REVIEW ARTICLE

Established practice may be disappointing, but a critical mind can always clarify problems: The Case of PEARS procedure

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After the introduction of Valve Sparing Root Replacement and its modifications, it has been a continuous challenge to improve results of standard procedures, in particular regarding overall survival and freedom from reoperation. Likewise, as aortic root surgery became safer, clinicians felt entitled to offer surgery earlier in natural history of patients with aortic dilatation. The development of a tailored, Personalized External Aortic Root Support (PEARS) offers the opportunity to re-think the process of care for patients at major risk for aortic dissection, especially those with connective tissue disorders and congenital heart defects. The distinctive features of procedure allow a truly preemptive management of these populations, lowering the aortic size at which surgery can be safely and electively performed. The radical idea that resulted in PEARS was to conserve, rather than remove, the diseased aortic wall. In addition, the opportunity to customize the device fulfills the goal to tailor the procedure to each patient, which is a key concept of modern surgery.

Key words: **Aortic root; Aortic dissection; Congenital heart defects; Connective tissue disorders; PEARS.**

Después de la introducción de las Valve Sparing Root Replacement y sus modificaciones, ha sido un desafío continuo mejorar los resultados de los procedimientos estándar, en particular con respecto a la supervivencia general y la ausencia de reoperación. Asimismo, a medida que la cirugía de la raíz aórtica se volvió más segura, los médicos se sintieron más comprometidos a ofrecer una cirugía más temprana en la historia natural de los pacientes con dilatación de la raíz aórtica. El desarrollo de un soporte de raíz aórtico externo personalizado (PEARS) ofrece la oportunidad de replantear el proceso de atención de los pacientes con mayor riesgo de disección aórtica, especialmente aquellos con trastornos del tejido conectivo y defectos cardíacos congénitos. Las características distintivas del procedimiento permiten un manejo verdaderamente preventivo en estos subgrupos, reduciendo el tamaño aórtico al que se puede realizar la cirugía de manera segura y electiva. La idea radical que dio lugar a PEARS fue conservar, en lugar de eliminar, la pared aórtica enferma. Además, la oportunidad de personalizar el dispositivo cumple con el objetivo de adaptar el procedimiento a cada paciente, que es un concepto clave en era de la cirugía moderna.

Palabras clave: **Raíz aórtica; Disección aórtica; Defectos congénitos cardiacos; desórdenes del tejido conectivo; PEARS.**

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Congenitally determined aortic root and wall pathologies carry a high risk of life-long complications, such as aortic valve regurgitation (AoR), aortic dissection (AD) or acute rupture [1].

All the aforementioned conditions are associated with a significant mortality rate in the mid- and long- term.

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Aortic dilatation is a well-recognized feature of a wide spectrum of inherited connective tissue disorders (CTD), including Marfan syndrome (MS), Turner syndrome (TS), Loeys-Dietz syndrome (LDS) and bicuspid aortic valve (BAV) [2]. In addition, congenital heart diseases (CHD) such as single ventricle (SV), common arterial trunk (CTA), transposition of the great arteries (TGA), aortic coarctation (CoA), hypoplastic left heart syndrome (HLHS), and tetralogy of Fallot (TOF) are associated with aortic medial abnormalities, aortic dilatation, and AoR [3].

In presence of an aortic root aneurysm, prophylactic surgery on the root with the aim of preventing catastrophic events (AD/aortic rupture) is the most important life prolonging treatment for patients, either with CHD or not [4]. This strategy, combined with early detection, diagnosis and monitoring have dramatically reduced deaths from dissection and increased the longevity of this population.

To date, three forms of surgery are commonly adopted to address this issue:

1) Total Root Replacement (TRR) with a valved conduit (Bentall procedure);

2) Valve-Sparing Root Replacement (VSRR);

3) the recently introduced Personalized External Root Support (PEARS) [5] with a microporous mesh sleeve.

After the introduction of VSRR and its further modifications, it has been a continuous challenge to improve the results of the consolidated and standard procedures, in particular with respect to overall survival and freedom from re-operation.

