

Table SI. Patient demographics (n = 1,239). Functional complement C<sub>1</sub> esterase inhibitor (C<sub>1</sub>INH) normal group

	Men, %	Women, %
Gender	33	67
Age		
0–19 years	6.9	7.4
20–39 years	23.6	32.1
40–59 years	43.6	38.3
60–79 years	24.6	20.4
≥80 years	1.2	1.8

Table SII. Functional complement  $C_1$  esterase inhibitor ( $C_1$ INH) test normal subgroup ( $C_1$ INH test  $\geq 50\%$ ). A subset of patients ( $n = 621$ ) reporting both swellings of the tongue and answered affirmatively to the question: "Have you ever had swellings of the lips, the tongue or the skin which developed quickly and disappeared after a few days?" The temporal pattern of angioedema: debut, remittance and relapse in patients

	<i>n</i> (%) <sup>a</sup>
Have you had hives as explained below?	
Yes	396 (63.8)
No	107 (17.2)
Don't know	99 (15.9)
Have you ever had angioedema and hives at the same time?	
Yes	235 (37.8)
No	219 (35.3)
Don't know	153 (24.6)
Do you still suffer from angioedema?	
Yes	356 (57.3)
No	92 (14.8)
Don't know	153 (24.6)
How many times do you think you have had angioedema? Number of attacks with >5-year interval?	
1 time	133 (21.4)
2 times	62 (10.0)
3 times	37 (6.0)
4 times	24 (3.9)
$\geq 5$ times	246 (39.6)
For how long have you had angioedema episodes?	
1 week	295 (47.5)
1–2 weeks	61 (9.8)
3–4 weeks	15 (2.4)
1–2 months	15 (2.4)
2–3 months	9 (1.4)
>3 months	52 (8.4)
How many years has past since your first attack?	
15 years	5 (10.5)
14–10 years	21 (25.0)
9–5 years	85 (20.9)
4–3 years	69 (34.8)
<2 years	110 (14.9)
Maximum interval between attacks	
$\geq 15$ years	11 (1.8)
14–10 years	7 (1.1)
9–5 years	40 (6.4)
4–3 years	44 (7.1)
2 years	38 (6.1)
1 year	65 (10.5)
6 months	81 (13.0)
3 months	79 (12.7)
1 month	57 (9.2)
2 weeks	43 (6.9)
Episodes every week	62 (10.0)
Age at first attack	
<1 year	3 (0.5)
1–2 years	8 (1.3)
3–4 years	10 (1.6)
5–10 years	24 (3.9)
11–20 years	88 (14.2)
21–40 years	193 (31.1)
41–60 years	223 (35.9)
61–80 years	63 (10.1)
>80 years	1 (0.2)

<sup>a</sup>Total does not add up to 100% due to persons giving multiple responses.

Table SIII. Functional complement  $C_1$  esterase inhibitor ( $C_1$  INH) test normal group. A selected group of patients ( $n = 621$ ) reporting both swellings of the tongue and answering affirmatively to the question: "Have you ever had swellings of the lips, the tongue or the skin which developed quickly and disappeared after a few days?". Spatial pattern and quality of symptoms ranked by frequency

	<i>n</i> (%) <sup>a</sup>
Location of symptoms	
Lips	13 (82.6)
Head and neck	753 (60.8)
Eyes	747 (60.3)
Tongue	621 (100.0)
Arms	257 (41.4)
Legs	249 (40.1)
Throat without choking sensation	293 (47.2)
Thorax	146 (23.7)
Abdominal area	136 (21.9)
Genitalia	147 (23.7)
Throat with choking sensation	147 (23.7)
Breasts	87 (14.0)
Symptoms during attacks	
Itching swellings	449 (72.3)
Difficulty breathing	396 (63.8)
Stinging or burning swellings	589 (47.6)
Dizzy	306 (49.3)
Choking sensation	217 (34.9)
Abdominal pain	138 (22.2)
Near fainting	123 (19.8)
Diarrhoea	103 (16.6)
Vomiting	68 (11.0)
Fainting	44 (7.1)

<sup>a</sup>Total does not add up to 100% due to persons giving multiple responses.