MCQ's

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I- Mark the correct statements about heme biosyntheses

1- Sideroblastic anemia is due to ALA synthase deficiency
2- In sideroblast anem serum iron is high
3- Patient with sideroblastic anemia have enlarged liver and spleen
4- Patient with sideroblastic anemia looks pale, weak
5- All of the above

II- Mark the correct statements about heme biosyntheses

Heme is a component of:

1- hemoglobin
2- myoglobin
3- liver cytochrome
4- respiratory cytochrome
5- peroxidases
6- catalases
7- 1 & 2
8- 3 & 4
9- 5 & 6
10- All of the above

III- Mark the correct statements in the following regarding heme:

1- is synthesized by every human cell
2- is synthesized mainly by liver
3- mainly synthesized in bone marrow
4- 2 & 3
5- All of the above

IV- For each numbered statement choose the correct figured one

1-	Heme biosynthesis process	A-	Results from the action of protoporphyrinogen oxidase on protoporphyrinogen
2-	Glycine plus succinyl CoA is acted upon by ALA synthase enzyme	B-	Results from the action of Coproporphyrinogen oxidase on Coproporphyrinogen III
3-	Delta amino levulonic acid	C-	Results from Ferrochelatase + Fe^{2+} enzyme deficiency
4-	Hydroxymethyl bilne synthase deficiency	D-	Results from protoporphyrinogen oxidase deficiency
5-	Porphobilinogen as a substrate	E-	Results from Coproporphyrinogen oxidase enzyme deficiency
6-	Uroporphyrinogen III synthase deficiency	F-	Results from action of UROD on Uroporphyrinogen III
7-	Hydroxymethyl belane	G-	Results from action of Uroporphyrinogen III synthase on hydroxymethjyl bilane
8-	Uroporphyrinogen decarboxylase deficiency	H-	Causes PCT
9-	Uroporphyrinogen III	I-	Results from the action of porphobilinogen deaminase on porphobilinogen
10-	Coproporphyrinogen III	J-	Leads to CEP
11-	HCP	K-	Is acted upon by hydroxymethyl bilane synthase
12-	VP	L-	Lead to AIP
13-	E.P.P.	M-	Is the substrate of ALA dehydratase enzyme
14-	Protoporphyrinogen	N-	To produce Delta ALA
15-	Protoporphyrin	O-	Is composed of 8 reactions each of which is catalyzed by a specific enzyme

V- Mark the following statements T for True and F for False

1- Hepatic erythropoetic porphyria (HEP) is AD ☐
2- Hepatic erythropoetic porphyria is due to UROD deficiency ☐
3- PCT is due to UROD deficiency ☐
4- CEP is AD ☐
5- CEP comes in acute attacks ☐
6- ALA dehydratase deficiency porphyria is not photosensitive ☐
7- AIP is photosensitive ☐
8- HEP is photosensitive ☐
9- Porphyria may present with acute attack in ALA dehydratase deficiency porphyria ☐
10- Porphyrin abnormalities occur in association with: ☐
   a- hemolytic anemia ☐
   b- liver disease ☐
   c- lead poisoning ☐
   d- Sideroblastic anemia ☐
   e- Renal failure ☐
   f- Iron deficiency ☐
11- Patient with acute attack of porphyria present with
   a- Photosensitivity ☐
   b- Diarrhea ☐
   c- Constipation ☐
   d- Abnormal cramp ☐
   e- Personality changes ☐
   f- Electrolytes are normal ☐
   g- Pain and tingling of arms and legs ☐
   h- Low blood pressure and shock ☐
   i- Gall bladder stone ☐
   j- Skin scarring ☐
12 About Erythrocoproporphyria mark the following statement T for true and F for false
   a- it is similar to congenital erythropoietic porphyria ☐
   b- it is rarest type of porphyria ☐
   c- it is seen in childhood ☐
   d- RBCS show increased Coproporphyrin and protoporphyrin ☐
   e- RBCS show iron crystals ☐
   f- It is not photosensitive ☐
   g- It comes in acute attack ☐
   h- It is a bone marrow porphyria ☐
   i- Patient may have hypochromic microcytic anemia ☐
   j- Iron treatment is useful in this porphyria ☐

VI- Name the items in the following headings:

A) Name of Bone marrow porphyrias ☐
B) Names of Hepatic porphyrias ☐
C) Name of the commonest porphyrias ☐
D) Name the acute episodes of porphyrias with no photosensitivity ☐
E) Name of acute episodes with photosensitivity ☐
F) Name Porphyrias with skin changes only ☐

VII- For each of the following numbered statements choose the correct figured answer:

1-	The porphyrin molecules by absorbing visible light	A-	Is located in cytosol
2-	The damage lead to blistering	B-	Is present in mitochondrion
3-	Burning and soreness of the skin on exposure to light	C-	Can be scanned for porphyrins by spectrofluorometry
4-	Plasmas taken from patients suffering from symptomatic porphyria	D-	Is due to accumulation of lipophilic protoporphyria
5-	ALA synthase enzyma	E-	Due to accumulation of water soluble uroporphyrins
6-	ALA dehydratase enzyme	F-	Generate free radicals and lipid peroxidation and cell membrane damage and cell death

7- In ALA dehydratase deficiency prophyria (Doss prophyria)	G- The urine shows increased ALA and coproporphyrin III
8- AIP	H- The urine shows ALA +++, PBG +++
9- CEP	I- Urine shows uroporphyrin-1 and Coproporphyrin-1
10- In PCT	J- Plasma shows uro +
11- In HEP	K- RBCS show protoporphyrin
12- Coproporphyrinogen oxidase, Protopor - phyrinogen oxidase and ferrochelatase + Fe enzymes	L- Are located in mitochondrion
13- Familial PCT	M- Is type II PCT
14- Sporadic PCT	N- Is type I PCT
15- Type I and II PCT are	O- PCT
16- Hepatocellular carcinoma and hematologic malignancy excite	P- Precipitated by hemochromotosis, iron over load and β Thalassemia
17- In PCT	Q- Are characteristic signs of PCT
18- Skin fragility hypertrichosis and plethora of central face and upper chest and shoulders	R- Uroprophyrins are excreted in urine and coproporphyrin and isocoproporphyrin in stool
19- 2 UROD gene mutations	S- Are positive to collage IV and PAS stain and are found in the roof of blister of EPP and PCT
20- Caterpillar bodies	T- Are recessively inherited in HEP
21- Liver biopsy in PCT	U- Reduces serum ferritin level and iron store
22- Plasmapheresis combined with somatostatin	V- Helps utilize iron body store in PCT
23- Desferioxamine	W- Is iron chelator and used to treat PCT
24- Erythropoietin	X- Are used to treat PCT
25- Phlebotomy	Y- The hepatocytes show needle like intracytoplasmic inclusion of uroporphyrin crystals

VIII-Mark the following statement T for true and F for false:

1- PCT improve when IFN alfa is given to treat hepatitis C ☐
2- Erythropoietin is given by mouth ☐
3- Dose of erythropoietin is 50 – 100 International units per kg body weight 3-times weekly ☐
4- Chloroquin phosphate pediatric dose is 12.5 mg/kg per os twice weekly ☐
5- Pediatric dose of plaquinil is 3mg/kg body weight per os twice weekly to treat PCT ☐
6- Phlebotomy in PCT leads to remission for 6-months up to 10-years of PCT ☐
7- Acute porphyrias are:
a- CEP ☐
b- HEP ☐
c- Erythrocoproporphyria (ECP) ☐

8- Acute porphyrias are precipitated by low carbohydrate diet ☐
9- Acute porphyrias are more common in men ☐
10- Acute porphyrias present with hypertension and arrythmas ☐
11- AIP onset is from puberty till age of 50 – 60 years ☐
12- AIP may lead to optic atrophy ☐
13- In AIP urinary retention is due to affection of sensory nerves ☐
14- Abdominal pain and dysphagia in AIP is due to autonomic nerve affection ☐
15- Sensory neuropathy in AIP lead to diffuse pain which is proximal or distal ☐
16- Acute porphyrias include:
b- AIP ☐
c- HCP ☐
d- V.P. ☐
17- Acute Porphyrias are treated with – high doses of glucose ☐
18- Hematin given in AIP may cause:
b- Anaphylaxis ☐
c- Coagulopathy ☐
19- In HCP one third of cases show photosensitivity similar to PCT ☐
20- All patients with VP have photosensitivity in sun exposed areas ☐

IX- Choose for numbered statements the correct figured one

1-	Plasma of VP	A-	Is used to treat VP
2-	Carriers of VP among patient relatives	B-	When a child screams on going outdoor in the sun
3-	86% of adult carrier of VP over age of 15	C-	Is the most common porphyria at age of 2 – 4 years
4-	35% of adult carrier of VP	D-	Is treated with intravenous hematin infusion
5-	Carbohydrate loading	E-	Have stools positive for porphyrin
6-	Variegate porphyria	F-	Show plasma fluorescence
7-	E.P.P.	G-	Are detected by finding protoporphyrinogen oxidase activity in fibroblast or lymphocyte.
8-	EPP is suspected	H-	Fluoresce at 625 – 627 nm using spectrophotometer, fitted with red sensitive photomultiplier
9-	In EPP	I-	Increases protoporphyrin excretion
10-	Hyperkeratosis and thickening and mild scarring on dorsum of hands	J-	Is used to treat EPP in the dose of 50 – 200 mg once daily
11-	By EM	K-	Is suppressed by iron and increase porphyrin synthesis
12-	Iron given	L-	Given to EPP patient exacerbate porphyria
13-	The red cell ALA synthase	M-	Iron is seen deposited in RBCS of EPP patient
14-	β caratine	N-	Is a manifestation of mild photosensitivity in EPP
15-	Cholestyramine used to treat E.P.P.	O-	Three is waxy thickening of skin of nose and knuckles

