Management of the bad atrioventricular valve in Fontan…time for a change

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Central Message

Bad atrioventricular valve function in Fontan circulation results in bad outcome. A re-examination of the *timing* of intervention and best *procedure* is needed.

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See Commentary on page 1649.

Successful long-term outcome following Fontan procedure depends on numerous factors, including normal ventricular systolic and diastolic function, adequate pulmonary artery size and distribution, low pulmonary artery pressures and resistance, sinus rhythm, and a *competent* atrioventricular valve.

The recent paper by King and colleagues[1] in the *Journal of the American College of Cardiology*, commented on by Driscoll and Cetta,[2] highlights a frequently encountered conundrum for the surgeon: valve disease in single-ventricle patients palliated by Fontan circulation. The study details what many surgeons have anecdotally experienced: a functional (competent) systemic atrioventricular valve (SAVV) is a key component to a successful Fontan circulation, and a durable, effective valve repair in these patients is very challenging. The study by King and colleagues[1] provides sobering data regarding the frequency of this problem and its clinical implications. In their cohort, atrioventricular valve failure more than doubled the rate of Fontan failure, with two-thirds of patients with a common atrioventricular valve and one-half with a single tricuspid valve experiencing valve failure by 30 years of age.[1]

Many unanswered questions remain in this regard: at what point along the single-ventricle palliation pathway should valve repair be undertaken? When is it permissible to perform valve repair at the time of Fontan procedure and when should it be staged in advance of Fontan? Are there certain valvuloplasty techniques that yield better outcomes? Which valves are beyond salvage and require replacement, and when should heart transplantation be considered?

The aim of this Expert Opinion is to challenge current thinking and provoke a mindset change as to how to best approach SAVV regurgitation in the single-ventricle patient. The important questions include: what is the valve morphology and the mechanism(s) of regurgitation; what is the ventricular morphology and function; what is the best time to intervene along the palliative single-ventricle pathway; and what procedure is best to apply? Based on these issues, a proposed management algorithm for valve intervention is provided.

What's Good

There are a select group of patients with Fontan circulation in whom SAVV anatomy and repair is ideal and preferred: the morphologic left ventricle with a morphologic mitral valve. As our experience has grown in adult mitral valve repair, select single (left) ventricle patients with mitral valve anatomy and dominant, well-functioning left ventricles are the subgroup that is best served with valve repair. This population has the advantage of a systemic left ventricle and valve morphology designed to withstand systemic output. As established in adult cardiac surgical patients, the etiology of regurgitation can be characterized by the Carpentier classification[3] and repair tailored to the underlying mechanism(s). Although isolated annular dilation (Carpentier Type I) can be treated with a variety of partial bands or complete annuloplasty rings in adults and older children, in younger children with somatic growth remaining, partial bands or rings may not be an option. In this

younger age bracket, alternative annuloplasty techniques that decrease the annular dimension while still allowing for somatic growth, such as the Wooler (eccentric annuloplasty) technique may be considered, or a partial annuloplasty band made from a variety of materials. There also has been interest in biodegradable annuloplasty bands that would allow for somatic growth.[4] Children may have normal leaflet motion (Carpentier Type I) with regurgitation secondary to a cleft, in which simple cleft closure may result in a competent valve. In cases of prolapse (Carpentier Type II), a variety of techniques have been used in adults that include triangular or quadrangular resection, chordal transfer, or artificial Gore-Tex (''neo-chords'') for posterior and/or anterior leaflet prolapse or flail.[5,6] The Alfieri stitch (ie, ''edge-to-edge'' repair) has also been used for central regurgitation or segmental prolapse of the anterior or posterior leaflet. In adults, such repairs are accompanied by placement of annuloplasty rings for ''stabilization.''

In patients with congenital heart disease, the mechanism of regurgitation is often multifactorial with abnormalities of the leaflet(s) and/or subvalvar apparatus, in combination with ventricular dilation. As a result, a combination of repair techniques is often required. In children, similar techniques as those detailed previously have also been employed and in selected situations chordal shortening has been used. Patients with restricted leaflet motion (Carpentier Type III) are more challenging. Depending on the cause of the restriction, leaflet augmentation can be employed to allow more area for coaptation or chordal elongation used in cases of chordal restriction. Studies on the results of mitral valve repair in children are largely limited to single-center series often with a range of underlying etiologies. In a group of 20 patients with congenital mitral disease operated on at <1 year of age, 10 with mitral regurgitation, Uva and colleagues[7] reported an actuarial freedom from reoperation of 58% at 7 years. Zias and colleagues[8] reported on a series of 26 mitral patients including 21 patients with mitral regurgitation and found at a mean follow-up of 31 months 95% of patients demonstrated improvement with 15% needing early re-repair and 1 patient requiring a valve replacement. In a study of 79 largely older children with mitral disease, 74 with regurgitation, and a wide range of etiologies including atrioventricular septal defects, acquired disease, and single-ventricle patients, Aharon and collagues[9] reported an actuarial survival of 82% at 5 years and freedom from reoperation of 89% at 8 years. Ohno and colleagues[10] reported on 49 patients with mitral regurgitation and found freedom from reoperation to be 86% at 13 years. Although the majority of these studies focus on mitral (or left atrioventricular valve) repair in the setting of a biventricular circulation, survival and durability can serve as a reference point and would be presumably better than in a single-ventricle circulation.

