NERVOUS SYSTEM

ENCEPHALON

A. PROSENCEPHALON (Anterior Vesicle)

Telencephalon

- Hemispheres
- Lateral part of Corpus callosum
- Corpus striatum
- Columna forcinis
- Basal ganglions
- Rhinencephalon
- Lateral ventricules

Diencephalon

- Thalamus
- Corpus pineale
- Tegmen ventriculi tertii
- Hypophysis
- Corpus mamillare
- Tuber cinereum
- Infundibulum
- Chiasma opticum
- Tractus opticus

B. MESENCEPHALON (Middle Vesicle)

- Crus cerebri
- Tectum mesencephali
- Tegmentum mesencephali
- Substantia nigra
- Cores of the 3rd and 4th cranial nerves
- Aquaductus mesencephali

C. RHOMBENCEPHALON (Anterior Vesicle)

Isthmus Rhombencephali

- Vellum medullare rostrale
- Crura cerebelli rostralia

Metencephalon

- Pons
- Cerebellum
- 5th cranial nerve

Myelencephalon

- Medulla oblongata
- Brachia cerebelli caudalia
- Tegmen fossa rhomboidea
- Ventriculus quartus
- 6th, 7th, 8th, 9th, 11th, 12th cranial nerves

MALFORMATIONS OF THE CENTRAL NERVOUS SYSTEM

- **DUPLICATION OF PROSENCEPHALON**
- **MICROENCEPHALY** (an abnormally small brain/The diminution affects particularly the cerebrum).
- **CORTICAL DYSPLASIA AND GYRUS MALFORMATIONS**
  - ✓**Neuronal Heterotopia** (The presence of clusters of nerve cells at a site where they are normally absent, such as subcortical white matter).
  - ✓**Microgyria** (**polymicrogyria**/ convolutions are small and unusually numerous, and the normal gyral pattern is lost in affected areas).
  - ✓**Ulegyria** (also a wrinkled appearance of the cortex but arises as a consequence of scarring and atrophy in otherwise topographically normal gyri/ It is a result of laminar necrosis of the deep sulcal cortex).

✓**Lissencephaly**
(**agyria**/ convolutions are almost entirely absent)

✓**Pachygyria**
(**macrogyria**/ excessively broad brain convolutions)

- **AGENESIS OF THE CORPUS CALLOSUM**

(Partial or complete absence (agenesis) of an area of the brain that connects the two cerebral hemispheres).

- **CYSTIC SEPTUM PELLUCIDUM**

(A cystic fluidfilled cavity between the lateral ventricles that varies from a thin slit to a rounder cavity).

- **HOLOPROSENCEPHALIA**

(Failure of the forebrain to separate normally into 2 discrete cerebral hemispheres)

- **CYCLOPIA**

✓ Single large median eye

✓ Failure of division of the optic primordium into paired symmetric optic stalks and vesicles, which therefore results in a single midline globe

✓ Ewes ingesting the plant "Veratrum californicum" on day 14 of gestation give birth to lambs with cyclopia

- **CEBOCEPHALY (monkey face)**

✓ Anatomically comparable to cyclopia

✓ There are 2 eyes, severely hypoplastic, in separate but approximated orbits.

- **DYSRAPHIC MALFORMATIONS**
  - ✓ **ANENCEPHALY** (Absence of brain)
    - **Acrania** (Complete failure of cranial development)
    - **Cranioschisis** (The cranium fails to close completely)
    - **Craniorhachischisis** (Both the brain and spinal cord fails to close; both anencephaly and spina bifida are present)

Cerebral aplasia (“anencephaly”) in a foal. The cerebral hemispheres have failed to form, and only brainstem and cerebellum are present. (Courtesy Noah’s Arkives, University of Georgia.)

- **DYSRAPHIC MALFORMATIONS**
  - ✓ **CRANIUM BIFIDUM AND RELATED DEFECTS**
    - **Encephalocele** (Protrusion of the brain through a defect in the cranium (**cranium bifidum**))
    - **Meningocele** (only fluid-filled meninges protrude)
    - **Meningoencephalocele** (meningeal and brain tissue protrusion).

