



**Weill Cornell Medicine**

# *Outcomes of Type B Dissection in Marfan Syndrome*

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# Disclosures

- None

# Objective (1)

- To highlight outcomes of type B dissection in Marfan syndrome

# Types A vs. B Dissection in MFS

**Type A Dissection (Cornell Aneurysm Registry, 2023): 336 MFS patients**

9%

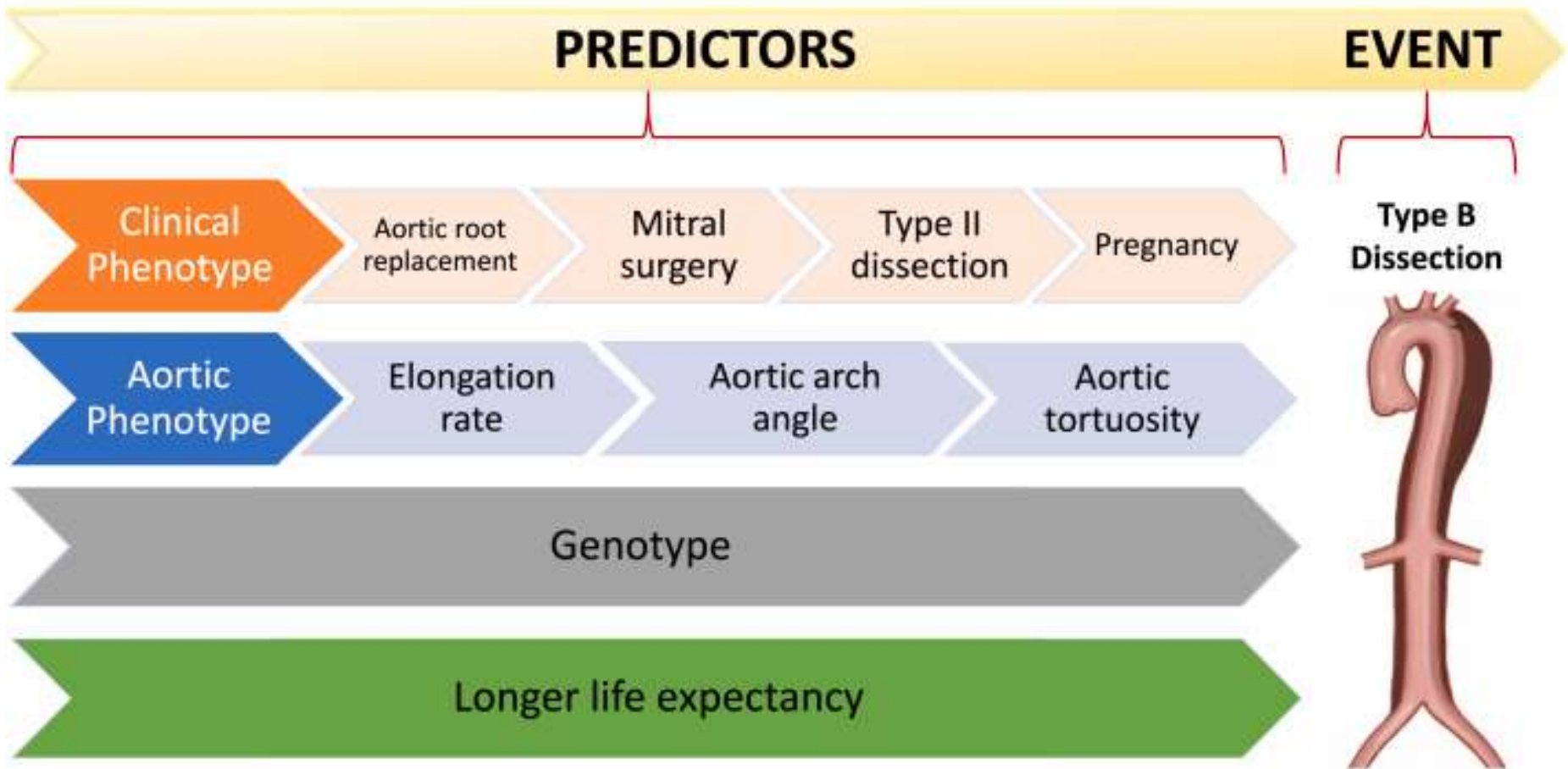
43% aware of MFS diagnosis: large size and lack of knowledge of diagnosis are biggest risk factors

