

Section VII: ECM

Extracellular Matrix

1. The most abundant protein in the human body is:

- (a) Elastin
- (b) Fibrillin
- (c) Collagen
- (d) Fibronectin

2. Collagens assume a gel-like consistency in:

- (a) Vitreous humor
- (b) Tendons
- (c) Bones
- (d) Cornea

3. The following is *not* true for collagens:

- (a) Fibrous proteins
- (b) Extracellular
- (c) Glycoproteins
- (d) Lack quaternary structure

4. Basement membranes mainly contain collagen type:

- (a) III
- (b) IV
- (c) VII
- (d) XII

5. Tendons and ligaments mainly contain collagen type:

- (a) III
- (b) IV
- (c) VII
- (d) XII

6. Fetal skin mainly contains collagen type:

- (a) III
- (b) IV
- (c) VII
- (d) XII

7. The commonest amino acid in collagens is:

- (a) Glycine
- (b) Proline
- (c) Hydroxyproline
- (d) Lysine

8. Collagens are synthesized by:

- (a) Osteoblasts
- (b) Chondroblasts
- (c) Fibroblasts
- (d) All of the above

9. The formation of interchain H-bonds in collagens depends on the presence of:

10. The formation of interchain covalent bonds in collagens depends on the presence of:

11. Lysyl oxidase requires:

12. Lysyl hydroxylase requires:

13. The following is *true* for extension peptides in pro-collagen:

- (a) Completely retained in mature collagens
 - (b) Removed intracellularly
 - (c) Removed extracellularly
 - (d) Partly retained in mature collagens

14. Osteogenesis imperfecta involves collagen type:

- (a) I (b) II (c) III (d) IV

15. A common cause of death in type II osteogenesis imperfecta is:

- (a) Fractures
 - (b) Epistaxis
 - (c) Hematuria
 - (d) Pulmonary hypoplasia

16. Desmosine cross-links are found in:

17. In elastin, every one in seven amino acids is:

- (a) Glycine (b) Lysine (c) Proline (d) Valine

18. In collagens, every one in three amino acids is:

- (a) Glycine (b) Lysine (c) Proline (d) Valine

19. The elasticity of elastin is due to:

- (a) Desmosine cross-links
- (b) Valine-rich domains
- (c) Both
- (d) None of the above

20. The following is *true* for Marfan's syndrome:

- (a) Floppy mitral valves
- (b) Arachnodactyly
- (c) Fibrillin gene mutation
- (d) All of the above

21. The following is *not true* for keratan sulphate:

- (a) Most homogeneous group of glycosaminoglycans
- (b) Keratan sulphate-I is found in cornea
- (c) May be N-linked or O-linked with proteins
- (d) Sulphate content is variable

22. Epimerization at C5 of glucuronate forms:

- | | |
|----------------|-----------------|
| (a) Iduronate | (b) Gulonate |
| (c) Guluronate | (d) Mannuronate |

23. The enzyme deficient in Hunter syndrome is:

- (a) Iduronate sulphatase
- (b) α -L-Iduronidase
- (c) Galactose-6-sulphatase
- (d) β -Galactosidase

24. The enzyme deficient in Hurler syndrome is:

- (a) Iduronate sulphatase
- (b) α -L-Iduronidase
- (c) Galactose-6-sulphatase
- (d) β -Galactosidase

25. The enzyme deficient in Morquio syndrome A is:

- (a) Iduronate sulphatase
- (b) α -L-Iduronidase
- (c) Galactose-6-sulphatase
- (d) β -Galactosidase

26. The enzyme deficient in Morquio syndrome B is:

- (a) Iduronate sulphatase
- (b) α -L-Iduronidase
- (c) Galactose-6-sulphatase
- (d) β -Galactosidase

27. The enzyme deficient in Sanfilippo syndrome A is:

- (a) Heparan sulphamidase
- (b) N-Acetyl glycosaminidase
- (c) Acetyl CoA: Glucosaminide N-acetyl transferase
- (d) N-Acetyl glucosamine-6-sulphate sulphatase

28. The enzyme deficient in Sanfilippo syndrome B is:

- (a) Heparan sulphamidase
- (b) N-Acetyl glycosaminidase
- (c) Acetyl CoA: Glucosaminide N-acetyl transferase
- (d) N-Acetyl glucosamine-6-sulphate sulphatase

