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Amendments procedure:

The current document will be held in the protocols section as a 'pdf'.

Proposed amendments can be made to a working 'word' copy.

When amendments have been accepted and signed off a new version will be saved as a pdf, with new amendments listed here. Previous versions will be saved in archive folder.

This document is to support effective management of the allergy and immunology clinical service and is to be used in conjunction with Trust policies, which are accessible on the Trust home page.

## Hereditary Angioedema Management Plan –Home Therapy

Immunology Nurse Specialist: 01782 672504  
Immunology Department Secretary: 01782 674284

### Management of Attacks

#### MILD

Peripheral swellings or other swellings that do not prevent attending work or usual activities.

#### Treatment plan:

- Painkillers if required

#### MODERATE: Abdominal attacks and peripheral attacks that require treatment

Peripheral attack that prevents attending work or usual activities, e.g. swollen feet or hands

#### Treatment plan:

- Give iv C1 inhibitor 1000units of Cinryze/Berinerst or 2,100-4,200U of Rocunest
- If C1 inhibitor is not available or cannot be given then icatibant 30mg subcutaneously may be used as an alternative
- If no improvement then seek medical help.

#### SEVERE: Head or neck attacks

**YOU MUST ATTEND THE EMERGENCY DEPARTMENT IF YOU HAVE A HEAD OR NECK ATTACK EVEN IF YOU HAVE HAD TREATMENT AT HOME.**

#### Treatment plan:

- If you are having any difficulty breathing or swallowing dial 999.
- Attend the Emergency Department as soon as possible.
- You can give the following or take with you to the emergency department:
  - iv C1 inhibitor (Cinryze/Berinerst) 20units per kilogram body weight. Usually 1000-1500u or 4,200U of Rocunest.
  - If C1 inhibitor is not available or cannot be given then icatibant 30mg subcutaneously may be used as an alternative

## General Reminders

- Make sure that your C1 inhibitor or icatibant is in date and you always have enough to treat an attack
- Keep your Emergency Department letter somewhere safe
- Before travelling abroad you may need to:
  - order extra medication.
  - get permission to bring attenuated androgens such as danazol into a country
  - get permission to carry medication and equipment on airlines
  - use a cool bag to maintain C1 inhibitor and icatibant between 0-25<sup>0</sup>c
- Discuss your treatment if you are planning a pregnancy. Androgens must not be taken during pregnancy.
- Avoid the following medication:
  - Blood pressure medication called ACE inhibitors
  - Oestrogen containing medication e.g. oral contraceptive

## What to do to prevent attacks during dental and other procedures

### Low risk procedures

No extra precautions are necessary for these procedures:

- minor dental procedures eg, routine cleaning, filling of cavities, crown placement, restorative work, etc
- local anaesthetic procedures (non-dental)
- normal delivery of baby (see below)

### Moderate/High risk procedures

If you are expecting to have any of these procedures then talk to the dentist or surgeon in plenty of time so that plans can be put in place with your immunology team.

- dental procedures that involve cutting of tissue or injection of anaesthetic and any procedure that has caused angioedema previously
- operative procedures requiring intubation (most general anaesthetics)
- normal deliveries are NOT high risk, but usually precautions are put in place as deliveries can be unpredictable