# Types A vs. B Dissection in MFS

Type A Dissection (Cornell Aneurysm Registry, 2023): 336 MFS patients	Type B Dissection (Cornell Aneurysm Registry, 2023): 336 MFS patients
9%	14%
43% aware of MFS diagnosis: Large size and lack of knowledge of diagnosis are biggest risk factors	Risk is unpredictable: 87% aware of MFS diagnosis, 88% had normal or mildly dilated descending aorta

# Types A vs. B Dissection in MFS

Type A Dissection (Cornell Aneurysm Registry, 2023): 336 MFS patients	Type B Dissection (Cornell Aneurysm Registry, 2023): 336 MFS patients
9%	14%
43% aware of MFS diagnosis: Large size and lack of knowledge of diagnosis are biggest risk factors	Risk is unpredictable: 87% aware of MFS diagnosis, 88% had normal or mildly dilated descending aorta
	Patients with TBD significantly more likely to have ARR (55% after ARR), significantly more likely to have independent MV surgery



**What happens to individuals with MFS who experience a type B dissection?**

# Outcomes of Type B Dissection in MFS: Characteristics

## Cornell Aneurysm Registry (362 patients)

50 (13.8%) with type B dissection

DeBakey type 3A in 5,  
type 3B in 42,  
isolated abdominal  
dissection in 3

# Outcomes of Type B Dissection in MFS: Characteristics

## Cornell Aneurysm Registry (362 patients)

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# Outcomes of Type B Dissection in MFS: Characteristics

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50 (13.8%) with type B dissection DeBakey type 3A in 5, type 3B in 42, isolated abdominal dissection in 3	Median post- dissection follow-up: 8.9 years (IQR: 3.9-15.4)	175 (48.3%) had ARR 45/50 patients with type B dissection had ARR: 30 after ARR, 15 before, 5 in absence of ARR
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# Outcomes of Type B Dissection in MFS

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14 (28%) were managed medically  
2 (4%) suffered rupture with death

# Outcomes of Type B Dissection in MFS

Indications for type B surgery  
(34 patients)



Large absolute diameter in 13 (38.2%)  
Rapid growth in 16 (47.1%)



End-organ malperfusion in 4 (11.8%)  
Rupture in 1 (2.9%)

## Surgery for Type B Dissection in MFS

- All had open thoracoabdominal aortic repair as initial procedure
  - 19: surgery for extent I TAA
  - 7: surgery for extent II TAA
  - 2: surgery for extent IV TAA
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  - 2: unknown
- 30-day mortality 0%, paraplegia 0%, mild and reversible AKI in 2, stroke 0%

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(36 years [IQR: 31-43] vs. 46 [IQR: 35-57],  $p=0.025$ )