What's Bad

Although valvuloplasty techniques for the *common atrioventricular valve* in atrioventricular septal defect have improved with time, this group continues to be challenging. A myriad of different approaches has been reported for the various pathologies of these valves ranging from leaflet resection or augmentation, artificial chordae, cleft (''zone of apposition'') closure, and suture or banded annuloplasty techniques with a 85% to 90% 10-year freedom from reoperation in patients with a biventricular circulation.[11,12] Those who do require reintervention following initial atrioventricular septal defect repair demonstrate disappointing durability even at experienced centers, with Stulak and colleagues[13] reporting a 48% 10-year freedom from reoperation after first reoperation.

In the single-ventricle population, the presence of a common atrioventricular valve has been identified as a risk factor for mortality after the bidirectional cavopulmonary shunt[14] and Fontan[15] and are less likely to reach Fontan completion.[16] In single-ventricle patients with common atrioventricular valve pathology, valve repair may be considered for significant regurgitation but certainly does not ensure improved outcomes with frequent repair failure. In a study by Buratto and colleagues[17] of unbalanced atrioventricular septal defects undergoing single-ventricle palliation, 32% needed valve repair and had an early mortality of 18%, and 32% needed further valve intervention. King and colleagues[18] reported on 114 patients with Fontan circulation with common atrioventricular valves and found that 28 patients required valve intervention, and 24 of those patients underwent repair at the first intervention. Of the patients who underwent repair, 67% experienced valve repair failure that was defined as the need for reintervention for moderate or more regurgitation.[18] The 4-year freedom from repair failure was a disappointing 50%.[18] Similarly in the study of King and colleagues,[1] common atrioventricular valve was a factor for valve repair failure on univariate analysis, with 68% failing as compared with 14% of mitral valve repairs. Ota and colleagues[19] studied valve repair in single-ventricle patients with heterotaxia presenting with moderate-to-severe or greater regurgitation and found 44% had a durable initial repair with a median follow-up of 6.2 years. Analysis of valve repair techniques used revealed that leaflet apposition techniques yielded improved freedom from significant regurgitation at most recent follow-up.[19]

What's Ugly

A subset of the single-ventricle population has atrioventricular valve pathology that is beyond salvage. As identified by King and colleagues[1] as an independent predictor of atrioventricular valve failure and valve intervention, a dominant right ventricle with a *morphologic tricuspid valve*

has also been identified as a risk factor for Fontan failure in other studies.[20] One half of patients with a single tricuspid valve in the series of King and colleauges[1] experienced valve failure by 30 years. Tricuspid valve pathologies have proven to be challenging to provide a durable repair even in a biventricular circulation with good baseline systolic function. This may be due in part to the absence of the normal crescent shape of the right ventricle that is lost with right ventricular dilation. Consequently, it is not surprising that tricuspid valve pathologies with underlying right ventricular morphology tasked to withstand systemic pressure are frequently the most challenging atrioventricular valves in the single-ventricle population. Durability of tricuspid repair in patients with congenitally corrected transposition (ie, morphologic tricuspid valve and morphologic right ventricle in the systemic circulation) has also been poor, raising the issue of preferential tricuspid replacement over repair.[21] In addition, the morphology of the tricuspid valve in some patients with hypoplastic left heart syndrome is significantly altered, with 12% having a bileaflet tricuspid valve and more than one third having some evidence of leaflet dysplasia.[22]

