Is Conservative Treatment Justified in Marfan Syndrome Patients with Non-Complicated Acute Type B Aortic Dissection?

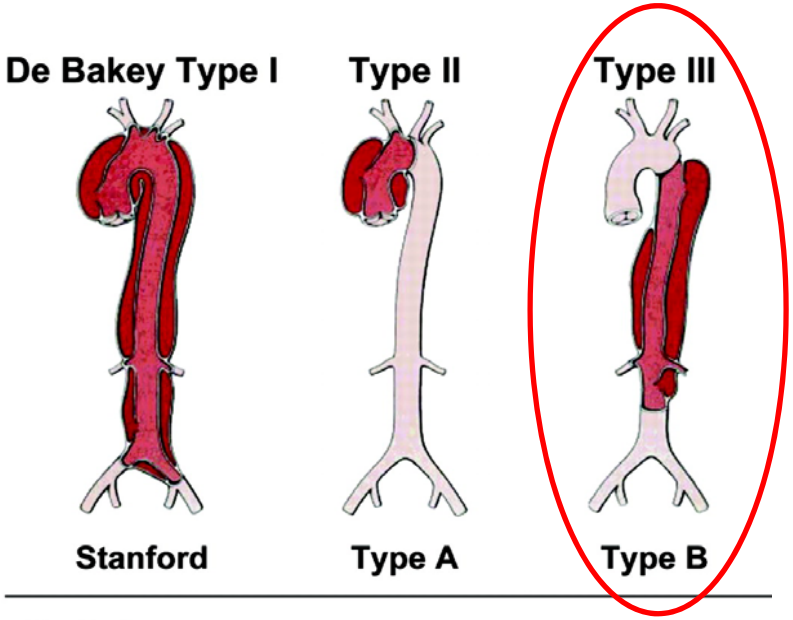
Insights from the International Registry of Acute Aortic Dissection

Gilon D, Pyeritz RE, Evangelista A, O'Gara P, Harris KM, Braverman AC, Peterson MD, Voehringer M, Suzuki T, Conklin LD, Montgomery DG, Isselbacher EM, Nienaber CA, Eagle KA, Korach A

