

Congenital Aortic Stenosis: Valve Repair after Valvuloplasty

Severe congenital aortic stenosis (CAS) often requires treatment in the early stages of life. Initial treatment is surgical commissurotomy or balloon valvuloplasty. Sometimes these approaches lead to aortic regurgitation. Once it occurs the only solution is valve replacement with mechanical prostheses or pulmonary autograft. Mechanical prostheses require anticoagulation and have a significant incidence of reintervention and mortality. (1) Replacement with pulmonary autograft does not require anticoagulation but is a more aggressive surgery involving a second valve and its long-term outcome has not yet been established. (2) “Bicuspidization” of unicuspid aortic valves has been proposed as a reconstructive surgery (3). Our hypothesis is that this concept is also applicable to a unicuspid valve that has become insufficient after treatment of the stenosis surgical or percutaneous. We describe a case in which “bicuspidization” was used in this scenario.

We present the case of a child diagnosed with CAS at birth. A commissurotomy was performed 14 days after birth. The gradient disappeared but moderate to severe aortic regurgitation appeared. He was reoperated at 33 months to rebuild the valve. Apparently, the fused tissue was divided between the valve cusps, tricuspidizing the valve and placing shortening sutures on the free leaflet edges.

Postoperative echocardiography revealed grade 2 regurgitation, with progressive center jet over the next two years, presenting severe regurgitation with left ventricular dilation at 4 years. Cardiac resonance imaging showed a 50 mm end-diastolic diameter and an ejection fraction of 50%. The patient had profuse sweating with exercise as symptom of heart failure, and was referred to our center.

Valve repair surgery was indicated and planned. Adherences were dissected by median sternotomy and cardiopulmonary bypass was initiated. A transverse aortotomy was performed. The valve was a unicuspid aortic valve reconstructed by tricuspidization. The two commissures of the right leaflet had insufficient height and the effective height of the three leaflets (4) was limited. The right leaflet was hypoplastic.

The valve was bicuspidized. The rudimentary right leaflet was resected and a new commissure was created in front of the left /noncoronary leaflet, connecting the remaining tissue of the left and noncoronary leaflets to the new commissure, with two patches of heterologous pericardium. The leaflets were adjusted achieving an effective height of 9 mm.

Intraoperative echocardiography showed a competent valve with planimetric area of 1.6 cm². The postoperative period was uneventful and the child was transferred to the general ward after two days and

discharged four days later, recovering his usual activities in 4 weeks. A year later he continues to be well.

The echocardiography at one year shows a competent valve with 34 mm end-diastolic and 21 mm end-systolic ventricular diameters and normal ejection fraction (71%). (Figures 1 and 2; Videos 1 and 2)

Treatment of CAS aims to eliminate the transvalvular gradient. After initial surgical or interventional procedure, significant valve regurgitation occasionally appears. Traditionally the only solution to this problem has been valve replacement. Valve reconstruction has not been applied uniformly or successfully, which is not surprising when in most cases the underlying anatomy is unicuspid. This is characterized by commissural and leaflet hypoplasia, (5) an anatomic condition leading to poor hemodynamic function.

Valve reconstruction has an important advantage in restoring hemodynamic valve function; it involves

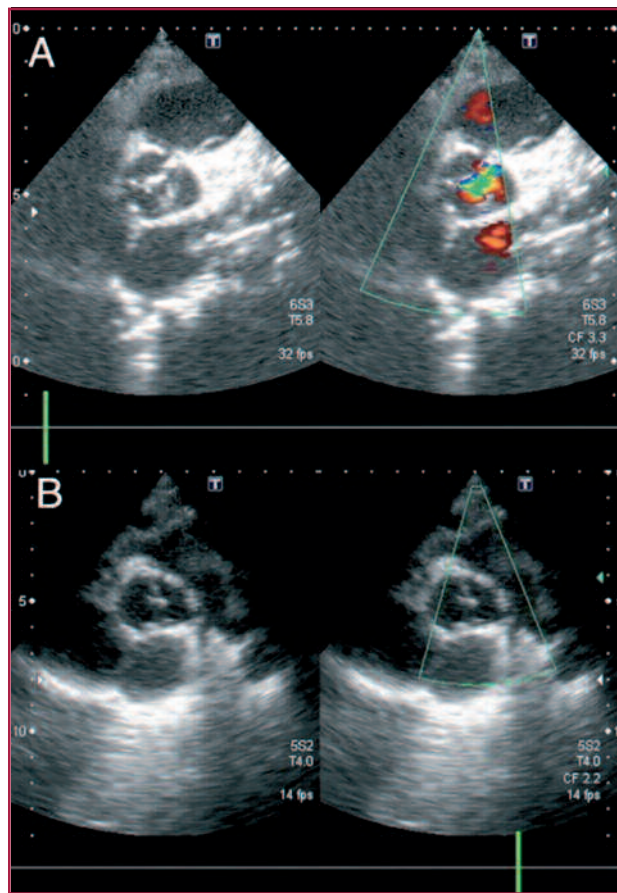


Fig. 1. Transthoracic echocardiography in diastole. **A.** Preoperative. **B.** One-year postoperative control