SPINA BIFIDA AND RELATED DEFECTS

- **Spina Bifida** (The vertebral counterpart of cranium bifidum. Frequently tends to affect the caudal spine and is characterized by a dorsal defect in the closure of one to several vertebral arches)
- **Total Myeloschisis** (Defect that involves the whole of the vertebral axis)
- **Amyelia** (Absence of spinal cord)
- **Local Myeloschisis** (Localized defect caused by failure of closure of the neural tube)

Spina Bifida Occulta

(No herniation of the meninges or spinal cord through the defect)

Spina Bifida Cystica

- ✓ **Meningocele** (Herniation of meninges)
- ✓ **Myelocele** (Herniation of spinal cord)
- ✓ **Meningomyelocele**

(Herniation of meninges and spinal cord)

MYELODYSPLASIA (Abnormal development of the spinal cord)

- **Diplomyelia** (the cord is duplicated completely within common leptomeninges and dura)
- **Diastomatomyelia** (the cord is duplicated within separate meningeal coverings and vertebral canals separated by a bony partition)
- **Syringomyelia** (tubular cavitation of the spinal cord that extends over several segments)
- **Syringobulbia** (When the cavitation involves the medulla)
- **Hydromyelia** (Dilation of the central canal of the spinal cord)

ARNOLD-CHIARI MALFORMATION

(Chiari type II malformation),

- ✓ **The cerebellum is herniated** into the foramen magnum and cranial spinal canal
- ✓ **Internal hydrocephalus** may be a secondary effect, and **spina bifida or meningomyelocele** may be concurrent.

DANDY-WALKER SYNDROME

- Midline defect of the cerebellum
- The cerebellar vermis largely absent
- The cerebellar hemispheres widely separated by a large fluid-filled cyst (reaches the 4th ventricle) in an enlarged caudal fossa.

SEGMENTAL APLASIA AND HYPOPLASIA OF MEDULLA SPINALIS

- **Perosomus elumbus** (partial agenesis of the spinal cord)
- **Sacrococcygeal agenesia**

HYDROCEPHALUS

- Excessive accumulation of fluid in the cranial cavity
  - ✓ **Internal hydocephalus** (the fluid is within the ventricular system)
  - ✓ **External hydrocephalus** (the fluid is in the arachnoid space)
  - ✓ **Communicating hydrocephalus** (the excess fluid is present in both locations)

- **Cerebrospinal fluid (CSF)** is produced by the ventricular choroid plexuses by means of filtration and secretion.

- The flow of CSF is from the **lateral ventricle** through the **interventricular foramen** to the **third ventricle** and then via the **cerebral aqueduct** to the **fourth ventricle.**

- From here, most of the fluid leaves the ventricular system and passes by way of the **lateral aperture of fourth ventricle** into the **subarachnoid space**.

- A small amount of CSF passes into the **central canal of the spinal cord** from the **fourth ventricle.**

- A disturbance of cerebrospinal fluid (CSF) **formation, flow, or absorption,** leads to excessive accumulation of CSF within the cerebral ventricles and/or subarachnoid spaces, resulting in ventricular dilation and increased intracranial pressure.

Depending on the aetiology, hydrocephalus is classified into congenital and acquired forms;

✓Congenital Hydrocephalus

- Most common in toy and brachycephalic breed dogs, calves and foals
- The causes include genetic factors, developmental anomalies, intrauterine or prenatal infections, dietary deficiency of vitamin A,..
- If hydrocephalus develops in infancy before closure of the cranial sutures, the head enlarges.

✓Acquired Hydrocephalus

- The causes are almost always obstructive;
- Causes of obstruction include compression by cerebral abscesses and neoplasms, and blockages by infectious/inflammatory disease resulting in a ventriculitis and, uncommonly, by cholesteatomas in the choroid plexus of the lateral ventricles of the horse.

