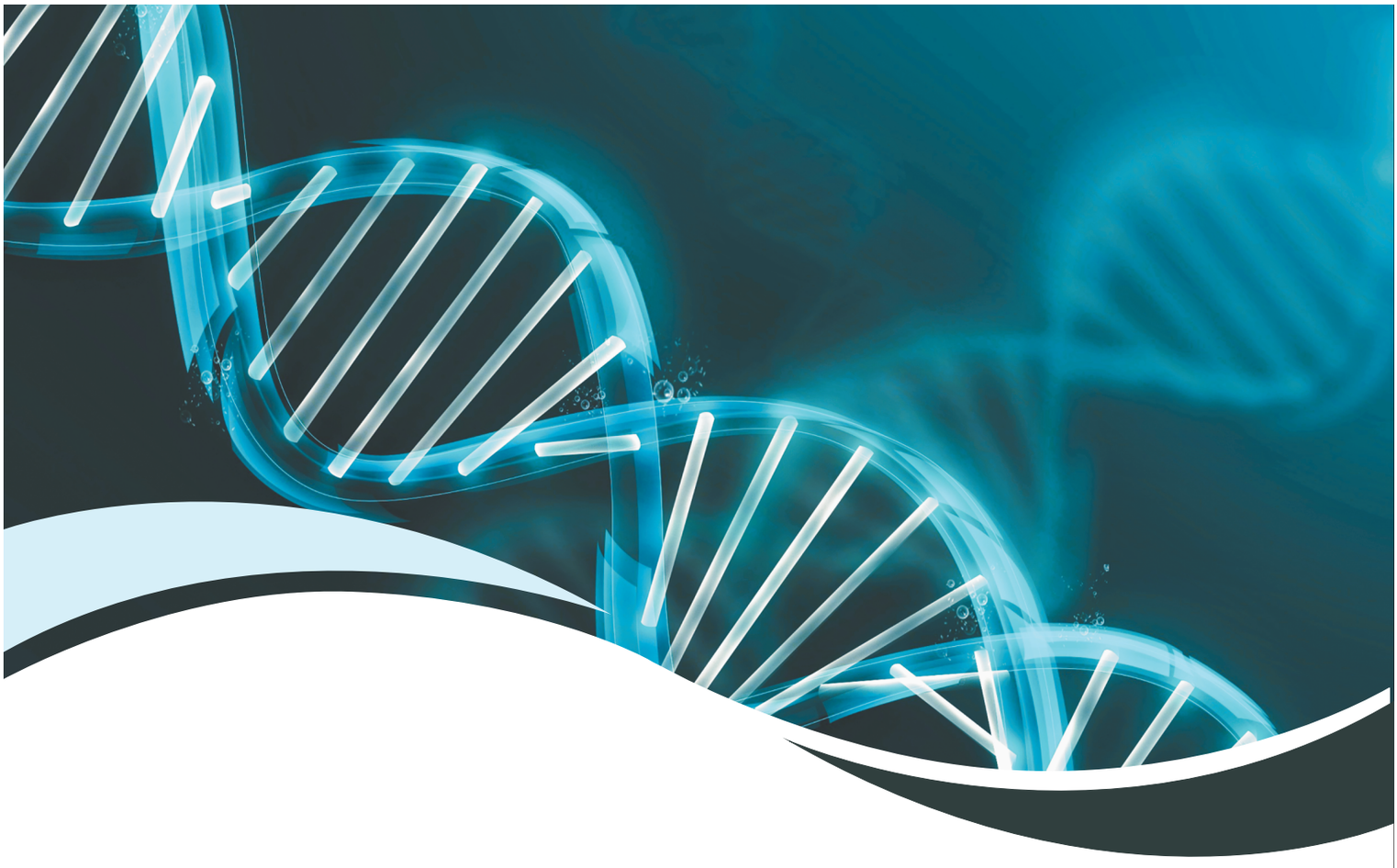
OneShot 4.0

Biochemistry

DBMCI · 2026

DBMCI one



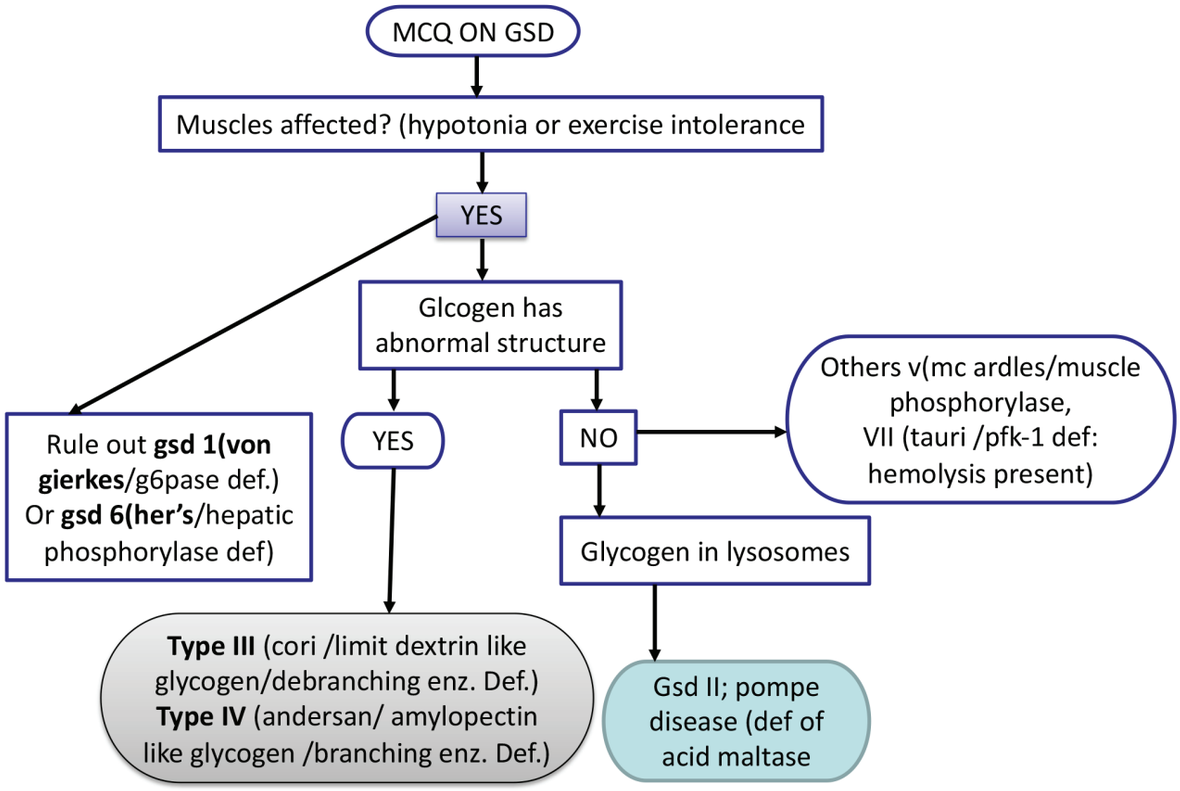
BIOCHEMISTRY

Success doesn't come
from what you do occasionally;
it comes from what you do
consistently. Every small effort
you make today is a step closer
to your dreams. Stay focused,
stay determined, and
remember that while the
journey may be tough,
the reward will be worth it.

