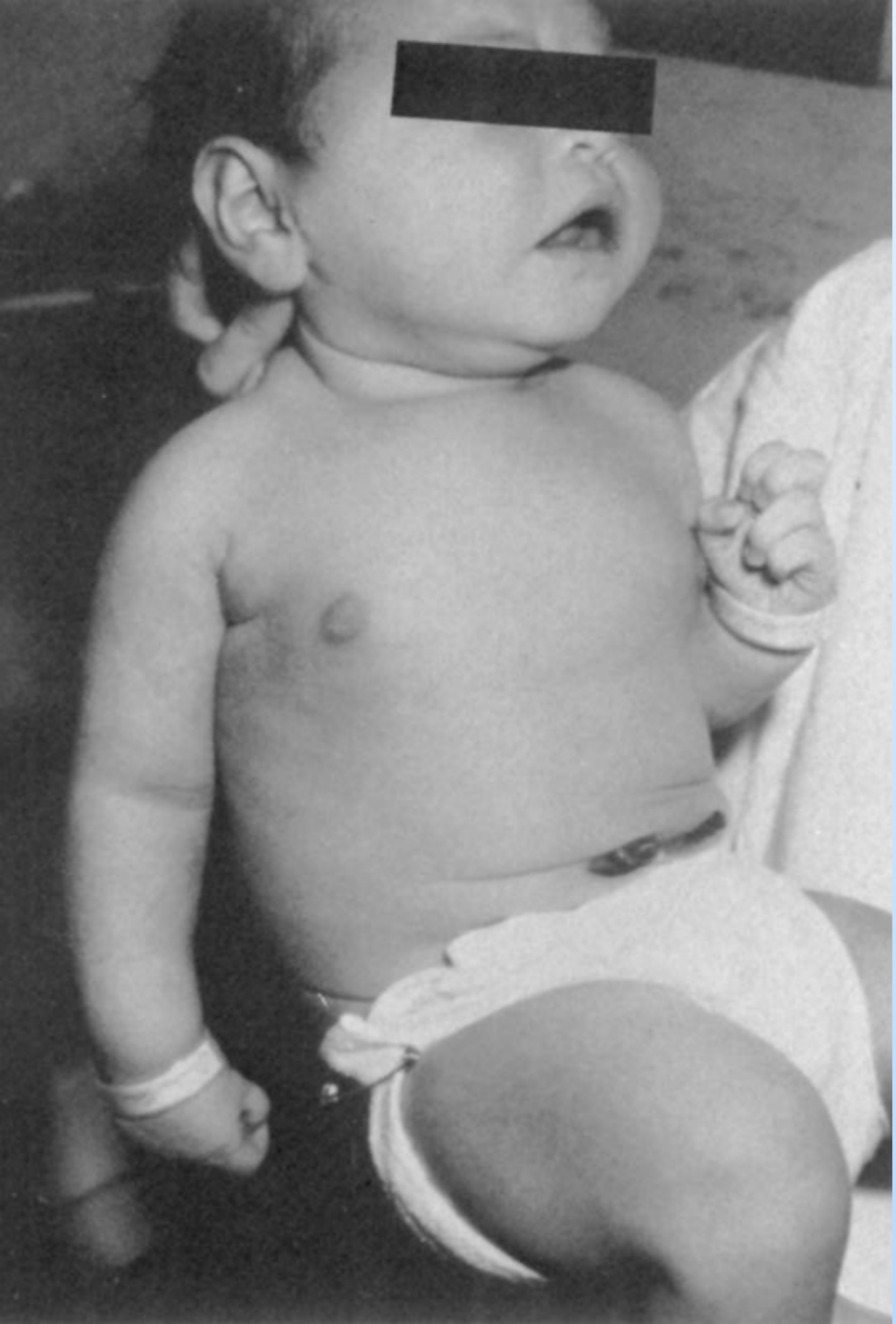
- Ankle clonus

Abnormal gaits

- *The spastic gait*
- *Circumduction gait*
- *Cerebellar ataxia*
- *waddling gait*
- *clumsy, tentative gait*

Movement disorders

- ***Paralysis (palsy)*** – the absence *of any* voluntary movements
- ***Paresis*** is incomplete paralysis
- ***Ataxia*** - gross uncoordination that may become worse with the eyes closed
- ***Athetosis*** - slow, writhing, wormlike, constant, grossly uncoordinated movements that increase on voluntary activity and decrease on relaxation



A newborn with **brachial right sided paralysis (palsy).** The arm hangs limp alongside the body and internally rotated, and the wrist is pronated hand (hangs limp downwards).