16-	Cystein in the dose of 500mg BID is given in E.P.P.	P-	Give I.V. hematin
17-	Patients with EPP not protected from visible light during operation	Q-	NBUVB (311 – 313nm) is used as a line of treatment
18-	Severe burn may occur to patients with EPP during operation	R-	Corrects the enzyme defect in EPP
19-	To increase epidermal thickening in E.P.P.	S-	Increases excretion of protoporphyrin
20-	To decrease hepatic accumulation of protoporphyrin in EPP	T-	Show erythrodontia and severe photoxic rash with mutilation of nose, ears, fingers and sun exposed areas
21-	Oral bile acid given to EPP	U-	Is AR and starts in infancy
22-	Gene treatment of ferrochelatase deficiency	V-	Causes fluorescence of teeth and RBCS
23-	CEP	W-	Urine is pink and stained nappies
24-	CEP patients	X-	If exposed to visible light
25-	Wood's light	Y-	May develop peripheral neuropathy post operative
26-	In CEP and HEP	Z-	Act as free radical scavenger

II- Mark the following statements T for true and F for false:

1- In CEP there is hemolytic anemia and hyperspleenism

2- In CEP there is thrombocytosis

3- In CEP three is scleromalacia

III- In investigating CEP complete the following:

Urine shows:

- _____
- _____
- _____

Stool shows:

- _____
- _____
- _____

RBSC show:

- _____
- _____
- _____

IV- Complete the following statements regarding treatment of CEP:

1- The solar wave length avoided to protect skin and eyes is of the wave length_____ _____

2- Oral superactivated charcoal is given to _____

3- Packed RBCS are transfused to _____ _____

4- Spleenectomy is done to _____ _____

5- Other lines of treatment include:

- _____
- _____
- _____

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Answers of MCQs (Porphyrias)

I- 5

II- 10

III- 5

IV-

1 = O	2 = N	3 = M	4 = L	5 = K
6 = J	7 = I	8 = H	9 = G	10 = F
11= E	12 = D	13= C	14 = B	15 = A

V-

1 = F	2 = T	3 = T	4 = F	5 = F
6 = T	7 = F	8 = T	9 = T	10-a = T
10-b = T	10-c = T	10-d = T	10-e = T	10-f = T
11-a = F	11-b = T	11-c = T	11-d = T	11-e = T
11-f = F	11-g = T	11-h = T	11-i = T	11-j = T
12-a = F	12-b = T	12-c = T	12-d = T	12-e = T
12-f = F	12-g = F	12-h = T	12-i = T	12-j = F

VI-

[A]- CEP, EPP, ECP
[B]- AIP, HCP, VP, PCT, HEP
[C]- ALA dehydratase deficiency porphyria
[D]- PCT, AIP, EPP
[E]- AIP, ALA dehydratase deficiency porphyria
[F]- HCP, VP
PCT, EPP, CEP, HEP

VII-

1 = F	2 = E	3 = D	4 = C	5 = B
6 = A	7 = G	8 = H	9 = I	10 = J
11 = K	12 = L	13 = M	14 = N	15 = P
16 = O	17 = R	18 = Q	19 = T	20 = S
21 = Y	22 = X	23 = W	24 = V	25 = U

VIII-

1 = T	2 = F	3 = T	4 = F	5 = T
6 = T	7-a = F	7-b = F	7-c = F	8 = T
9 = F	10 = T	11 = T	12 = T	13 = F
14 = T	15 = T	16-a = T	16-b = T	16-c = T
17 = T	18-a = T	18-b = T	19 = T	20 = T

IX-

1 = H	2 = G	3 = F	4 = E	5 = A
6 = D	7 = C	8 = B	9 = O	10 = N
11 = M	12 = L	13 = K	14 = J	15 = I
16 = Z	17 = Y	18 = X	19 = Q	20 = P
21 = S	22 = R	23 = U	24 = T	25 = V
26 = W				

X- 1 = T 2 = F 3 = T

XI-

Urine shows - (uroporphyrin I coprophyrin I)
Stools - coproporphyrin I
RBCS - Uroporphyrin I Coproporphyrin I
Protoporphyrin

XII-

(1) Wave length 360 – 500 nm
(2) Charcoal reduces entropathic porphyrin circulation
(3) Reduce porphyrins
(4) Reduce hemolysis and platelet consumption
(5) Desferrioxamine, bone marrow transplant, Gene treatment, beta carotene