Treatment of Hereditary Angioedema

Accreditation requested at the "Consejo Catalán de Formación Continuada de las Profesiones Sanitarias – Comisión de Formación Continuada del Sistema Nacional de Salud"

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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 Xla
   d. Plasmin, tissue plasminogen activator (tPA)

3. Which of the following molecules is released from high-molecular-weight kininogen 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 short-term 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 rhC1NH (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 kininogen is one of the strategies