Likewise, as aortic root surgery became safer, clinicians felt entitled to offer surgery earlier in the progression of various diseases associated with aortic dilatation, especially CTD. However, all the solutions described so far are major operations with protracted operative time, as well as inherent high morbidity and mortality, which means that they carry risks related either to the procedure itself or, later in life, to the possibility of re-intervention and the side-effects of the required medical therapy.

As history teaches, innovation in cardiovascular surgery can be sometimes driven by an interdisciplinary collaboration, for example between surgeons and engineers, as for the latest developments in the field of aortic surgery.

The engineer Tal Golesworthy from Gloucester, moved by the need to fix his own diseased Marfanoid aorta, used the spatial data from high-resolution digital images to construct an exact replica of the vessel, a process known as computer assisted design (CAD), so as to fashion a dedicated support before opening the chest, rather than attempting its construction from relatively stiff material during surgery [6]. This rapid prototyping allowed revising the idea of wrapping the aorta, involving this time a bespoke "jacket", perfectly fitting the individuals' vessel, from the LV to the proximal arch [7].

Magnetic Resonance Imaging (MRI) digital information can be used as a starting point to create this replica of the patient's aorta (including the sinuses and the coronary arteries origin). On this model, it is then possible to build an external support with an exact match to the contours of the diseased vessel [8]. This support, placed around the patient's aorta at time of surgery, subsequently gets incorporated into the adventitia and forms a composite (**Fig. 1**).

The development of a tailored, Personalized External Aortic Root Support (PEARS) offered the opportunity to radically re-think the process of care for patients at major risk for AD, especially those with CTD [9]. Not by chance, the inspiring model for this procedure comes from MS.

Figure 1. Surgical view of PEARS procedure (personal drawings of the authors).

The distinctive and unique features of the graft and the technical details of the operation imply the opportunity of a truly preemptive management of these special populations, lowering the aortic size at which surgery can be safely and electively performed [10].

Another important potential use of this new technique is in combination with the standard Ross procedure [11]. The rationale for this surgery, which is usually referred to as Ross-PEARS, came from the externally supported Ross procedure described by Slater and Colleagues [12], which uses a Dacron (DuPont, Wilmington, DE) graft to support the neo-aortic root with the aim of preventing its progressive dilatation, a known pitfall of the standard technique. It is possible to achieve the same result combining PEARS with the pulmonary autograft of the Ross procedure (**Fig. 2**).

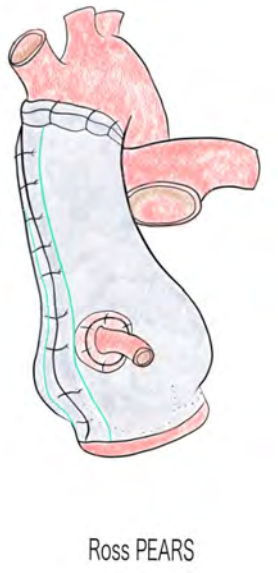

Figure 2. Schematic representation of Ross-PEARS (personal drawings of the authors).

In addition, it has been found that the external mesh, closely fitting the aorta, becomes fully incorporated in the adventitia and preserves the vascular architecture, in contrast to the conventional wrapping with low porosity and poorly fitting Dacron grafts also when used in combination with Ross procedure [13,14].

Future investigations on the impact of mesh implantation around the PA could bring further insights regarding this novel application of the PEARS, which so far has not been extensively explored in the clinical practice.

In conclusion, the radical idea that resulted in PEARS was to conserve, rather than remove, the diseased aortic wall. This straight away avoids all the problems related to the blood/endovascular interface and greatly reduces the magnitude of surgery. In addition, the opportunity to customize, individualize or personalize the device, made possible by modern image acquisition, manipulation and going from images to a three dimensional product, perfectly match with the goal to tailor the procedure to each patient, which is a key concept of modern surgery [5]. In order to make the procedure as safe as possible and get the best final result, a dedicated Team is mandatory, in particular considering that the surgical handling of a diseased and weak aortic tissue is challenging, especially at the level of root and coronary arteries. For the same reason, operators must be familiar with all complications and options in aortic root surgery. Accordingly, CPB should be always available in the operating room, in order to manage promptly and properly an unexpected dangerous scenario.

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