Conflict of Interests

- None

Background



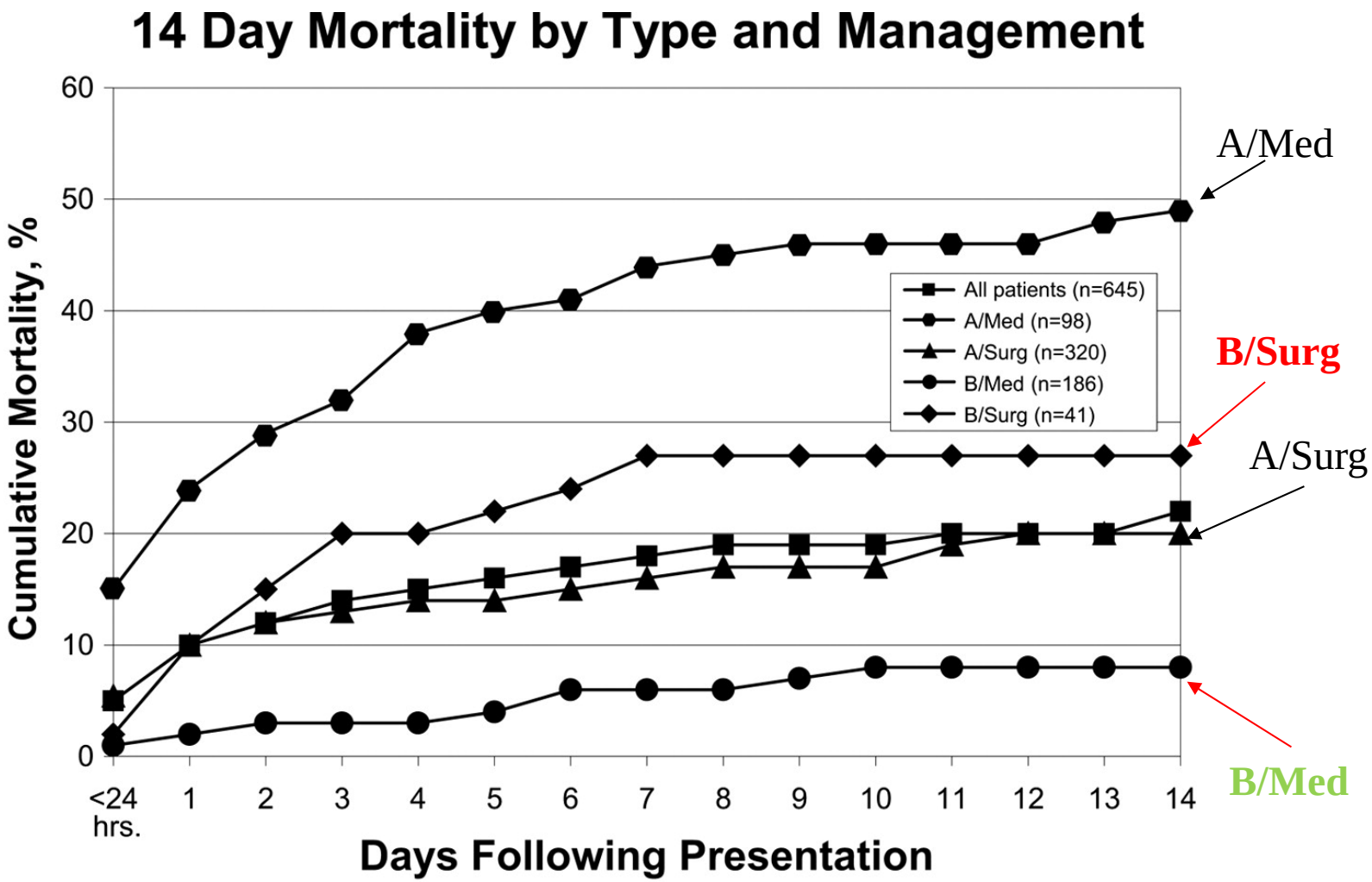
De Bakey

Type I	Originates in the ascending aorta, propagates at least to the aortic arch and often beyond it distally.
Type II	Originates in and as confined to the ascending aorta.
Type III	Originates in the descending aorta and extends distally down the aorta or, rarely retrograde into the aortic arch and ascending aorta.

