

45 NOTES TO PG

—Dr. Ankit Yadavendra, M.B.B.S., Dr.V.M.G.M.C., Solapur

05-BIOCHEMISTRY

K_m(Micaelis Menten/Brig Haldane cons)

↑K_m → ↓attraction

competitive inh-↑K_m, V_{max} same

non competitive inh-K_m same, ↓V_{max}

uncompet inh-↓K_m, ↓V_{max}

D-glu-pyranose

D-Fructose-Furannose

enantiomer-mirror image

anomer-differ in C1-αD-glu, βD-glu

epimer-differ in other C(only1)(MAGI)

C2-D-Mannose

C3-D-Allose

C4-D-Galactose

C5-L-Idose

diastereomer-other term not applicable

glu oxidase-glu → glucuronic acid

disaccharide

nonreducing

glu-glu, C1-C1, trehalose(trehalase)

glu-glu, C1-C4, maltose(maltase/αglucosidase)

glu-fru, C1-C2, sucrose(sucrase)

reducing

glu-glu, C1-C6, isomaltose(isomaltase/αlimit dextrinase)

βglu-βgal, lactose(lactase/βglucosidase)

cellobiose

melibiose

disaccharide digestion-SI brush bord

maltotriose-glu-glu-glu

sugar—aldose—ketose

triose(3)-glyceraldehyde-diOHacetone

tetrose(4)-erythrose-erythrulose

pentose(5)-ribose,xylose-ribulose,xylulose

hexose(6)-glu,gal,mannose-fru

seduheptulose(7)-glucoheptose-sedo heptulose

polysaccharide

(fru)n-inulin

(GAl)n-aGAr

(glu)n,lin $\alpha 1 \rightarrow 4$ -amylose

(glu)n,br $\alpha 1 \rightarrow 4, 1 \rightarrow 6$ -amylopectin

(glu)n,br $\alpha 1 \rightarrow 4, 1 \rightarrow 6$ (more)-glycogen

(glu)n,lin $\beta 1 \rightarrow 4$ -cellulose

20%amylose+80%amylopectin=starch

artific glu-dextrin

(glu)n, $1 \rightarrow 3$ -dextran

N-acetyl Dglucosamine, $\beta 1 \rightarrow 4$ -chitin

dietary fibre rich in-pectin

glycosaminoglycan

(msamine+msuronic acid)n

(glu+glu)-hyaluronic acid-no SO₄

(glu+ido)-heparin-max SO₄,max -ve charge

(glu+glu)-heparan SO₄

(gal+glu)-chondroitin SO₄

(gal+ido)-dermatan SO₄(max in cartil)

(gal+glucosamine)-keratan SO₄

hyaluronic acid-synovial fluid,vitreous humor,loose connective ts

heparin-mast cell

heparan SO₄-skin fibroblast,aortic wall

Chondroitin SO₄-Cartil,bone,tendon,lig, Cornea

dermatan SO₄-skin,heart valve

keratan SO₄-hair,nail,claw,horn,hoof, cornea

lignin

highly complic br polymer of aromat alcohol

compound-bond

carb-glycosidic

prot-peptide

lipid-ester

nucleic acid-ester

glycerol+fatty acid(FA)=triglyceride

triglyceride+phosphoric acid=phosphatidic acid(PA)

PA+cholin=phosphatidylcholin/lecithin
 PA+ethanolamine=cephalin
 PA+PA=cardiolipin
 Palmitic acid+Serine=SPhingol
 ceramide+choline=sphingomyelin
 ceramide+glu=glucosylceramide
 ceramide+gal=galactosylceramide
 ceramide+glu+gal=globoside
 PEP+N-acetyl mannosamine=N-acetyl neuraminic acid(NANA)
 cer+glu+gal+NANA=ganglioside(GM3), smallest
 cer+glu+gal+NANA+gal=GM2
 cer+glu+gal+NANA+gal+gal=GM1

C16:0-palmitic acid
 C18:0-stearic acid
 C18:1 Δ 9-oleic acid
 C18:2 Δ 9,12-linoleic acid
 C18:3 Δ 6,9,12- γ linolenic acid
 C18:3 Δ 9,12,15-linolenic acid
 C20:4 Δ 5,8,11,14-arachidonic acid
 omega no.=total C-last double bond

eicosanoid(20C compound)
 cyclic
 5mem-PG
 6mem-oxane(thromboxane)
 noncyclic
 3double bond-leukotriene
 4double bond-lipoxin

hemoprot
 Hb,Mb,cytoch(C,P450),catalase,peroxidase,Try pyrrolase,NO synthase
 nonheme Fe containing prot
 ferritin

ETC-inner mitoch memb
 flow of electron fr low $\rightarrow$ high redox potential
 $-0.3 \rightarrow \text{NADH} \rightarrow \text{FMN, FeS} \rightarrow \text{coQ} \rightarrow \text{b} \rightarrow \text{c1} \rightarrow \text{c} \rightarrow \text{aa3} \rightarrow \text{O}_2 \rightarrow +0.18$

inhibitor&uncoupler
 1)inh of ETC(CAR BAP CHAC)
 I-NADH coQ oxidoreductase-Chlorpromazine, Aminobarbital, Rotenone(CAR)
 II-succinateDH(no ATP prod)-malonate

