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ORIGINAL ARTICLE

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Surgical repair of truncal valve regurgitation†

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CONGENITAL

Abstract

OBJECTIVES: Truncal valve regurgitation remains a short- and long-term risk factor for patients with truncus arteriosus. There are limited data available on techniques and outcomes of truncal valve repair (TVR). The aim of this study was to report our experience with TVR in patients of all ages.

METHODS: From 1997 to 2012, 36 patients (13 neonates, 30 children and 3 adults) underwent TVR for significant regurgitation.

RESULTS: There were 3 early deaths (8%), all of which were in neonates. Twenty-two patients had a quadricuspid, 13 a tricuspid and 1 a bicuspid truncal valve before repair. Valve repair improved regurgitation in 31 of 36 repairs. The median regurgitation decreased from moderate-severe to mild ($P < 0.001$). During a mean follow-up of 38.3 ± 44.9 months (range 1 month–15 years), there was 1 late death, 16 patients required reoperation on the truncal valve and 1 required a second reoperation. Freedom from reoperation was 91.4 ± 4.8% at 1 year, 55.0 ± 10.4% at 5 and 22.9 ± 12.2% at 10 years. A quadricuspid valve after repair tended to worsen freedom from reoperation ($P = 0.15$), and tricuspidization tended to improve freedom from reoperation ($P = 0.19$). Neonatal repair (hazards ratio (HR) 4.1, $P = 0.03$) and leaflet thinning (HR 22.5, $P = 0.002$) were independent predictors of reoperation.

CONCLUSIONS: Valve repair for truncal valve regurgitation is feasible, with good results. Surgical creation of a tricuspid truncal valve seems to provide the best outcomes in this challenging population.

Keywords: Truncus arteriosus • Truncal valve regurgitation • Truncal valve repair • Valve repair

INTRODUCTION

Truncus arteriosus is a rare cardiovascular malformation that represents 2–4% of all congenital heart defects [1]. Since the first description of successful repair by McGoon *et al.* [2] in 1968, the outcome has evolved from a fatal condition early in life in 50% of patients to one in which neonatal repair is increasingly common and successful [3]. However, associated lesions such as interrupted aortic arch (IAA), coronary anomalies and truncal valve insufficiency (TVI) remain major risks factors for reoperation and survival [4].

TVI is estimated to occur in 25–50% of patients with truncus arteriosus [5]. In this condition, the semilunar valve can be dysplastic or dysfunctional or have an abnormal number of leaflets, usually four. Moreover, associated abnormalities such as conal septum hypoplasia, ventricular septal defect (VSD) causing valve prolapse and the relationship of the pulmonary arteries with a consistently dilated aorta may affect the truncal valve function. In addition, TVI may present in the neonatal period or throughout childhood. Therefore, the surgical management of moderate or severe TVI is challenging, particularly in the neonatal period. Traditional strategies for the operative management of these patients have included temporization of this problem or attempts at valve replacement with a homograft valve or a mechanical prosthesis [5, 6]. However, none of these options have been shown to be very successful or desirable. Moreover, in growing children, the ability to repair a regurgitant semilunar valve may be even more important to avoid the need to replace or up-size a prosthetic valve or allograft. Thus, repair of the regurgitant truncal valve constitutes the 'ideal' surgical strategy. However, reports on technical feasibility or durability of truncal valve repair (TVR) are scarce and limited to case reports or small series of patients [7–10]. No standard approach, with regard to timing and surgical technique, for the treatment of this condition currently exists.

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The aim of this study was to report our recent experience with truncal valve surgery in neonates, children and young adults, focusing on the surgical strategy, timing of operation and durability of the repair.

METHODS

Study design

This study is a retrospective review of all patients managed at our institution with the diagnosis of truncus arteriosus and moderate or severe truncal valve regurgitation during follow-up from 1997 to 2012. The cardiology and cardiac surgery databases were queried for patients with both these diagnoses, and all of them were included in the study. The study was approved by the Boston Children's Hospital Institutional Review Board, and individual patient consent was waived.

The primary endpoints were truncal valve reoperation, late valve function and death. Clinical or treatment variables were recorded to determine the predictors of reoperation. All patients underwent follow-up to death or March 2012.

Operative techniques

The techniques used to repair the truncal valve were planned ahead of time with the help of 2D and 3D echocardiography and tailored to the underlying mechanism of valve dysfunction. Preoperative datapoints of interest for planning the repair included the number, size and shape, thickness and mobility of the leaflets, number and location of commissures and cusps, regurgitant jets and size of the truncal valve annulus, aortic root and sinotubular junction. Specific techniques varied according to the anatomy of the valve and the cause of insufficiency. A combination of suture closure of adjacent leaflets with resection or exclusion of rudimentary leaflets, leaflet extension with autologous pericardium, annular plasties, commissuroplasties and aortoplasties were used to repair the truncal valve (see Fig. 1).

The most common surgical scenario in our series was a quadricuspid truncal valve with thick edges and one deficient and one flail leaflet usually separated by a raphe. These leaflets were mostly located at the left or non-coronary sinuses of Valsalva. Repair was accomplished by thinning the free edges of the leaflets, and the raphe was then taken down to improve mobility and the flail leaflet was resuspended either by suturing it to the adjacent leaflet or commissure, making the valve trileaflet [9, 11]. In patients with a dilated aortic root, the non-coronary sinus was excised and the aortic wall sutured back together. Taking advantage of the existence of a subannular conus in truncal valves, an annuloplasty was also used, particularly in neonates [12].