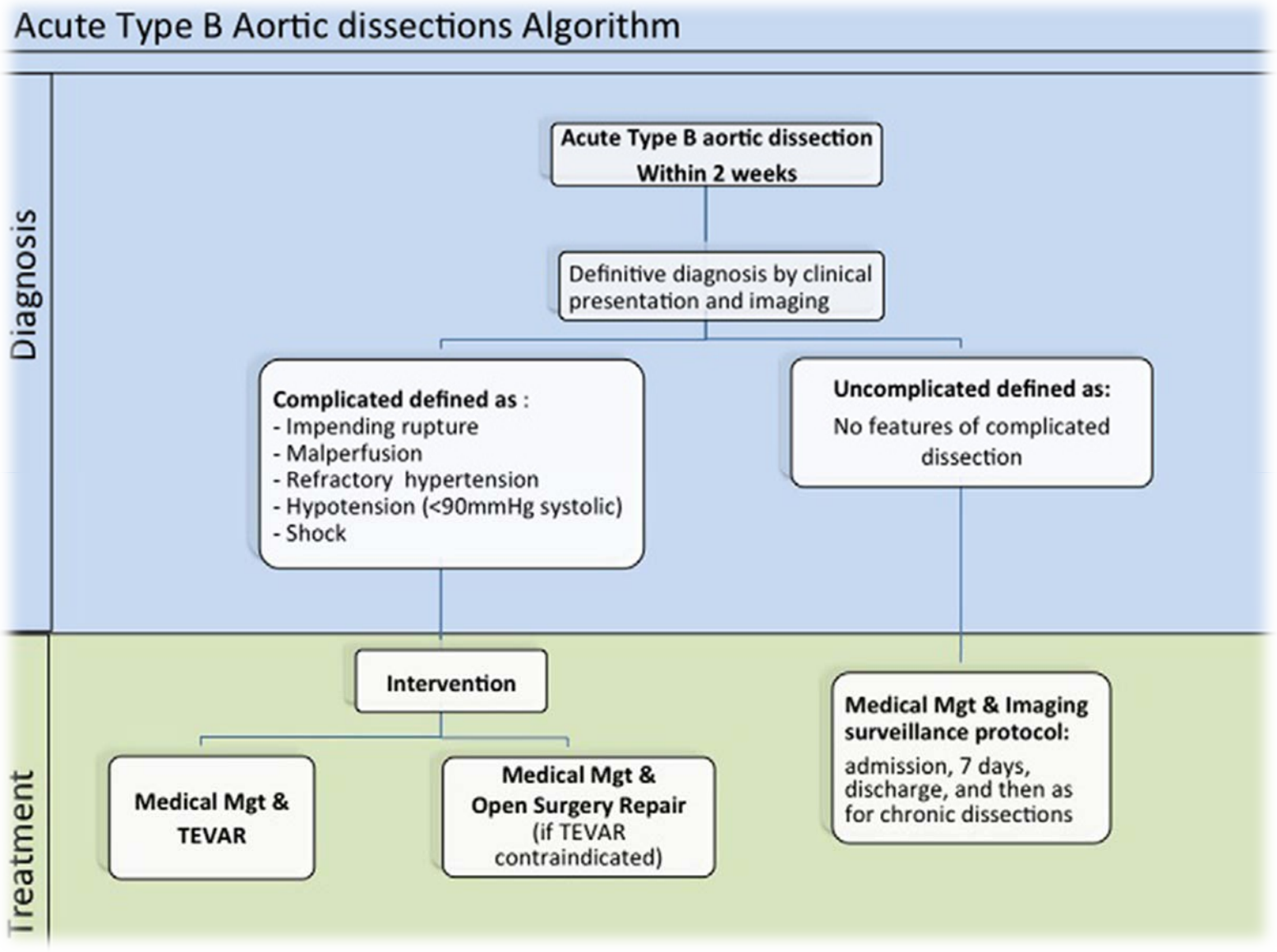
Stanford

Type A	All dissections involving the ascending aorta, regardless of the site of origin.
Type B	All dissections not involving the ascending aorta.

International Registry of Acute Aortic Dissection
IRAD



IRAD Investigators, 2005



Fattori R. et al. JACC 2013

Marfan & AAD

- Aortopathy
- Type B: 20-25%
- Limited information
- Single institution
- Marfan patients ~ 5%

Aim

- To analyze the in-hospital and long-term implications of conservative management in non-complicated Type B acute aortic dissection patients with Marfan Syndrome

