

Supplemental table 2 Prevalence, event rates and age at onset of individual outcomes †					
	All (213)	Men (83)		Women (130)	
		Classical (57)	Non-classical (26)	Classical (98)	Non-classical (32)
1. Sudden cardiac death					
Number of patients	4 (2%)	4 (7%)	0 (0%)	0 (0%)	0 (0%)
Event rate, with 95%-CI	0.6 (0.2-1.4)	2.6 (0.8-6.2)	0 (-)	0 (-)	0 (-)
Age at death	48 (47-56)	48 (47-56)	(-)	(-)	(-)
2. Sudden cardiac arrest					
Number of patients	6 (3%)	1 (2%)	3 (12%)	2 (2%)	0 (0%)
Event rate, with 95%-CI	0.8 (0.3-1.8)	0.6 (0.03-3.2)	2.5 (0.6-6.9)	0.6 (0.1-2.1)	0 (-)
Age at first event	66 (46-73)	46 (-)	67 (64-67)	65 (57-73)	(-)
3. Sustained ventricular tachycardia					
Number of patients	4 (2%)	0 (0%)	2 (8%)	2 (2%)	0 (0%)

Event rate, with 95%-CI	0.6 (0.2-1.4)	0 (-)	1.7 (0.3-5.6)	0.6 (0.1-2.1)	0 (-)
Age at first event	66 (51-73)	(-)	66 (64-67)	62 (51-73)	(-)
4. Ventricular fibrillation					
Number of patients	2 (1%)	1 (2%)	0 (0%)	1 (1%)	0 (0%)
Event rate, with 95%-CI	0.3 (0.1-0.9)	0.6 (0.03-3.2)	0 (-)	0.3 (0.02-1.5)	0 (-)
Age at first event	59 (46-73)	46 (-)	(-)	73 (-)	(-)
5. Second- degree AV block, Mobitz II					
Number of patients	1(1%)	0 (0%)	1 (4%)	0 (0%)	0 (0%)
Event rate, with 95%-CI	0.1 (0.01-0.7)	0 (-)	0.8 (0.04-4.1)	0 (-)	0 (-)
Age at first documentation	65 (-)	(-)	65 (-)	(-)	(-)
6. Third- degree AV block					
Number of patients	9 (4%)	1 (2%)	4 (15%)	4 (4%)	0 (0%)
Event rate, with 95%-CI	1.3 (0.6-2.3)	0.6 (0.03-3.2)	3.4 (1.1-8.1)	1.2 (0.4-3.0)	0 (-)

Age at first documentation	60 (50-72)	60 (-)	58 (50-63)	59 (54-72)	(-)
7. Sinusarrest					
Number of patients	7 (3%)	3 (5%)	2 (8%)	2 (2%)	0 (0%)
Event rate, with 95%-CI	1.0 (0.4-2.0)	1.9 (0.5-5.3)	1.7 (0.3-5.6)	0.6 (0.1-2.1)	0 (-)
Age at first documentation	58 (48-69)	58 (48-60)	60 (58-62)	59 (49-69)	(-)
8. PM, ICD and CRT-D implantation					
Number of patients	36 (17%)	9 (16%)	11 (42%)	14 (14%)	2 (6%)
PM	14 (7%)	4 (7%)	3 (12%)	5 (5%)	2 (6%)
PM + ICD upgrade at later stage	3 (1%)	0 (0%)	2 (8%)	1 (1%)	0 (0%)
ICD	7 (3%)	0 (0%)	3 (12%)	4 (4%)	0 (0%)
DDD-ICD	9 (4%)	3 (5%)	2 (8%)	4 (4%)	0 (0%)
CRT-D	2 (1%)	2 (4%)	0 (0%)	0 (0%)	0 (0%)

PM + CRT-D upgrade at later stage	1 (0.5%)	0 (0%)	1 (4%)	0 (0%)	0 (0%)
9. Coronary artery disease					
Number of patients	18 (9%)	9 (16%)	3 (12%)	6 (6%)	0 (0%)
Event rate, with 95%-CI	2.5 (1.6-3.9)	5.8 (2.8-10.7)	2.5 (0.6-6.9)	1.9 (0.8-3.9)	0 (-)
Age at first documentation	55 (39-77)	51 (46-64)	55 (53-67)	57 (39-77)	(-)
10. Percutaneous coronary intervention					
Number of patients	7 (3%)	3 (5%)	2 (8%)	2 (2%)	0 (0%)
Event rate, with 95%-CI	1.0 (0.4-2.0)	1.9 (0.5-5.3)	1.7 (0.3-5.6)	0.6 (0.1-2.1)	0 (-)
Age at first documentation	53 (39-67)	50 (48-64)	61 (55-67)	46 (39-53)	(-)
11. Coronary artery bypass graft surgery					
Number of patients	4 (2%)	3 (5%)	1 (4%)	0 (0%)	0 (0%)
Event rate, with 95%-CI	0.6 (0.2-1.4)	1.9 (0.5-5.3)	0.8 (0.04-4.1)	0 (-)	0 (-)
Age at first surgery	58 (54-69)	58 (54-58)	69	(-)	(-)

12. Systolic dysfunction on MRI or echocardiography					
Number of patients	28 (13%)	11 (19%)	8 (31%)	8 (8%)	1 (3%)
Event rate, with 95%-CI	3.9 (2.7-5.6)	7.1 (3.7-12.4)	6.7 (3.1-12.8)	2.5 (1.2-4.7)	0.9 (0.04-4.3)
Age at first documentation	60 (29-77)	52 (29-64)	64 (53-69)	65 (54-77)	72 (-)
13. Left ventricular outflow tract obstruction					
Number of patients	8 (4%)	2 (4%)	1 (4%)	4 (4%)	1 (3%)
Event rate, with 95%-CI	1.1 (0.5-2.1)	1.3 (0.2-4.3)	0.8 (0.04-4.1)	1.2 (0.4-3.0)	0.9 (0.04-4.3)
Age at first documentation	56 (31-65)	43 (31-55)	58 (-)	57 (46-61)	65 (-)
14. Moderate or severe heart valve disease					
Number of patients	39 (18%)	14 (25%)	6 (23%)	16 (16%)	3 (9%)
Event rate, with 95%-CI	5.5 (4.0-7.4)	9.1 (5.2-14.8)	5.0 (2.0-10.5)	5.0 (3.0-7.9)	2.6 (0.7-7.2)
Age at first documentation	58 (36-77)	51 (36-61)	65 (51-70)	61 (47-77)	74 (65-76)

15. Heart surgery/ intervention*					
Number of patients	10 (5%)	2 (4%)	3 (12%)	5 (5 %)	0 (0%)
Event rate, with 95%-CI	1.4 (0.7-2.5)	1.3 (0.2-4.3)	2.5 (0.6-6.9)	1.6 (0.6-3.5)	0 (-)
Age at first surgery/ intervention	63 (49-72)	53 (49-57)	64 (64-68)	61 (52-72)	(-)

Data are presented as number (percentage) or median (range). All event rates are per 1000 patient years from age 15 onwards.

† Events per 1000 person years from age of 15 = (Number of events/ Cumulative follow up from age 15)x1000

* heart surgery or intervention other than percutaneous coronary intervention or coronary artery bypass graft surgery

AV = atrioventricular; CRT = cardiac resynchronization therapy; ICD = implantable cardiac defibrillator; PM = pacemaker