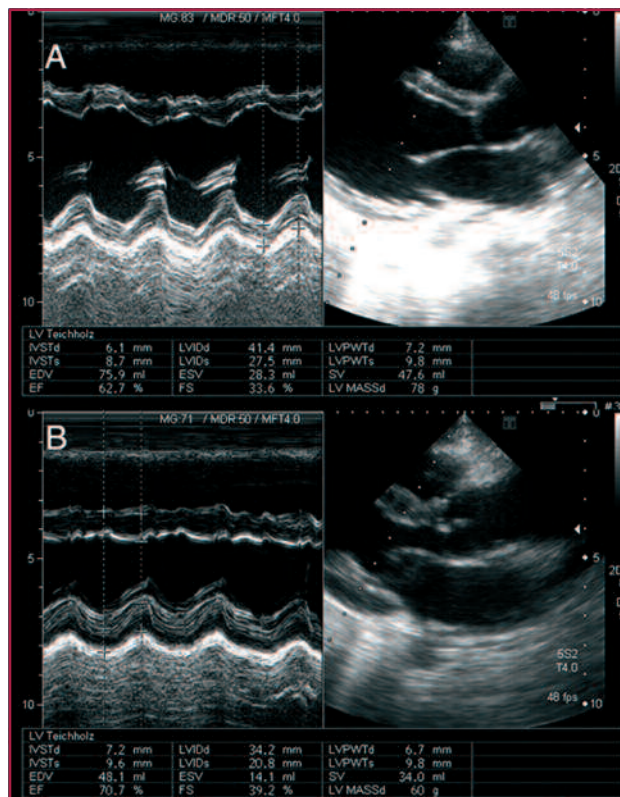


Fig. 2. Transthoracic echocardiographic measurements. **A.** Pre-operative. **B.** One-year postoperative control

a low rate of endocarditis, limiting surgery to the diseased valve and allowing growth of the ventricular outflow tract together with the somatic growth of the patient.

We had already described the concept of “bicuspidization” of unicuspid valves, creating a new commissure with normal height and adding pericardial tissue to replace the lack of native valve tissue (3). We have recently proposed a technical modification that acquires a more symmetrical bicuspid design (6).

The case we report demonstrates that this concept also applies to regurgitant valves due to CAS treatment. The initial functional result is close to that of a normal valve. As with any novel approach mid-term results are still limited and no data on long-term durability is available. Even if it was limited to 5 or 10 years, this would allow the child time to grow before applying a more definitive solution. Furthermore, a new valve reconstruction could be performed if the somatic growth of the child caused the used pericardial patches to become “small”.

The reconstruction of the aortic valve should be the technique of choice if significant aortic regurgitation occurs after CAS surgical or interventional treatment.

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REFERENCES

1. Alsoufi B, Al-Halees Z, Manliot C, McCrindle BW, Al-Ahmadi M, Sallehuddin A, et al. Mechanical valves versus the Ross procedure for aortic valve replacement in children: propensity-adjusted comparison of long-term outcomes. *J Thorac Cardiovasc Surg* 2009;137:362-70. <http://doi.org/bjg5c9>
2. Mokhles MM, Rizopoulos D, Andrinopoulou ER, Bekkers JA, Roos-Hesselink JW, Lesaffre E, et al. Autograft and pulmonary allograft performance in the second post-operative decade after the Ross procedure: insights from the Rotterdam Prospective Cohort Study. *Eur Heart J* 2012;33:2213-24. <http://doi.org/rzj>
3. Schäfers HJ, Aicher D, Riodioncheva S, Lindinger A, Rädle-Hurst T, Langer F, et al. Bicuspidization of the unicuspid aortic valve: a new reconstructive approach. *Ann Thorac Surg* 2008;85:2012-8. <http://doi.org/bq4kq4>
4. Bierbach BO, Aicher D, Issa OA, Bomberg H, Gräber S, Glombitza P, et al. Aortic root and cusp configuration determine aortic valve function. *Eur J Cardiothorac Surg* 2010;38:400-6. <http://doi.org/cp29hp>
5. Anderson RH. Understanding the structure of the unicuspid and unicommissural aortic valve. *J Heart Valve Dis* 2003;12:670-3.
6. Aicher D, Bewarader M, Kindermann M, Abdul-Khalique H, Schäfers HJ. Aortic valve function after bicuspidization of the unicuspid aortic valve. *Ann Thorac Surg* 2013;95:1545-50.

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Aortopulmonary Window Associated with Tetralogy of Fallot

The aortopulmonary window (APW) is a rare congenital malformation which occurs in 1 out of each 100,000 live births, representing 0.15% of all congenital heart defects. It is defined as an abnormal communication between the aorta and the pulmonary artery and / or right pulmonary branch, above the two separate normally formed semilunar valves. (1) It may occur alone, but it is commonly associated with another congenital heart disease. It varies in size and location, resulting in different classifications, those of Robinson and Mori being the most commonly employed at present.

This communication produces a large left - right (L-R) shortcircuit similar to a large patent ductus arteriosus (PDA), leading to congestive heart failure and early development of pulmonary hypertension. Surgical treatment is indicated at diagnosis. The prognosis of untreated patients is poor with 40% mortality in the first year of life. (2)

We describe the case of a male patient, with a diabetic mother, born at term by cesarean section with a birth weight of 3090 g. At 9 days old he is referred to cardiology as a result of a heart murmur. Tetralogy of Fallot is diagnosed and he continues with cardiovascular ambulatory follow-up with atypical evolution, tachypnea, radiological cardiomegaly, dilated left heart chambers and echocardiographic pulmonary hy-