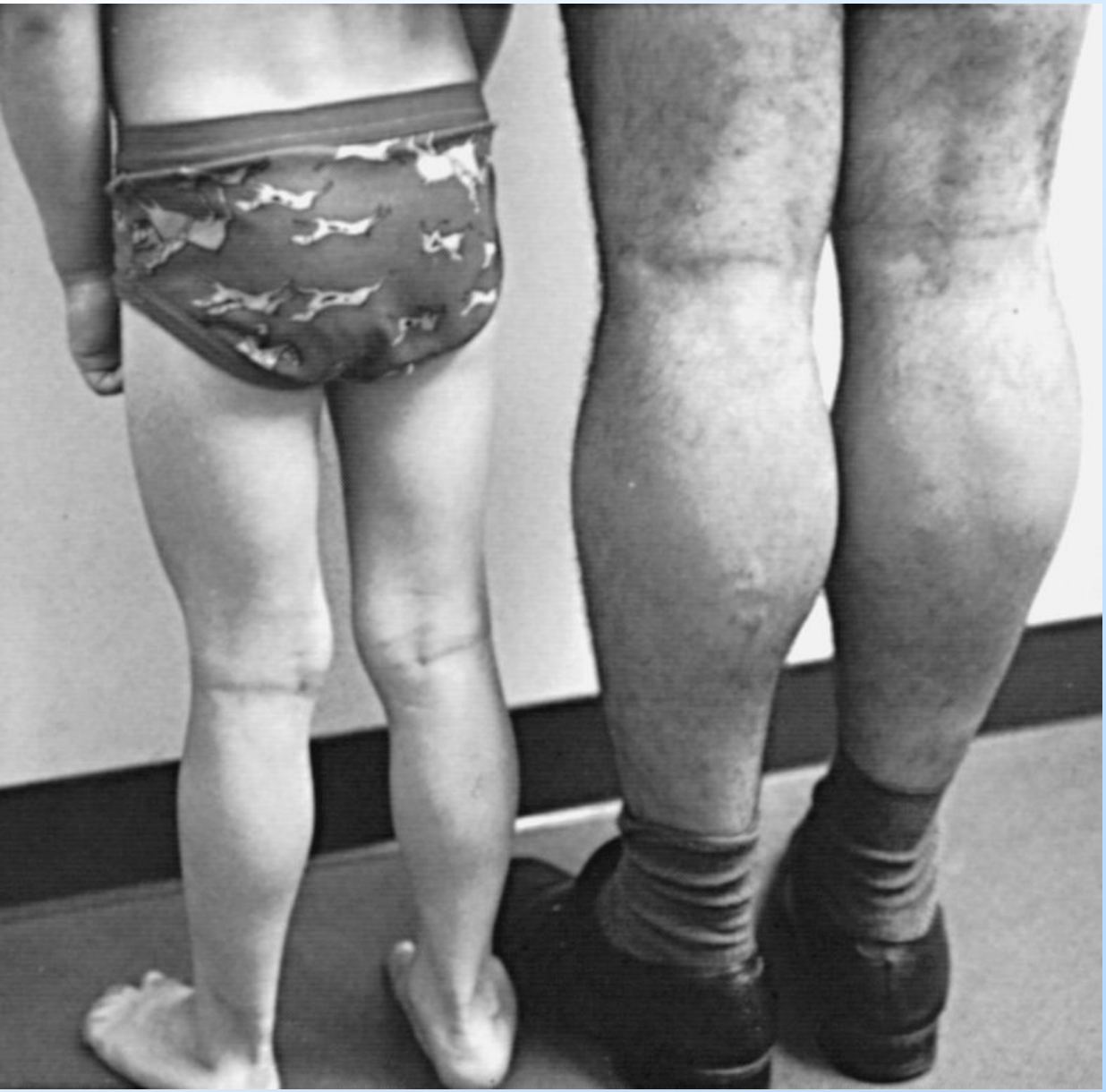
Movement disorders

- ***Dystonia*** - slow twisting movements of limbs or trunk (alternation of a hypotonia with rigidity, formation of elaborate postures)
- ***Tics*** - involuntary, compulsive, stereotyped movements of an associated group of muscles (can be suppressed by strong-willed effort).
- ***Tremors*** - constant small very fast involuntary movements.

