Surgery for Truncus Arteriosus: Contemporary Practice

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Surgery for truncus arteriosus has an early mortality of 3% to 20%, with a long-term survival of approximately 75% at 20 years. Nowadays, truncus arteriosus repair is mostly done in the neonatal period together with a single-staged repair of concomitant cardiovascular anomalies. There are many challenging subgroups of patients with truncus arteriosus, including those with clinically significant truncal valve insufficiency, an interrupted aortic arch, or a coronary artery anomaly. In fact, truncal valve competency appears to be the most important factor influencing the outcomes after truncus arteriosus repair. The use of a conduit during truncus arteriosus repair invariably requires reoperation on the right ventricular outflow tract. Through improvements in perioperative techniques over time, many children are now living well into adulthood after repair of truncus arteriosus, albeit with a high rate of reoperation. Despite this, the long-term outcomes of truncus arteriosus repair are good, with many patients being asymptomatic and with a quality of life comparable to the general population.

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Truncus arteriosus (TA) was first reported in an autopsy case by Wilson[1] in 1798, and the anatomical details were later described by Buchanan[2] in a 6-month-old infant. Although TA has an incidence of 3 to 10 per 100,000 live births, it accounts for 4% of all critical congenital cardiac anomalies.[3,4] Despite improvement in perioperative management of TA, the reported operative mortality during the last decade remains between 3% and 20%.[5-16] The challenging patients with TA are those with truncal valve (TV) insufficiency, concomitant interrupted aortic arch (IAA), or anomalous coronary anatomy. The optimal methods of reconstruction of the right ventricular-to-pulmonary artery connection that could minimize the operation rate is yet to be found. We reviewed current outcomes of TA repair, focusing on evolving strategies to decrease mortality and reoperation rates.

Methods

A structured review of the literature was performed using PubMed and MEDLINE databases. The search strategy involved the terms "truncus arteriosus" or "common arterial trunk" or "truncal valve" in the title or keywords. We mostly selected publications from the last 20 years; however, we did not exclude commonly referenced and highly regarded older publications. We also searched the reference list of articles identified by this search strategy and selected those we judged relevant. The last search was conducted in January 2020.

Current Outcomes of TA Repair

Early mortality for TA repair is 3% to 20%, depending on the perioperative status and the presence of concomitant anomalies.[5-17] Long-term survival after TA repair has been reported to be approximately 75% at 20 years.[5-19] Most deaths appear to occur within the first year after repair.[13,16,17,20-24] Rajasinghe and colleagues[22] reported that 57% (13 of 23) of their late deaths occurred within the first year after repair. Similarly, Tlaskal and colleagues[13] reported that 88% (7 of 8) of late deaths occurred within the first year.

We recently demonstrated that patients who survive to 1 year after TA repair have excellent outcomes, with 92.5% survival at 20 years.[17] The mortality after TA repair is summarized in Table 1.[5-16,21-23,25-30] Nowadays, it is well accepted that complete surgical repair of TA is done within the first weeks of life.[5,13,15,16,31-33] Although it is not uncommon to operate within the first days of life, these neonates may be at higher individual risk because they may require emergent surgery due to clinically significant TV insufficiency or concomitant anomalies and thus be in a critical state.

Most patients require reoperation throughout their lives because a conduit is often used to reconstruct their right ventricular outflow tract (RVOT). We have previously reported a freedom from reoperation of 3% at 20 years with median time to reoperation of 4 years.[16] Of our 171 patients, 62% (106 of 171) have required at least 1 RVOT reoperation. The optimal method of RVOT

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Table 1. Outcomes of Truncus Arteriosus Repair

