

Table IVS - Severity of the disease in the total cohort, in first HAE patients in family and in subsequent HAE patients in family.

Characteristics	Total (N = 126)	First in family* (N = 52)	Not first in family (N = 74)
Severity classification			
Mean \pm SD (minimum - maximum)	2.6 \pm 1.3 (1.0 -5.0)	2.6 \pm 1.4 (1.0 -5.0)	2.6 \pm 1.3 (1.0 -5.0)
Median (Q1-Q3)	2.0 (2.0 - 3.0)	2.0 (2.0 - 3.2)	2.0 (2.0 - 3.0)
Severity classification, n (%)			
1	20 (15.9)	10 (19.2)	10 (13.5)
2	63 (50.0)	26 (50.0)	37 (50.0)
3	13 (10.3)	3 (5.8)	10 (13.5)
4	6 (4.8)	2 (3.8)	4 (5.4)
5	24 (19.0)	11 (21.2)	13 (17.6)

HAE: hereditary angioedema; N: total number of patients; n: number of patients with available data; Q1: 1st quartile; Q3: 3rd quartile; SD, standard deviation; *first patients diagnosed in a family (if 2 or more patients are diagnosed in the same initial year, they are all considered as "First in Family").