✓Physiologic Hydrocephalus

(Hydrocephalus is “physiologic” in the early fetus when the hemispheres are largely thin-walled vesicles)

HYDRANENCEPHALY and PORENCEPHALY

The formation of fluid filled cavities in the brain, termed **porencephaly** (small cavities) and **hydranencephaly** (large cavities), usually occurs in utero during gestation.

HYDRANENCEPHALY

- Complete or almost complete absence of the cerebral hemispheres, leaving only membranous sacs filled with CSF and enclosed by leptomeninges.
- The cavitation results from **destruction of immature neuroblasts** whose loss prevents normal development as a result of faulty or aberrant neuroblast migration.
- The cranial cavity is always complete, in contrast to hydrocephalus
- Hydranencephaly occurs in all species but is more common in calves and lambs.
- Viral causes of hydranencephaly: **Akabane virus, Bluetongue virus, Bovine viral diarrhea virus, Border disease virus,** etc….

CEREBELLAR DEFECTS

- ✓ **Cerebellar Hypoplasia** (the size of the cerebellum is reduced)
- ✓ **Cerebellar Abiotrophia** (premature or accelerated degeneration of fully formed cerebellar neurons)
- ✓ **Hereditary Striatonigral & Cerebelo-Oliver Degeneration**
- ✓ **Hereditary Convulsion and Ataxia of cattle**
- ✓ **Gomen Disease**

INTRAUTERINE VIRAL CAUSES OF DEVELOPMENTAL DEFECTS OF THE CENTRAL NERVOUS SYSTEM

- AKABANE VIRUS INFECTION
- CHUZAN VIRUS INFECTION
- CACHE VALLEY VIRUS INFECTION
- BLUE TONGUE VIRUS INFECTION
- RIFT VALLEY FEVER AND VESSEL BRON DISEASES
- BOVINE VIRAL DIARRHEA
- BORDER DISEASE
- HOG CHOLERA
- FELINE PARVOVIRUS INFECTION

LYSOSOMAL STORAGE DISEASES OF THE NERVOUS SYSTEM

- A group of hereditary disorders characterized by accumulation of lipid metabolites or mucopolysaccharides in the the cells (Lipidosis and saccharidiosis).
- Results from dysfunction of lysosome-mediated degradation of products (substrates) of normal cellular metabolism. These substrates cannot be degraded by lysosomes, and the accumulated substrate eventually results in death of the affected cells.
- Lysosomes are present within all cells, except erythrocytes.

- Lysosomal acid hydrolases are capable of digesting completely the complex macromolecules synthesized for cell membranes, organelles, secretory products, and so forth.

Proteins → Amino acids

Polysaccharides → Monosaccharides

Glycolipids → Simple lipids

Lysosomal storage diseases develop because of:

- Overloading the cell above the normal enzymatic capacity
- One or more hydrolase deficiency or absence
- Inhibition of enzyme activity; especially in some plant poisoning
- Cellular uptake of unnatural substances that lysosomes are unable to degrade

- In **lipidosis** neurons are the most affected. This is due to the excess lipid content in the central nervous system.
- In **Mucopolysaccharidoses** also neurons are the most effected. Besides the neurons, it also accumulates in the liver, spleen, connective tissue and other organs.
- In these disorders, due to the collected material, neurons have a granular appearance and contain thin vacuoles.