Study, First Author	Year	Study Period	Patients (n)	Age at Repair (median, d)	Early Mortality, % (n/N)	Overall Survival, %	Follow-up (mean, y)
Jahangiri[6]	2000	1992-1998	50	14	4.0 (2/50)	96 at 3 y	1.9[b]
Schrieber[28]	2000	1976-1998	106	134	17.9 (19/106)	61 at 10 y	6.0
Alexiou[7]	2001	1974-1994	23	51	17.4 (4/23)	79 at 10 y	14.8
Brown[30]	2001	1978-2000	60	76[a]	16.7 (10/60)	82 at 10 y	9.4
Danton[9]	2001	1988-2000	61	34	13.1 (8/61)	73 at 5 y	4.2
Thompson[10]	2001	1992-1999	65	10	4.6 (3/65)	92 at 3 y	2.7[b]
Kalavrouziotis[18]	2006	1993-2005	29	28	3.4 (1/29)	93 at 6 y	6.2
Henaine[11]	2008	1986-2003	153	35	17.0 (26/153)	79 at 10 y	8.1
Raisky[12]	2009	2000-2006	32	74[a]	15.6 (5/32)	76 at 5 y	3.3
Hawkins[19]	2010	1999-2008	42	31	9.5 (4/42)	82 at 5 y	5.0
Tlaskal[13]	2010	1981-2009	83	54	22.9 (19/83)	75 at 10 y	10.9
Vohra[29]	2010	1964-2008	32	14	9.4 (3/32)	91 at 20 y	24.5[b]
Xu[14]	2010	2000-2006	23	350	8.7 (2/23)	91 at 5 y	4.1
Buckley[21]	2019	2009-2016	216	10	6.9 (15/216)	87 at 5 y	2.9[b]
Ivanov[26]	2019	1997-2017	97	32	17.5 (17/97)	68 at 10 y	2.5[b]

[a]Denotes mean age at repair; [b]Denotes median follow-up time.

reconstruction has yet to be found, as will be discussed in more detail later.

Despite a high rate of reoperation, the long-term functional state of patients after TA repair is good. Most patients are in New York Heart Association Functional Class I or II, and most patients are reported to have mild or less TV insufficiency.

TV Surgery

TV insufficiency is one of the most important factors influencing outcomes of patients with TA.[11,15,17,34-36] Mild TV insufficiency is often well tolerated and often does not require surgical intervention at the time of TA repair. However, a small proportion of patients with mild TV insufficiency may progress to moderate or greater insufficiency, requiring surgical intervention on the valve. We previously reported that 7.2% (11 of 159) of patients with mild or less TV insufficiency have progressed to moderate or greater insufficiency and have a freedom from TV surgery of 85% at 20 years.[15]

One of the most challenging patient cohorts are neonates with clinically significant TV insufficiency. Unfortunately, the proportion of neonates with TV insufficiency and their associated outcomes are seldom reported in the literature. Neonates with a higher degree of TV insufficiency often present earlier, with more overt signs of heart failure necessitating urgent surgical intervention. Clearly, operating on neonates in this critical state would increase the risk of early death. We previously reported outcomes of 11 neonates who underwent concomitant TV surgery, with an early mortality of 27% (3 of 11) compared with 10% (1 of 10) in older patients.[15] Similarly, Russell and colleagues[35] reported outcomes of 5 neonates who underwent concomitant TV repair between 1979 and 2012, with an early mortality of 40% (2 of 5). This illustrates that management of neonates becomes much more complicated when concomitant TV surgery is required because they are often in a critical condition. However, given the poor quality of the TV and overt signs of heart failure, there is often no choice but to operate on the TV.

Several studies showed that moderate or greater TV insufficiency was a risk factor for early reoperation for TV replacement,[13] early mortality, and generally poorer long-term outcomes if not adequately addressed during the initial operation.[22,34,36-40] However, Tlaskal and colleagues[13] 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 TV replacement. Despite a number of different techniques in a surgeon's armamentarium, deciding when to address TV insufficiency is often difficult.

