COMPONENTS OF BLOOD

Date

preparation should start ē in 6 Hrs...

Whole blood components	Storage temperature	Shelf life
Red blood cells	2-6°C	42 days
Platelets	20-24°C (ROOM TEMP.)	5 days (ē continuous AGITATION)
Fresh frozen plasma (FFP)	-18 to -30°C	1 year
Cryoprecipitate	-18 to -30°C	1 year

*Cryoprecipitate rich in vWF, factors 8,13 and fibrinogen and FFP has other clotting factors

ADDITIVES → BLOOD BAG

Anti coagulants	Name	Shelf life
ACD	Acid citrate dextrose	21days } 3 WKS
CPD	Citrate phosphate dextrose	21days
CPD-A	Citrate phosphate dextrose adenine	35days ♀ 5 WKS
SAGM	Saline adenine glucose mannitol (citrate & phosphate too)	42days 6 WKS

Cerebellum Coupon code:-NEETPG

ADENINE → REASON FOR ↑ LIFE SPAN

ACD
↓
Acidified

1. GLUCOSE / DEXTROSE → Nutrition to RBC
2. PHOSPHATE → BUFFER : Ⓝ pH Maintained
3. CITRATE → CHELATION OF Ca^{2+} → ∴ PREVENT CLOT
4. ADENINE → ATP ⇝ stabilise RBC Membrane
5. SALINE → ISOTONICITY
6. MANNITOL → PREVENT HEMOLYSIS

★ EXCESSIVE CITRATE → can result in ↓ Ca^{2+} TETANY

BLOOD TRANSFUSION

Date

INDICATIONS OF BT

1. WHOLE BLOOD : HYPOVOLEMIC SHOCK in Trauma Patient
2. pRBC : ANEMIA
3. FFP : MULTIPLE CLOT FACTOR DEFICIENCY
4. CRYOPRECIPITATE : vWF / VIII / XIII / FIBRINOGEN
   - vWF → vWD
   - VIII → HEMOPHILIA-'A' → CAN'T BE GIVEN IN ALL HEMOPHILIAS
   - FIBRINOGEN → AFIBRINOGENEMIA

TRANSFUSION PROTOCOL → WHOLE BLOOD / pRBC

* START TRANSFUSION c̄ in 30 Minutes
* COMPLETE TRANSFUSION c̄ in 4 Hours

Cerebellum Coupon code:-NEETPG

± if started >30 Minutes → FNHTR

MASSIVE BT → >1 BLOOD VOLUME c̄ in 24 Hours or

5L = 10 BLOOD BAGS

50% BLOOD VOLUME c̄ in 3 Hours

COMPLICATIONS

1. HYPOTHERMIA → prevented by WARMING BLOOD ON WARMER → if NA → Slow Transfusion
2. CITRATE ↑ → ↓↓ Ca^{2+} TETANY → Rx : Ca^{2+} GLUCONATE
   - metabolized by LIVER
3. ↑↑ HCO_3^-
4. METABOLIC ALKALOSIS
5. RBC LYSIS → ↑↑ K^+

HYPOKALEMIA >>> HYPERKALEMIA

MOST © CAUSE OF DEATH

- in Massive BT → DILUTIONAL COAGULOPATHY (↓Plts / ↓ Clotting factors)
- in BT → TACO

COMPONENTS OF BLOOD

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Cerebellum Coupon code:-NEETPG

APPROACH TO TRANSFUSION

Date

TIMING

ACUTE (<24 Hr)

FEVER

⊕ FEVER

- **AHTR** (ABO INCOMPATIBILITY)
  - PREFORMED ANTIBODIES
  - IV HEMOLYSIS (DCT ⊕)
    - HEMOGLOBINURIA (dark colored urine) → flank pain, back pain
    - AKI
    - DIC (coagulopathy) → ↓ Platelets, ↑ PT/INR
  - OT Patients
    - ↑ OOZING
    - ↑ BLEEDING from Surgical site
    - ↓ BP
  - Rx: SUPPORTIVE (IV Fluids)
- **FNHTR** (ANTI-HLA)
  - DONOR WBCs
  - CYTOKINE RELEASE
  - FEVER | CHILLS
  - NO HEMOLYSIS
  - Rx: SUPPORTIVE
  - I°: LEUKOREDUCTION
    - INDICATIONS
      - i) FNHTR
      - ii) Regular Transfusions
      - iii) Transplant candidates
- **TTI** (Bacterial | Viral)
  - GPC → PLATELETS
  - GNR → pRBC
  - MOST © CAUSE POST-TRANSFUSION HEPATITIS
  - HBV > HCV
- **TRALI** (<6 Hrs)
  - FEVER ⊕
  - NON-♡ PULMONARY EDEMA
    - JVP ⓝ
    - BNP ⓝ
    - PCWP ⓟ
    - ↓ BP
  - DONOR ANTI-HLA Ab → RECIPIENT WBC
  - FFP >>> pRBC

⊖

- **ALLERGIC**
  - MINOR → URTICARIA
    - Rx: Anti-Histaminics
  - MAJOR → ANAPHYLAXIS
    - Urticaria
    - Angioedema
    - SOB | Strider
    - ↓ BP
    - ± Rx: ADRENALINE + ANTI-HISTAMINICS + DEXA + IV fluids
    - ± If Recurrent → look for IgA deficiency; use WASHED pRBC

DELAYED (>24 Hrs)

- **DHTR**
  - extremely similar to AHTR
  - BUT ↓↓↓ SEVERITY
  - Minor Ag incompatibility (DUFFY / KIDD Ag)
- **PTP**
  - Ab: ANTI-HPA1a
  - Rx: IVIG (if severe)
- **TA-GVHD**
  - POST-HEMATOPOETIC STEM CELL TRANSPLANT
  - SCID
  - BM ↓↓ d/t DONOR T-CELLS
  - ∴ Avoid IRRADIATED BLOOD PRODUCTS

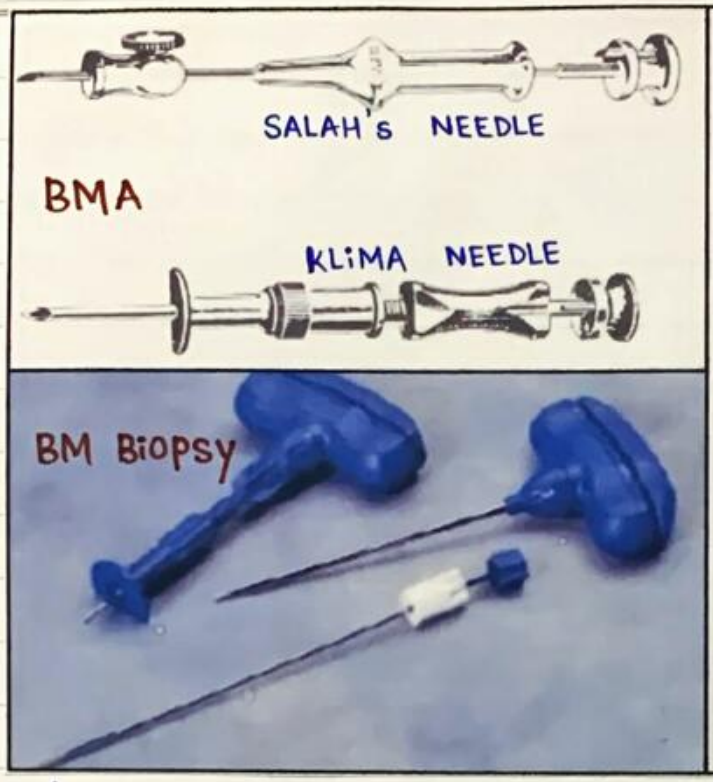
REACTIONS → NEXT STEP : STOP TRANSFUSION

Date

- **TACO** (<12 Hrs)
  - FEVER ⊖
  - ♡ PULMONARY EDEMA
    - JVP ↑↑
    - BNP ↑↑
    - PCWP ↑↑
    - BP ⓝ / ↑↑
    - pRBC >>> FFP
- * HAV → NOT CHECKED IN BLOOD BANK
- * MALARIA → transmitted through ALL BLOOD COMPONENTS → Transfusion transmitted MALARIA → TROPHOZOITES (resistant)

Cerebellum Coupon code:-NEETPG

Date



SAHLi HEMOGLOBiNOMETER

SiTE

* PSIS
* ASIS in OBESE ADULT
* TiBiA in INFANTS

Cerebellum Coupon code:-NEETPG

PLEURIPOTENT HSC

COMMON MYELOID PROGENITOR → EPO & Fe^{2+} INDEPENDENT

BFU-E ← EPO DEPENDENT

EPO (+) → CFU-E

PRONORMOBLAST (= PROERYTHROBLAST) → Hb 1st Appears in EM

NORMOBLAST

- EARLY (BASOPHILIC)
- INTERMEDIATE (POLYCHROMATOPHILIC) → Hb 1st in L/M
- LATE (ORTHOCHROMATIC) → NUCLEUS REMOVAL

RETICULOCYTES → 1-2 Days → MATURE RBC

BPO

Date

* Giant Pro-Erythroblast → seen in PARVO Big INFECTION

RETICULOCYTE COUNT → (N) = 0.5 - 1.5% Q

PB ⇒ Marker of BM → HEMATOPOEITIC ACTIVITY Q

↑ RETIC. COUNT
↓
HEMOLYTIC ANEMIAS
eg. HS

↓ RETIC. COUNT
↓
1. APLASTIC ANEMIA
2. NUTRITION ANEMIA
↓
eg. IDA / Megaloblastic Anemia

* RETICULOCYTE COUNT has to be corrected for '2' REASONS
- Hb level
- Maturation Time

Cerebellum Coupon code:-NEETPG

* After CORRECTION, we get RPI

$$\text{RPI} = \text{RETIC. COUNT} \times \frac{\text{Pt's Hb}}{\text{DESIRED Hb}}$$

- RPI < 2.5 → HYPOPROLIFERATIVE ANEMIA
- RPI > 2.5 → HYPERPROLIFERATIVE ANEMIA

* Stain → ROMANOWSKY STAIN (NON-SPECIFIC) can be used

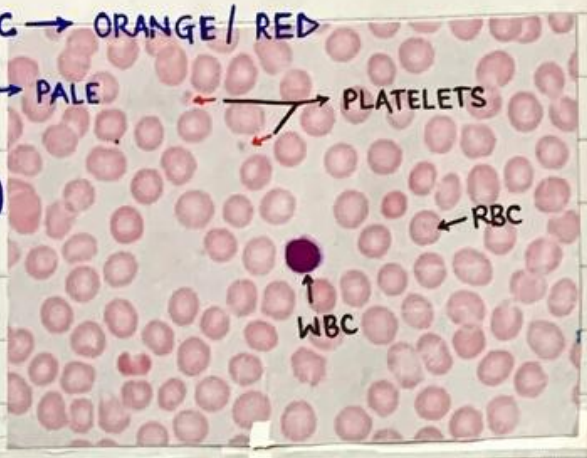
* Best Stain → SUPRAVITAL STAIN
- NEW METHYLENE BLUE (BEST)
- BRILLIANT CRESYL BLUE

→ Live RBC used

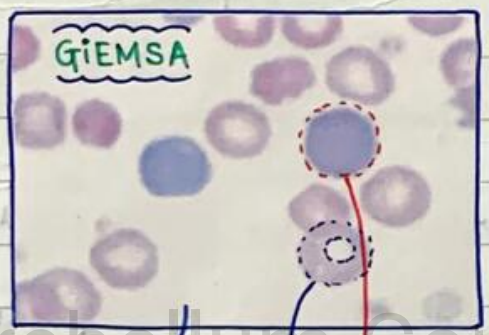
ate

ROUTINE STAIN for 'PS'

ROMANOWSKY STAIN (family)
- RBC → ORANGE / RED
- B/g → PALE

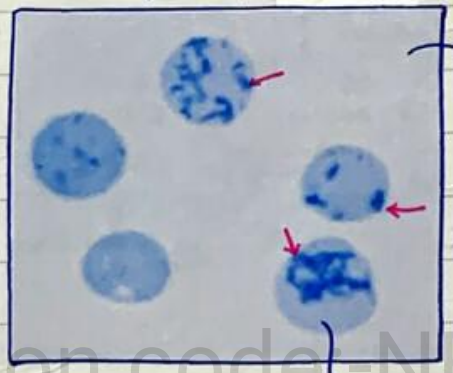


1. GIEMSA STAIN (MC Used / BEST)
2. Jenner
3. Wright
4. Leishmann
5. FIELD STAIN → used for P. Falciparum



PALE B/g

RETICULOCYTES
- Ⓝ RBC
- * larger than RBC
- * non-nucleated



NEW METHYLENE BLUE → BEST

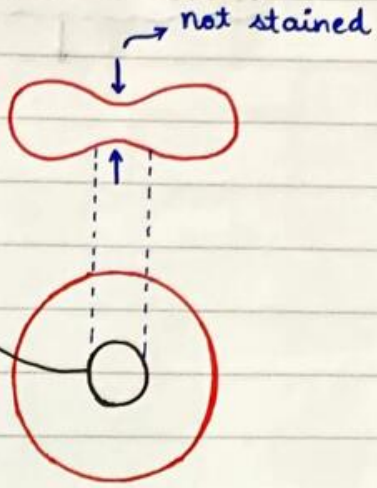
RETICULOCYTES
- (xx / xx) → Reticulum like structure → RNA