III-cytochrome reductase-BAL, Antimycin A, Phenformin(BAP)
 IV-cytochrome oxidase(CHAC)-CN, H₂S, Azide, CO
 2)inh of proton translocation/F₀F₁/ATP synthase-Oligomycin
 3)inh of nucleotide transporter-ATP-ADP
 4)uncoupler-↓ATP synth, ↑ETC
 natural-UCP/thermogenin, thyroxine
 synth-2,4-dinitrophenol, 2,4-dinitroresol, ChloroCarbonylCyanide
 Phenylhydrazine(CCCP)
 CN bind Fe⁺⁺⁺ of cyt a₃

enzyme

International Union Biochemistry(1964)

1st digit-class

2nd digit-subclass

3rd digit-subsubclass

4th digit-individual enzyme

(OTHLIL)

Oxidoreductase

Transferase

Hydrolase-urease

Lyase(synthase)

Isomerase

Ligase(synthetase)-req ATP

↑10°C temp-↑rate by doubl till opt temp

cofactor-enzyme

Cu(TDCL)-Tyrosinase, Dopamine βhydroxylase,Cytochrome oxidase, Lysyl oxidase

Mg-cholesterol synth, glutamyl cysteine synthetase, glutathione synthetase

(CTPARK)Carboxylase, Transketolase, Phosphatase, Adenyl cyclase, Ribonuclease,
 Kinase

Mn-arginase, pyruvate carboxylase, isocitrateDH, superoxide dismutase, peptidase

Mb-sulphite oxidase, xanthine oxidase

Se-glutathione peroxidase

Zn-carbonic anhydrase, glutamateDH, LDH, alcoholDH, superoxide dismutase,

leucine aminopeptidase, retinene reductase,alk phosphatase, DNA polymerase, RNA
 polymerase, d-ALA dehydratase

isoenzyme

multiple form of same enzyme, in single species, catalyze same chemical rxn

differ in struct, electrophoretic,immunolog, pH opt, Km, V_{max}, AA seq, quart struct, ts
 conc, physical propert

no. on basis of electroph mobility towards anode-LDH1-highest mobility

LDH1-heart(max),RBC

LDH2(max)-heart,RBC
LDH3-brain,kidn
LDH4-liver,sk m
LDH5-sk m,liver
CPK1(BB)-Brain
CPK2(MB)-Heart
CPK3(MM)-sk m
SGPT-cytosol
SGOT-cytOsOl+ mitOchOndria(bOth)
Liver-ALT,muScle-AST
 α 2HEat labile ALP-HEpatitis
pre β ALP-bone ds

oxygenase
dioxygenase(true oxygenase)-homogentisate oxidase, L-Try pyrrolase
Monooxygenase(Mixed fn oxidase)-cyt P450, biosynth steroid

zymogen(proenzyme)
pepsinogen
trypsinogen
chymotrypsinogen
proelastase
procarboxypeptidase
proaminopeptidase
fact II,VII,IX,X,XI,XII
plasminogen
angiotensinogen

PAGHADI
PyruvateDH complex
Acetyl coA carboxylase
Glycogen synthase
HMG coA reductase
-Active-Dephosphorylated form, Insulin dephosphorylat, maintain phosphorylase b in inactive form

CGH-active-phosphorylated form, epinephrine, NE, glucagon phosphorylate
Citrate lyase
Glycogen phosphorylase
H sensitive lipase

acid phosphatase(PT)
pH=5-6

Prostate-Tartarate sensitive
RBC-formaldehyde, cupric ion sensitive
band1-prostate
band2,4-granulocyte
band3-PLT, RBC, monocyte
band5-osteoclast, monocyte, Kupffer cell

suicide enzyme-cyclooxygenase

fluorescence
release of longer wavelength on absorb short wavelength

organ—fed—fasting—starvation
brain-glu-glu-ketone body
heart-FA-FA-ketone body
liver-glu-FA-AA
m,adipose ts-glu-FA-FA,ketone body
RBC-glu-glu

hypoglycemia → ↓insulin → ↑glucagon → ↑epinephrine → ↑cortisol,GH

high energy compound(≥ 7 kcal/mol)
PEP(14.8) > carbamoyl P > 1,3BPG > creatine P(10.3) > ATP → ADP(7.3) > ATP → AMP
(7.7) > G1P > G6P > glycerol1P

glycolysis/Embden Meyerhof pathway (cytosol)
glu → (glucokinase/hexokinase+ATP+Mg) → G6P+ADP → (phosphohexose
isomerase+Mg) → F6P → (PFK1+ATP+Mg) → F1,6BP+ADP → (aldolaseA) →
DHAP,G3P → (phosphotriose isomerase) → G3P → (G3PDH+iP+NAD) →
1,3BPG+NADH → (phosphoglycerate kinase+ADP+Mg) → 3PG+ATP → 2PG →
(enolase+Mg) → PEP → (pyruvate kinase+ADP+Mg) → pyruvate+ATP →
(LDH+NADH) → lactate+NAD
irreversible rxn-hexokinase, PFK, pyruvate kinase
substrate level phosphorylation-phosphoglycerate kinase, pyruvate kinase
Brain,all ts-Hexokinase(BHeja)
↓Km(0.1mM),inh by G6P
liver,pancr-glucokinase
↑Km(10mM),not inh by G6P
ATP-inh,AMP-activator
1 glycolysis=9-2=7ATP
RBC,cornea,lens=2ATP
RBC-hypoxia → ↑release all glycolytic enz fr band3 RBC memb

