

# Presentation and Outcomes of Patients with Brugada Syndrome in the Arab Gulf Region: *Preliminary Findings from the Gulf Brugada Registry*

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*VeniceArrhythmias 18<sup>th</sup> October , 2015*

# Disclosure

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**NO CONFLICT OF  
INTEREST TO  
DECLARE**

# Right Bundle Branch Block, Persistent ST Segment Elevation and Sudden Cardiac Death: A Distinct Clinical and Electrocardiographic Syndrome

## A Multicenter Report

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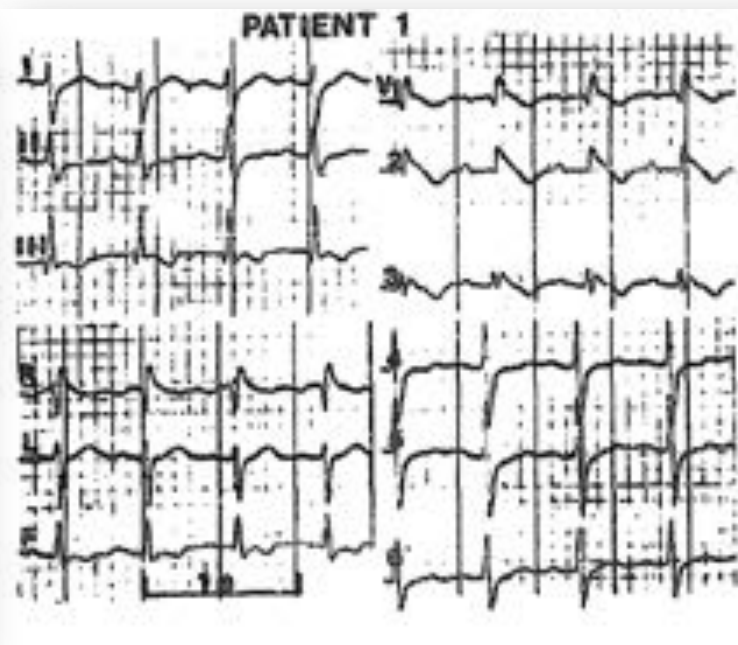
*Aalst, Belgium and Barcelona, Spain*

**Table 1.** Clinical and Electrocardiographic Characteristics of the Eight Patients Studied

Pt No.	Age (yr)*/ Gender	Cycle Length (ms)	Interval (ms)		
			PR	QRS	QT
1	2/M	600	170	130	340
2	2/F	740	130	120	360
3	46/M	705	180	145	410
4	48/M	750	180	150	380
5	44/M	660	160	130	345
6	53/M	840	190	140	405
7	8/F	740	180	135	330
8	26/M	760	170	120	340

\*Age at the time of the first episode of (aborted) sudden cardiac death.

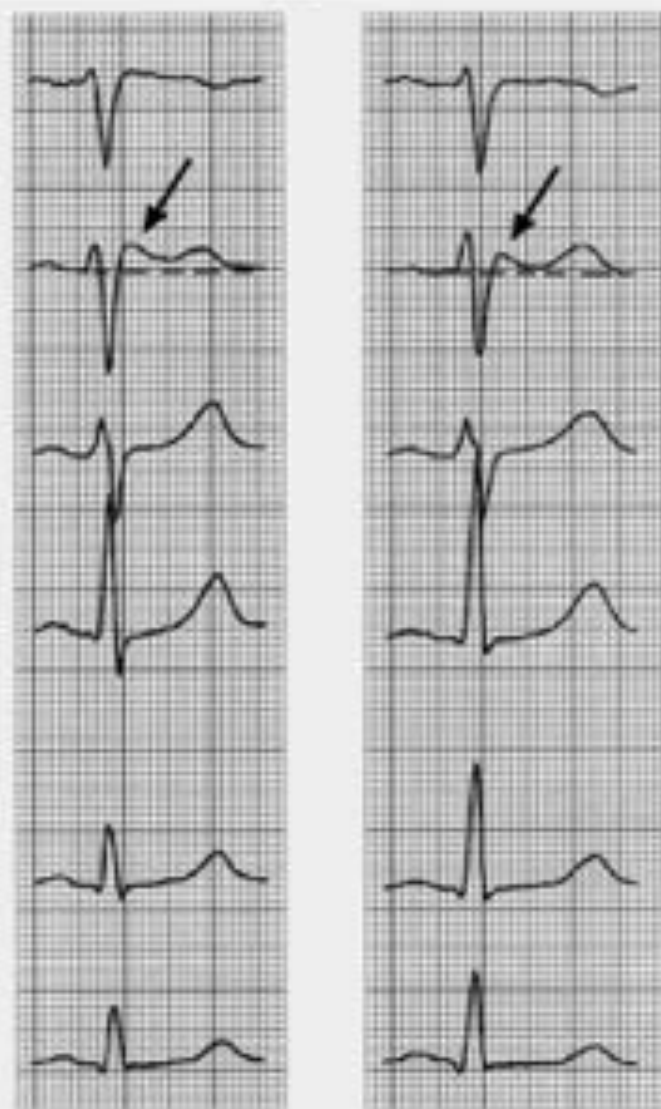
F = female; M = male; Pt = Patient.



