Differentiating HAE from other types of angioedema

	HAE-C1-INH[1–5]	HAE-nC1-INH[1,2,6,7]	Mast cell-mediated angioedema*[1,2]	Acquired angioedema with low C1-INH[1,2,8]	ACEi-induced angioedema[9]
Onset of attacks	**Begins during childhood[1,2]**	Begins during or after the second decade of life[7]	All ages[2]	Usually begins after 30 years of age[1,2]	Variable (50% within first week of treatment)[9]
Response to antihistamines and corticosteroids	**No response[1,2]**	No response[2]	Response[1]	No response[2]	No response[9]
Family history	**Yes, ~75% of cases[1]**	Yes[1]	No[1,2]	No[1]	No[1]
Gender-based differences	**Male to female ratio 1:1[3]**	More prevalent in females[6]	Not reported	Not reported	More prevalent in females[9]
Duration of attacks	**2–5 days[5]**	2–5 days[7]	1 day[2]	2–5 days[8]	Resolves within 2 days upon ACEi discontinuation in the majority of cases[9]
Abdominal attacks	**Yes[1,2]**	Yes[7]	Not reported	Yes[8]	Yes[9]
Urticaria and pruritus	**No[1,2]**	No[7]	Often[2]	No[8]	No[9]
Prodromal symptoms	**84% of patients[4]**	Not reported	Not reported	Not reported	Not reported

*Includes histamine. ACEi, angiotensin-converting enzyme inhibitor; HAE, hereditary angioedema; HAE-C1-INH, HAE with C1-esterase inhibitor deficiency;

HAE-nC1-INH, HAE with normal C1-esterase inhibitor.

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