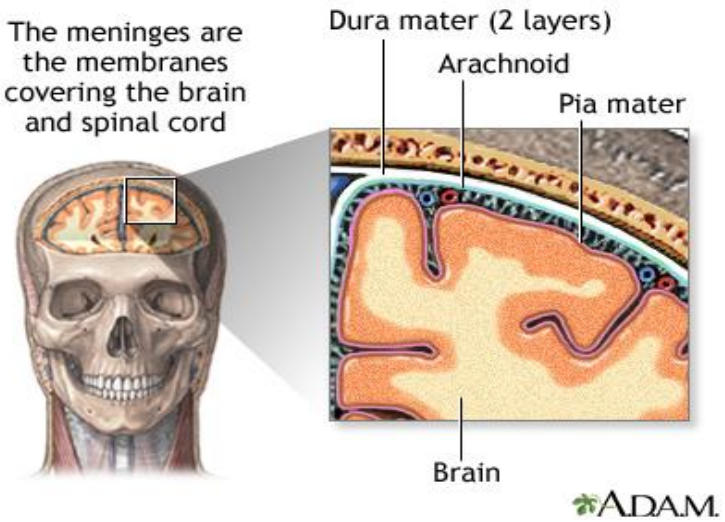
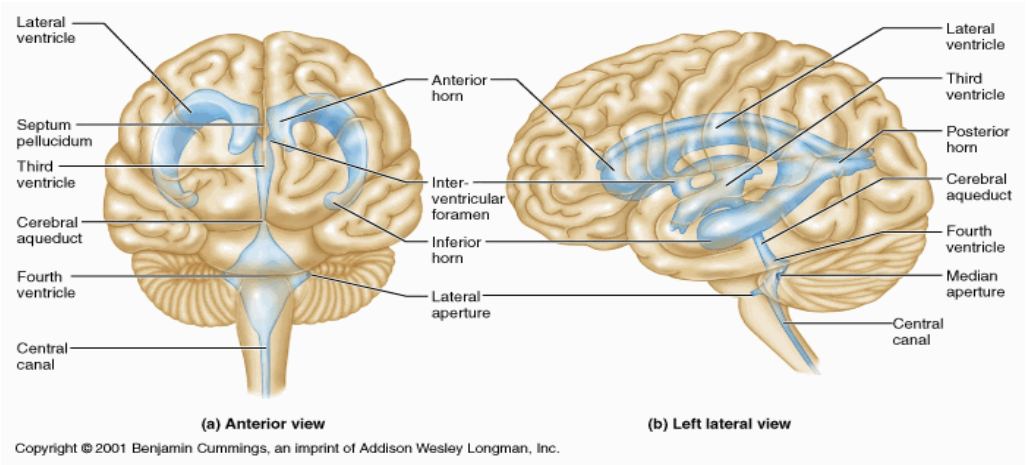
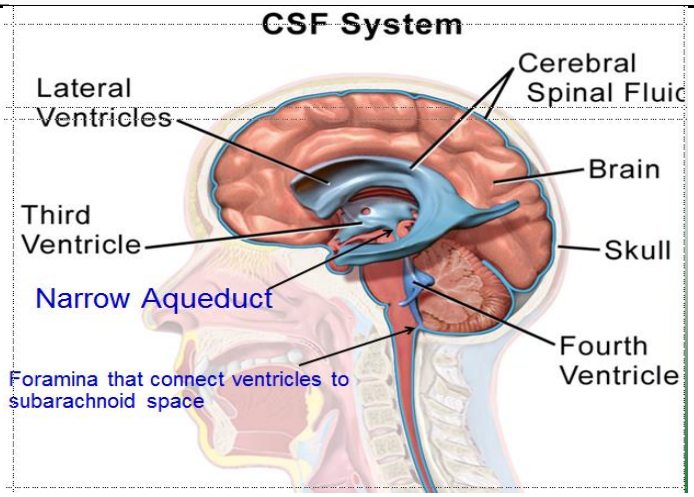
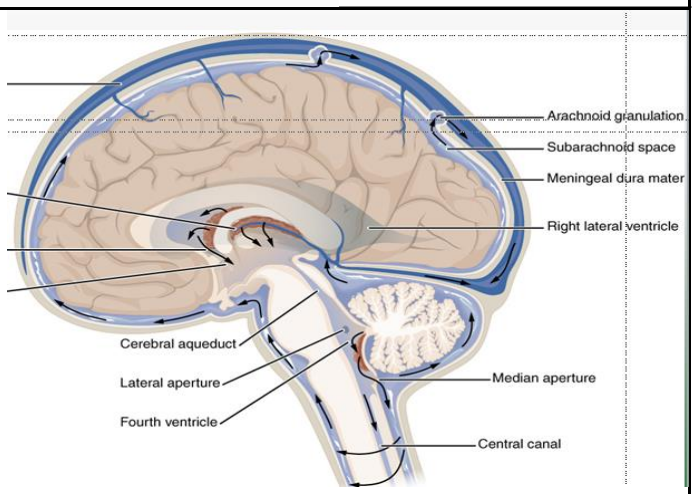
BIOS 2015 ... CHAPTER 14- Neurological Disorders