When TV insufficiency is severe, the decision is obvious. However, contention arises when discussing moderate TV insufficiency. At The Royal Children's Hospital Melbourne, we routinely aim to repair the TV in the setting of moderate or greater insufficiency. Furthermore, we have previously reported no apparent association between the degree of TV insufficiency or

concomitant TV surgery and mortality,[15,16] perhaps owing to our approach to the TV. We previously reported outcomes of 17 patients with moderate TV insufficiency, of whom 13 underwent TV surgery. In the remaining 4 patients, 2 have died, and only 2 are alive without TV intervention.[15]

Unfortunately, although concomitant TV surgery may be necessary, it has limited long-term durability. We previously reported that freedom from TV reoperation was 19.2% at 20 years after concomitant TV surgery.[15] Kaza and colleagues[36] reported 29% (5 of 14) of patients who underwent concomitant TV repair required TV reoperation, with a freedom from TV reoperation of 50% at 7 years. Like others, we have used several surgical techniques for TV repair (Figure 1). These include suturing partially developed commissures, resuspension of leaflets, resection of redundant portion of leaflets, pericardial leaflet extension, tricuspidization of a quadricuspid valve (Figures 1A, 1B), or reduction of the annulus (Figures 1C, 1D). The outcomes of these methods are varied, with early mortality as high as 30%.[34] There is now an evolving appreciation for the need of reduction of the diameter of the truncal annulus that may be a key for successful repair.[15,41-46]

The Quadricuspid Truncal Valve

Interestingly, the most common subgroup of patients requiring TV surgery appears to be those with a quadricuspid TV. Russell and colleagues[35] reported that most patients who underwent TV surgery had a quadricuspid TV. Of our patient cohort who required TV surgery, 57% (12 of 21) had a quadricuspid TV. Furthermore, 79% (15 of 19) of patients with moderate or greater insufficiency had a quadricuspid TV.[15,41] In contrast, most patients with a quadricuspid TV and mild or less insufficiency did not require TV surgery. In fact, only 16.7% (7 of 42) of patients who had a quadricuspid TV had progression of insufficiency requiring surgery, of whom 5 had mild or less TV insufficiency.[15,43]

Our preferred method of TV repair, which appears to give the best long-term results, is tricuspidization with reduction of the annulus (Figures 1A, 1B).[43] This can be achieved by resection of a leaflet and annulus reduction, which was first described by Imamura and colleagues[45] in 1999, cusp reconstruction and annulus reduction, or cusp reconstruction. Interestingly, we have shown durability of tricuspidization to be superior to non-tricuspidization techniques. In our recent study, there were only 3 reoperations in 11 patients who underwent tricuspidization compared with 4 reoperations in 6 patients who underwent repair by non-tricuspidization.[43] 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 TV repair by non-tricuspidization.[43] Furthermore, tricuspidization provided better long-term outcomes even if the non-tricuspidization group included younger children (aged <6 years) in whom TV replacement was performed.[43] Similarly, Myers and colleagues[44] reported tricuspidization methods of TV repair tended to improve freedom from reoperation on the TV.

Interrupted Aortic Arch

Another significant issue in patients with TA is an IAA. An IAA occurs in approximately 10% to 20% of patients with TA and is commonly type B.[8,30,31,33,47-49] Several previous studies have shown an IAA to be a risk factor for mortality[6,8,13,22,31,34,50]; however, many of these studies were assessing patients who underwent surgery in the 1980s and 1990s. McCrindle and colleagues[50] reported that outcomes of TA with IAA were worse than IAA alone. More contemporary studies have mitigated the risk of death likely due to improved surgical techniques and perioperative management of these complex patients. However, many of these patients present early in the neonatal period and still pose a significant surgical challenge. We previously reported no association between mortality in TA with IAA and TA alone.[16] In fact, overall survival in patients with TA with IAA was 83% at 20 years compared with 74% at 20 years in patients with TA without IAA. Thus, it would appear that IAA per se no longer increases the risk of death in patients with TA who undergo operations in the modern era.[51] Rather, the competence of the TV may be the most important factor influencing outcomes.