CYTOPATHOLOGY OF NEURONS

- **Nuclear margination** (The neuronal nucleus is usually single and centrally located, and its margination can indicate nonspecific degeneration).
- **Chromatolysis** (change in appearance of the soma brought about by the dispersal of the rough endoplasmic reticulum (Nissl granules) and is subclassified as central or peripheral according to its locus within the cell body.)
  - ✓ **Central chromatolysis**
  - ✓ **Peripheral chromatolysis**
- **Neuronal atrophy** (Loss of cytoplasmic bulk and reduction in size)
- **Liquefactive necrosis**
- **Satellitosis** (gathering of phagocytes around the neuron) **and neuronophagia** (phagocytosis of dead neurons by macrophages)

CYTOPATHOLOGY OF NEURONS

- Ischemic necrosis
- Chronic neuronal injury
- Vacuolar degeneration
- Storage of pigments and other materials
- Siderotic pigmentation of neurons
- Viral inclusion bodies
- Nonviral eosinophilic cytoplasmic inclusion bodies
- Lafora bodies

WALLERIAN DEGENERATION

- Wallerian degeneration denotes the changes that follow acute focal injury to a myelinated axon.

❑First, Focal eosinophilic swellings occur, often containing accumulations of degenerate organelles, and then fragmentation happens.

❑The myelin itself condenses into aggregates and fragments and, together with remaining axonal debris, becomes the target of invading macrophages.

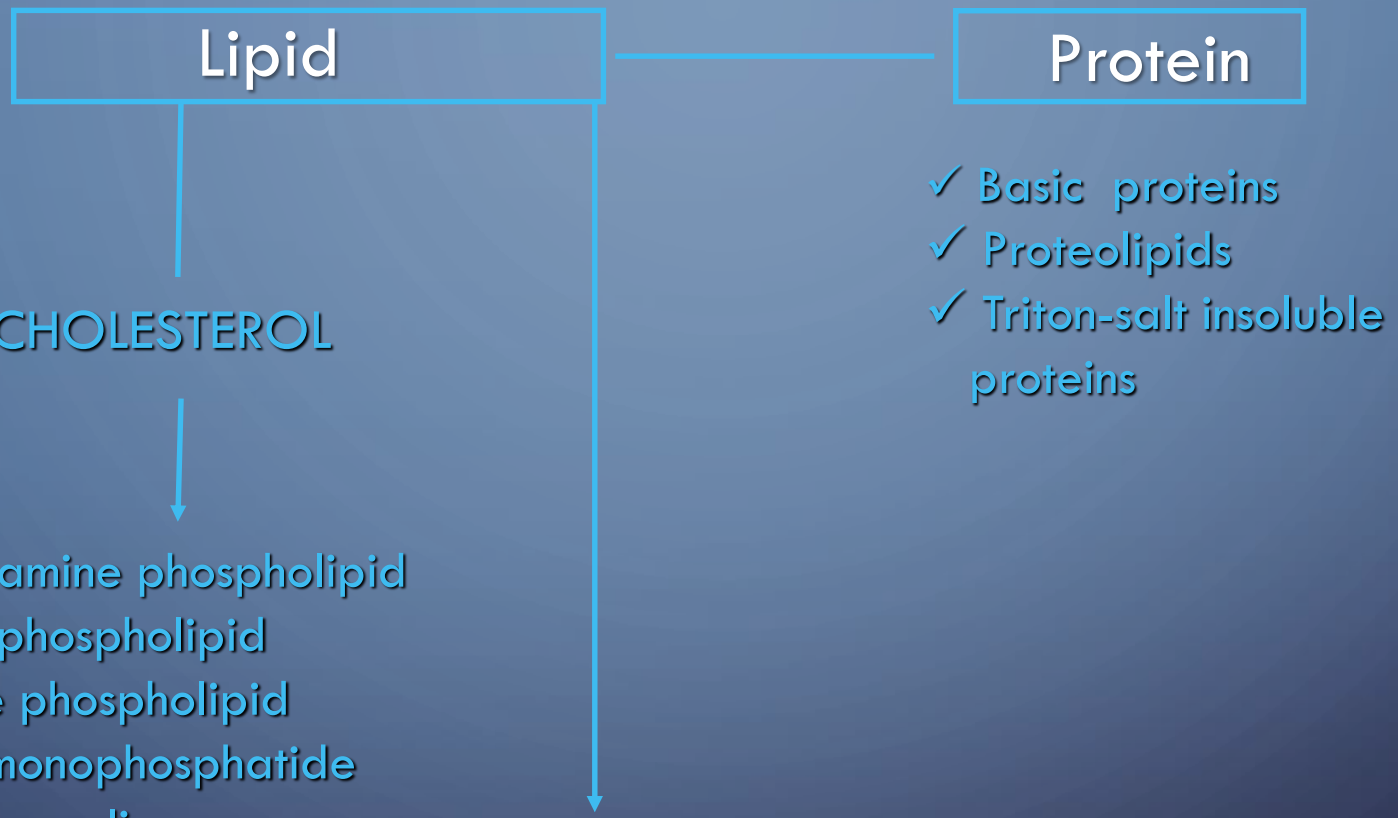
❑Some of the myelin debris is phagocytosed by Schwann cells themselves, and they begin to proliferate. As the debris is cleared away, proliferating Schwann cells form bands along the myelinated axons.

