



# Endovascular Management in Marfan Syndrome



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## Introduction

Marfan syndrome (MFS) is the most common inherited connective tissue disease and is characterised by mutations of the fibrillin 1 (FBN1) gene encoding the protein fibrillin-1 [1]. MFS carries an increased risk of aortic dilatation, dissection and rupture, which are responsible for increased and early mortality rate. Prevention of aortic dissection is one of the cornerstones of care in patients with Marfan syndrome and prophylactic surgical replacement of dilated aortic root and/or ascending aorta has significantly increased life expectancy of patients with Marfan syndrome. In a worst case scenario of aortic dissection, open surgery is the treatment of choice in the setting of MFS regardless of type and location [2].

## Synopsis

Endovascular treatment as an alternative to open surgery in patients with MFS is controversial. Recommendations from the current guidelines and expert consensus papers are hesitant to promote elective endovascular treatment. Essentially only emergencies like aortic rupture, or re-interventions with unfavourable anatomy for open repair or a graft-to-graft approach may be considered for endovascular management [3]. Recently extracted data from the IRAD on all patients with MFS suffering from dissection collaborated either conservative or open surgical management in MFS [4].

Recent IRAD data [5] in patients with Marfan syndrome and type B aortic dissection were treated with open surgery in 28.7% vs 9.7% in non-MFS patients ( $P < 0.001$ ) and less frequently with medical management (50.0% vs 62.6%;  $P = 0.014$ ); endovascular and hybrid management were less common (19.1% and 2.1% respectively). Another very important finding from this registry is that patients with MFS more frequently needed to undergo re-interventions beyond the first year of follow-up; 5-year freedom from re-intervention was 54.4% vs. 88.0% in non-MFS patients ( $P < 0.001$ ) for patients initially treated with open surgery. For those with initial endovascular treatment freedom from re-intervention was 32.0% vs. 71.5% ( $P < 0.001$ ).

The main reason why the endovascular treatment has not been embraced yet was not the early outcomes, but rather the non-

sustained results and bad late outcomes. While hospital mortality varies between 2.5% to 12.5% and the stroke and spinal cord ischemic injury rates were 2.5% and 2.5% respectively [6-8] the endovascular approach appears to be associated with an early treatment failure rates between 25.0% to 43.8% primarily due to endoleaks [6].

A catastrophic complication associated with the use of endovascular stent grafts for patients with type B aortic dissection include retrograde aortic dissection and stent-graft induced distal new entry (SINE). The frequency of retrograde dissection in patients with MFS varies from 13.5% to 50% [9]. On the other hand, the presence of SINE among Marfan patients was found to vary between 27% and 33.3% [10,11].

The experience with endovascular treatment of type B dissection in Marfan syndrome is still limited both within IRAD and in view of only few published cases. Currently, experts believe that only after previous graft replacement of parts of the aorta and thereby creation of a safe landing endovascular therapy may have a role in Marfan syndrome. General belief is that when medical management fails, open surgery should be first-line therapy for Marfan syndrome patients with type B dissection, even in the endovascular era, unless the risk of open surgery is considered prohibitive or for life-threatening acute complications [12].

There is, however, a long-standing experience with endovascular management of certain peculiar cases of MFS. Hence, in our centre, every case requires an extensive discussion in a multidisciplinary meeting, favouring eventually a personalized approach in MFS. For instance endovascular appears a safe option as a graft to graft approach even in patients with MFS. For example, a previously implanted elephant trunk is a suitable landing pad for a deployment of stent-graft and reconstruction of distal dissection even in MFS. Patients with MFS and previous multiple operations and a relatively high risk for any open surgical procedure could also be considered candidates for an endovascular approach. In addition, hybrid approaches or entirely endovascular approach by the use of a fenestrated stent-graft with additional side branch stenting are emerging as more complex procedures that could offer acceptable long-term results without the need for thoracotomies in

patients with MFS [13]. Finally percutaneous “neo-branching” could be a safe and sustained solution and more important a minimally invasive technique achieving a total exclusion of an aneurysm by anchoring a graft snugly within the target vessel [14].

## Conclusion

Modern endovascular management is emerging as an additional option in the management of patients with MFS. A broader application of endovascular techniques is possible with advances in stent-graft technology and increasing skills. Appropriate pre-procedure planning, careful selection of favourable cases and an individualised approach is needed in order to grant even patients with MFS the benefits of an endovascular approach.

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