

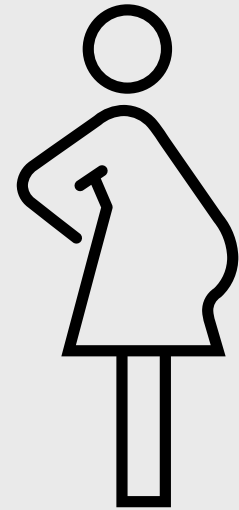
Treating HAE during pregnancy

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Disclosures

- Speaking and/or consultancy fees from Biocryst, CSL Behring, Ionis Pharmaceuticals, KalVistaTakeda, Pharming, Pharvaris and Sanofi/Genzyme

Disclaimer

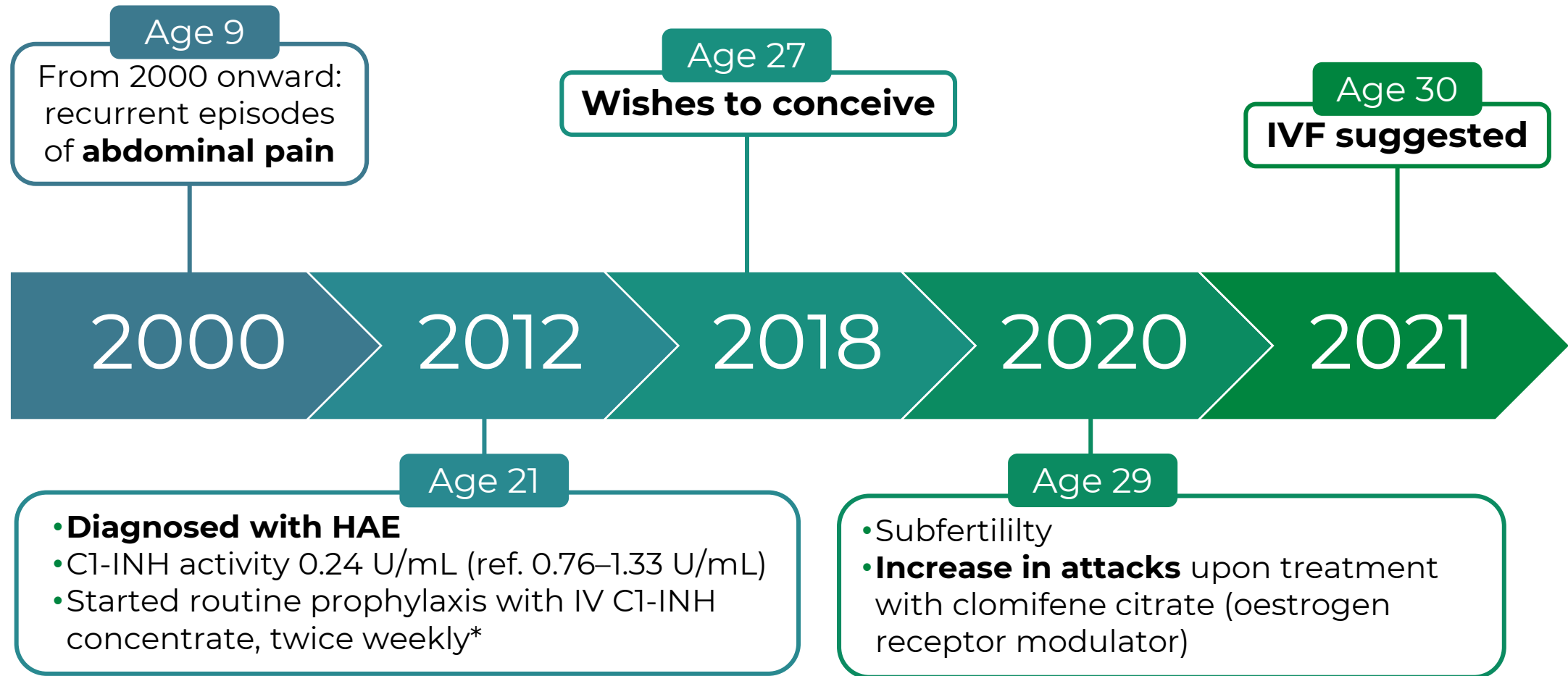
- The views, information or opinions expressed herein are those of the speaker; they do not necessarily reflect those of CSL Behring
- Slides may contain off-label content

Patient case

- 30-year-old female with HAE
- Frequent **abdominal attacks** despite routine prophylaxis with IV C1-INH concentrate*
- Wish to conceive
- **Emotional distress**

**The use of intravenous C1 inhibitor for long-term prophylaxis is dependent on manufacturer's Summary of Product Characteristics*

