

1. **Ocular manifestations of Fabry disease may involve:**
  - A. Cornea
  - B. Conjunctiva
  - C. Retina
  - D. All of the above
2. **Which of the following statements regarding the corneal lesion (cornea verticillata) in Fabry disease are true?**
  1. The corneal swirls rarely impair vision.
  2. The corneal findings are more readily appreciated in affected males than in carrier females (heterozygotes).
  3. The corneal lesions may resolve with enzyme replacement therapy.
  4. Corneal swirls are pathognomonic for Fabry disease.
  - A. 1 and 3 only
  - B. 2 and 4 only
  - C. 1, 2, and 3
  - D. 4 only
3. **Frequent systemic complications of Fabry disease include all of the following except:**
  - A. Exercise intolerance
  - B. Stroke
  - C. Tremor
  - D. Renal failure
4. **Enzyme replacement therapy for Fabry disease has been shown to be of benefit for:**
  - A. Retinal vascular tortuosity
  - B. Impairment of renal function
  - C. Angiokeratoma corporis diffusum
  - D. There are no benefits for Fabry disease with enzyme replacement therapy.
5. **Fabry disease follows which hereditary pattern?**
  - A. Autosomal dominant with high penetrance
  - B. Autosomal dominant with low penetrance
  - C. Autosomal recessive
  - D. X-linked
6. **Childhood symptoms of Fabry disease include all of the following except:**
  - A. Pain in the hands and feet
  - B. Seizures
  - C. Fatigue
  - D. Postprandial gastrointestinal pain
7. **Symptoms of Fabry disease occurring in adolescence include all of the following except:**
  - A. Blindness
  - B. Gastrointestinal distress
  - C. Angiokeratomas on the trunk
  - D. Microalbuminuria or proteinuria
8. **Which of the following is the most common ocular clinical manifestation of Fabry disease?**
  - A. Blindness
  - B. Increased tortuosity of the conjunctival vasculature
  - C. Corneal verticillata
  - D. Swelling of the eyelids
9. **Which of the following statements is accurate?**
  - A. Neuro-ophthalmic presentations, ranging from nystagmus to diplopia, usually due to extraocular muscle palsies that accompany Fabry disease, have rarely been reported in the literature.
  - B. Neuro-ophthalmic presentations, ranging from nystagmus to diplopia, usually due to extraocular muscle palsies that accompany Fabry disease, have been over-reported in the literature.
  - C. Neuro-ophthalmic presentations, ranging from nystagmus to diplopia, usually due to extraocular muscle palsies that accompany Fabry disease, have often been reported in the literature.
  - D. None of the above statements is accurate.
10. **Patients diagnosed with Fabry disease should undergo all except which of the following tests during a baseline exam?**
  - A. Visual acuity assessment
  - B. 24-2 SITA standard test
  - C. Slit lamp exam
  - D. Dilated fundus exam
11. **Confounding diagnoses for Fabry disease include all of the following except:**
  - A. Corneal deposits from available drugs such as amiodarone hydrochloride, chloroquine, chlorpromazine or indomethacin
  - B. Eyelid edema caused by cardiac or renal disease
  - C. Conjunctival vessel changes caused by hereditary hemorrhagic telangiectasia and ataxia telangiectasia
  - D. Diabetes
12. **Fabry disease may result in all of the following ocular complications except:**
  - A. Poorer prognosis in patients who undergo penetrating keratoplasty
  - B. Disc edema from uncontrolled hypertension
  - C. Anterior ischemic optic neuropathy
  - D. Blindness
13. **Barriers to educating patients with Fabry disease include all of the following except:**
  - A. Lack of familiarity with the disease
  - B. Denial as a result of a lack of symptoms
  - C. Understanding the complex nature of genetic disorders
  - D. All of the above are barriers to patient education.

**14. Which of the following statements is FALSE regarding the importance of blanching vs. non-blanching when it comes to conjunctival saccular aneurysm-like changes?**

- A. Conjunctival vasculature changes occur in the blood vessels; subsequently, when pressed, they should blanch.
- B. A keratinized lesion that includes some red blood cells will not blanch.
- C. Angiokeratomatous changes look like small microaneurysms or aneurysms and will blanch when pressure is applied.
- D. When the vessel can be compressed or the blood flow stopped, this indicates if the vasculature will blanch.

**15. Which of the following statements is accurate?**

- A. Although the literature indicates a propeller cataract and a Fabry cataract would be two separate entities, a propeller cataract could very well be a continuation of a dendritic cataract.
- B. A propeller cataract and a Fabry cataract are two separate and distinct entities, with no possibility for correlation.
- C. Cataract extraction is simpler in patients with Fabry disease than in any other cataract patient.
- D. Typically whitish, granular, and dendritic or spoke-like, a posterior subcapsular cataract develops more often in women than in men, and often affects vision.

**16. Limited physical ability affects patients with Fabry disease socially in which of the following ways?**

- A. Patients with Fabry disease are completely unaffected socially by limited physical ability.
- B. Patients with Fabry disease experience a social decline because of their limited physical ability.
- C. Despite painful episodes, patients will not alter their lifestyle.
- D. Patients with Fabry disease do not have a limited physical ability.

**17. Which of the following is not a risk for which patients undergoing treatment for Fabry disease should be counseled?**

- A. Inconvenience of ongoing IV infusions lasting 2-4 hours and occurring every 2 weeks
- B. Allergic reactions which may occur after the first four to seven infusions
- C. Localized infections at the needle site
- D. All of the above are risks patients undergoing treatment for Fabry disease should be made aware of.

**18. Which of the following are basic facts regarding Fabry disease of which patients should be made aware?**

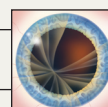
- A. Fabry disease is an abnormality of metabolism.
- B. Fabry disease results from an inability to break down normally occurring molecules that lead to their accumulation in tissues.
- C. Fabry disease is an X-linked inherited condition.
- D. All of the above are basic facts regarding Fabry disease of which patients should be made aware.

**19. Which of the following is a location of the body on which the angiokeratomatous changes associated with Fabry disease are characteristically found?**

- A. Bathing trunk area
- B. Umbilicus
- C. Penis/scrotum
- D. All of the above are locations of the body on which angiokeratomatous changes associated with Fabry disease are characteristically found.

**20. When do corneal findings typically occur in patients with Fabry disease?**

- A. Visible at birth
- B. After age 30 in females
- C. After age 30 in males
- D. Prior to age 10 in most males and females



## CE Instructions

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PRIMARY CARE OPTOMETRY NEWS  
NOVEMBER 2007  
**The Optometrist's Role in Diagnosing and Managing Fabry Disease**

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THE MONOGRAPH AND TEST EXPIRE NOVEMBER 2009.

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