In older patients, a technique of truncal valve annulus remodelling, inspired by Yacoub *et al.*'s technique of transaortic primary repair of subpulmonary VSD with aortic cusp prolapse [13] was used (see Fig. 2), as the truncal valve annulus below the right coronary cusp was found to balloon out and cause prolapse of the right coronary cusp. A series of interrupted pledgeted multifilament braided sutures were inserted in a transaortic approach on the left aspect of the crest of the ventricular septum, passed through the truncal valve annulus and out the aortic root wall adjacent to the infundibulum, where they were tied. As in Yacoub *et al.'s* technique, this results in closing the VSD, placating redundant tissues towards the media of the aortic sinus and bringing the right coronary sinus, cusp and truncal annulus down to the VSD crest, effectively remodelling the truncal annulus.

Statistical methods

Statistical analyses were performed with the SPSS software (version 20, SPSS, Inc., Chicago, IL, USA). Data are presented as

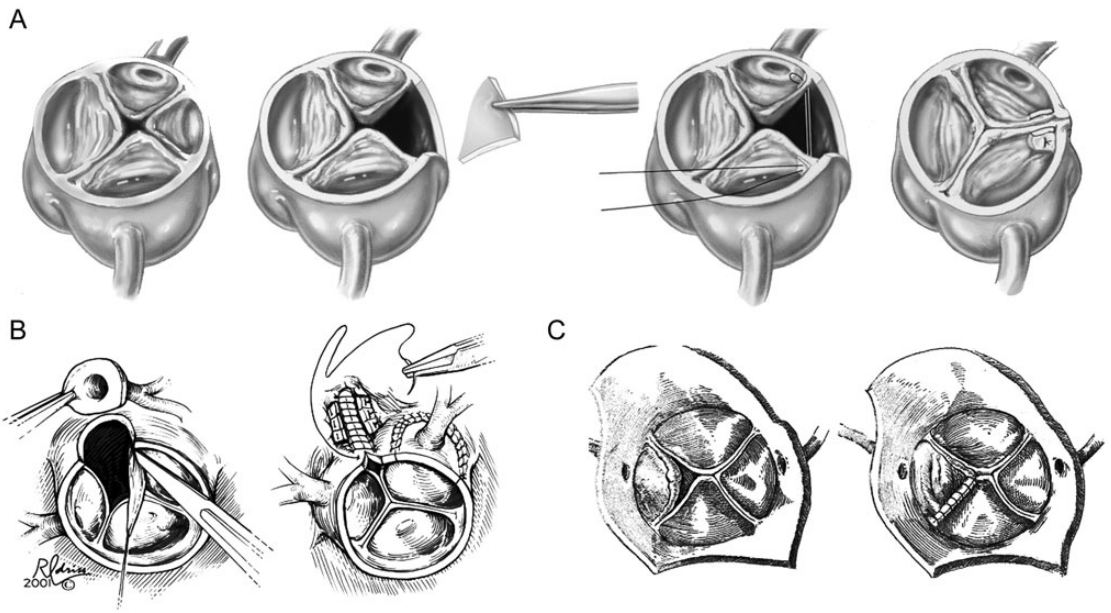

Figure 1: TVR tricuspidzation techniques. (**A**) Resection of rudimentary leaflet and tricuspidization. Reproduced with permission from Imamura *et al.* [22]. (**B**) Rudimentary leaflet and sinus resection with coronary transfer. Reproduced with permission from Mavroudis and Backer [16]. (**C**) Leaflet approximation in a quadricuspid truncal valve. Reproduced with permission from Kaza *et al.* [9].

