Atrioventricular valve surgery: Restoration of the fibrous skeleton of the heart

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Millions of years of relentless evolution have shaped the human heart into a durable and incredibly efficient pump. This high durability and efficiency are impossible without the normal fibrous skeleton of the heart. To achieve optimal results in our surgical work, we must emulate the work of nature as closely as possible. A perfect example of this would be surgery for the atrioventricular valve (AVV). Although the results of AVV repair have improved over time, AVV regurgitation remains a formidable challenge, particularly, in the univentricular circulation.¹⁻⁷ Although more than 20% of patients with univentricular circulation develop moderate or greater AVV regurgitation of mitral, tricuspid, or common AVV by 20 years, the rate of failure of common AVV in the univentricular circulation is greater than 50% by 20 years.³ Furthermore, it is unequivocal that AVV regurgitation is associated with poorer survival in patients with univentricular circulation.⁵ Achieving a durable repair of AVV, including that in common AVV, is often difficult in univentricular circulation, especially in patients with right ventricular dominance.³ In this focused review, we summarize the recent literature on the outcomes of AVV surgery in patients with univentricular circulation with emphasis on the importance of restoring the fibrous skeleton of the heart.

NORMAL AND MALFORMED FIBROUS SKELETON OF THE HEART

The structurally normal human heart has a well-developed fibrous skeleton (Figure 1, A) that provides support for the myocardium and the valves. The aortic valve is in fibrous continuity with the mitral and tricuspid valves. The confluence of these 3 valves forms the central fibrous body. This structure is malformed in the spectrum of patients with common AVV. Patients with common AVV and balanced ventricles (Figure 1, B) or various degrees of right

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CENTRAL MESSAGE

Restoration of the fibrous skeleton of the heart appears to be crucial for durable repair of common AVV in univentricular circulation.

ventricular hypoplasia (Figure 1, C) still have a normally formed aortic valve. In contrast, in patients with common AVV and hypoplasia of the left ventricle with various degrees of aortic valve hypoplasia, the fibrous skeleton is even less supported (Figure 1, D). The extreme end of the spectrum of fibrous skeleton maldevelopment would occur in patients with aortic atresia with common AVV. Unsurprisingly, it has been demonstrated that right ventricular dominance is a risk factor for death⁸ and AVV insufficiency.³ Repair of AVV in right ventricle dominant univentricular circulation is notoriously difficult. Thus, patients with right ventricle dilatation and AVV insufficiency may be referred to heart transplantation before an attempt of repair is undertaken. However, even patients with poor single ventricular function may have dramatic improvement when the competence of the AVV is restored.⁹ In contrast, patients with tricuspid atresia have well-formed mitral valve and fibrous skeleton of the heart and rarely have AVV insufficiency.³

INCIDENCE OF ATRIOVENTRICULAR VALVE DYSFUNCTION IN PATIENTS WITH SINGLE-VENTRICLE PHYSIOLOGY

It appears that failure of AVV affects approximately 20% of patients with single-ventricle physiology by 20 years of

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FIGURE 1. Schematic of the fibrous skeleton of the heart. Well-developed fibrous skeleton in normal heart (A). A spectrum of maldevelopment of fibrous skeleton of the heart in patients with complete atrioventricular septal defects with balanced ventricles (B), right-sided hypoplasia (C), and left-sided hypoplasia (D), with extreme end of the spectrum represented in aortic atresia.

age.³ Single-ventricle palliation is performed for a broad range of congenital anomalies, and the rate of AVV failure is, predictably, related to the morphology of the AVV and, thus, to the extent to which the fibrous skeleton of the heart is developed.^{3,10} Consequently, patients with tricuspid atresia have the lowest rate of AVV failure, with 8% having valve failure at 25 years, because they have a mitral valve as their AVV. The rate of AVV failure is higher in those with mitral atresia reaching 46% at 25 years. Yet, the highest rate of AVV failure is observed in those with a common AVV, affecting 56% of these patients by 25 years of age.³

OUTCOMES OF UNIVENTRICULAR REPAIR IN PATIENTS WITH COMMON ATRIOVENTRICULAR VALVE

Great progress has been made in repair of balanced complete atrioventricular septal defect (AVSD) in recent years, with operative mortality reported of 1% to 3% and 20-year survival of approximately 90%.¹¹⁻¹⁵ Conversely, little progress has been made in the surgical management of unbalanced AVSD. Early reports suggested that only one-third of patients could survive to Fontan, and only one-half of these patients with Fontan circulation went on to be long-term survivors.^{16,17} Recently, we demonstrated that the outcomes of these patients were better than previously thought, with 60% of patients reaching Fontan

completion and a 25-year survival of 80% in those patients who reached Fontan completion.^{1,18} We have learned from this experience that it is crucial to have AVV competence to achieve a long-term survival in these challenging patients² as transplantation-free survival of those left with moderate or great AVV insufficiency was approximately 30% at 2 years after attempted AVV repair.² In sharp contrast, those who had common AVV replacement had 75% survival at 10 years.² A recently proposed strategy of biventricular recruitment and conversion has been described, but early results appear similar to those achieved with conventional single ventricular palliation, and long-term follow-up is lacking.¹⁹

