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Editorial
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Preventive Aortic Root Intervention for Marfan Patients - How Much Longer are we Still Waiting?

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Editorial

Marfan syndrome is an autosomal dominant disorder affecting skeletal, ocular and cardiovascular systems with an occurrence estimated to range from 1 in 3,000 to 1 in 10,000 people [1]. One of the most prominent cardiovascular manifestations is aneurysmal dilatation of the aorta, particularly at the level of the valsalva sinuses, with a predisposition for aortic tearing and rupture. With rapidly progressive aortic dilatation or aortic regurgitation, aortic root repair becomes inevitable, with surgical options for composite graft replacement, valve-sparing operation, or homograft root replacement [2]. Personalized External Aortic Root Support (PEARS) is a preventive surgery that has been made available to Marfan patients with asymptomatic aortic dilation since 2004 [3]. Despite being available for 16 years it remains contentious within the cardio-thoracic community. The ExoVasc® PEARS is a computer-aided design and 3D printed mesh implantation created to fit a patient-specific aortic root morphology [4]. The operation has undergone (British) Health Technology Appraisal under the auspices of National Institute of Care and Excellent (NICE) in England and successfully obtained approval for selected patients [5]. In April 2020 a audit was conducted on all PEARS procedures, it revealed that there had been 321 cases successfully preformed across 29 surgical centers in Europe, Australia and New Zealand. An internal review of these cases revealed excellent results: Low peri-operative mortality rate of 0.03%, shorter procedural time, a large proportion of off-pump, and faster recovery comparing with the standard cardiac surgeries which normally require cardiac arrest and cardiac pulmonary bypass. During the 16 years interim clinical monitoring and follow-up imaging studies no indication of long-term complications from the PEARS procedure [1,6]. Arguably, over several years, PEARS is still capable to prevent further expansion of a slightly dilated aortic root/ascending aorta, similarly preventing any expansion of the aortic annulus or sinotubular junction and eventual aortic valve regurgitation as well as Stanford type A aortic dissection [7]. The follow-up data obtained from the PEARS patients so far has been overwhelmingly positive and convincing in terms of stopping dilation and as a result of minimizing the risk of dissection in the future. At present there is an absence of any clinical trial designed for evaluating PEARS procedure. One reason is the impossible of randomization such as blinding due to the sham thoracotomy would not be ethically viable [8]. Another prime reason is that Marfan syndrome is excitedly rare and as a result there is a lack of individual patients who face the option of which operation to have at a point in their lives. Although a comparative trial remains non excitant, a potential way to scientifically justify PEARS procedure could be a meticulous compulsory registry [9] indiscriminately documenting all PEARS cases from all centers worldwide. An external body should be appointed to supervise the compulsory registry as well as full data submission and audit from any participating centers. The surgeons or centers, who are interested in embracing PEARS technique, should be offered to be trained in a large volume centre with a standardized protocol [10]. By this means, sufficient evidence can be given over time to recommend this new technology and surgery to selected patients, even without any randomized controlled clinical trials. The interaction of clinical observation, inference based on biological understanding and surgeons' experience and skills often plays an important role in medical decision making when clinical evidence such as randomized clinical trial alone is not feasible. Optimal judgment in such a scenario may rely on the integration of observational evidence, understanding of human biology and patient preference as well as surgical skills.

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