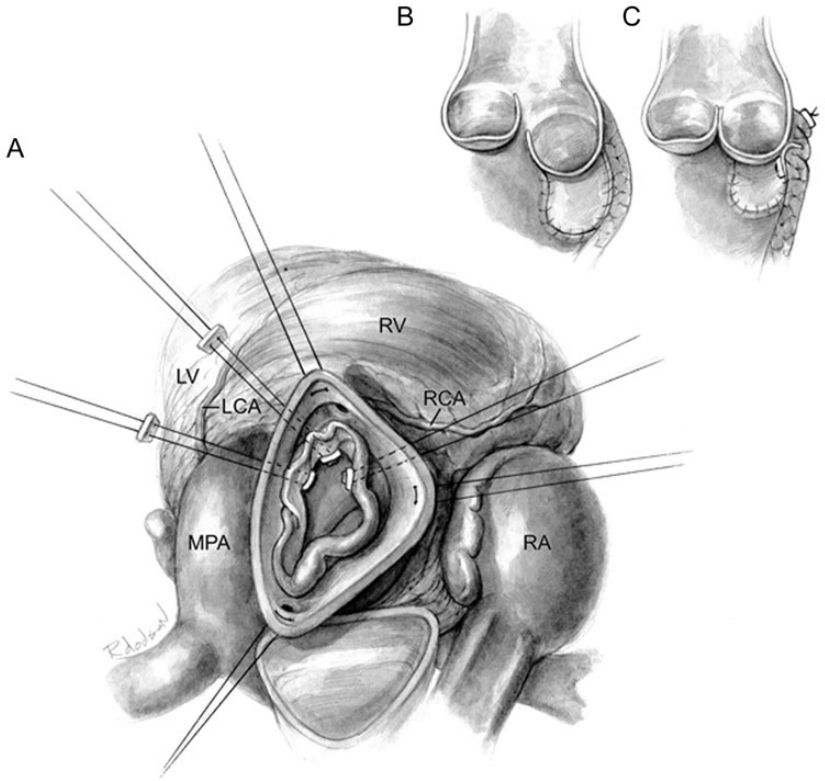

Figure 2: Truncal annulus remodelling technique by bringing the VSD crest up to the truncal annulus. (**A**) View from the head of the patient (at the bottom of the image), down the barrel of the transected ascending aorta towards the truncal valve. (**B**) Cross-sectional representation of the truncal root and VSD patch before repair, with a prolapsing truncal cusp. (**C**) View after repair, eliminating prolapse. LCA: left coronary artery; LV: left ventricle; MPA: main pulmonary artery; RA: right atrium; RCA: right coronary artery; RV: right ventricle.

mean ± SD or median (range) where appropriate. Continuous variables were analysed with the Student's *t*-test for normally distributed variables, or the related samples Wilcoxon signed-rank test for non-normally distributed variables, and categorical variables using Fisher's exact test. Normality of continuous data was evaluated with the Kolmogorov–Smirnov test. Actuarial estimates were calculated using the Kaplan–Meier method and differences between curves assessed by the log-rank test. Univariable and stepwise Cox multivariable regression analyses were used to identify the predictors of late outcomes. Preoperative and operative variables with a univariable $P < 0.1$ or those judged to be clinically important were entered into the Cox models. Hazards ratios (HRs) for reoperation of risk factors were calculated using univariable and multivariable Cox proportional hazards estimates. All statistical tests were two-tailed and $P < 0.05$ were taken as significant.

RESULTS

Demographics

Thirty-six patients underwent a total of 53 truncal valve procedures during the study period for moderate or severe regurgitation. Forty-nine procedures were valve repairs, and 4 were valve replacement. The initial procedure was TVR in all 36 patients. Patient baseline characteristics are summarized in Table 1. The mean age at TVR was 6.9 ± 6.8 years. Sixty per cent of patients had a quadricuspid truncal valve prior to repair, and 33% had associated cardiac lesions.

TVR techniques

A functional tricuspid valve was accomplished in 21 patients (58%), quadricuspid in 11 (31%) and bicuspid in 4 (11%) by a combination of commissuroplasties, suture closure of adjacent leaflets, pericardial patch extension and leaflet resuspension and thinning. Surgical valve repair techniques used are detailed in Table 2. The techniques used among the 13 neonates did not differ significantly from the overall cohort of patients, which included commissurotomy in 2 patients (15.4%), commissuroplasty in 7 (53.8%), leaflet augmentation in 2 (15.4%), leaflet thinning in 3 (23.1%), annuloplasty in 2 (15.4%), single sinus excision in 1 (7.7%) and tricuspidization in 4 (30.8%).

Outcomes

There were 4 deaths (11.1%). Three neonates died in hospital after primary repair following extracorporeal membrane oxygenation (ECMO). The first patient collapsed and was transferred to our institution with significant right ventricular dysfunction and tricuspid insufficiency that persisted after repair. The second patient had significant truncal valve regurgitation after repair and areas of duskiness that precluded the patient from weaning from by-pass. Interestingly, these 2 patients had IAA and single coronary artery as associated lesions. The third patient had persistent significant truncal valve regurgitation, which required truncal valve re-repair. Although this improved the regurgitation to mild, weaning from cardiopulmonary bypass remained impossible and the patient was transitioned to ECMO, and subsequently developed multiorgan

Table 1: Baseline patient characteristics

Characteristic	Entire cohort ($n = 36$)	Reoperation free ($n = 20$)	Reoperations ($n = 16$)	P
Age at TVR (years)	6.9 ± 6.8	9.3 ± 6.6	4.0 ± 6.6	0.02
Neonates	13 (36.1%)	4 (20.0%)	9 (56.2%)	0.04
Infants	0 (0%)	0 (0%)	0 (0%)	1.00
Children	20 (55.6%)	14 (70.0%)	6 (37.5%)	0.09
Adults	3 (8.3%)	2 (10%)	1 (6.2%)	1.00
Associated anomalies	12 (33.3%)	8 (40%)	4 (254%)	0.48
IAA	5 (13.9%)	3 (15%)	2 (12.5%)	1.00
Coronary anomalies	5 (13.9%)	3 (15%)	2 (12.5%)	1.00
Aortic coarctation	1 (2.8%)	1 (5%)	0 (0%)	1.00
DCRV	1 (2.8%)	1 (5%)	0 (0%)	1.00
Multiple VSDs	1 (2.8%)	1 (5%)	0 (0%)	0.44
TVR at truncus arteriosus repair	11 (30.6%)	4 (20%)	7 (43.8%)	0.16
Truncal valve anatomy before repair				
Quadricuspid	22 (61.1%)	10 (50%)	12 (75%)	0.18
Tricuspid	13 (36.1%)	10 (50%)	3 (18.8%)	0.08
Bicuspid	1 (2.8%)	0 (0%)	1 (6.2%)	0.44
Truncal valve anatomy after repair				
Quadricuspid	11 (30.6%)	3 (15%)	8 (50%)	0.03
Tricuspid	21 (58.3%)	15 (75%)	6 (37.5%)	0.04
Bicuspid	4 (11.1%)	2 (10%)	2 (12.5%)	1.00

Associated lesions: a patent foramen ovale, the usual malalignement VSD for truncus arteriosus and/or a right aortic arch were not considered as significant associated lesion, as these are frequent in truncus arteriosus.
IAA: interrupted aortic arch; DCRV: double chamber right ventricle; VSD: ventricular septal defect; TVR: truncal valve repair.

