Patients with HAE and comorbidities

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Disclosures

- Speaker/advisor fees received from AbbVie, Allergika, Almirall-Hermal, Amgen, Beiersdorf, Biocryst, BMS, Boehringer-Ingelheim, Celgene, CSL-Behring, Eli-Lilly, Galderma, Hexal, Janssen, Klinge, Klosterfrau, LEO-Pharma, LETI-Pharma, L´Oreal, Novartis, Octapharma, Pfizer, Pflüger, Pharming, Regeneron, Shire, Takeda, Regeneron, Sanofi-Genzyme und UCB Pharma

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- The views, information or opinions expressed herein are those of the speaker; they do not necessarily reflect those of CSL Behring

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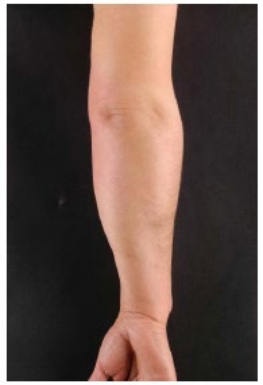
Patient description

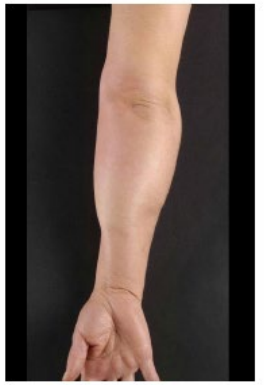
- Elderly, female patient aged 62
- Diagnosed with HAE type I
- Experiences severe HAE, with a high frequency of attacks in the last 40+ years
- Suffers from comorbidities such as **depression** and **hypertension**
- Profession: Cleaner (6 hours per day)

Case history

1978 (first symptoms aged 19)

- 3 months after pregnancy
- Swelling of the skin and abdomen biweekly





Treatment before diagnosis:

- Celestamine/antihistamines on-demand therapy (approx. 30 doses) – unsuccessful
- Celestamine oral tablets (1 per day for over 1 year)
- Celestan depot IV administrations (approx. 10 injections during treatment)

HAE, hereditary angioedema; IV, intravenous

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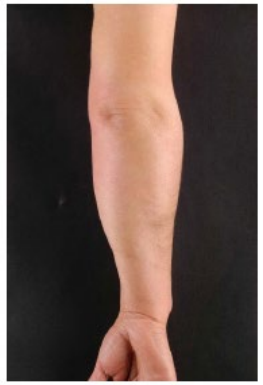

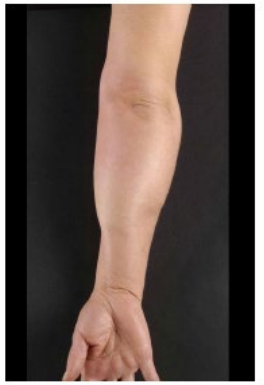

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1989 (diagnosis aged 30)

- HAE type I diagnosis
- Family history
  - Lives with husband, and 2 adult children live nearby
  - Has 5 siblings
  - No other family members with HAE or any who have displayed similar symptoms

HAE, hereditary angioedema; IV, intravenous

Treatment history

1989–1994

Danazol

(100 mg daily, 60 kg)

- Weight gain (>10 kg)
- Headaches
- Weakness
- Adynamia
- Increasing depressive episodes
- >30 attacks per year (medium–severe)

**The indications for use and dosing of intravenous C1 inhibitor are dependent on manufacturer's Summary of Product Characteristics. Berinert® 500/1500 is licensed for the treatment and pre-procedure prevention of acute episodes of HAE. The approved dose of Berinert® 500/1500 for the treatment of acute attacks is 20 IU/kg. CSL Behring does not suggest or recommend the use of C1-INH (IV) in any way other than as described in the Summary of Product Characteristics*

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1998–1999

C1-INH IV

(500 IU/kg, twice weekly in hospital*)