Page	Note
	Nervous System - two major divisions:
	1. Central nervous system - Brain and spinal cord
	2. Peripheral nervous system - Cranial and spinal nerves - Sensory neurons - Neuromuscular junctions
	The Brain is protected by:
	1. The skull bone
	2. The meinges
	3. Cerebrospinal Fluid (CSF).
	The meninges have three components:
	 Pachymeninges: **1. Dura mater** ("tough mother") thick layer, adherent to bone, forms sac that holds brain. Leptomeninges: **2. Pia and 3.Arachnoid** ("spidery") layers, delicate, thin, transparent membranes that are closely applied to the brain.
	Hemorrhage occurs in three spaces:
	1. Epidural - between skull bone and dura,usually from a ruptured meningeal artery secondary to a skull fracture.
	2. Subdural - between the dura and arachnoid, from rupured bridging veins that extend from the dura to the arachnoid.
	3. Subaracnoid - between the arachnoid and pia, from ruptured vessels in the subarachnoid space, ofter from hypertension.
	CSF - made in ventricles from choroid plexus, blood-csf baarrier controls constituents.
	- CSF circulates from ventricles into subarachnoid space and is absorbed at arachnoid granulations back into the blood stream. Blockage of this pathway causes hydrocephalus - large ventricles resulting in a leage head (see below)





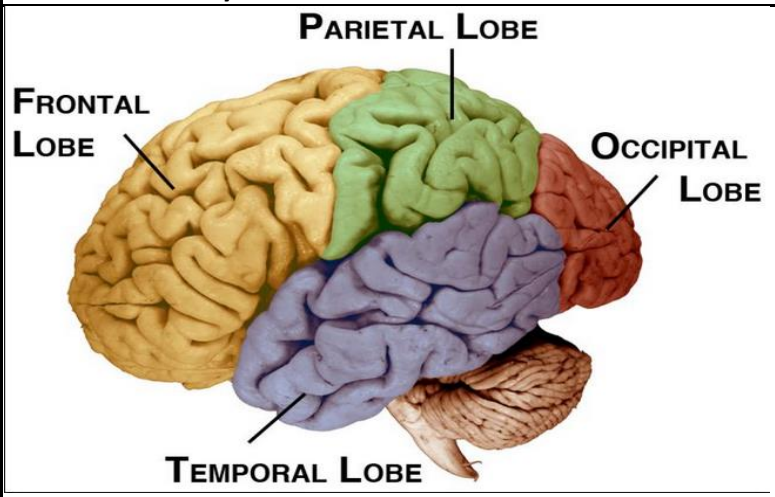


Above - the fourth ventricle is a narrow point in the ventricular system and is often the site of stenosis that results in "non-communicating" hydrocephalus.

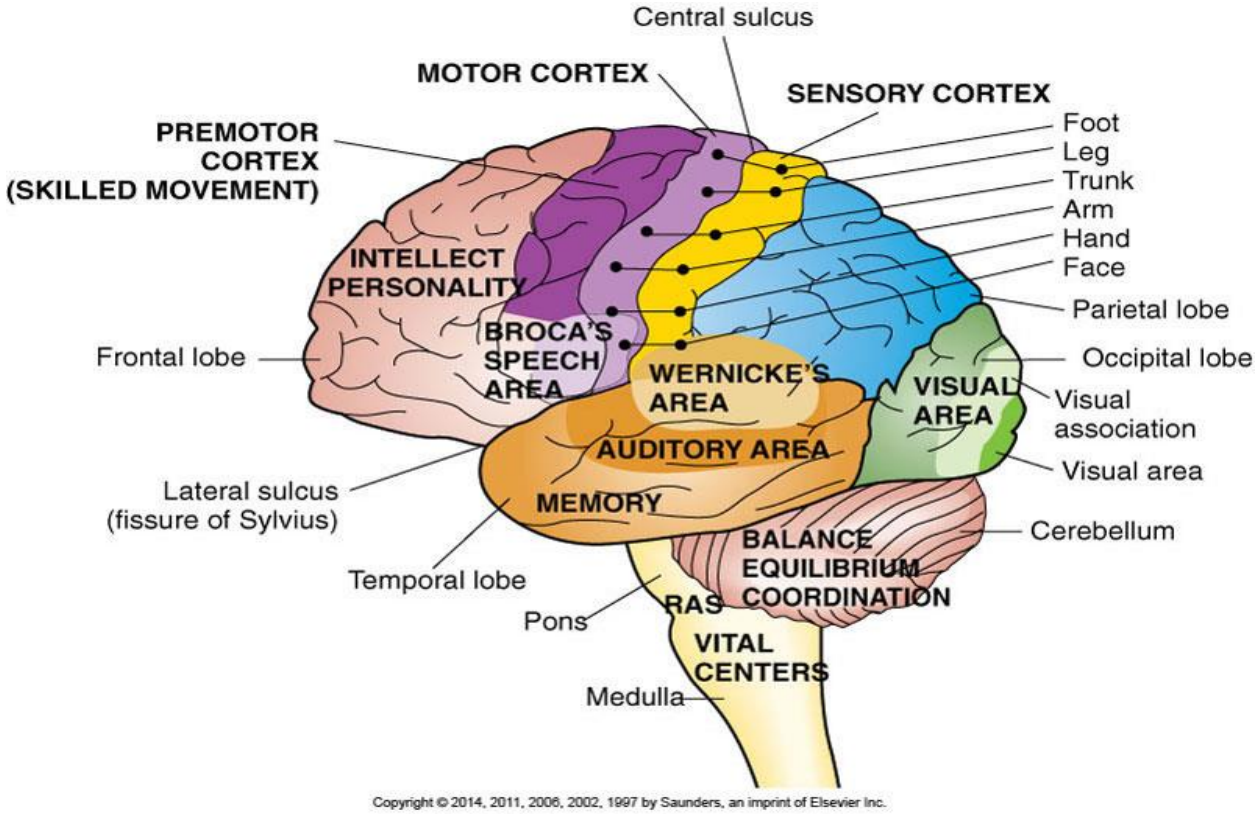
Blood Brain Barrier: Controls what crosses from the circulation into the brain.