Table 2: Operative techniques used in truncal valve repair

Technique	Entire cohort ($n = 36$)	Reoperation free ($n = 20$)	Reoperations ($n = 16$)	P
Leaflets				
Commissurotomy	3 (8.3%)	1 (5.0%)	2 (12.5%)	0.57
Commissuroplasty	22 (61.1%)	11 (55.0%)	11 (68.8%)	0.5
Leaflet augmentation	8 (22.2%)	6 (30.0%)	2 (12.5%)	0.26
Leaflet thinning	9 (25.0%)	5 (25.0%)	4 (25.0%)	1.00
Raphe takedown	1 (2.8%)	1 (5.0%)	0 (0%)	1.00
Tricuspidization	11 (30.6%)	7 (35.0%)	4 (25.0%)	0.72
Annular or root remodelling				
Truncal annuloplasty	5 (13.9%)	4 (20.0%)	1 (6.2%)	0.36
Annular remodelling using VSD crest	3 (8.3%)	3 (15.0%)	0 (0%)	0.24
Single sinus of Valsalva reduction	6 (16.7%)	6 (30.0%)	0 (0%)	0.02
Other procedures				
Subaortic resection	1 (2.8%)	1 (5.0%)	0 (0%)	1.00

system failure on full flow ECMO and expired. The post-mortem examination showed obstruction of left main coronary ostium. In addition, a 4.5-year old child died from a massive stroke after reoperation for severe TVI that was unsuccessfully re-repaired and ultimately required a prosthetic replacement.

Valve repair improved regurgitation in 31 of 36 repairs (86%) and was less than moderate in 27 patients (75%) after repair. The median regurgitation decreased from moderate-severe to mild ($P < 0.001$). Although 9 patients (25%) presented a significant truncal valve peak gradient prior to repair, no patient exhibited truncal valve stenosis after repair. During a mean follow-up of 38.3 ± 44.9 months (range from 1 month to 15 years), 16 patients required a reoperation on the truncal valve, one of whom required a second reoperation. No patients required >2 reoperations. Nine of the 13 patients who had their initial TVR as neonates required a reoperation (69%). Among reoperations, 13 were for truncal valve re-repair, 2 for prosthetic replacement after attempted re-repair and 2 for direct prosthetic replacement. Freedom from reoperation for TVI was 91.4 ± 4.8% at 1 year, 87.2 ± 6.1% at 2 years, 55.0 ± 10.4% at 5 years and 22.9 ± 12.2% at 10 years (see Fig. 3).

One patient required placement of a permanent pacemaker for postoperative atrioventricular block after reoperation for valve replacement. This patient's initial repair included only leaflet-level repairs, and no annuloplasty or truncal root remodelling techniques.

Predictors of truncal valve reoperation

The Kaplan–Meier survival analysis stratified by age category showed that neonates and adults had significantly more reoperations than children ($P = 0.039$; see Fig. 3). A quadricuspid