**Type 1**



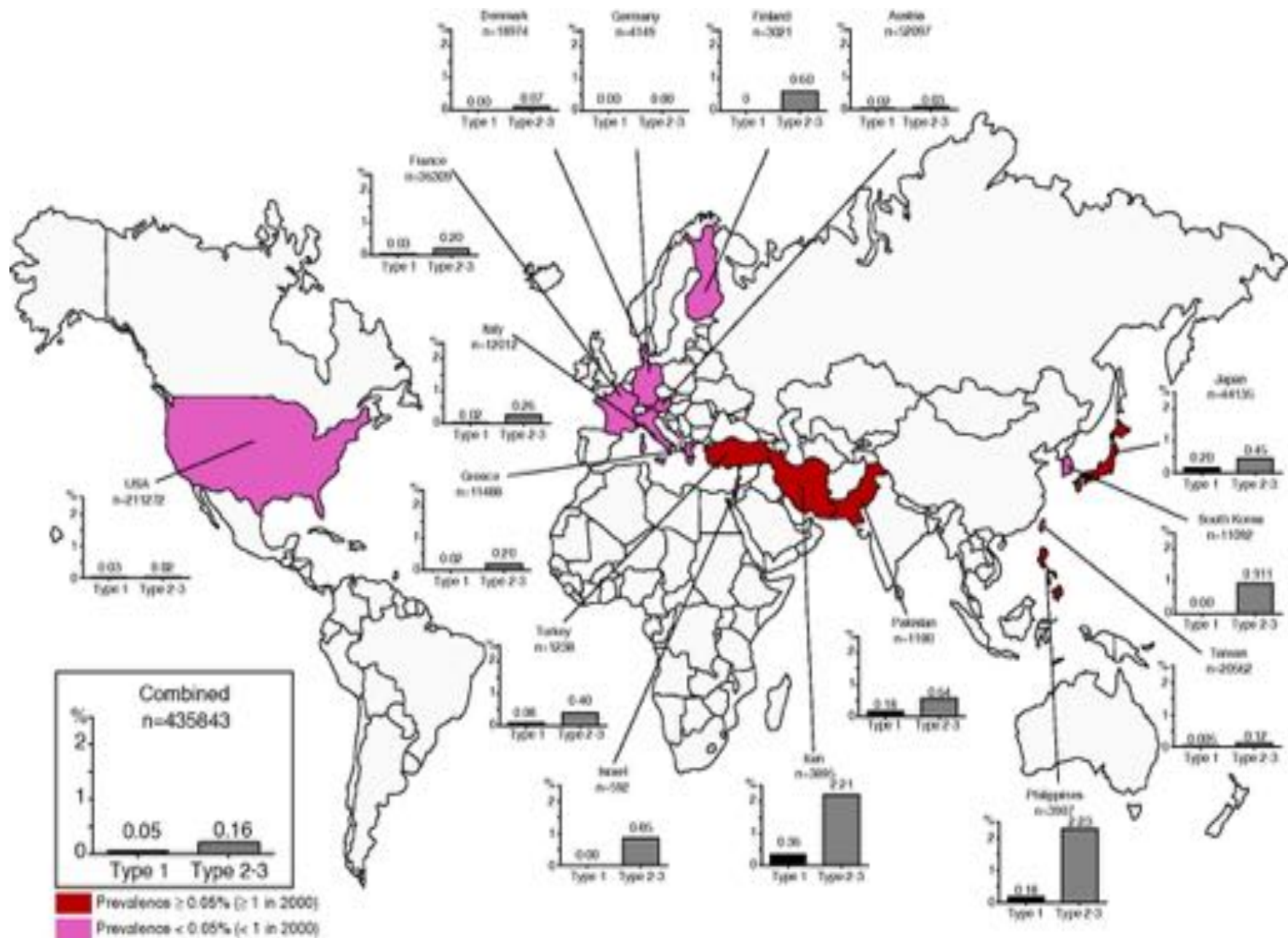
**Type 2 & 3**



# Diagnosis

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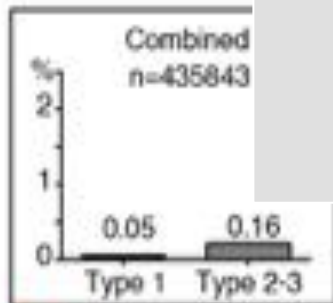
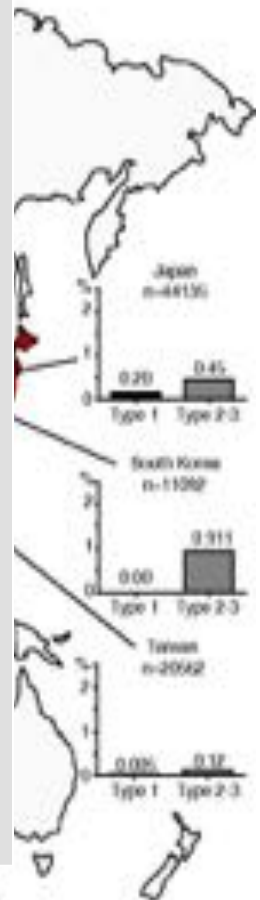
- ST-segment elevation with type I morphology  $\geq 2$  mm in  $\geq 1$  lead among the right precordial leads V1, V2 positioned in the 2nd, 3rd, or 4th ICS occurring either spontaneously or after provocative drug test with intravenous administration of Class I antiarrhythmic drugs.