In patients with balanced ventricles who undergo complete repair, the risk of AVV reoperation is approximately 25% at 20 years.¹¹ Conversely, in patients with unbalanced AVSD, the rate of AVV reoperation is in the range of 40% to 50% at 25 years follow-up. AVV failure in these patients is associated with increased risk of mortality.^{2,20}

RESULTS OF ATRIOVENTRICULAR VALVE SURGERY

Traditional Repair Techniques

In general, the results of reoperation for AVV regurgitation are not durable across the spectrum of AVSDs. In

patients with balanced complete AVSD, the freedom from reoperation is approximately 25% at 20 years.^{11,13} In patients with unbalanced AVSD, the results are even more disappointing with 40% requiring reoperation by 15 years.^{1,4}

Imai and colleagues²¹ reported 28 patients with unbalanced AVSD and univentricular physiology who underwent concomitant AVV repair and Fontan procedure between 1985 and 1998. The techniques used in this series were circular suture annuloplasty and cleft closure, and they reported a decrease in the mean grade of AVVR from 1.8 to 0.6 after repair. This study did not describe early mortality

or long-term survival of patients who underwent repair of their common AVV.

Kotani and colleagues²² reported 66 patients with single-ventricle physiology who underwent AVV repair from 1998 to 2011, of whom 10 patients had common AVV. The mean grade of AVV regurgitation decreased from 2.1 to 1.3 after repair. At 5 years follow-up, freedom from reoperation was 75%, whereas survival was 76%. Significant residual AVV regurgitation on intraoperative echocardiography was associated with increased risk of reoperation.

Laux and colleagues²³ reported 31 patients with single-ventricle physiology who underwent AVV repair between

FIGURE 2. Schematic of common AVV repair techniques aimed at restoration of fibrous skeleton of the heart in univentricular circulation. Septation of the common AVV in univentricular circulation (A) is not feasible; thus, repair is often limited to annuloplasty and edge-to-edge approximation to achieve good central coaptation and central stabilization (B). To achieve durability of the repair, the fibrous skeleton of the heart should be emulated as close as possible, for instance, with a bridging annuloplasty (C). Common AVV repair in patients with left-sided hypoplasia is particularly difficult, partly due to septal deviation and severe malformation of fibrous skeleton of the heart (D). Thus, in addition to annuloplasty and central stabilization (E), the bridging annuloplasty (F) or complete closure of hypoplastic component of the common AVV is often the only way to ensure long-term durability.

1998 and 2014, of whom 32% (10/31) had a common AVV. Using a range of repair techniques including annuloplasty, cleft closure, and edge-to-edge repair, they achieved freedom from AVV reintervention of 62% at 10 years.

Our group²⁴ reported 66 patients with single-ventricle physiology who underwent AVV repair between 1988 and 2010, of whom 18% (14/66) had a common AVV. Common AVV was a risk factor for reoperation, with freedom from reoperation of only 30% at 10 years.

Nakata and colleagues²⁵ reported 65 patients with single-ventricle physiology who underwent AVV repair between 1999 to 2008, of whom 54% (35/65) had common AVV. Freedom from reoperation on the AVV was 57% at 5 years.

Closure of the Hypoplastic Component of the Common Atrioventricular Valve

In patients with significant imbalance of the ventricles, closure of the hypoplastic component of the valve may restore competence without compromising the valve orifice. This may be achieved by suture approximation of the leaflets or use of a patch. Our group²⁶ reported 38 patients with Fontan who underwent closure of the hypoplastic component of common AVV between 1975 and 2018 from a binational registry. Freedom from reoperation was 83% at 18 years.

Restoration of the Fibrous Skeleton of the Heart

In an attempt to address the high rate of failure observed after repair of common AVV in the univentricular circulation, several groups have described using bridging annuloplasty (Figure 2) in these patients.²⁷⁻³⁰ In this technique, a strip of synthetic material or pericardium is placed transversely across the common valve annulus above the ventricular septum and sutured to the annulus, with the aim of reducing the anteroposterior dimension of the annulus and replicating the normal fibrous skeleton.²⁷⁻³⁰ Although there have only been a small number of studies so far, results appear promising.

Misumi and colleagues³¹ reported 38 patients who underwent common AVV surgery between 1995 and 2012, of whom 27 underwent repair using the bridging technique. They demonstrated a 15-year freedom from reoperation of 45.3%, which was similar to other repair techniques, although with a small number of patients. Furthermore, they found the technique to be reproducible, and they did not need to replace any valves after introduction of the technique.

He and colleagues³² reported 37 patients who underwent repair of common AVV between 2007 and 2018. The bridge technique was used in 51% (19/37) of patients. Use of the bridge technique was associated with superior freedom from common AVV failure at 10-year follow-up (62% vs 35%, $P = .01$).

CONCLUSIONS

Common AVV regurgitation is a challenging problem, especially in the setting of univentricular physiology. Although current repair techniques have a high rate of long-term failure, the ideal technique, which is yet to be determined, should emulate the normal fibrous skeleton of the heart as closely as possible.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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