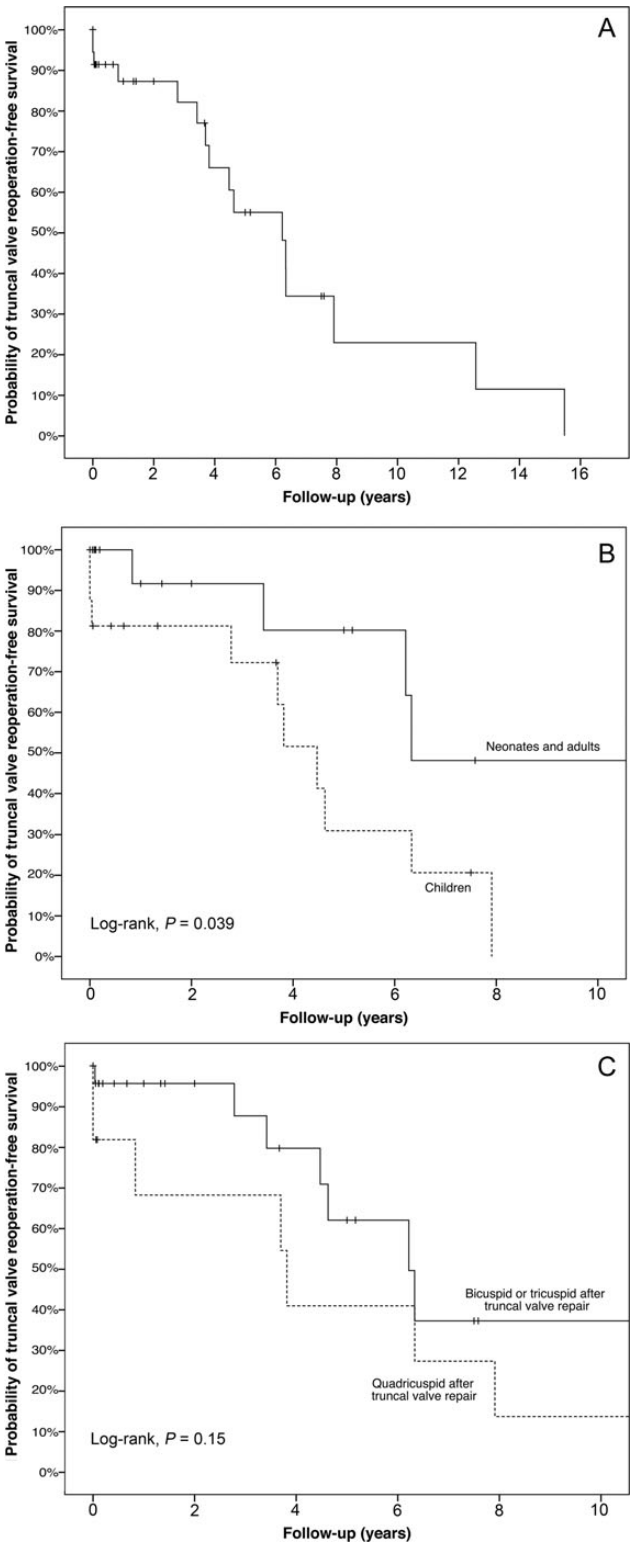

Figure 3: Kaplan–Meier estimates of freedom from truncal valve reoperation after TVR. (**A**) Entire cohort. (**B**) Analysis stratified by age at TVR. (**C**) Analysis stratified by truncal valve anatomy after TVR.

anatomy after repair tended to have worse freedom from reoperation, however, not to a significant level ($P = 0.15$), and tricuspidization also tended towards better freedom from reoperation ($P = 0.19$). At univariable and multivariable Cox regression analysis, neonatal TVR and valve repair using leaflet thinning were significant predictors of truncal valve reoperation (see Table 3).

Although limited by the number of patients and not reaching statistical significance, repair techniques appeared to differ in neonates ($n = 13$) who required a truncal valve reoperation (9/13), who had more leaflet-level repairs (commissuroplasties 5, commissurotomies 1 and pericardial patch leaflet augmentation 1) and fewer annulus-level repairs (annuloplasty 1), compared with neonates who did not require a reoperation (4/13), who had more truncal root or annulus repairs (root remodelling 1, resection of nodular leaflet, annuloplasty and translocation of coronary ostium 1 and truncal annulus remodelling in 1) and fewer leaflet-level repairs (commissuroplasty 1 and leaflet thinning 1).

DISCUSSION

Despite improved surgical management of patients with truncus arteriosus, semilunar valve regurgitation remains a risk factor for early and late morbidity and mortality [8, 14]. However, there is no standard surgical approach to TVI. A recent review of the Society of Thoracic Surgeons Congenital Heart Surgery Database results [10], reporting 572 truncus arteriosus repairs from 2000 to 2009, included 27 patients with truncal valve surgeries (23 at truncus arteriosus repair, 4 later). The mortality of truncus repair with a concomitant truncal valve procedure (30%) was significantly increased compared with controls without truncal valve surgery at truncus arteriosus repair (10%, $P = 0.0002$), while all 4 patients who had delayed truncal valve procedures died. The association of truncus arteriosus repair, IAA repair and TVR had a mortality of 60%. Truncal valve procedures also increased the risk of mechanical support and a longer hospital stay. This study was limited to the initial neonatal admission, and provided no follow-up on late reoperations and truncal valve dysfunction after repair. Kaza *et al.* [9] reviewed their single-centre experience in TVR in 17 patients from 1995 to 2008. This study, although on a more limited number of patients, has the advantage of including 3 non-neonatal patients and follow-up data. Three patients had 1 re-repair, and 1 had 2 re-repairs before undergoing a prosthetic valve replacement at age 13 years. Freedom from truncal valve reoperation was 70% at 5 years and 50% at 7 years, with only 1 patient (6%) requiring valve replacement.

Anomalies associated with truncus arteriosus such as significant truncal valve regurgitation, IAA and/or coronary anomalies remain a risk factor for morbidity and mortality. In our series, 5 patients presented with IAA, 5 with coronary anomalies and 3 exhibited both conditions. Two of the early deaths were in patients with significant TVI, IAA and single coronary artery. Those deaths seemed related to myocardial ischaemia rather than the valve repair itself, because of the inability to wean from cardiopulmonary bypass and myocardial dysfunction depicted by echocardiography. In our experience, the association of TVI, IAA and coronary anomalies remains a challenging condition with high mortality.