Our center has been using end-to-side repair since the early 1980s, and we aim for end-to-side anastomosis of the aortic arch when possible.[24,51,52] Repair can be undertaken with the use of deep hypothermic circulatory arrest or isolated cerebral perfusion. Anastomosis of the ascending and descending aorta is then performed, with or without the use of a patch or interposition graft. If, after mobilization and approximation there appears to be undue distortion of the aortic arch, or potential for compression of the left pulmonary artery (Figure 2), a patch can be used to augment the anastomosis.[24,51,53]

It is not uncommon that patients with concomitant IAA require reoperation for aortic arch obstruction. The Congenital Heart Surgeon's Society[31] reported an aortic arch reoperation rate of 13.2% (5 of 38), with a survival of only 28% at 5 years without reoperation. McCrindle and associates[50] reported that aortic arch reoperation was more likely for those who had IAA repair by a method other than direct anastomosis with patch augmentation. A previous review of IAA repairs in isolation or with TA repair at our center demonstrated a low aortic arch reoperation rate using the end-to-side technique.[24,46,52] Of note, however, 5 of 6 patients who required aortic arch reoperation in our series on TA and IAA had initial repair without the use of a patch.[51] In those patients with TA and an IAA, we reported an overall freedom from aortic reoperation of 68% at 10 years, with 3 patients requiring reoperation within 30 days of the initial repair.[51]

Coronary Artery Anomalies

A concomitant coronary artery anomaly has been reported to be a risk factor for mortality.[8,9,16,17,28,54-56]