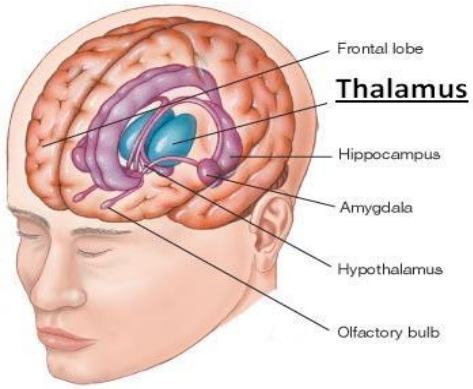
- lipid soluble compounds readily pass through the blod brain barrier. Many drugs will not pass through unless modified to be lipid soluble.

Brain anatomically divided into lobes as below:

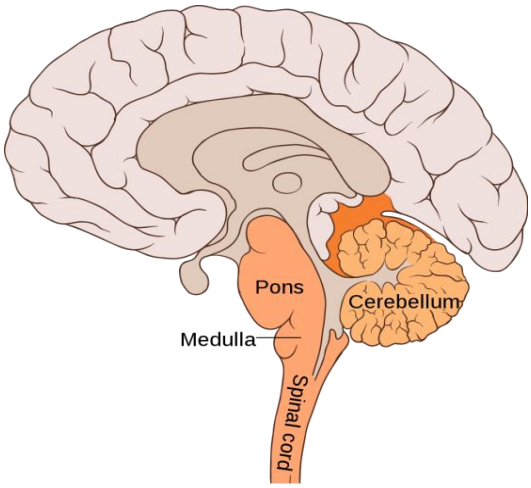


Different areas have specific functions:





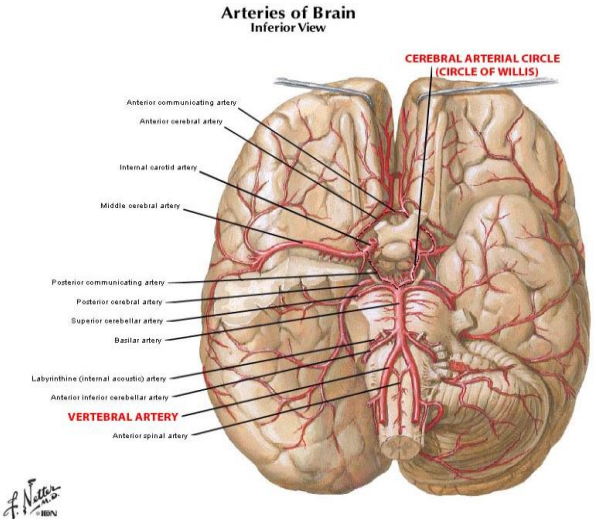
Thalamus
Relay station for incoming sensory impulses
Hypothalamus
Key role in maintaining **homeostasis** of the body
Controls autonomic nervous system and much of the **endocrine system**
Regulates body temperature, fluid and food intake, regulation of sleep cycles, stress response, **emotional responses, sex drive**

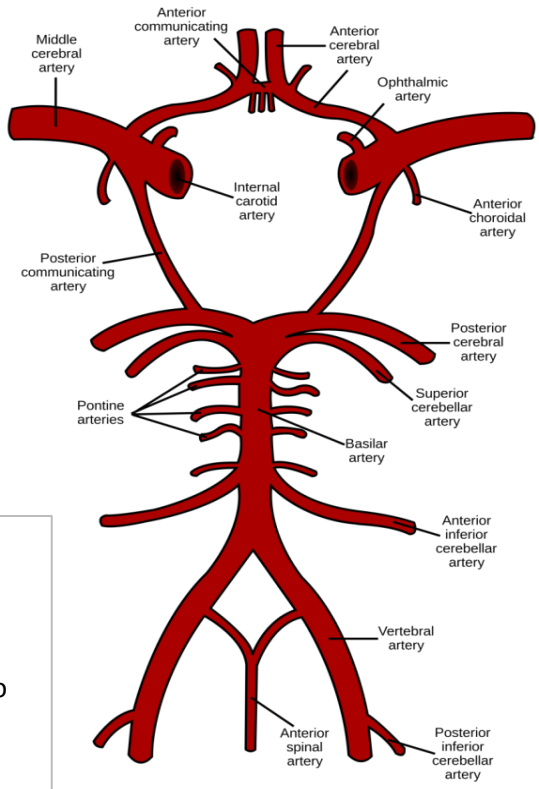


Cerebellum:
Maintenance of posture
Maintenance of equilibrium

BrainStem (Pons and medulla oblongata):
Medulla oblongata
- Control center for **respiratory and cardiovascular function**
- Coordination of **cough reflex, swallowing, vomiting**

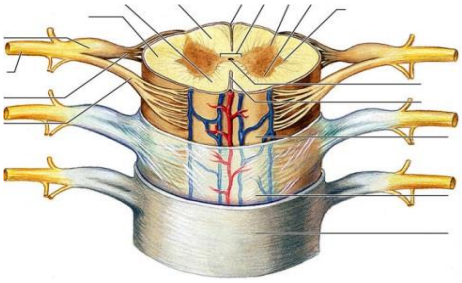
Blood Supply to Brain:

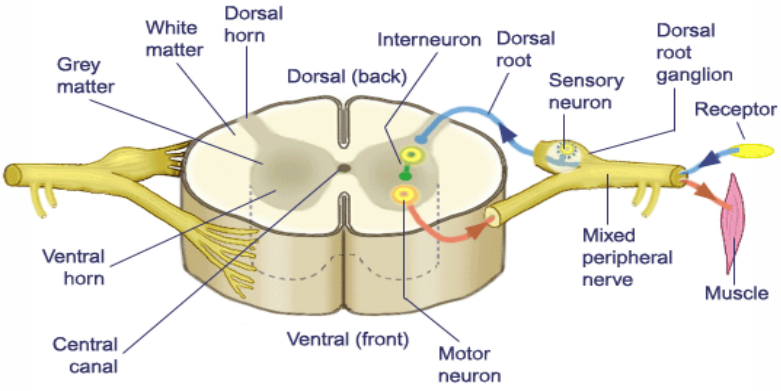




Two main supplies to the brain:
1. Internal carotid arteries.
2. Vertebral arteries (branches off subclavians).
Both feed the circle of Willis.
- Vertebral arteries join to make the basilar artery that gives rise to cerebellar arteries and terminates in a bifurcation giving rise to posterior cerebral arteries.
- The internal carotid arteries give rise to the middle and anterior cerebral arteries.

