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Understanding **Hereditary Angioedema (HAE)**

What is HAE?

Hereditary angioedema (HAE) is a **rare genetic disease.**[1]

People living with HAE experience **unpredictable, painful, and debilitating attacks** of tissue swelling in various locations of the body that can be life-threatening depending on the location(s) affected.[1-2]

The Most Common Symptoms of HAE are Attacks of:

96%	**93%**	**52%**	**21%**
Diffuse Skin Swelling[3]	Submucosal Edema[3]	Larynx Swelling[3]	Oropharynx Swelling[3]

Multiple locations may be affected within the same attack and may occur simultaneously or consecutively.[3-4]

If left untreated, attack symptoms will typically intensify over the first **12-36 hours,** then gradually subside **over 2-5 days**.[5]

Severe **upper airway attacks may be fatal** without prompt administration of an appropriate on-demand treatment.[5-7]

Who Does it Affect?

HAE occurs in about **1 in 10,000** to **1 in 50,000** people.[8]

50% of people with HAE first experience an attack **before age 10.**[9]

75% of people living with HAE have a known family history with an autosomal-dominant inheritance pattern; **in up to 25% of people**, it may result from **spontaneous mutations.**[5,10-12]

What Causes HAE?

Mutations in the serpin family G member 1 (SERPING1) gene resulting in deficiency (type 1) or dysfunction (type 2) in the C1 inhibitor (C1INH) protein[1,5,6,10,11,13,14] and subsequent uncontrolled activation of the kallikrein-kinin system (KKS).[15-19]

Identifiable triggers for HAE attacks include trauma, infection, or exposure to estrogen-containing medications.[17]

How is it Diagnosed?

Laboratory analysis of blood samples, or genetic samples, are required to establish an HAE diagnosis.[8]

Frequent and severe abdominal pain associated with HAE can be misdiagnosed; these inaccurate diagnoses can result in ineffective treatment or unnecessary surgery.[20]

How Does HAE Impact People's Lives?

HAE has a significant impact on quality of life, work/school productivity, social activities, and career/educational achievements.[21-24]

People living with HAE miss **approximately 20 days of work / school per year.**[25]

People living with HAE may be **unable to use their hands or walk** during attacks.[26-27]

≥50% indicated that HAE had **hindered their career/educational advancement.**[25]

Children and adolescents who experience HAE attacks report **lower health-related quality of life** (HRQoL).[28]

What Do Guidelines Recommend?

International guidelines reinforce four key recommendations for the treatment of HAE attacks.[1,10,29]

- Treat attacks as **early** as possible after recognition of onset.
- Treatment should be **considered for all attacks,** regardless of anatomic location or severity.
- Train all patients in the **self-administration** of on-demand treatment.*
- Ensure all patients have ready access to, and carry, sufficient on-demand medication to treat at least two attacks.

Early treatment prevents **progression of swelling, shortens the time to attack resolution, and minimizes daily disruptions.**[1,10,29]

Self-administration of treatment reduces:[1,10,29]

- **Severity and duration of HAE attacks**
- **Morbidity**
- **Cost of care**

Access to on-demand medication for **at least two attacks** ensures readiness for successive or potentially **life-threatening laryngeal attacks.**[1,10,29]

*If it is licensed in their country for self-administration.[1,10,29]

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