IRAD Sites

St. Michael's Hospital, Toronto

Brigham & Women's Hospital

Massachusetts General Hospital

Dartmouth-Hitchcock Medical Center

Tromsø University

University of Rostock

Minneapolis Heart Institute

University of Massachusetts

Mayo Clinic

University of Calgary

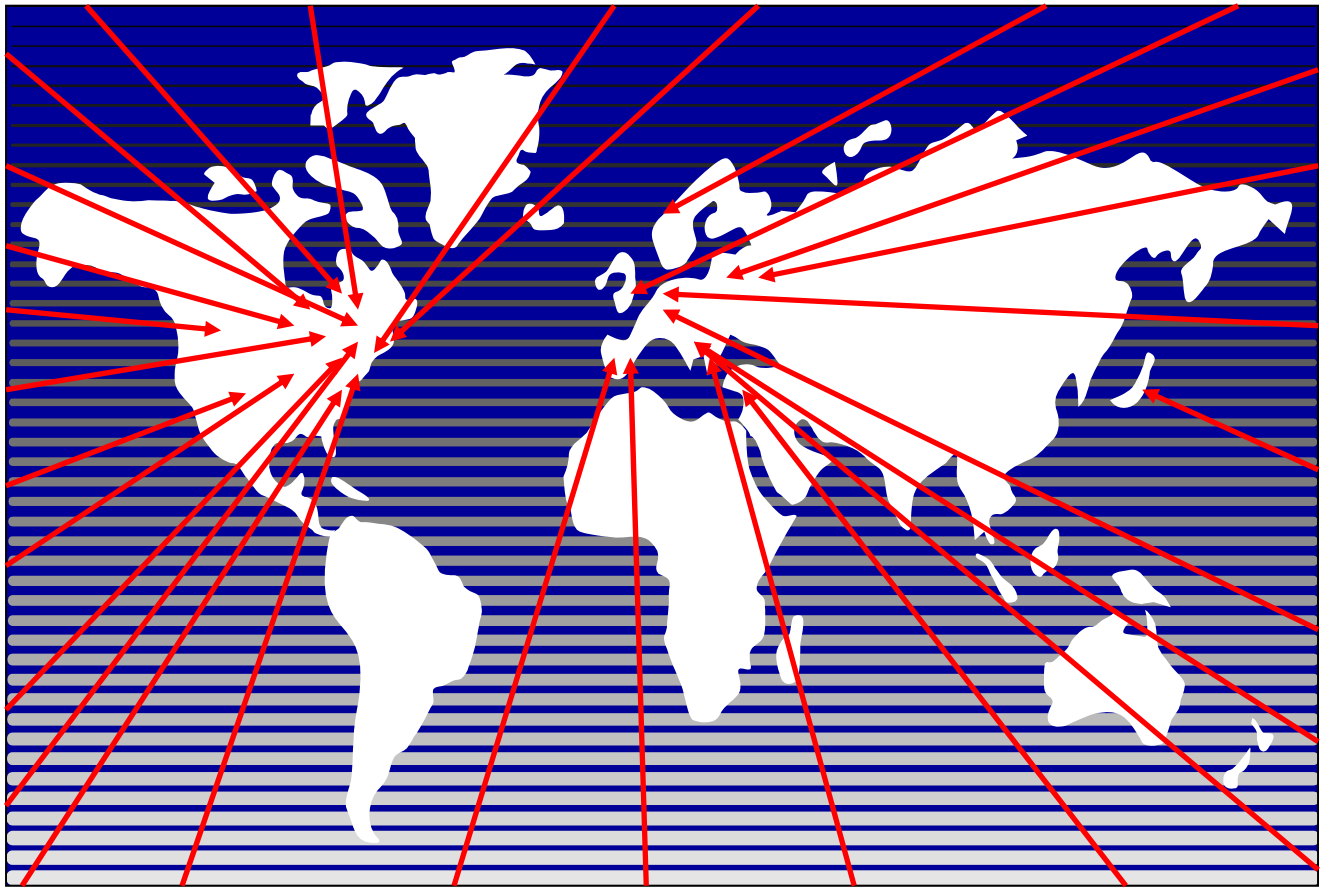
University of Michigan

University of Colorado

Washington University, St. Louis

University of Pennsylvania

University of Pittsburgh



Medical School Graz, Austria

University of Vienna

