

## MCQ's

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Mark the correct answer in the following :

**1. All of the following are true about Cholesterol Sulfate except:**

- a) Cholesterol Sulfate comprise 50% of the total lipid of stratum granulosum in normal skin.
- b) Cholesterol sulfate is lower in the outer stratum corneum than it is in the whole horny layer.
- c) In x-linked recessive ichthyosis (XI) the steroid sulfatase enzyme activity is absent in all nucleated epidermal layer and in the stratum corneum, resulting in accumulation of cholesterol sulfate in the skin.
- d) The scales in (XI) contain five folds increase in cholesterol sulfate and a 50% decrease in free sterol content.

**2. Which of the following is true about Harlequin ichthyosis (HI):**

- a) It is an autosomal dominant with chance of 1:2 of each subsequent child to become affected.
- b) HI fetus are affected in utero and prenatal ultrasonographic diagnosis is not possible.
- c) Prenatal diagnosis (PND) of HI is helped by fetal skin biopsy at 21-22 weeks of gestational age.
- d) The characteristic abnormalities of HI are known to be more expressed in the hair canal.
- e) c and d are correct.

**3. HI is characterized at birth by :**

- a) Polydactyly and renal abnormalities, micromelia, Patent ductus arteriosus and enlarged thymus.

- b) Ectropion, eclabium, distortion of the nose and absence of ears or rudimentary canal, swollen and crumpled hands and feet.
- c) Examination of blood smear from all cases of ichthyosis as a screening test reveals presence of lipid vacuoles in Red-blood cells.
- d) All of above is correct.
- e) a and b are correct

**4. The characteristic abnormality of (HI) are known to be more expressed in the hair canal, which of the following is correct:**

- a) Electron microscopic (EM) examination of fetal skin biopsy demonstrated abnormal vacuoles in keratinized cells
- b) Malformation of lamellar granules in the hair canal
- c) Clumps of aberrant keratinized cells containing lipid droplets in the amniotic fluid
- d) Immunoblast study of epidermal extracts revealed the filaggrin to be more prominent than profilaggrin in all the hairy skin regions where hair canals were extensively keratinized
- e) a, b and c are correct
- f) All of the above are correct

**5. Early prenatal diagnosis of Harlequin ichthyosis (HI) is possible at:**

- a) Prenatal diagnosis (PND) of (HI) is possible at 19 weeks of expected gestational age (EGA).
- b) PND of (HI) is possible at 16 weeks of (EGA)
- c) PND of (HI) is possible at 22 weeks of (EGA)
- d) All of the above are correct.

**6. Choose the correct answer**

- a) Lack of desquamation in (HI) is due to a defect in lamellar body secretion.

- b) Lamellar granules are lipid rich organelles present in the stratum corneum.
- c) The keratinocytes contained giant mitochondria which may be related to abnormal lipid metabolism of keratinocytes which affect composition of lamellar granules.
- d) Keratinocytes in (HI) have reduced activity of the serine / threonine protein phosphatase
- e) All of the above are correct
- f) a, c, and d are correct

**7. Study of the epidermal protein shows that**

- a) K5 & K14 are present in cells of the stratum basale
- b) K1 & K10 in stratum corneum cells
- c) K6 & K16 are decreased in hyperproliferative conditions
- d) Profilaggrin is a major constituent of keratohyaline in the granular cell layer
- e) a, b and c are correct
- f) a & d are correct

**8. The causative genetic defect which was found in (HI) include:**

- a) Transaminase 1 gene mutation
- b) Abnormal lamellar granules, and filaggrin processing abnormalities
- c) a and b are correct
- d) a is correct
- e) b is correct

**9. All of the following are true about protein phosphatase 2A (PP2A) except:**

- a) PP2A acts on profilaggrin and cleaves it to filaggrin during transition from granular layer to horny layer.
- b) The PP 2A conversion of phosphorylated profilaggrin to nonphosphorylated filaggrin is blocked in (HI)
- c) Keratinocytes from (HI) showed lower protein phosphatase activity in culture
- d) All of the above are correct
- e) Only a and b are correct

**10. Protein phosphatase 2A (PP2A):**

- a) Regulates cholesterol synthesis by activating the enzyme 3 hydroxy 3 methyl glutaryl coenzyme A reductase (HMGCoA).
- b) PP2A could be a link between lamellar granule synthesis and profilaggrin processing.
- c) The gene that controls PP2A is located in Chromosome 16.
- d) All of the above are correct.
- e) Only a and b are correct

**11. (HI) was found to evolve to :**

- a) Erythrodermic ichthyosis (non-bullous) congenited ichthyosiform erythroderma) (NBCIE)
- b) Lamellar ichthyosis (LI)
- c) Ichthyosis Vulgaris (IV)
- d) Bullous Ichthyosis

Answers to MCQ's		
1. (a)	2. (e)	3. (e)
4. (e)	5. (a)	6. (f)
7. (f)	8. (c)	9. (d)
10. (e)	11. (a)	