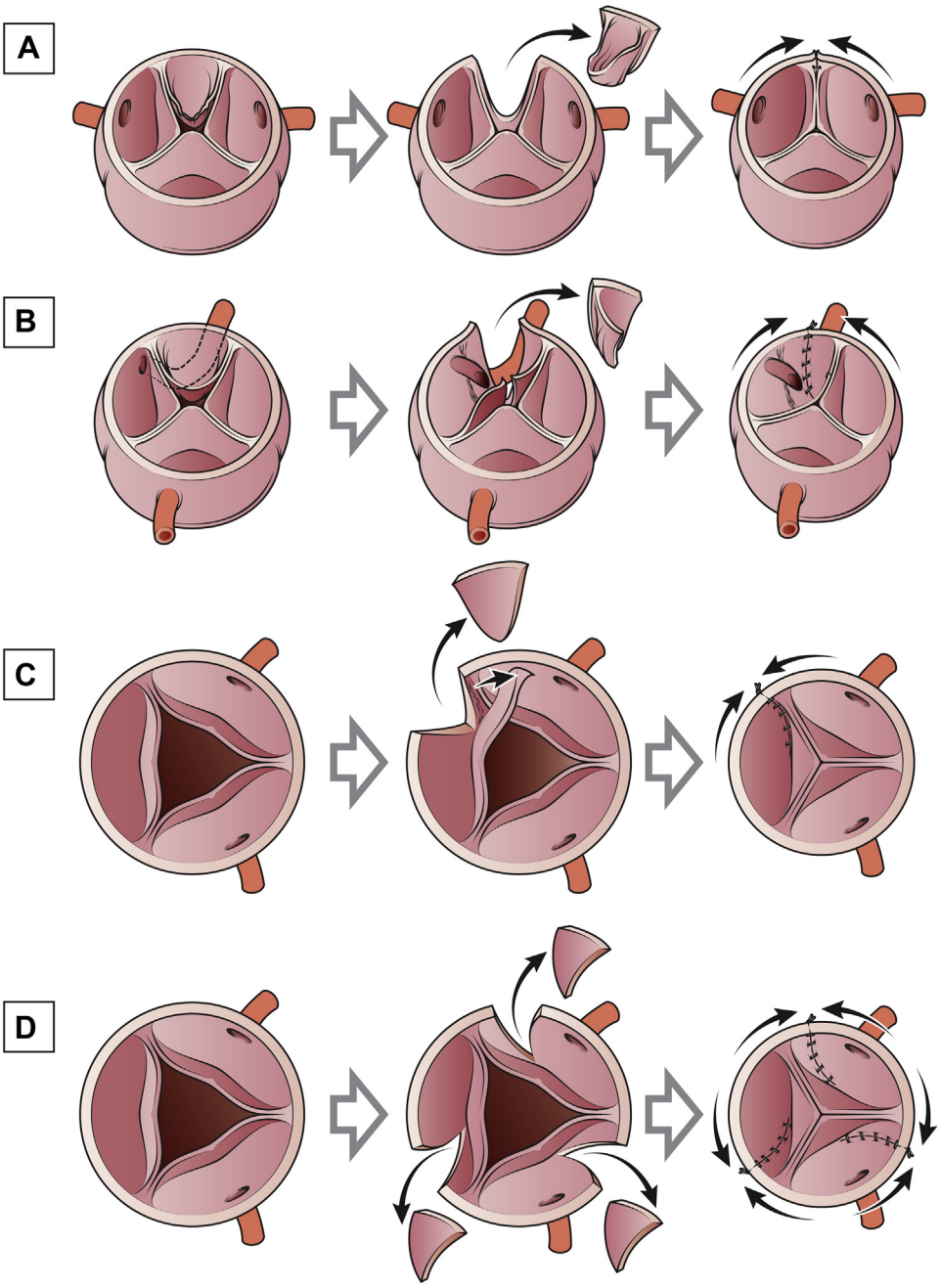

Figure 1. Surgical approaches to addressing significant truncal valve (TV) insufficiency in patients with truncus arteriosus. (A) Tricuspidization of the TV with resection of the smallest or most redundant leaflet and reduction of the annulus. (B) In the case of an intramural coronary artery crossing underneath the commissure, the redundant leaflet is resected along with the commissure, and the remaining valve leaflet is reconstructed and the coronary artery unroofed. (C and D) A large tricuspid TV can be repaired by reduction of the annulus, allowing for a larger surface for coaptation.

Schreiber and colleagues[28] reported the outcomes of 13 patients with a coronary artery anomaly and concluded that a coronary artery anomaly was associated with poorer surgical outcomes related to compression or distortion of the anomalous coronary artery. Coronary artery anomalies have been reported in 5% to 20% of patients with TA, but ascertaining the precise incidence of this anomaly is difficult.[8,9,16,28] It is not uncommon for a coronary artery anomaly to be missed on preoperative echocardiogram in patients with TA and subsequently diagnosed intraoperatively. Abnormalities of the coronary arteries mostly consist of abnormal origin of a coronary artery, with or without an intramural course, a single coronary artery giving rise to the entire coronary circulation, or variable epicardial courses of these arteries.[57]

Although there is a limited cumulative experience with coronary artery anomalies and TA, it appears that many deaths occurred in patients with a major branch coronary artery that was crossing the RVOT.[16] The reason for these deaths cannot be definitively explored but could potentially have resulted from compression of the branch coronary artery by the overlying conduit. This is a difficult predicament, because the overlying conduit would

require sufficient spacing from the underlying coronary artery to avoid compression as well as being compact enough to avoid kinking or distortion when the sternum is closed.

Rarely, patients with TA may have an intramural coronary artery (Figure 3A), which may increase the risk of myocardial ischemia. A recent study by Patrick and colleagues[58] reported that an intramural coronary artery occurred in 15% of their patients. Myocardial ischemia may be caused by compression of the intramural segment, kinking due to an acute take-off angle, or the compression by an ostial ridge. Surgical management of TA and an intramural coronary artery involves unroofing the intramural segment and formation of an unobstructed neo-ostium (Figured 3B, 3C). The intramural segment may course behind the truncal valve commissure and may need to be reconstructed or resuspended to facilitate unroofing.[41] We have previously described our technique for coronary artery unroofing in detail and provided a video of the unroofing technique.[41]

RVOT Reconstruction

Reconstruction of the RVOT can be established with a conduit or direct anastomosis of the pulmonary artery to the right ventricle. The best method of RVOT reconstruction is yet to be determined. The use of a conduit to reconstruct the RVOT is the most common method. However, conduits have limited durability because they cannot grow or regenerate and, therefore, inevitably require reoperation.[59-61] Additionally, as with any foreign material, they are prone to infection and thrombosis.[60]

