IMMUNOLOGY

HEREDITARY ANGIOEDEMA

***Archan Sil**
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Abstract: *Hereditary angioedema is an uncommon disorder with autosomal dominant mode of inheritance and is clinically characterized by recurrent episodic swelling of face, limbs, genitals, airway and gastrointestinal tract. Because of lack of awareness, most patients with hereditary angioedema remain undiagnosed and untreated. Swelling episodes in patients with hereditary angioedema are mediated by bradykinin. Excess bradykinin due to defective C1 inhibitor protein is the basic fault. While in type 1 HAE, C1 inhibitor protein levels are low, HAE type 2 is characterized by normal levels of C1 inhibitor protein that is functionally defective. C1 inhibitor protein levels. and function are normal in type 3 hereditary angioedema Treatment of acute attacks, short term prophylaxis and long-term prophylaxis are the mainstay in management. C1 inhibitor protein concentrate is the preferred treatment for patients with hereditary angioedema in the developed countries. However, because of non-availability of this drug in India and many other developing countries, most patients are treated with fresh frozen plasma, attenuated androgens and tranexamic acid. In this review, we update on the pathogenesis, clinical features, diagnosis and management of hereditary angioedema.*

Keywords: *Hereditary angioedema, Bradykinin, C1 inhibitor, Acute attacks, prophylaxis, Attenuated androgens, Tranexamic acid.*

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Points to Remember

- ***Hereditary angioedema (HAE) is an uncommon disorder characterized by episodic edema.***
- ***Because of lack of awareness, the disease remains undiagnosed for several years.***
- ***HAE should be suspected in all patients who present with episodic edema without urticaria.***
- ***In patients with suspected HAE, C4, C1-INH levels and C1-INH function should be assessed.***
- ***Most patients have diseases onset in childhood. Hence, pediatricians have an important role to play in the early diagnosis of HAE.***
- ***Patients with HAE in most of the developing countries including India are managed using fresh frozen plasma, attenuated androgens and tranexamic acid because all 1st line treatments are not available.***

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