enzyme-inh

G3PDH-iodoacetate, arsenite

PFK1-ATP, citrate, H⁺

pyruvate kinase-ATP

phosphoglycerate kinase-arsenite

enzyme-activator

PFK1-F2,6BP(MI), ADP, AMP, iP

pyruvate kinase-F1,6BP

Pasteur effect-O₂ → ↓glycolysis

Crabtree effect-glu → ↓O₂ consumption

Rapaport Leubering cycle

glu → → 1,3BPG → (2,3BPG mutase) → 2,3 BPG → (2,3BPG phosphatase) → 3PG → → pyruvate

ATP=1

Cori cycle

RBC,m(glu → lactate) → liver(lactate → glu) → RBC,m

any monosaccharide attached to P cant cross plasma memb

lactate → Cori cycle, TCA cycle

glu-Ala cycle

m[pyruvate → (ALT/SGPT) → Ala] → liver [Ala → (ALT/SGPT) → pyruvate → G6P → glu]

gluconeogenesis(mitochondria+ cytosol)

substrate-lactate, pyruvate, AA(except Leu, Lys), propionate, glycerol

liver=90%(1kg/d), kidn=10%

mitoch[pyruvate → (pyruvate carboxylase+ATP+CO₂) → OAA+ADP →

(malateDH+NADH) → malate+NAD] → cytosol[malate → (malateDH+NAD) →

OAA+NADH → (PEP carboxykinase+GTP) → PEP+GDP+CO₂ → → F1,6BP →

(F1,6BPase+H₂O) → F6P+iP → G6P → (G6Pase+H₂O) → glu+iP

AcoA deriv fr FA oxidase allosteric activat-pyruvate carboxylase

↑-after 72h starvation

ts mainly dependent on glu-brain, RBC, testes, kidn medulla

fasting>24h-gluconeogenesis

G6Pase present-liver,kidn, absent-m,brain,adipose ts

glycerol → (glycerol kinase+ATP) → glycerol3P+ADP → (glycerol3PDH+NAD) →

DHAP+NADH

glycerol kinase present-liver,kidn ,absent-adipose ts

odd chain FA → propionic acid → methyl malonylcoA → (methylmalonylcoA mutase+ 5deoxyadenosyl cobalamin) → succinylcoA

enz comm to glycolys& gluconeogenesis-fru2,6bisphosphatase

TCA/citric acid/Krebs cycle(mitochondr matrix)

pyruvate → (PDH+NAD+coA) → AcoA+NADH+CO₂ → (citrate synthase+OAA) →

citrate → (aconitase) → cisaconitate+H₂O → (aconitase+H₂O) → isocitrate →

(isocitrateDH+NAD) → αKG+NADH+CO₂ → (αKGDH+NAD+coA) →

ScoA+NADH+CO₂ → (succinate thiokinase+GDP) → succinate +GTP+coA →

(succinateDH+FAD) → fumarate+FADH → (fumarase+H₂O) → malate →

(malateDH+NAD) → OAA+NADH

3NADH+1FADH+1GTP

1TCA cycle=10ATP, total=12.5ATP

1glu(ATP)=7+12.5×2=32(net), 34(total)

OAA remain constant, play catalytic role

pyruvate dehydrogenase(3enzyme)

5coenzyme(TLCCFN)

TPP,Lipoic acid,CoA,FAD,NAD

AA → TCA cycle

His,Pro,glutamine,Arg → Glu → αKG

Iso,Met,Val,propionate → succinyl coA

Tyr,Phe → fumarate

Try → Ala → pyruvate

OHPro,Ser,cysteine,Thr,Gly → pyruvate

enzyme-inh

aconitase-fluoroacetate(noncomp)

AlpKGDH-Arsenite(noncomp), hyperAmmonemia, succinylcoA, NADH

succinateDH-malonate(compet)

enolase-fluoride

citrate synthase-ATP, NADH, AcoA, succinylcoA

isocitrateDH-ATP, NADH

enzyme-activator

isocitrateDH-ADP

HMP/hexose monoP/pentose P/Dicken Horecker/shunt/phosphogluconate oxidative pathway(cytosol-liver, adipose ts, adrenal gld, RBC, testis, lactating breast)

glu → G6P → (G6PD+NADP) → → ribose5 P(5C) → (transaldolase,transketolase-3C) →

xylulose5P(5C),seduheptulose(7C),erythrose(4C) → glu

2transketolat, 1transaldolat rxn

90%glu → glycolysis, 10%glu → HMP path

liver,RBC-30%glu utilizat

fructose metab

fru → (fructokinase) → F1P → (aldolaseB) → DHAP,glyceraldehyd → (triose kinase)