Bioprosthetic Conduits

The choice of conduit depends on several factors, including but not limited to availability, patient size and hemodynamics, and surgeon preference. The Contegra (Medtronic, Minneapolis, MN) bovine jugular vein conduit is predictable in performance and lacks immunogenic properties.[62] Herrmann and colleagues[63] reported outcomes of 100 TA patients between 1981 and 2018 with a median follow-up of 15.6 years. They demonstrated longer freedom from reoperation with the bovine jugular vein conduit compared with an aortic homograft but no difference compared with a pulmonary homograft.[63] Furthermore, they showed that larger conduit size was associated with longer freedom from reoperation. Interestingly, Vitanova and colleagues[64] reported RVOT reconstruction in 145 patients and found that the overall durability of homograft, bovine jugular vein conduits, and porcine-valved conduits to be similar; however, the bovine jugular vein conduit appeared to develop insufficiency and stenosis earlier than other types.

In contrast to these more favorable results, Buckley and colleagues[21] reported outcomes of 216 neonates that showed a 2-fold increase risk of reintervention when the Contegra conduit was used compared with a homograft, regardless of size. Furthermore, several studies have reported an increased rate of endocarditis in patients with a Contegra conduit.[63,65,66]

Homografts

An alternative to the bioprosthetic conduits are the pulmonary and aortic homografts. Homografts have been the conduit of choice in recent decades but have been limited due to smaller sizes, availability, and potential for obstruction. Homografts reportedly have better hemodynamic properties and increased longevity.[67] Vohra and colleagues[29] in 2010 reported outcomes of 32 patients who underwent TA repair with 24 aortic homograft and 8 pulmonary homograft reconstructions, resulting in a freedom from reoperation of 68% at 10 years and 37% at 20 years. As with bioprosthetic conduits, larger homograft sizes increase longevity and improve freedom from reoperation. Several studies have suggested homografts less than 12 mm in diameter fail earlier, requiring earlier replacement.[13,16,22,68,69] Vohra and colleagues[29] showed that oversizing the homograft increased longevity of the graft for up to 12 years. Care must be taken, however, to not excessively oversize the conduit. Mastropietro and colleagues[70] reported outcomes of 216 neonates in a multicenter study and showed that conduit type did not impact reoperation rates, whereas a conduit size exceeding 50 mm/m^2 had a 5-fold increase in mortality. Furthermore, larger-sized conduits appear to have an increased risk for coronary artery compression, TV or pulmonary artery distortion, and they require a larger ventriculotomy.[23]

Polytetrafluoroethylene Conduit

More recently, polytetrafluoroethylene (PTFE) conduits have become increasingly used due to their availability, ease of construction, lower cost, being immunologically inert, and having comparable results to other conduits.[71,72] Seese and colleagues[71] reported outcomes of 28 neonates who underwent RVOT reconstruction between 2004 and 2016 with a homograft (n = 7) or a PTFE conduit (n = 18). They reported that the rates of reintervention and time to reintervention on the RVOT were similar between PTFE conduits and homografts. Interestingly, Seese and colleagues[71] reported that at both the 5- and 10-year follow-up assessment, patients who had a PTFE conduit had better survival than those who had a homograft. Similarly, Mercer and colleagues[72] reported outcomes of RVOT reconstruction in 55 patients aged younger than 2 years between 2004 and 2015 and found that reintervention rates and time to reintervention were similar between patients who received a homograft and those who had a PTFE conduit. Furthermore, as with other conduits, larger conduit size resulted in longer freedom from reintervention on the RVOT. Interestingly, it appears that the primary mode of failure with the PTFE conduits may be fibrosis or narrowing at the distal anastomosis over time rather than fibrous tissue formation within the conduit.[72]