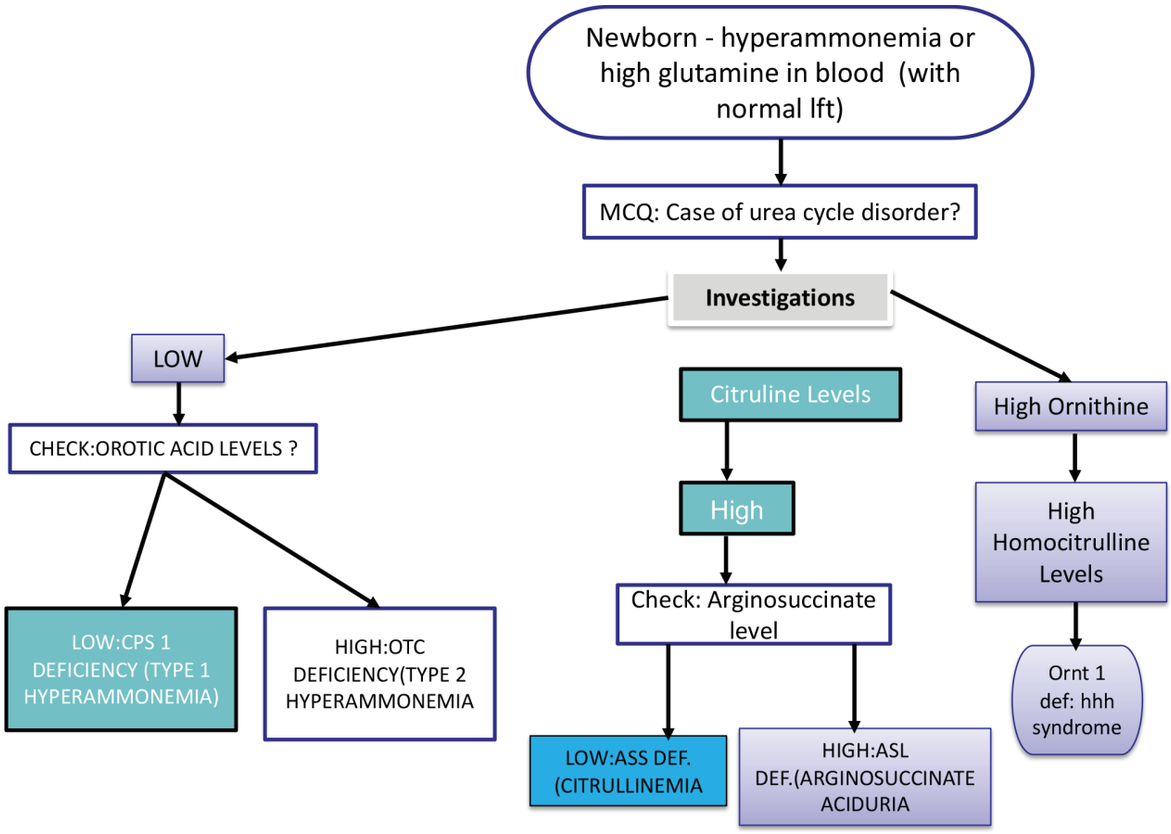
- Dr. Amit Jain

HOW TO APPROACH MCQ ON CLINICAL DISORDERS

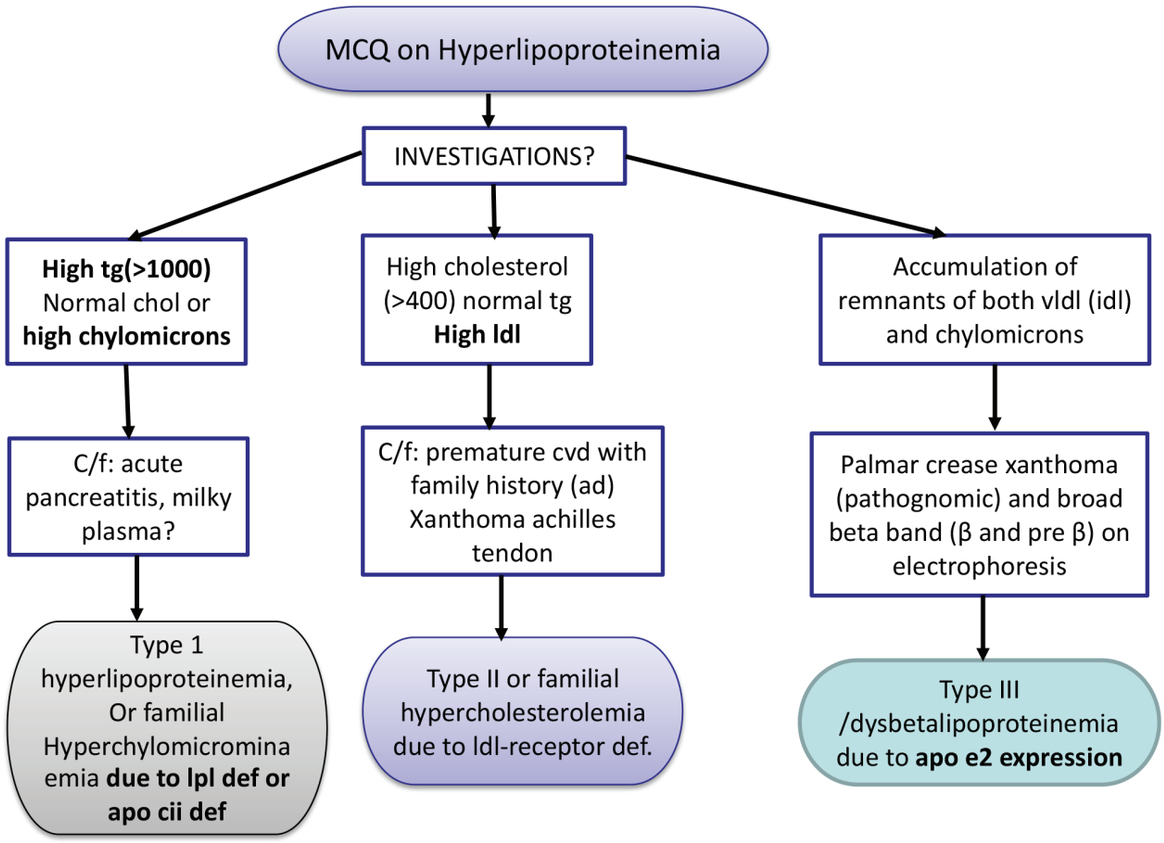
Glycogen storage disorders



Urea cycle disorders



Hyperlipoproteinemia



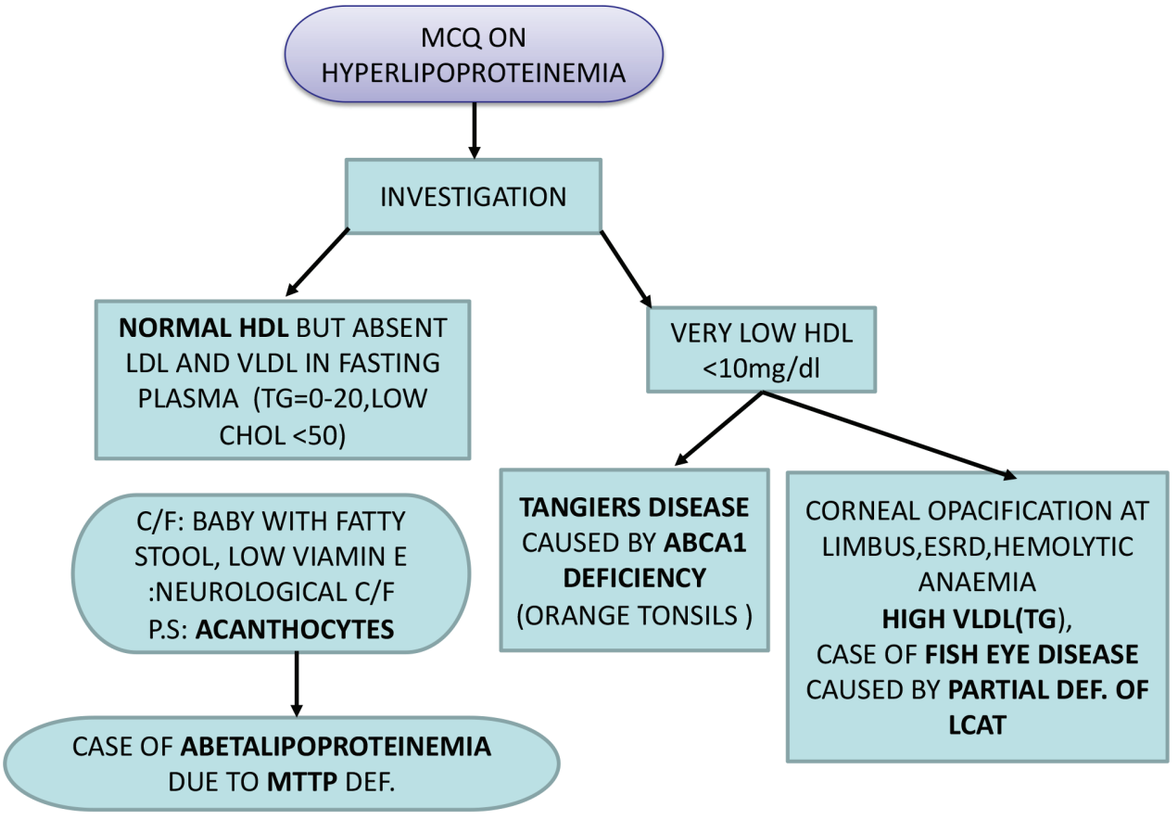
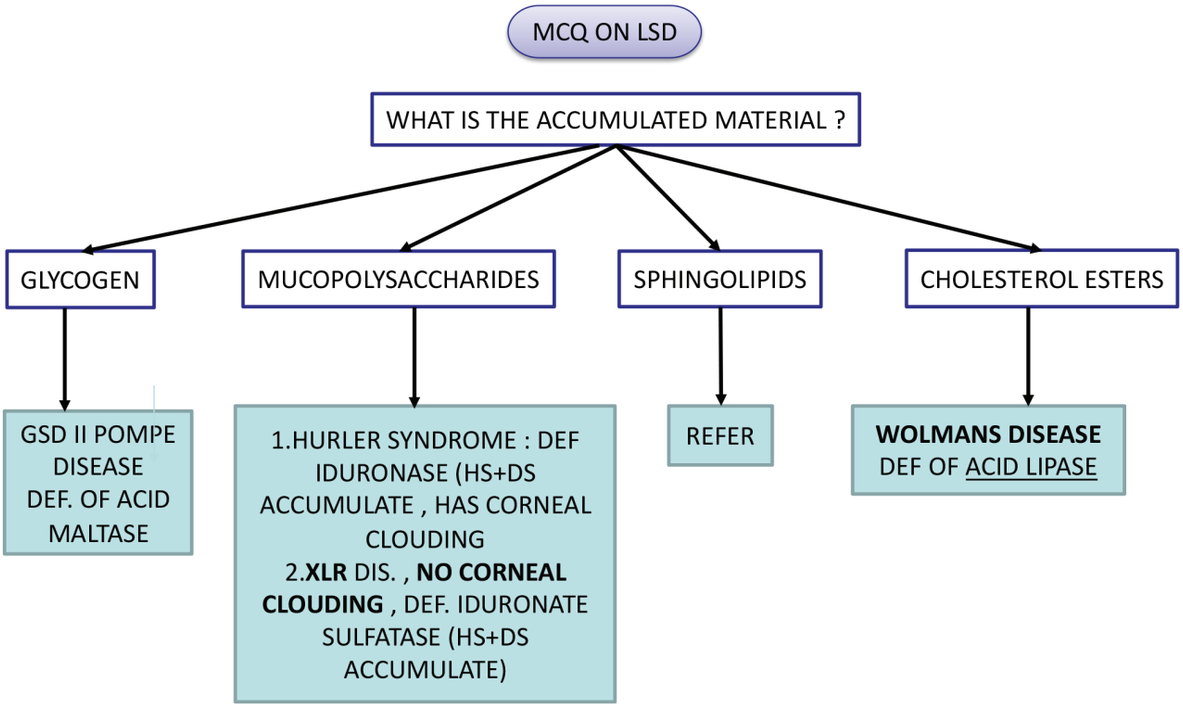
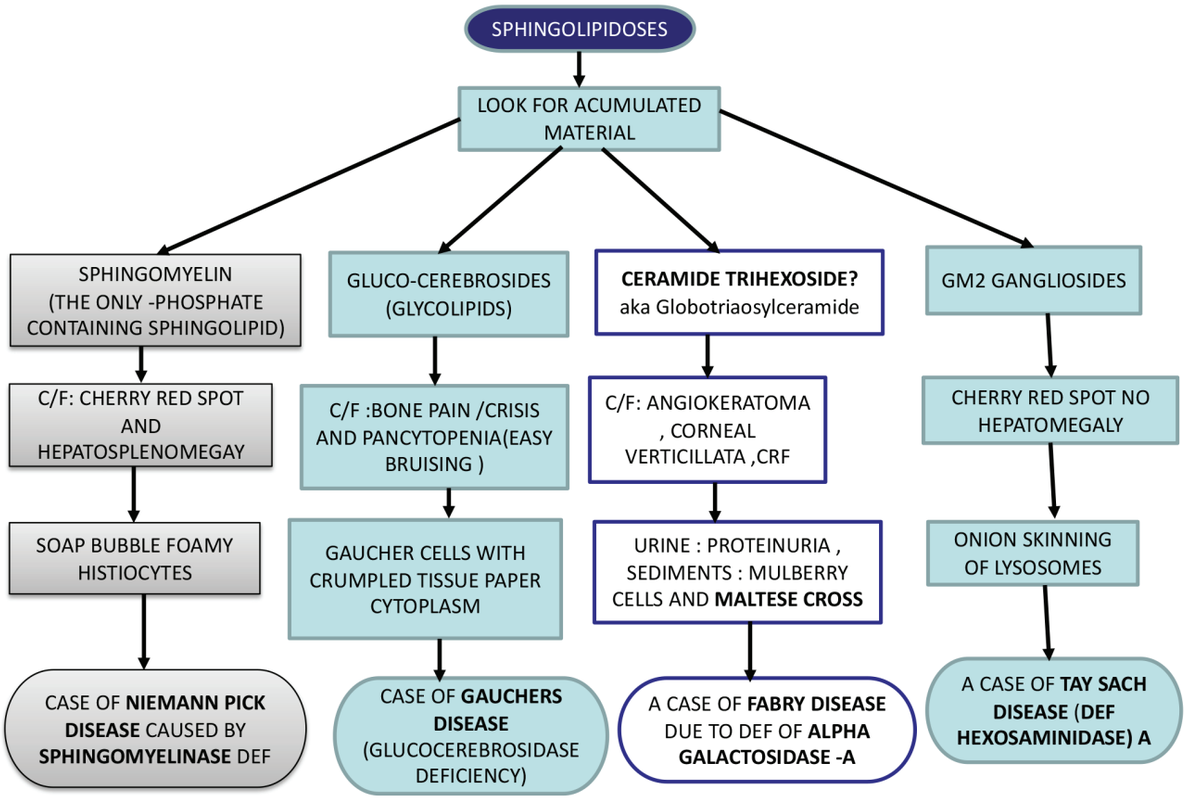
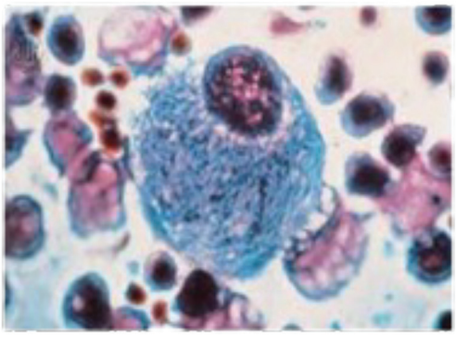

MCQ ON HYPERLIPOPROTEINEMIA
INVESTIGATION
NORMAL HDL BUT ABSENT LDL AND VLDL IN FASTING PLASMA (TG=0-20,LOW CHOL <50)
VERY LOW HDL <10mg/dl
C/F: BABY WITH FATTY STOOL, LOW VIAMIN E :NEUROLOGICAL C/F
P.S: ACANTHOCYTES
TANGIERS DISEASE CAUSED BY ABCA1 DEFICIENCY (ORANGE TONSILS)
