Truncal valve repair in children

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Truncal valve insufficiency presents significant short- and long-term challenges for congenital cardiac surgeons. Occurring in up to 43% of patients with truncus arteriosus, truncal valve insufficiency is one of the most important factors influencing the outcomes in these patients.¹⁻⁹ Although an array of surgical techniques has been described, consistently durable results have been difficult to achieve. This is a testament to the fact that the ideal repair technique is yet to be determined. The purpose of this focused review is to highlight recent key publications on the topic.

IMPACT OF TRUNCAL VALVE INSUFFICIENCY ON SURGICAL OUTCOMES

Mild truncal valve insufficiency is often well tolerated, nonprogressive in the long-term, and does not necessitate truncal valve surgery.^{1,5,7-13} However, moderate or greater truncal valve insufficiency has been associated with greater mortality and often requires truncal valve surgery.^{1,3,7,8,13-17} Several studies have shown that moderate-to-severe truncal valve insufficiency is a risk factor for early truncal valve replacement,⁶ early mortality, and, generally, poorer long-term outcomes if not adequately addressed during the initial operation.^{3,6,13-17} Russell and colleagues³ reported 23 truncal valve operations in 572 patients operated between 2000 and 2009, concluding that failure to address significant truncal valve insufficiency was associated with poor outcomes. Conversely, Tlaskal and colleagues¹⁰ determined that persistent moderate insufficiency is usually well tolerated and does not lead to an increase in early mortality, although it was associated with the need for eventual truncal valve replacement. We have recently reported a 20-year overall survival of 79% in children with no truncal valve insufficiency at the time of initial surgery, 77% with mild insufficiency, 77% with moderate insufficiency, but only 22% at 10 years in those with severe insufficiency.⁹

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CENTRAL MESSAGE

It appears that reduction of the annulus is essential to improve the durability of truncal valve repair.

See Commentaries on pages 1343 and 1345.

TECHNIQUES OF TRUNCAL VALVE REPAIR

A number of surgical techniques to repair the truncal valve have been described. Rarely, the truncal valve may be bicuspid and stenotic, requiring commissurotomy.¹ However, truncal valve insufficiency is more commonly encountered and has been addressed by suturing together partially developed commissures, resuspension of leaflets, resection of redundant portion of leaflets, annuloplasty of commissures and pericardial leaflet extension, and tricuspidization of a quadricuspid valve. Each of these techniques has been reported with varying success.^{3,5,6,18} As the truncal annulus is often large, annular reduction appears to be an efficient way to deal with truncal valve incompetence. Such a reduction of the truncal valve annulus could be achieved by sub-commissural (Figure 1, A and A1) or circular suture annuloplasty with reduction of sinotubular junction diameter (Figure 1, A1) or by resection of part of the truncal root with complete (Figure 1, B and B1) or partial resection of the adjacent cusp (Figure 1, C and C1). The resection technique was first described in 1999 by Roger Mee's group¹⁹ and appears to be effective in neonates and infants with severe truncal valve insufficiency,^{2,8,20} and is more applicable to quadricuspid truncal valves. Although truncal valve repair is generally not durable in the long-term,¹ it appears that tricuspidization of the truncal valve with⁸ or without²¹ resection of a segment of the truncal root and annular reduction provides the most durable results. Most children who require truncal valve surgery have a quadricuspid truncal valve.^{1,4,8} A detailed video demonstrating the technique of tricuspidization of quadricuspid truncal valve with annular reduction in a neonate has been

FIGURE 1. Techniques for tricuspidization and annulus reduction in the quadricuspid truncal valve. Tricuspidization can be achieved by suture annuloplasty of the truncal root (A) and approximation of the prolapsing cusp to the adjacent cusp (A1) with obliteration of interleaflet triangles. Alternatively, a cusp can be completely (B) or partially (C) resected, with the root reapproximated (B1 and C1). This achieves both tricuspidization and annular reduction.

previously published.² Because approximately one third of children with truncus arteriosus have quadricuspid truncal valve^{9,18} and approximately one third of children with a quadricuspid valve require valve surgery,⁸ this is a useful technique in armamentarium of congenital heart surgeon.

Unsurprisingly, concomitant truncal valve surgery is more challenging, as it is often required in the youngest and most critically ill children.⁹ However, it is important to repair the truncal valve at this stage, as not addressing severe truncal valve insufficiency has been associated with poor outcomes.^{1,3,13-17}