- Similar Wallerian changes occur at the opposite end of injured axon.

If conditions are favorable at the site of injury, sprouts from the axonal stump will reach to their correct destinations along the Schwann cell bands .

- Finally, a new axon arise and is remyelinated by Schwann cells.

CHEMICAL COMPOSITION OF MYELINE



MYELINOPATHIES

- **Hypomyelinogenesis** (a process in which there is underdevelopment of myelin)
- **Dysmyelination** (the formation of biochemically defective myelin)
- **Demyelination** (degeneration and loss of myelin already formed)

- **Intramyelinic edema (Status spongiosus)**

Spongy degeneration or Status spongiosus is a group of diseases of young animals characterized by a motheaten appearance (referred to here as status spongiosus) that primarily occurs in the white matter of the CNS but also extends into the gray matter.

NEUROGLIAL CELLS

Neuroectodermal

- Astrocyte
- Oligodendroglia
- Ependymal cells

Mesodermal

- Microglia

INCREASED INTRACRANIAL PRESSURE CEREBRAL SWELLING, AND EDEMA

- Brain edema is an increase in water content of brain tissue

- Edema may be **diffuse** or **localized**

  - ✓ **Diffuse edema** (acquired hydrocephalus and vitamin A deficiency in young animals can be responsible for diffuse brain edema)
  - ✓ **Local edema** (Lesions that may result in local edema of the brain or spinal cord include neoplasms, inflammations, parasitic cysts, focal necrosis of various causes, trauma, hemorrhages of parenchyma and meninges, and space-occupying lesions of the meninges that cause pressure on the brain)

- **Vasogenic edema, or tissue swelling:**
  - ✓ The most common type of edema in the central nervous system.
  - ✓ Common complication of traumatic, inflammatory, neoplastic, and haemorrhagic lesions of the nervous system.
  - ✓ Injury to vascular endothelium and leakage of plasma constituents into the perivascular extracellular space, particularly that of the white matter

- **Cytotoxic edema, or cell swelling:**
  - ✓ The accumulation of fluid intracellularly in neurons, astrocytes, oligodendroglia, and endothelial cells (called hydropic degeneration in other cells of the body) as a result of altered cellular metabolism, often caused by ischemia.

- **Interstitial (Hydrostatic) edema** is characterized by the accumulation of fluid in the extracellular space of the brain because of elevated ventricular hydrostatic pressure.

  Hydrocephalus → **Interstitial edema** that affects the central white matter

  Hydromyelia and syringomyelia → **Interstitial edema** that affects the spinal cord

LESIONS OF BLOOD VESSELS AND CIRCULATORY DISTURBANCES

The blood supply to the brain is derived from the **internal carotid** and **vertebral arteries**

Circulatory disturbances		
Ischemic lesions	**Hemorrhagic lesions**	**Microcirculatory lesions**
Arteriosclerotic changes	Spontaneous hemorrhages	Diapedesis of red cells
Hyaline necrosis	Isolated hemorrhages or hematomas	Leakage of plasma
Amyloid degeneration	Epidural/subdural meningeal hemorrhages	
Cerebrospinal vasculitis		
Thrombosis and embolism		
Result = arterial venous infarcts		

TRAUMATIC INJURIES

- **Concussion**

Concussion is a transient loss of consciousness and reflex activity following a sudden injury to the head.