CORNEAL OPACIFICATION AT LIMBUS,ESRD,HEMOLYTIC ANAEMIA
HIGH VLDL(TG),
CASE OF FISH EYE DISEASE CAUSED BY PARTIAL DEF. OF LCAT
CASE OF ABETALIPOPROTEINEMIA DUE TO MTTP DEF.

Figure1: Bilateral Achilles Tendon Xanthomas

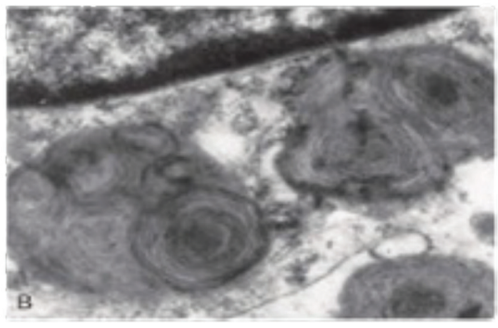

MCQ ON LSD
WHAT IS THE ACCUMULATED MATERIAL ?
GLYCOGEN
MUCOPOLYSACCHARIDES
SPHINGOLIPIDS
CHOLESTEROL ESTERS
GSD II POMPE DISEASE
DEF. OF ACID MALTASE
1.HURLER SYNDROME : DEF IDURONASE (HS+DS ACCUMULATE , HAS CORNEAL CLOUDING
2.XLR DIS. , NO CORNEAL CLOUDING , DEF. IDURONATE SULFATASE (HS+DS ACCUMULATE)
REFER
WOLMANS DISEASE
DEF OF ACID LIPASE