Robert Bosch Krankenhaus, Stuttgart

University of Tokyo

Hôpital Bichat, Paris

San Donato, Milan

Univ. Hospital S. Orsola, Bologna

Duke University

University of Virginia

University Hospital Vall d'Hebron, Barcelona

Hospital "12 de Octubre", Madrid

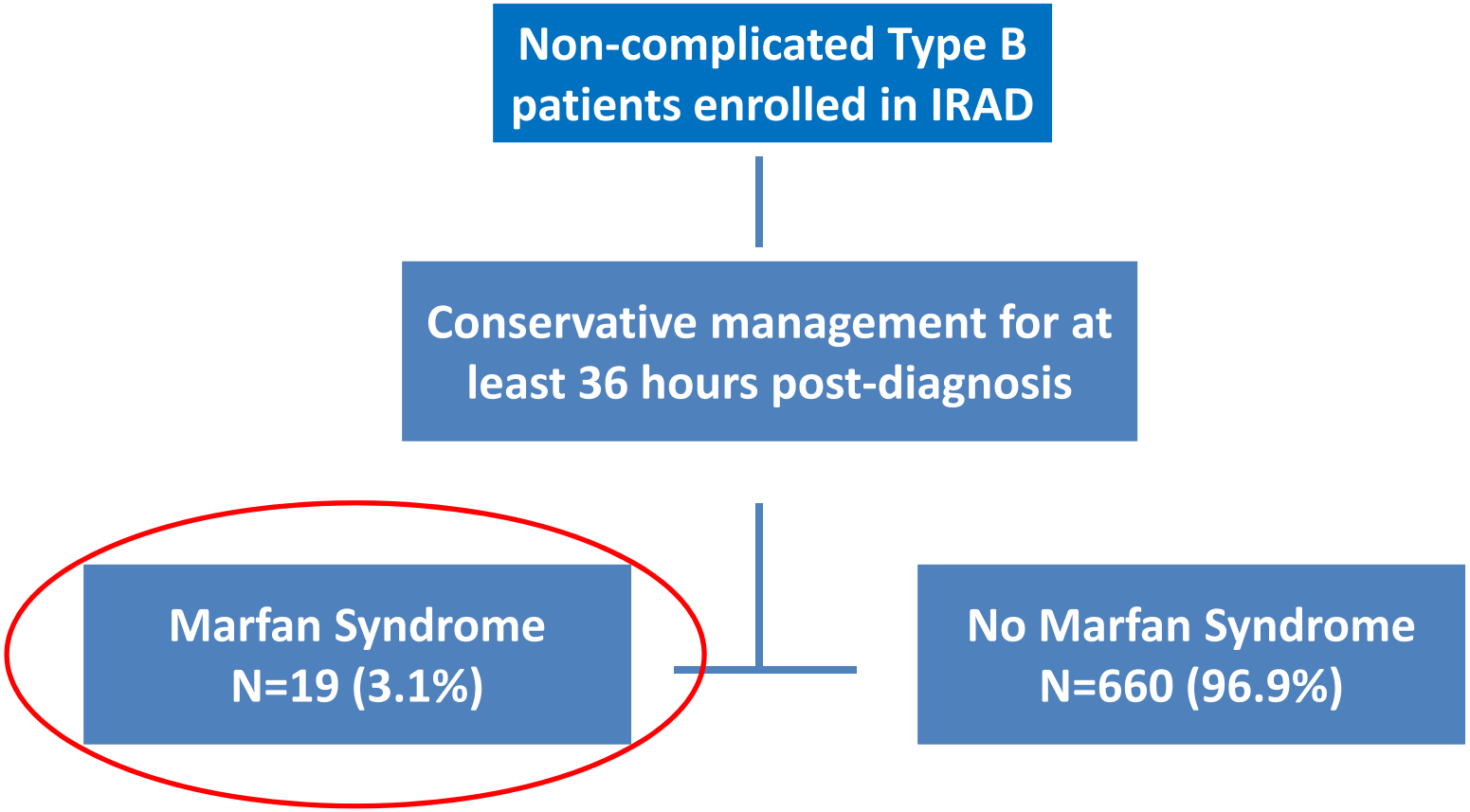
San Giovanni e Ruggi, Salerno

Hadassah Univ. Hospital, Jerusalem

Methods

- Marfan
- Stanford Type B AAD
- Complicated
  - ✓Persistent pain / Uncontrolled hypertension
  - ✓End organ malperfusion / progression
  - ✓Bleeding / impending rupture

