

How to Use This Series

Chapter Head

1 Cell Biology

Question Difficulty Key

Green box = Easy question
Yellow box = Medium question
Red box = Hard question

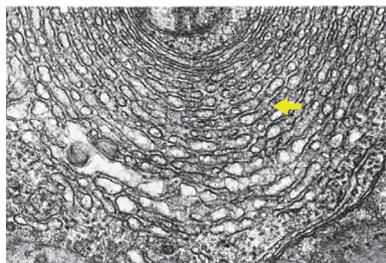
Section Header

1.1 Questions

Easy Medium Hard

Consider the following case for questions 1 to 2:

A 26-year-old woman presents to the clinic with complaints of severe abdominal pain. She is known to be 6 weeks pregnant and did not have any problems with previous pregnancies. An ultrasound is performed and demonstrates a corpus luteum cyst. The following figure is obtained from the biopsy of the cyst following surgical exploration.



Question Stem

1. Based on the predominance of the organelle indicated by the arrow within the cell in the figure, which other cell in the body might have a similar microscopic appearance?

- A. Goblet cell
- B. Adrenal cortical cell
- C. Exocrine pancreatic cell
- D. Parotid glandular cell
- E. Lacrimal glandular cell

2. What is the function of the organelle indicated by the arrow within the cell in the photomicrograph?

- A. Synthesis of proteins
- B. Post-translational modification of proteins
- C. Storing and packaging of proteins
- D. Synthesis of steroid hormones
- E. β -oxidation of fatty acids

Answer Options

3. A 44-year-old woman presents with flaccid blisters filled with clear fluid that arise from otherwise healthy skin. She reports painful oral mucosal erosions that preceded the cutaneous blisters. Histopathology, from the edge of a blister, shows suprabasal epidermal cells separated from each other and from the basal cells to form clefts and blisters. Direct immunofluorescence (DIF) testing performed on perilesional skin shows intercellular deposition of immunoglobulin G (IgG) throughout the epidermal skin. Which of the following proteins are targets of these autoantibodies in the patient?

- A. Keratin
- B. Claudin
- C. Integrin
- D. Desmoglein
- E. Connexin

4. A 36-year-old male presents with chest pain that started three days ago. Physical examination reveals abnormally long limbs relative to the trunk, arachnodactyly, joint laxity, thoracic scoliosis, pectus excavatum, and a diastolic murmur. Which of the following proteins is most likely defective in this patient?

- A. Cytoskeletal protein
- B. Extracellular matrix protein
- C. Adhesive protein
- D. Nucleoprotein
- E. Membrane-bound protein

5. A 32-year-old male presents with hypertension and signs of arterial insufficiency in the lower limbs. His father and a paternal uncle died from coronary vascular disease. His lab test results came back with elevated total and low-density lipoprotein (LDL) cholesterol and reduced high-density (HDL) cholesterol levels. Which of the following proteins is most likely defective in this patient?

- A. Cytoskeletal protein
- B. Extracellular matrix protein
- C. Adhesive protein
- D. Nucleoprotein
- E. Membrane-bound protein

2

Difficulty Level Icon

In general, an easy question requires only a diagnosis based upon the question stem.

A medium question requires not just the diagnosis of the condition based upon the question stem, but knowledge about that diagnosis so as to answer a question about it.

Hard questions are a combination of medium questions with less commonly known material about the diagnosis.

1.2 Answers and Explanations

Easy Medium Hard

1. Correct: Adrenal cortical cell (B)

The arrow indicates smooth or agranular endoplasmic reticulum (SER). This usually takes the form of a tightly woven network of branched tubules with no ribosomes attached [point of difference with the rough endoplasmic reticulum (RER)]. RER proliferates to SER, where the synthesis of lipid and steroid molecules occurs. Therefore, there is pronounced expansion of SER in steroid hormone-producing cells, such as the one in the figure, obtained from corpus luteum (progesterone-secreting granulosa lutein cells).

Cells of the adrenal cortex have a characteristic steroid-synthesizing cell structure. These cells produce mineralocorticoids, glucocorticoids, and androgens.