→ G3P → → pyruvate
fructokinase-liver, kidn, intest

galactose metab

gal → (galactokinase) → gal1P → (gal1P uridyl transferase) → UDPgal → UDPglu → glycogen

glycogen synthesis

UDPglu + glycogen/glycogenin → (glycogen synthase) → linear glycogen → (branching enzyme/1 → 4,1 → 6 transglycosylase) → glycogen

glycogenolysis

glycogen → (glycogen phosphorylase) → smaller glycogen + G1P → (debranching enz/1 → 4,1 → 4,7 glucan transferase) → (1,6glucosidase) → glu

lipid metab

FA → TG-esterificat

TG → FA-lipolysis

FA → AcoA-βoxidat

AcoA → FA-FA synthes/lipogenesis

even chain FA is never converted to glu

βoxidation FA (mitoch matrix)

FA → (thiokinase/AcoA synthetase + ATP) → acylCoA + AMP → [carnitine acyltransferase I (outer surf inner mitoch memb)] → carrier prot → [carnitine acyltransferase II (inner surf inner mitoch memb)]

FA + CoA + ATP → FA-CoA + AMP

16CFA (palmitic acid) - 7 cycle = $4 \times 7 = 28 + 8 \text{ AcoA} = 8 \times 10 = 80$, net ATP = $28 + 80 - 2 = 106$ ATP

18CFA (stearic acid) - $8 \times 4 + 9 \times 10 - 2 = 120$ ATP

for unsaturated FA - subtract 2 ATP for each double bond

oleic acid = 118 ATP

linoleic acid = 116 ATP

linolenic acid = 114 ATP

arachidonic acid = 126 ATP

peroxisome βoxidation - long chain FA → 8CFA (octanoyl CoA)

αoxid-br chain FA (phytanic acid)

organ not use FA for energy - brain, RBC, adren medulla

enzyme-inh

carnitine acyltransferase I - malonyl CoA

odd chain FA → propionic acid + acetic acid

lipogenesis

1AcoA+7malonylcoA+14NADPH= palmitic acid(16C)+7H₂O+7CO₂+14NADP
allosteric inh of AcoA-palmitic acid
allosteric activator of AcoA-citric acid
malonyl acyl transferase-2enz activity
sat FA(≤16C) assemble in-cytoplasm

esterification

glycerol → (glycerol kinase,abs in adipose ts)+ DHAP(adipose ts) → TG

lipolysis

TG → (lipoprot lipase-bld, HS lipoprot lipase-adipose ts) → glycerol+3FA

cholesterol(27C)

esterif(linoleic acid, palmitic acid, stearic acid)=70%, nonesterif=30%

synth

AcoA+AcoA → acetoacetyl coA → (HMG coA synthase)+AcoA → HMGcoA(6C) → (HMGcoA reductase) → mevalonate(6C) → isoprenoid(5C)×3 → farnosyl(15C)×2 → squalene(30C) → lanosterol → → 19enz → cholesterol

ketone body synth(liver,mitochondria)

AcoA+AcoA → acetoacetyl coA → (HMG coA synthas)+AcoA → HMGcoA → (lyase) → acetoacetate+AcoA → βOH butyrate(MC),acetone
prim ketone body-acetoacetate
sec ketone body-βOHbutyrate,acetone

AA metab

AA → KA

glutamate → αKG

Ala → (SGPT/ALT) → pyruvate

Asp → (SGOT/AST) → OAA

inborn error in AA metab-urine odour

glutaric acidemia, isovaleric acidemia-Sweaty feet, acrid

haWkINSuria-SWimming pool

MSUD-maple syrup

Met malabsorption-cabbage

multiple carboxylase def-tomcat urine

PKU-mousy/musty

trimethylaminuria-rotting fish

tyrosinemia-rancid butter, fishy, boiled cabbage

urea cycle

CO₂+NH₃ → (carbamoyl P synthase1) → carbamoyl P → citrullin → cytoplasm →

(arginosuccinate synthase) → arginosuccinate → (arginosuccinate lyase) → Arg → (arginase, hydrolysis) → ornithine & urea → citrullin
most powerful activator of carbamoyl P synthetase I is N-acetyl glutamate (NAG) - activated by Arg
2 NH₂ groups in urea derive from Asp + NH₃
NH₃ > urea transport by Glu
N₂ carried from muscle → liver, kidney - Ala
urea cycle → TCA cycle - fumarate

bilirubin metabolism

heme → (microsome) → bilirubin
hemoprotein, senescent RBC → monocyte (liver, spleen, bone marrow) → (heme oxygenase) → biliverdin → (biliverdin reductase) → bind to albumin → hepatocyte → bind to ligandin → [UDP glucuronyl transferase (UGT1A1)] → conjugate with glucuronic acid → conjugate bilirubin → bile → 20% reabsorb in ileum & colon, rest excreted in feces (stercobilinogen) & urine (urobilinogen)