Valvuloplasty techniques employed in tricuspid valves are similar to those previously discussed and can include purse-string or banded annuloplasty, eccentric annuloplasty and commissuroplasty, closure of clefts, edge-to-edge repair, artificial chords or chordal shortening, and patch augmentation of deficient leaflet(s).[23] Studies have shown a durable tricuspid repair to be challenging in the single-ventricle population. For instance, in the study by Ohye and colleauges,[24] 63% of patients who underwent valve repair had moderate regurgitation at a median follow-up of 26 months. However, when a successful repair can be achieved with preserved ventricular function, Honjo and colleagues[25] showed that such patients had equivalent survival as the control group. Independent predictors of death or transplant in that study included indexed valve annulus diameter (which highly correlated with ventricular dilation) and decreased ventricular function post-repair.[25] In contrast, 75% of patients in that study who had moderate or greater reduced ventricular function on post-repair transesophageal echocardiogram died or underwent transplant.[25] Similarly, Sugiura and colleagues[26] demonstrated that patients who develop ''early'' tricuspid regurgitation (around time of Norwood Stage I palliation) had substantially worse prognosis than those who developed late tricuspid regurgitation (5-year survival of 43% vs 93%, $P = .003$); however, performing tricuspid repair significantly improved survival compared with those who did not have a valve intervention (52% vs 23% at 5 years, $P = .043$). Surgery improved the degree of regurgitation and diastolic diameter but did not improve ventricular systolic function.[26] Some evidence suggests that those patients who require valve reintervention have a poor prognosis. Ohye and colleague[24] reported on a subset of patients with an early successful tricuspid repair who subsequently developed late valve failure. That subset of patients had poor ventricular function and the survival of that subgroup was only 25%. In contrast, in the Boston Children's experience, 48% of patients required tricuspid valve re-repair and freedom from transplant or tricuspid valve replacement was 97% with a median follow-up of 38 months.[27]

Despite a concerted effort by surgeons to preserve native valves with valvuloplasty in almost all circumstances, there are some valve pathologies that are simply beyond repair. A recent study of *atrioventricular replacement* among Japanese single-ventricle patients reported poor outcomes, with 20 of the 56 patients dying within a median 5.0-year follow-up and an almost 50% mortality among the 11 patients who required redo valve replacements.[28] In a relatively large study by Menon and colleagues[28] from the Mayo Clinic, 61 patients with a median age of 14 years underwent atrioventricular valve repair or replacement for right, left, and common atrioventricular valves following Fontan procedure. The median duration between Fontan and atrioventricular valve surgery was 4.7 years and median follow-up was 9 years. There were 32 (52%) deaths, with 8 (13%) dying within 30 days of surgery. The 5-, 10-, and 15-year survival rates were 67%, 57%, and 45%, respectively. At late follow-up, 44 of 61 (72%) had arrhythmias, 21 of 29 (72%) were symptomatic, and 12 of 61 (20%) developed protein-losing enteropathy.[28] Independent risk factors for mortality were reduced ventricular function and development of protein-losing enteropathy.[28]

Studies of mitral valve replacement in children with a biventricular circulation have also demonstrated poor results, with early mortality rates that range from 10% to 20%.[29,30] Alsoufi and colleagues[30] examined outcome in a very young group of 79 patients (median age 24 months) and on competing risk analysis at 10 years, mortality was 40% without repeat mitral valve replacement, 20% with repeat mitral valve replacement, and 40% were alive without need for reoperation. In the recent Pediatric Cardiac Care Consortium study that examined a large number of children (n = 441; median age 4.3 years), transplant-free survival at 20 years was 76%.[29]

Time for a Change

Given the disappointing results of survival and durability of valve repair and replacement in the single-ventricle setting, *earlier* attention for valve intervention—as early as mild-moderate regurgitation develops—should be considered, particularly when there is ventricular dilation or any degree of systolic dysfunction. In the pediatric population, symptoms can be hard to assess and are often absent, and an emphasis is placed on preserving ventricular function, which may entail earlier intervention before significant ventricular dilation/dysfunction and/or organ

dysfunction that may lead to symptoms. Therefore, in practice, the timing of intervention is highly dependent on imaging, particularly the degree of valvular regurgitation and/or stenosis and ventricular function/dilation. That said, high or increasing doses of diuretics and afterload-reducing agents in patients are warning signs that an intervention should be performed.

In general, all would agree that an initial attempt at valve repair is reasonable and preferred, regardless of valve morphology and particularly in a child. The issue and controversy surround the need for a second valve intervention when the initial valvuloplasty fails. It is at this time point when stronger consideration should be given to valve replacement, particularly if a second valvuloplasty is suboptimal. The importance of a dilated or dysfunctional systemic ventricle to successfully remodel cannot be overemphasized if candidacy for Fontan is the goal. In an older patient with a functioning Fontan circulation, greater consideration should be made toward valve replacement, particularly when valve anatomy is not a morphologic mitral valve, to preserve and maintain ventricular size and function. General principles to be contemplated are outlined to follow, and a management strategy is summarized in Table 1.

Goal.

1. In general, Fontan completion should consist of just (only) that, "Fontan completion." When other significant abnormalities are present, eg, pulmonary artery stenosis, systemic ventricular outflow tract obstruction, and atrioventricular valve regurgitation that will require attention, strong consideration should be given to staging to Fontan and correcting the abnormality(s) *prior to* Fontan. Of these abnormalities, atrioventricular valve regurgitation is the most concerning, probably because it adversely affects ventricular size and function. For these reasons, intervention before Fontan seems most prudent if we want to optimize the success of the eventual Fontan circulation.