Results


Non-complicated Type B patients enrolled in IRAD
Conservative management for at least 36 hours post-diagnosis
Marfan Syndrome N=19 (3.1%)
No Marfan Syndrome N=660 (96.9%)

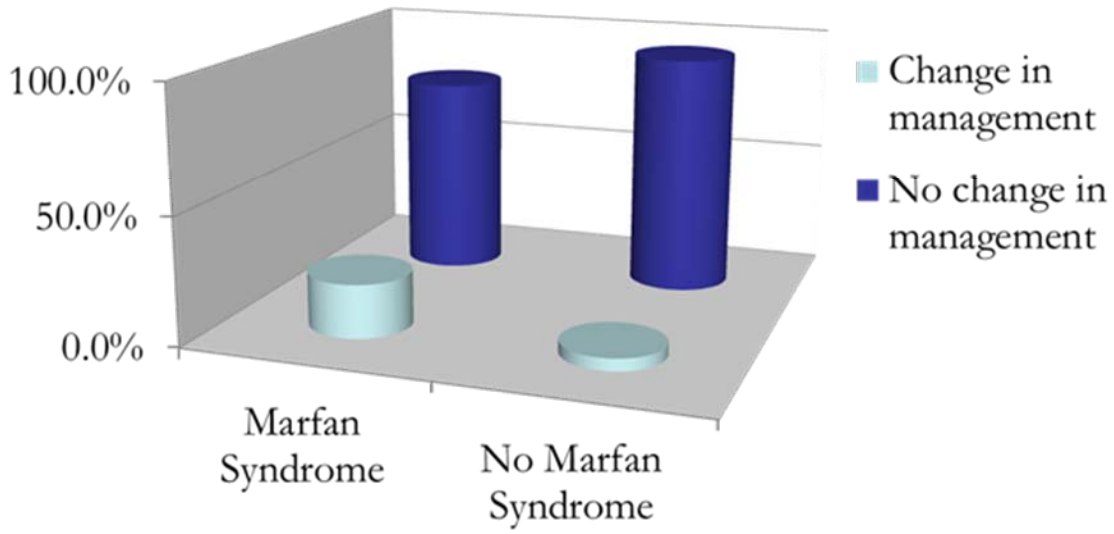
Demographics

	Marfan Syndrome	No Marfan Syndrome	p-value
Age (mean±SD)	40.9 ± 9.4	64.8 ± 13.5	<0.001
Gender- male	8 (42.1%)	370 (61.7%)	0.085
Race – white	16 (88.9%)	476 (82.8%)	0.751
History hypertension	4 (22.2%)	479 (80.0%)	<0.001
History diabetes	0 (0.0%)	40 (6.8%)	0.623
History atherosclerosis	2 (11.8%)	186 (31.3%)	0.085
Known aortic aneurysm	11 (61.1%)	111 (18.7%)	<0.001
Prior aortic dissection	11 (64.7%)	38 (6.4%)	<0.001
History aortic valve disease	8 (42.1%)	35 (5.9%)	<0.001
Family history aortic disease	7 (70.0%)	21 (10.8%)	<0.001