SPHINGOLIPIDOSES
LOOK FOR ACUMULATED MATERIAL
SPHINGOMYELIN (THE ONLY -PHOSPHATE CONTAINING SPHINGOLIPID)
C/F: CHERRY RED SPOT AND HEPATOSPLENOMEGAY
SOAP BUBBLE FOAMY HISTIOCYTES
CASE OF NIEMANN PICK DISEASE CAUSED BY SPHINGOMYELINASE DEF
GLUCO-CEREBROSIDES (GLYCOLIPIDS)
C/F :BONE PAIN /CRISIS AND PANCYTOPENIA(EASY BRUISING)
GAUCHER CELLS WITH CRUMPLED TISSUE PAPER CYTOPLASM
CASE OF GAUCHERS DISEASE (GLUCOCEREBROSIDASE DEFICIENCY)
CERAMIDE TRIHEXOSIDE? aka Globotriaosylceramide
C/F: ANGIOKERATOMA , CORNEAL VERTICILLATA ,CRF
URINE : PROTEINURIA , SEDIMENTS : MULBERRY CELLS AND MALTESE CROSS
A CASE OF FABRY DISEASE DUE TO DEF OF ALPHA GALACTOSIDASE -A
GM2 GANGLIOSIDES
CHERRY RED SPOT NO HEPATOMEGALY
ONION SKINNING OF LYSOSOMES
A CASE OF TAY SACH DISEASE (DEF HEXOSAMINIDASE) A

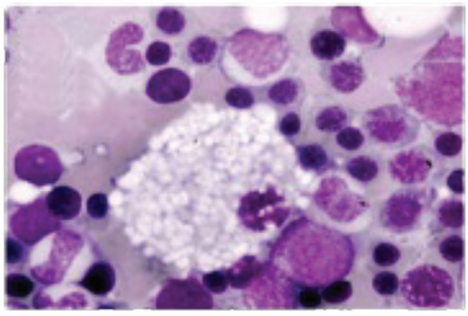
Gauchers disease : *wrinkled or crumpled tissue paper appearance of histiocytes*



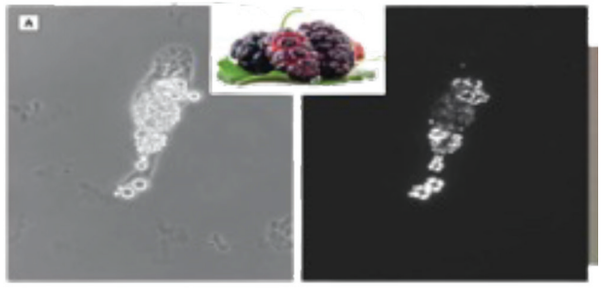
Tay sach disease: *lysosomes with onion skin*

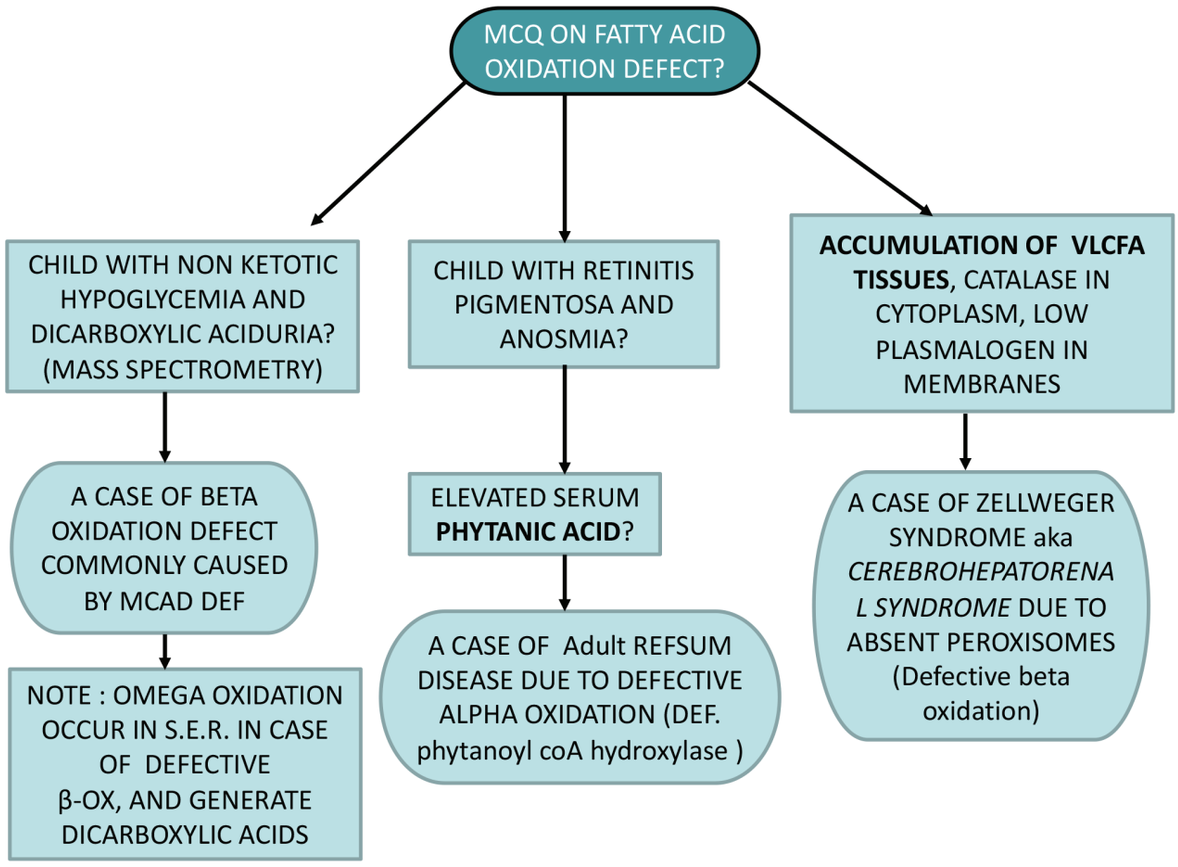


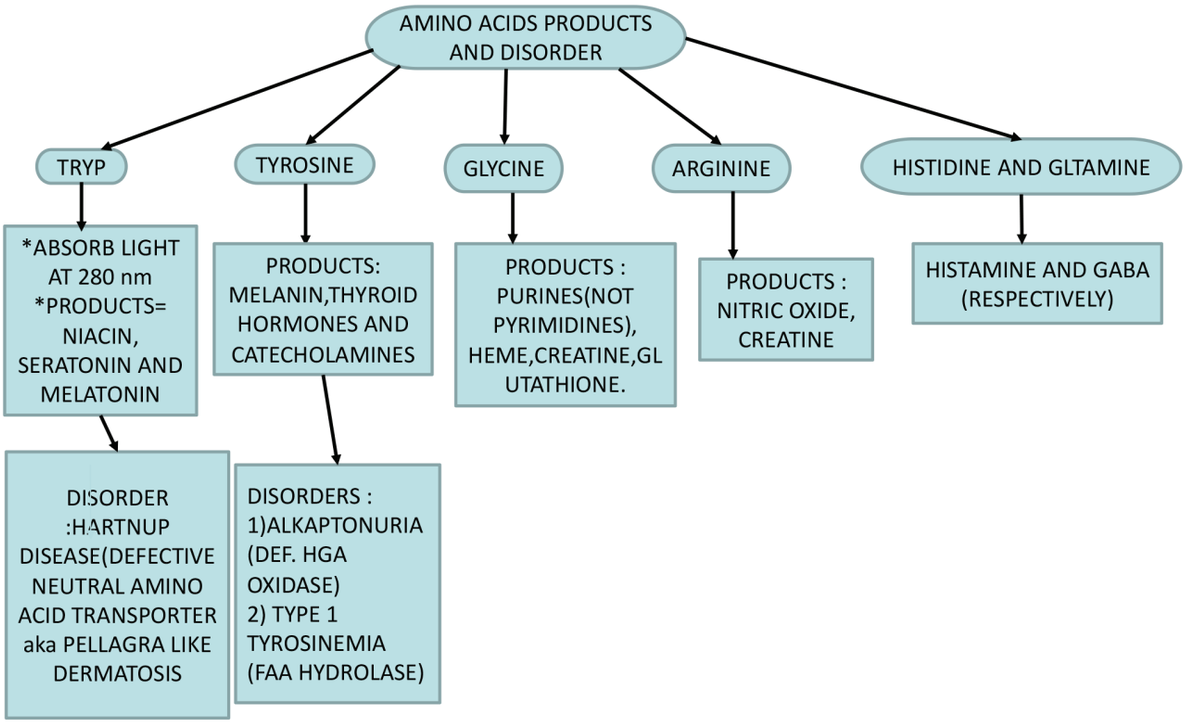
Niemann pick disease : *Foamy histiocytes/ foam cell with bubbly appearance*