pathway-rate limiting enzyme

ACh synth - choline uptake
bile acid synth - 7 α hydroxylase
catecholamine synth - Tyr hydroxylase
cholesterol synth - HMGCoA reductase
FA oxidation - carnitine Acyl transferase I
FA synth - ACoA carboxylase
gluconeogenesis - pyruvate carboxylase > fructose 1,6-bisphosphate > PEP carboxylase
glycogenesis - glycogen synthase
glycogenolysis - glycogen phosphorylase
glycolysis - phosphofructokinase I
heme synth - Liver - δ -amino Levulinate synthase, erythroid cell - uroporphyrinogen synthase, ferrochelatase
HMP shunt - G6PD
ketone body synth - HMGCoA synthase
Krebs cycle - isocitrate DH
porphyrin synth - ALA synthetase
purine biosynth - PRPP synthetase
pyrim synth - carbamoyl P synthase II
urea synth - carbamoyl P synthase I
uric acid synth - xanthine oxidase
vitD synth - 1 α hydroxylase (kidney)

cytosol

glycolysis
HMP shunt

transamination
glycogenolysis
glycogen synthesis
FA synthesis
prot synthesis
gluconeogenesis(part)
pyrimidine synthesis(part)
heme synthesis(part)
urea synthesis(part)
mitochondria
TCA cycle
ETC
 β oxidation FA
ATP synthesis
ketone body synthesis
pyrimidine synthesis(part)
urea synthesis(part)
heme synthesis(part)
gluconeogenesis(part)
nucleus
DNA replication
transcription
ER
drug metabolism
ethanol oxidation
heme $\rightarrow$ bilirubin
FA chain elongation
protein synthesis(part)
glycoprotein synthesis(part)
lipoprotein synthesis(part)
cholesterol synthesis(part)
golgi body/dictyosome
maturation of synthesised protein
glycosylat
lysosome
protein degradation
carbohydrate degradation
lipid degradation
nucleotide degradation
peroxisome
 α oxidation long chain FA
ribosome
cytosolic/free/polyribosome

cytosolic prot(Hb)
cytoskeletal prot
mitochondr prot
nuclear prot
peroxisomal prot
bound/rough ER polyribosome
mitochondr memb
ER memb
golgi memb
plasma memb
secretory prot
lysosomal enzyme

organelle-marker
cytosol-LDH
endoplasmic reticulum-G6Pase
Golgi apparatus-Galactosyl transferase
lysosome-acid phosphatase
mitochondria-glutaMATEdH(MATrix), cytochrome oxidase(MEmb)
nucleus-DNA
peroxisome-catalase, uric acid oxidase
pl memb-Na⁺-K⁺ ATPase, 5'-nucleotidase, adenyl cyclase
Ribosome-RNA

glycogen storage ds
(VPCAMHaiTu-GADiBhiMeriHaiPar)
I-VonGierke-Glucose6phosphatase
II-Pompe-Acid maltase/Alp 1-4 glucosidase
III-Cori-Debranching enz
IV-Anderson-Branching enz
V-McArdle-Myophosphorylase
VI-Hers-Hepatic phosphorylase
VII-Tauri-Phosphofructokinase
VIII-—-Phosphorylase kinase
m-II,V,VII,fatal-II,IV
exercise intolerance,2nd wind phenom-V

lipid storage disorder
(TumSabNahiGayeKyaFirFaridabad-HaHarSaalGaavGaavGaavCiye)
Tay Sach-βHexosaminidaseA
Sandoff-βHexosaminidase A+B
Neiman Pick-Sphingomyelinase
Gaucher-βGlucosidase

Krabbe(GM1 gangliosidase)- β Galactosidase
Fabry- α Galactosidase
Farber-Ceramidase
MR-NGKF,ren ds-Fabry

Phe-Tyr metab(PTNAT-HTHOH)
Phenylketonuria(hyperphe I)-Phe OHLase
Tyrosinemia II(Richer Hanhart synd)-Tyr OHLase
Neonatal tyrosinemia-Hydroxyphenyl pyruvate OHLase
Alkaptonuria-homogentisate Oxidase
Tyrosinemia I(tyrosinosis,hepatorenal synd)-fumaryl acetoacetyl Hydrolase
atypic phenylketonuria(hyperphe II,III)-dihydrobiopterin reductase
hyperphe IV,V-dihydrobiopterin synth

porphyria-enzyme def(ACPEHaiVaha)
A/c intermittent-uroporphyrinogen1 synthase
Cong erythropoeitic(Gunther ds)-uroporphyrinogen3 cosynthase
Porphyri cutan tarda-uroporphyrinogen decarboxylase
Erythropoeitic/Proto-ferrochelatae
Heredit copro-coproporphyrinogen oxidase
Variegate-protoporphyrinogen oxidase

urea cycle(AACHH-AAAOC)
Argininuria-Arginase
Arginosuccinaturia-Arginosuccinate lyase
Citrullinemia-Arginosuccinat synthetase
HyperAmMONemia I-Ornithine transcarbAMOylase
Hyperammonemia II-CarbamoylP synthetase-I

mucopolysaccharidoses/lysosomal storage ds
I-Hurler synd- α Lp L iduronidase
II-Hunter synd-iduronate sulfatase
III-SanFilipp synd-heparan SulFamidase
VI-MaroteauxLamysynd-arylsulfataseB
VII-Sly synd- β glucuronidase
IX-Natowicz synd-hyaluronidase

ds-enzyme def
albinism-tyrosinase
amyotrophic lat Sclerosis-Superoxide dismutase
BruTTon agammaglobulinem-Tyr kinase
Canavan ds-aspartoacylase
c/c granulomatous ds-NADPH oxidase

