The Nervous System
Introduction

The nervous system is divided into 2 components:

- ***<u>The central nervous system (CNS):</u>*** (the parts of the nervous system protected by bone; the skull & the vertebral column) consists of **the intracranial part** and **the spinal part**.

  The intracranial part: formed of 2 cerebral hemispheres, the brain stem (midbrain, pones and the medulla) and the cerebellum.

  The cerebral hemisphere is divided by sulci into four main lobes: the frontal, parietal, temporal and the occipital lobes. The 2 cerebral hemispheres are connected to each other by the corpus callosum and to the upper part of the brain stem by the two cerebral peduncles.

  The spinal part: is formed of the spinal cord (which ends at the lower border of the first lumbar vertebra) and the cauda equina.

- ***<u>The peripheral nervous system (PNS):</u>*** The anterior horn cell, the peripheral nerve, the motor end plate, and the muscles (or glands).

The cavities of the CNS (the ventricles): there are 4 ventricles in the brain; two lateral ventricles, a third ventricle and a fourth ventricle

Congenital Anomalies of the Central Nervous System

- ***Cranial Defects:***

Anencephaly or Encephalocele: Neonates with anencephaly have a rudimentary brainstem or midbrain, but no cortex or cranium. This is a rapidly fatal condition. Patients with encephalocele usually have a skull defect and exposure of meninges alone or meninges and brain.

- ***Congenital Malformations of the Brain:***

- **Macrocephaly:** Macrocephaly represents a head circumference above the 97th percentile and may be the result of ***macrocrania*** (increased skull thickness), ***hydrocephalus*** (enlargement of the ventricles), or ***megalencephaly*** (enlargement of the brain).
- **Megalencephaly:** (or large brain) may be the result of an embryologic disorder
- **Microcephaly:** represents a head circumference below the 3rd percentile.
- **Hydrocephalus** represents enlargement of the ventricular system.

- ***Congenital Anomalies of The Spinal Cord:***

- **Spina bifida:** Defective closure of the caudal neural tube at the end of week 4 of gestation results in anomalies of the lumbar and sacral vertebrae or spinal cord. In **spina bifida occulta**, the skin of the back is apparently intact, but defects of the underlying bone or spinal canal are present. These defects include *tethering of the cord to a thick filum terminale, a lipoma or dermoid cyst*, or *a tiny epithelial tract extending from the skin surface to the meninges*. *A small dimple or tuft of hair* may be present over the affected vertebra.

  Patients with spina bifida occulta may have difficulties controlling bowel or bladder, weakness and numbness in the feet. Bladder dysfunction may result in repeated episodes of urinary tract infections, reflux nephropathy, and renal insufficiency.
- **Meningomyelocele:** These anomalies range in severity from clinically insignificant defects of the L5 or S1 vertebral arches to major malformations of the spinal cord that lies uncovered by

skin or bone on the infant's back. The latter severe defect, results in total paralysis and loss of sensation in the legs and incontinence of bowel and bladder

Macrocephaly (Large Head)

- Head circumference is above the 97th percentile for age & sex.

Hydrocephalus

Definition: Dilatation of the ventricular system due to either **obstruction** of CSF flow within the ventricular system or interference with it's **absorption** in the subarachnoid space.

Types & Etiology:

1- Obstructive hydrocephalus:

- *Congenital:*
  - Congenital absence of foramina of Monro.
  - Aqueduct of Sylvius's stenosis (toxoplasmosis) – malformation of vein of Galen.
  - Foramen of Lushka and Magendi membrane at the outlet of the 4th ventricle) → cystic expansion of the 4th ventricle that compress the cerebellum.
- *Traumatic*: Intra-ventricular hemorrhage.
- *Inflammatory*:
  - Post meningitic gliosis.
  - Viral infection (Mumps).
- *Neoplastic*: Brain tumor (posterior fossa tumors).