Cerebellum Coupon code-NEETPG

Date

RBC

* DIAMETER → 7-8 μm
* BICONCAVE / DUMBELL SHAPED
  - SPECTRIN



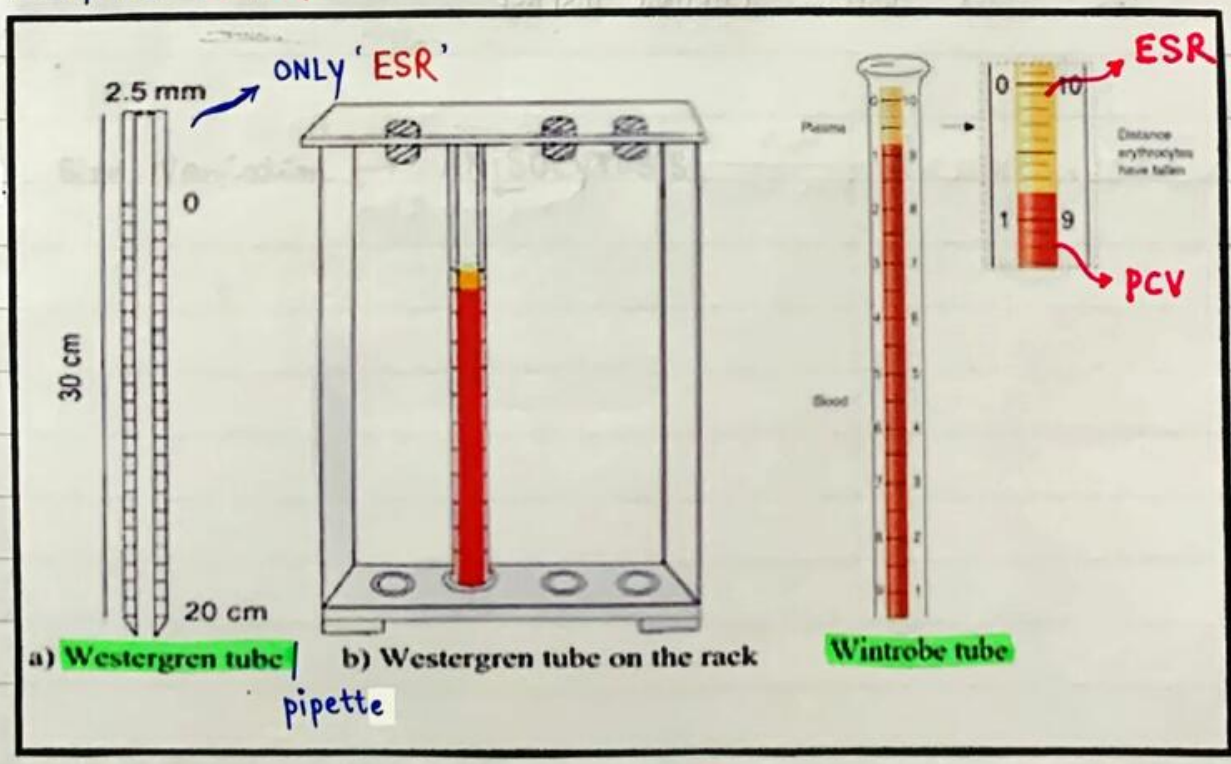
CENTRAL PALLOR → { Ⓝ = CP = $\frac{1}{3}$ d_{RBC} }

- HYPOCHROMIC ANEMIA → CP > $\frac{1}{2}$ d_{RBC}

RBC INDICES

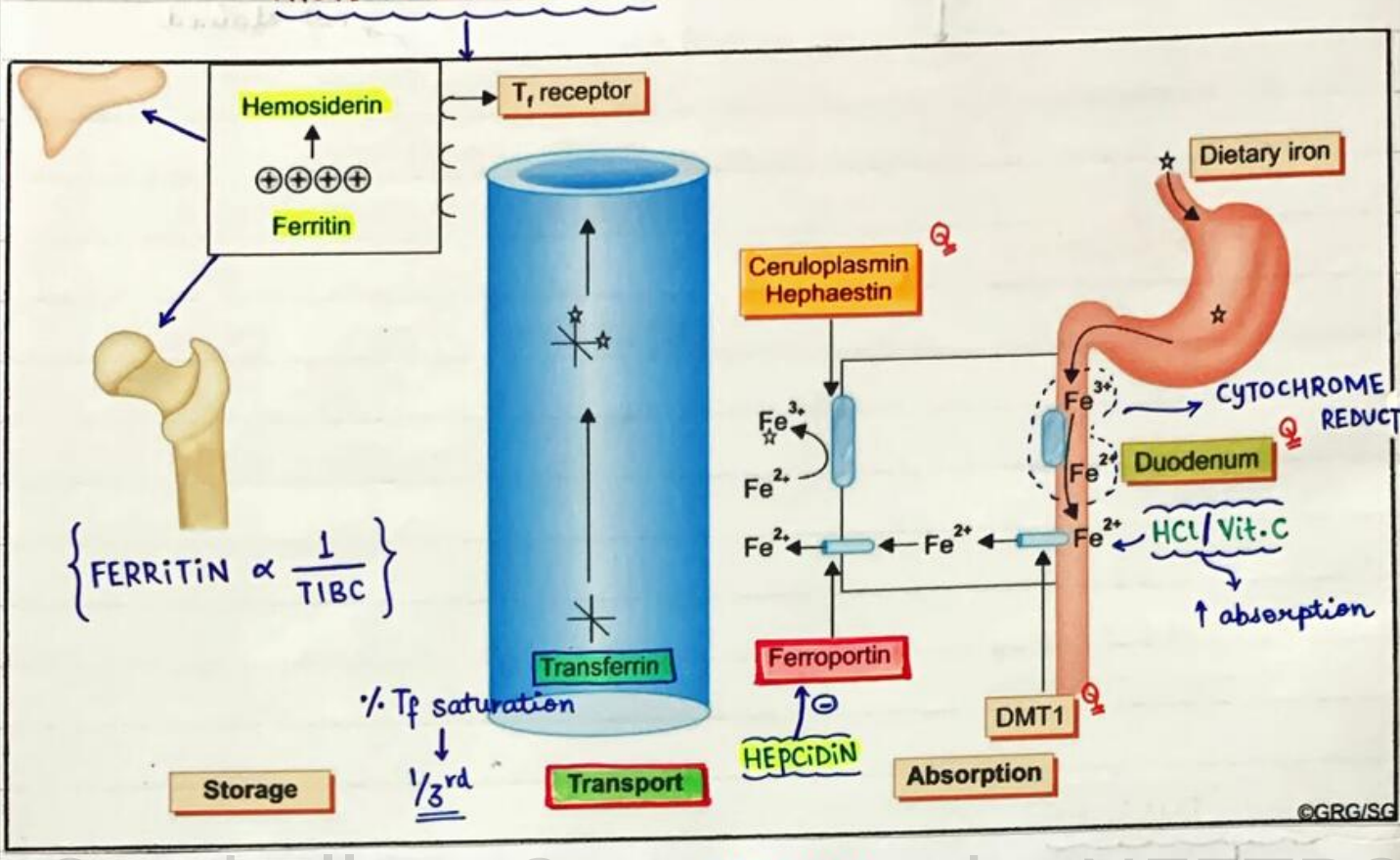
Cerebellum Coupon code:-NEETPG

1. MCV (size) → 80 - 100 fL
2. MCH (Hb) = 27 - 32 pg
3. MCHC (Hb Concenteration) = 33 - 37 g/dL → Ⓝ in MEGALOBLASTIC ANEMIA
4. RDW (Variation in Size) = 11.5 - 14.5 %
5. PCV / Hct ≃ 45 %



Date

IRON METABOLISM



★ MASTER REGULATOR OF IRON : HEPCIDIN
(Source → LIVER)

Cerebellum Coupon code:-NEETPG

T I B C
Total Iron Binding Capacity
↓ ↓ ↓ ↓
Top IDA Bottom CHRONIC DISEASE

$$\text{MENTZER INDEX} = \frac{\text{MCV}}{\text{RBC Count}}$$

< 13 → ThaLESSemia Trait
> 13 → IDA

INTEGRATED APPROACH TO ANEMIA

Date

↓ RBC Mass

- >100 : MACROCYTIC
  - Liver Disease
  - Hypothyroidism
  - Myelodysplasia / MEGALOBLASTIC ANEMIA
  - Cell Maturation Defect
- 80-100 : NORMOCYTIC
  - Renal Disease
  - AOCD
  - Myelofibrosis / Myelophthisis

MCV [<80] → MICROCYTIC

Hb ↓
- ↓ Heme
  - Fe
    - IDA
    - AOCD
  - Porphyrin → SIDEROBLASTIC ANEMIA
- Globin ↓ → THALASSEMIA (α/β)

IDA → Fe intake / Fe loss (Blood Loss)

AOCD → ↓ Fe absorption, ↓ Fe release → ~~FOR~~ → Hepcidin ↑↑

IRON OVERLOAD STATES

I. IDA → MCC : Blood Loss

- ↓ S. Ferritin (↑↑Sp / ↓Sn) (+ APR)
- TSAT = Fe ↓ / TIBC ↑ ⇒ ↓ <18%
- S.Tfr ↑ / log Ferritin ↓ ⇒ >2
- GOLD STANDARD : BMA Cytology (⊖ Fe stores) | PERL'S STAIN

EVALUATION ? → ♂ / POST-MENOPAUSAL ♀
POOR RESPONSE TO Fe SUPPLEMENTS

MALABSORPTION / CANCER (GIT) → r/o by UGIE / Colonoscopy

℞ : ORAL Fe / PARENTERAL Fe / pRBC

ORAL Fe (Before MEALS)
- GLUCONATE (12%)
- FeSO4 (20%)
- FUMARATE (33%)
- LIPOSOMAL Fe (Ferisome)

PARENTERAL Fe
- Fe DEXTROSE
- Fe SUCROSE
- FCM (MC Used)
- Fe ISOMALTOSE (Max. Fe)
- FERRUMOXYTOL → Bright Signals in MRI (1-3 Months)

- * MOST © S/E : GI S/E
- * ↓ ABSORPTION → PPI | H2r⊖ / Phytates / Tannates / Malabsorptive States
- * ↑ ABSORPTION → Vit. C

RESPONSE

CLINICAL (Earliest) <12-24 Hrs
- * 1st : ↓ Subjective Symptoms (Pica / fatigue)
- * Epithelial : ANGULAR CHEILITIS (~4-6 Wks)
- * KOILONYCHIA (~6 Months)

LABS
- * 1st : BM → ERYTHROID HYPERPLASIA (<48 Hrs)
- * ↑ RETIC. Hb (3-4 days)
- * ↑ RETICULOCYTOSIS (~1 WK)
- * ↑ Hb → ↔ RDW
- ↔ BM Fe Stores (~6-12 Months)

II. AOCD : ↑ HEPCIDIN (+ APR)

⊖ FPN
- ↓ Fe absorption
- ↓ Fe release from FERRITIN

- * ↑↑ S. Ferritin (+ APR)
- * TSAT = Fe ↓ / TIBC ↓ ⇒ ↓ >18%
- * S.Tfr ↔ / log Ferritin ↑ ⇒ <2

℞ : ℞ of underlying cause
± IV Fe (if TSAT < 20%)
± ESA (if S. EPO < 500)

III. SIDEROBLASTIC

- * GENETIC : X-linked → ALA Synthase ⊖
- * ACQUIRED : PYRIDOXINE DEFICIENCY (INH / Alcohol)
- * IDIOPATHIC : Pb Poisoning

LABS → ↑↑ S. Ferritin
TSAT = Fe ↑ / TIBC ↔ ⇒ ↑↑
BMA Cytology : RINGED SIDEROBLASTS

℞ : ℞ underlying cause but if IDIOPATHIC → High Dose Vit. B6

MICROCYTIC (<80fL)

Challenge for ORAL IRON for 1 WEEK

RETICULOCYTE COUNT → ↑↑ : IDA → CONTINUE ORAL IRON FOR 6-12 MONTHS

Doesn't Increase → IRON PROFILE

- * All ↓ except TIBC, ↑ S.Tfr, ↓ Protoporphyrins → IDA
- * All ↓ except S. Ferritin, ↑ TLC / CRP / ESR → AOCD
- * All ↑ except TIBC, BMA SIDEROBLASTS → SIDEROBLASTIC ANEMIA

Cerebellum Coupon code:-NEETPG

Cerebellum Coupon code: NEETPG

IV. THALASSEMIA (CHAIN IMBALANCE)

α → DELETIONS (can present BEFORE / AT BIRTH)