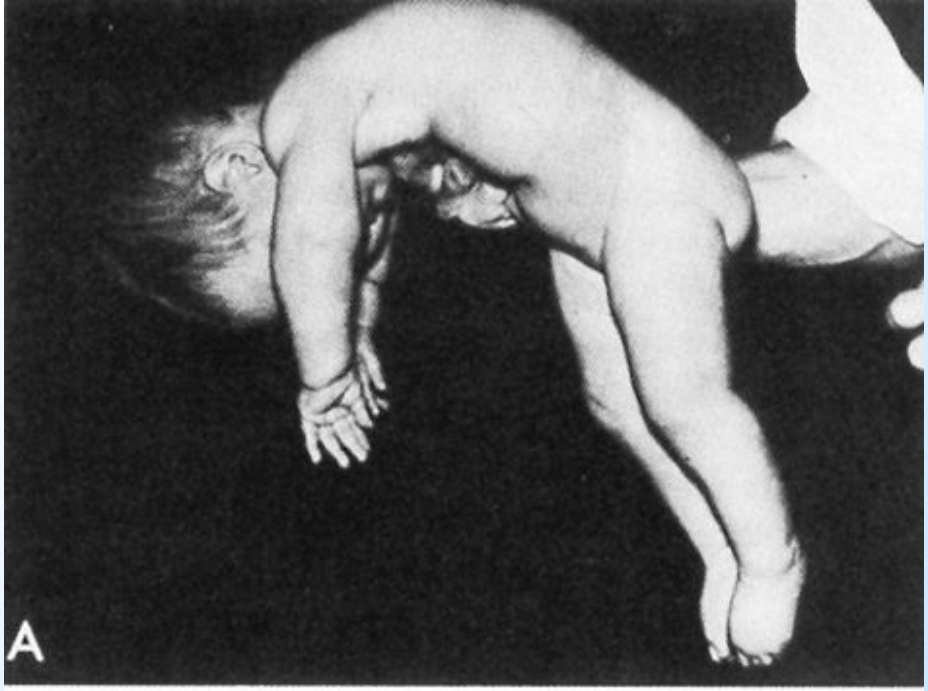
Muscles

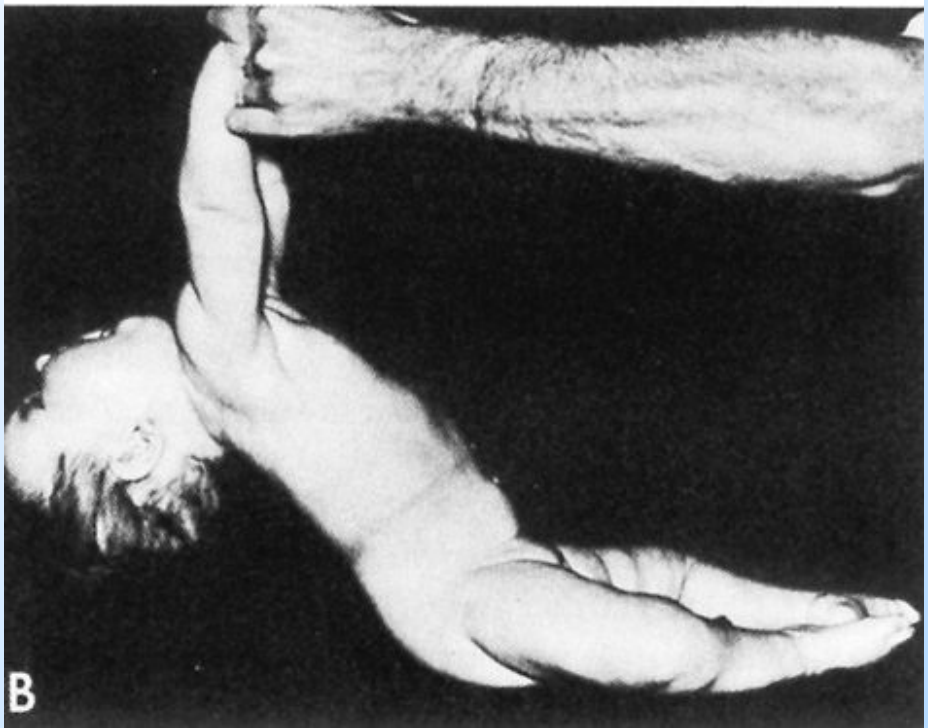
Examination includes assessment of

- *muscles' development*: wasting, pseudohypertrophy
- *Tone*: hypotonia, hypertonia
- *Strength*: increase, decrease



Posterior aspect of the legs of a father and his 6-year-old son with a rare autosomal dominant