Children with a quadricuspid truncal valve are more likely to undergo truncal valve surgery than those with any other valve morphology.¹ We have previously reported that patients with a quadricuspid truncal valve and moderate or greater truncal insufficiency were most likely to require truncal valve surgery¹ and described a tricuspidization technique in a low birth weight neonate in detail.² In contrast, most patients with a quadricuspid truncal valve and mild or less truncal valve insufficiency are free from truncal valve surgery in the long term. Only about 16% of patients with mild or less insufficiency have progressive truncal valve regurgitation, which may require truncal valve surgery.^{8,9} The preferred method of repair in those with moderate or greater insufficiency that appears to give best long-term results is tricuspidization of the quadricuspid valve with reduction of the annulus.^{8,9,21} In children with significant truncal valve insufficiency, the large diameter

of the annulus and relative deficiency of valvar tissue to cover truncal orifice often necessitates reduction of the truncal valve annulus. This can be achieved by resection of a cusp extending onto the annulus.^{2,19} Such resection leading to tricuspidization appears to be more durable than any other methods of repair.⁸ Tricuspidization with base-preserving cusp extension with autologous pericardium in infants has been recently described,²² yet long-term outcomes of such a repair remain to be seen. The importance of correcting significant truncal valve insufficiency cannot be overemphasized. In a contemporary multicenter study, 20% of children who underwent repair of truncus arteriosus experienced major adverse cardiac events.²³ However, such events did not occur more frequently in children, who underwent concomitant surgical intervention on the truncal valves. In contrast, residual significant truncal valve insufficiency, which required an early reoperation, had uniformly poor outcomes.⁴ Furthermore, children with significant truncal valve insufficiency and interrupted aortic arch have particularly poor outcomes.^{4,24}

RESULTS

Generally, repair of truncal valve is associated with poor durability (Table 1).^{5,21} Kaza and colleagues⁵ in 2010 showed that in 14 patients who underwent truncal valve repair by leaflet approximation and suture annuloplasty, freedom from truncal valve reintervention was 70% at 5 years and 50% at 7 years. We have recently reported a

TABLE 1. Outcomes of concomitant repair of truncus arteriosus and truncal valve in neonates and infants

Study	Study period	n	Median age	Repair	Replacement	Follow-up (mean y)	Early deaths	Late deaths	Truncal valve reoperations
Black, 2002 ¹³	1996-1998	5	Neonates	80% (4/5)	20% (1/5)	NR	0%	20% (1/5)	NR
Henaine et al, 2008 ⁶	1986-2003	9	35 d	67% (6/9)	33% (3/9)	8.1	44% (4/9)	20% (1/5)	60% (3/5)
Kaza et al, 2010 ⁵	1995-2008	17	24 d	100% (17/17)	0%	2	0%	6% (1/17)	29% (5/17)
Russell et al, 2012 ³	2000-2009	23	12 d	96% (22/23)	4% (1/23)	NR	26% (6/23)	NR	NR
Russell et al, 2012 ⁴	1979-2011	5	8 d	100% (5/5)	0%	5.5	40% (2/5)	0%	33% (1/3)
Myers et al, 2013 ²¹	1997-2012	13	NR	100% (11/11)	0%	3.2	27% (3/11)	NR	88% (7/8)
Naimo et al, 2018 ¹	1979-2016	21	52 d	95% (20/21)	5% (1/21)	18	19% (4/21)	11% (2/19)	68% (13/19)

NR, Not reported.

freedom from truncal valve reoperation of 65% at 5 years, 51% at 10 years, and 26% at 20 years after concomitant truncal valve repair.¹ Concomitant truncal valve surgery has been shown to be a risk factor for late truncal valve surgery.⁷

We have previously reported that most patients (57%, 12/21) who required concomitant truncal valve repair had a quadricuspid valve.¹ Furthermore, most (79%, 15/19) patients who had moderate and severe truncal valve insufficiency had a quadricuspid truncal valve.¹ Freedom from truncal valve reoperation was 64% at 10 years after tricuspidization of the quadricuspid truncal valve.⁸ In fact, freedom from truncal valve reoperation after tricuspidization was superior to truncal valve replacement in children younger than 6 years of age in whom a mechanical prosthesis may not be feasible.⁸ Importantly, freedom from reoperation in patients who underwent concomitant tricuspidization was 64% at 10 years compared with 0% at 6 years in patients who underwent concomitant truncal valve repair by nontricuspidization.⁸ Similarly, Myers and colleagues²¹ reported tricuspidization methods of truncal valve repair tended to improve freedom from reoperation on the truncal valve.

Replacement with a large prosthesis may not be feasible in younger children, and avoidance of a mechanical prosthesis may obviate the need for life-long anticoagulation. Thus, tricuspidization of the quadricuspid truncal valve is desirable whenever possible. While the durability of repair remains questionable, it is preferable over replacement, particularly during the neonatal period.

CONCLUSIONS

Concomitant truncal valve surgery is most commonly required for quadricuspid truncal valve. Overall, truncal valve repair is not durable. It appears that tricuspidization and annular reduction with complete or partial resection of one cusp give most durable result.

Conflict of Interest Statement

The authors reported no conflicts of interest.

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Tricuspidization

1. Total cusp resection and annulus reduction

2. Cusp reconstruction and annulus reduction

3. Cusp reconstruction

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