When and How to Enlarge the Small Aortic Root

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Successful enlargement of the small aortic root in children has remained a management challenge, particularly in the neonates and small infants. Achieving this aim requires thorough understanding of the anatomic features of the left ventricular outflow tract, careful patient selection, and skilful execution of complex surgery. This article reviews the anatomical principles upon which the surgical techniques rely, the decision-making, the timing of surgery, the surgical options, and the outcomes.
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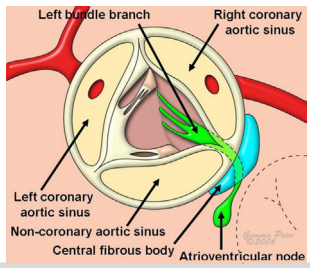
Introduction

Left ventricular outflow tract (LVOT) obstruction in neonates, infants, and small children is one of the remaining challenges to which there is no good surgical solution. Although the LVOT is a very short channel, the anatomy, mechanical function, and performance of the aortic root are highly complex with perfect integration. To restore the functional integrity in the case of surgery on the hypoplastic aortic annulus, often surrounded by diseased LVOT, robust pre-operative work-up would be required to select those suitable for timely aortic root/LVOT interventions. It is well accepted that the use of valve (and annulus) conserving operations to allow the children to grow, instead of applying the Ross +/− Konno operation in early infancy, can minimise risk.[1–3] However, the real question of when to enlarge a small aortic root, which is 'how small is too small,' must be addressed. Its untimely delay can risk the development of circulatory failure and excessive ventricular hypertrophy.

It comes as no surprise that neonates/small infants with congenital aortic stenosis with a hypoplastic aortic root and/or multiple levels of LVOT obstruction can carry many pitfalls, such as a borderline left ventricle, the presence of endocardial fibroelastosis, smallish mitral valve, arch hypoplasia, and pulmonary hypertension. These are real challenges. On the other hand, the current root enlargement technique of choice is the Ross-Konno procedure, apart from some institutionally related technical modifications including the concomitant resection of endocardial fibroelastosis, which can benefit these children with a reconstructed, wide-open, and competent LVOT without growth limitation. This at least provides a robust physiological platform to address other important haemodynamic lesions as part of a staged management of complex LVOT syndrome.



The morphology of the aortic root and complex topography of adjacent structures including, the sinotubular junction, the sinus of Valsalva, the tricuspid and mitral valves, the coronary arteries and the conduction system.

Central Message

Understanding the anatomy and physiology of the aortic root is the key to successful enlargement.

The Aortic Root is a Highly Sophisticated Structure

The two main functions of the aortic root are: 1) to act as an anchor for the synchronised movement of the inlet mitral valve and outlet semilunar valve; and 2) to support the intricate movements of the aortic valve cusps. The fact that our best

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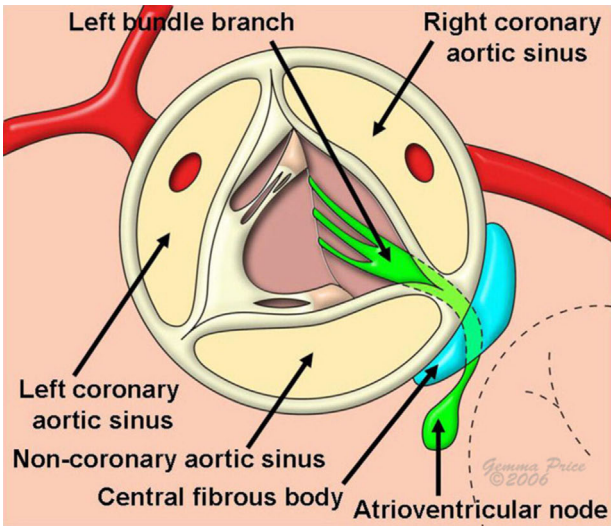

Figure 1 Illustration of the complex anatomy of the aortic root. (Reprinted with permission from Anderson RH. The surgical anatomy of the aortic root. Multimed Man Cardiothorac Surg 2007;2007 (102).[10])

cardiac morphologists are still not in agreement on how to define the existence of the 'aortic annulus' suggests that this is a structure of exquisite design. The aortic annulus is best described as crown shaped, and it is to this shape and structure that the aortic valve is attached and supported in the root.[4,5] For every heartbeat, studies applying cinematography and contrast injections have visualized aortic root motion wherein the root is displaced downward during systole and returns to its previous position in diastole.[6] Magnetic resonance imaging (MRI) studies have elegantly revealed an axial downward motion and a clockwise twist during systole.[7–9] It has been suggested that this motion retracts the aortic root into the LVOT and presumably protects the root itself from unnecessary dilatation during systole.

The challenge for the surgeon is to understand this exquisite morphology and function of the aortic root and then try to surgically reproduce as many aspects of it as possible. In this endeavour, the surgeon will also need to have a precise understanding of the topography of adjacent structures, including the sinotubular junction, the sinus of Valsalva, the tricuspid and mitral valves, the coronary arteries, and the conduction system (Fig. 1).[10]

Essential Pre-Operative Work-Up

Echocardiography

The cardiac ultrasound examination is the fundament of all pre-operative work-ups. In this specific context, it can determine the degree and impact of obstructions at multiple levels along the entire LVOT. The aortic valve morphology, size, and function can be assessed. A cardiac ultrasound can also provide important assessments of the mitral valve size and function, left ventricular volume, and the possible presence of endocardial fibroelastosis. Associated malformations, including arch hypoplasia or interruption and coarctation, and pulmonary vascular issues such as pulmonary hypertension can be identified. The management of neonates/small infants with a borderline left ventricle can be very challenging in the assessment of its adequacy to support a systemic circulation.[11]

MRI

In our experience, the main limitation of MRI is its availability of expert acquisition of data and its interpretation, and then there is the need of sedation or general anaesthesia in young patients for quality imaging. MRI can otherwise provide valuable flow and volume data to provide additional information on the size of the left ventricle and its hemodynamic performance. These data are particularly important in cases of borderline left ventricle and presence of intra and extra cardiac shunts, where the assessment of left ventricular systolic and importantly diastolic function is difficult. In the absence of a reliable non-invasive test, MRI combined with ultrasound may increase the sensitivity to the diagnosis of endocardial fibroelastosis. MRI can also provide additional information on quality and size of the pulmonary valve.

Computer Tomography

Modern computed tomography (CT) examination is very fast and normally does not require any sedation or general anaesthesia. Its popularity is further enhanced with anatomical and 3D reconstruction in complex associated malformations, including total anomalous venous return, interrupted aortic arch, and coarctation.

Cardiac Catheterization

In the presence of MRI and CT, conventional catheterization is of limited value from a diagnostic point of view. However, many newborn and younger children will have undergone a conventional catheterization in a prior attempt to dilate the critical aortic stenosis. This has become the main cause of aortic regurgitation.

Decision-Making and Timing

In children with significant aortic valve/annular hypoplasia, the most important issue is to achieve effective delay of the inevitable valve replacement without undue compromise of the myocardial function. When the aortic root structures display small dimensions with Z-score $<$ -2.5 to -3.0,[12] a root enlargement procedure would be considered. The Z- score would only be a guide and its measurement and translation from real dimension can be skewed in very small children. Obviously, the smaller the dimension the stronger is the indication for root enlargement procedure. However, a too small aortic root may, from a surgical technical point of view, be non-permissive of safe coronary transfer and therefore outweigh the advantages of the pulmonary autograft implantation procedure. There may be some discrepancy between the smallish aortic valve annulus and the larger pulmonary valve. The latter is usable as long as the valve function does not