The optimal approach to management of TVI remains controversial [8, 9, 15, 16]. Previous studies have reported initial moderate or severe TVI as a risk factor for truncal valve reoperation or late development of severe insufficiency [6]. Moreover, we previously reported increased mortality in neonates with significant TVI not undergoing TVR [8]. Thus, our approach is not to

Table 3: Cox regression analysis of predictors of truncal valve reoperation

Variable	Univariable			Multivariable		
	HR	*P*	95% CI	HR	*P*	95% CI
Neonatal TVR	2.61	0.09	0.87–7.82	4.06	0.03	1.12–14.69
Associated cardiac lesions	0.77	0.67	0.23–2.54			
Pre-repair truncal valve anatomy		0.76				
Bicuspid	1*					
Tricuspid	0.44	0.49	0.04–4.50			
Quadricuspid	0.46	0.47	0.06–3.81			
Post-repair truncal valve anatomy		0.23			0.51	
Bicuspid	1*			1*		
Tricuspid	0.37	0.25	0.07–2.00	0.39	0.28	0.07–2.19
Quadricuspid	0.94	0.94	0.18–4.82	0.66	0.64	0.11–3.82
Commissurotomy	0.79	0.76	0.17–3.67			
Commissuroplasty	1.10	0.85	0.37–3.34			
Leaflet patch augmentation	1.85	0.46	0.36–9.51			
Leaflet thinning	8.07	0.008	1.71–37.95	22.49	0.002	3.07–164
Raphe takedown	0.05	0.84	$0.00–4.6 \times 10^{11}$			
Tricuspidization	0.44	0.21	0.12–1.57	2.57	0.39	0.30–22.40
Annuloplasty	1.66	0.64	0.19–14.18	0.12	0.12	0.008–1.68
Annular remodelling using VSD crest	0.04	0.71	0.00–485 504			
Single sinus of Valsalva reduction	0.03	0.23	0.00–9.16			

*Reference group.

temporize with the problem but to deal with moderate or severe TVI at the time of presentation either at neonatal repair or later, during follow-up, usually concomitantly with a conduit change or pulmonary valve replacement surgery. Timing of intervention is key because the techniques used to repair the truncal valve differ slightly based on age at repair. For neonates, we prefer to use annuloplasty, aortoplasty and commissuroplasty techniques, to minimize the risk of damaging the extremely delicate neonatal leaflets. Nevertheless, for infants and older children, thinning out of the edges of the leaflets, with suture closure or resuspension of flail leaflets and pericardial augmentation of deficient leaflets, is utilized more frequently. A limitation of the present study is that we included all patients with diagnoses of truncus arteriosus and truncal valve regurgitation. This study thus did not aim to include patients with truncal valve stenosis, and cannot comment on the surgical management of this patient group, although 9 patients (25%) presented significant stenosis before repair.

An important application of 3D echocardiography is the understanding of the semilunar valves [17]. Unlike 2D imaging, 3D echocardiography demonstrates the entire surface area of the leaflets and therefore the zones of coaptation and the extent and location of valvar deficiency and prolapse. In addition, it details the dynamic and coordinated movements of the leaflets and helps in assessing the regurgitant jets. In our experience, 3D echocardiography is key for preoperative planning of valve repair as well as for intraoperative assessment of the repair, and is therefore being applied on a routine basis for surgical planning for aortic, truncal and/or mitral valve repair at our institution.

Systemic semilunar valve repair is thought to provide a better haemodynamic result and allow for growth in a neonate or child, compared with valve replacement. However, outcomes of children undergoing surgical systemic semilunar valvuloplasty are limited to few reports with small numbers and limited follow-up [7, 8, 18, 19]. Moreover, little is known about the outcomes of the repair of systemic semilunar valves in neonates [20] and specifically, durability of TVR is mostly unknown [9, 15, 16]. McElhinney *et al.* [21] reported 66% early mortality in truncal valve replacement, and 25% early mortality in truncal root replacement with a homograft, compared with 0% early mortality in truncal valvuloplasty. Furthermore, they found that TVI conferred a survival disadvantage, with 1- and 5-year survivals of 56 and 52%. We have recently reported our experience with aortic valve repair in children [18]. From 1989 to 2005, 81 patients younger than 19 years with moderate or severe aortic regurgitation underwent surgical valvuloplasty. Regurgitation improved in 77 patients (95%), was mild or less in 68 (84%) and 33 (41%) required reoperation during follow-up. Estimated freedom from reoperation was 72% at 5 years and 54% at 7.5 years. In our series of 21 patients undergoing TVR from 1997 to 2008, TVI improved in 27/30 procedures (90%) and was less than moderate in 24/30 procedures (80%). Reoperation was required in 11 patients (47%) during follow-up and estimated freedom from reoperation was 100, 77% and 34% at 1, 5 and 10 years, respectively. Therefore, in our experience, contemporary results of TVR are similar to those reported for the aortic valve. However, this statement is limited by the retrospective nature of our study, our single-institution approach and the relatively small series of patients. Moreover, the larger and older timeframe reported in the aortic valve repair study could also bias the comparison. However, in our present study, ~40% of the patients were neonates, in whom valve repair is generally considered less durable. Finally, it should be noted that it was not always possible to improve regurgitation with TVR, as 9 patients (25%) still presented moderate regurgitation after repair. This result is suboptimal, and perhaps reflects the fact that surgeries were included over a relatively long time-period and involved surgeons of varying expertise in valve repair. We currently would not accept more than mild regurgitation after repair.

One point of interest is the identification of leaflet thinning as an independent risk factor for reoperation after TVR. This is a classic example of statistical significance needing to be interpreted. Should leaflet thinning be eliminated from TVR? Leaflet thinning improves the mobility of thickened leaflets, which allows for more normal leaflet curvature and coaptation, increasing the coaptation height, a parameter of paramount importance for valve competence in the aortic position. We would argue that the multivariable analysis identified leaflet thinning as an independent predictor of reoperation, because this technique was used in a subgroup of patients with thickened, fused leaflets that were more difficult to repair.