- ASYMPTOMATIC + N Hb on ELECTROPHORESIS
  - αα/α- → CARRIERS [Hb / MCV N]
  - αα/-- → MINOR [↓MCV]
- Symptoms @ BIRTH + ABNORMAL Hb on ELECTROPHORESIS
  - α-/-- → INTERMEDIA [↓↓Hb / ↓MCV / Splenomegaly]
  - --/-- → MAJOR → require IU Transfusions
    - if not → DIE IN-UTERO (HYDROPS)
    - [BART's Hb – γ4 Tetramers] → MCC of NON-IMMUNO HYDROPS FETALIS

HbH (β4 Tetramers)
→ Vital Stain (New Methylene Blue)
→ PS → GOLF BALL APPEARANCE

MAJOR : Symptomatic + Transfusion Dependent (Severe Anemia)

INTERMEDIATE : Symptomatic + not Transfusion Dependent (Moderate Anemia)

MINOR : Asymptomatic (Trait) (No / Mild Anemia)

β → FRAME SHIFT MUTATIONS (never presents BEFORE / AT BIRTH)

- β0 → NO β-CHAINS → EXON MUTATION
- β+ → SOME β-CHAINS → INTRON DEFECT (splicing)

	HOMOZYGOUS	HETEROZYGOUS	COMPOUND HETERO.
β0	β0β0 → MAJOR	β0β → (N) MINOR [HETEROZYGOUS β-THALASSEMIA]	β0β+ → INTERMEDIA / MAJOR
β+	β+β+ → INTERMEDIA (MC) → MAJOR / MINOR	β+β	HbS β0 → Sickle defect, β-Thal defect → PS → SICKLE CELLS ± ↓↓MCV → SPLENOMEGALY

- HbA2 > 3.5% ♀
- Asymptomatic
- Hb = 10–12 gm%

★ MAJOR : Pediatrics / Hb < 3 gm% / Repeated BT ⊕ → IRON OVERLOAD → CHF (MC COD) [DCM > RCM] (10–20 Yrs)
HbF ↑ = 30–90%

★ INTERMEDIA : Hb > 7 gm% / Pediatrics ⟶ Adults → BT DEPENDENT
HbF ↑ = 10–30%

Rx : MAJOR → Regular BT (Hypertransfusion Regime) → Hb > 10 gm% ensure (N) growth
(FRESH & LEUCOREDUCED) > 10–15 U pRBC's
START ± IRON CHELATION
- DEFEROXAMINE (iv) → ↑↑ S/E
- DEFERIPRONE (PO) → TID
- DEFERASIROX (PO) → OD

↓ S/E

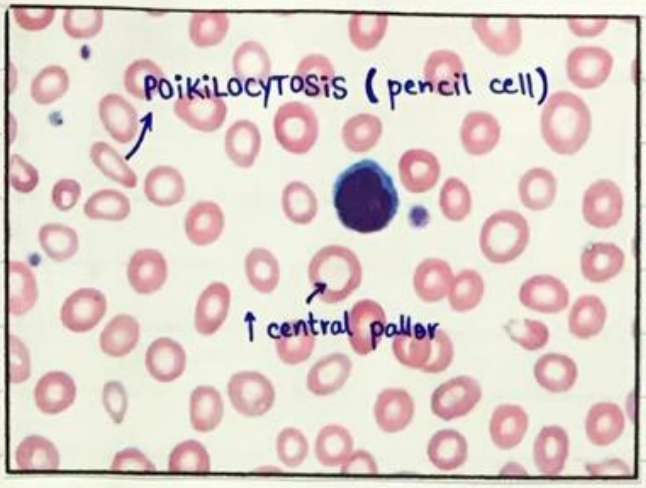
Date

MICROCYTIC HYPOCHROMIC ANEMIA

I. IDA

↓

ANISOCYTOSIS
POIKILOCYTOSIS



MOST DEFINITE | SPECIFIC

↓

BMA | B

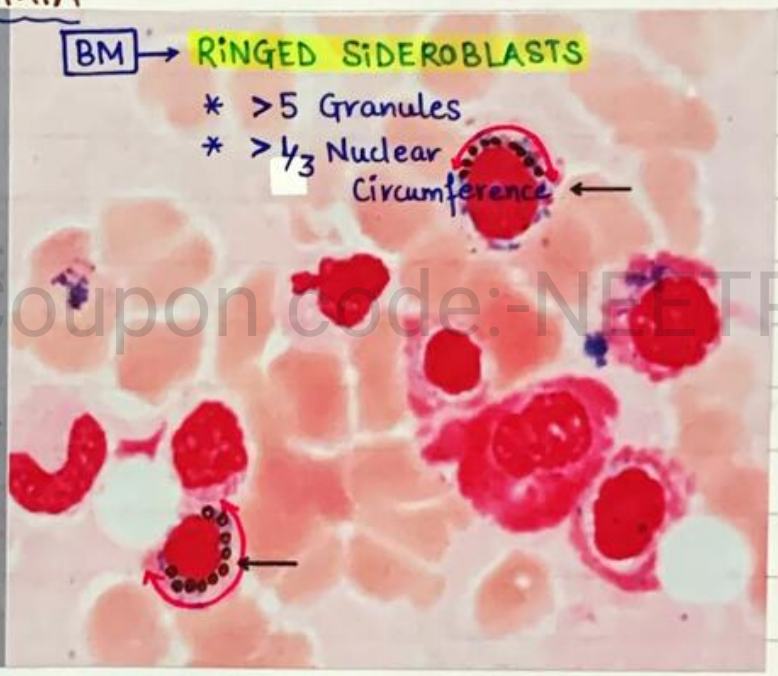
↓

PERLS STAIN for Fe

↓

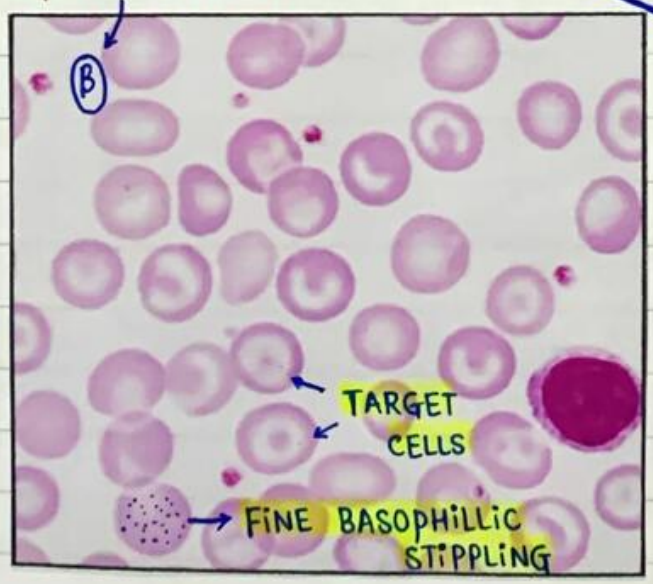
⊖ Fe STORES

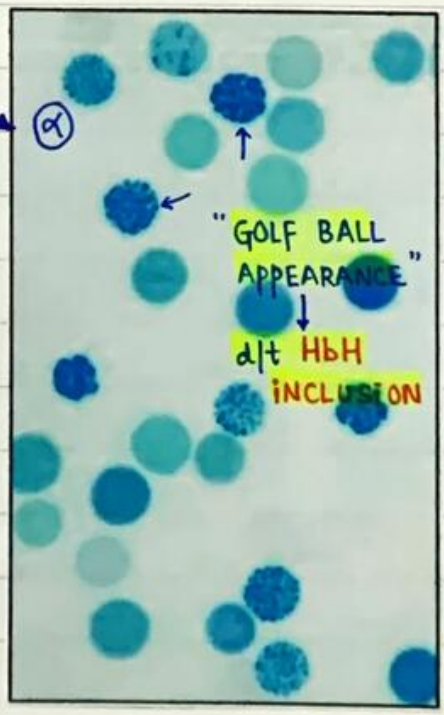
II. SIDEROBLASTIC ANEMIA



Cerebellum Coupon code:-NEETPG

III. THALASSEMIA





Date

[>100] → MACROCYTIC → MEGALOBLASTIC

NO → ALCOHOLIC LIVER DISEASE (↑GGT)
NO → HYPOTHYROIDISM
NO → DRUGS [ZIDOVUDINE / AZT]
NO → MDS / APLASTIC ANEMIA

YES → Vit. B_{12} Deficiency ⟷ FOLATE Deficiency (Jejunum)

Vit. B_{12} Deficiency → Terminal ileum

↓Hb | ↑MCV → >120 ⇒ CONFIRMS Dx

VEGAN

↓ / ⊖ GREENS in DIET

NEUROLOGICAL ⊕
- PN → Axonal
- SACD → PC/CST
- DEMENTIA

HYPERPIGMENTATION ⊕ (KNUCKLES)

HOMOCYSTEINE ↑↑
MMA ↑↑
BEST Invx

HOMOCYSTEINE ↑↑
MMA (N)
BEST Invx → RBC FOLATE

* MDS / APLASTIC ANEMIA
  * OLD / ♂ ¢ CYTOPENIA
    - MC: ANEMIA
  * ↑↑ MCV → 100–110
  - BM | PS → DYSPLASTIC CELLS (≥10%)
  - BLASTS < 20%
  - CYTOGENETIC ABNORMALITY
  * Rx → SUPPORTIVE
  - ISOLATED 5q DELETION (♀ ¢ (N) / ↑ Platelets) → LENALIDOMIDE
  - OTHERS → DNMT ⊖
    * AZACITIDINE
    * DECITABINE

ETIOLOGY
- ↓ ABSORPTION
  - i) MALABSORPTION
    * Sprue (Tropical > Celiac)
    * CD
  - ii) AUTOIMMUNE ⇝ PERNICIOUS ANEMIA
    - watch for other AI Diseases
    - ATROPHIC GLOSSITIS (↑R/O Gastric Adeno Ca)
  - iii) GASTRECTOMY / ILEAL RESECTION
- ↓ INTAKE → VEGANS / ALCOHOLICS
- COMPETITION
  * SIBO
  * D. latum (Fish Tapeworm)

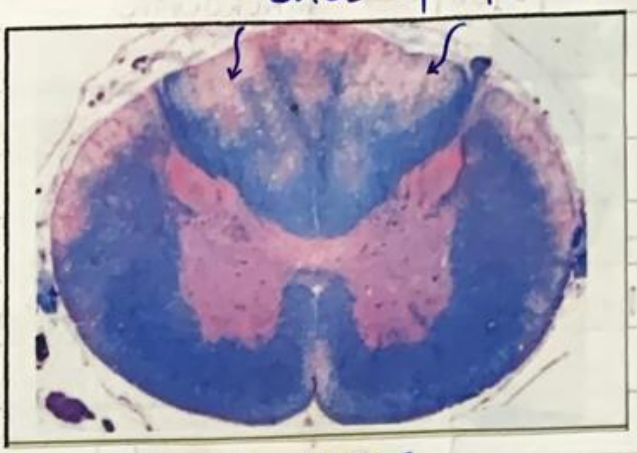
Cerebellum Coupon code: NEETPG

Date

↓

MEGALOBLASTIC ANEMIA

SACD of Spinal Cord



SACD ↓ Vit. B_{12}
↓ HIV/AIDS
↓ Copper Deficiency

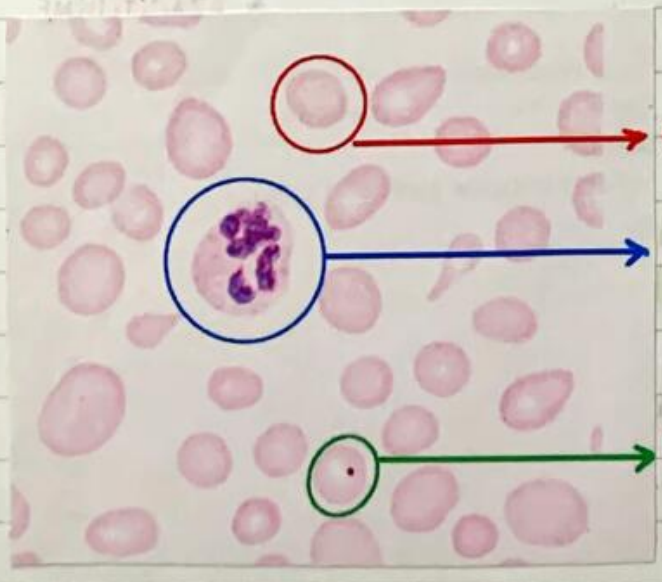
★ FIGLU TEST
↓ FOLIC ACID TEST
URINE → SAMPLE