Case history



**The use of intravenous C1 inhibitor for long-term prophylaxis is dependent on manufacturer's Summary of Product Characteristics*

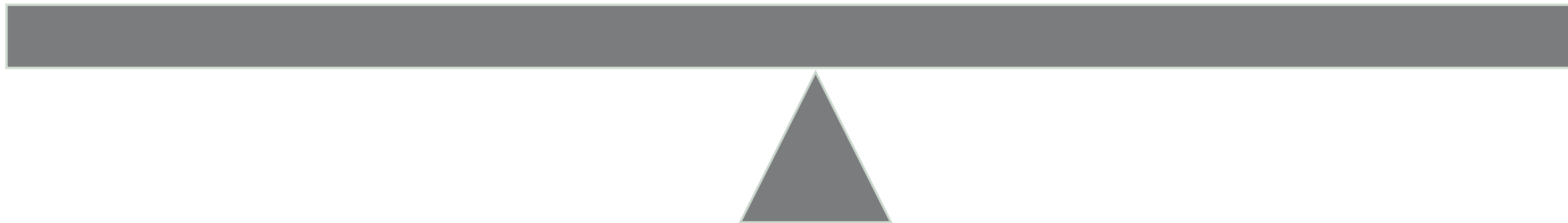
Conflicting treatment goals in pregnancy

IVF/pregnancy

- Stimulation of ovulation may induce angioedema attacks
- Emotional distress may even further evoke angioedema attacks
- Treatment options limited to pdC1-INH concentrates

Improved HAE control

- More treatment options available for both prophylaxis and acute treatment
- Aimed at reduced oestrogen levels



Patient's concerns regarding IVF and pregnancy



Poll: Which strategy would you recommend for this patient?

- A. Increase dose of IV C1-INH concentrate
- B. Reduce dosing interval of IV C1-INH concentrate
- C. Consider other route of administration of C1-INH concentrate
- D. Refer to psychologist with HAE expertise
- E. Wait and see

Course 2021

- IVF postponed
- Continued angioedema attacks despite frequent IV C1-INH concentrate use
- Poor disease control (AECT 7)



Course 2021

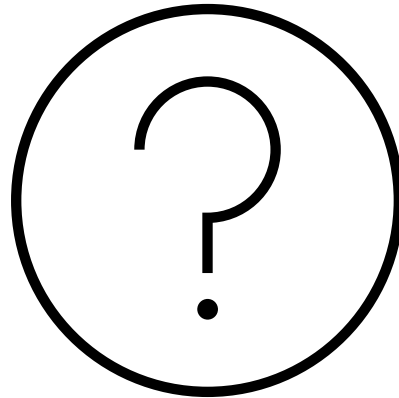
- IVF postponed
- Continued angioedema attacks despite frequent IV C1-INH concentrate use
- Poor disease control (AECT 7)

- Compassionate use of SC C1-INH granted
- 2 minor attacks in three months, AECT improved (AECT 14)
- Increased confidence in HAE course during a future pregnancy

