

External aortic root support in Marfan syndrome

Interventional procedures guidance

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[nice.org.uk/guidance/ipg394](https://www.nice.org.uk/guidance/ipg394)

Your responsibility

This guidance represents the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, healthcare professionals are expected to take this guidance fully into account. However, the guidance does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or guardian or carer.

Commissioners and/or providers have a responsibility to implement the guidance, in their local context, in light of their duties to have due regard to the need to eliminate unlawful discrimination, advance equality of opportunity, and foster good relations. Nothing in this guidance should be interpreted in a way that would be inconsistent with compliance with those duties.

Commissioners and providers have a responsibility to promote an environmentally sustainable health and care system and should assess and reduce the environmental impact of implementing NICE recommendations wherever possible.

1 Guidance

- 1.1 Current evidence on external aortic root support in Marfan syndrome is based on small numbers of patients. The evidence on safety shows occasional serious adverse events and the evidence on efficacy is limited to the short term. Therefore this procedure should only be used with special arrangements for clinical governance, consent and audit or research.

- 1.2 Clinicians wishing to undertake external aortic root support in Marfan syndrome should take the following actions.
- Inform the clinical governance leads in their Trusts.
 - Ensure that patients and their carers understand the uncertainty about the procedure's safety and efficacy and provide them with clear written information. In addition, the use of NICE's information for patients ('Understanding NICE guidance') is recommended (available from www.nice.org.uk/guidance/IPG304/publicinfo).
 - Audit and review clinical outcomes of all patients having external aortic root support in Marfan syndrome (see section 3.1).
- 1.3 Further research should report on long-term outcomes, particularly the occurrence of dissection and aortic dilatation, and the need for further procedures.

2 The procedure

2.1 *Indications and current treatments*

- 2.1.1 Marfan syndrome is a genetic connective tissue disorder typically associated with long limbs, lens dislocation and aortic root dilatation.
- 2.1.2 In patients with Marfan syndrome the proximal aorta progressively dilates and is prone to dissection and rupture. Conventional management involves preventative surgery to replace the ascending aorta with a prosthetic graft. The aortic valve may also be replaced, or the native valve reimplemented.

2.2 *Outline of the procedure*

- 2.2.1 The aim of external aortic root support in Marfan syndrome is to prevent enlargement and subsequent dissection or rupture, without the need for graft replacement and aortic valve surgery.
- 2.2.2 Magnetic resonance imaging (MRI) is used to map the precise anatomy of the patient's aorta. Computer-aided design is used to make a bespoke external polymer mesh support.

- 2.2.3 The mesh support is sutured into place around the aorta via a median sternotomy. Cardiopulmonary bypass is usually not needed.

Sections 2.3 and 2.4 describe efficacy and safety outcomes from the published literature that the Committee considered as part of the evidence about this procedure. For more detailed information on the evidence, see the overview, available at www.nice.org.uk/guidance/IP/885/overview

2.3 *Efficacy*

- 2.3.1 In a case series of 20 patients with a median follow-up of 20 months, post-operative changes in aortic root diameter (assessed by MRI) were reported in 16 patients, ranging from a decrease of 6 mm to an increase of 3 mm, with the median change being a decrease in diameter of 1 mm.
- 2.3.2 The case series of 20 patients reported that cardiopulmonary bypass was used for 20 minutes only in the first patient treated with external aortic root support. Among the comparator group of 7 patients treated by aortic root replacement, the average bypass time was 149 minutes (range 139–243 minutes).
- 2.3.3 In the case series of 20 patients, all were alive and well (not otherwise described) at the time of the last follow-up (median 20 months).
- 2.3.4 The operation was completed as planned in 95% (19/20) of patients in the case series of 20 patients.
- 2.3.5 The case series of 20 patients compared operative characteristics with those of 7 patients treated by aortic root replacement with a prosthetic graft. The median operation times were 148 minutes (range 125–415 minutes) and 374 minutes (range 240–493 minutes) respectively.
- 2.3.6 The Specialist Advisers listed key efficacy outcomes as avoidance of cardiopulmonary bypass, reduced bleeding, prevention of ascending aortic rupture or dissection, and prevention of ascending aortic enlargement. They noted that there is uncertainty about whether the procedure will prevent aortic dissection in the long term, and whether it will prevent deterioration of the aortic valve.

2.4 *Safety*

- 2.4.1 In the case series of 20 patients, 1 had a post-operative cardiac arrest with ventricular fibrillation. The circulation was restored after removing the anterior closing suture on the aortic root support. The authors observed that the location of one of the coronary origins might have been misinterpreted on the MRI scan. The patient made a good recovery.
- 2.4.2 Among the first 10 patients in the case series of 20 patients, 2 had transient atrial fibrillation after the procedure.
- 2.4.3 The Specialist Advisers considered theoretical adverse events to include infection, haemorrhage, migration of the mesh support and possible constriction of the coronary arteries. They listed an additional theoretical possibility of thinning of the aortic wall inside the external support.

2.5 *Other comments*

- 2.5.1 The Committee noted that this procedure has the potential for more rapid recovery than aortic replacement surgery, and that treatment may confer psychological benefits and improve quality of life. Commentary from several patients reported a positive effect on their emotional health and wellbeing, stating that the procedure had given them peace of mind.
- 2.5.2 However, the Committee noted that this is a preventative procedure for a condition with an unpredictable natural history, in terms of the progress and extent of aortic dilatation and the occurrence of aortic dissection. Uncertainties about these long-term outcomes underlie the recommendation for further research.

3 **Further information**

- 3.1 This guidance requires that clinicians undertaking the procedure make special arrangements for audit. NICE has identified relevant audit criteria and has developed an audit tool (which is for use at local discretion), available from www.nice.org.uk/guidance/IPG394
- 3.2 For related NICE guidance see www.nice.org.uk

Information for patients

NICE has produced information on this procedure for patients and carers ('Understanding NICE guidance'). It explains the nature of the procedure and the guidance issued by NICE, and has been written with patient consent in mind. See www.nice.org.uk/guidance/IPG394/publicinfo

Endorsing organisation

This guidance has been endorsed by [Healthcare Improvement Scotland](#).

Accreditation

