Commissioning Policy Summary for the Treatment of acute attacks of hereditary angioedema

What is hereditary angioedema (HAE)?

HAE is usually an inherited condition, meaning it runs in families. Sometimes it can also occur when there is no family history and this is due to a spontaneous gene mutation.

Acquired angioedema (AAE) has the same symptoms as HAE and it occurs as a result of an underlying condition such as lymphoma.

HAE is recognised by huge swellings called angioedema. These are caused by the lack of a chemical, known as C1 inhibitor, in the blood. The swellings last from three to five days. They can happen on any part of the body, hands, feet, arms and legs, trunk, intestines, genital organs, face, tongue and airway. Swelling of the abdomen causes severe pain, vomiting and diarrhoea, and swelling of the airway can be fatal.

How rare is HAE & AAE?

These are very rare conditions. HAE occurs in 1 in 50,000 of the population and AAE occurs in 1 in 500,000 of the population.

What this policy covers

The NHS England policy defines services that will be provided for the treatment of acute attacks of angioedema caused by C1 Inhibitor deficiency.

This policy covers treatment available for both adults and children with HAE.

The aim of the policy is to help patients move towards managing their symptoms so that their personal safety is maintained with minimal disruption to living healthy and productive lives.

HAE patients will be treated in accredited centres by specialists who are able to provide treatment in line with the new policy.

What this policy means to you as a patient

The frequency and severity of HAE attacks varies considerably from patient to patient and the pattern of attacks can change at different stages of life.

The recommendations are that:

- All severe attacks of HAE causing pain due to internal abdominal swelling or attacks threatening the airway should be treated at the earliest possible stage with C1 inhibitor or Icatibant.

  C1 inhibitor is given by an injection into a vein.
  Icatibant is given by an injection just under the skin.

- Preventative treatment with C1 inhibitor is given before surgery, for obstetrics including pregnancy, and dental work.

- Patients who experience very few attacks of HAE may receive treatment for their acute attacks in A&E.

- Patients who meet certain criteria, such as frequent severe attacks should be offered training to give their own medication in a home setting. This allows them to treat attacks at the earliest stage in order to minimize the impact on their lives.

- Icatibant may be given as an alternative to C1 inhibitor where a patient meets the criteria for home therapy but would have difficulties in managing injections into a vein.

- Recombinant C1 inhibitor (Ruconest) is a type of C1 Inhibitor that is not derived from human blood. This will be available for patients who for religious or medical reasons cannot use other C1 inhibitor products.

What this policy does not cover

Long-term preventative treatment with C1 Inhibitor is not covered in this policy. In the very severe cases of HAE / AAE where the physician considers this necessary a special individual funding request must be made.

How this policy fits in with the wider services for HAE

The services for acute attacks of Hereditary Angioedema outlined in this policy are provided within a wider service pathway for the management of HAE. This would include:

- Referral from a GP to a Specialist HAE Centre
- Diagnosis of HAE / AAE
- Patient information provided explaining the condition
- An individual HAE management plan set up including:

    - Emergency treatment plan;
    - Preventative treatment plan;
    - Management plan for special circumstances including dental treatment and operations; and
    - Regular checks on liver function and scans.

All patients with HAE types 1 and 2 and AEE will be able to access these service provisions.

If patients experience difficulties accessing these services they will need to discuss their treatment with their HAE specialist.

If they still have issues regarding treatment they should contact PALs and their patient support organization HAE UK (www.haeuk.org) or PID UK (www.piduk.org) for advice.

Glossary

Accredited centres	Treatment centres that meet all the requirements for treating HAE patients according to the NHS England policy for managing acute attacks of HAE.
Acute attacks	Sudden onset attacks.
Lymphoma	A type of blood cancer.
C1 Inhibitor	A medication containing the blood protein that people with HAE lack.
Icatibant	A medication that blocks the action of a substance called bradykinin, which causes swelling of the tissues.
Ruconest	A type of C1 Inhibitor that does not come from human blood. This product is suitable for patients who for religious or medical reasons cannot use other C1 inhibitor products.
Obstetrics	The branch of healthcare that cares for women during pregnancy and childbirth.
PALs	Patient Advice and Liaison Service, which will give help and advice to patients.
Spontaneous gene mutation	A change in the genetic make-up in an individual, occurring for the first time in a family.

Related document

NHS England Clinical Commissioning Policy: Treatment of acute attacks of hereditary angioedema April 2013 Reference: NHSCB/B09/P/b