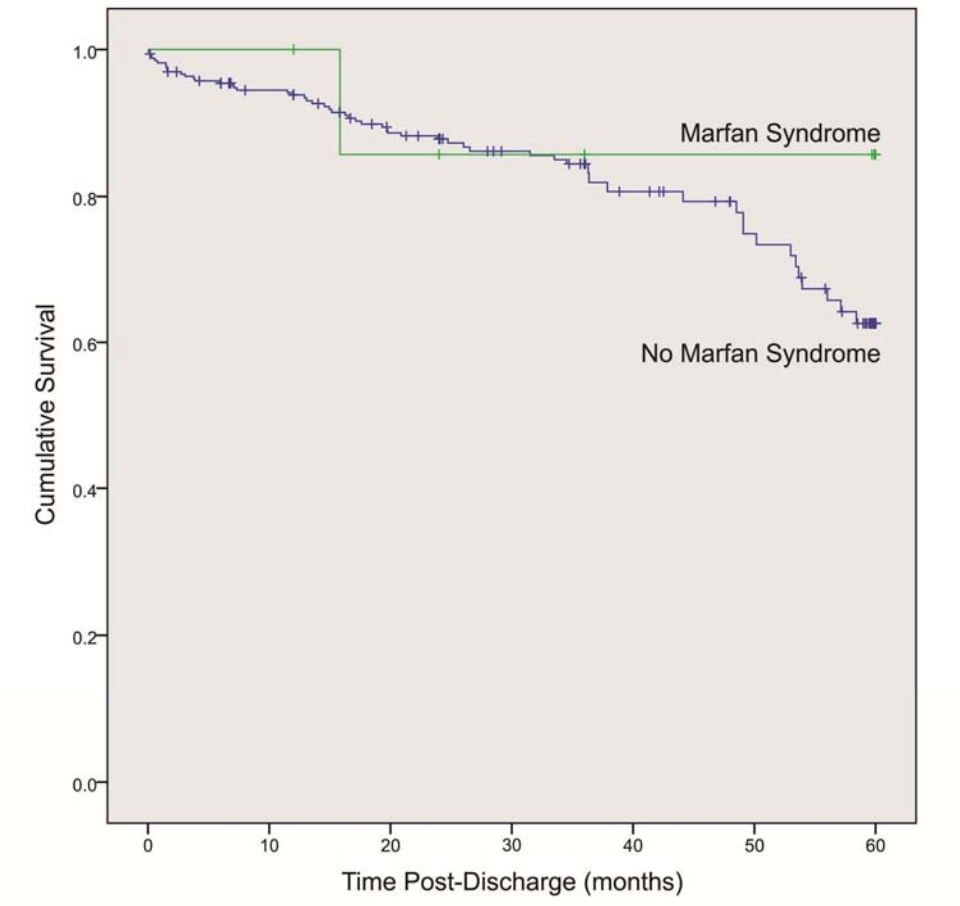
In Hospital Outcome

	Marfan Syndrome	No Marfan Syndrome	p-value
CVA	0 (0.0%)	7 (1.3%)	1.000
Coma	0 (0.0%)	7 (1.3%)	1.000
Spinal Cord Ischemia	0 (0.0%)	4 (0.8%)	1.000
Myocardial Ischemia	0 (0.0%)	14 (2.5%)	1.000
Myocardial Infarction	0 (0.0%)	10 (1.8%)	1.000
Mesenteric Ischemia/Infarction	0 (0.0%)	3 (0.5%)	1.000
Acute Renal Failure	1 (5.3%)	4 (0.7%)	0.156
Extension of Dissection	4 (21.1%)	26 (4.7%)	0.014
Hypotension	0 (0.0%)	21 (3.8%)	1.000
Cardiac Tamponade	1 (5.3%)	2 (0.4%)	0.097
Limb Ischemia	0 (0.0%)	1 (0.2%)	1.000
Mortality	0 (0.0%)	25 (4.2%)	1.000

Change in Management



Long Term Survival



Conclusions

- Our study shows conservative management of non-complicated Type B aortic dissection is safe in Marfan Syndrome patients
- Heighted clinical awareness of the increased possibility of extended dissection is necessary in patients with Marfan Syndrome

Limitations

- Retrospective
- Small sample
- Variable treatment