Criggle Najja-UDPglucuronyl transferase
 cystathionuria-cystathionine lyase
 essential pentosuria-xylitolDH
 fructosuria(nonfatal)-fructokinase B
 galactossemia1/gal intolerance(fatal)-gal1P uridyl transferase
 gyrate atrophy of retina-ornithine d transaminase
 heredit angioneurotic edema-C1 esterase
 heredit fru intoler(fatal)-ALdoase B
 HHH synd-ornithine permease
 histidinemia-His NH₃ lyase
 homoCYSTinuri-CYSTathione synthetase
 hyperlipoproteinemia I-lipoprotein lipase
 I cell ds-N acetyl glucosamine1P transferase
 Lesch Nyhan synd-HGPRT
 maple syrup urine ds-α-ketoacid decarboxylase
 Menkes ds(YYYY)-IYsYI hYdroxYlase
 metAchromatic leukodystrophy-arylsulfatase A
 mucopolysaccharidosis1(sialidosis)-sialidase/neuraminidase
 osteopetrosis-carbonic anhydrase II
 recur Mburia-carnitine palmitoyl transferase II
 Refsum ds-phytanic acid oxidase
 SCID-adenosine deaminase
 sudden infant death synd-med chain acyl coADH
 TTP-ADAMTSB(vWFmetalloproteinase)
 Werner synd-DNA helicase
 Wolman ds-Lysosomal acid Lipase
 xanthinuria-xanthine oxidase
 Xeroderma pigmentosa-DNA exonuclease

enzyme replacement therapy
 Gaucher ds(α-glucuronidase/cerebrosidase)
 Hurler synd
 Hunter synd
 Maroteaux Lamy synd
 Fabry ds
 Pompe ds

lipid
 chylomicron(B48,origin) > VLDL(B100,preβ) > LDL(B100,E,β) > HDL(A1,A11,α) →
 (DLT) Diameter, Lipid, Triglyceride
 C < V < L < H → (DPP) Density, Protein, Phospholipid
 L > H > V > C → cholesterol
 endogenous/sr TG-VLDL

RNA editing

total cholesterol:HDL<3.5-prevent CAD

↑intracellular cholesterol

↓HMGCoA synthase, HMGCoA reductase

↓LDL receptor

↑ACAT, cholesterol esterification

LDL receptor present on Liver & extrahepatic tissues, present on cytosolic side (clathrin coated)

LCAT-esterification of cholesterol

apoA1-activate LCAT

apoC1,2-activate lipoprotein lipase

apoA2, C3-inhibit lipoprotein lipase

apoE-activate of cholesterol

macrophages have affinity for oxidized LDL

fatty liver-↑TG

Cholesteryl Ester Transfer Protein (CETP)-transport protein from HDL → VLDL, chylomicron

hyperlipoproteinemia (CLLIVV)

1-C-familial lipoprotein lipase defect

2a(familial hyperβlipoproteinemia, hypercholesterolemia)-L-LDL receptor defect

2b(familial combined hyperlipoproteinemia)-L+V-↑apoB, ↓LDL receptor

3(broad β distribution/familial dysβlipoproteinemia/remnant removal disorder)-I-apoE defect

4(familial hypertriglyceridemia)-V-↑endogenous TG-apo V defect

5(endogenous hypertriglyceridemia)-V+C-severe apoV defect

hypolipoproteinemia

familial hypoβlipoproteinemia-↓apoB

familial αlipoproteinemia (Tangier disorder)-no HDL absent

test for carbohydrate

Molisch-Identify carb

Barfoed-Differ b/n mono&disaccharide

Benedict-identify monosaccharide

Benzidine-pentose

Selivanoff-identify Fru

Roth's-ketone body

Bial's-pentose

Ehrlich-bilirubin in urine

Fouchet-bile pigment

Hay Sulphur-bile Salt

rxn given by cholesterol

Salkowski
Liebermann Burchard
Zak
Zimmerman

meth for prot struct

named after Person/reagent-Primary(trans)-mass spectrometry(MALDI-Matrix Associated Laser Desorption Ionisation, H⁺ ion)

named after tEchnology-sECondary(Pauling& Corey-no. of AA per helix=3.6, αhelix>βpleated)& tertiary(MI bond-hydrophobic, max no.-van der Waal)

struct by default-tertiary-Xray crystallography, NMR

separation technique-quarternary(Hb, Ig, immune complex, myosin)-PAGE electrophoresis, HPLC chromatography, centrifugation, isoelectric focussing
total prot estimation-Biuret assay, Lowry assay, Bradford assay

prot denaturing agent

physical-heat, violent shaking, Xray, UV

chemical-acid, alkali, organic solvent(ether, alcohol, acetone), salt of heavy metal(Pb,Hg), urea, salicylate, guanidine