29. The enzyme deficient in Sanfilippo syndrome C is:

- (a) Heparan sulphamidase
- (b) N-Acetyl glycosaminidase
- (c) Acetyl CoA: Glucosaminide N-acetyl transferase
- (d) N-Acetyl glucosamine-6-sulphate sulphatase

30. The enzyme deficient in Sanfilippo syndrome D is:

- (a) Heparan sulphamidase
- (b) N-Acetyl glycosaminidase
- (c) Acetyl CoA: Glucosaminide N-acetyl transferase
- (d) N-Acetyl glucosamine-6-sulphate sulphatase

31. The enzyme deficient in Sly syndrome is:

- (a) β -Glucuronidase
- (b) β -Glucosidase
- (c) β -Galactosidase
- (d) β -Galacturonidase

32. The following is a podocyte cell surface protein:

- (a) Nephrin
- (b) Podocin
- (c) Both
- (d) None of the above

33. The following is *not* a feature of Liddle's syndrome:

- (a) Abnormal apical membrane Na^+ -channel in renal cortical collecting ducts
- (b) Hypotension
- (c) Hypokalemia
- (d) Metabolic alkalosis

34. Skeletal muscle membrane Na^+, K^+ -ATPase is stimulated by:

- (a) Insulin
- (b) Epinephrine
- (c) Both
- (d) None of the above

35. Digitalis over dose causes hyperkalemia by:

- (a) Inhibiting muscle Na^+, K^+ -ATPase
- (b) Stimulating muscle Na^+, K^+ -ATPase
- (c) Inhibiting liver Na^+, K^+ -ATPase
- (d) Stimulating liver Na^+, K^+ -ATPase

36. The following is an adaptor protein:

- (a) Spectrin
- (b) Ankyrin
- (c) Actin
- (d) Calmodulin

37. Hereditary spherocytosis is due to defective:

- (a) Ankyrin R
- (b) Ankyrin B
- (c) Ankyrin G
- (d) None of the above

38. Spectrin and ankyrin function together to:

- (a) Maintain RBC shape
- (b) Expression of antigens on T lymphocytes
- (c) Assembly of axonal membrane ion transporters
- (d) All of the above

39. Defect in RBC membrane glycosylphosphatidylinositol results in:

- (a) Paroxysmal nocturnal hemoglobinuria
- (b) Sickle cell anemia
- (c) α -Thalassemia
- (d) β -Thalassemia

40. Extracellular fibrillar amyloid deposits in Alzheimer's disease are rich in:

- (a) Amylin
- (b) Amyloid β -peptide ($A\beta 42$)
- (c) Amyloid precursor protein
- (d) α -Synuclein

41. Which of the following organelle is involved with glycosylation and sorting of proteins?

- (a) Endoplasmic reticulum
- (b) Golgi apparatus
- (c) Mitochondria
- (d) Peroxisomes

42. Which of the following acts as a signal for transporting enzymes to lysosomes?

- (a) KDEL sequence
- (b) Signal peptide sequence
- (c) Mannose-6-phosphate
- (d) COPI vesicles

43. Which of the following protein is involved with import and export of various macromolecules across the nucleus?

- (a) Heat shock protein Hsp 70
- (b) Rho protein
- (c) Ran GTPsase
- (d) Cyt P450

44. Which of the following is *not* true about Zellweger syndrome?

- (a) Accumulation of very long fatty acids
- (b) Defect in bile acid synthesis
- (c) Reduction of plasmalogens
- (d) Accumulation of phytanic acid

45. Which of the following is *not* a peroxisomal abnormality?

- (a) Zellweger syndrome
- (b) Neonatal adrenoleukodystrophy
- (c) Infantile Refsum disease
- (d) I-cell disease

46. All of the following is true about Ubiquitinated protein degradation process except.

- (a) It is ATP dependent process
- (b) Ubiquitin dependent degradation of proteins occurs in the proteasomes
- (c) Linkage of ubiquitin always mark for degradation of that protein
- (d) Ubiquitin is recycled after the process

Answer key

1. c	2. a	3. d	4. b	5. d	6. a
7. a	8. d	9. c	10. d	11. d	12. d
13. c	14. a	15. d	16. b	17. d	18. a
19. c	20. d	21. a	22. a	23. a	24. b
25. c	26. d	27. a	28. b	29. c	30. d
31. a	32. c	33. b	34. c	35. a	36. b
37. a	38. d	39. a	40. b	41. b	42. c
43. c	44. d	45. d	46. c		