Fabry disease: *maltese cross and mulberry cell in urine: a tell tale sign*




MCQ ON FATTY ACID OXIDATION DEFECT?
CHILD WITH NON KETOTIC HYPOGLYCEMIA AND DICARBOXYLIC ACIDURIA? (MASS SPECTROMETRY)
A CASE OF BETA OXIDATION DEFECT COMMONLY CAUSED BY MCAD DEF
NOTE : OMEGA OXIDATION OCCUR IN S.E.R. IN CASE OF DEFECTIVE β-OX, AND GENERATE DICARBOXYLIC ACIDS
CHILD WITH RETINITIS PIGMENTOSA AND ANOSMIA?
ELEVATED SERUM **PHYTANIC ACID**?
A CASE OF Adult REFSUM DISEASE DUE TO DEFECTIVE ALPHA OXIDATION (DEF. phytanoyl coA hydroxylase)
ACCUMULATION OF VLCFA TISSUES, CATALASE IN CYTOPLASM, LOW PLASMALOGEN IN MEMBRANES
A CASE OF ZELLWEGER SYNDROME aka *CEREBROHEPATORENAL SYNDROME* DUE TO ABSENT PEROXISOMES (Defective beta oxidation)


AMINO ACIDS PRODUCTS AND DISORDER
TRYP
TYROSINE
GLYCINE
ARGININE
HISTIDINE AND GLTAMINE
*ABSORB LIGHT AT 280 nm *PRODUCTS= NIACIN, SERATONIN AND MELATONIN
PRODUCTS: MELANIN,THYROID HORMONES AND CATECHOLAMINES
PRODUCTS : PURINES(NOT PYRIMIDINES), HEME,CREATINE,GLUTATHIONE.
PRODUCTS : NITRIC OXIDE, CREATINE
HISTAMINE AND GABA (RESPECTIVELY)
DISORDER :HARTNUP DISEASE(DEFECTIVE NEUTRAL AMINO ACID TRANSPORTER aka PELLAGRA LIKE DERMATOSIS
DISORDERS : 1)ALKAPTONURIA (DEF. HGA OXIDASE) 2) TYPE 1 TYROSINEMIA (FAA HYDROLASE)

Name the Porphyria presenting with ?

- *Acute symptoms but no sun sensitivity =*
- *Acute C/F and sun sensitivity =*
- *Erythrodontia and severe sun sensitivity =*

Quick formula for numericals based Q (enzymes /total cholesterol)

- Michaelis menton equation $V = \frac{Vmax \ X \ S}{Km + S}$
- Enzyme efficiency = $\frac{Kcat}{Km}$
- Friedwalds formula : LDL = Total chol – (HDL + = $\frac{TG}{5}$)

Metabolic Inhibitors as MCQ

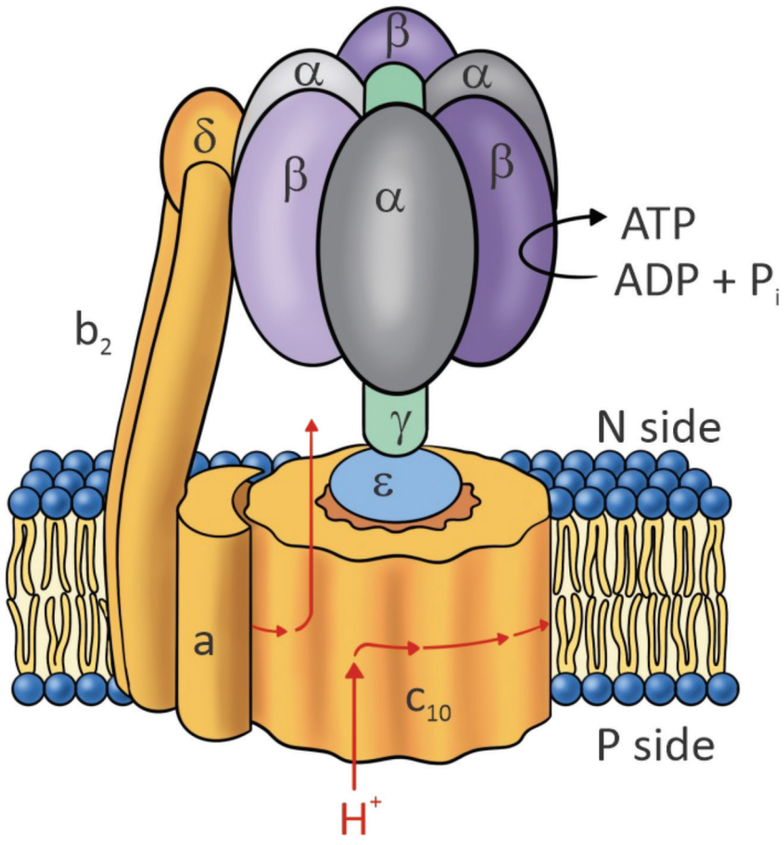
	Inhibitor	Enzyme
1	Iodoacetate	G3PD (glycolysis)
2	Fluoride	Enolase (glycolysis)
3	Arsenic	PDH complex, alpha KGDH (TCA) Uncoupler of SLP by PGKinase (glycolysis)
4	Lead	ALA dehydratase, ferrochelatase (heme synthesis)
5	Flouroacetate	Aconitase (TCA)
6	Malonate and carboxin	Succinase dehydrogenase (TCA / complex II ETC)
7	Ricin	28S rRNA (peptidyl transferase)
8	Alpha amnatin	RNA polymerase II
9	ETC complexes I	Rotenone
10	II	Malonate, carboxin
11	III	Dimeric paralol (BAL), Antimyc in
12	IV	CN, CO, H2S, azide
13	ATP ADP TRANSCLOC ASE	Atractyloside
14	UNCOUPLERS of oxidative phosphorylation	Dinitrophenol, (thyroid hormones, Long Chain fatty acid and UCP-1 are physiological uncouplers)
15	ATP synthase	Oligomycin

Obligate Allosteric Activators	Enzyme
Acetyl coenzyme A	Pyruvate carboxylase (gluconeogenesis)
N-acetylglutamate	CPS-I (urea cycle)

Non vitamin coenzyme	Enzyme
BH4	NO SYNTHASE, HYDROXYLASE OF TRYPTOPHAN, PHE
SAM	SERATONIN → MELATONIN, NE → EPINEPHRINE, CREATINE SYNTH
THIOREDOXIN	Ribonucleotide diphosphate reductase reductase
Heme	Cytochromes

ATP produced	ATP USED
Glycolysis = 7	Glyconeogenesis = 6
Acetyl coenzyme A (TCA) = 10	**Urea cycle = 4**
Beta oxidation (of 16C saturated fatty acid palmitic acid) = 106	

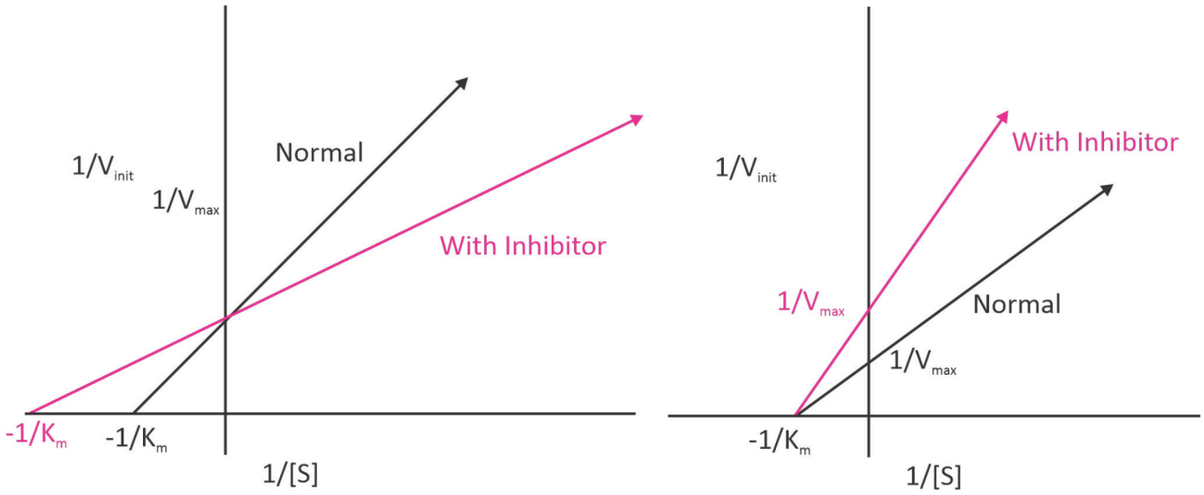
Atp synthase : β subunit is catalytic



All biochemistry test (LFT, KFT, hormones, lipids, drugs, amino acids : serum sample (plain vial)