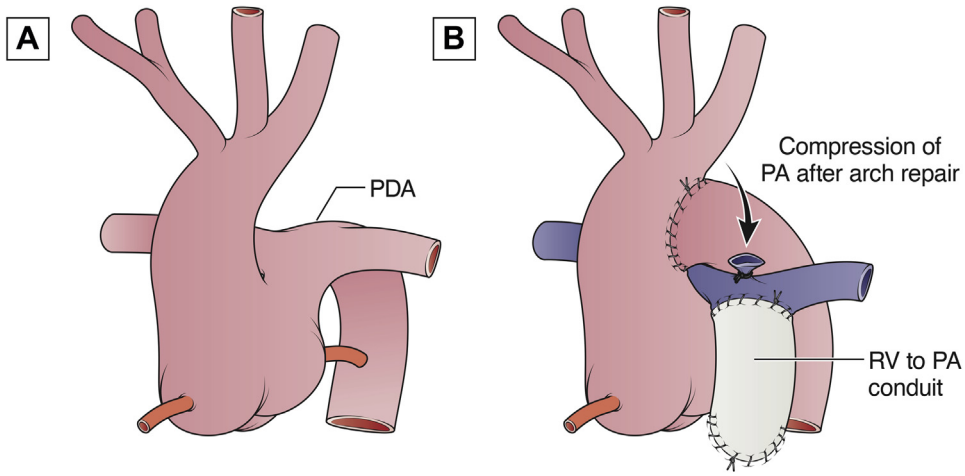

Figure 2. Surgical repair of truncus arteriosus (TA) and interrupted aortic arch (IAA). (A) Schematic of TA with IAA. (B) Surgical repair of TA with IAA. The descending aorta is extensively dissected and mobilized, advanced, and directly anastomosed end-to-side to the ascending aorta. The patent ductus arteriosus (PDA) is ligated, and all ductal tissues are resected. When the descending aorta cannot be adequately mobilized for the advancement procedure as described, the aortic arch can be repaired using patch augmentation of the arch or interposition graft to the descending aorta. Care should be taken to avoid compression of the right pulmonary artery. (RV to PA, right ventricle to pulmonary artery.)

Direct Right Ventricle-to-Pulmonary Artery Anastomosis

The biggest hindrance to long-term freedom from conduit reoperation is the lack of living material with the capacity for growth or regeneration. Any child who requires a conduit as part of their TA repair will invariably require reoperation. In an attempt to circumvent this issue, direct anastomosis of the pulmonary artery to the right ventricle, with or without the use of autologous tissues, has been explored.[9,12,14,27] In the case of TA type 1, the main pulmonary artery can be sutured to the right ventriculotomy with the anterior portion of the RVOT being reconstructed with a pericardial patch and a monocusp valve created of autologous pericardium. Danton and colleagues[9] reported a freedom from RVOT reoperation of 89% at 4 years in patients who underwent direct anastomosis of the main pulmonary artery to the right ventricle compared with 58% at 4 years in patients who underwent conduit repair.

An issue with direct anastomosis of the RVOT is that without favorable anatomy, it often leads to pulmonary artery distortion, which necessitates early reoperation.[73] Another drawback to this method is the potential for severe pulmonary regurgitation, which has not yet been observed, likely due to short follow-up.[9,12,14,27] Augmentation of the anastomosis with a patch anteriorly has been suggested to avoid pulmonary artery distortion, which would also allow for interval growth and avoid or delay obstruction to the RVOT or the branch pulmonary arteries.[9] Although early mortality is similar between direct anastomosis and using a conduit, there are limited long-term data.[9,12,14]