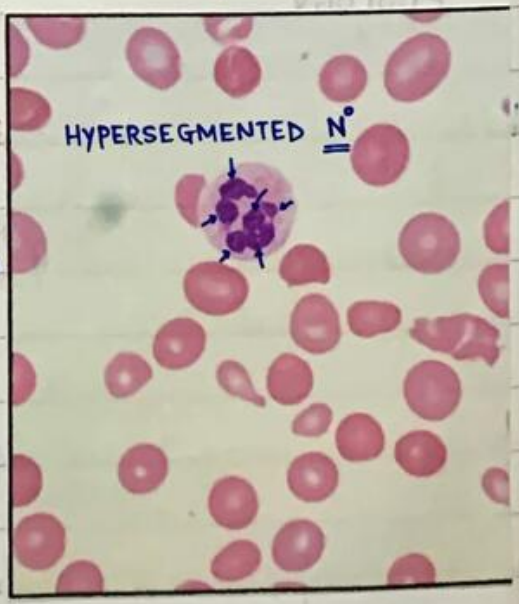
SCHILLING TEST
↳ done to Know CAUSE OF MEGALOBLASTIC ANEMIA

PS Findings of MEGALOBLASTIC ANEMIA

Cerebellum Coupon code:-NEETPG

1. EARLIEST : MACRO - OVALOCYTOSIS
2. MOST SPECIFIC : HYPERSEGMENTED NEUTROPHIL (>5 Nuclear lobes in 5% N° or ≥ 6 lobes in 1 N°)
3. HOWELL - JOLLY BODIES → Sign of DYSERYTHROPOEISIS
   - * NUCLEAR REMNANTS made up of 'DNA'
   - * SINGLE DOT
   - * SEEN IN MEGALOBLASTIC ANEMIA & POST - SPLENECTOMY





MCV

[80 – 100]

NORMOCYTIC → RI

$$\frac{\text{Retic Count} \times \frac{\text{Hct}}{\text{N Hct}}}{\text{Maturity Index}} = \frac{\text{CRC}}{\text{Maturity Index}}$$

Date

RI

<2%

* ACUTE BLOOD LOSS

* APLASTIC ANEMIA

INHERITED

eg. FANCONI ANEMIA

* RADIUS | THUMB Ab N Q
* UROGENITAL Ab N → eg. HORSE SHOE
* ↑ R/O CANCERS → MDS / AML

SCREENING : DEB / MMC

Di-Epoxy Butane Mitomycin – C

CONFIRM : Genetic Testing

>2%

* RECENT BLOOD LOSS
* HEMOLYTIC ANEMIA → ↑LDH

ACQUIRED

1°

* Young Adults
* IDIOPATHIC
* BMA → Hypocellularity → DRY TAP (↑ fat)

2°

INFECTIONS → PARVO B19

DRUGS

DOSE DEPENDENT → CHEMOTHERAPY

DOSE INDEPENDENT

* ABx : SULPHA DRUGS, CHLORAMPHENICOL
* AED
* ATD's (MC S/E : RASH)
* COLCHICINE (MC S/E : Diarrhoea) → AGRANULOCYTOSIS

Rx : <40 yr

if MATCHED SIBLING DONOR → ALLOSTERIC HSCT

if No MSD → IMMUNOSUPPRESSION (Triple)

1. HORSE Anti-Thymocyte Globulin
2. CNI → Cyc | Tac
3. ELTROMBOPAG

Cerebellum Coupon code:-NEETPG

HEMOLYTIC ANEMIAS

Date

I. HS

* AD >>> AR
* RBC Membrane Defect
  - ANKYRIN → MC MUTATION
  - SPECTRIN
  - BAND-3 → Pincer Cells
  - GLYCOPHORIN (MOST ABUNDANT)
* AR-HS if SPECTRIN ↓ (SEVERE)

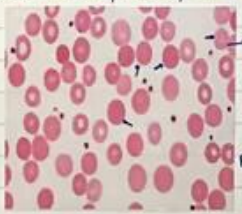
C/F : Jaundice / Splenomegaly
Gall Stones

LABS : Retic. Count ↑
MCV ↓
MCHC ↑

K/C/O HS ± ↓ Retic. Count
→ PARVO B19 INFECTION
→ APLASTIC CRISIS → Rx = rest + plenty of fluids

SCREENING : OSMOTIC FRAGILITY TEST
(OF ↑ → > 0.5% NaCl)

Δ : FLOW CYTOMETRY → EMA TEST



II. G6PD DEFICIENCY

causes OXIDATIVE STRESS
→ DENATURE / DAMAGE RBC
→ HEINZ BODIES / BITE CELLS

* XLR (M >>> F)
* PRECIPITATING FACTORS
  - DRUGS → PRIMAQUINE (MC), DAPSONE
  - FOOD → FAVA BEANS
  - BACTERIAL INFECTIONS
* INTERMITTENT DISORDER
* NO SPLENOMEGALY

NOT STAINED ± ROMANOWSKY
∴ CRYSTAL VIOLET

aka DEGMACYTES
MOST CHARACTERSTIC ON PS...

MISNOMER

III. PNH (IVH) → Rx : ECULIZUMAB

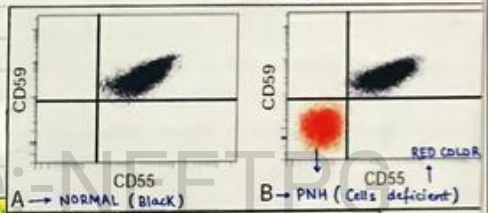
PIGA Gene Defect
→ GPI Anchors ✗ → CD55/DAF ✗, CD59/MIRL ✗ → CRP
CRP = COMPLEMENT REGULATORY PROTEIN → INACTIVATE MAC

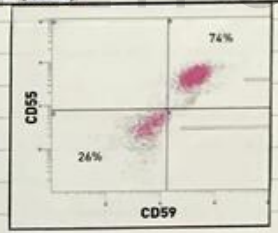
PATHOGENESIS
ACIDIC pH → ⊕ COMPLEMENT PATHWAY → C_{5b-9} (MAC)
CRP ⊗, GPI-AP ⊗
→ RBC / WBC / Plts
→ 1. HEMOLYTIC ANEMIA
→ 2. PANCYTOPENIA
→ 3. THROMBOSIS
→ TRIAD OF PNH
→ HEPATIC VEIN THROMBOSIS
* BUDD CHIARI SYNDROME
* MC SITE / MCC OF DEATH IN PNH

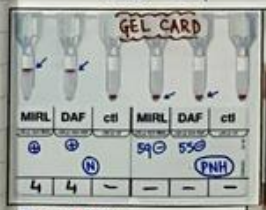
SCREENING : GEL CARD / SUCROSE LYSIS TEST

Δ : FLOW CYTOMETRY / FLAER
CD55 CD59
Test GPI-AP

PNH
- Stem Cell Disorder
- PRE-LEUKEMIC CONDITION → PNH is Moody → Aplastic Anemia, Acute Leukemia, MDS
- TRIAD OF PNH







Top : Positive
Neeche : Negative

Cerebellum Coupon code:NEETPG

HEMOGLOBINOPATHIES

Date

QUALITATIVE

→ SCA

→ β- Chain → 6th Position

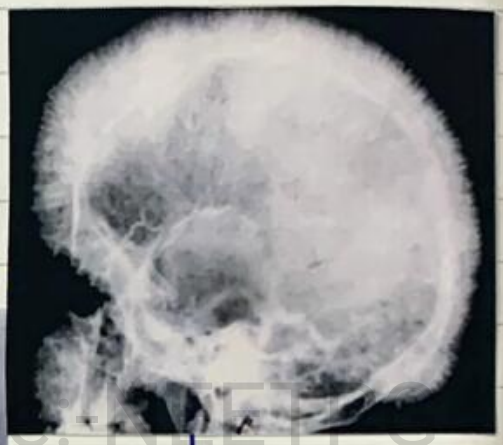
β_6

HbA → GLUTAMIC ACID (GO) — replaced — VALINE (VELCOME) ← HbS

* MOST © MUTATION : POINT MUTATION (MISSENSE)

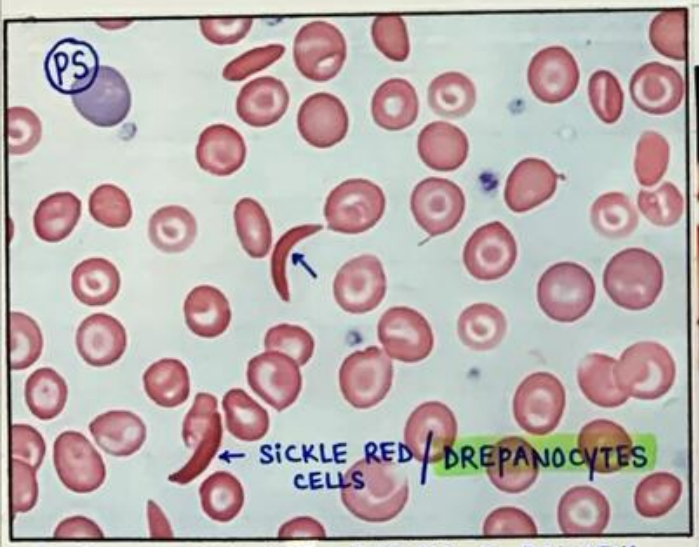
Mn → [SICK ARE MISSED]

C/F
- SKULL → CREW CUT APPEARANCE
- VERTEBRAE → FISH MOUTH VERTEBRA
- SPLEEN → AUTOSPLENECTOMY



→ NOT DIAGNOSTIC

→ Thalassemia
- SCA
- HA



AFRICAN CHILD c ANEMIA c PAINFUL SWELLING OF DIGITS (DACTYLITIS) ABDOMINAL PAIN & CHRONIC FATIGUE

TESTS :

i) SICKLING TEST : 2% Sodium Metabisulphite

ii) Hb ELECTROPHORESIS : H/A/F/S/A

iii) GOLD STANDARD : HPLC

Cerebellum Coupon Code: KNEE15

AIHA

SPLENOMEGALLY

Date

WARM AIHA

@ 37°c , IgG Ab (garam)

Imp. ONE LINERS

Q_1: MC a/w CLL

Q_2: MC a/w which AI Disease : SLE

Q_3: DRUGS : PENICILLIN / α-METHYL DOPA

EVH

* SPLENOMEGALLY

Rx : CS

((if SEVERE) → IVIG | PEX)

± RITUXIMAB

± SPLENECTOMY

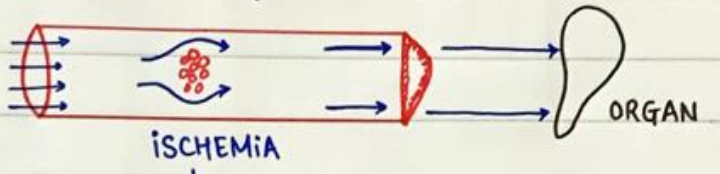
COLD AIHA @ 4°c

COLD AGGLUTININ

IgM → CLUMPS

* MYCOPLASMA
* SYPHILIS

CLUMPS OF RBC



ISCHEMIA

[PALLOR → CYANOSIS → REDNESS]