Beer law-↑amount transmit light-↓conc of absorbing material

Lambert law-↑amount transmit light-↓ thickness of absorb material

strength of bond(CDEHαιVaha)

Covalent> Disulfide> Electrostatic> H₂> vdW

def vitamin—enzyme/substrate marker

B1-RBC transketolase

B2-RBC glutathione peroxidase

B3-urine N methyl nicotinamide

B6-urine xanthurenic acid

B9-urine FIGLU

B12-urine methylmalonic acid

vitA

follicular hyperkeratosis

def in famine

metaplasia

vitD

7deOHcholesterol → (UV light,skin) → cholecalciferol(calcidiol) → (25OHase,liver) → 25OHcholecalciferol(calcidiol) → (1αOHase,kidn) → 1,25OHcholecalciferol(calcitriol)
synth by-keratinocyte

daily req=400-600IU

measurment in serum-vitD2(25OHvitD)

vitE

prevent chain rxn in lipid peroxidation(chain breaking aqueous antioxidant), cant prevent CAD

vitK

K1(phytonadione,phylloquinone)-plant

K2(menaquinone)-bact

K3(menadione)-synth

carboxyl of γ Glu in fact II,VII,IX,X,protC,S

vitC

posttranslation modificat(OHlation of Pro&Lys $\rightarrow$ OHPro&OHLys) collagen

NE synth[dopamine $\rightarrow$ (dopamine β monooxygenase/hydroxylase) $\rightarrow$ NE]

carnitine synth

bile acid synth(7 α hydroxylase)

Fe absorpt(Fe $^{+++}$ $\rightarrow$ Fe $^{++}$)

adrenal steroid synth

Tyr metab(para OHphenylpyruvate $\rightarrow$ homogentisate)

vitB9 metab(vitB9 $\rightarrow$ THF)

cant be synth in human d/t def of-L gulonolactone oxidase

vitB1(thiamine)

thiamine pyrophosphate-PDH, α KGDH, transketolase

vitB2(riboflavin)

FAD-succinateDH(compl II), FMN-NADH DH(compl I), photophobia, blurred vision, burning, itching eye, corneal vascularisation, petechiae, poor growth

vitB3(niacin)

NAD,NADP-G6PD

vitB5(pantothenic acid)

coA(adenylic acid+ pantothenic acid+ β mercaptoethanolamine)

(thioethanolamine)-carrier of acyl grp

burning feet synd

vitB6(pyridoxine)

pyridoxal P(carrier of NH $_2$ grp) req by all transaminase& some decarboxylase,

cystathione lyase(synthase), glycogen phosphorylase

vitB7(biotin)

carboxylat of pyruvat(3C), propionyl coA (3C), acetyl coA(2C)

vitB9(folic/pteroylglutamic acid)

folic acid $\rightarrow$ F reductase $\rightarrow$ THF(1C carrier)

vitB12(cyanocobalamine)(HaM)

counteract action of phenobarbitone

homocysteine+N5methylTHF $\rightarrow$ (homocysteine methyl transferase/methionine

synthase+methylcobalamine) $\rightarrow$ methionine+THF

γ Glu carboxypeptidase- $\uparrow$ vitB9 absorpt

enzyme elevated in CSF

adenosine deaminase-TB

creatine kinase-hydrocephalus, infarction, tm, SAH, trauma

LDH-ICH, bact meningitis

lysozyme-bact meningitis, TB meningitis, cerebral atrophy, tm, multiple sclerosis, ICH, epilepsy

organ dep on insulin for glu(BASH)

Breast

Adipose ts

Skeletal m

Heart

not dep(BRAKLIPP)

Brain

RBC

Adrenal

Kidney

Liver

Intestine

Pancreas

Placenta

bile acid

½ of body cholesterol metab → bile acid

prim-cholic acid, chenodeoxycholic acid

conjugate-glycocholic acid, taurocholic acid

sec-deoxycholic acid, lithocholic acid

intest bact-prim BA → sec BA by deconjugate & 7α dehydroxylation

7αhydroxylase

activator-cholesterol, thy h

inhibitor-bile acid

source of energy for m activity

immediat-1-2s-ATP, 5-7s-creatine P

short burst(100m sprint, wt lifting)-ATP, creatine P, anaerobic process

long durat(200m sprint)-glycogen anaerobic metab

longer durat(marathon, jogging)-aerobic metab, FA

starvation

initial stage(2-3d)-liver glycogen(18h), gluconeogenesis(m prot breakdown → Ala, Glu), ketone body, FA

intermediate stage(24d)-FA, ketone body

advanced stage(>24d)-breakdown ts prot

HarTnup ds

defect in Transport across Tubule of Tryptophan & neutral AA
pellagra, neurologic sympt, aminoaciduria

cystinosis-lysosomal defect

branched chain AA (Val, Leu, Iso) are major N₂ source for Glu & Ala synth

Arg + Gly → (kidn) → guanidoacetate + SAM (liver) → SAH + creatine → creatineP → (m) → creatinine

