Aortic repair in Marfan’s –

should we lower the threshold for treatment?

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Antoine Bernard-Jean Marfan
(1858 -1942)
In 1896, Marfan presented his 1st case at the Société Médicale des Hôpitaux de Paris

A 5 year old girl, Gabriel P., presented with ‘pattes d'araignée‘ (= spider legs); her mother had noticed the abnormalities already at birth. Marfan called the condition dolicostenomely (stenos = narrow; melos = limb).

Gabrielle P’s striking skeletal abnormalities progressed to the time of her death (probably from tuberculosis) in early adolescence.
Normal Aortic Diameter

Upper limit of the ascending and descending aorta related to age

Sinus of Valsalva diameter, by body surface area

American College of Cardiology Foundation, J Am Coll Cardiol 2010;55:e27- e129
Figure 2  Schematic presentation of a sternal view with four regions measured for follow-up after repair: LV = left ventricle; LA = left atrium; 1 = valve annulus; 2 = aortic sinuses; 3 = sinotubular junction; 4 = proximal ascending artery. (Reprinted from Am J Cardiol, Volume 64, Roman MJ, Devereux RB, Kramer, Fox R, O’Loughlin J. Two-dimensional echocardiographic aortic dimensions in children and adults, pp. 507–512 with permission from Excerpta Medica Inc.)
<table>
<thead>
<tr>
<th>Table 1 Normal aortic dimensions in adults</th>
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<tr>
<td>Diameter</td>
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<td>Aortic annulus</td>
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<td>Male</td>
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<td>Female</td>
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<tr>
<td>Sinus of Valsalva</td>
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<td>Male</td>
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<td>Female</td>
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<td>Aortic root</td>
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<td>Proximal ascending aorta</td>
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<td>Male</td>
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<td>Ascending aorta</td>
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<td>Aortic wall</td>
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Figure 4  Schematic drawing of aortic dissection class 1, subdivided into DeBakey types I, II and III. Also depicted are Stanford classes A and B with subtypes a and b (subtype depends on the thoracic or abdominal involvement according to Reul and Cooley).
Figure 5  Differentiation of classes 1–5 of aortic dissection. Class 1: classic aortic dissection with true and false lumen without communication of the two lumina; class 2: intramural haemorrhage or haematoma; class 3: ulceration of aortic plaque following plaque rupture; class 4: subtle or discrete aortic dissection with bulging of the aortic wall; class 5: iatrogenic or traumatic aortic dissection, illustrated by a catheter induced separation of the intima. (From Svensson LG, Labib SB, Eisenhauser AC, Butterfly JR. Intimal tear without haematoma. Circulation 99: 1331–6, 1999; American Heart Association; reproduced with permission.)
Class IIa

If the maximal cross-sectional area in square centimeters of the ascending aorta or root divided by the patient's height in meters exceeds a ratio of 10, surgical repair is reasonable because shorter patients have dissection at a smaller size and 15% of patients with Marfan syndrome have dissection at a size less than 5.0 cm.

*(Level of Evidence: C)*


Svensson LG, Khitin L. Aortic cross-sectional area/height ratio timing of aortic surgery in asymptomatic patients with Marfan syndrome J Thorac Cardiovasc Surg 2002;123:360-361
Effect of Aortic Diameter on Risk of Complication

Aortic Cx in Marfan’s

- 90% of M&M in Marfan’s due to aortic pathology
- aortic dissection is #1 cause of mortality
- risk of dissection correlates with diameter, but occurs at small diameters as well
- hospital mortality:
  - elective repair – 1 - 2%
  - emergencies – 10 - 20%


Young Marfan’s patient with *isolated ascending* enlargement

*female, 13 years*

13 year old girl (185 m tall) and her 43 year old father – both have Marfan’s

Presenting with *isolated ascending aortic enlargement*

Diameter (CT-angio): 4.2 cm
How far should we go?