RAYNAUD'S PHENOMENON

EVH

On P. Smear → CLUMPS

COLD HEMOLYSIN

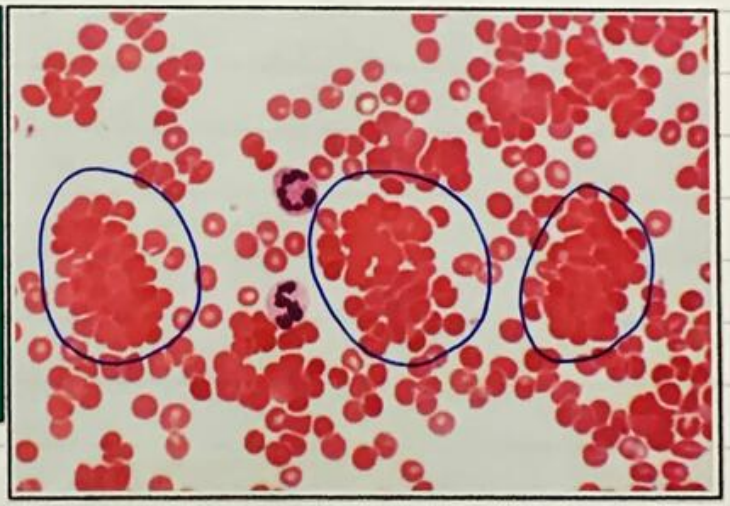
aka PCH

IgG

DONATH LANDSTEINER Ab

a/w VIRAL INFECTIONS

IVH



EVH → HS | SCA | Thal

IVH → PNH / PCH

G6PD

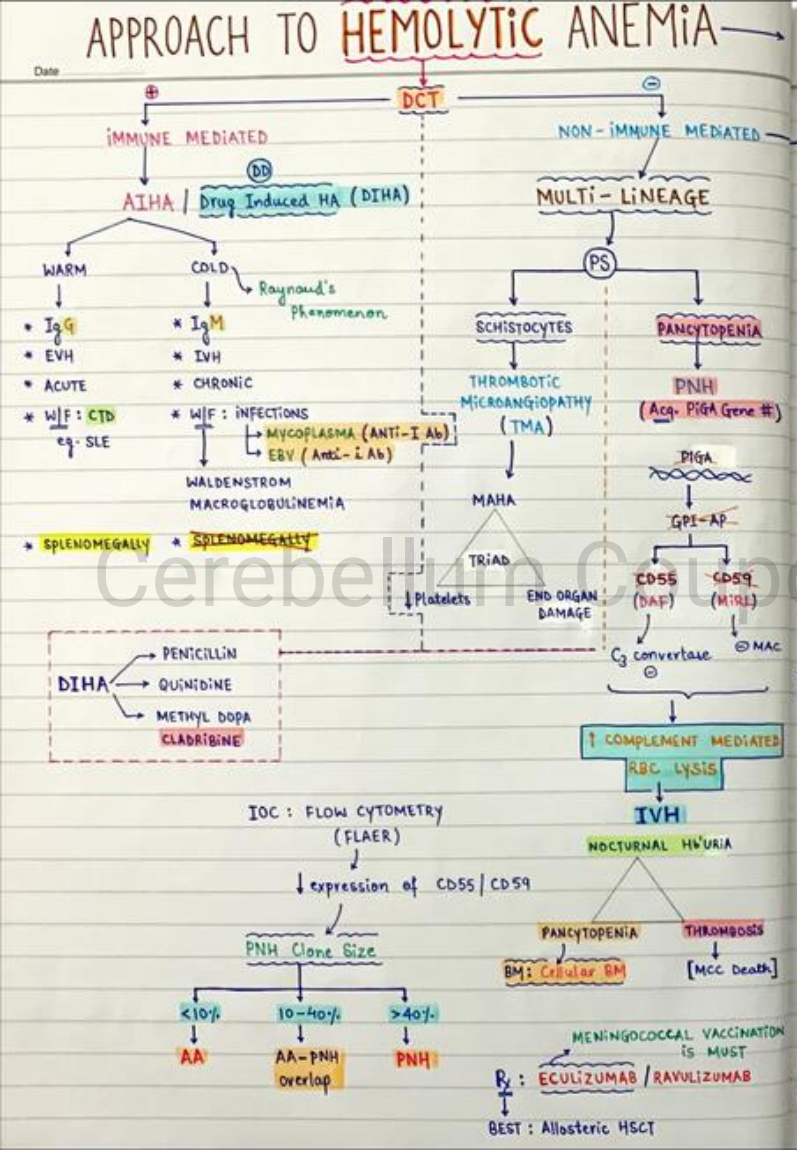
Rx : ~~CS~~

AVOID COLD EXPOSURE ± RITUXIMAB

Cerebellum Coupon code:-NEETPG

APPROACH TO HEMOLYTIC ANEMIA

Date



Cerebellum Coupon code:-NEETPG

iVH → ↑ Bilirubin (MILD) / S. Haptoglobulin ↓↓ / Hb'emia / Hb'uria / HEMOSIDERINURIA

EVH (Spleen) → ↑↑ Bilirubin / S. Haptoglobulin Ⓝ / MOST PATIENTS : SPLENOMEGALY

↑ Gallstones (pigment)

ONLY RBC → PS

- SICKLE CELLS → S.C.D
  - * POINT MUTATION (β_6)
  - CAG → CTG
  - glutamate → valine
  - C/F
  - a) ANEMIA : ↓Hb / Ⓝ MCV
    - ↳ DREPANOCYTES on P.Smear
  - b) VASO-OCCLUSION
    - DACTYLITIS (1^{st} Sx)
      - * CHILDREN
      - * HAND & FOOT
    - SEQUESTERATION CRISIS (Med. Emergency of SCA)
      - * ACUTE CHEST SYNDROME (Pneumonia like)
  - Rx : IVF / AB_x / Exchange Transfusion (ET)
    - * SPLENIC SEQUESTERATION (<4yr) → ↓Hb / MASSIVE SPLENOMEGALY
    - >4yrs → Autosplenectomy : ASPLENIA
  - c) INFECTIONS → OM in SCA d/t "SALMONELLA"
  - d) OTHERS
    - STROKE → may be Asymptomatic → DEMENTIA
    - PRIAPISM
    - * CHRONIC LEG ULCERS
    - * RENAL PAPILLARY NECROSIS [↓ Urinary Conc^n]
    - * PAH
    - * HEMATURIA
- BITE / BLISTER CELLS → G6PD Deficiency
- SPHEROCYTES → HS
- G6PD Deficiency / HS → P.T.O

IOC : Hb ELECTROPHORESIS

SCD			
HbSS			
HbA	0%	SCT	ASYMPTOMATIC
HbF	5-25%	HbAS	R.P.N during
HbS	75-95%	60%	* major surgery
>60% → SICKLING		0%	* high altitude
		40%	* Pregnancy

- * MC Sx : HEMATURIA ↓Uconc
- * Protective for P. Falciparum

Rx : TRANSFUSIONS / HU

↑ HbF levels

- ET
  - * GOAL : ↓HbS (<30%)
  - * Eg. Acute Chest Synd. / Acute Stroke / Major Surgery
- Simple B.T
  - * GOAL : Hb → 9-10gm%
  - * Eg. Anemia Minor Surgery / Aplastic Crisis (Parvo B_{19}) / Splenic Seq. Crisis

* CRIZANLIZUMAB → for VASO-OCCLUSIVE CRISIS

* VOXELOTOR → ⊖ HbS Polymerisation

Date

G6PD DEFICIENCY

↓ NADPH

↓ reduced GLUTATHIONE

↑ oxidative stress

↓

IV HEMOLYSIS

C/F : DRUGS

* PRIMAQUINE
* SULFONAMIDES
* ɣ- URICASE
* IV METHYLENE BLUE
* NITROFURANTOIN for UTI's
* DOXORUBICIN

OTHERS

* DIET → FAVA BEANS
* DKA | INFECTIONS

PS : HEINZ BODY (NEW METHYLENE BLUE)
BITE | BLISTER CELLS (WRIGHT GIEMSA STAIN)

In Acute cases of G6PD Deficiency

↓

G6PD levels can be Normal...

HS

MC ← ANKYRIN (AD > AR) + Family H/O ⊕ & SPHEROCYTES in PS

↓

DCT ⊖

C/F : ↓Hb | Ⓝ MCV | ↑↑ MCHC

↓

Anemia | ↓ Growth (Short Stature) | Splenomegaly | Chronic Jaundice

Chronic Leg Ulcers

Pigmented Gallstone

AIIMS Q → Family H/O CHOLECYSTECTOMY

IOC : FLOW CYTOMETRY (↓ EMA Binding) Q

↓

Eosin-5-Maleimide

SCREENING : OFT (↓Sn | ↓Sp)

AGLT (↑Sn)

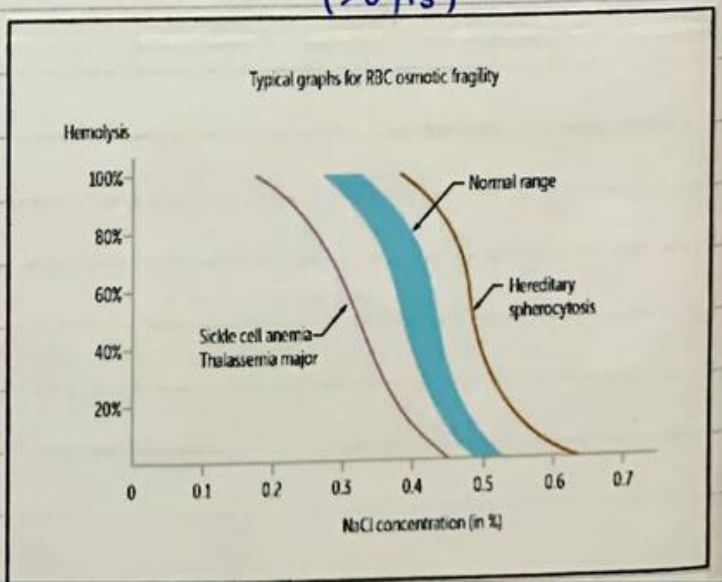
(Acidified Glycerol Lysis Test)

Rx : REGULAR BT's + FOLATE

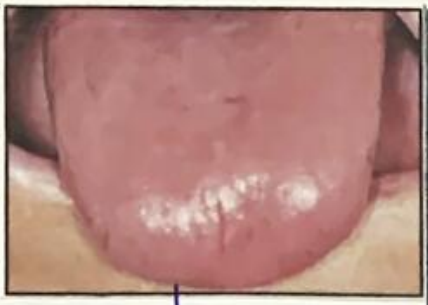
± SPLENECTOMY (in Transfusion Dependent)

↓

(>6 Yrs)



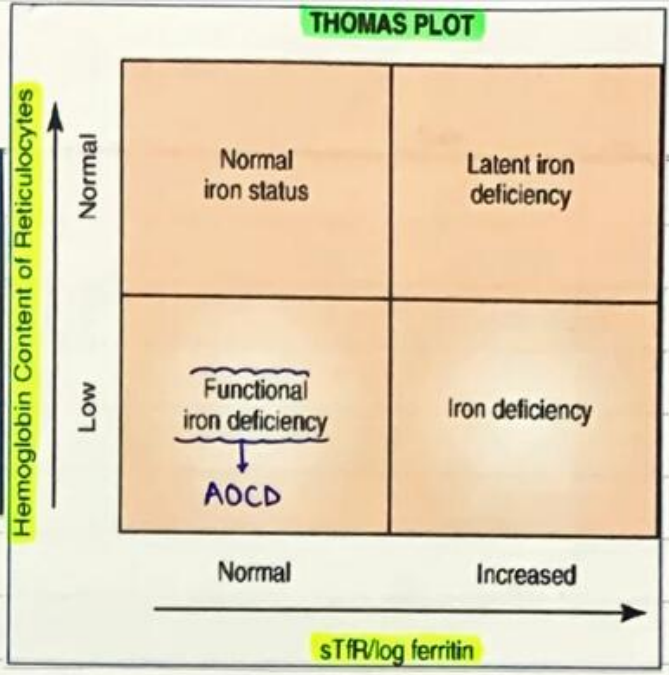
Cerebellum Coupon code: NEETPG

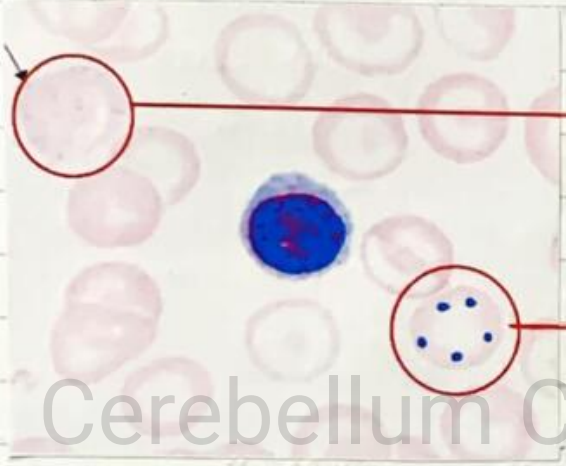


BALDED TONGUE

KOILONYCHIA

IDA





FINE BASOPHILLIC STIPPLING → ⊖ in IDA

MEGALOBLASTIC ANEMIA >> THALASSEMIA

COARSE BASOPHILLIC STIPPLING

↓

LEAD POISONING

Cerebellum Coupon code:-NEETPG

NESTROF Test

Naked Eye
Single Tube
RBC OFT

↓

0.3% NaCl

THALASSEMIA → incomplete hemolysis

NORMAL → complete hemolysis

{RINGED SIDEROBLASTS} → also ⊕ in MDS...

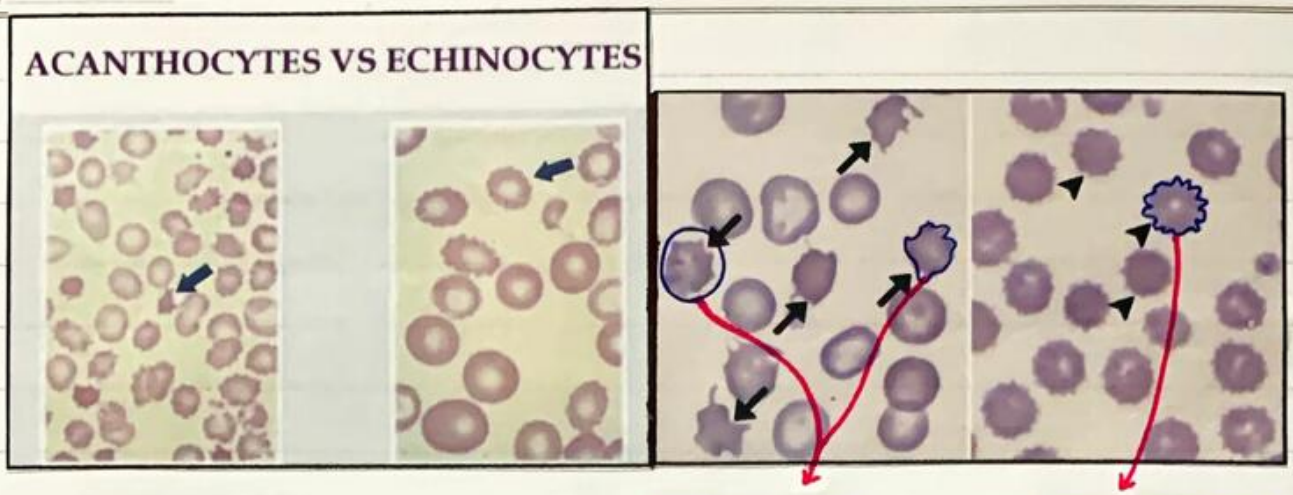
BURTON's / BURTONIAL LINE

↓

CHRONIC LEAD POISONING

Date

ACANTHOCYTES VS ECHINOCYTES



ACANTHOCYTES

SPUR CELLS / SPIKE

↓

Seen in

↓

A-betalipoproteinemia

ECHINOCYTES [B]

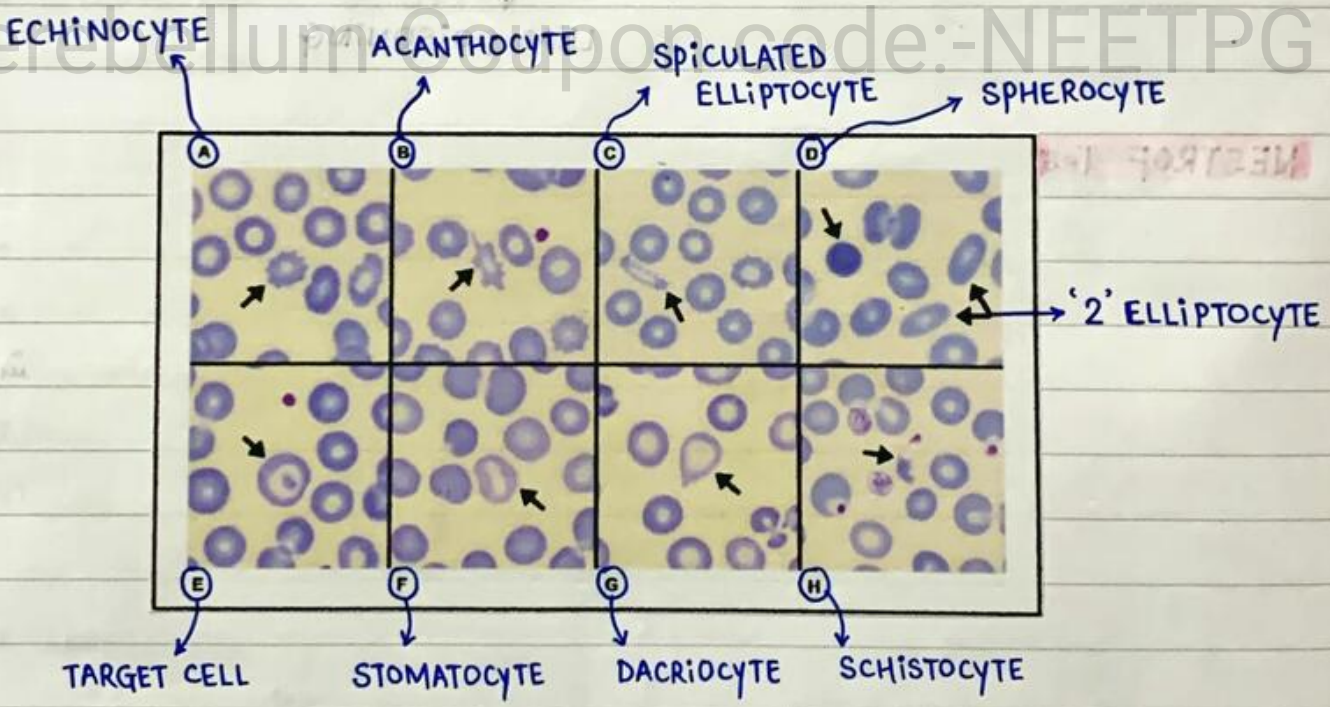
BURR CELLS

↓

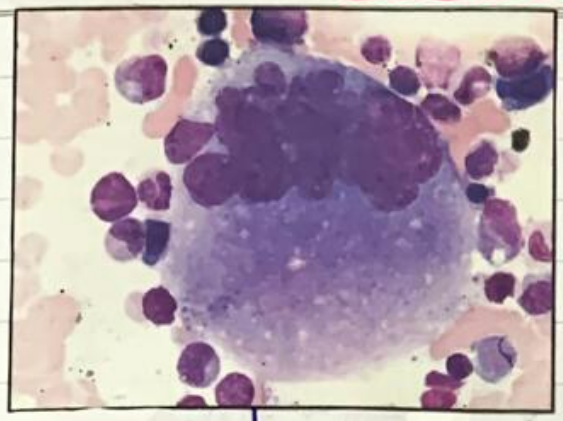
Seen in

* Burns
* Uremia
* EDTA changes

Cerebellum Coupon code:-NEETPG

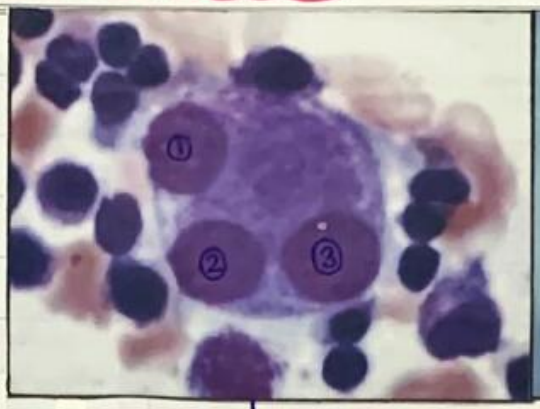


MEGAKARYOCYTE



MEGAKARYOCYTE BM ASPIRATION

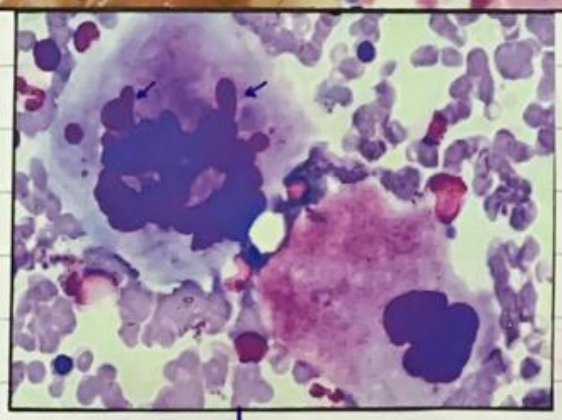
NORMAL : Mother of Platelets



PAWN BALL MEGAKARYOCYTE

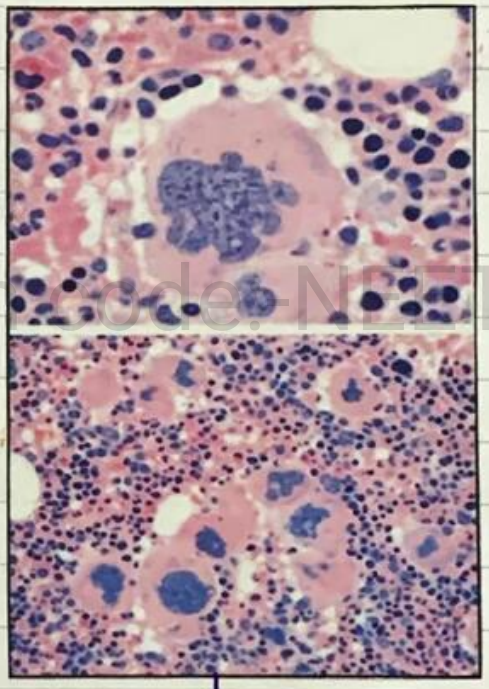
Nuclei are seen as THREE BALLS

seen in MDS ...



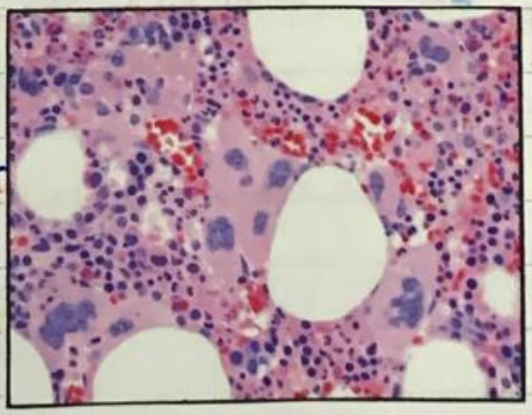
STAGHORN MEGAKARYOCYTE

Nuclei in the shape of Staghorn, seen in ET (Essential Thrombocythemia)



CLOUD LIKE MEGAKARYOCYTE

Nuclei is like cloud → seen in 1° MYELOFIBROSIS



DWARF MEGAKARYOCYTE

Nuclei is very very small; seen in CML

Cerebellum Coupon code- NEETPG

PLATELETS & COAGULATION

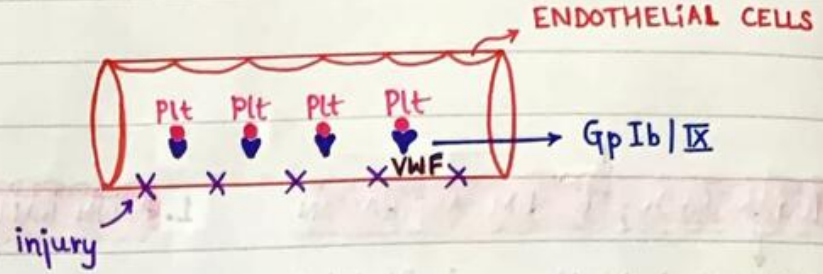
Date

* PRECURSOR : MEGAKARYOCYTES
* (N) Plt. Count : 1.5 – 4.5 Lac / mm^3

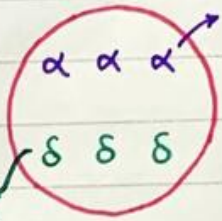
CLOT FORMATION

1 PLATELET ADHESION

ONE → GpIb/IX – VWF



2 PLATELET SECRETION



F

ALPHA GRANULES

* P-Selectin
* PF_4 (Plt. Factor 4)
* PDGF
* Factor 5/8
* Fibrinogen

DELTA / DENSE GRANULES

DENSE

ADENOSINE (ADP/ATP)