muscular dystrophy. **Hypertrophy of the calves** resembles **Duchenne muscular dystrophy**





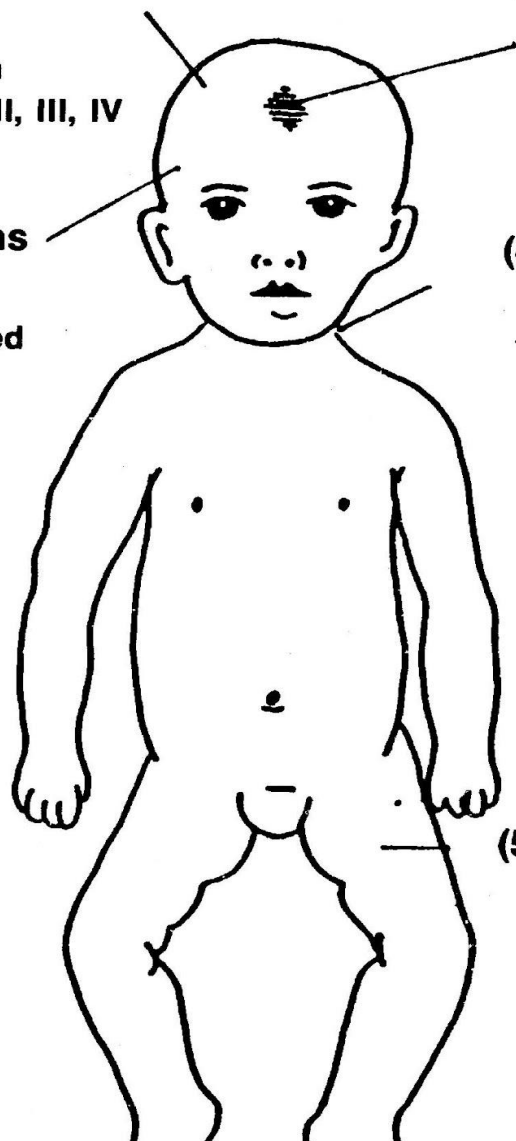
Hypotonia

On ventral suspension, the baby assumes the position of a rag doll.

When pulled up from the supine to the sitting position, the head of the baby lags.

Main semiotics of CNS disorders. Meningitis.