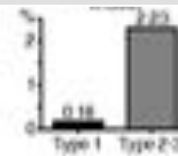


- **No precise data** are available on the epidemiology of BrS.
- Prevalence is much higher in Asian and Southeast Asian countries, especially Thailand, Philippines and Japan, reaching **0.5–1 per 1000**
- 8–10 times **more prevalent in males** (more prominent transient outward current (Ito) in males, higher testosterone levels also may have a significant role in the male predominance).

Denmark n=1814 Germany n=4145 Finland n=321 Austria n=2007



Prevalence  $\geq 0.05\%$  ( $\geq 1$  in 2000)  
 Prevalence  $< 0.05\%$  ( $< 1$  in 2000)



# Outcome after implantable cardioverter-defibrillator in patients with Brugada syndrome: the Gulf Brugada syndrome registry

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**Background and Objective:** Among patients with Brugada syndrome (BS) and aborted cardiac arrest, syncope, or inducible ventricular fibrillation at electrophysiologic study (EPS), the only currently recommended therapy is an implantable cardioverter-defibrillator (ICD), but these are not without complications. We assessed the total number of shocks (appropriate and inappropriate) and complications related to ICD in patients with BS.

**Methods and Results:** Twenty-five patients implanted with ICD for BS in 6 Gulf centers between January 1, 2002, and December 31, 2010, were reviewed. Implantable cardioverter-defibrillator indication was based on aborted cardiac arrest (24%), syncope (56%), or in asymptomatic patients with positive EPS (20%). During a follow-up of  $41.2 \pm 17.6$  months, 3 patients (all with prior cardiac arrest) had appropriate device therapy. Four patients developed complications; 3 of them had inappropriate shocks.

**Conclusion:** In our cohort, appropriate device therapy was limited to cardiac arrest survivors, whereas none of those with syncope and/or positive EPS had arrhythmias. Overall complication rate was relatively high, including inappropriate ICD shocks.



# Gulf Brugada Registry

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## Objective

- To describe the presentation and prognosis of patients with BrS in the Arab Gulf countries.

## Methods

- Consecutive patients with BrS were enrolled from 5 centers in the Arab Gulf region (2012 to present).
- Patients were included if they had a diagnostic type 1 ECG spontaneously or provoked with a Class I antiarrhythmic drug, and after exclusion of diseases that may mimic BrS.



# Gulf Brugada Registry

	All Patients (N=131)	Cardiac Arrest (N=16)	Syncope (N=36)	Asymptomatic/minimally symptomatic (N=79)
<b>Age</b>	33 ± 13	32 ± 9	34 ± 11	33 ± 14
<b>Gender (%Male)</b>	88	94	97	86
<b>LVEF</b>	58 ± 5	56 ± 8	58 ± 3	59 ± 3
<b>Type 1 ECG on Presentation (%)</b>	50	50	61	46
<b>Presenting ECG (%)</b>				
<b>Normal</b>	7	31	3	4
<b>Type 1</b>	50	50	61	46
<b>Type 2</b>	39	19	33	45
<b>Type 3</b>	4	0	3	5
<b>History of Atrial Fibrillation (%)</b>	5	19	0	4