Plasma glucose : grey vial

Enzyme Inhibitors



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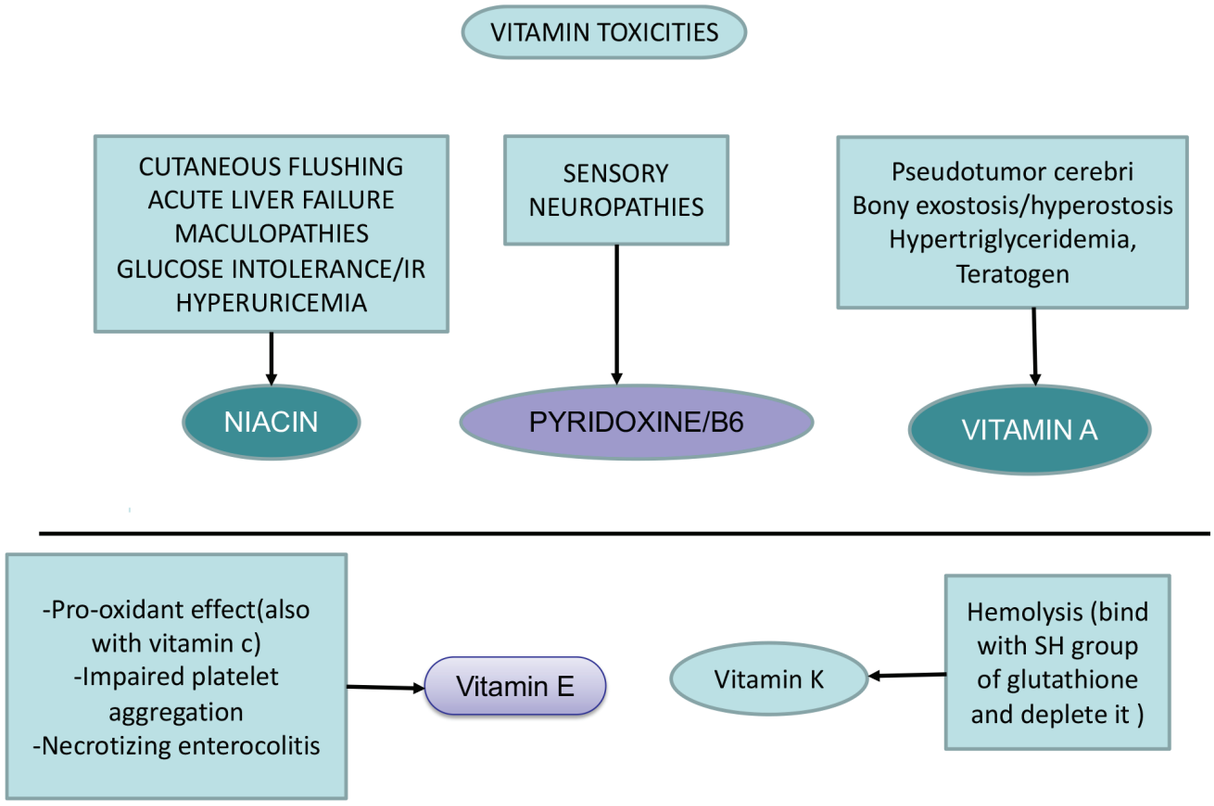
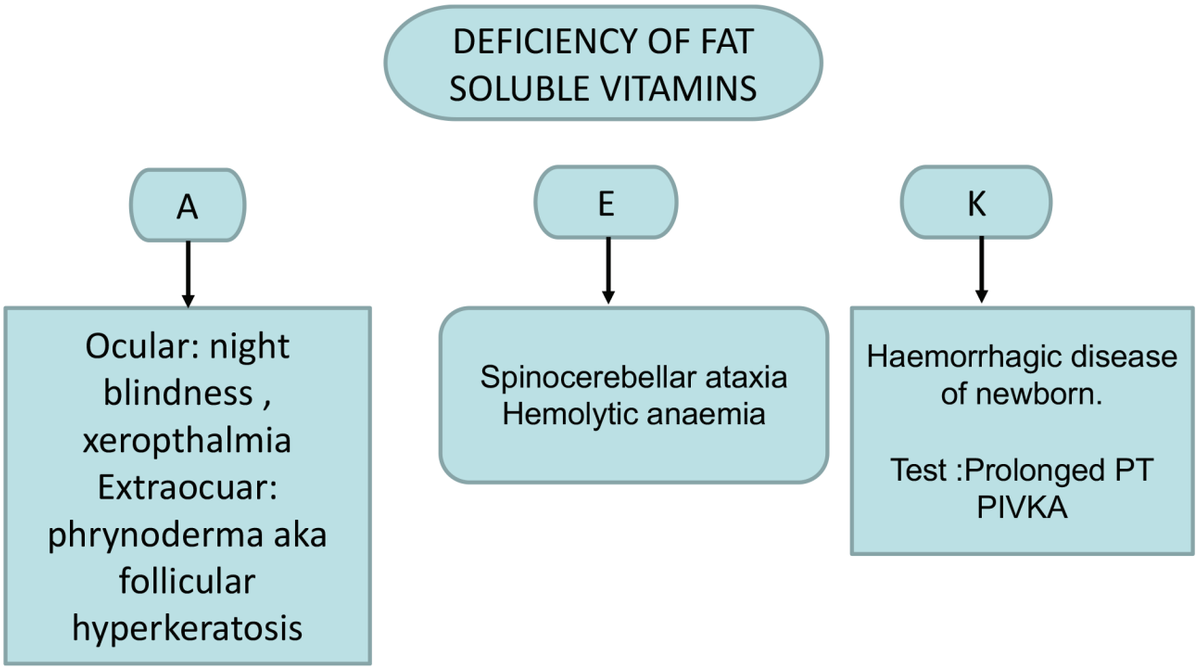
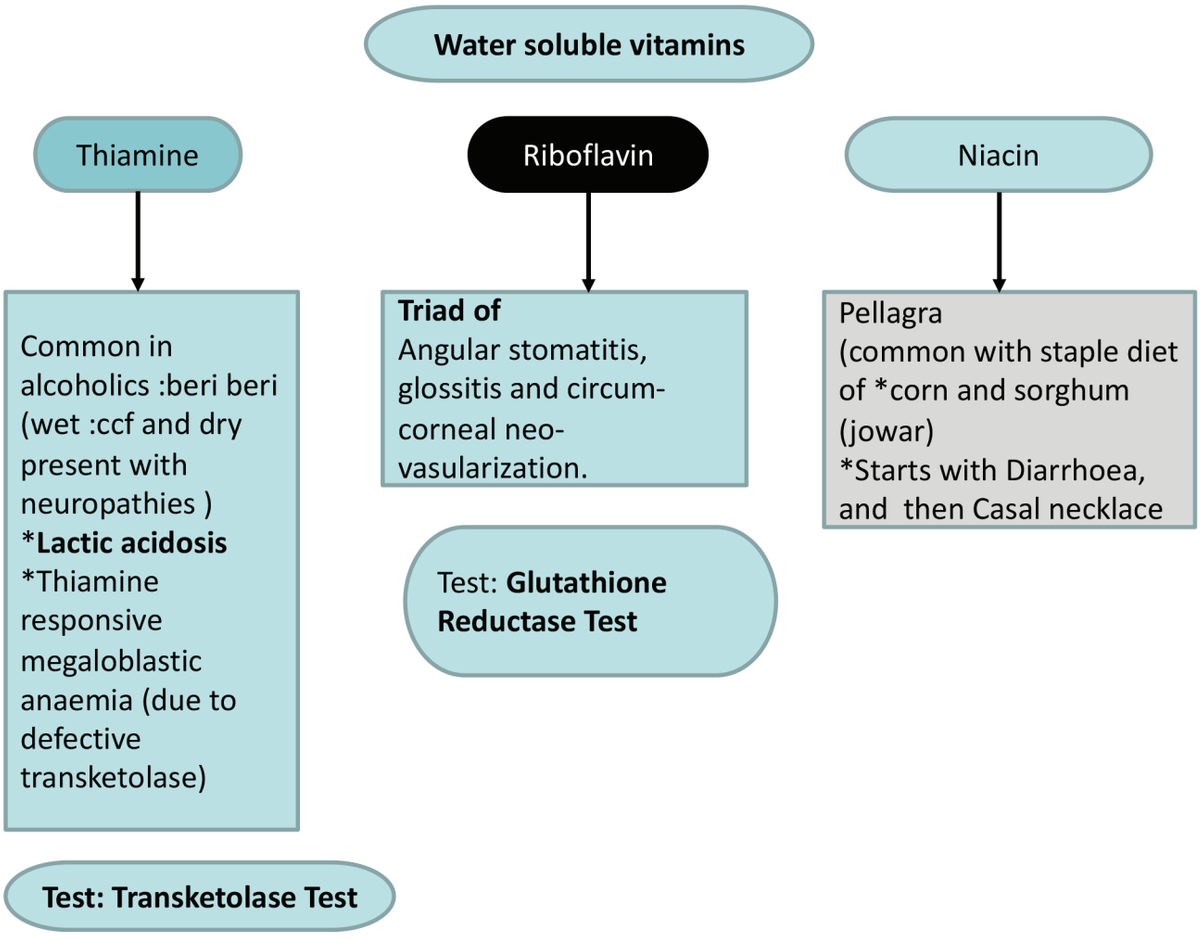
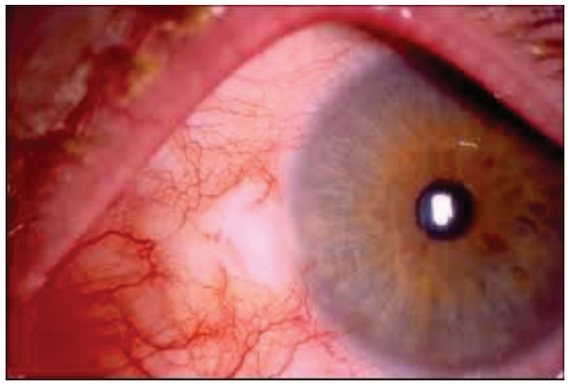
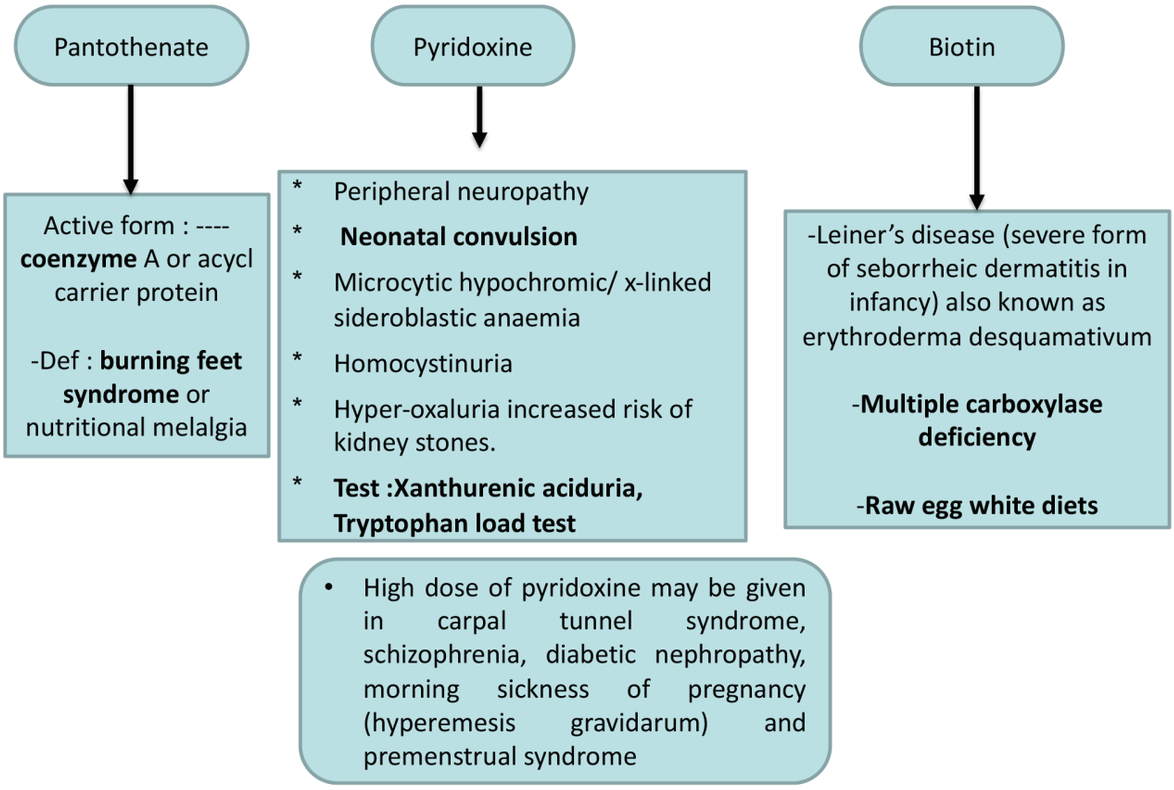
VITAMINS