In summary, we report the largest series of patients to date undergoing TVR within all age groups. Outcomes are comparable to those reported for children undergoing aortic valve repair. Neonatal repair was a significant independent predictor of reoperation, and creation of a trileaflet valve provides the most durable results in this challenging population. New techniques, such as truncal root and annulus remodelling, are promising and require further evaluation.

Conflict of interest: none declared.

REFERENCES

[1] Calder L, Van Praagh R, Van Praagh S, Sears WP, Corwin R, Levy A *et al*. Truncus arteriosus communis. Clinical, angiocardiographic, and pathologic findings in 100 patients. Am Heart J 1976;92:23–38.
[2] McGoon DC, Rastelli GC, Ongley PA. An operation for the correction of truncus arteriosus. JAMA 1968;205:69–73.
[3] Thompson LD, McElhinney DB, Reddy M, Petrossian E, Silverman NH, Hanley FL. Neonatal repair of truncus arteriosus: continuing improvement in outcomes. Ann Thorac Surg 2001;72:391–5.
[4] Rodefeld MD, Hanley FL. Neonatal truncus arteriosus repair: surgical techniques and clinical management. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2002;5:212–7.
[5] De Leval MR, McGoon DC, Wallace RB, Danielson GK, Mair DD. Management of truncal valvular regurgitation. Ann Surg 1974;180:427–32.
[6] Elkins RC, Steinberg JB, Razook JD, Ward KE, Overholt ED, Thompson WM Jr. Correction of truncus arteriosus with truncal valvar stenosis or insufficiency using two homografts. Ann Thorac Surg 1990;50:728–33.
[7] Hawkins JA, Kouretas PC, Holubkov R, Williams RV, Tani LY, Su JT *et al*. Intermediate-term results of repair for aortic, neoaortic, and truncal valve insufficiency in children. J Thorac Cardiovasc Surg 2007;133:1311–7.
[8] Jahangiri M, Zurakowski D, Mayer JE, del Nido PJ, Jonas RA. Repair of the truncal valve and associated interrupted arch in neonates with truncus arteriosus. J Thorac Cardiovasc Surg 2000;119:508–14.
[9] Kaza AK, Burch PT, Pinto N, Minich LL, Tani LY, Hawkins JA. Durability of truncal valve repair. Ann Thorac Surg 2010;90:1307–12; discussion 12.
[10] Russell HM, Pasquali SK, Jacobs JP, Jacobs ML, O'Brien SM, Mavroudis C *et al*. Outcomes of repair of common arterial trunk with truncal valve surgery: a review of the society of thoracic surgeons congenital heart surgery database. Ann Thorac Surg 2012;93:164–9; discussion 9.
[11] Schmidt KI, Jeserich M, Aicher D, Schafers HJ. Tricuspidization of the quadricuspid aortic valve. Ann Thorac Surg 2008;85:1087–9.
[12] Pigula FA, Mahnke CB, Anagnostopoulos PV, Casta A, Munoz R, Gandhi SK. Closed correction of systemic semilunar valve insufficiency in the neonate. J Thorac Cardiovasc Surg 2003;126:1650–2.
[13] Yacoub MH, Khan H, Stavri G, Shinebourne E, Radley-Smith R. Anatomic correction of the syndrome of prolapsing right coronary aortic cusp, dilatation of the sinus of Valsalva, and ventricular septal defect. J Thorac Cardiovasc Surg 1997;113:253–60; discussion 61.
[14] Henaine R, Azarnoush K, Belli E, Capderou A, Roussin R, Planche C *et al*. Fate of the truncal valve in truncus arteriosus. Ann Thorac Surg 2008;85:172–8.
[15] Black MD, Adatia I, Freedom RM. Truncal valve repair: initial experience in neonates. Ann Thorac Surg 1998;65:1737–40.
[16] Mavroudis C, Backer CL. Surgical management of severe truncal insufficiency: experience with truncal valve remodeling techniques. Ann Thorac Surg 2001;72:396–400.
[17] Marx GR, Su X. Three-dimensional echocardiography in congenital heart disease. Cardiol Clin 2007;25:357–65.
[18] Bacha EA, McElhinney DB, Guleserian KJ, Colan SD, Jonas RA, del Nido PJ *et al*. Surgical aortic valvuloplasty in children and adolescents with aortic regurgitation: acute and intermediate effects on aortic valve function and left ventricular dimensions. J Thorac Cardiovasc Surg 2008;135:552–9.
[19] Myers PO, Tissot C, Christenson JT, Cikirikcioglu M, Aggoun Y, Kalangos A. Aortic valve repair by cusp extension for rheumatic aortic insufficiency in children: Long-term results and impact of extension material. J Thorac Cardiovasc Surg 2010;140:836–44.
[20] Baird CW, Myers PO, Del Nido PJ. Aortic valve reconstruction in the young infants and children. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2012;15:9–19.
[21] McElhinney DB, Reddy VM, Rajasinghe HA, Mora BN, Silverman NH, Hanley FL. Trends in the management of truncal valve insufficiency. Ann Thorac Surg 1998;65:517–24.
[22] Imamura M, Drummond-Webb JJ, Sarris GE, Mee RB. Improving early and intermediate results of truncus arteriosus repair: a new technique of truncal valve repair. Ann Thorac Surg 1999;67:1142–6.

