Treatment of Hereditary Angioedema

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CME Items

- 1. Which of the following is the final mediator in C1-INH-HAE and the cause of the angioedema attacks?
 - a. Kallikrein
 - b. C2 kinin
 - c. Bradykinin
 - d. C1-esterase inhibitor
- 2. Which of the following does the C1 esterase inhibitor inhibit to regulate the kallikrein-kinin system?
 - a. C1r, C1s, MASPs
 - b. Factor XIIa, kallikrein
 - c. Factor XIa
 - d. Plasmin, tissue plasminogen activator (tPA)
- 3. Which of the following molecules is released from high-molecular-weight kiningen as a result of the action of kallikrein?
 - a. C2-kinin
 - b. Plasmin
 - c. Bradykinin
 - d. Coagulation factor XII
- 4. Which of the following enzymes catabolize bradykinin and contribute to decreasing its levels?
 - a. Carboxypeptidase
 - b. Aminopeptidase P
 - c. Angiotensin-converting enzyme
 - d. All the above are true
- 5. Icatibant acetate is used for the treatment of C1-INH-HAE. Which of the following is its mechanism of action?
 - a. Kallikrein antagonism
 - b. C1-esterase inhibitor replacement
 - c. Blockage of the bradykinin type 2 receptor (B2R)
 - d. Stabilization of high-molecular-weight kininogen (HK)
- 6. Which of the following applies to lanadelumab?
 - a. It is a blocker of the type 2 bradykinin receptor (B2R).
 - b. It is a potent and selective inhibitor of human plasma kallikrein.
 - c. It is approved for long-term prophylaxis in patients of any age with C1-INH-HAE.
 - d. It is administered intravenously.

- 7. Which of the following applies to a patient with C1-INH-HAE under long-term prophylaxis with subcutaneous plasma-derived human C1 inhibitor (60 IU/kg twice a week) and good control of the disease (0 angioedema attacks in the last 3 months)?
 - a. He/she should have specific on-demand treatment for acute angioedema attacks available at home
 - b. He/she can treat any angioedema attack with subcutaneous plasma-derived C1 inhibitor (20 U/kg)
 - c. He/she can undergo surgery without prior shortterm prophylaxis
 - d. None of the above are true
- 8. Which of the following is true of intravenous pdC1INH?
 - a. It is approved for the treatment of acute attacks in children of any age by the EMA
 - b. It is approved for long-term prophylaxis by the FDA and the EMA
 - c. There are 2 marketed products (Berinert, CSL-Behring; Cinryze, Takeda Pharmaceutical Company Ltd), but their regulatory status is different
 - d. All the above are true
- 9. Which of the following is true of rhC1INH (Ruconest, Pharming Group NV)?
 - a. It is produced in transgenic cows
 - b. It is approved for the treatment of acute angioedema attacks
 - c. It is administered subcutaneously
 - d. Answers B and C are true
- 10. Which of the following applies to treatments under development for C1-INH-HAE?
 - a. Most drugs are aimed at blocking the bradykinin type 2 receptor (B2R)
 - b. Gene therapy is very advanced and expected to be on the market in 2021
 - c. Some drugs block activated FXII
 - d. Direct blockage of high-molecular-weight kiningen is one of the strategies