M C Q's

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Mark one correct answer:

1- Alkaptonuria becomes manifest not before the third to fourth decade of life of the patient because

a) Homogentisic acid (HGA) is less produced in childhood
b) HGA is more produced in adults
c) HGA production is related to hormonal changes with advance of age
d) There is difference in the rate of excretion of HGA by kidneys

2- Fishberg test is positive when

a) urine turns black on exposure to air
b) a photographic paper turns dark when exposed to alkalinized ochronotic urine
c) when alkapton level in urine is high
d) when the diaper of an ochronotic baby is stained after being washed with soap

3- Ascorbic acid in treatment of endogenous ochronosis is characterized by:

a) is given as long term course with a dose of 100mg per Kg per day
b) reduce excretion of homogentisic acid
c) diminish serum benzoquinone acetic acid
d) inhibit oxidation, polymerization and binding of homogentisic acid to collagen
e) all of the above
f) none of the above

4- The main cause of exogenous ochronosis is:

a) The use of high concentration hydroquinone cream
b) Affect white skin more than dark skin
c) Systemic deposition of Ochre pigment in the skin
d) Extended use of bleaching creams
e) All of the above

5- What agents could produce exogenous ochronosis:

a) hydroquinone creams and topical phenal
b) Topical mercury preparations and Topical picric acid and Topical resorcinol
c) Quinine injection and anti malarial drugs
d) All of the above
e) None of the above

6- Exogenous ochronosis is to be differentiated from which of the following:

a) endogenous ochronosis
b) localized argyria
c) therapy with levodopa and methyldopa
d) minocycline pigmentation
e) all of the above

7- Hereditary alkaptonuria affects approximately:

a) one in 6000
b) one in 16000
c) one in 100,000
d) one in a million

8- Hereditary alkaptonuria is:

a) autosomal dominant
b) x-linked dominant
c) autosomal recessive
d) x-linked recessive

9- Homogentisic acid is a metabolite of:

a) Homocysteine
b) Ornithine
c) Phenyle alanine and tyrosine
d) Aminoglycans

10- Alkaptonuria may be diagnoses by:

a) Benedict reagent
b) acidifying the urine
c) adding homogentisic acid oxidase to urine
d) exposing urine to ultra violet light

11- Oslers sign in ochronosis is:

a) Palmoplantar pigmentation with thickening and pitting
b) bluish hyperpigmentation of medial and lateral aspects of sclera of both eyes.
c) Bluish pigmentation of the auricle with black cerumen
d) Bluish nail discoloration with black sweat

12- Treatment of ochronosis includes:

a) 4% hydroquinone topically
b) Topical hydrogen peroxide
c) Q-switched laser
d) Systemic steroid

13- The alkapton is:

a) 2, 5 hydroquinone acetic acid
b) Fumaric acid
c) Maloyl acetoacetic acid
d) Porphobilinogen

14- Hereditary alkaptonuria is due to:

a) Catalase deficiency
b) Homogentisic acid oxidase deficiency
c) Protease deficiency
d) Tryptase deficiency

15- Normal catabolism of Phenylalamine end in formation of

a) uric acid
b) Maloylaceto acetic acid
c) Melanin formation
d) Yellow pigment formation

16- High performance liquid chromolography is used to detect:

a) Maloylaceto acetic acid in plasma
b) Homogentisic acid oxidase enzyme level in plasma
c) Homogentisic acid level in plasma
d) Phenol oxidase enzyme activity in the skin

17- Magnetic resonance spectroscopy is used to detect homogentisic acid level in

a) aortic valve
b) in urine
c) in vertebral disk
d) in articular cartilage

18- Mark the following statements T for true / F for false

a) high dorsal radicular medullary compression is seen in a patient with ochronotic arthropathy
(T) (F)

b) Human gene for alkaptonuria is on X chromosome
(T) (F)

c) Human gene for endogenous alkaptonuria is on chromosome 3q23
(T) (F)

d) Stenosis of aortic valve is reported in ochronosis
(T) (F)

e) Ochronosis does not affect sclera
(T) (F)

19- The clinical features of alkaptonuria are characterized by the following – Mark T for true F for false

a) manifestations are related to age
(T) (F)

b) at birth there is dark urine and dark cerumen
(T) (F)

c) axillary pigmentation is seen in infants
(T) (F)

d) Ear lobe pigmentation is seen at age of 10-15 years
(T) (F)

e) Pigmentation of sclera at age of 20-40
(T) (F)

f) Aortic stenosis is seen at age of 20-years
(T) (F)

g) Ochronotic arthropathy is seen at age of 30-40 years
(T) (F)

h) Renal failure is rare in late stage Ochronosis
(T) (F)

i) Ochronosis should be considered in low back ache in young individuals
(T) (F)

j) Prostatic lithrosis and renal stones with obstruction of urinary tract is a known complication of ochronosis
(T) (F)

20- Mark the following statements: T for true / F for false

a) Long term Ascorbic acid is a known treatment for achronosis
(T) (F)

b) differential diagnosis of ocular ochronosis includes melanoma
(T) (F)

c) Restriction of protein intake to 1 gram per kg per day is recommended in treatment of ochronosis
(T) (F)

d) Substitution therapy with recombinantly obtained homogentisic acid oxidase enzyme is the aim in treatment of ochronosis
(T) (F)

e) impaired cellular immune function is reported with ochronotic valvular disease
(T) (F)

f) Ochronosis leads to blue nail discoloration
(T) (F)

g) The extensive pigmentation in ochronosis is due to melanin pigment
(T) (F)

h) Ochronosis is associated with palmoplantar pigmentation and thickening
(T) (F)

i) The urine of patients with alkaptonuria becomes dark because of formation of 2,5 hydroxyquinone acetic acid
(T) (F)

Answers of the MCQ's :

1 - (d) 2 - (b) 3 - (e) 4 - (d)
5 - (d) 6 - (e) 7 - (d) 8 - (c)
9 - (c) 10- (a) 11- (b) 12- (c)
13- (a) 14- (b) 15- (b) 16- (c) 17- (b)
18- a (T) b (F) c (T) d (T) e (F)
19- a (T) b (T) c (F) d (F) e (T)
f (F) g (T) h (T) I (T) J (T)
20- a (T) b (T) c (T) d (T)
e (T) f (T) g (F) h (T)
I (T)