**Department of Immunology**

Manchester Royal Infirmary

Oxford Road

Manchester

M13 9WL

Direct Line 0161 276 6686 Office Hours 09:00 to 17:00

To Whom It May Concern:

* This patient has hereditary angioedema/acquired angioedema (HAE/AAE). This can cause recurrent attacks of abdominal pain, limb swelling or laryngeal obstruction.
* Please read this in conjunction with the patient’s recent clinic letter (carried by the patient) that confirms the patient details, diagnosis, and recommended dose and product of C1 esterase inhibitor (C1INH) and / or icatibant. Patients will usually carry their own treatment and will often be trained to self-administer.
* Treatment of laryngeal HAE attacks is a **medical emergency**. The symptoms of HAE are unlikely to respond to adrenaline, antihistamines or steroids.
* Treatment of choice is C1INH given intravenously over five to ten minutes. If the symptoms do not improve a further dose can be given or contact an immunology consultant on the telephone number below (no guaranteed out of hours provision).
* Patients will be prescribed one of 3 C1INH products, Berinert, Cinryze or Ruconest. All products can be stored at room temperature. If you have to give a different C1INH product to the patient’s usual treatment we ask you to take a serum sample to be saved (send to Immunology Laboratory at the Manchester Royal Infirmary – not required if you use Ruconest).
* Berinert is supplied as either 500 unit or 1500 unit vials with its own diluent. Use the diluent provided to reconstitute, 10mls of water for injection for each 500 unit vial, 3 mls for a 1500 unit vial. It may take longer to dissolve the 1500 unit vial. Cinryze is supplied as 500 unit vials with 5ml of water for injection. Ruconest is supplied as 2100 unit vials with 14ml solvent (water for injections to be used if no solvent provided). All products should be administered as a slow intravenous bolus injection over five to ten minutes. We would recommend using a butterfly needle rather than a Venflon. C1INH can be pushed by hand and does not need to be given by a pump or syringe driver.
* C1INH will improve most symptoms over thirty to one hundred and twenty minutes.
* C1INH should also be given preventatively before any invasive (endoscopy / bronchoscopy) or surgical procedures including dental extractions.
* C1INH is licensed for the treatment of HAE.
* Icatibant is an alternative treatment that is licenced for use in acute attacks of HAE and 30 mg is administered subcutaneously into the abdomen, slowly over 30 seconds or longer.
* If the patient does not have their own supply of C1 esterase inhibitor or icatibant please use the most accessible product locally held in your hospital. You may need to contact the Pharmacy Department.

Contact numbers:

Office hours – 0161 276 6486

Out of hours via MFT switchboard – 0161 276 1234

**Immunology Department**