

PRECISION AND FUTURE MEDICINE

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Supplementary Table 2. Distribution of general, clinical characteristics, and causes of death and death risk for HHT (n=2,874)

Variable	HHT	Crude HR (95% CI)	Adjusted HR (95% CI) ^{a)}
Age (yr)	50.9±20.3		
0–9	107 (3.72)	0.00 (0.00–999)	0.00 (0.00–999)
10–19	148 (5.15)	3.36 (0.31–37.0)	3.18 (0.28–35.2)
20–29	274 (9.53)	1.0	1.0
30–39	226 (7.86)	7.81 (0.96–63.4)	8.08 (0.99–65.7)
40–49	408 (14.2)	8.45 (1.11–64.6) ^{c)}	7.97 (1.04–61.0) ^{d)}
50–59	617 (21.5)	13.4 (1.82–98.1) ^{c)}	12.4 (1.68–91.2) ^{d)}
60–69	530 (18.4)	32.2 (4.46–232.2) ^{b)}	30.4 (4.20–220.1) ^{b)}
70–79	440 (15.3)	96.8 (13.5–692.9) ^{b)}	92.6 (12.9–665.7) ^{b)}
≥80	124 (4.31)	287 (39.8–2,065.3) ^{b)}	274.6 (37.9–1,988) ^{b)}
Male sex	1,117 (38.8)	1.49 (1.19–1.86) ^{b)}	1.70 (1.35–2.14) ^{b)}
Income level			
Upper	1,269 (44.2)	1.0	1.0
Medium	767 (26.7)	0.74 (0.55–0.99) ^{c)}	1.07 (0.78–1.46)
Lower	838 (29.1)	1.18 (0.92–1.52)	1.34 (1.03–1.75) ^{d)}
Comorbidities			
Hypertension	448 (15.6)	2.09 (1.62–2.70) ^{b)}	1.12 (0.85–1.48)
Diabetes mellitus	290 (10.1)	1.88 (1.39–2.54) ^{b)}	0.97 (0.71–1.32)
Dyslipidemia	155 (5.39)	0.98 (0.55–1.74)	0.65 (0.36–1.20)
Myocardial infarction	12 (0.42)	5.11 (2.11–12.3) ^{b)}	2.08 (0.84–5.17)
Heart failure	27 (0.94)	5.91 (3.14–11.1) ^{b)}	1.82 (0.93–3.54)
Atrial fibrillation	16 (0.56)	2.87 (0.71–11.5)	0.98 (0.24–4.04)
Ischemic stroke	43 (1.50)	2.91 (1.55–5.46) ^{b)}	0.82 (0.42–1.58)
Hemorrhagic stroke	5 (0.17)	9.91 (3.18–30.8) ^{b)}	6.16 (1.87–20.2) ^{d)}
Transient ischemic attack	7 (0.24)	3.31 (0.83–13.2)	1.68 (0.40–7.03)
Chronic kidney disease	12 (0.42)	2.47 (0.79–7.69)	2.67 (0.83–8.56)
Malignancy neoplasm	28 (0.97)	7.32 (4.00–13.3) ^{b)}	4.86 (2.55–9.28) ^{b)}
Pulmonary arterial hypertension	4 (0.14)	14.9 (3.69–60.1) ^{b)}	7.36 (1.68–32.2) ^{d)}
Epistaxis	57 (1.98)	-	-
Hemoptysis	10 (0.35)	-	-
Arteriovenous malformation	28 (0.97)	2.88 (1.77–4.71) ^{b)}	1.82 (1.09–3.05) ^{d)}
Anemia	52 (1.81)	9.44 (3.57–25.5) ^{b)}	2.95 (0.99–8.80)
Liver disease	79 (2.75)	2.88 (1.77–4.71) ^{b)}	1.82 (1.09–3.05) ^{d)}
Bile disease	9 (0.31)	9.44 (3.57–25.5) ^{b)}	2.95 (0.99–8.80)
Death	306 (10.7)	-	-
Cause of death			
Certain infections and parasitic diseases (A00-B99)	2 (0.65)	-	-
Malignancy neoplasm (C00-C97)	67 (21.9)	-	-
Benign neoplasm (D00-D48) & Diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism (D50-D89)	7 (2.29)	-	-

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PRECISION AND FUTURE MEDICINE

Survival and death risk for aPAH

Supplementary Table 2. Continued

Variable	HHT	Crude HR (95% CI)	Adjusted HR (95% CI) ^{a)}
Endocrine, nutritional, and metabolic diseases (E00-E90)	10 (3.27)	-	-
Mental and behavioral disorders (F01-F99)	3 (0.98)	-	-
Diseases of the nervous system (G00-G98)	9 (2.94)	-	-
Diseases of the circulatory system (I00-I99)	70 (22.9)	-	-
Diseases of the respiratory system (J00-J98)	27 (8.82)	-	-
Diseases of the digestive system (K00-K92)	23 (7.52)	-	-
Diseases of the skin and subcutaneous tissue (L00-L99)	0	-	-
Diseases of the musculoskeletal system and connective tissue (M00-M99)	0	-	-
Diseases of the genitourinary system (N00-N99)	9 (2.94)	-	-
Pregnancy, childbirth and the puerperium (O00-O99)	0	-	-
Certain conditions originating in the perinatal period (P00-P96)	0	-	-
Congenital malformation, deformations and chromosomal abnormalities (Q00-Q99)	1 (0.323)	-	-
Symptoms, signs, and abnormal clinical and laboratory findings, not elsewhere classified (R00-R99)	13 (4.25)	-	-
Injury, poisoning, and certain other consequences of external causes (S00-T98)	12 (3.92)	-	-
Not provided	53 (17.3)	-	-

Values are presented as mean \pm standard deviation or number (%).

HHT, hereditary hemorrhagic telangiectasia; HR, hazard ratio; CI, confidence interval.

^{a)}Estimated by a Cox proportional hazard model analysis using the variables indicated in the table; ^{b)}P<0.001; ^{c)}P<0.05.