18th February 2015

Deputy Eoghan Murphy TD
Dáil Éireann
Leinster House
Dublin 2

PQ No. 33815/14

* To ask the Minister for Health his views on implementing best practices regarding hereditary angioedema (HAE) which have been outlined in a report (details supplied), and adopted in most European Union countries, specifically to make home infusions of C1-INH protein standard practice; his views on the report's positive cost-benefit analysis of said practice and to educate medical professionals of the symptoms of HAE.

Dear Deputy Murphy,

The Health Service Executive has been requested to reply directly to you in the context of the above Parliamentary Question, which you submitted to the Minister for Health for response. I have examined the matter and the following outlines the position.

Response:

Background

Hereditary angioedema is a rare inherited disorder characterised by dysregulation of the complement and kinin pathways leading to unpredictable episodes of swelling of the skin and mucous membranes. Swellings can be temporarily disfiguring – in the case of skin and facial swellings, painful – in the case of intestinal angioedema, and potentially life-threatening – in the case of laryngeal oedema. Diagnostic delay is common. Even when the diagnosis has been made the delay in administration of effective treatment when a patient presents to an emergency department can be considerable. It is well recognised that delay in treatment is associated with adverse outcomes. Most patients are under the care of one of 4 full-time adult clinical immunologists or the single consultant paediatric immunologist.

Management options in Ireland

- Patients and their families are educated about the condition
- Patients are offered prophylaxis to prevent swelling – options include tranexamic acid and danazol. Danazol is an androgen and is relatively contraindicated in young females due to its virilising effects. Prophylaxis is often ineffective.
- Most commonly, patients that experience orofacial swelling or abdominal pain will attend an emergency department (or if possible their clinical immunology department) for administration of C1 esterase inhibitor concentrate (IV) or icatibant (SC). Patients carry their documentation to inform ED staff of the correct management protocol and local protocols are available in many major emergency departments. Despite this, anecdotal reports tell of significant treatment delays.
• Some patients have access to icatibant as an on-demand medication at home. These patients must have received a dose of icatibant in hospital. This is not yet available to all patients but is being rolled out to those with a history of unstable swellings.
• Some patients are particularly difficult to control with repeat emergency admissions require long term prophylaxis with regular C1 inhibitor concentrate (2-3 times/week). This is best administered in a home setting.

International practice
• Recent World Allergy Organisation guidelines (among other position papers and publications) state that all patients with hereditary angioedema should be considered for home treatment. Home treatment prevents undertreatment of attacks, delayed treatment and inappropriate treatment.
• In the UK all HAE patients are offered C1 inhibitor concentrate to keep at home to be used in the event of a significant swelling episode. They are either trained in self (or partner assisted) administration or can bring the treatment to a local emergency department or primary care facility.
• In the UK all patients with unstable swellings are offered C1 inhibitor concentrate prophylaxis home therapy.
• Some centres are evaluating icatibant and other treatment alternatives such as recombinant C1-inhibitor
• A variety of sources confirm that on-demand home-therapy is cost effective but controversy exists over which product is most cost-effective

Gap analysis
• Access to on-demand medication for the emergency treatment of angioedema swellings at home is not standard in the Irish context.
• Home C1 inhibitor prophylaxis has been arranged for individual patients in Ireland, however, it is difficult to arrange and is not standard in the Irish context.
• Some patients do have home possession of icatibant for emergency use, this requires further work.
• Attempts (through the Irish Association of Allergy and Clinical Immunology; formerly IAI) to offer ensure these international standards of care are made available to all hereditary angioedema patients have not been successful so far.

Educational issues
• An understanding management of hereditary angioedema and the other much more common forms of angioedema (allergic, ACE inhibitor induced and chronic spontaneous) is essential for primary care and emergency department physicians
• Clinical Immunology services in St. James’s Hospital, Beaumont and Galway provide information to patients, physicians, health-care professionals and students on a regular basis and are committed to the dissemination of knowledge on this and other immunological and allergic conditions.

I trust this answers your question to your satisfaction.

Yours sincerely,

Helen Byrne
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Acute Hospitals Division