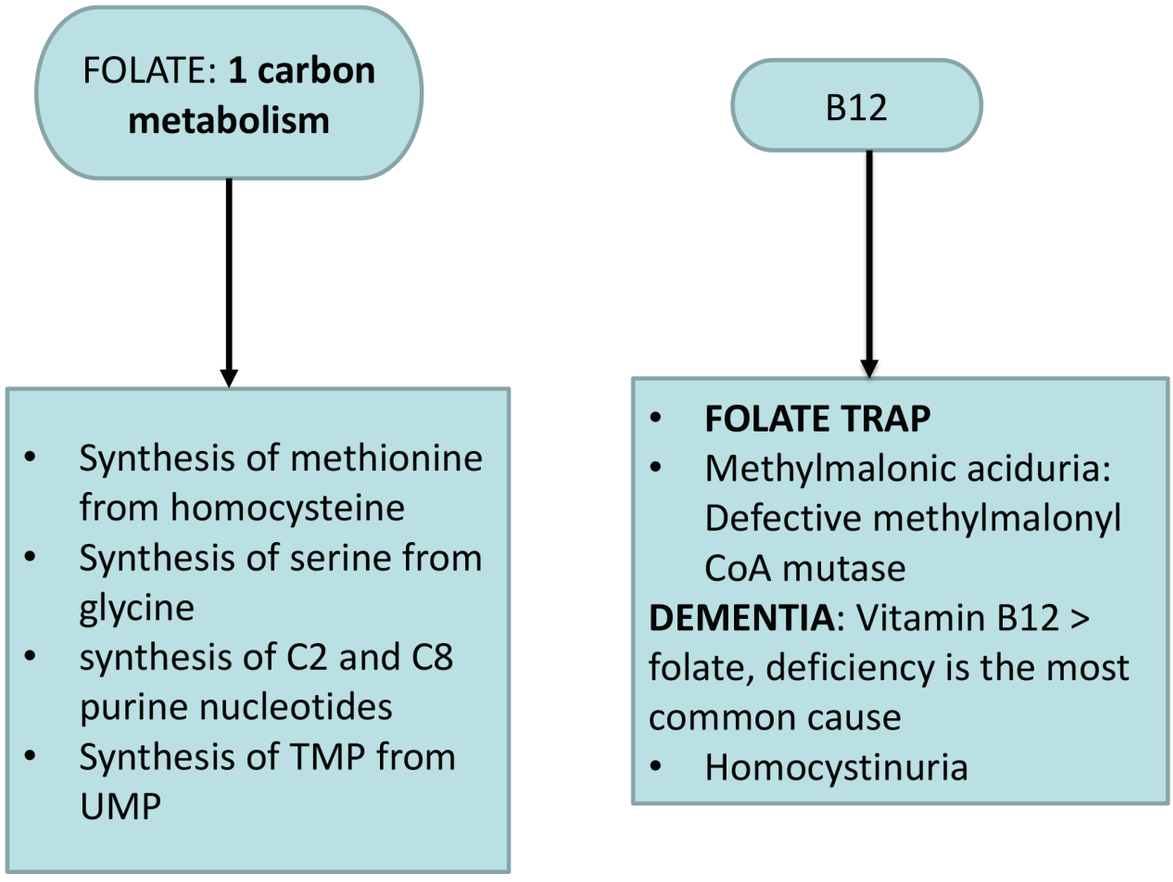

VITAMIN TOXICITIES
CUTANEOUS FLUSHING
ACUTE LIVER FAILURE
MACULOPATHIES
GLUCOSE INTOLERANCE/IR
HYPERURICEMIA
SENSORY NEUROPATHIES
Pseudotumor cerebri
Bony exostosis/hyperostosis
Hypertriglyceridemia,
Teratogen
NIACIN
PYRIDOXINE/B6
VITAMIN A
-Pro-oxidant effect(also with vitamin c)
-Impaired platelet aggregation
-Necrotizing enterocolitis
Vitamin E
Vitamin K
Hemolysis (bind with SH group of glutathione and deplete it)