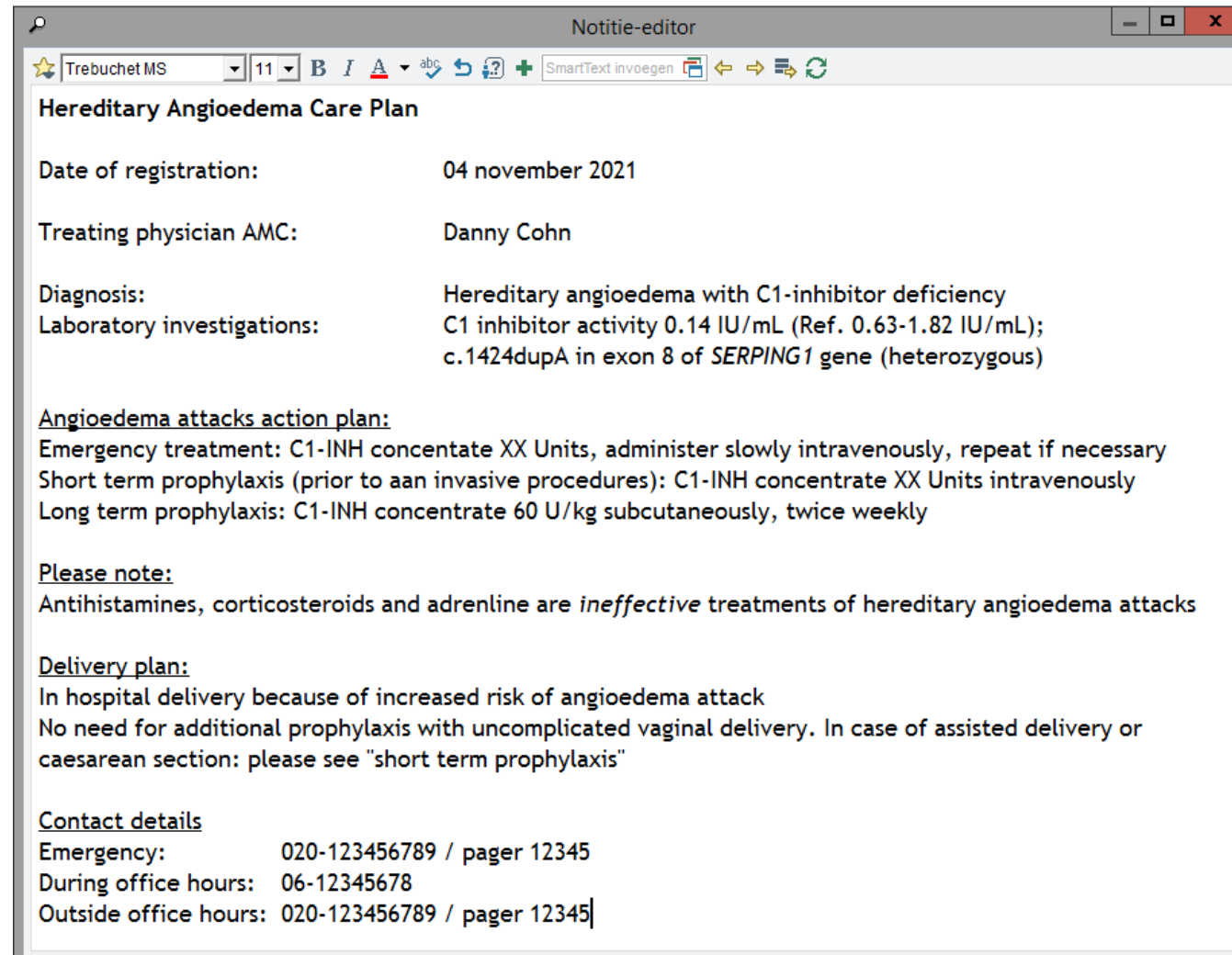
**According to the Summary of Product Characteristics of SC C1-INH (Berinert® 2000/3000) or IV C1-INH (Berinert® 500/1500), there are limited data that suggests no increased risk from the use of C1-INH products in pregnant women*

Question to the panel

What special recommendations do you have for an individualised care plan regarding pregnancy and delivery?



Individual care plan



The image shows a screenshot of a 'Notitie-editor' (note editor) window. The window title is 'Notitie-editor'. The toolbar includes a font dropdown set to 'Trebuchet MS', a size dropdown set to '11', and various text formatting icons (bold, italic, underline, text color, background color, bulleted list, numbered list, link, unlink, redo, undo). The main text area contains the following content:

Hereditary Angioedema Care Plan

Date of registration: 04 november 2021

Treating physician AMC: Danny Cohn

Diagnosis: Hereditary angioedema with C1-inhibitor deficiency

Laboratory investigations: C1 inhibitor activity 0.14 IU/mL (Ref. 0.63-1.82 IU/mL);
c.1424dupA in exon 8 of *SERPING1* gene (heterozygous)

Angioedema attacks action plan:
Emergency treatment: C1-INH concentrate XX Units, administer slowly intravenously, repeat if necessary
Short term prophylaxis (prior to an invasive procedure): C1-INH concentrate XX Units intravenously
Long term prophylaxis: C1-INH concentrate 60 U/kg subcutaneously, twice weekly

Please note:
Antihistamines, corticosteroids and adrenaline are *ineffective* treatments of hereditary angioedema attacks

Delivery plan:
In hospital delivery because of increased risk of angioedema attack
No need for additional prophylaxis with uncomplicated vaginal delivery. In case of assisted delivery or caesarean section: please see "short term prophylaxis"

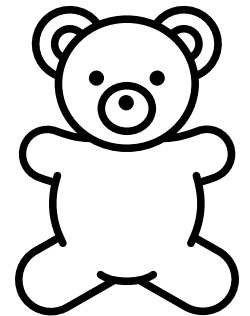
Contact details
Emergency: 020-123456789 / pager 12345
During office hours: 06-12345678
Outside office hours: 020-123456789 / pager 12345

Delivery and HAE

In-hospital delivery recommended

No need for preprocedural prophylaxis if unassisted vaginal delivery

Acute treatment readily available



Take-home messages

IVF and pregnancy may increase angioedema attacks for multiple reasons:

- Emotional distress
- Stimulation of ovulation
- Increased oestrogen levels

HAE treatment options are limited to pdC1-INH concentrate in pregnancy

Personalised treatment and delivery plan highly recommended