In the young Marfan’s patient with *ascending enlargement* aim for —

- prophylactic repair to prevent acute type A dissection and downstream Cx
- maintained AV function
AV reimplantation with ‘neosinus’ (David V - procedure)
Valve sparing root replacement

native root

Valve motions

remodeling (Yacoub)

reimplantation (David)
Results of Valve-Sparing Aortic Root Reconstruction in 158 Consecutive Patients

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Background. This study assesses the durability and clinical outcome of valve-sparing aortic root reconstruction using the reimplantation technique in a single center cohort.

Methods. From July 1993 to July 2001, 158 patients underwent replacement of the ascending aorta with native valve reimplantation. Mean age of patients was 52 ± 17 years (9 to 84 years), 103 were men (65%). Thirty-four patients (22%) suffered from Marfan’s syndrome. Aortic dissection Stanford type A was present in 29 patients (19%) (22 acute, 7 chronic), and concomitant partial or total arch replacement was necessary in 57 patients (36%). One or more additional procedures were performed in 28 patients (18%). Mean follow-up was 36 ± 25 months (0.4 to 96 months).

Results. Thirty-day mortality was 3.8% (6 patients), but only 2.2% in elective patients. Mean bypass time was 169 ± 50 minutes (99 to 440 minutes), aortic cross-clamp time was 129 ± 31 minutes (79 to 205 minutes). In patients undergoing arch replacement, circulatory arrest was 26 ± 18 minutes (7 to 99 minutes). During follow-up, there were 5 (3.3%) cardiac-related late deaths. Grade of aortic insufficiency (AI) decreased from 2.3 ± 1.1 (0 to 4) preoperatively to 0.23 ± 0.44 (0 to 2) postoperatively (p < 0.0001). Six patients required aortic valve replacement, 4 of those due to progressive AI. Average grade of AI increased significantly to 0.42 ± 0.61 (0 to 3) at latest evaluation (p = 0.002). Two patients experienced a transient ischemic attack within the first postoperative week. No further thromboembolic complications were noticed. All patients presented with a favorable exercise tolerance.

Conclusions. The aortic valve reimplantation technique achieves excellent clinical outcome with few complications even in complex pathologies. Lack of anticoagulation and favorable durability encourage wider and earlier use of this technique.

David – Long-term results

Fig 2. Freedom from reoperation due to valve morbidity.

Fig 3. Freedom from moderate or severe aortic valve insufficiency. Reoperation was required for the first four events.

Ann Thorac Surg 2002;74:2026-33
David procedure / Leipzig

- 2006 – 2009: n = 93 (David I / V)
- perioperative mortality: 2.2%
  - type A dissection: 2/18 = 11%
  - Non-diss: 0/75 = 0%
- Aortic regurgitation (@ discharge)
  - none-to-mild AR – 98%
  - mild-to-moderate AR – 2%
Long-term results of aortic valve–sparing operations in patients with Marfan syndrome

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Objective: The appropriateness of aortic valve–sparing operations in patients with Marfan syndrome has been questioned. This study examines the long-term results of these operations in patients with Marfan syndrome.

Methods: From 1988 to 2006, 103 consecutive patients with Marfan syndrome (mean age, 37 ± 12 years) and aortic root aneurysm had aortic valve–sparing operations. Emergency surgery was performed in 11 patients: 8 for acute type A aortic dissection and 3 for unexplained persistent chest pain. Fourteen patients also had mitral valve surgery. The technique of aortic valve reimplantation was used in 77 patients, and aortic root remodeling was used in 26 patients. Patients were followed prospectively and underwent annual echocardiographic studies. The mean follow-up was 7.3 ± 4.2 years and 100% complete.

Results: There was 1 operative death and 5 late deaths. Four of the 6 deaths were due to complications of aortic dissections. The patients’ survival at 15 years was 87.2% compared with 95.6% for the general population of Ontario matched for age and sex. Seven patients had important aortic insufficiency: 4 mild to moderate, 2 moderate, and 1 moderate to severe. Freedom from greater than mild aortic insufficiency at 15 years was 79.2%. Three patients, all after aortic root remodeling, had aortic valve replacement, 2 for aortic insufficiency and 1 for endocarditis. At the most recent follow-up, 97 patients were alive: 86 were in functional class I, and 11 were in functional class II.