Timing and threshold for intervention.

2. The *timing* of valve intervention is critical to optimize longer valve durability and preserve (or allow recovery) of ventricular function. In general, we advise operation when moderate atrioventricular valve regurgitation is present, particularly when there is annular or ventricular dilation, ie, we stage at bidirectional cavopulmonary shunt, or in between cavopulmonary shunt and Fontan but *before* Fontan completion. Although some[31] choose to leave moderate regurgitation alone at the time of bidirectional cavopulmonary shunt because improvement in regurgitation is noted in some patients, we believe the better course of action is to be proactive. Our approach is to routinely repair the valve for moderate regurgitation, given the poor results that are now well documented in the literature and outlined previously.

TABLE 1. Strategy for management of the bad atrioventricular valve in the single-ventricle patient*

- Mechanism of mitral morphology
  - Structural – chordal, cleft, leaflet (minor) → repair > replace
    marked dysplasia → replace > repair
  - Functional – annular/ventricular dilation → repair ≈ replace
- Mechanism of regurgitation for tricuspid and common atrioventricular valve morphology
  - Structural – chordal, cleft, leaflet (minor) → repair ≈ replace
    marked dysplasia → replace > repair
  - Functional – annular/ventricular dilation → replace ≈ repair
- Valve morphology
  - mitral → repair > replace
  - tricuspid → replace ≈ repair
  - common → replace ≈ repair
- Ventricular size/function
  - normal/normal → repair > replace
  - RV (dominant) dilation/normal → replace ≈ repair
  - LV (dominant) dilation/normal → repair ≈ replace
  - Univentricular, unbalanced normal/normal → replace > repair
  - Dilation/reduced function → replace vs Tx

RV, Right ventricular; *LV*, left ventricular; *Tx*, transplant. *Strategies are general guidelines to consider and ultimate decision-making is individualized depending on the patient and clinical situation. In a child, an initial valvuloplasty is preferred regardless of valve pathology; however, stronger consideration to replacement is given when initial valve repair fails.

3. If moderate (functional) regurgitation develops after the bidirectional cavopulmonary shunt and is present at the time Fontan completion is being considered, we would delay Fontan and stage valve repair.
4. A dilated ventricle with moderate or more regurgitation and a structurally normal valve (ie, functional regurgitation) is the most concerning, and caution is advised before proceeding to Fontan completion. In some settings, the elimination of a volume loading lesion, eg, a systemic-to-pulmonary artery (Blalock–Taussig shunt) shunt, can be expected to result in favorable ventricular remodeling (reduction in ventricular dilation), but our threshold to perform concomitant valve repair at the time of Blalock–Taussig shunt takedown and bidirectional cavopulmonary shunt is low to optimize/preserve ventricular remodeling.
5. Valvuloplasty at the time of Fontan completion can be considered under the following circumstances: moderate regurgitation with a morphologic left ventricle that is not dilated and has preserved function with a *structural* abnormality with the mitral valve, eg, prolapse or cleft.

Techniques and morphology considerations.

6. Improved valvuloplasty *techniques* for acquired mitral and tricuspid valve disease and in selected subgroups of congenital heart disease, eg, Ebstein anomaly, have demonstrated improved durability and augment the surgical armamentarium; some of these techniques can and should be applied in the setting of congenital heart

disease. Importantly, many of these valvuloplasty techniques involve attention to abnormalities of the *subvalvar* apparatus in addition to leaflet and annular maneuvers that are most commonly used.

7. Morphologic tricuspid and common atrioventricular valves are the most difficult to achieve a sustainable long-term result. It is reasonable to begin with an initial valvuloplasty. However, if initial valvuloplasty fails, a lower threshold to proceed with valve replacement at the subsequent operation should be strongly considered. Although mechanical valve replacement and the concomitant need for warfarin anticoagulation are not an ideal alternative,[29,30] the dismal results documented with a "bad atrioventricular valve, bad Fontan circulation" is precisely the reason it should be considered more seriously…hence, a different mindset to address this problem.

Transplant pathway.

8. Severe regurgitation with severe ventricular dilation *and* dysfunction should undergo transplant treatment pathway.
9. If valve replacement is deemed inappropriate because of inability to safely use warfarin anticoagulation or the risk of the valve operation is felt to be prohibitive, then the patient should undergo transplant pathway.

SUMMARY

Successful long-term outcome following Fontan procedure depends largely on a competent atrioventricular valve. Contemporary valvuloplasty techniques have evolved for different pathologies and have improved durability. The *timing* of and threshold for valve repair may be the most important factor to be considered, with earlier intervention being applied for lesser degrees of regurgitation and before the Fontan procedure is completed. In selected situations, valve replacement should be more strongly considered.

Conflict of Interest Statement

Authors have nothing to disclose with regard to commercial support.

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