Full recovery is expected, and it is assumed that in mild cases there is no morphologic injury.

More severe forms of diffuse brain injury can have generalized acute brain swelling caused by unregulated vasodilation.

- **Contusion**

Contusion means bruising; the architecture of the nervous tissue is retained, but there is hemorrhage into the meninges and about the blood vessels in the parenchyma.

Contusions may be diffuse or focal injuries, although often those coexist.

Same pathogenetic factors as those causing concussion, but of greater magnitude.

A **coup** contusion is located at the impact site, and a **contrecoup** contusion at a location on the opposite side of the brain.

- **Laceration**

Laceration is a traumatic injury in which there is disruption of the architecture of the tissue.

The mechanics of lacerations are, in general, the same as those of contusion.

Lacerations caused by penetrating injuries are always liable to secondary infections.

In the absence of infection, repair takes place in the manner that is usual for defects of nervous tissue.

- **Fracture of the skull**

Fractures of the skull can provide a pathway of infection to the sinuses, meninges, and brain.

Fractures of the base of the skull may involve the middle ear and allow the escape of cerebrospinal fluid (CSF) and the entrance of infection.

Frontal fractures involving the cribriform plate may allow CSF to escape into the nasal cavity.

Fractures of the skull are usually quite easy to detect by virtue of the displacement of bone and meningeal hemorrhage.

Injuries to the spinal cord

It is possible for direct injuries to the spinal cord to occur without obvious injury to the vertebrae.

Much more common are indirect injuries to the cord acquired in the course of vertebral luxations or fractures with dislocation.

The most common of indirect injuries to the cord are produced by extruded nucleus pulposus in dogs, and by compression of the cervical cord in the syndrome known as “wobbler”

Subluxations are largely restricted to the cervical column, where there is relative mobility of the ligaments.

In the thoracic and lumbar spine, comparable forces are more likely to cause fracture because of the brevity of the ligaments.

Traumatic injuries to the spinal cord may be slight enough to be recoverable, or they may, cause transection and an extensive length of necrosis.

DEGENERATION IN THE NERVOUS SYSTEM

- Meninges
  - Collagenous and osseous metaplasia
  - Spherical mineralized nodules (psammoma bodies)
  - Hyalinization of dural collagen
  - Ossifying pachymeningitis

- Choroid plexuses
  - Hyaline degeneration
  - Cholesteatosis, cholesteatomas (or *cholesterol granulomas,* occurs in 15-20% of old horses. Those in the lateral ventricules cause hydrocephalus by obstructing the interventricular foramen)

- Atrophy in the brain and spinal cord

- **Anoxia and anoxic poisons**

The pathologic effects of anoxia on nervous tissue are of 2 types;

- **Ischemic neuronal necrosis** followed by glial repair
- Greater degrees of anoxia, sufficient to kill astroglia as well as neurons, result in **softening**.

Histotoxic: respiratory enzyme blockage
Hypoxic: lack of oxygen in inhaled air
Anoxic: hemoglobin is not free
Ischemic: decreased blood volume (common)

- Anoxia and anoxic poisons

✓Cyanide poisoning

✓Nitrate/nitrite poisoning

✓Fluoroacetate poisoning

✓Carbon monoxide poisoning

✓Hypoglycemia

MALACIA AND MALACIC DISEASES

Malacia means grossly observable **softening**, and is used to signify necrosis of tissue in the Central nervous system.