# Gulf Brugada Registry

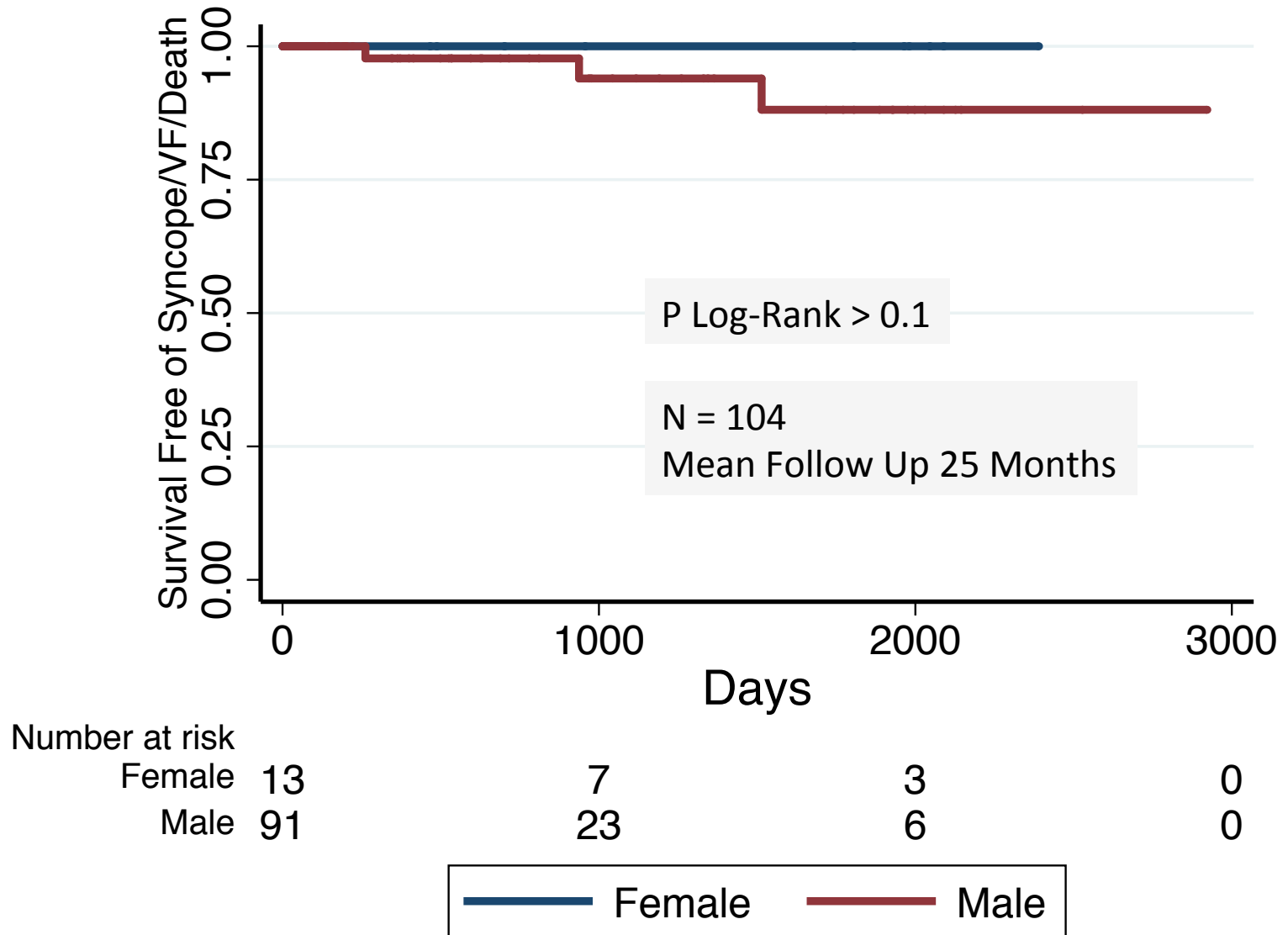
	All Patients (N=131)	Cardiac Arrest (N=16)	Syncope (N=36)	Asymptomatic/minimally symptomatic (N=79)
<b>Family History of SCD (%)</b>				
First Degree Relative	14	7	14	16
Any Relative	29	7	17	29
<b>Class I Drug Challenge</b>				
Performed	59/129 (46%)	5/16 (31%)	19/36 (53%)	35/77 (45%)
<b>Type 1 Provoked</b>	36/36 (100%)	5/5	19/19	35/35
<b>Drug Used (%)</b>				
<i>Ajmaline</i>	83	80	74	89
<i>Procainamide</i>	8	0	21	3
<i>Flecainide</i>	7	0	5	8
<i>Amiodarone</i>	2	20	0	0
<b>Exercise ECG</b>				
Performed	49/129 (38%)	2/16 (13%)	114/36 (39%)	33/77 (43%)
Type 1 Provoked in Recovery	11/49 (23%)	0/2 (0%)	2/14 (14%)	9/33 (27%)
<b>Genetic Testing</b>				
Performed	18/125 (14%)	1/16 (6%)	2/35 (6%)	15/74 (20%)
SCN5A Positive	14/18 (78%)	0/1	1/2	13/15
<b>EPS</b>				
Performed	23/131 (18%)	1/16 (6%)	10/36 (28%)	12/79 (15%)
VF Induced	12/23 (54%)	1/1	8/10	3/12