Spinal Nerves:





31 pairs of spinal nerves
Named by location in the vertebral column where they emerge

Each nerve connected to spinal cord by roots
Dorsal (posterior) roots are :
- Sensory (afferent) fibers

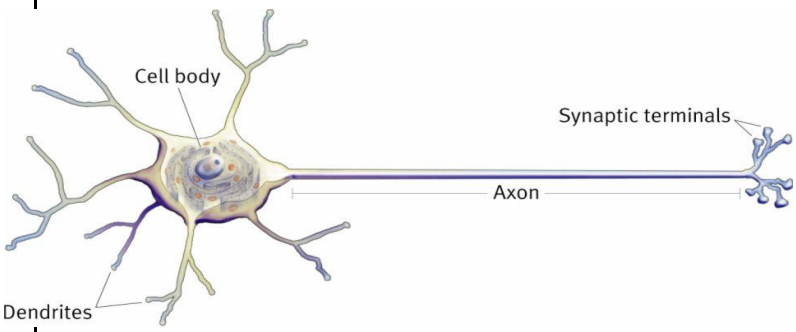
Ventral (anterior) roots are:
- Motor (efferent) fibers

Note the reflex arc:
1. Sensory neuron in dorsal root ganglion senses strech in stretch receptor in patellar tendon in knee (after tendon is stretched when hit with reflex hammer).
2. The sensory neuron stimulates an interneuron.
3. The interneuron stimulates the alpha motor neuron in the ventral horn, and it sends a signal to the muscle to contract.

Note:

- the reflex arc operates without input from the brain, only the spinal cord is required.

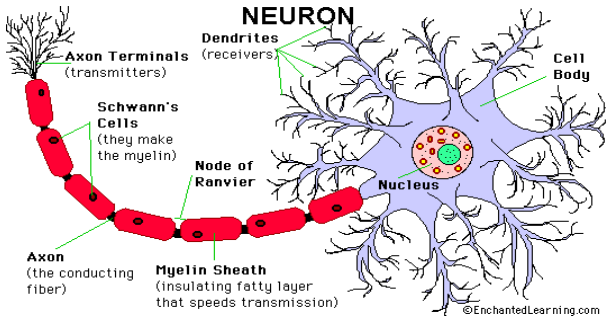
- the motor neuron in this spinal cord is a **"lower motor neuron"** as opposed to motor neurons in the brain, **"upper motor neurons" that allow conscious movement of the muscle, in contrast to a reflex.**



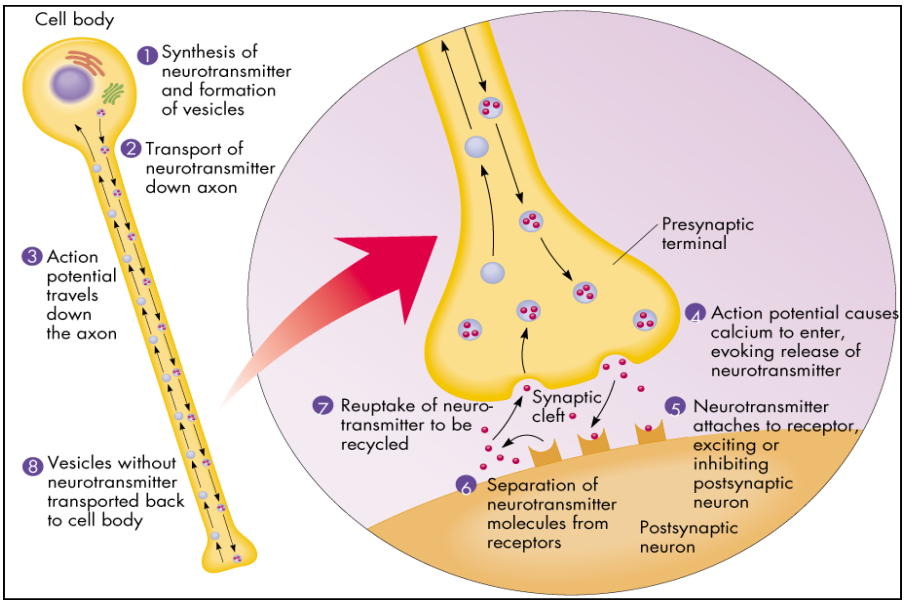
NEURON:

Axons:
Conduct impulses away from cell body

Dendrites:
Conducts impulses toward cell body



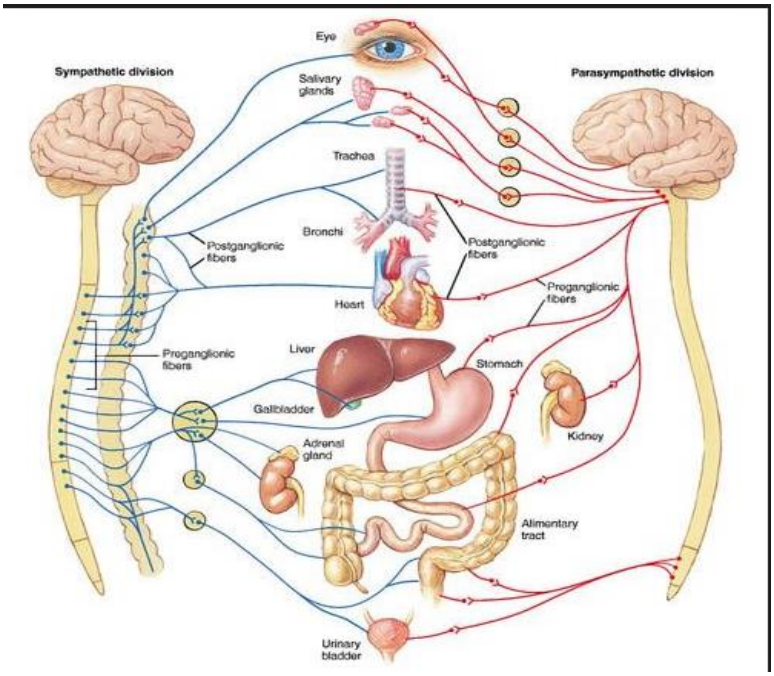
Nerve fibers may be covered by a myelin sheath.
- Insulates fiber
- Speeds up rate of conduction
- Formed by Schwann cells in the PNS
- Formed by oligodendrocytes in the CNS



Neurotransmitters:

1. made in cell body.
2. packaged in synaptic vesicles
3. transported down axon to swelling at the end of the axon - the presynaptic terminal.
4. A nerve impulse causes the vesicle to fuse to the plasma membrane of the presynapic terminal and release the neurotransmitter into the synaptic space.
5. The neurotransmitter binds a receptor on the post synapic neuron or muscle cell and transmissin is done.
6. The neurotransmitter is either degraded in the synaptic space or taken back up into the presynaptic termainal.

Autonomic Nervous System

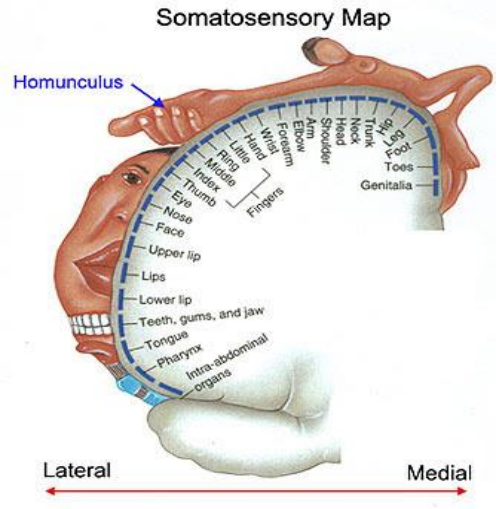


<u>Sympathetic and parasympathetic</u>

- Motor and sensory innervation
- Involuntary
- Both have presynaptic neurons in the brain and spinal cord. These send nerves that terminate in a ganglion (collection of nerve cell bodies, discrete structure).

Sympathetic ganglia are in a chain adjacent to the vertebrae.

Parasympathetic ganglia are more distant from the spine and are adjacent to or within the organ being serviced.



Homunculus is a drawing of the body parts serviced by the sensory and motor strips of the brain drawn onto a picture of the brain (frontal sections) that makes several points:

1. Some body parts (hands and face) get an disproportionate amount of attention vs their true size.

2. The homunculus shows how focal lesions in the brain result in focal deficits.

3. The anatomy is crossed. Cells on the right drive body parts on the left.

Damage to left hemisphere
- Loss of logical thinking ability, analytical skills, other intellectual abilities, communication skills

Damage to right hemisphere
- Impairs appreciation of music and art
- Causes behavioral problems

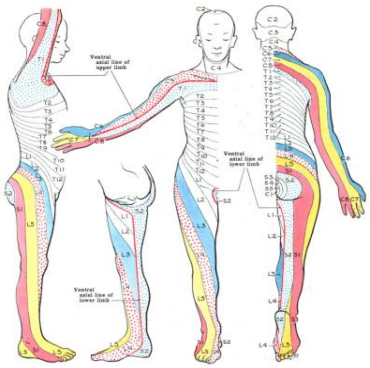
Vegetative state
- Loss of awareness and mental capabilities
- Result of diffuse brain damage
- Person unresponsive to external stimuli

Locked-in syndrome
- Individual is aware and capable of thinking but is paralyzed and cannot communicate

Criteria for brain death
- Cessation of brain function, the cortex and the brainstem.
- Requires a ventilator for breathing.
- Evaluation by two different physicians to declare "brain dead".

Damage to upper motor neurons (in brain)
- Weakness or paralysis on the contralateral side of the body

Damage to lower motor neurons (in spinal cord)
- Weakness or paralysis on the same side of the body
- At and below the level of spinal cord damage



Dermatomes
- Assists in evaluation of spinal cord lesions

- Touch, pain, temperature tested in skin regions to identify spinal nerve and/or spinal cord level that is affected.

Aphasia
Inability to comprehend or express language
Receptive—damage to Wernicke's area (**they speak, but it makes no sense to them or others**).
Expressive—damage to Broca's area (**they know what they want to say but can not get it out**).

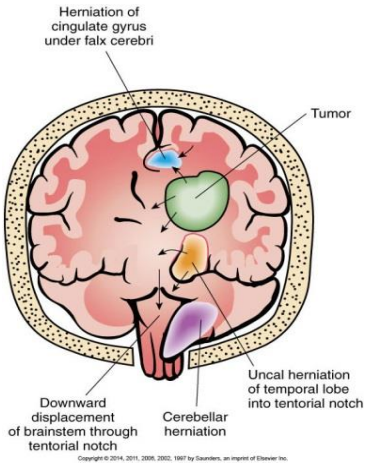
Dysarthria
Motor dysfunction affecting muscles used in speech. Can not speak clearly.