SEROTONIN

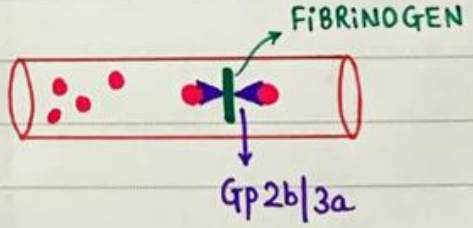
S. Ca^{2+}

EPINEPHRINE

Cerebellum Coupon code: NEETPG

3 PLATELET AGGREGATION

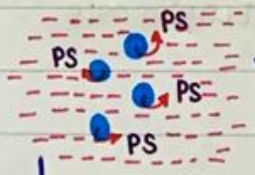
Gp 2b/3a — FIBRINOGEN



PLATELET ADHESION → PLATELET SECRETION → PLATELET AGGREGATION

↓

1° HEMOSTATIC PLUG



Platelets show FLIPPING OF PHOSPHATIDYL SERINE

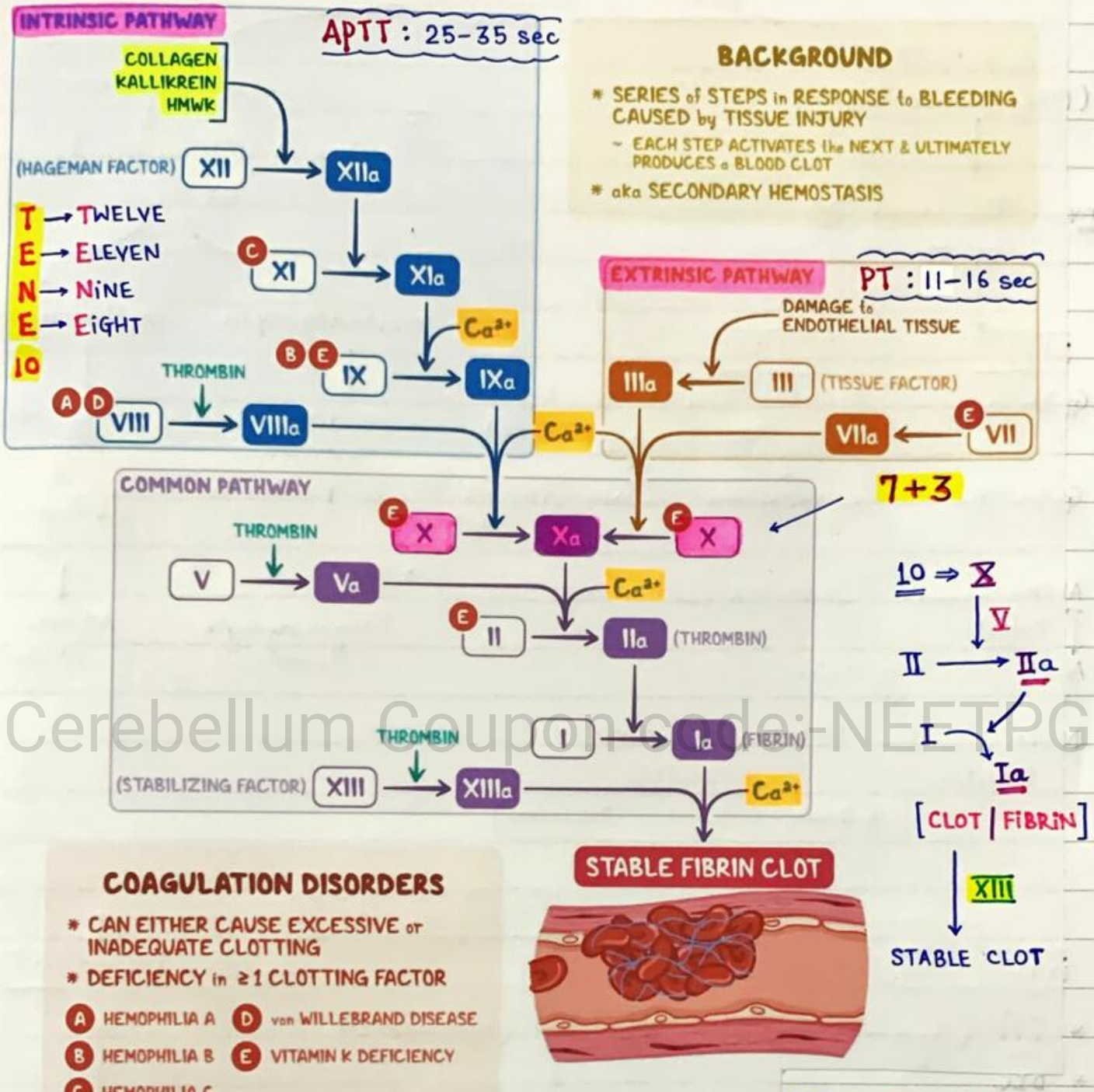
⊖ve charge → ACTIVATE CLOTTING SYSTEM

↓

2° PLUG (CLOT)

PATHWAY

Date



BACKGROUND

* SERIES of STEPS in RESPONSE to BLEEDING CAUSED by TISSUE INJURY
  - EACH STEP ACTIVATES the NEXT & ULTIMATELY PRODUCES a BLOOD CLOT
* aka SECONDARY HEMOSTASIS

COAGULATION DISORDERS

* CAN EITHER CAUSE EXCESSIVE or INADEQUATE CLOTTING
* DEFICIENCY in ≥1 CLOTTING FACTOR

A HEMOPHILIA A
B HEMOPHILIA B
C HEMOPHILIA C
D von WILLEBRAND DISEASE
E VITAMIN K DEFICIENCY

XIII : LAKI LORAND FACTOR
↓
FIBRINOGEN STABILIZING FACTOR

Cerebellum Coupon code:-NEETPG

* BLEEDING TIME : measure of platelets (2-9 mint.)
* CLOTTING TIME ~ 8-15 mints.
* THROMBIN TIME : measure of Factor I (FIBRINOGEN)
* UREA CLOT SOLUBILITY TEST → Factor XIII (LOKI LORAND FACTOR)

PLATELETS & COAGULATION PATHWAY

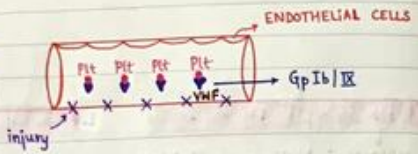
Date

* PRECURSOR : MEGAKARYOCYTES
* (N) Plt. Count : 1.5–4.5 Lac / mm^3

CLOT FORMATION

1 PLATELET ADHESION

ONE → GpIb/IX – VWF



2 PLATELET SECRETION

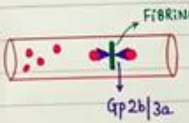
DENSE
- ADENOSINE (ADP/ATP)
- SEROTONIN
- S. Ca^{2+}
- EPINEPHRINE

→ DELTA/DENSE GRANULES

ALPHA GRANULES
- * P-Selectin
- * PF_4 (Plt. Factor 4)
- * PDGF
- * Factor 5/8
- * Fibrinogen

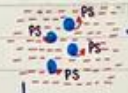
3 PLATELET AGGREGATION

Gp 2b/3a — FIBRINOGEN



PLATELET ADHESION → PLATELET SECRETION → PLATELET AGGREGATION

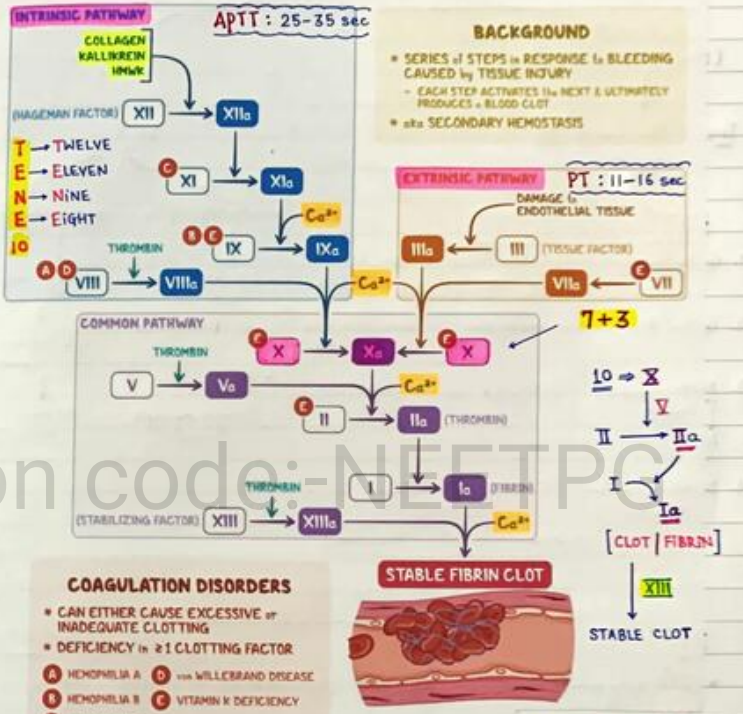
→ 1° HEMOSTATIC PLUG



Platelets show FLIPPING OF PHOSPHATIDYL SERINE

⊖ve charge → ACTIVATE CLOTTING SYSTEM

→ 2° PLUG (CLOT)



BACKGROUND

- * SERIES of STEPS in RESPONSE to BLEEDING CAUSED by TISSUE INJURY
  - EACH STEP ACTIVATES the NEXT & ULTIMATELY PRODUCES a BLOOD CLOT
- * aka SECONDARY HEMOSTASIS

COAGULATION DISORDERS

- * CAN EITHER CAUSE EXCESSIVE or INADEQUATE CLOTTING
- * DEFICIENCY in ≥1 CLOTTING FACTOR
  - A HEMOPHILIA A
  - B HEMOPHILIA B
  - C HEMOPHILIA C
  - D von WILLEBRAND DISEASE
  - C VITAMIN K DEFICIENCY

XIII : LAKI LORAND FACTOR

→ FIBRINOGEN STABILIZING FACTOR

* BLEEDING TIME : measure of platelets (2-9 mint.)
* CLOTTING TIME ~ 8–15 mints.
* THROMBIN TIME : measure of Factor I (FIBRINOGEN)
* UREA CLOT SOLUBILITY TEST → Factor XIII (LOKI LORAND FACTOR)

Cerebellum Coupon code:-NEETPG

Date

PLATELET DISORDERS

QUALITATIVE
(Plts. Function Defect)

Invx : Plt. count Ⓝ
BT ↑↑

DEFECT	DISEASE
1. GpIb/IX	BERNARD SOULIER DISEASE (AR) → Big/बड़ा Platelet
2. GpIIb/IIIa	GLANZMANN THROMBASTHENIA (AR)
3. ALPHA Granule Defect → Aged ⟿ Gray Hairs	GRAY PLATELET SYNDROME
4. DELTA Granule Defect	HERMANSKY PUDLAK SYNDROME (Oculo-cutaneous Albinism)

QUANTITATIVE
(aka THROMBOCYTOPENIA)

Invx : Plt. count ↓↓
BT ↑↑

MEGAKARYOCYTIC
* ↑ Platelet destruction in P. Blood...
* BM → ↑ Megakaryocyte

AMEGAKARYOCYTIC
BM : abⓃ → ↓ Platelet count → APLASTIC ANEMIA

Cerebellum Coupon code:-NEETPG

MEGAKARYOCYTIC THROMBOCYTOPENIA

IMMUNOGENIC
* ITP (IgG → MOST ©)
* DIC

NON-IMMUNOGENIC

TTP (THROMBOTIC)
↓ADAM TS-13 → Abⓝ platelet aggregation → * Platelet destruction * Thrombi → PLATELET COUNT ↓

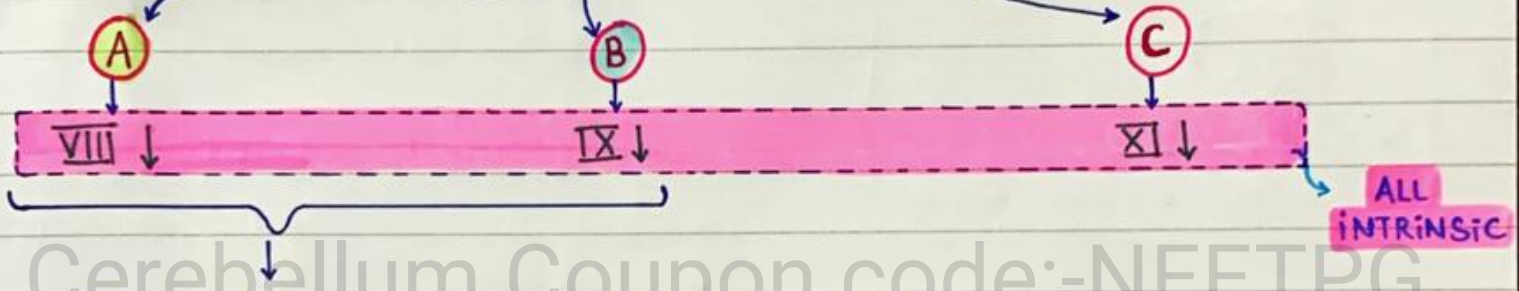
HUS
* Ⓝ ADAM TS-13
* Pediatric Age
* GIT infection → E. Coli (O157 : H7) → VEROTOXIN (Shiga like) → ↓ PLATELET COUNT

Date

ITP → Ab against GpIb/IX or GpIIb/IIIa

ACUTE ITP	FEATURE	CHRONIC ITP
CHILD	AGE	ADULT
⊕	H/O VIRAL INFECTION	⊖
<50,000	PLATELET COUNT	↓↓↓↓↓
<6 Months	RESOLUTION	>6 Months
SELF LIMITING	Rx	STEROIDS

HEMOPHILIA



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* XLR → M >> F
* AR
* MOST © PRESENTATION : HEMARTHROSIS
* ASYMPTOMATIC
* Invx : ISOLATED aPTT ↑

IOC to distinguish all HEMOPHILIAS → FACTOR ASSAY

Rx : CRYOPRECIPITATE Rx : FFP

VON-WILLEBRAND DISEASE

N VWF has '2' functions

- PLATELET ADHESION (↓ GpIb/IX) → defect : BT ↑
- FACTOR VIII STABILISATION (Most Important function of vWF) → defect : aPTT ↑

↑BT PT N aPTT ↑↑

INTEGRATED APPROACH TO THROM

Date

↓ PLATELETS

CLUMPS on PS

NO

YES

ABNORMAL PS ?

NO

ISOLATED ↓ PLATELETS

[NO SYSTEMIC MANIFESTATION]

CULPRITS

NO CULPRITS

2° ITP

AUTOIMMUNE [SLE / APS]

INFECTIONS [HIV / HCV]

DITP

* GOLD / HEPARIN
* VANCOMYCIN / PROCAINAMIDE

1° ITP

[Δ OF EXCLUSION]

* ♀ Young / Middle Aged
* NO SPECIFIC TEST !
* NO SPLENOMEGALLY

Rx

EARLY (<3 Months)

* CS
* if very severe → IVIG ± Plt ± TXA

LATE (>3 Months)

* TPO r ⊕ → P/O
* TPO Mimetic → (S/C)
* RITUXIMAB
* NEW DRUGS →
* TOC : SPLENECTOMY

Cerebellum Coupon code:-NEETPG

HIT

Ab: PF_4 - Heparin complex

TYPE-1

* NON-IMMUNE
* ↓ Plt (>1 Lac)
* NO THROMBO-EMBOLIC MANIFESTATIONS
* Day : 1-4
* CONTINUE HEPARIN

TYPE-2 [HITTS]

* IMMUNE → [HIT Ab] → Non-Sp
* ↓↓ Plt (<1 Lac) → MODERATE ≃ 60,000
* TE RISK → HIGH
* Day : 5-10
* DISCONTINUE HEPARIN for LIFELONG...