Asp + glutamine + ATP → asparagine-glutamic acid + AMP

AA derivative

phenylalanine → tyrosine → dopa

→ dopamine → melanin

→ norepinephrine → epinephrine

→ thyroxine

tryptophan

→ kynurenine → xanthurate

niacin

serotonin

melatonin

5-HIAA

histidine

histamine (histidine decarboxylase)

ergothionine

carnosine

anserine

glycine

porphyrin → heme (succinic acid)

purine

creatine (GAM) (Gly + Arg + Met)

glutathione (Gly + Cys + Glu) - Cys-reductase property, transport of AA in intest & kidn tubule
via γ-glutamyl (Meister) cycle, detoxification

glycocholic acid

hippuric acid

arginine

urea

creatine

NO

ornithine

→ (ornithin decarboxylase) → putrescine

→ spermine

→ spermidine

methionine(no SH grp, cant form disulphide bond)

choline

spermine

spermidine

βalanine

cysteine

coA

carnosine

carnitine(Lys+Met)

cysteine(SH grp)

coA

taurine(present in human milk → brain developm)

glutamine

GABA

homocysteine

alkaptonuria

thrombosis,CAD,stroke

osteoporosis,#

neuropsych manifestat

developm delay

visual disturbance

microalbuminuria

lysine

carnitine(βOH γCH₃ aminobutyrate)

selenocysteine

glutathione peroxidase(antioxidant)

deiodinase(convert T₄ → T₃)

thioredoxin reductase

Gly reductase

formateDH

common interm in synth of Arg, Ser& Asp

glyceraldehyde3P

in chromatography, nonpolar AA faster than polar AA

most nonpolar(ILVA)-Isoleu> Leu> Val> Ala

in reverse chromatography& electrophoresis polar faster than nonpolar

AMINO ACID(Lα)

nature AA>300, body>50, protein=20

essential(AVHILLMPTT)

Arginine(semi)

Valine

Histidine(semi)

Isoleucine

Leucine

Lysine

Methionine

Phenylalanine

Tyrosine

Tryptophan

ketogenic(LL)

Leucine

Lysine

mixed(PITT)

Phenylalanine

Isoleucine

Tyrosine

Tryptophan(largest)

polar(all OH,acidic,basic)

acidic/-ve/anionic

glutamic acid(oxidat deaminat)

aspartic acid

basic/+ve/cationic(HisbasicAreLoose)

Histidine(Heterocyclic, most stable at physiolog pH, extra NH₂ grp)

ArGinine(most polar d/t Guanido grp)

Lysine

imino acid

proline(=NH, amino side chain)

aliphatic(GAVLI)

Glycine(smallest, simplest, flexibil prot, optic inactive, detoxif benzoic acid)

Alanine

Valine

Leucine

Isoleucine

aromat(TryToPushHer)(absorb-280nm)

Tryptophan

Tyrosine

Phenylalanine

Histidine

S containing

cysteine

methionine

hydroxyl AA(SleeperThreeTier)(can be phosphorylated by prot kinase)

Serine(d glycoSylation)

Threonine

Tyrosine

AA forming succinyl coA(MVIT)

Methionine

Valine

Isoleucine

Threonine

AA forming α KG(PHArG)

Proline

Histidine

Arginine

Glutamine

nonpolar(AVLI Met PPT)

Alanine

Valine

Leucine

Isoleucine

Methionine

Proline

Phenylalanine

Tyrosine

heterocyclic

Proline

Glycine

cystinuria(COAL)

Cysteine

Ornithine

Arginine

Lysine

Selenocysteine(21stAA-UGA)(GAS)

pyrolysine(22ndAA-UAG)

Abbreviations

a-artery, AA-amino acid, abtc-antibiotic, AI-autoimmune

bef-before, bel-below, b/l-bilateral, bld-blood, b/n-between, bn-benign, br-branch,

Bx-biopsy

ca-carcinoma, carb-carbohydrate, c/i-contraindication, c/l-contralateral,

conc-concentration, cong-congenital, Cx-cervix

d-day, def-deficient, ds-disease, d/t-due to, Dx-diagnosis

E-estrogen

fem-female, fr-from

gld-gland, glu-glucose

h-hormone

idiop-idiopathic, i/l-ipsilateral, inf-infection, inj-injury
lig-ligament, LL-lower limb, l/t-leading to
m-muscle, maj-major, mal-male, MC-most common, met-metastasis, min-minor,
mtx-methotrexate, Mx-management
n-nerve, norm-normal
P-progesterone, pl-plasma, prot-protein, pt-patient
Rx-treatment
SCC-squamous cell carcinoma, sr-serum, Sx-surgery, sz-seizure
tm-tumour, ts-tissue
UL-upper limb, u/l-unilateral
vag-vagina, VC-vocal cord, vel-velocity, vert-vertebra, vit-vitamin, vol-volume
w-week, wt-weight
Xr-X ray
y-year
#-fracture
°-degree

-(N.B.-THESE NOTES ARE ONLY FOR THE PURPOSE OF GUIDANCE AND HELP
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dr.ankityadavendra@gmail.com)