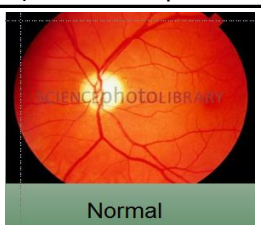
Agraphia
- Impaired writing ability
Alexia
- Impaired reading ability

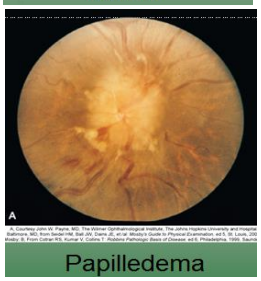
Seizures or convulsions
- Caused by spontaneous, excessive discharge of neurons in the brain

- May be focal or generalized.

- Generalized seizures may be mild (absence or peit mal seizures) or severe (tonic-clonic or grand mal seizures).







A severe increase in intracranial pressure may cause the cerebellum to herniate into the foramen magnum, compressing the brain stem, and shutting down vital functions like the heart beat and respirations.

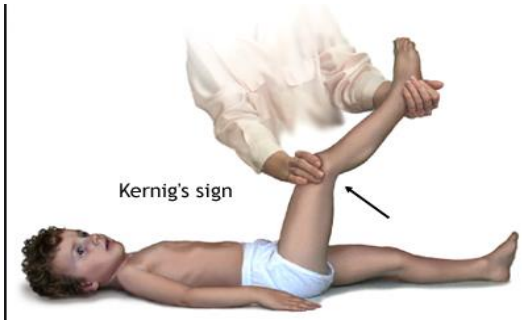
One may see papilledema when examining the retina with an ophthalmoscope.

Brain Tumors

Benign and malignant tumors occur.
Danger is related to location and type.
- Well demarcated benign tumors at edge of brain are easily removed.
- Malignant tumor at the edge may not have defined margins, hard to know if you got it all out.
- Tumor of any kind deep in brain (or in brain stem) may not be operable.
-Tumors can cause hemorrhage and swelling producing fatal edema.

Transient Ischemic Attacks (TIAs), like a mini-stroke with no permanent damage.
- Result from temporary localized reduction of blood flow in the brain

- Impaired function
- Visual disturbances
- Numbness and paresthesia in face
- Transient aphasia or confusion

	Cerebrovascular Accidents (CVAs) or Stroke
	- is an infarction of brain tissue that results from lack of blood.
	- occlusion of a blood vessel (from atherosclerosis or an embolus) or rupture of an aneurysm.
	- Lack of voluntary movement or sensation on opposite side of the body. - Initially flaccid paralysis - Spastic paralysis develops weeks later
	CNS Infections - Meningitis
	- Different age groups are susceptible to infection by different causative organisms.
	- Kernig and Brudzinski signs of meningitis:
	 
	- may also do a lumbar puncture to study and culture the CSF.
	Encephalitis
	- Infection of the parenchymal or connective tissue in the brain and spinal cord
	- usually viral in origin.
	- Western equine encephalitis, St. Louis Encephalitis, West Nile fever are **transmitted by mosquitos**
	- Herpes Simplex Encephalitis
	- Neuroborreliosis (Lyme Disease) **transmitted by a tick**
	Other Infections:
	- Rabies, virus transmitted by the bite of an infected animal.
	-Poliomyelitis (infantile paralysis)
	- Tetanus (“Lock Jaw”) Caused by Clostridium tetani Exotoxin enters nervous system
	Infection-Related Syndromes
	Reye’s syndrome
	- Linked to viral infection in children treated with aspirin - Pathological changes in brain and liver
	Guillain-Barre syndrome
	- Postinfection polyneuritis
	- Progressive muscle weakness with **ascending flaccid paralysis**
	- can be fatal.

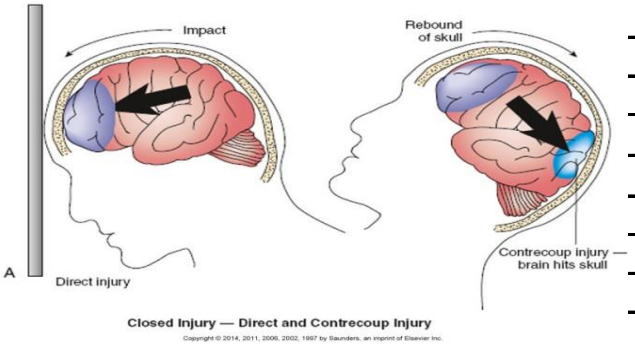
Head Injuries

Concussion (minimal brain trauma)
- Reversible interference with brain function (temporary loss of consciousness).
- Result of mild blow to the head or whiplash-type injury
- Amnesia and headaches may follow

Contusion:
- bruising of brain.

Closed head injury
- Skull bone intact, brain injured.
- May produce "coup" and "contre coup" lesions.

Open Head Injury
- Damage to skull bone.

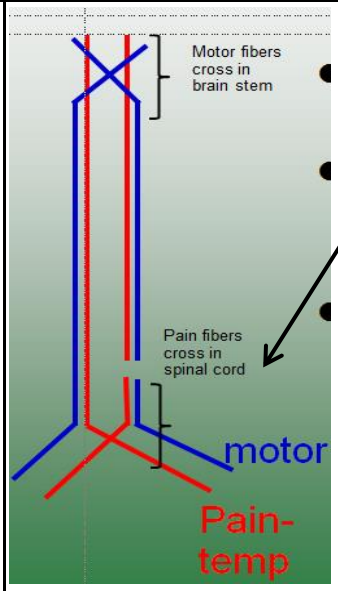


Closed Injury — Direct and Contrecoup Injury

Spinal cord lesion

Spinal shock
Initial period after injury—reflexes absent, flacid paralysis

Recovery - reflexes return, return of function is variable.



