

Supplementary Table 1. Survival rate (%) and 95% confidence interval overall, by sex, by age group, by cause of death, by HHT, by SLE, and by AIDS for associated pulmonary artery hypertension (ICD-10: I27.2)

Year	Overall	By sex		By age group										By cause of death			By HHT		By SLE		By AIDS	
		Female	Male	0-9 years	10-19 years	20-29 years	30-39 years	40-49 years	50-59 years	60-69 years	70-79 years	≥80 years	Non-cardiovascular	Cardiovascular	Non-HHT	HHT	Non-SLE	SLE	Non-AIDS	AIDS		
1	81.1 (80.5-81.7)	83.6 (82.8-84.3)	77.3 (76.2-78.3)	87.5 (85.5-89.3)	91.9 (86.5-95.2)	95.8 (93.4-97.4)	95.5 (91.5-95.0)	91.6 (89.9-93.0)	88.5 (87.0-89.8)	82.0 (80.5-83.4)	77.9 (76.6-79.2)	67.2 (65.6-68.9)	51.9 (50.4-53.4)	49.4 (46.9-51.8)	81.1 (80.5-81.7)	75.0 (12.7-96.0)	80.7 (80.0-81.3)	90.2 (87.6-92.2)	81.1 (80.5-81.7)	90.0 (47.0-98.0)		
2	73.9 (73.2-74.6)	77.3 (76.4-78.2)	68.6 (67.4-69.8)	86.3 (84.3-88.2)	89.1 (83.1-93.1)	94.8 (92.2-96.6)	90.2 (87.9-92.1)	88.0 (86.2-89.8)	83.6 (81.9-85.2)	75.0 (73.3-76.7)	69.0 (67.5-70.5)	54.6 (52.8-56.3)	34.1 (32.7-35.5)	33.8 (31.5-36.2)	73.9 (73.2-74.6)	75.0 (12.7-96.0)	73.4 (72.6-74.1)	86.4 (83.5-88.9)	73.9 (73.2-74.6)	90.0 (47.0-98.0)		
3	68.6 (67.8-69.4)	72.1 (71.1-73.0)	63.3 (62.0-64.5)	86.1 (84.0-88.0)	87.7 (81.4-92.0)	92.4 (89.2-94.6)	88.3 (85.8-90.4)	85.0 (82.8-86.9)	80.2 (78.3-81.9)	70.2 (68.3-72.0)	62.2 (60.6-63.8)	44.9 (43.0-46.7)	23.8 (22.6-25.1)	22.4 (20.3-24.5)	68.6 (67.8-69.4)	75.0 (12.7-96.0)	68.0 (67.2-68.8)	82.6 (79.3-85.4)	68.6 (67.8-69.4)	90.0 (47.0-98.0)		
4	64.0 (63.2-64.9)	67.6 (66.5-68.6)	58.5 (57.2-59.9)	85.6 (83.5-87.5)	86.7 (80.1-91.3)	91.6 (88.3-94.1)	87.3 (84.6-89.5)	81.5 (79.0-83.7)	76.8 (74.7-78.7)	65.2 (63.1-67.1)	56.3 (54.6-58.0)	37.0 (35.0-38.9)	16.1 (15.0-17.2)	15.1 (13.4-17.0)	64.0 (63.2-64.8)	37.5 (1.00-80.0)	63.4 (62.5-64.2)	78.7 (75.1-81.9)	64.0 (63.2-64.8)	78.7 (38.0-94.2)		
5	60.3 (59.4-61.1)	63.8 (62.7-64.9)	54.8 (53.4-56.2)	85.4 (83.2-87.3)	86.7 (80.1-91.3)	90.8 (87.2-93.4)	86.0 (83.1-88.4)	79.8 (77.3-82.2)	74.1 (71.9-76.2)	61.5 (59.3-63.6)	50.7 (48.8-52.5)	30.5 (28.5-32.5)	10.9 (10.0-11.8)	9.37 (7.97-10.9)	60.3 (59.4-61.1)	-	59.6 (58.7-60.5)	75.6 (71.7-79.1)	60.2 (59.4-61.1)	78.7 (38.0-94.2)		