Meningitis



(1) altered consciousness

- lethargy
- confusion
- coma : I, II, III, IV

(2) convulsions

- focal
- generalized

- fever
- vomiting
- constipation

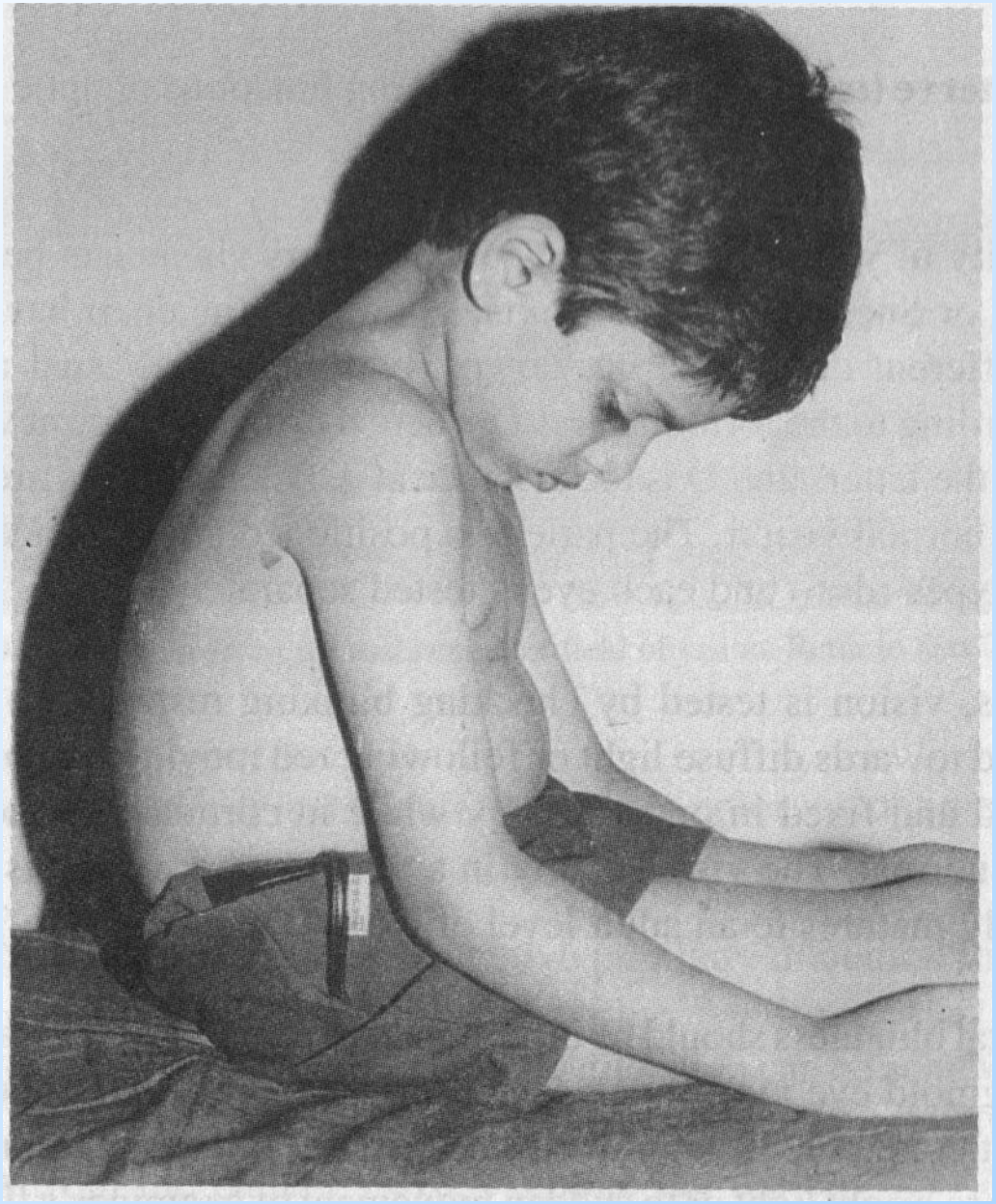
(3) ↑ I.C.P.

