Specialised Services Circular

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NHS England Clinical Commissioning Policy 16045/P: Plasma-derived C1-esterase inhibitor for prophylactic treatment of hereditary angioedema (HAE) types I and II

Circulation

For action	**For information**
Local Team Assistant Directors of Specialised Commissioning Local Team Pharmacists	Regional Team IFR Leads Regional Directors of Specialised Commissioning Regional Clinical Directors of Specialised Commissioning Regional Directors of Nursing (Specialised Commissioning)

Background

1. Hereditary angioedema (HAE) is a rare condition arising from a genetic deficiency of C1-esterase inhibitor, also called C1-inhibitor, a regulator of inflammatory pathways. Intravenous administration of reconstituted plasma-derived C1-inhibitor (human) replaces the C1-inhibitor regulatory protein.
2. In individuals without HAE, this protein controls enzyme cascade reactions so that uncontrolled swelling of the subcutaneous and submucosal tissues does not normally occur. In HAE, the absence of a functional control protein leads to episodes of uncontrolled swelling. Swellings can be disabling, cause severe pain and can be fatal if occurring in the airways.
3. Most people with HAE have low concentrations of C1-inhibitor (HAE Type I); around 15% have normal or high concentrations of non-functional C1-inhibitor protein (HAE Type II).
4. Most patients require C1-inhibitor, or icatibant, as emergency treatment for acute clinically significant attacks and C1-inhibitor for short term (generally single dose) prophylaxis prior to known triggers which include, for example, dental work or surgery. For the majority of people with HAE, attacks are either infrequent or can be controlled adequately using oral prophylactic medications together with a plan to treat acute attacks as above.
5. A minority of people who experience two or more clinically significant attacks of swelling per week, for whom oral prophylaxis is not tolerated or is ineffective, may benefit from prophylactic C1-inhibitor injections on a regular basis to reduce the frequency of attacks and the need for emergency treatment.
6. A clinically significant attack is one which is i) potentially life threatening because it affects the head or neck or ii) causes pain or disability such that the patient cannot continue their normal activities. This may be due to disabling cutaneous swelling, sufficient to prevent the patient from undertaking normal activities or severe abdominal pain which will not respond to oral analgesia. Varying treatment pathways do not imply that an attack requiring hospital treatment is necessarily more significant than one which can be treated with self-administered therapies.
7. C1-inhibitor is a blood product, extracted from pooled donated plasma, which is then purified to eliminate the risk of contamination with pathogens, especially blood-borne viruses.

Summary

8. NHS England has approved the use of C1 esterase inhibitor for the prophylactic treatment of HAE types I and II for:
   a) Individuals who fail, or are intolerant of oral prophylaxis and who experience two or more clinically significant attacks per week, despite oral prophylaxis (see Definitions Policy 16045 here), over a period of at least 56 days requiring

treatment with C1 esterase inhibitor or icatibant.

b) Individuals in whom oral prophylaxis is contraindicated for example pregnant women, recognising that there are currently no other prophylactic treatment options during pregnancy and that there is increased risk of rapid deterioration in condition and additional risks to women during pregnancy.

9. Patients should already be receiving on demand treatment with either a C1 esterase inhibitor or icatibant.

10. Patients who require prophylactic treatment with a C1 esterase inhibitor will require registration via a Blueteq form. A form has been produced and enabled on the system for providers currently commissioned for this service.

11. It is estimated that circa 55-110 patients may access prophylactic treatment in the next 5 years. It is considered that this is cost neutral as patients requiring prophylaxis would have been accessing on demand treatment to the same level.

Action

12. Sub Regional Teams to note the policy recommendations and complete and circulate the draft provider letter provided as relevant.

13. Regional teams to produce and provide Contract Variation for providers

14. Sub Regional Teams to ensure that Trusts are registering prophylactic C1 esterase inhibitor use on Blueteq.

15. Ensure reimbursement for prophylactic C1 esterase inhibitor is on the basis of completed Blueteq forms only.

Further Information

NHS England Clinical Commissioning Policy 16045/P: Plasma-derived C1-esterase inhibitor for prophylactic treatment of hereditary angioedema (HAE) types I and II can be found at:

https://www.england.nhs.uk/commissioning/spec-services/npc-crg/blood-and-infection-group-f/f06/

	
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