

HEMOPHILIA

(1)

Hemophilia → an inherited bleeding disorder

↓ Resulting
from congenital deficiency
in factor VIII or IX

* linked disorder ~~is~~ recessive genetic

Pattern

↓ so

Males are commonly affected
as they have X Y
Single X

But

Females are usually carriers of the
disease
X X Double X

When both X chromosomes are
affected they may have Haemophilia

Haemophilia comes in 2 types

Haemophilia - A - factor VIII

Haemophilia - B - factor IX

where the clotting factors are deficient or absent
which helps in clotting

↓

Its a rare genetic disorder in which blood
clots - so slowly

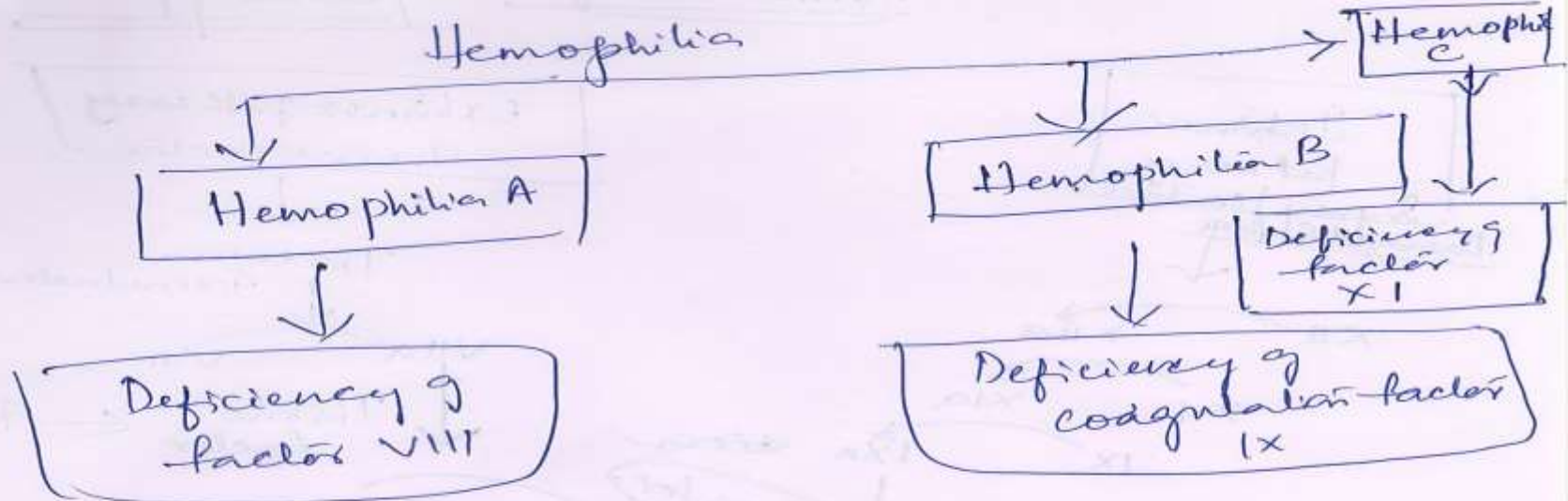
↓

due to
deficit clotting factors
VIII & IX

* the person can bleed to death with
a small injury as the veins don't
clot at all.

Haemophilia - A - Classical Hemophilia
B - Christmas disease.

"

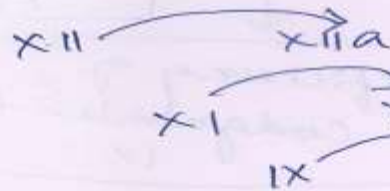


Epidemiology : More common in Males
 Seen in } 1 : 4000 to 1 : 5000 Males
 Ratio of }

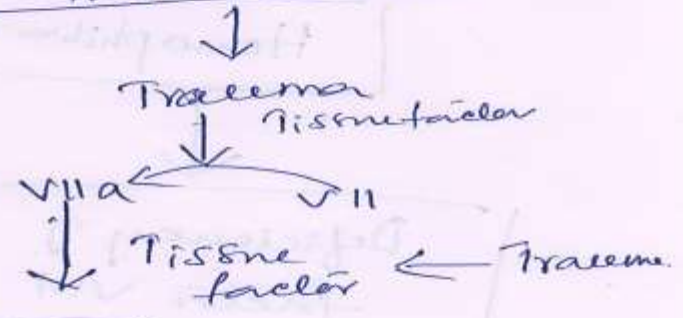
- Etiology :
1. Hereditary from parent to child through
 2. Medical conditions like
 1. Liver Disease
 2. Vit K deficiency
 3. Side effect of certain Medicines