bulging fontanel

(4) meningeal irritation

- neck rigidity
- + ve Kernig
- + ve Brudzinski

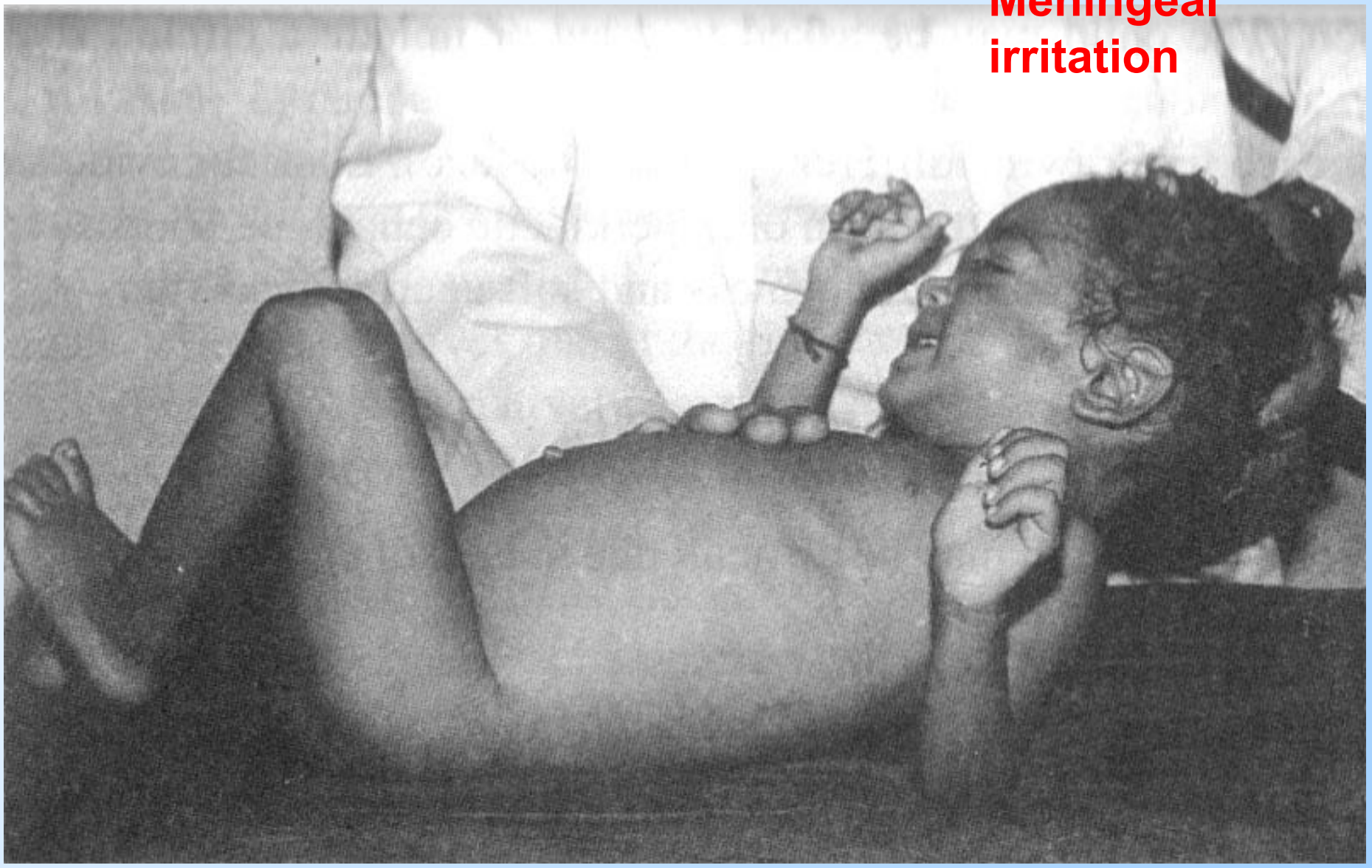