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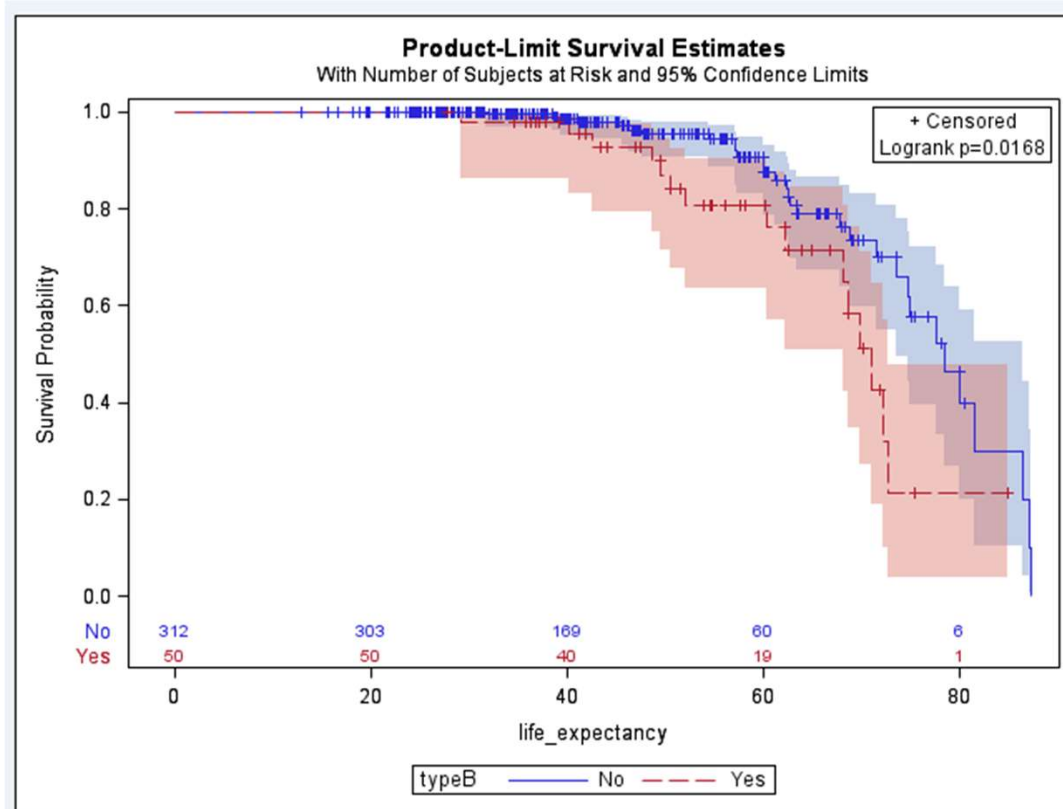
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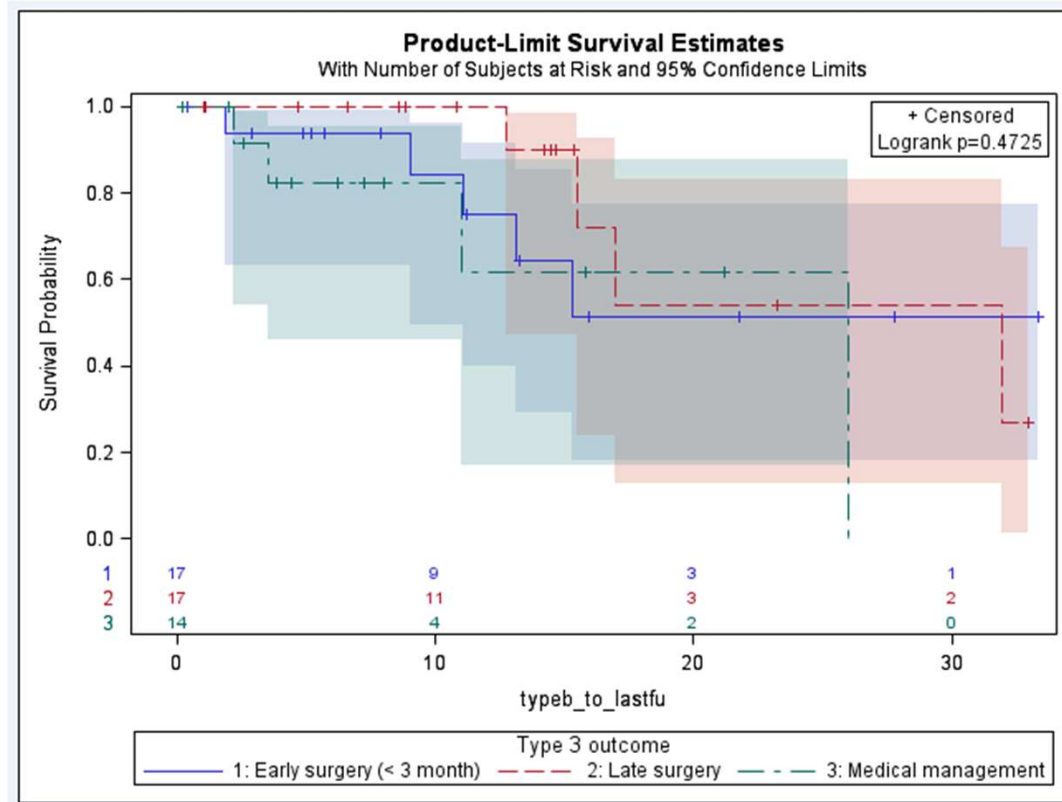
Indications for surgery different ( $p=0.03$ ): Driven by rapid growth (11/17 or 64.7%)  
in men and absolute aortic diameter (8/17 or 47.1%) in women



# Life Expectancy



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# Summary

1. In contemporary MFS cohorts, prevalence of type B dissection (13.8%) *exceeds* that of type A
2. Type B dissection necessitates aortic surgery in the majority
3. There are key sex specific differences in the indication for, and timing of, type B dissection surgery  
*Should this alter / influence surveillance guidelines?*
4. Type B dissection is associated with significantly reduced life expectancy but only in men due to earlier age at type B dissection
5. Outcomes following type B dissection do not differ by management strategy

# THANK YOU



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