Normal coagulation/clotting process (4)

Intrinsic Pathway
Injury/contact
Damaged surface



Extrinsic pathway
Tissue factor



VIII cofactor for Xa

Prothrombin II

Thrombin IIa
fibrinogen → fibrin

XIIIa XIII
cross linked
fibrin
clot
(Stable fibrin)

Common Pathway →

Intrinsic system
Surface damage

Extrinsic pathway (5)

Tissue factor

XII → XIIa

VII → VIIa

Factor Eleven Deficit
H¹C² → Rosenthal's Disease

XI → XIa

Christmas disease
H¹B¹ Deficit
IX

IX → IXa

VIII → VIIIa

Factor VIII Deficiency
H¹A

X

Xa

V → Va

Prothrombin II

IIa Thrombin

I Fibrinogen

IIa Fibrin

What is Van Willebrand disease ???

???

Pathogenesis

1. Factors deficiency
2. Factors dysfunction
3. Factor inhibitors

↓ lead to
Disruption of clotting cascade

↓ ↓
Resulting Spontaneous H[']age
in response to trauma

H[']age site includes :

1. Joints
2. Muscles
3. CNS
4. GIT
5. Genito urinary
6. Pulmonary
7. CVS

When a blood vessel is injured

- First → Blood vessel constricts to limit the loss of blood
- Secondly → Circulating platelets form plug at the injured site
- Finally → Coagulation occurs by activation of mechanism of clotting

(1)

Proteins in a sequence allows the platelet plug to be stabilized by fibrin matrix

Which forms over the damaged part ensuring that vessel wall to heal.

Factor VIII & IX are the only 2 of 13 proteins involved in cascade process of coagulation

Absence or deficiency — Leads to Bleeding disorders

Treatment : is determined by severity of the disease

Mild & Moderate Hemophilia 'A'

: Require only temporary treatment to correct clotting defect

Preoperatively / following trauma

or before dental extractions

Desmopressin → Synthetic form of Vasopressin

but it is ineffective in Hemophilia 'B'

as they require replacement treatment
of factor IX

⑧. For Severe Hemophilia pat's : Effective treatment is replacing missing clotting factors.

freeze dried preparation, comes in various vial sizes

250, 500 or 1000 units

of factor VIII or IX

Dose depends on severity / pat's wt / site of bleeding.

Most of children :

Treated at regular intervals
(three times a week) as
Prophylactic Measures.

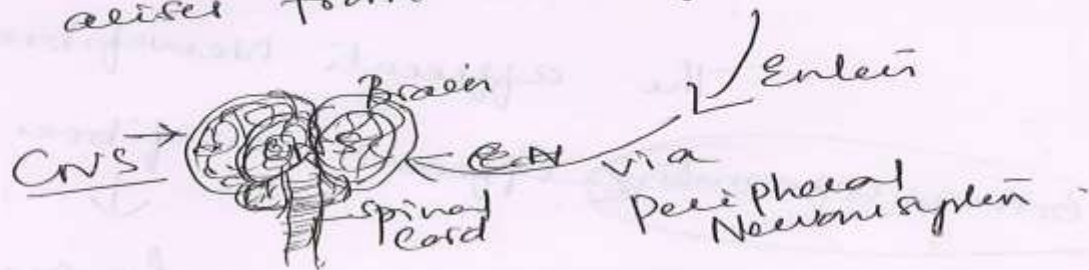
Central Nervous System ①

Nervous system is essential control system of Human body.

Consists of

- CNS (Central)
- Peripheral Nervous system.