(5) focal paralysis



Meningeal irritation

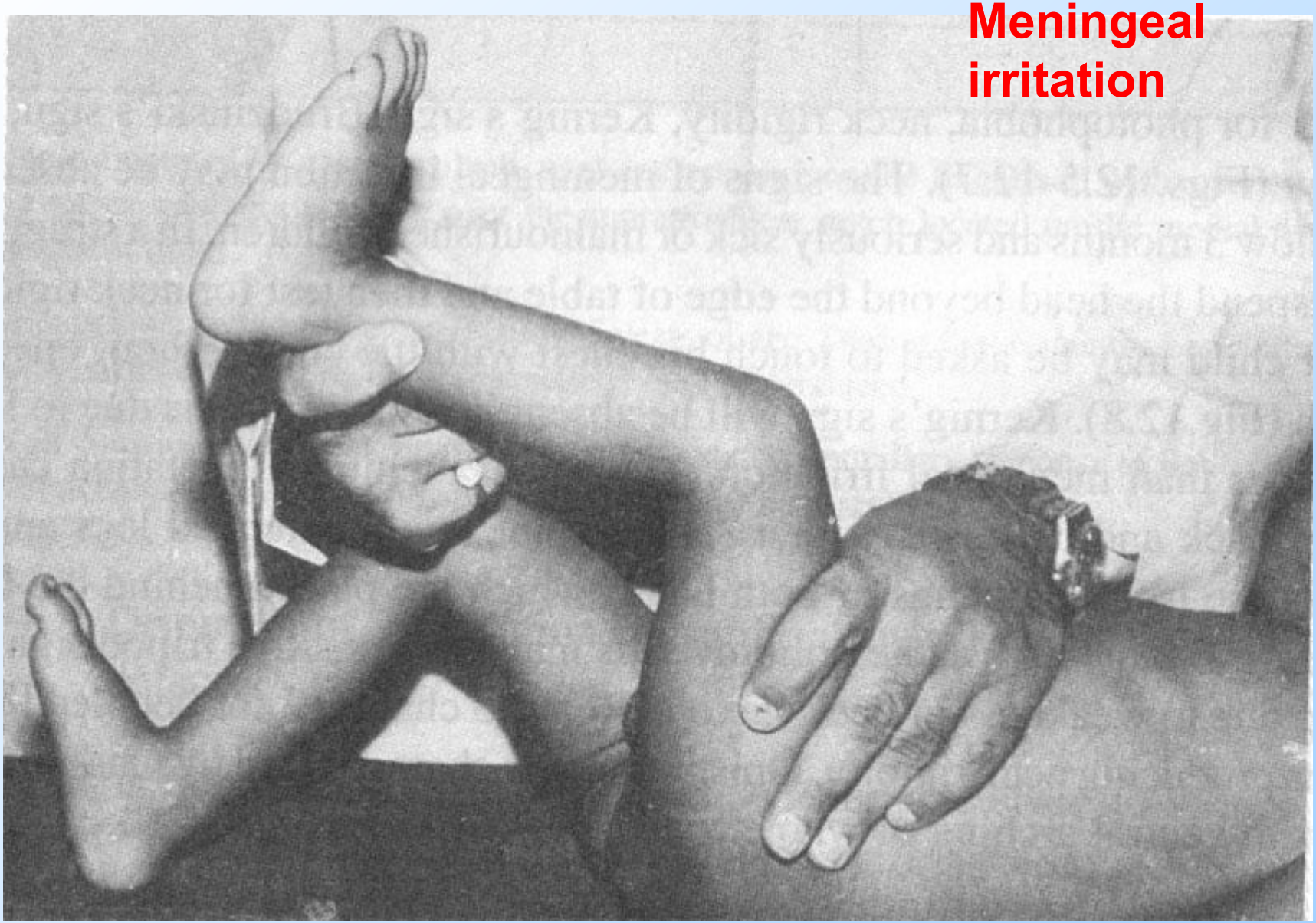
Examination for neck rigidity in older child