# Gulf Brugada Registry

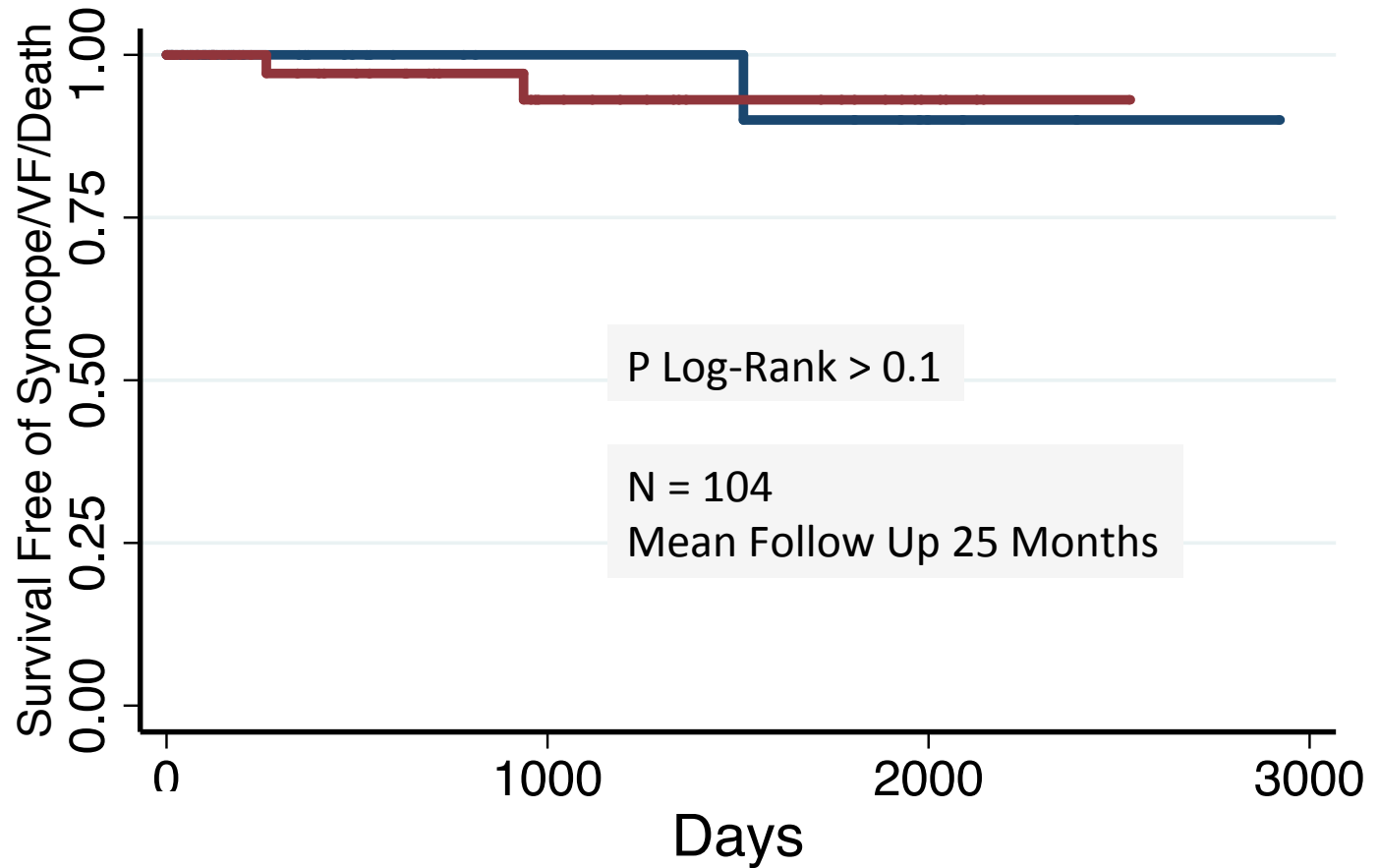
	All Patients (N=131)	Cardiac Arrest (N=16)	Syncope (N=36)	Asymptomatic/minimally symptomatic (N=79)
<b>Implantable Cardioverted Defibrillator</b>				
<b>Implanted</b>	46/131 (35%)	13/16 (81%)	27/36 (75%)	6/79 (8%)
<b>Type (% Single Chamber)</b>	98			
<b>In-Hospital Complication (%)</b>	4			
<b>Venous Access (%)</b>				
<i>Subclavian</i>	55			
<i>Cephalic</i>	41			
<i>Axillary</i>	4			
<b>Insurance (%)</b>				
<b>Government</b>	38	33	31	43
<b>Military</b>	20	7	11	27
<b>Private</b>	26	27	30	24
<b>Uninsured</b>	15	33	28	6
<b>Ethnicity (%)</b>				
<b>Gulf National</b>	45	13	37	54
<b>Indian Subcontinent</b>	36	56	43	29
<b>Other Arab</b>	11	13	17	8
<b>South East Asia</b>	5	13	3	4
<b>Other</b>	3	5	0	5



