NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

Interventional procedures consultation document

Personalised external aortic root support (PEARS) using mesh to prevent aortic root expansion and aortic dissection in people with Marfan syndrome

Marfan syndrome is a genetic disorder in which the large artery from the left side of the heart (the aorta) can expand to the point where the inner lining can tear (dissection), risking a fatal rupture. In this procedure, which is done under general anaesthesia, the chest is opened through the breastbone. A mesh is then wrapped around the outside of the aorta at the part closest to the heart (the aortic root). The aim is to support the aorta to stop it from expanding and reduce the risk of rupture.

This is a review of NICE's interventional procedures guidance on external aortic root support in Marfan syndrome.

NICE's interventional procedures advisory committee met to consider the evidence and the opinions of professional experts, who are consultants with knowledge of the procedure.

This document contains the draft guidance for consultation. Your views are welcome, particularly:

- comments on the draft recommendations
- information about factual inaccuracies
- additional relevant evidence, with references if possible.

NICE is committed to promoting equality of opportunity, eliminating unlawful discrimination and fostering good relations between people with particular protected characteristics and others.

This is not NICE's final guidance on this procedure. The draft guidance may change after this consultation.

Issue date: November 2021

After consultation ends, the committee will:

- meet again to consider the consultation comments, review the evidence and make appropriate changes to the draft guidance
- prepare a second draft, which will go through a resolution process before the final guidance is agreed.

Please note that we reserve the right to summarise and edit comments received during consultation or not to publish them at all if, in the reasonable opinion of NICE, there are a lot of comments or if publishing the comments would be unlawful or otherwise inappropriate.

Closing date for comments: 16 December 2021

Target date for publication of guidance: April 2022

1 Draft recommendations

1.1 Evidence is adequate on the short-term safety and efficacy of personalised external aortic root support (PEARS) using mesh to prevent aortic root expansion and aortic dissection in people with Marfan syndrome. Evidence is limited on long-term outcomes in quantity and quality. Therefore, this procedure should only be used with special arrangements for clinical governance, consent, and audit or research. Find out what special arrangements mean on the NICE interventional procedures guidance page.

1.2 Clinicians wanting to do PEARS using mesh to prevent aortic root expansion and aortic dissection in people with Marfan syndrome should:

- Inform the clinical governance leads in their healthcare organisation.
- Give patients (and their families and carers as appropriate) clear written information to support shared decision making, including NICE's information for the public.
- Ensure that patients (and their families and carers as appropriate) understand the procedure's safety and efficacy, and any uncertainties about these.
- Audit and review clinical outcomes of all patients having the procedure. The main efficacy and safety outcomes identified in this guidance can be entered into NICE's interventional procedure outcomes audit tool (for use at local discretion).
- Discuss the outcomes of the procedure during their annual appraisal to reflect, learn and improve.

1.3 Healthcare organisations should:

- Ensure systems are in place that support clinicians to collect and report data on outcomes and safety for every patient having this procedure.
- Regularly review data on outcomes and safety for this procedure.

1.4 Patient selection should be done by a multidisciplinary team.

1.5 The procedure should only be done in specialist centres with experience of managing this condition, by surgeons trained and experienced in aortic root surgery.

1.6 Further research should report details of patient selection, including aortic diameter, and long-term outcomes, including evidence of disease progression, such as dilation and dissection of the aortic root.

2 The condition, current treatments and procedure

The condition

2.1 Marfan syndrome is a genetic disorder of the connective tissues. One effect of it is that the wall of the aorta can weaken and progressively widen. The wall can tear (dissection) and possibly rupture, which is often fatal. The strongest predictors of dissection are the aortic root size and the rate of change in size over time.

Current treatments

2.2 Conventional treatment involves pre-emptive surgery to replace the ascending aorta with an artificial fabric graft. Some clinicians recommend this when the aortic diameter is 45 mm or more. The aortic valve is also usually replaced but may be conserved. Patients can experience considerable anxiety waiting for their aorta to reach the size threshold recommended for surgery.

2.3 If the patient has a mechanical valve implanted, they need lifelong anticoagulation. If a bioprosthetic valve is used, it is likely to eventually fail and the patient will need another operation. Valve-sparing root replacement surgery, in which the aorta is replaced with a tube graft and the native aortic valve is conserved, is also suitable for some patients with normal valve function. This is technically more challenging, and patients may need further surgery to replace the aortic valve at a later date.

The procedure

2.4 The aim of personalised external aortic root support (PEARS) using mesh in people with Marfan syndrome is to reinforce the aortic root and ascending aorta to prevent enlargement and subsequent dissection or rupture. The native aortic valve is left intact so there is no need for lifelong anticoagulation after the procedure. This is a particular advantage for young women considering future conception. Cardiopulmonary bypass is usually not needed, and the operative time is shorter than traditional aortic root replacement.

2.5 The first step of the procedure is to do imaging studies of the patient's ascending aorta and aortic root. Computer-aided design is used to create a 3-dimensional model of the aorta, which is then used to make a bespoke external polymer mesh support. The mesh is soft, flexible and porous. Openings for the coronary arteries are fashioned into the mesh support.

2.6 Under general anaesthesia, a median sternotomy is done and the aorta is dissected away from adjacent structures and proximal to the coronary arteries. The mesh support is passed behind the aorta, sutured up the front and secured to the aortoventricular junction. It fully encircles the aortic root and extends from the

region of the valve annulus to the origin of the brachiocephalic artery.

3 Committee considerations

The evidence

3.1 NICE did a rapid review of the published literature on the efficacy and safety of this procedure. This comprised a comprehensive literature search and detailed review of the evidence from 8 sources, which was discussed by the committee. The evidence included 3 cohort studies, 2 non-randomised comparative studies, 2 case series and 1 case report. It is presented in the summary of key evidence section in the interventional procedures overview. Other relevant literature is in the appendix of the overview.

3.2 The professional experts and the committee considered the key efficacy outcomes to be: aortic-related pathology, including dissection and dilation, need for aortic valve replacement, and anxiety about progressive aortic disease.

3.3 The professional experts and the committee considered the key safety outcomes to be: bleeding, infection and atrial fibrillation.

3.4 Thirteen commentaries from patients who have had this procedure were discussed by the committee. All the patients responded that they would recommend the procedure to another patient with their condition and several noted the benefit of not having to take anticoagulants for the rest of their lives.

Committee comments

3.5 The committee was informed that surgeons receive proctoring for their first few procedures.

3.6 The committee encourages the establishment of a registry for this procedure.

3.7 The procedure is used for other aortopathies as well as Marfan syndrome.

Tom Clutton-Brock
Chair, interventional procedures advisory committee
November 2021

ISBN: