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SCIENTIFIC EDITORIAL

Aortic valve reconstruction in children: A new string to our bow



La reconstruction valvulaire aortique chez l'enfant : une nouvelle corde à notre arc

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Background

During the Renaissance, one of greatest Italian anatomists of the time, Leonardo da Vinci, surprisingly reproduced the anatomy of the left ventricular outflow tract (LVOT); since then, its sumptuous architecture has been a source of inspiration to many geniuses. However, surgical repair of the aortic valve remains a challenge for modern-day cardiac surgery.

Over time, numerous anatomical and physiological studies have revealed, down to the last detail, the secrets of how the aortic valve functions, and embryological knowledge of its morphogenesis has been almost totally acquired. Consequently, conservative options for aortic valve treatment have evolved. Over the past two decades, this knowledge has facilitated progress in paediatric interventional procedures, and has improved and standardized our surgical techniques.

Abbreviations: AVR, Aortic Valve Replacement; LVOT, Left Ventricular Outflow Tract.

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Following Alain Carpentier's "French correction" for mitral valve disease, El Khoury et al., in 2005, proposed a functional classification of aortic valve abnormalities based on the movement of the aortic leaflets (type I, normal; type II, excess; and type III, restricted leaflet motion) [1]; an appropriate surgical repair (or, in the extreme, a valve replacement) is consequently advised. For the first time, we had a clear, standard, predetermined surgical strategy guided by physiopathological analysis of the aortic valve.

Although the results of this new methodology were encouraging, Polain de Waroux et al. identified some key points for valve reparability [2]: smooth, thin and large leaflets with redundant tissue were considered as repairable, whereas small, restrictive, fibrous or thickened leaflets were thought to preclude surgical repair. As we can see, the "quality of the repair" is determined by the "quality of the tissue".

Unfortunately, this valid tool developed for adult aortic pathology is not optimal for children's disease, for two reasons: (1) the aortic valve is usually congenitally malformed (endocarditis, root dilatation and calcification are rare in childhood), with the most common lesion being aortic stenosis; isolated aortic regurgitation is rare, and a mixed aortic valve disease is often the result of previous balloon or surgical commissurotomy, reasonably tolerated for years; and (2) in case of valvuloplasty failure, a rescue therapy is not achievable by a bioprosthesis substitution [3], and a mechanical valve replacement or a Ross procedure is the only acceptable alternative.

Another crucial point is "surgical timing" – an imperative variable in childhood. Certainly, the valve phenotype (tricuspid < bicuspid < unicuspid) plays an important role in prediction of freedom from reoperation, but – above all – younger age at surgery is a hindrance to durability of repair [4]. Finally, having moderate aortic regurgitation (\leq grade 2) on discharge or a mean transvalvular gradient \leq 20 mmHg was found not to be predictive of late valve failure [4].

So, what is the best strategy for the management of aortic valve diseases in children?

Surgical reasoning in children

The most common congenital heart defect is bicuspid aortic valve, which manifests mainly in infants and young children with isolated aortic stenosis – leaflet deformity and commissural fusion being the most common mechanism [5]. In this case, the treatment is well established, and the debate between surgery and percutaneous intervention is not within the scope of this editorial.

Excluding isolated aortic stenosis, the discussion remains open: aortic valve pathology in childhood is a broad spectrum of mixed aetiologies, and the real mechanism is often misunderstood until the aorta is opened. In this situation, the optimal operative technique is still a "surgical dilemma".

As mentioned previously, the long-term results of congenital aortic valve repair depend on the "quality of the tissue".

Regarding the "quality of the tissue", Mastrobuoni and El Khoury [6] proposed the classification of two scenarios from

a surgical point of view: (1) lesions with enough or an excess of tissue (or, in any case, with good quality of leaflets); and (2) lesions with a lack of tissue (such as unicuspid and postballoon valvotomy valves).

In the first scenario, several surgical options usually allow a successful primary repair by means of free margin shortening, such as central plication [7], without the need for exogenous material. Trusler et al., in 1992, described the interesting option of "lateral" leaflet plication for treatment of chronic aortic cusp prolapse, often seen in outlet-type ventricular septal defects [8]. In all these cases, excellent and durable results are expected.

In the second scenario, lesions with a lack of tissue are a challenge to manage surgically, and are the key issue under debate. This subgroup of congenital pathologies should be named "complex" aortic valve disease. Aortic regurgitation or a mixed disease represents the typical clinical presentation, and the surgical reasoning should be governed by the risk of failure and the expected long-term results. Two strategies are possible: replacement or "attempt" to repair.

Replacement strategies in childhood

Aortic valve replacement (AVR) in children is realized by mechanical prosthesis or autologous pulmonary graft substitution (Ross procedure). These procedures are always achievable, long-term results are similar and studies have shown good outcomes and an acceptable complication rate with both valve choices [9].

Lifestyle limitations and morbidity resulting from anticoagulation therapy are the main disadvantages of mechanical AVR. Furthermore, younger age (< 6 years) has been shown to be an independent predictive factor for reoperation [10], given the significantly increased risk of early and late death in children receiving smaller mechanical valves (patient-prosthesis size mismatch).

Even if pulmonary autograft AVR confers a survival advantage in this younger population [9], age between 0 and 10 years (and possibly 15 years) remains the "Achilles' heel" of the Ross procedure [11]. Pulmonary autograft and right ventricular outflow tract intervention (1.28%/year and 1.97%/year, respectively) are a source of mandatory reoperation in young children [12]. We do believe that the "modified Ross operation", described by Alain Prat in 2010 [13], with reinforcement of the pulmonary autograft (not indicated for growing structures), and a bigger right ventricular outflow tract conduit are the secrets behind the better results in the adult population compared with the paediatric population [11].

For these multiple reasons, when it comes to "complex" aortic valve treatment, even if the correction is not "perfect", congenital surgeons are forced to repair the aortic valve in order to delay the "gold standard" Ross procedure or an "adult-sized" mechanical valve replacement.

Conservative strategies for "complex" aortic valve disease

Aortic valve repair in children is an interesting option, but "attempts" may involve a cost associated with converting

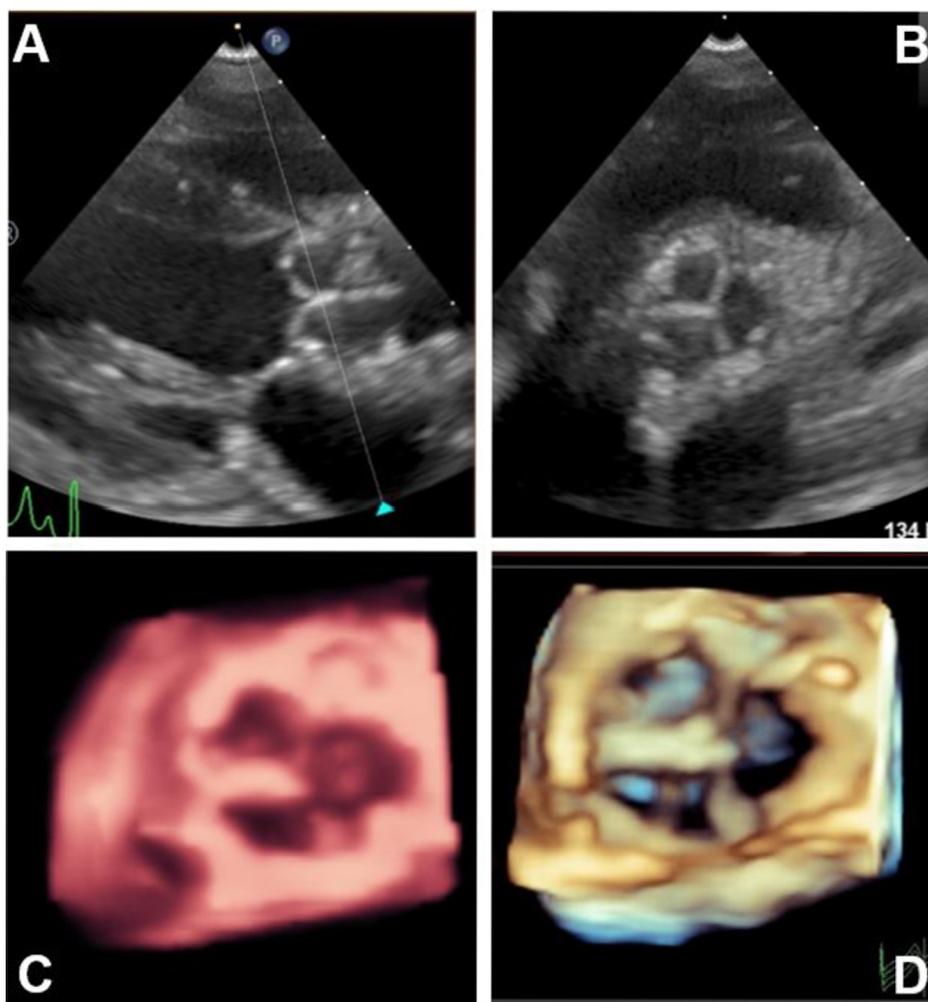


Figure 1. A and B. Two-dimensional transthoracic echocardiography of the new aortic valve (after the Ozaki procedure), from (A) the long-axis view and (B) the short-axis view. C and D. Three-dimensional transthoracic echocardiography of the aortic valve, from (C) the aortic view and (D) the left ventricular outflow tract view.

the initial procedure to AVR. Furthermore, the use of foreign materials is mandatory to recreate a good coaptation.

Tricuspidization, described by Pretre et al. in 2006 [14], could be an option in bicuspid aortic valve with at least one relatively normal leaflet. The new cusp should be created from the autologous pericardium. In the other cases, surgeons treat the “lack of tissue” by using patch cusp extension (associated with several combinations of techniques), cautiously tailored to minimize the risk of coronary ostial obstruction [15].

All of these procedures are clearly operator dependent, and suboptimal results are accepted, even if this is not necessarily satisfactory. In addition to the “well known” use of foreign materials, the durability depends on the morphology of the final reconstruction.

Absence of prolapse (coaptation tip below the level of the aortic annulus), coaptation length (> 4 mm) and symmetry of leaflets are considered universally to be predictors of durability [16]. Although the heterogeneous population makes data difficult to interpret, less satisfying outcomes are related to the non-tricuspid postrepair arrangement [17], and the degree of coaptation asymmetry (often

associated with complex lesion repair) seems to be strongly related to early reoperation rate [16]. Anyway, the state of the art and the longevity of complex repair are far from perfect!

A new option of “in between” repair and replacement is now available for the treatment of complex lesions: aortic valve reconstruction with glutaraldehyde-treated autologous pericardium. This “adult” technique, described by Ozaki et al. in 2014 [18], may offer some advantages in the paediatric population: surgeons create a new biological tricuspid aortic valve presenting all the morphological criteria for good outcomes.

Ozaki procedure: preliminary experience and conclusions

We recently started our “aortic valve reconstruction programme” at the Children’s Hospital of Toulouse, treating two cases (both aged 4 years), each presenting with a complex aortic valve disease, a lack of tissue resulting from a previous balloon angioplasty or a three-leaflet retraction.

The reconstruction was carried out as described meticulously by Ozaki et al. [18], without complications, and with a clamping time shorter than a “classical” Ross procedure.

Early postoperative results are encouraging, with less than trivial central valve regurgitation and a mean gradient < 20 mmHg, even in a smaller annulus. Echocardiographic evaluation showed perfect symmetry of the “new” leaflets, with a coaptation length > 10 mm without prolapse (Fig. 1).

We decided to perform the Ozaki procedure in children aged < 10 years, and even if some teams have associated this technique with annulus enlargement to treat patients with annular hypoplasia, we do believe that, at the moment, severe LVOT obstruction should be considered as a contraindication.

Definitely, the Ozaki procedure is a reproducible technique. Questions remain regarding the durability of the “foreign” material used for plasty, the feasibility in an annulus that is very small in size (< 13 mm) and the long-term effect of growing LVOT structures. The decision to treat the pericardium for a maximum of 5 minutes and future “customization”, derived from advances in new technologies (three-dimensional simulation, bioprinting), could lead to further evolution of the technique.

A multicentre study with long-term follow-up is required to define the limits of this procedure and, consequently, to improve our decision making.

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Disclosure of interest

The authors declare that they have no competing interest.

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