Encephalomalacia: necrosis in the brain
Myelomalacia: necrosis in the cord
Encephalomyelomalacia: cerebral and spinal necrosis

Poliomalacia: softening of gray matter
Leukomalacia: softening of white matter

MALACIA AND MALACIC DISEASES

- Focal symmetrical poliomyelomalacia syndromes
- **Polioencephalomalacia of ruminants**
- **Thiamine deficiency**
- Nigropallidal encephalomalacia of horses
- Salt (NaCl) poisoning
- Mycotoxic leukoencephalomalacia of horses
- **Lead poisoning**

Thiamine (Vitamine B1) deficiency in ruminants

- Thiamine deficiency in cattle and sheep has been termed polioencephalomalacia.

- Rumen microbes are able to synthesize thiamine.

- Only very young ruminants, whose rumens have not yet been populated by thiamine-producing microbes, are susceptible to thiamine deficiency.

- The disease is seen most commonly in cattle 6 to 18 months of age fed concentrated rations. In sheep, most cases occur in younger age groups (2 to 7 months).

- Clinical signs in cattle and small ruminants may include depression, stupor, ataxia, head pressing, apparent cortical blindness, opisthotonos, convulsions, and recumbency with paddling and then death.

- **Gross lesions**, if present, are limited primarily to the cerebral cortex.

- Initially, 2 days after onset : cerebral edema

- In rare cases with more severe brain swelling: brain displacement with herniation of the parahippocampal gyri beneath the tentorium cerebelli and the vermis of the cerebellum into the foramen magnum can occur.

- 4 days after onset, yellow discoloration of the cerebrocortical gray matter
- Eight to 10 days after onset, edematous separation and cortical necrosis
- In advanced cases with prolonged survival, atrophy of cerebral gyri

Microscopically;

- Neuronal necrosis and edema
- Macrophages and gitter cells to phagocytose necrotic debris
- Laminar separation (at the gray matter–white matter interface) in which there are prominent accumulations of macrophages

LEAD POISONING

The most consistently important poison in **farm animals**.

- ✓Common and fatal in **cattle**;
- ✓Less common but fatal in sheep;
- ✓Occasionally observed in horses, dogs, and cats;
- ✓Rare in swine.

The disease in cattle is probably always **acute**,
in horses is virtually always **chronic**.

Usual sources of lead for cattle: ***paint* and *metallic lead in storage batteries*.**

Lead is usually obtained by **ingestion**, but only a small proportion **(1-2%)** of the ingested dose is **absorbed**.

Absorbed lead is slowly excreted in bile, milk, and urine;

and is deposited in tissues, → in liver and kidneys in acute poisoning

↘ in bones in chronic poisoning.

Clinical syndromes are chiefly neurologic:
Acute poisoning in cattle usually leads to death in 12-24 hours
Staggering, muscle tremors, recumbency, convulsions, opisthotonos, champing of the jaws, hyperesthesia

When the poisoning is less acute;
Dullness and immobility predominate,
Cattle are apparently blind, and without appetite.

The diagnosis of lead poisoning is necessarily **chemical** because lesions are either absent or nonspecific;

Possible **lesions;**

- ✓ The lower gut may contain a small volume of dark fetid feces, attributed to lead sulfide.
- ✓ moderate brain swelling
- ✓ The capillaries and venules are congested
- ✓ Endothelial swelling and proliferation
- ✓ Laminar cortical necrosis
- ✓ Irregular, acid-fast, intranuclear inclusion bodies can be found in the renal tubules

NEURODEGENERATIVE DISEASES

- CENTRAL NEURONOPATHIES AND AXONOPATHIES

Compressive optic neuropathy

Organomercurial poisoning (Minamata disease)

- CENTRAL AND PERIPHERAL NEURONOPATHIES AND AXONOPATHIES

Organophosphate poisoning

Arsenic poisoning

Neonatal copper deficiency (swayback, enzootic ataxia)

PERIPHERAL AXONOPATHIES

Mononeuropathy: lesion involving single peripheral nerve

Mononeuropathy multiplex: several nerves are randomly involved

Polyneuropathy: bilaterally symmetrical involvement of several nerves. It carries the implication of a systemic disturbance.