2- Non obstruction (communicating) hydrocephalus:-

- **Impaired absorption** of CSF:

- *Congenital:* Arnold chiari malformation: obstruction of subarachoid pathway around the brain stem by downward displacement of medulla and cerebellum usually with meningomyelocele.
- *Traumatic:* subarachnoid hemorrhage.
- *Inflammatory:* post meningitic gliosis.
  - **Overproduction** of CSF: Choroid plexus adenoma.

Clinical picture:-

1- Before closure of sutures and fontanels (infancy):

- *General Examination:*
  - Progressive increase in all skull diameters.
  - Scalp skin: thin and shiny.
  - Scalp veins: prominent.
  - Anterior fontanel: widely opened, tense or bulging.
  - Sutures: widely separated
  - Macewen (cracked pot sign): resonant note on percussion due to suture separation.
  - Craniotabes.
  - Face: Globular and prominent forehead.
  - Eye: Sunset appearance with a rim of sclera above the iris - forward & downward displacement of the globes.
- *Back Examination*: Meningocele or meningomyelocele (Arnold- Chiari syndrome).
- *Neurological*:
  - Motor system: spasticity of limbs due to compression on motor area or long tract.
  - Power: diminished.
  - Reflexes: exaggerated.
  - Cranial nerves: Optic atrophy in chronic cases + 6th nerve palsy +/- Convulsions.

2- After closure of sutures (older children):

- Head enlargement is less marked due to closure of anterior fontanel.
- Neurological:
  Signs of increased ICT are marked - Variable neurological deficits (long tract).

Clinical Assessments:

- Serial measurement of head circumference.
- Fundus examination.

Laboratory:

CSF analysis is to rule out congenital infections as a cause.

Imaging:

X-ray skull: **in infancy** (before closure of suture):

- Fontanel: Craniofacial disproportion.
- Wide separation of sutures.

Older children (after closure of sutures): silver beaten appearance.

Cranial U/S (before closure of the fontanels).

CT scan the most important:

- Degree of ventricular dilatation - Site of obstruction - Exclude brain tumors.

Management: **-** In obstructive hydrocephalus a shunt operation is performed (ventriculojugular or ventriculoperitoneal).

Microcephaly (Small Head)

Definition:-

It is a small head with skull circumference less than the 3rd percentile.

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Seizures (Convulsions) in Childhood

Definition:

- A seizure is the clinical event that result from abnormal excessive neuronal activity & characterized by any of the following:
- Alteration of consciousness, motor activity, behavior, sensation or autonomic function.
- It may be viewed as a symptom of an underlying disease process.

- Seizures are common in the pediatric age group and occur in ≈10% of children.
- Less than one third of seizures in children are caused by **epilepsy**.
- Epilepsy is considered to be present when two or more unprovoked seizures occur at an interval greater than 24 hr apart.

Etiology:

- **Acute non recurrent convulsions**:-
  One or more convulsive fits that occur during the same acute illness & don't recur after recovery:
  - Febrile convulsions. - CNS infections: meningitis, encephalitis.
  - ICH: spontaneous, traumatic. - Toxic: e.g. tetanus. - Anoxic: sudden severe asphyxia.
  - Metabolic: hypoglycemia, hypocalcemia, hypo or hypernatremia, hypomagnesemia.
- **Chronic recurrent convulsions:-**
  Recurrent attacks of convulsions with symptoms free intervals:
  - Epilepsy: idiopathic.
  - Organic secondary to brain insult: post infection, post anoxic, post traumatic, post hypoxic, post toxic.
  - Degenerative brain disease.
  - Congenital cerebral malformation.

Generalized Tonic-Clonic (Grand Mal) Epilepsy

The commonest **form** of childhood convulsions 60%:

- **An aura:** unusual behavior recognized by the mother.
- **Tonic phase:** powerful sustained contraction (5 minutes)
  - The patient falls to the ground stiff due to powerful sustained contraction of all muscles.
  - Arm flexed - Legs extended
- **Clonic phase:** Rhythmical contraction and relaxation of muscles of limbs and face. Biting the tongue and incontinence may occur during the clonic phase. Duration of the attack is variable but if exceed 20 minutes it considered status epilepticus.
- **Post epileptic phase**: The child falls in deep sleep and afterwards he may be confused or irritable.