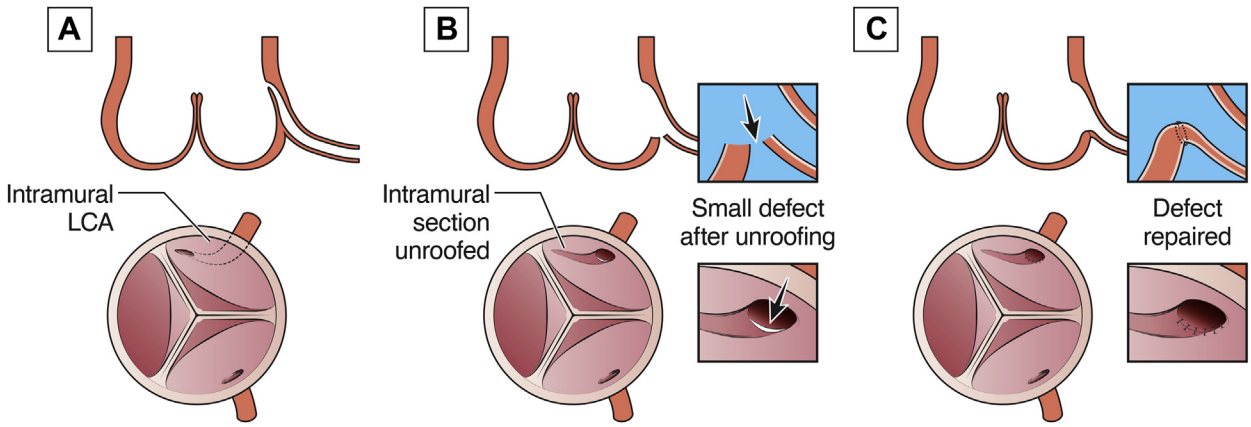

Figure 3. Truncus arteriosus (TA) with concomitant intramural coronary artery. (A) TA with an intramural coronary artery, often with an acute angle and ridge of tissue. (B) The coronary artery should be unroofed with the creation of a generous neo-ostium. (C) If a small defect occurs after the unroofing procedure, the vascular wall is reapproximated by running suture. (LCA, left coronary artery.)

Long-term Quality of Life After TA Repair

An ever-growing population of TA patients are reaching adulthood, yet there is little information about their quality of life. Because most patients who undergo TA repair require reoperation throughout their lifetime, whether this negatively affects their quality of life is unclear. O'Byrne and colleagues[74] assessed the health status of 25 patients with a median age of 11.8 years who underwent TA repair. Factors such as exercise tolerance, maximal oxygen consumption, maximal work, and forced vital capacity were all lower than normal for age and sex.[74] Additionally, health-related quality of life was diminished and comparable with that of children with severe heart disease, represented by the Fontan population.[74] However, psychosocial functional status was not significantly diminished. These findings collectively represent moderate morbidity and disability.[74] By understanding the health-related quality of life after TA repair, we may be able to assess the overall impact of this condition on the individual and potentially provide a reference for management to address physical or psychosocial issues that may arise.

We recently reported on the quality of life in adult survivors of childhood TA repair.[75] Using the 36-Item Short Form Health Survey (SF-36) questionnaire, we observed that adults with TA had a similar quality of life compared with age-matched controls. Furthermore, the quality of life in patients with TA was compared with that of a patient who underwent the arterial switch operation, neonatal procedure with traditionally low reoperation rates. Despite a higher reoperation rate in TA patients, they have a similar quality of life as patients who underwent the arterial switch. Interestingly, when each of the domains of the SF-36 questionnaire was assessed, physical function was consistently lower than in age-matched controls in all age groups. As the SF-36 questionnaire does not provide an objective measure of physical capabilities, it is difficult to ascertain whether there are limitations due to a sedentary lifestyle or whether adults with TA have physical limitations despite physical activity. Further quantitative assessment of physical functioning through exercise testing is required to elucidate potential underlying cardiac limitations of physical activity.

Conclusion

Repair of TA is associated with excellent survival beyond the first year after repair. However, reoperation rates remain high due to the use of a conduit for the RVOT. The long-term functional state of survivors is excellent, and their quality of life similar to the age-matched control. The competence of the TV appears to be the most important factor influencing early outcomes. Children with mild TV insufficiency are free from TV surgery for up to 25 years. However, children with moderate or greater TV insufficiency, particularly those with a quadricuspid TV, will likely require TV surgery throughout their lifetime.

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