

Case 2 MCQs

Please refer to both the case study and required reading article to assist you in answering these questions.

Question 1

Which of the following is NOT a feature of iridocorneal endothelial syndrome?

- A. **Posterior polar cataract**
- B. Peripheral anterior synechiae
- C. Elevated IOP
- D. Hammered silver appearance to corneal endothelium
- E. Glaucomatous optic neuropathy

Correct answer question 1: A

Question 2

Which of the following is NOT a variant of ICE syndrome?

- A. Progressive iris atrophy
- B. Chandler syndrome
- C. **Iridoschisis**
- D. Cogan Reese Syndrome
- E. None of the above

Correct answer question 2: C

Question 3

Which cell type is abnormal in ICE syndrome?

- A. Keratocytes
- B. Corneal epithelium
- C. Trabecular meshwork
- D. **Corneal endothelium**
- E. Descemet's membrane

Correct answer question 3: D

Question 4

Regarding Chandler syndrome, which statement is false?

- A. Glaucoma tends to be less severe than in other variants of ICE syndrome
- B. It is the most common ICE syndrome presentation
- C. Iris atrophy is universally present**
- D. The patient may present with blurred vision
- E. Corneal oedema may be observed

Correct answer question 4: C

Question 5

Which of the following is a mechanism of raised IOP in ICE syndrome?

- A. Anterior bowing of the iris due to the formation of posterior synechiae
- B. Hypersecretion of aqueous humour
- C. Mature cataract causing angle closure
- D. Proliferating endothelial cells obstructing the trabecular meshwork**
- E. Impaired drainage via the uveoscleral pathway

Correct answer question 5: D

Question 6

Which condition is associated with a prominent Schwalbe's line?

- A. Axenfeld-Rieger syndrome**
- B. Acute anterior uveitis
- C. Fuchs' endothelial dystrophy
- D. Posterior polymorphous dystrophy
- E. Epithelial basement membrane dystrophy

Correct answer question 6: A

Question 7

What is the inheritance pattern of ICE syndrome?

- A. Autosomal dominant
- B. Autosomal recessive
- C. X-linked recessive
- D. Mitochondrial
- E. It is not inherited**

Correct answer question 7: E

Question 8

ICE syndrome more commonly presents:

- A. Unilaterally in children in their first decade
- B. Unilaterally in women in their third to fifth decade**
- C. Bilaterally in men over 80
- D. Bilaterally in teenage boys
- E. Bilaterally in middle age, with no sex predilection

Correct answer question 8: B

Question 9

Posterior polymorphous dystrophy is generally inherited via an autosomal dominant pattern

TRUE or FALSE

Correct answer question 9: TRUE

Question 10

The aetiology of ICE syndrome is not fully understood. Which of the following has been hypothesized to be a causative agent?

- A. Fungus
- B. Virus**
- C. Bacteria
- D. Protozoa
- E. Radiation exposure

Correct answer question 10: B

Question 11

Which of the following treatment modalities would be most effective in treating raised IOP in early glaucoma associated with ICE syndrome?

- A. Argon laser trabeculoplasty 360 degrees
- B. Selective laser trabeculoplasty to the unaffected trabecular meshwork**
- C. Topical rho-kinase inhibitors

- D. Cypass Micro-Stent
- E. Inhibition of aqueous secretion with topical antihypertensive therapy**

Correct answer question 11: E

Question 12

Trabeculectomy surgery with mitomycin C has a higher success rate in ICE syndrome than in POAG?

True or **False**

Correct answer question 12: FALSE

Question 13

Which of the following disorders is associated with the lowest risk of glaucoma?

- A. Peters anomaly
- B. Chandler syndrome
- C. Axenfeld-Rieger anomaly
- D. Fuchs endothelial dystrophy**
- E. Cogan-Reese syndrome

Correct answer question 13: D

Question 14

Which of the following topical anti-glaucoma medications would be most appropriate in a patient with ICE syndrome?

- A. Latanoprostene bunod
- B. Pilocarpine 4%
- C. Timolol 0.5%**
- D. Netarsudil 0.02%
- E. Bimatoprost 0.03%

Correct answer question 14: C

Question 15

Approximately what percentage of people with ICE syndrome develop glaucoma?

- A. <5%
- B. 12-25%
- C. 28-42%
- D. 46-82%**
- E. >85%

Correct answer question 15: D

Question 16

In psuedopolyopia, each pupil has its own sphincter muscle

TRUE OR FALSE

Correct answer question 16: False

Question 17

Which of the following statements about diffuse iris melanoma is false?

- A. Sentinel scleral vessels may be present
- B. Peripheral anterior synechiae are rare
- C. They tend to be bilateral**
- D. It is a differential diagnosis for Cogan-Reese syndrome
- E. The surface of the iris tends to be thick and dark

Correct answer question 17: C

Question 18

Iris reconstruction may help to alleviate visual and cosmetic symptoms in patients with progressive iris atrophy

TRUE or FALSE

Correct answer question 18: TRUE

Question 19

Which of the following is a glaucoma drainage device that can be used in the surgical treatment of patients with ICE syndrome?

- A. Baerveldt tube

- B. InnFocus Micro Shunt
- C. Molteno Tube
- D. Ahmed valve
- E. **All of the above**

Correct answer question 19: E

Question 20

Which of the following is NOT a differential diagnosis for Progressive iris atrophy?

- A. Axenfeld-Reiger Syndrome
- B. **Diffuse iris melanoma**
- C. Iridoschisis
- D. Aniridia
- E. Chandler syndrome

Correct answer question 20: B

Question 21

What is iridoschisis?

- A. Sloughing off of the posterior layers of iris stroma via irido-zonular contact
- B. Hypoplasia of the iris sphincter
- C. Progressive enlargement of the pupil due to iris atrophy
- D. Pupillary miosis resulting from the use of parasympathetic agonists
- E. **Progressive separation of the anterior layers of the iris stroma**

Correct answer question 21: E

Question 22

Regarding the histopathology of ICE syndrome, please select the statement that is TRUE

- A. On light microscopy, abnormal endothelial cells are shorter than adjacent normal cells and form a monolayer
- B. Abnormal endothelial cells have low metabolic activity
- C. Electron microscopy in progressive iris atrophy shows normal basement membrane on the surface of the trabecular meshwork
- D. **In Cogan-Reese syndrome, iris nodules are only found in the endothelialised portion of the iris**
- E. Scanning electron microscopy in Cogan-Reese syndrome shows a continuous sheet of regular cuboid endothelial cells on the iris surface

Correct answer question 22: D

Question 23

What is the reported male to female ratio in progressive iris atrophy?

- A. **1:2 – 1:5**
- B. 1:6 – 1:10
- C. 3:1 – 5:1
- D. 1:20
- E. 8:1

Correct answer question 23: A

Question 24

Which of the following statements regarding *in vivo* corneal confocal microscopy and Chandler Syndrome is FALSE?

- A. Corneal confocal microscopy is superior to specular microscopy in the presence of corneal oedema
- B. **Corneal changes on confocal microscopy are limited to the endothelial layer**
- C. Corneal confocal microscopy can be used for both static and dynamic imaging
- D. There is loss of normal hexagonality of the endothelial cells
- E. Hyper-reflective nuclei may be visible

Correct answer question 24: B

Question 25

Which of the following can be used to treat corneal oedema or consequences of corneal oedema in ICE syndrome?

- A. IOP lowering with topical aqueous suppressant medication
- B. Hypertonic saline solution
- C. Penetrating keratoplasty
- D. Descemet' stripping endothelial keratoplasty
- E. **All of the above**

Correct answer question 25: E