Grand mal epilepsy has good prognosis if the first attack starts after the age of 3 years and the mental development is normal.

Myoclonic Epilepsy

- Occurs at any age but is more seen in infants and young children.
- Usually associated with mental retardation.
- The attack which is very frequent, present with sudden symmetrical mass jerking involving all limbs.

Febrile Seizures

- Febrile convulsions, the most common **seizure disorder** during childhood, generally have an excellent prognosis but may also signify a serious underlying acute infectious disease such as sepsis or bacterial meningitis. Therefore, each child with a seizure associated with fever must be carefully examined and appropriately investigated for the cause of the fever, especially when it is the 1st seizure.
- Febrile seizures are age dependent and are rare before 9 mo and after 5 yr of age. The peak age of onset is ≈14–18 mo of age.
- The incidence approaches 3–4% of young children.

Coma

Coma is a medical emergency; prompt diagnosis and treatment may be life saving.

Immediate examination

- Air way - Cardio respiratory system - Record vital signs

If shocked or cardio respiratory arrest resuscitation as follow:

Airway: lie flat, clear air way, mouth to mouth, mask and bag.

Breathing: oxygen, endo-tracheal intubation.

Circulation: External cardiac massage + cardioversion.

I.V infusion: Ringer's lactate + $NaHCO_3$

Drugs

ECG monitor

Causes of coma:

1- Head injury.
2- Vascular accidents.
3- Hypoxic ischemic encephalopathy (HIE), Cardio respiratory arrest.
4- Meningitis – Encephalitis.
5- Status epilepticus
6- Increased intracranial pressure.

7- Hypertensive encephalopathy.
8- Drug poisoning.
9- Metabolic: - Hypoglycemia - Uremia - Diabetic Ketoacidosis (DKA)
- Hepatic coma - Water intoxication

Diagnosis:

History: Trauma, chronic illness, drug or household product ingestion.
Gradual change in personality or behavior.
Diabetes Mellitus: Type, amount, and time of last dose of insulin.
Headache – Vomiting – Change in personality (brain tumor).
Chronic heart disease (brain abscess).

In newborn: with vomiting, convulsion, failure to thrive (Inborn errors of metabolism)

In toddler: review of medications, their location at home to diagnose intoxication or finding pills or empty medication containers.

Examination:

1- Mouth odor: Acetone: (DKA, Kerosene, Alcohol).
Foetor hepaticus: (Hepatic Failure).
Garlic: (Arsenic).
2- A- Pupil constricted: Pontine hemorrhage, organophosphorous poisoning, morphia intoxication.
B- Pupil dilated: Atropine, Barbiturate, and Cyanide.
3- Signs of meningeal irritation (complete neurological examination).
4- Skin examination: Rash : Encephalitis.
Purpura: Intracranial hemorrhage, meningococcemia.
Flushed: Atropine, Cyanide intoxication.
5- Vital signs: - Bradycardia and hypertension (increased ICP).
- Slow respiration: Respiratory depression or sedation.
- Irregular respiration: Cerebellar herniation.

Investigations:

1- Blood Glucose – Serum electrolyte 2- CT – M R I (brain) 3- CSF exam.
4- EEG 5- Angiogra 6- Blood and urine analysis for toxic substances
7- Renal and hepatic functions. 8- Metabolic screen

Response	**Score**
Best Motor Response	
Nil (flaccid)	1
Extensor response	2
Abnormal flexion	3
Withdrawal	4
Localization of pain	5
Obeys commands	6
Best Verbal Response	
Nil	1
Incomprehensible sounds	2
Inappropriate words	3
Confused conversation	4
Oriented, fluent speech	5

Glasgow coma scale:

Best Eye Opening Response	
Nil	1
To pain	2
To speech	3
Spontaneous	4

- It is a clinical score to assess
the progression of the condition and its response to the management given.

- **T**he score is from 3 – 15. Normal is 15, less than 8 means coma.