# Risk of Syncope/VF/Death by Gender



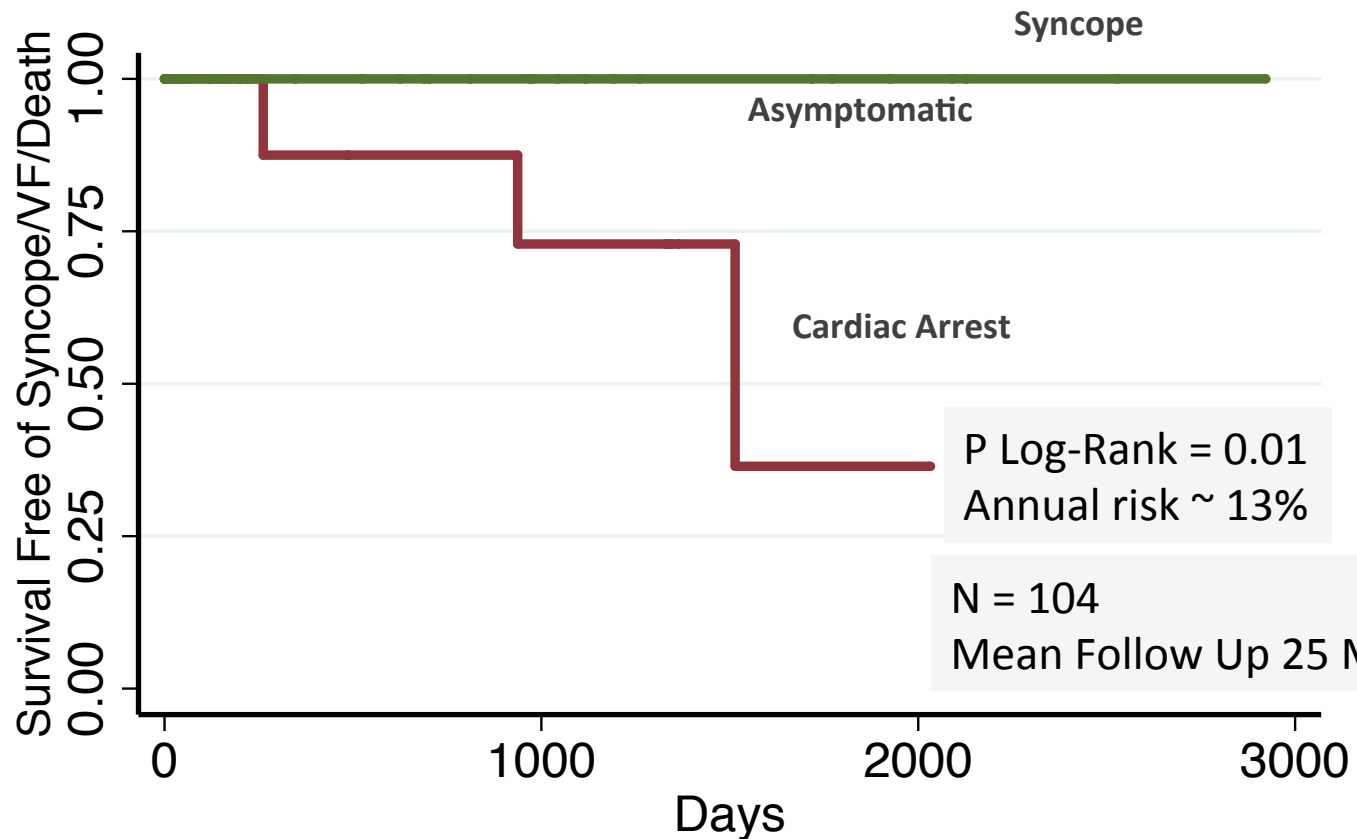
# Risk of Syncope/VF/Death by Type 1 ECG



	0	1000	2000	3000
Number at risk				
Drug Induced Type 1	51	10	4	0
Spontaneous Type 1	53	20	5	0