Conclusions: Aortic valve–sparing operations provided excellent clinical outcomes in this series of patients with Marfan syndrome. Postoperatively, complications of aortic dissections were the leading cause of death.
Long-term follow up

Aortic valve sparing operations

Ontario population matched for age and sex

5 yr.  95.7 ± 2%
10 yr.  93.4 ± 3%
15 yr.  87.2 ± 6.6%

98.5%
97.0%
95.6%

5 yr. 100%
10 yr. 94.9 ± 3.5%
15 yr. 87.6 ± 7.7%

Patients at risk

0  5  10  15
103  71  30  11

0  5  10  15
102  70  29  9

Years Postoperatively

Survival (%)

Freedom from reoperation (%)

JTCVS 2009;138:859-64
S/P life saving scAA repair for acute type A – now *presenting with arch / TAAA*

S/P supracoronary ascending replacement for acute type A aortic dissection

Now presenting with *downstream* aortic disease and *progressive aneurysm of the transverse arch* and the *TAAA*
After prophylactic repair of the ascending aorta, the arch and descending aorta are sites for later-onset aneurysms and dissections in patients with Marfan syndrome, prompting the need for routine imaging of the arch and descending aorta. Survival in patients with Marfan syndrome has been significantly improved with medical and surgical management of the aortic disease (76,92,93). The David valve sparing reimplantation operation for suitable patients undergoing elective aortic root surgery at centers with a high volume of these cases has become standard practice (76,92–99), although some have reported less-optimal long-term results with valve-sparing procedures (100,101).
Distal Aortic Reinterventions After Root Surgery in Marfan Patients

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Background. Distal aortic disease may evolve in Marfan patients after aortic root surgery. The aim of this study was to analyze the results of distal aortic reoperations in Marfan patients after proximal aortic surgery.

Methods. A total of 95 Marfan patients (56 male; mean age 34.5 ± 10.9 years) have been followed or operated on at our institution between October 1994 and December 2007. Results of patients who required distal aortic reinterventions after root surgery were collected and analyzed.

Results. Fifteen Marfan patients (9 male; mean age 39.1 ± 7.5 years) underwent distal aortic reinterventions after aortic root surgery. The indications for distal reoperations consisted of dissecting aneurysm after type A dissection surgery in 12 patients (80%) and nondissecting aneurysm in 3 patients (20%). Type A dissection during initial aortic root surgery was the only independent predictor of distal aortic reoperation (hazard ratio 3.8). One patient (6.7%) died perioperatively. Neurologic morbidity consisted of 1 patient with paraplegia and 1 with temporary paraparesis, and no strokes. Two patients died during a mean follow-up of 36.2 ± 25.5 months. Survival was 91.7% ± 1.6% at 1 and 5 years postoperatively. Three patients undergoing reoperative arch procedures required additional interventions on the distal aorta during follow-up.

Conclusions. Distal aortic reoperations in Marfan patients are more common among those presenting with type A dissection. Although technically challenging, such procedures can be performed with good short- and long-term results. Complete aortic arch replacement with an elephant trunk technique, if it can be safely performed, should be considered for Marfan patients presenting with type A dissection.

Fig 1. Freedom from distal reoperations after aortic root surgery.
INVITED COMMENTARY

Most of the improvement in the life expectancy of Marfan syndrome (MFS) patients over the last 30 years is attributable to improved recognition and prophylactic replacement of ascending aorta and root aneurysm, an operation with low operative risk in the current era. However, as Girdauskas and colleagues [1] demonstrate, root replacement still leaves the patient with a diseased arch and descending thoracoabdominal aorta, particularly if dissection has already occurred before root replacement. Twenty-eight percent (16 of 58) of their root replacement patients required reoperation within 4 years, and of course the number is likely to rise with longer follow-up. Dissection before root surgery was the strongest predictor of reoperation, and reoperation was usually for aneurysm in the descending aorta. Although early reoperative mortality was low (1 patient, 20%), mortality was 30% in this group at 3 years, and 3 more patients needed yet another reoperation.