Equine laryngeal hemiplegia

Neonatal copper deficiency (swayback, enzootic ataxia)

- ✓ Characteristic neurologic disease of lambs, goat kids, and piglets.
- ✓ Caused by maternal/fetal copper deficiency.
- ✓ Clinical swayback in lambs occurs in a congenital form and a delayed form, also called “enzootic ataxia” in which, after being normal at birth, lambs suddenly develop signs at any time between 1 week and several months of age.
- ✓ Affected lambs can be born dead, weak, or unable to stand. If mobile, they are ataxic.

- ✓ Lesions occur in the cerebrum, brainstem, and spinal cord in the congenital form, but only in the brainstem and spinal cord in cases with a postnatal onset.
- ✓ Bilateral and symmetrical gelatinous softening or cavitation.
- ✓ Microscopically, a variable astrogliosis is associated with the degeneration of white matter.

MYELINOPATHIES

- **Hypomyelination/dysmyelination**

Ovine and caprine hypomyelinogenesis

- **Leukodystrophic and myelinolytic diseases**

Globoid cell leukodystrophy (Krabbe disease)

- **Spongy myelinopathies**

Idiopathic spongiform myelinopathies

Toxic/metabolic spongiform myelinopathies

- **Spongy Encephalomyelopathies**

Idiopathic - Toxic/metabolic spongiform myelinopathies

- Branched-chain α-ketoacid decarboxylase deficiency

(Maple syrup urine disease)

- Hepatic and renal encephalopathy
- Hexachlorophene toxicosis
- Halogenated salicylanilide toxicosis
- *Stypandra* toxicosis
- *Helichrysum* toxicosis

Nonmyelinic Spongiform Encephalomyelopathies

- Citrullinemia
- Scrapie of sheep and goats,
- Bovine spongiform encephalopathy (BSE),
- Chronic wasting disease (CWD) of deer and elk,
- Transmissible mink encephalopathy (TME),
- Feline spongiform encephalopathy (FSE),
- Exotic ungulate encephalopathy of captive wild ruminants,
- Neuronal vacuolar degeneration of Angora goats,
- Multifocal spongy encephalomyelopathy in dogs

- Human prion diseases include

✓ *Kuru,*
✓ Creutzfeldt-Jakob disease CJD,
✓ *Gerstmann-Straussler-Scheinker disease* (GSS),
✓ *Fatal familial insomnia* (FFI).

SCRAPIE

- The first **prion disease** to be recognized and described.
- Clinically affected animals are usually in the 2- to 5-year age group

- **Pathogenesis;**

➢Infection occurs probably via ingestion

➢The agent proliferates in the lymphoid tissues and lower intestine

➢May take up to 2 years to reach the nervous system.

Clinical signs;

✓ Affected sheep are initially alert but excitable, tremble when excited, and may have seizures.

✓ Agitated rubbing against posts and trees, behavior that gave rise to the colloquial name “scrapie.”

✓ Self-trauma can cause extensive loss of wool and abrasions of the skin.

✓ Progressive dysmetria and emaciation, and finally paralysis and death.

- No significant **gross lesions**, and no inflammatory changes.

The most characteristic finding is the presence of **large intraneuronal vacuoles** in the medullary reticular, medial vestibular, lateral cuneate, and papilliform nuclei.

Spongy vacuolation of the neuropil in gray matter is the result of vacuolation of neuronal processes

V

BOVINE SPONGIFORM ENCEPHALOPATHY (BSE)

- Ingestion of feed contaminated with **infectious prion** is the most likely origin for BSE.

- Cattle 3-6 years of age become **apprehensive, hyperesthetic, and dysmetric**. Display **fear** and **aggressive behavior**, with progressive gait disturbances leading to frequent falling.

- The pathologic features of BSE resemble those of scrapie

- Vacuolation of neuronal cell bodies and processes is prominent in the dorsal vagal, medullary reticular, vestibular, solitary, spinal trigeminal, and red nuclei;
- accompanied by moderate degrees of spongiform change in neuropil.