— Spontaneous Type 1     
 — Drug Induced Type 1

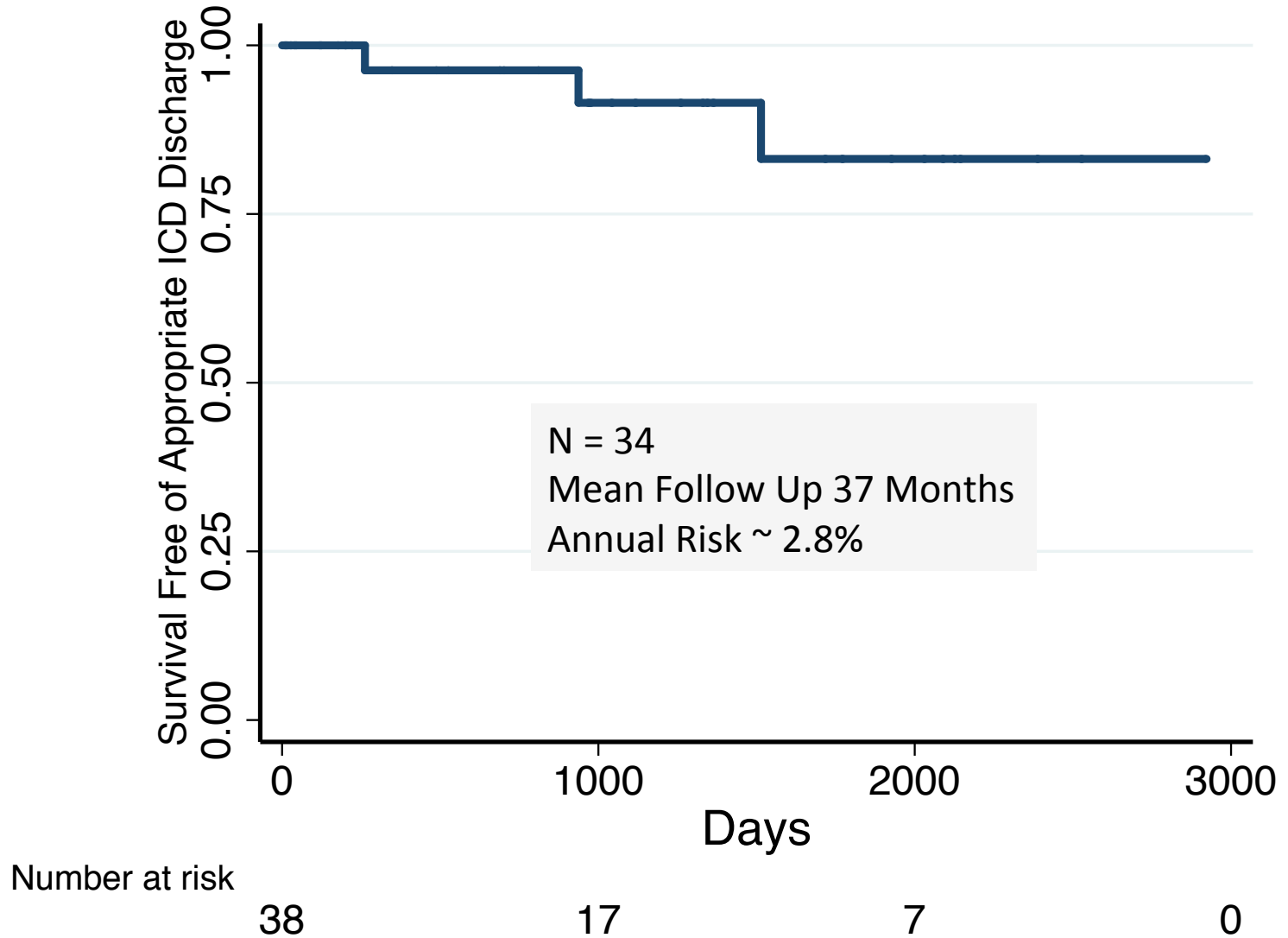
# Risk of Syncope/VF/Death by Symptoms



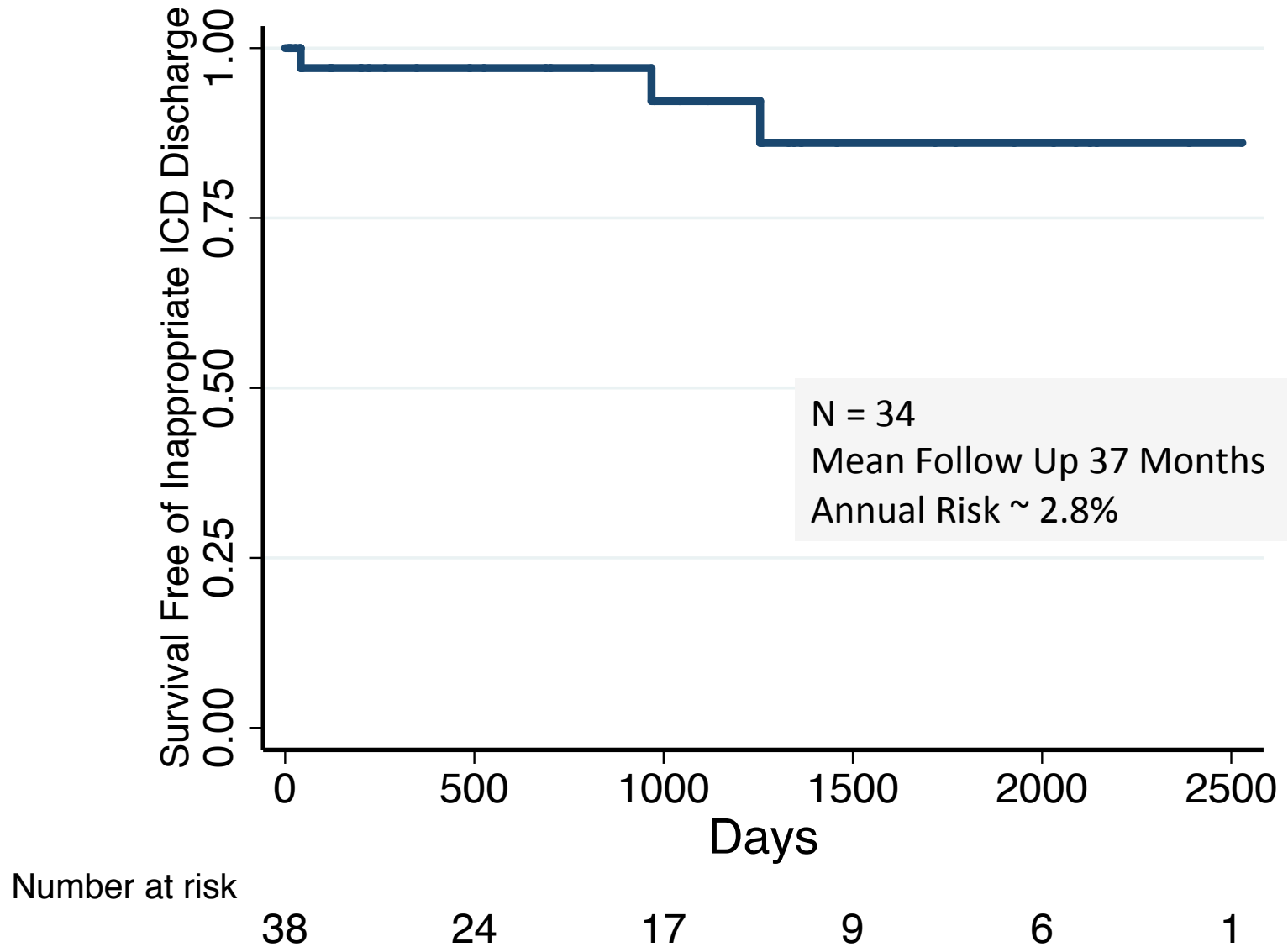
Number at risk

Asymptomatic	62	14	4	0
Cardiac Arrest	10	5	1	0
Syncope	32	11	4	0

# Risk of Appropriate ICD Discharge

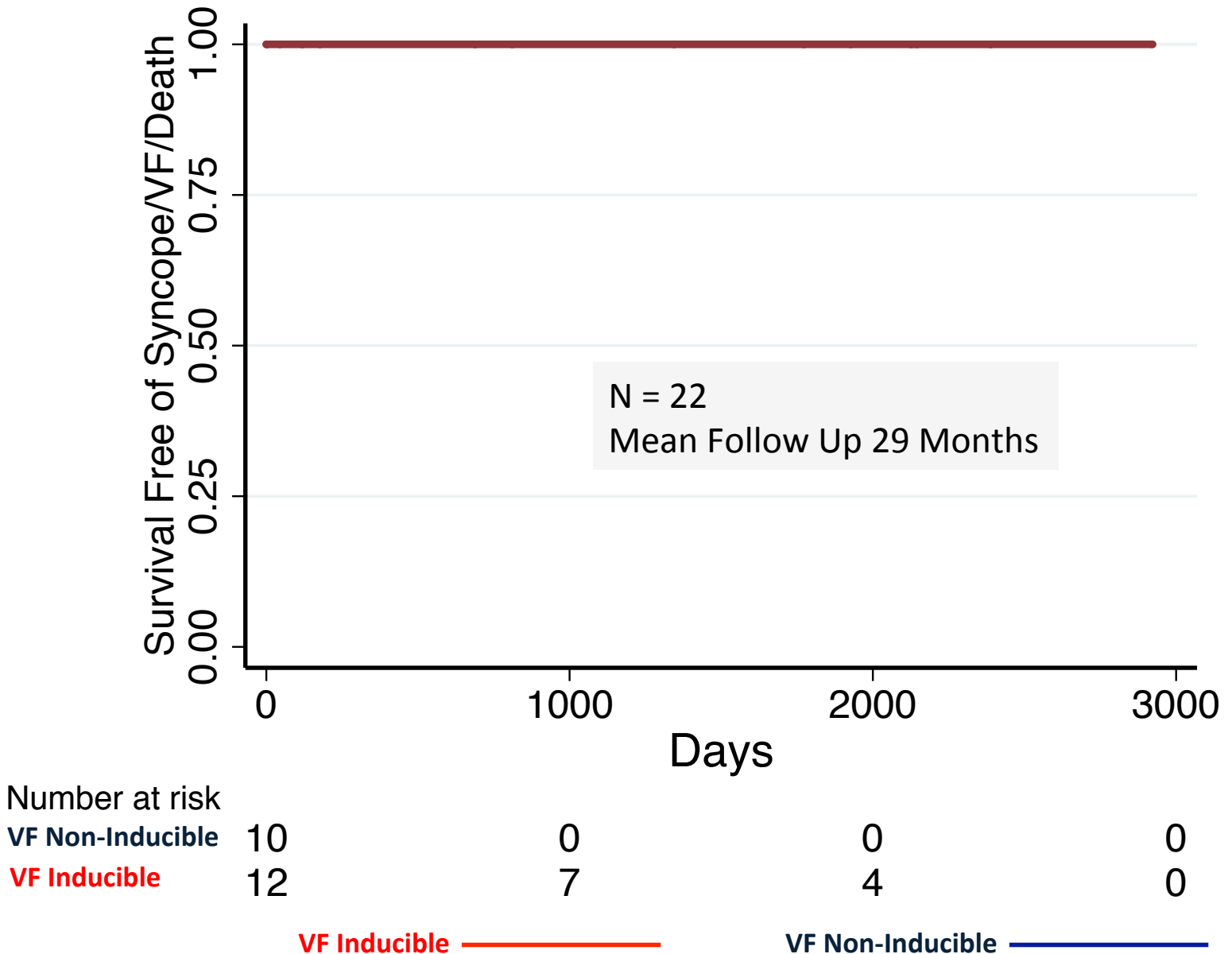


# Risk of Inappropriate ICD Discharge





# Risk of Syncope/VF/Death by EPS Results



# Gulf Brugada Registry

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- The present study is the largest BrS report from the Arab countries.
- The frequency by which BrS is diagnosed in clinical practice in the Gulf suggests a higher prevalence compared to other regions.
- Life-threatening cardiac events during follow-up were limited to patients presenting with cardiac arrest.
- Longer follow up and a larger sample size are needed to validate these findings.