

Test yourself in hereditary angioedema in pediatrics

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Choose only one answer:

- 1- Which of the following is the underlying etiopathogenesis of hereditary angioedema type I?**
 - A- Excessive mast cell degranulation
 - B- Immune complex vasculitis
 - C- C1-esterase inhibitor deficiency
 - D- Complement 4 deficiency

- 2- A 6-year old girl presented to the emergency room with facial swelling and mild stridor. This was the first attack. There was no urticaria. Her father had recurrent attacks of swelling of the extremities and genitalia especially with stress. What is the first line of treatment of this girl?**
 - A- Epinephrine.
 - B- Tracheostomy
 - C- Fresh frozen plasma
 - D- IM antihistamine

- 3- Which of the following is the recommended long-term prophylaxis in a 9-year old girl with frequent attacks of hereditary angioedema type I?**
 - A- Oral antihistamines
 - B- Attenuated androgens
 - C- Fresh frozen plasma
 - D- Tranexamic acid

- 4- A 10- year old boy presented with recurrent attacks of facial swelling but no rash or itching. No family history of similar condition. What is the recommended initial investigation to reach the suspected diagnosis?**
 - A- Erythrocyte sedimentation rate
 - B- Complement -4 level
 - C- Total immunoglobulin E
 - D- Genetic sequencing

- 5- In patients with hereditary angioedema type II, what are the expected C1-esterase inhibitor results?**
 - A- Normal level and function
 - B- Decreased level and function
 - C- Increased activity with normal level
 - D- Decreased function but normal level

- 6- Which of the following is the recommended long-term prophylaxis for hereditary angioedema type I and II?**
 - A- Injected C1 esterase inhibitor
 - B- Cetirizine
 - C- Corticosteroids
 - D- Fresh frozen plasma

- 7- Which of the following investigations are recommended to be done periodically for hereditary angioedema pediatric patients on danazol?**
 - A- Serum testosterone
 - B- Liver function tests
 - C- Complement -4 level
 - D- Erythrocyte sedimentation rate

- 8- What is the most important indicator of improvement and dose adjustment for a patient with hereditary angioedema type I/II on long term prophylaxis?**
 - A- Serum complement-4 level
 - B- Plasma C1-INH level
 - C- Frequency and severity of the attacks
 - D- Patient's compliance.

- 9- What is the mechanism of action of Icatibant?**
 - A- Kallikrein inhibitor
 - B- C1- esterase inhibitor
 - C- Bradykinin receptor antagonist
 - D- Increase degradation of bradykinin.

(Answers on page 100)