Goblet cell (A) is a mucus-producing cell with pronounced RER and perinuclear Golgi complex, with typical apical secretory granules containing mucinogen. Exocrine pancreatic cell (C), parotid glandular cell (D), and lacrimal glandular cell (E) are protein (enzyme)-secreting cells characterized by abundant RER and pronounced perinuclear Golgi complex. These cells also feature apical secretory granules.

2. Correct: Synthesis of steroid hormones (D)

Synthesis of steroid hormones is an important function of SER. It also plays major roles in lipid synthesis (including cholesterol) and metabolism of xenobiotics (drugs, carcinogens, etc.).

Synthesis of proteins (A) is the function of RER. Post-translational modification (B) and storing and packaging (C) of proteins are functions of the Golgi apparatus. β -oxidation of fatty acids (E) occurs in the mitochondria, although those with very long chains are oxidized in the peroxisomes.

3. Correct: Desmoglein (D)

The facts that the suprabasal epidermal cells are separated from each other and from the basal cells and that there is intercellular deposition of antibodies (IgG) throughout the epidermis point toward disruption of epithelial cell junctions in the lateral domain. The patient is suffering from pemphigus vulgaris (PV), which is a mucocutaneous blistering disease that predominantly affects patients > 40 years of age. Patients with PV have IgG autoantibodies to desmogleins, transmembrane desmosomal proteins that belong to the cadherin family of calcium-dependent adhesion molecules.

Keratin (A) is an intermediate filament present in epithelial cells, and mutations of keratin genes cause epidermolysis bullosa simplex (EBS), a disease group

characterized by intraepidermal blistering. The common EBS types are dominantly inherited and present at birth, in infancy, or at the latest during early childhood. Claudin (B) is a transmembrane protein that forms occluding junctions (zonula occludens) in epithelial cells. These confer tightness or leakiness and help establish functional domains (apical versus lateral) in epithelia. Integrin (C) is a transmembrane protein that forms hemidesmosomes, anchoring junctions that affect the basal domain of epithelial cells. A disruption in these junctions would result in a dermal-epidermal dissociation, as occurs in bullous pemphigoid. Connexin (E) is a unit protein that forms nexus or gap junctions. These are communicating junctions involved in molecular transport between adjacent cells that need to be highly coordinated. These do not directly participate in cell anchorage.

4. Correct: Extracellular matrix protein (B)

The patient presents with classical features of Marfan's syndrome. The defect is due to a mutation of the *FBN1* gene that codes for the protein fibrillin. Fibrillin is an essential extracellular matrix protein that forms microfibrils and plays a role in subsequent assembly of elastic fibers. Abnormalities in this protein cause distinct clinical problems involving multiple systems, of which the musculoskeletal, cardiac, and ocular predominate.

Cytoskeletal proteins (actin, etc.) (A), adhesive proteins (desmosomal and hemidesmosomal proteins, etc.) (C), nucleoproteins (histones, transcription factors, etc.) (D), or membrane-bound proteins (ion channels, receptors, etc.) (E) are not involved in the pathophysiology of Marfan's syndrome.

5. Correct: Membrane-bound protein (E)

Familial hypercholesterolemia (FH) is an autosomal dominant disorder that causes severe elevations in total and LDL cholesterol, with decreased HDL. It is associated with a high risk for premature coronary artery disease. It is a disorder of absent or defective LDL receptors, the gene for which is located on the short arm of chromosome 19. The LDL receptor (LDLR) gene family consists of cell surface proteins involved in receptor-mediated endocytosis of specific ligands. LDL is normally bound at the cell membrane and taken into the cell, ending up in lysosomes, where the protein is degraded and the cholesterol is made available for repression of microsomal enzyme HMG CoA reductase, the rate-limiting step in cholesterol synthesis.

Cytoskeletal proteins (actin, etc.) (A), extracellular matrix proteins (collagen, elastin, etc.) (B), adhesive proteins (desmosomal and hemidesmosomal proteins, etc.) (C), or nucleoproteins (histones, transcription factors, etc.) (D) are not involved in the pathophysiology of FH.

Indicates Question Difficulty

Correct Answer

Correct Answer Explanation

Incorrect Answer Explanation