6	56.5 (55.6-57.5)	60.1 (58.9-61.2)	51.0 (49.5-52.5)	85.0 (82.8-87.0)	85.5 (78.4-90.4)	90.8 (87.2-93.4)	85.4 (82.5-87.9)	78.4 (75.7-80.8)	70.6 (68.1-72.9)	56.9 (54.5-59.1)	45.1 (43.1-47.0)	25.1 (23.0-27.1)	6.77 (6.06-7.54)	4.95 (3.94-6.12)	56.6 (55.6-57.5)	-	55.7 (54.8-56.7)	74.3 (70.2-77.9)	56.5 (55.6-57.5)	78.7 (38.0-94.2)		
7	53.8 (52.8-54.8)	57.3 (56.0-58.5)	48.3 (46.7-49.9)	84.8 (82.5-86.8)	83.7 (75.5-89.3)	88.9 (84.7-92.1)	84.1 (80.9-86.8)	76.8 (73.9-79.4)	67.4 (64.7-69.9)	54.6 (52.2-57.0)	40.7 (38.6-42.8)	21.3 (19.2-23.4)	4.11 (3.55-4.72)	2.97 (2.20-3.91)	53.8 (52.8-54.8)	-	52.9 (51.9-53.9)	72.1 (67.7-76.0)	53.7 (52.8-54.7)	78.7 (38.0-94.2)		
8	50.7 (49.7-51.8)	54.4 (53.1-55.8)	45.0 (43.2-46.7)	84.8 (82.5-86.8)	83.7 (75.5-89.3)	88.2 (83.7-91.5)	82.1 (78.5-85.2)	75.7 (72.7-78.5)	63.4 (60.3-66.4)	51.1 (48.5-53.7)	36.2 (33.8-38.5)	16.6 (14.5-18.9)	2.12 (1.73-2.58)	0.92 (0.53-0.15)	50.8 (49.7-51.8)	-	49.8 (48.7-50.9)	70.0 (65.3-74.1)	50.7 (49.6-51.8)	78.7 (38.0-94.2)		
9	48.3 (47.1-49.5)	52.1 (50.6-53.6)	42.4 (40.5-44.2)	84.4 (82.0-86.5)	83.7 (75.5-89.3)	88.4 (77.7-88.4)	81.0 (77.0-84.4)	73.6 (70.1-76.7)	61.2 (57.7-64.4)	47.9 (45.0-50.8)	32.7 (30.0-35.3)	13.6 (11.4-18.9)	0.94 (0.69-1.27)	0.06 (0.006-0.36)	48.3 (47.2-49.5)	-	47.6 (46.3-48.8)	65.3 (60.0-70.1)	48.3 (47.1-49.5)	78.7 (38.0-94.2)		
10	46.3 (45.0-47.6)	50.3 (48.6-52.0)	40.1 (38.0-42.2)	84.4 (82.0-86.5)	79.3 (66.2-87.7)	82.3 (75.3-87.4)	81.0 (77.0-84.4)	73.1 (69.6-76.4)	59.8 (55.9-63.5)	44.9 (41.4-48.3)	27.9 (24.6-31.3)	11.3 (8.60-14.3)	0.22 (0.11-0.40)	0.00 (0.00-0.00)	46.3 (45.0-47.6)	-	45.3 (43.9-46.7)	64.5 (59.1-69.5)	46.5 (45.1-47.8)	78.7 (38.0-94.2)		

Values are presented as survival rate (95% confidence interval).

HHT, hereditary hemorrhagic telangiectasia; SLE, systemic lupus erythematosus; AIDS, acquired immune deficiency syndrome; ICD-10, 10th revision of the International Statistical Classification of Diseases.