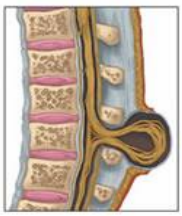
Tetraplegia (quadriplegia)
- Paralysis of all four extremities

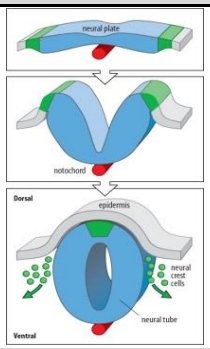
Paraplegia
- Paralysis of the lower part of the trunk and legs

Hemisection (damage to one side of cord)
Ipsilateral paralysis and contralateral loss of pain and temperature sensation

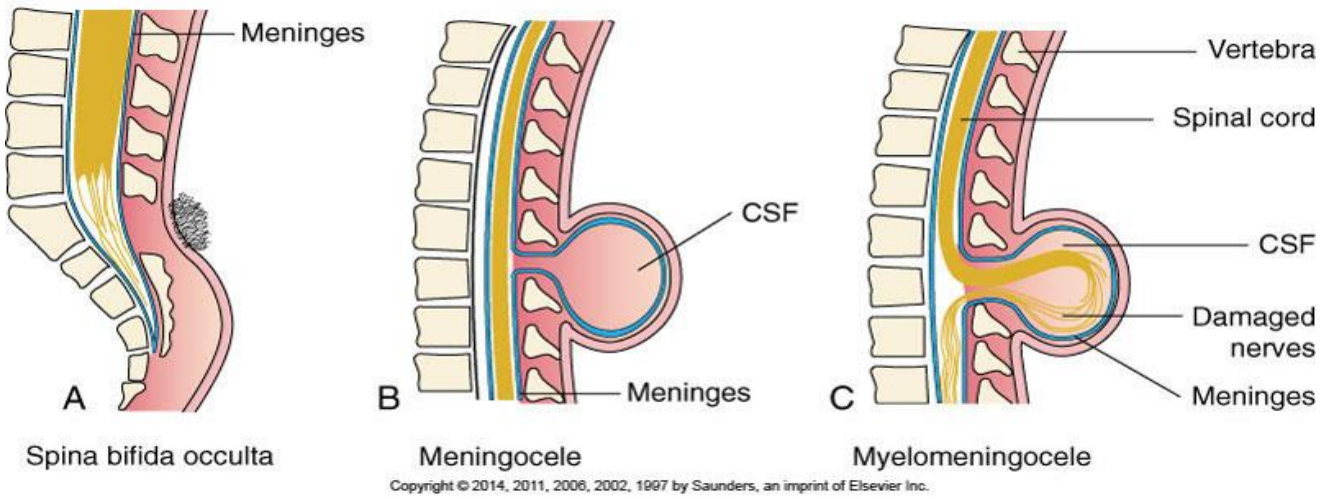
Complications of Spinal Cord Injury
- Urinary tract infections
- Pneumonia
- Skin breakdown
- Spasm and pain
- Depression

SPINA BIFIDA:





- Group of neural tube defects
- Common developmental abnormality
- The neural tube is the progenitor of the spinal cord. It develops from a plate that folds and makes a tube. If the fold does not fuse into a tube, development procedes with a “neural tube defect”



Spina bifida occulta: Bone defect only , skin intact, spinal cord intact.
Meningocele: Bone defect and meningeal sac covered by skin, spinal cord intact.
Myelomeningocele: Like meningocele, but spinal cord disrupted and herniates into sac.
There is also a type where there is a skin defect and exposed neural tissue.

Cerebral Palsy

- a disorder marked by motor impairment.

- many causes: Genetic mutations, abnormal fetal formation of functional brain areas, infection, hypoxia or brain damage in the perinatal period

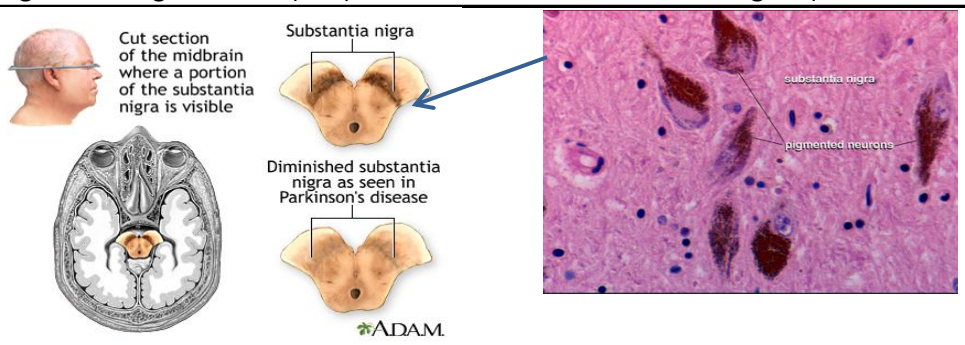
Chronic Degenerative Disorders

Multiple Sclerosis:

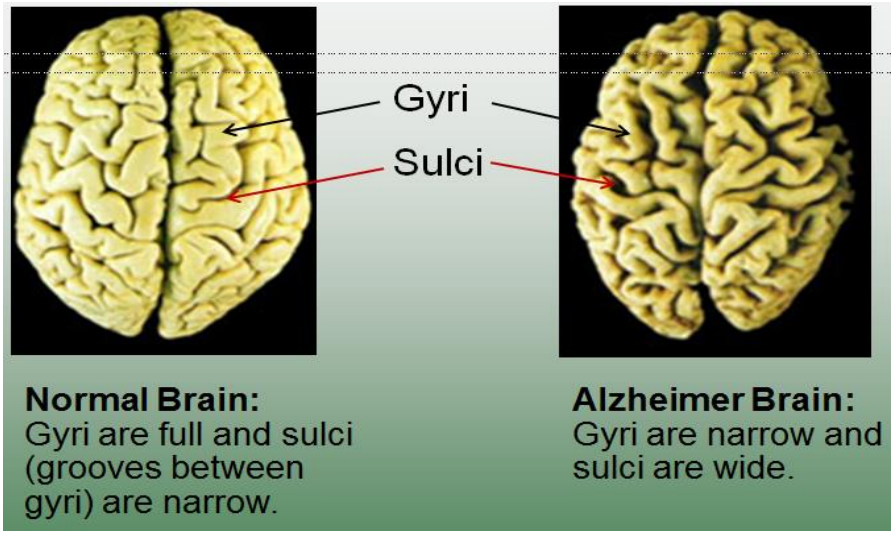
- Progressive demyelination of neurons

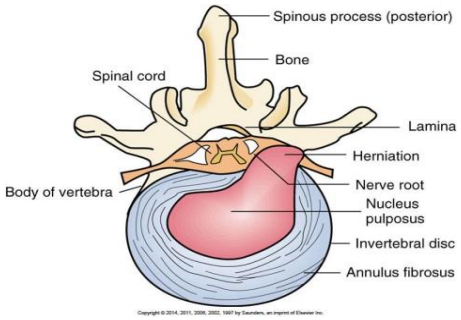
- Onset between 20 to 40 years of age.
- Cause unknown
- May be an autoimmune disease
- Increased risk for close relatives of affected individuals

- Manifestations determined by areas of demyelination
- Blurred vision, weakness in legs
- Diplopia (double vision), scotoma (spot in visual field)
- Dysarthria
- Paresthesia, areas of numbness, burning, tingling
- Progressive weakness and paralysis extending to the upper limbs
- Loss of coordination, bladder, bowel and sexual dysfunction, chronic fatigue

	Parkinson's Disease
	Progressive degeneration (loss) of neurons in the "substantia niagra" (in the midbrain) that make dopamine.
	
	Primary or idiopathic Parkinson's disease Usually develops after age 60 **Secondary parkinsonism caused by:** Encephalitis, Trauma (e.g., sports injury), Vascular disease, Drug-induced (e.g., phenothiazine tranquilizers)
	Early signs and symptoms Fatigue Muscle weakness, muscle aching, Decreased flexibility Less spontaneous changes in facial expression **Tremors in the hands at rest, repetitive pill-rolling motions of hands**
	Later signs and symptoms Tremors affect hands, feet, face, tongue, lips Increased muscle rigidity Difficulty initiating movements Slow movements Characteristic standing posture is stooped, leaning forward Propulsive gait Complex activities become slow and difficult.
	Treatment Removal of cause, if known **Dopamine replacement therapy** - **Levodopa—dopamine precursor** **Anticholinergic drugs** Speech and language pathologist, Physical therapy, Occupational therapy Monitoring and treatment of respiratory and urinary tract infections
	Amyotrophic Lateral Sclerosis (ALS) - aka Lou Gehrig's disease
	No identified cause **Progressive degenerative disease affecting upper motor neurons in the cerebral cortex and lower motor neurons in brainstem and spinal cord** No indication of inflammation around the nerves **Cognition unimpaired**
	Spastic Paralysis from loss of upper motor neurons.
	Flacid Paralysis from loss of lower motor neurons.
	Death is secondary to respirator failure.

	Treatment **No specific treatment to slow degeneratio**n Stem cell therapy under investigation Pharmaceutical treatment (e.g., with Riluzole [Rilutek]) to slow further damage to neurons
	Myasthenia Gravis
	Autoimmune disorder **Autoantibodies to acetylcholine (ACh) receptors destroys receptors.** Skeletal muscle weakness **Facial and ocular muscles usually affected first** **Note: Dysphagia and aspiration are significant problems**
	Diagnostic tests - Electromyography - Serum antibody test
	Treatment Anticholinesterase agents for temporary improvement of neuromuscular transmission Glucocorticoids to suppress the immune system Plasmapheresis to remove antibodies from the blood Thymectomy
	Huntington's Disease
	Inherited disease, Autosomal dominant gene, Carried on chromosome 4 **Does not usually manifest until individual is older than 40 years** **Progressive atrophy of brain** **Particularly in basal ganglia (nuclei) and frontal cortex** **Depletion of gamma-aminobutyric acid (GABA) in the basal nuclei**
	Signs and symptoms Mood swings, personality changes **Restlessness, choreiform movements in arms and face** Diagnostic tests, DNA analysis Treatment, Currently no therapy to slow progression of disease
	Dementia
	Progressive chronic disease **Cortical function is decreased.** - Impaired cognitive skills - Impaired thinking, judgment, and learning - Memory loss - Confusion - Behavioral and personality changes **Many causes of dementia** - Vascular disease - Infections - Genetic disorders

	Alzheimer's Disease
	
	<u>Signs and symptoms of Alzheimer's Disease</u> Extend over 10 to 20 years Behavioral changes, Irritability, hostility, mood swings Gradual loss of memory and lack of concentration Impaired learning, poor judgment Decline of cognitive function, memory, language Change in food intake Inability to recognize family, lack of environmental awareness, incontinence, inability to function
	Treatment No specific treatment Anticholinesterase drugs, Some temporary improvement
	Other Forms of Dementia
	Vascular dementia Caused by cerebrovascular disease
	Creutzfeldt-Jacob disease (CJD) **Rare, rapidly progressive** **Caused by prion (an infectious protein)** ingested or transmitted through contaminated blood
	AIDS Dementia , Common in later stages of AIDS

	Spinal Cord Problems	
		**Herniated intervertebral disk** Tear in capsule may occur suddenly or develop gradually with aging or obesity. Nucleus pulposus protrudes through ear and compresses nerve roots. Sensory, motor, or autonomic function may be impaired. Most common location—lumbosacral disks