

SUPPLEMENTARY MATERIAL

Supplemental material 1. Basal characteristics of patients. C1-C1-INH, C4 and C1 INH activity values before SC C1INH 1 LTP. On demand treatment and LTP in the previous year of the use of SC C1INH. Frequency and severity of attacks before use of SC C1INH LTP.

Case number/ Variable	1	2	3	4	5	6	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	
Age of beginning SC C1INH LTP	48	47	60	28	38	57	12	29	20	28	71	36	43	49	58	32	56	58	65	4	34	
Gender	F	M	F	F	F	F	M	F	F	M	F	M	M	F	M	F	F	F	F	F	F	F
Weight (kg)	60	84	69	65	53	65	65	60	110	136	80	65	102	61	90	63	54	82	65	25	62	
BMI	21.5	32.0	25.3	23.9	20.7	26.0	28.1	20.3	31.1	44.4	30.1	22.2	28.5	23.8	31.1	23.1	22.8	33.7	27.5	21.6	24.2	
HAE type	I	I	I	I	I	I	I	I	I	I	II	I	I	I	I	I	I	I	I	I	I	I
Co-morbidities or other relevant conditions																						
Pregnancy during LTP																	X					X
Hypothyroidism																			X			X
Anxiety	X	X					X	X	X	X	X	X					X					
Depression	X										X						X					
Bipolar disorder	X																					
Cognitive impairment				X																		
Phobic disorder																						
Adjustment disorder							X															
Obesity		X					X		X	X					X			X	X			
Type2 diabetes mellitus				X																		
Dyslipidemia		X	X	X					X	X	X			X	X			X	X			X
Fatty liver disease															X		X					
Gallstone														X								
Hyperuricemia																		X				
Arterial hypertension						X				X	X		X						X			
Ovarian endometrioma				X																		

Irritable bowel syndrome	X																				
Atopia/allergy						X	X		X		X		X		X						X
Psoriasis														X							
Vascular disorders											X										
Previous stroke											X										
Myopericarditis										X											
Bilateral hypoacusia				X																	
Bilateral oophorectomy								X													
C1 (mg/dL)	6.1-7.4	7.8-20.0	7.6-22.5	3.0-17.6	3.1-8.1	2.9-8.7	5.7-11.2	5.7-16.5	6.5-11.1	9.9-11.6	25.1-49.1	2.8-14.9	5.3-11.9	0.1-13.2	7.6-16.1	2.9-12.0	2.8-9.4	6.1-6.5	8.9	14.0	9.1-11.0
C1 activity (%)	7.2-23.3	8.7-23.9	33.0	25.5-71.8	41.2	10.5-12.2	5.1-17.2	7.4-20.0	20.1-24.9	8.9	25.3-36.0	1.6	1.9-49.3	15.3-20.7	15.1-18.2	20.1-22.9	22.1-36.0	25.9-33.3	20.9-25.1	33	30.0-36.0
C4 (mg/dL)	3.2-9.0	5.00-10.9	14.4-21.9	6.2-19.9	4.7-8.9	3.4-15.2	5.1-14.2	8.5-20.6	3.7-20.2	8.3-19.9	2.5-17.9	2.1-32.3	3.0-14.0	7.7-20.3	14.4-32.0	2.3-9.1	1.4-12.0	2.70-9.0	8.0-8.7	0.1	8.9
On demand treatment or LTP during the previous year																					
IRT with IV C1-INH					X					X	X				X						X
LTP with IV C1-INH	X	X	X	X		X	X	X	X			X	X	X			X				
Tranexamic acid							X				X									X	X
Attenuated androgens		X				X		X		X	X	X	X			X		X	X		
Attacks during the previous year																					
≥1/week	X	X	X	X	X		X		X		X		X			X				X	X
<1/week - ≥1/month								X				X		X	X		X	X			
<1/month						X				X											
Severity of attacks																					
Mild																					X
Moderate										X											
Severe	X	X	X	X	X	X	X	X	X		X	X	X	X	X	X	X	X	X	X	X
Location of attacks																					
Abdominal	X	X	X	X	X	X	X	X	X		X	X		X		X	X			X	X
Facial	X	X		X		X	X	X		X					X		X				

Peripheral	X	X	X	X	X	X	X	X	X	X	X	X	X	X	X	X	X	X	X	X	X
Cervical				X																	
Oropharyngeal/ laryngeal					X	X	X		X		X					X		X	X		
Genital		X				X	X		X						X	X					X

BMI, Body mass index; C1-INH, C1-esterase inhibitor; HAE, Hereditary angioedema; IRT, Individual replacement therapy; IV, Intravenous; LTP, Long-term prophylaxis