- Controlled disease and side-effects disappeared (except depression)

↓

- Treatment switched due to other issues

Tranexamic acid

(3 g per day – ineffective at controlling attacks)

Additional treatment used for breakthrough attacks:

- Danazol (100–200 mg per day)
- C1-INH IV on-demand
- Icatibant

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Persistent side-effects leading to reduced working:

- Increasing depressive episodes
- Weakness
- Nausea

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2010–2021

C1-INH IV

(500–1000 IU/kg, administered at home by husband as soon as attack appears*)

- Husband completed training programme for home administration
- HAE symptoms controlled after the second week

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2020

C1-INH SC long-term prophylaxis

(2000/3000 administered for two weeks)

- Patient unable to adhere to LTP regimen

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Current treatment plan

On-demand therapy

- Patient has only been interested in on-demand treatment for acute attacks
- She has a long history (approx. 23 years) of treatment with IV C1-INH (500–1000 IU/kg)
- Patient still declines long-term prophylaxis therapy – options discussed at every visit

- HAE attacks are managed with IV C1-INH treatment
  - Experiences moderate attacks approx. twice per month (abdomen and/or skin)
  - Laryngeal attacks occur 3/4 times per year (managed in hospital)
  - >1000 IV C1-INH injections – treatment is still effective at managing acute attacks at the same dose

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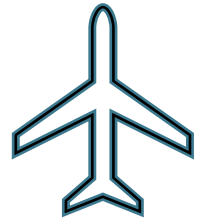
Issues encountered during treatment

- Language barrier
- Travel to homeland, where C1-INH was not available at that time
- Not able to drive to the hospital
- GP will not administer C1-INH IV on demand or prophylaxis
- Husband works and cannot help administer
- Injection site reactions with SC injection



C1-INH, C1-esterase inhibitor; GP, general practitioner; IV, intravenous; SC, subcutaneous

Treatment and patient reported outcomes

Treatment outcome

- **No loss of efficacy** present after 20+ years of receiving treatment with C1-INH
- No interaction with additional medication

Patient reported outcomes

- Angioedema control test (AECT) was used to assess disease control (score: 14)
- Angioedema quality of life questionnaire (AEQoL) was not feasible due to language barriers

C1-INH, C1-esterase inhibitor

Treating patients with comorbidities

- The presence of comorbidities may affect the clinical course of HAE in patients ≥65 years old[1]
  - Annual mean attack frequency has shown to be greater in patients with concomitant diseases, particularly in women
- Anxiety and depression are common comorbidities that affect 35% and 21% of patients with HAE[2]

HAE, hereditary angioedema

1. Bernstein JA. *et al., Ann Allergy Asthma Immunol.* 2020; 125(3):334–340.e1. 2. Banerji A. *et al., Ann Allergy Asthma Immunol.* 2020; 124(6):600–607.

Poll: Which quality of life tools are more appropriate for elderly patients?

A. Angioedema control test (AECT)

B. Angioedema quality of life questionnaire (AEQoL)

C. Angioedema activity score (AAS)

D. HAE activity score (HAE-AS)

Personalised medicine

Burden of disease = disease activity + quality of life

Quality of life may differ in presentation for patients across all ages

Elderly patients may be unable to complete long questionnaires to assess quality of life and disease control

According to a recent Delphi consensus, achieving total control of HAE and normalisation of patients' lives should be the goal of treatment in HAE[1]

HAE, hereditary angioedema

1. Maurer M. *et al., J Allergy Clin Immunol.* 2021; S0091–6749(21)00821–6

Take-home messages

- Treatment should be individualised to suit a patient's needs and preference
  - Despite having the option of long-term prophylaxis, on-demand therapy may be preferred, as seen in this case

- Comorbidities can affect the course of the disease and may influence the patient's treatment preference
  - These should be taken into account when developing a treatment plan

- Discussions around other treatment options at least once per year
  - Important to make patient aware of other/newer treatment options that are available