Information arrives from sensory receptors

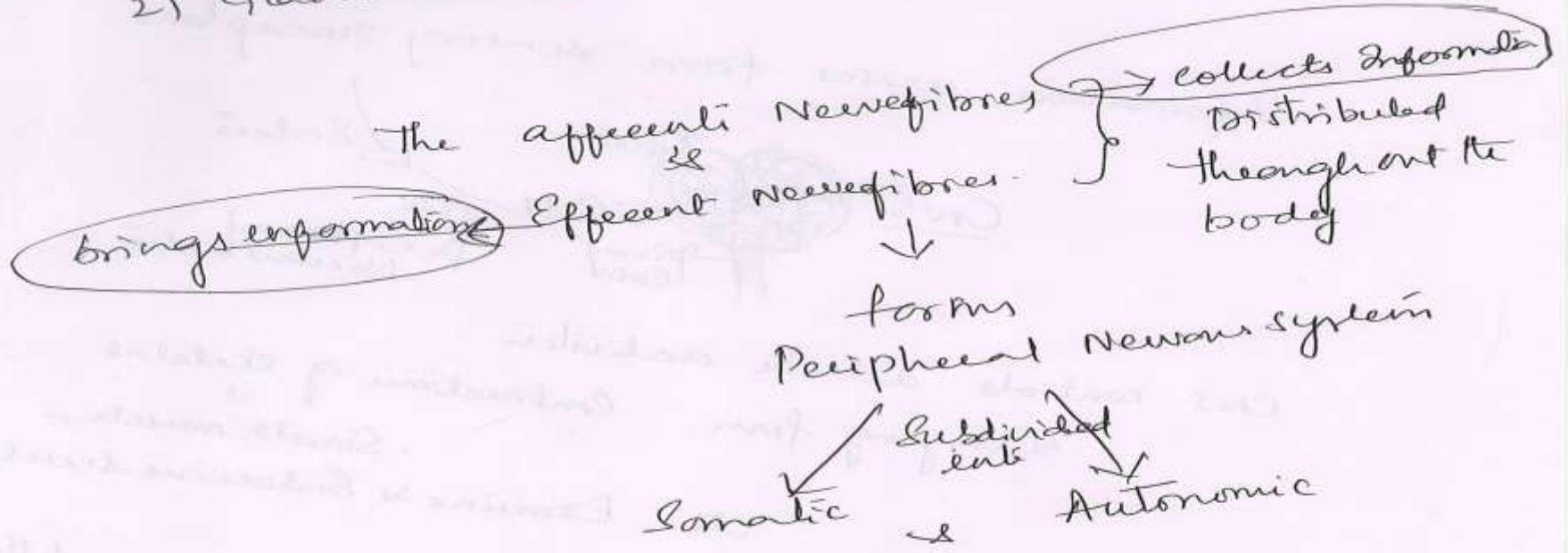


CNS controls all the activities ranging from contractions of skeletal & smooth muscles to Exocrine & Endocrine secretion.

Sensory receptors detect events in the body and the outside world. Signals or action potential from sense organs travel through peripheral afferent nerves to CNS,

They are processed at CNS ⁽²⁾ which controls the various activities of the body by Motor Mechanisms that generate movements and glandular secretions

- ↓
Generates
- 1) Movements
 - 2) Glandular secretions
- } through Efferent nerves.



Neurons → Highly specialized cells that are

- ① Excitatory
- ② Inhibitory
- ③ Neurosecretory

} - sometimes

③
Neurons receive and transmit signals to other
Neurons.
or
Effectors

Neuronal network accounts for

- Information in memory
- Evaluation of available knowledge
- Decision making
- Transmission of response signals

to appropriate effectors.

Brain controls "involuntary" activities such as

- Heart beat
- Respiration
- Digestion
- Secretion
- Movement

Brain also controls → higher order activities like

- thought
- Reasoning
- Abstraction (Dealing
e.g. gears.)

Human brain is ^{regarded as} more capable of
higher order activities than
any other species.

EPILEPSY

Def: Epilepsy is a group of neurological disorders characterized by seizures.

* It's a neurological disorder marked by sudden loss of consciousness or convulsions associated with abnormal electrical activity in the brain.

* It's ch. disorder of brain which causes recurrent seizures.

* Common in children to have - especially associated with high fever.

* Estimated that 1 in 10 people will experience in life.

Epilepsy is a condition characterized by recurrent recurrent seizures that may include repetitive muscle jerking called convulsions.

* Seizures are sudden disruption of brain's normal electrical activity accompanied by altered

consciousness & other Neurological & behavioural Manifestations (14) (15)

Types of seizures

1) Generalized Epileptic seizures :

* Occur when electrical abnormalities exist throughout the brain

* Partial seizure begins in an area called an epileptic focus and may spread to other parts of the brain and causes generalized seizure.

* Various parts of body involves to jerk repeatedly which last for few minutes to $\frac{1}{2}$ hr or < 1 hr \rightarrow Motor attacks
& Sensory attacks : Begin in one area.

\rightarrow Sensation may move along one side of the body before subsiding

* Visual seizures : Affect the area of the brain that controls sight \rightarrow & cause to see things that are not there

* Auditory " \rightarrow affect part of brain that controls hearing and cause patients to