The lessons are clear. First, operation before dissection remains the goal, because the “cat is out of the bag” once dissection occurs and a new, unfavorable “natural” history begins. The solution is not necessarily to operate at a smaller aortic diameter, but rather smarter, more selective use of the operation in patients identified to be at high risk, a strategy being investigated fervently. In contrast with Girdauskas’ series in which nearly half of root replacements were performed emergently for acute type A dissection, our experience at Johns Hopkins is heavily weighted toward elective root replacement in the nondissected aorta, and the reoperation rate at 10 years is significantly lower (15%). Second, after root replacement, all MFS patients, and particularly those with dissection, need meticulous surveillance and follow-up. Whether arch replacement is advisable at the time of root surgery remains controversial. Several studies have shown that concomitant arch replacement is probably not warranted in the elective, nondissected MFS patient, a position we share. However, benefit might be seen in acutely dissected patients, but they are the patients at greater risk for complications of concomitant arch replacement. We hope that future studies will resolve this paradox and tell whether the cost of arch replacement at the first operation will be offset by a less complex reoperation.

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Reference
Operation before dissection!
Prepare for a second OP

Acute Type A in a Marfan patient

Aim for

- Hemiarch replacement for better long-term FU with equivalent hospital mortality
- Prepare for downstream procedures place an ET!
Limited role of aortic size in the genesis of acute type A aortic dissection

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Abstract

Objective: Increased dimension of the aortic root and proximal aorta is considered a significant risk factor for catastrophic events that involve the ascending aorta. The objective of this study was to determine the possible correlation between pre-dissection aortic diameter and the occurrence of Stanford type A aortic dissection. Methods: Samples of dissected ascending aortas were obtained from 220 patients at the time of their operation. Two groups were identified: patients with connective tissue disorders (Group 1, n = 94) and those without (Group 2, n = 126). Measurements of the true (intimal) lumen were conducted and extrapolated as reliable approximation of pre-dissection aortic diameter. The possible association of intimal diameter with anthropometric and demographic data was analyzed. Results: Median aortic diameter was, respectively, 41.8 and 41.3 mm for patients with and without connective tissue disorders (41.4 mm for the entire cohort). Data analysis indicated that 57% of patients had aortic diameter above 40 mm, while patients with frank aneurysm accounted only for 10%; this proportion was higher in Group 1 compared to Group 2 (17.2% vs 4.7%). Poor or no correlation was demonstrated between aortic size and any of the anthropometric or demographic variables essayed. Significant subgroup differences were found among patients with a history of cigarette smoking, hypertension, diabetes, chronic renal insufficiency, and bicuspid aortic valve. Conclusion: Although aortic diameter remains a strong indication for preventive surgery in patients with inherited connective tissue disorders, acute aortic dissection occurs rarely in the setting of true ascending aortic aneurysms, and despite normal or near-normal aortic size in more than one-third of subjects. Dissection superimposing on small aortic diameters can be regarded as an expression of substantial functional tissue susceptibility to aortic catastrophic events.

Keywords: Aorta; Dissection; Marfan syndrome
In Marfan’s with Aortic disease, you should look for experts, and …

1. lower your threshold for repair of isolated ascending enlargement

2. lower your threshold for (hemi) arch Replacement in acute type A dissection

3. be prepared for downstream pathology

4. Close patient surveillance!
When should my bell ring … ?

Sudanese-born NBA player Manute Bol, tallest (2.31 m) player ever to appear in the NBA.
Sergei Rachmaninov 1873-1943
classical composer, thought to have had Marfan Syndrome

His long fingers, thought to be due to Marfan Syndrome, gave him and **exceptional reach on the piano**
Aortic repair in Marfan’s – should we lower the threshold for treatment?

Antoine Bernard-Jean Marfan (1858 -1942)

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