Meningeal irritation



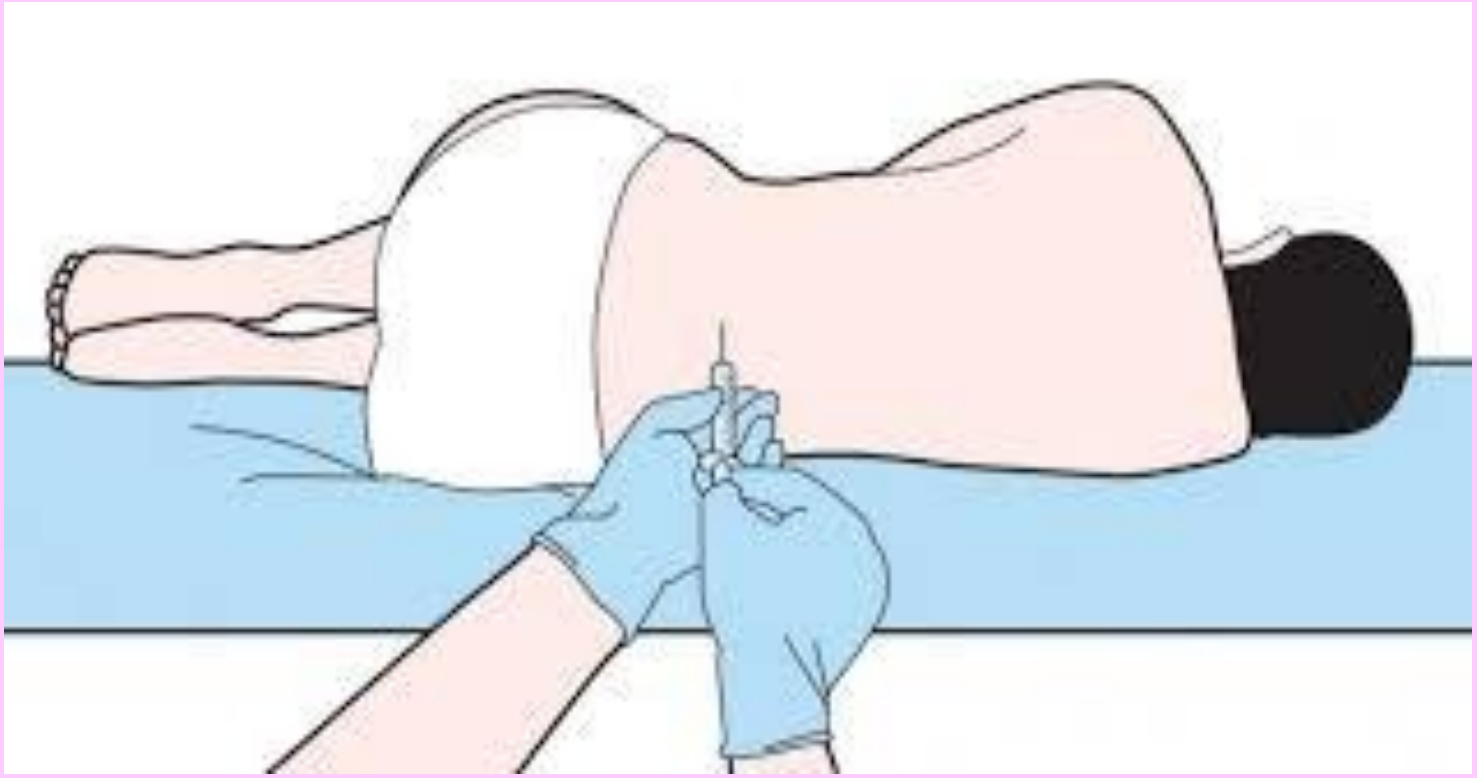
Brudzinski's sign

Meningeal irritation



Kernig’s sign

LP



- **The lumbar punction confirms the meningitis**

Normal Values for Cerebrospinal Fluid (CSF)

	Neonate	Infant/child
Pressure	50-60 mm H2O	50 -150 mm H2O
Cytosis (cells)	Below ≤ 20-30/ mkl	Below ≤ 10 /mkl
Cell type	neutrophils ≤40% Lymphocytes 60 %	- Lymphocytes 90 -100 %
Protein	0.25 – 0.5 g/l	0.16 – 0.25 g/l
Glucose] (% of the Serum Glucose)	80 -100 %	≥ 50 -60 %
Color	Xanthochromic	Water

CSF finding in bacterial meningitis

- **ICP** - increased
- **White blood cell count, µL** - 100 – 10000
- **Cell type** - neutrophiles 100%
- **Protein content** - ≥ 40 mg/dl (0.4 g/l)
- **Glucose** - ≤ 40 mg/dl (≤ 50% blood glucose)
- **Culture** - positive

Meningizm

- If the analysis of a *cerebrospinal fluid* finds inflammatory changes, the child *has* meningitis even having negative or doubtful clinical symptoms.
- If there are meningeal irritation symptoms, but no inflammatory changes in a *cerebrospinal fluid, there is no* meningitis. Such condition is called meningizm, it means **non inflammatory irritation of meninges** in various diseases in children.