APPENDIX. CONFERENCE DISCUSSION

Dr V. Hraska *(Sankt Augustin, Germany)*: The operation for truncus has become pretty standard; however, the need to deal with associated lesions such as truncal valve regurgitation still remains a risk factor affecting survival and reoperation. So I have two questions. First, three neonates died after truncus repair and truncal valve reconstruction. All these three patients ended up on ECMO in OR due to severe myocardial dysfunction. So the question is, do you think it is justifiable to put the patient on ECMO while a neoaortic valve is still leaky, which was the case, and there is no antegrade ejection?

Dr Myers: That is a very good and important question. That is absolutely not an acceptable situation. Obviously with a leaking truncal valve, coronary perfusion will not be sufficient. And in these three patients, we had evidence that we could not wean from ECMO. Ventricular function was decreased even when loading the heart. And so obviously it was not a good option. We even saw that one of the patients had an ostial obstruction of the left main coronary artery (at post-mortem examination) so that was obviously a suboptimal result of the repair.

Dr Hraska: Can you speculate how to get out of this very complex problem in the OR? What would you recommend?

Dr Myers: I think one of the factors was allowing coronary ischaemia while it was evidenced and temporizing on that and trying to see if these patients would improve with time. When we have ischaemia, it is not going to improve with time. And so I think a more aggressive approach should be taken rather than going onto ECMO and waiting over time, and waiting maybe one or two days to go to the cath lab to get evidence of what is happening.

Dr Hraska: Maybe one can try homograft as a choice here.

Dr Myers: That would be an option although the results are quite poor in this patient population.

Dr Hraska: So you have demonstrated that reconstruction may be effective across the age spectrum including neonates; however, effectiveness of reconstruction expressed through the freedom from reoperation is pretty low. Two risk factors for reoperation were identified by uni- and multivariate analysis, namely, neonatal age, which makes complete sense, and shaving of the valve, which is unclear for me. Why is shaving of the cusp a risk factor for failure? If, on the contrary, shaving is one of the most important techniques, how do you improve mobility of cusp? In other words, your data showed that shaving of the cusp should be abandoned; that is the message.

Dr Myers: I would not like that to be the message from this study. It is what came out from our analysis, from statistics. However, the quote of 'lies, damned lies, and statistics,' applies here. In our hands, we feel can improve results with valve shaving.

Maybe there is a patient selection issue here that is evidenced through leaflet shaving. We have also seen it in other studies, that we were in the process of going through, on the mitral valve and the tricuspid valve. And perhaps these are patients who have more long-standing, more severe

regurgitation, and more severe disease, and that it is not the actual shaving that is the issue, but rather the fact that we have to shave this valve or thin this valve to improve its mobility, and that these patients are sicker and have a more difficult valve to repair adequately.

Dr Hraska: Yes. It could be very misleading if just shaving is stated as a risk factor.

Dr Myers: Absolutely.

Dr M. Hazekamp *(Leiden, Netherlands)*: I have a technical question. You spoke about this truncal valve annuloplasty, and the question is, when exactly do you see an indication for that? And, secondly, you did it only under one commissure and not under more commissures?

Dr Myers: Are you talking about the technique that I described in the video?

Dr Hazekamp: Yes, in the video.

Dr Myers: Indeed, that is usually under the right coronary cusp and close to the left noncoronary commissure. It is not a full annuloplasty, but that is because our feeling, although I do not have any echo or morphologic data to prove this, our feeling was that in these patients due to this subarterial VSD that has been previously repaired, the annulus has a tendency to dilate with time. And we found that to be involved in the mechanism. Now, I do not have any hard data to prove that, to show that that segment of the annulus is particularly dilated, but that is what we have been going after and are pursuing this further.

Dr Hazekamp: But you never felt a need to do this under other commissures as well?

Dr Myers: Not using this technique, no.

Dr D. Anderson *(London, UK)*: When you talk about relocating the coronary, I presume you are referring to the left coronary?

Dr Myers: Yes.

Dr Anderson: Just a note of caution, I have seen at least three intramural left coronaries in the context of common arterial trunk, and obviously to try and relocate that would be a very dangerous exercise. Also, it needs to be borne in mind that it runs very low down towards what you might call the annulus. So in the event of replacement, it can very easily get caught up with the sutures to hold the valve in place. I mention that as a trap for the unwary. Have you made that observation yourself?

Dr Myers: I must say no. In our series, there were just two patients who had the left main coronary that was involved just above the rudimentary leaflet that was being excised. Both of those patients had that coronary button translocated, and with that, we had good outcomes, no mortality in those patients. And we did not observe that, but again, our patient number is very small. This is a very small subset of patients.

Dr Anderson: This is obviously the problem. This is such a variable morphology of the source of regurgitation that any statistical analysis of it is inevitably plagued by the heterogeneity of the underlying problem even though it is the same, as it were, fundamental lesion, i.e. common arterial trunk. But the nature of the regurgitation is well known, impossible to analyse statistically unless you have thousands of cases which, thankfully, we do not.

Dr Myers: Absolutely.