DEFICIENCY OF FAT SOLUBLE VITAMINS
A
Ocular: night blindness , xeropthalmia
Extraocuar: phrynoderma aka follicular hyperkeratosis
E
Spinocerebellar ataxia
Hemolytic anaemia
K
Haemorrhagic disease of newborn.
Test :Prolonged PT
PIVKA


Water soluble vitamins
Thiamine
Riboflavin
Niacin
Common in alcoholics :beri beri (wet :ccf and dry present with neuropathies)
*Lactic acidosis
*Thiamine responsive megaloblastic anaemia (due to defective transketolase)
Triad of
Angular stomatitis, glossitis and circum-corneal neo-vasularization.
Pellagra
(common with staple diet of *corn and sorghum (jowar)
*Starts with Diarrhoea, and then Casal necklace
Test: Glutathione Reductase Test
Test: Transketolase Test

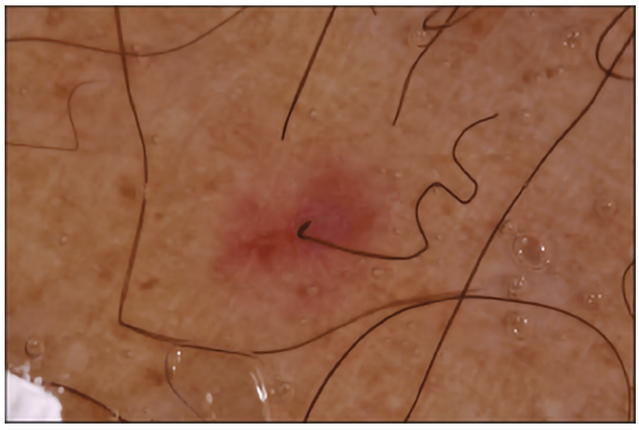


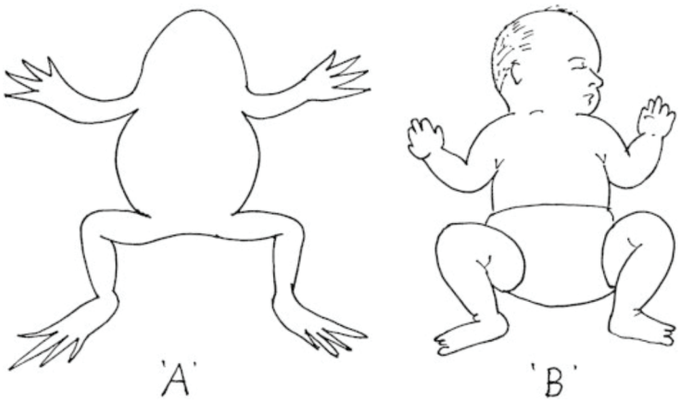

Pantothenate
Active form : ---- **coenzyme** A or acycl carrier protein
-Def : **burning feet syndrome** or nutritional melalgia
Pyridoxine
* Peripheral neuropathy
* **Neonatal convulsion**
* Microcytic hypochromic/ x-linked sideroblastic anaemia
* Homocystinuria
* Hyper-oxaluria increased risk of kidney stones.
* **Test :Xanthurenic aciduria, Tryptophan load test**
• High dose of pyridoxine may be given in carpal tunnel syndrome, schizophrenia, diabetic nephropathy, morning sickness of pregnancy (hyperemesis gravidarum) and premenstrual syndrome
Biotin
-Leiner's disease (severe form of seborrheic dermatitis in infancy) also known as erythroderma desquamativum
-**Multiple carboxylase deficiency**
-**Raw egg white diets**


FOLATE: **1 carbon metabolism**
- Synthesis of methionine from homocysteine
- Synthesis of serine from glycine
- synthesis of C2 and C8 purine nucleotides
- Synthesis of TMP from UMP
B12
- **FOLATE TRAP**
- Methylmalonic aciduria: Defective methylmalonyl CoA mutase

DEMENTIA: Vitamin B12 > folate, deficiency is the most common cause
- Homocystinuria

Vitamin C





MINERALS

1. Copper

Enzymes :

1. Cytochrome oxidase
2. Tyrosinase
3. Dopamine beta hydroxylase
4. Lysyl oxidase

Deficiency : Menke disease due to ATP 7A defects in intestine

2. Zinc

Enzymes :

1. Alcohol dehydrogenase
2. Carbonic anhydrase
3. Alkaline phosphatase

Zinc as mineral :

1. Important for insulin structure
2. Part of Zinc fingers (steroid receptors)
3. Deficiency
    a. Hypogonadism
    b. Acrodermatitis enteropathica
    c. Hypogeusia

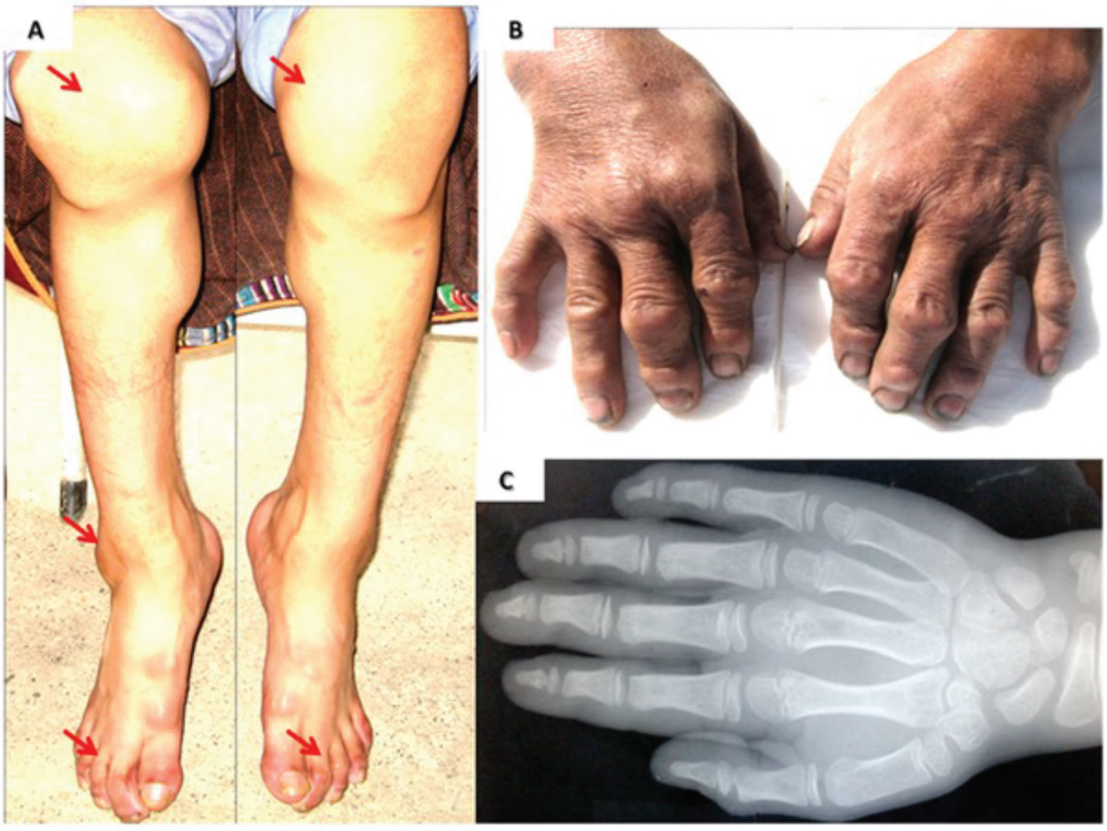
Acrodermatitis Enteropathica on 15 month old female characterised by periorificial and acral dermatitis and alopecia. There are annular, well demarcated, erythematous, crusted plaques and vesicles distributed symmetrically around orifices and hands.

3. Selenium

Enzymes :

1. Glutathione peroxidase
2. Deiodinase
3. Thioredoxin reductase

Deficiency : Keshan disease (cardiomyopathy) and Kashinbecks disease (osteoarthropathy caused by selenium and iodine deficiency)


A
B
C

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