'4T' SCORE → TIMING / SEVERITY OF THROMBOCYTOPENIA / THROMBOSIS / OTHER

Rx : STOP HEPARIN (+ WARFARIN) → reverse ē Vit. K

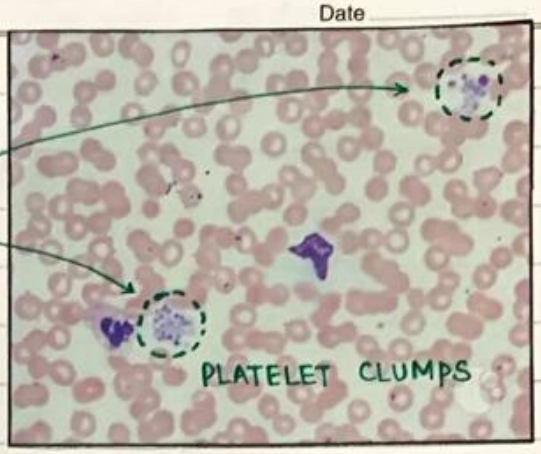
Alternative Anticoagulants →

* ARGATROBAN (iv)
* FONDAPARINUX (SC)
* DOAC

→ Warfarin can be RESTARTED once PLATELETS >1.5 Lac

BOCYTOPENIA (↓ PLATELETS)

Date



YES → PSEUDO - THROMBOCYTOPENIA (EDTA induced)

↓

REPEAT c HEPARIN / Na^+ CITRATE

WHAT ABNORMALITY

→ SCHISTOCYTES : TMA → {DD → DIC}

D-Dimer & PT / PTT

→ SPHEROCYTES : EVANS SYNDROME

↓

(AIHA + ITP)

↓

DCT & SLE Serology

→ LEUKOERYTHROBLASTIC PICTURE

↓

SOME NON-HEMATOPOEITIC MATERIAL

↓

REPLACING BM...

↓

* 1° Myelofibrosis
* Granuloma / S.O.T / Lymphoma

↓

BMA / B_X

DACROCYTES (TEAR DROP CELLS)

TRIAD + HSM

NUCLEATED RBC's

IMMATURE WBC's

P/O : ELTROMBOPAG
AVATROMBOPAG

→ ROMIPLOSTIM

→ FOSTAMATINIB (SYK ⊖)
[Splenic Tyrosine Kinase ⊖]
RILZABRUTINIB (BTK ⊖)

Cerebellum Coupon code: NEETPG

PANCYTOPENIA

MEGALOBLASTIC

↓

* B_{12} deficiency
* Folate deficiency

↓

MMA + Homocysteine ± RBC folate

DYSPLASTIC

↓

MDS / LEUKEMIA

↓

BMA_X

FEATURELESS

↓

AA / PNH

↓

BMA_X / FC_X

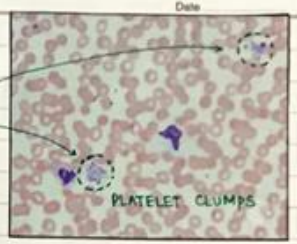
INTEGRATED APPROACH TO THROMBOCYTOPENIA (↓ PLATELETS)

Date

↓ PLATELETS

CLUMPS on PS — NO → ABNORMAL PS ? — NO → ISOLATED ↓ PLATELETS [NO SYSTEMIC MANIFESTATION]

CLUMPS on PS — YES → PSEUDO-THROMBOCYTOPENIA (EDTA induced) → REPEAT ± HEPARIN / Na^+ CITRATE



ABNORMAL PS ? — YES → WHAT ABNORMALITY

ISOLATED ↓ PLATELETS [NO SYSTEMIC MANIFESTATION]

CULPRITS → 2° ITP
- AUTOIMMUNE [SLE / APS]
- INFECTIONS [HIV / HCV]

NO CULPRITS → 1° ITP [Δ OF EXCLUSION]
- ♀ Young / Middle Aged
- NO SPECIFIC TEST !
- NO SPLENOMEGALLY

DITP
- GOLD / HEPARIN
- VANCOMYCIN / PROCAINAMIDE

HIT

TYPE-1
- NON-IMMUNE
- ↓ Plt (>1 Lac)
- NO THROMBO-EMBOLIC MANIFESTATIONS
- Day: 1-4
- CONTINUE HEPARIN

TYPE-2 [HITTS] — Ab: PF_4 - Heparin complex
- IMMUNE → [HIT Ab] → Non-Sp
- ↓↓ Plt (<1 Lac) → ≃ 60,000 MODERATE
- TE RISK → HIGH
- Day: 5-10
- DISCONTINUE HEPARIN for LIFELONG...

'4T' SCORE → TIMING / SEVERITY OF THROMBOCYTOPENIA / THROMBOSIS / OTHER

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Alternative Anticoagulants:
- ARGATROBAN (iv)
- FONDAPARINUX (SC)
- DOAC

→ Warfarin can be RESTARTED once PLATELETS >1.5 Lac

Rx

EARLY (<3 Months) ← Rx → LATE (>3 Months)
- CS
- if very severe → IVIG ± Plt ± TXA
- TPO r ⊖ → P/O : ELTROMBOPAG, AVATROMBOPAG
- TPO Mimetic (S/C) → ROMIPLOSTIM
- RITUXIMAB
- NEW DRUGS → FOSTAMATINIB (SYK ⊖) [Splenic Tyrosine Kinase ⊖]; RILZABRUTINIB (BTK ⊖)
- TOC : SPLENECTOMY

WHAT ABNORMALITY

- SCHISTOCYTES : TMA → {DD → DIC} (D-Dimer ↑, PT / PTT ↵)
- SPHEROCYTES : EVANS SYNDROME (AIHA + ITP) → DCT ± SLE Serology
- LEUKOERYTHROBLASTIC PICTURE
  - DACROCYTES (TEAR DROP CELLS)
  - SOME NON-HEMATOPOETIC MATERIAL → REPLACING BM... (TRIAD + HSM: NUCLEATED RBC's, IMMATURE WBC's)
    - I° Myelofibrosis
    - Granuloma / S.O.T / Lymphoma
    - → BMA / Bx

PANCYTOPENIA

MEGALOBLASTIC
- B_{12} deficiency
- Folate deficiency
- → MMA + Homocysteine ± RBC folate

DYSPLASTIC → MDS / LEUKEMIA → BMA_x

FEATURELESS → AA / PNH → BMA_x / FC_x

Cerebellum Coupon Code: NEETPG

OTHER BLEEDING DISORDERS

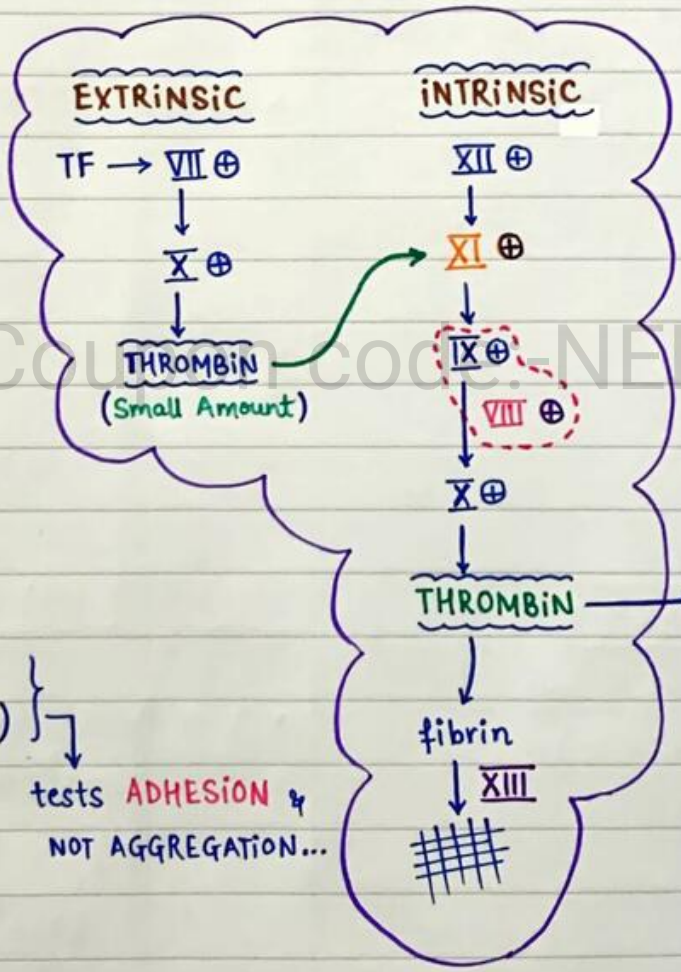
Date

VWD → MC INHERITED BLEEDING DISORDER

85%

- **TYPE 1** (AD) → PARTIAL QUANTITATIVE DEFECT → SUPERFICIAL BLEEDING → DENTAL, ENT
- **TYPE 2** → QUALITATIVE DEFECT → [2A / 2B / 2C / 2N] (2A, 2B, 2C: AD; 2N: AR) → HEMOPHILIA like
- **TYPE 3** (AR) → Rare → COMPLETE QUANTITATIVE DEFICIENCY → HEMOPHILIA like

♀: MENORRHAGIA



- * CBC: (N)
- * PT/INR: (N)
- * aPTT: Mild ↑
- * BT ↑↑ (NOT DONE NOWADAYS)
- * CT (N) / Mild ↑
- * PFA-100 → ↑ CLOSURE TIMES (Plt. Funct^{n} Assay)
- * RIPA (RISTOCETIN induced Platelet Aggregation) → ↓↓ / absent → tests ADHESION & NOT AGGREGATION...

℞: DESMOPRESSIN (DDAVP)

Q: Very High aPTT but patient is ASYMPTOMATIC & NOT BLEEDING at all

↓

FACTOR XII DEFICIENCY

Q: BLEEDING ⊕ but (N) aPTT

↓

FACTOR XIII DEFICIENCY

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Date

BERNARD - SOULIER SYNDROME

↓ (AR)

GpIb / IX #

(ADHESION)

↓

* ↓ Platelets
* बड़ा (Bada) Plts.
* IOC : FCx

↓

↓ / ⊖ RiPA

Ⓝ

GLANZMANN THROMBASTHENIA

↓ (AR)

Gp IIb / IIIa #

(AGGREGATION)

↓

* Ⓝ PS
* IOC : FCx

RiPA : Ⓝ

ADH / Epi / Collagen / AA

(test for Aggregation)

↓

↓↓ / absent

GRAY PLATELET SYNDROME

↓

NBEAL-2

↓

⊖ ALPHA granules

↓

* ↓ Platelets
* GIANT PLATELETS
* IOC : EM

↓

↑↑ PDGF

↓

↑ BM fibrosis

↓

HSM

HERMANSKY PUDLAK SYNDROME

↓

MULTIPLE GENES

↓

⊖ DELTA granules

↓

* Ⓝ PS
* IOC : EM

↓

* Pulmonary Fibrosis
* Oculo - Cutaneous Albinism

Cerebellum Coupon code:-NEETPG

COAGULATION DISORDERS

Date

Painful

Hemophilia Arthropathy

MOST © INHERITED COAGULATION DISORDER

↓

HEMOPHILIA (XLR) → ♂

A (80%) → VIII ↓

B (20%) → IX ↓

d/t INTRON 22 INVERSION

C/F : HEMARTHROSIS / IM Hematomas
LARGE ECCHYMOSIS (limb spanning)

Inv$_x$: CBC, PT/INR, BT } Ⓝ

CT ↑↑

aPTT ↑↑ → 50:50 Mix ⇝ aPTT CORRECTED

↓

Next : Factor VIII levels ↓↓

CerebellumCoupon code:-NEETPG

Rx : ACUTE BLEEDING → ↑F VIII concenterate

	Major	Minor
	Major Sx / ICH	Hemarthrosis, Minor Surgery
LEVELS :	80-100%	40-50%
VIII →	50 U/Kg	25 U/Kg
IX →	100 U/Kg	50 U/Kg

NEW : EMICIZUMAB (Bispecific) → work like F-VIII

Date

SOME PATIENTS → develop Ab's against F-VIII : ACQUIRED F-VIII INHIBITORS

ACUTE BLEED

* rF VIIa → TOC

* FEIBA
  { F VIIa / IX / X / II }
  ↓
  { Vit. K dependent + Ca^{2+} dependent }
  ↓
  Vit. K
  ↳ γ- carboxylation of CLOTTING FACTOR
  by adding Carboxylase group...
  ↓
  ↑ affinity of Ca^{2+} towards Clotting factors

CHRONIC BLEED

* IMMUNO - SUPPRESSION
* IMMUNE - TOLERANCE

Cerebellum Coupon code:-NEETPG

F-XIII DEFICIENCY

DELAYED ← Bleeding ⊕
PTT Ⓝ

UMBILICAL CORD STUMP BLEED H/O
@ BIRTH...

UREA CLOT LYSIS TEST
(5 Mole UREA)
↓
Now a days we use MONO-CHLORO ACETIC ACID (MCA)