ARQUIVOS DE ASMA, ALERGIA E IMUNOLOGIA

The new hereditary angioedema guidelines: what is its role?

As novas diretrizes de angioedema hereditário: qual é o seu papel?

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Progress in the knowledge and diagnosis of rare diseases has been extraordinary in recent years. In parallel, the interest in establishing treatments for these situations also showed a noticeable improvement. Guidelines on hereditary angioedema (HAE) have been published for over 20 years. Initially, these documents were developed from the experience of specialists and without a methodological system.[1,2] However, there was a need to establish specific guidelines for patients with hereditary angioedema, whose risk of death from asphyxia was from 25 to 40%. There was no way to ignore a clinical entity that was increasingly diagnosed and which had no appropriate therapeutic resources. It is important to highlight that plasma-derived C1 inhibitors have been available in European countries for decades, despite limited supply in most countries, including some developed countries.[3]

The recognition of the kinin-bradykinin system as the main mechanism involved in edema represented a significant change in the treatment of HAE. The need to treat attacks and reduce or even eliminate HAE mortality has boosted the development of drugs to treat the disease. The guidelines are beginning to take shape, with comparative studies demonstrating the effectiveness of newer treatments over conventional ones, such as plasma infusion or the use of plasmin inhibitors. Protocols with more adequate methodology are also included, although the recommendations are not yet necessarily supported by an adequate level of certainty in the evidence.[4] The main objective of expert panel recommendations is to advise clinicians on the best possible and acceptable way to approach a given decision making in the area of diagnosis, management or treatment.[5]

The treatment of angioedema attacks has been expanded with access to new therapeutic resources. Self-administration and early application of medications reduced emergency room visits, or even hospitalization, significantly reducing the duration of attacks. Considering that there would be adequate drugs for attacks, crisis prophylaxis became the new goal to be achieved. For this reason, several studies have demonstrated the effectiveness of new drugs and the impact on quality of life. It is important to emphasize that patients with properly treated hereditary angioedema have the same survival rate as the general population, and the disease does not cause relevant adverse effects, allowing a productive life. According to a recent publication that evaluated the situation of HAE management in 28 countries, there are inequalities in the services and treatments around the world, and access to appropriate treatments is still restricted to developed countries.[3] Mortality from hereditary angioedema in our country, recorded by the Association of patients with HAE (ABRANGHE), still impacts the profile of our patients (ABRANGHE personal communication).[6,7]

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There are numerous ways to synthesize available biomedical information[8] so that health care professionals can make decisions from heterogeneous sources. Clinical practice guidelines are documents that generally cover the context-specific information needed to make explicit and ideally transparent recommendations.[4,8] Low- and middle-income countries have been slowly and progressively adjusting to the standards of developed countries, but always striving for a global approach to the patient. It is difficult to ignore advances in patient assessment with the use of quality of life questionnaires, action plans and diagnostic tests. Clinical practice guidelines are the most important documents for incorporating scientific evidence into health decision-making, however, it is necessary to recognize some limitations of this process, mainly in developing countries. However, not knowing the evolution in the treatment of hereditary angioedema would be to deny the relevant role of new therapeutic resources.

With the advances described, consensus was largely replaced by guidelines, incorporating scientific evidence.[9,10] Clinical practice guidelines are not cookbooks as they may have limitations in their availability and applicability in the local context. However, they serve as an update so that the clinical diagnostic and therapeutic protocols (PCDT), essential instruments for the implementation of new resources, are reviewed. The Ministry of Health has used the instrument called AGREE II (Appraisal of Guidelines for Research & Evaluation II) that evaluates six domains: the scope and purpose, participation of those involved, rigor in its elaboration, clarity and specificity of the recommendations and applicability of the proposal.[11-13] Thus, the expectation is that the guidelines published here in the "Archives of Asthma, Allergy and Immunology" will contribute to a better diagnosis and treatment of the patient with hereditary angioedema, reaching the main recommendation of the latest guidelines of the World Allergy Organization, which is the normalization of the patient's life.

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