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Original Article

Repairing the tricuspid valve in congenital heart diseases other than Ebstein's*

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Abstract The tricuspid valve is being increasingly recognised as an important safeguard to the heart with congenital heart disease. Both structural anomalies of the valve and functional burdens from other malformations of the right heart can lead to major haemodynamic consequences both upstream and downstream. The indications to surgically intervene on the tricuspid valve are evolving and vary depending on the malformation. The extant surgical techniques and their applications to corresponding frequent congenital anomalies of the tricuspid valve are reviewed.

Keywords: Tetralogy of Fallot; functionally univentricular heart; single ventricle; atrioventricular septal defect; Uhl's anomaly; Marfan syndrome; hypoplastic left heart syndrome

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THE TRICUSPID VALVE IS ANATOMICALLY AND physiologically the first door of the heart after blood flows into the right atrium. The importance of tricuspid valve competence or lack thereof cannot be underestimated. As an active safeguard and primary port of entry, it may affect all the cardiac structures and physiology downstream, or, by reciprocity, be influenced by cardiopulmonary physiological and/or anatomic structural abnormalities, such as left-to-right shunts at the atrial or ventricular level, pulmonary valve abnormalities, right ventricular dysfunction, arrhythmia, or abnormal interventricular septal interaction. Furthermore, for congenital heart surgeons, it is a unique window of access to the heart, through which many structures can be reached and repaired, such as a ventricular septal defect closure, in isolation or during tetralogy of Fallot and atrioventricular septal defect repair, thereby placing it at risk for iatrogenic injury through mechanisms of stretch, tear, or inadvertent suturing.

In patients with a morphologically normal tricuspid valve, functional tricuspid insufficiency may occur frequently, relating to abnormal right ventricular geometry and the alterations it creates on the annulus or subvalvar apparatus of the valve. In patients with congenital heart disease other than Ebstein's anomaly, malfunctioning of a dysplastic tricuspid valve is not so uncommon. Examples include straddling or tethering of the valve in various congenital abnormalities, Uhl's anomaly, double-orifice tricuspid valve, clefts as with atrioventricular septal defect, Marfan's syndrome, hypoplastic left heart syndrome, and hypoplastic or near atretic valves seen with pulmonary atresia/intact ventricular septum, and tetralogy of Fallot.

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In this review article, indications and techniques will be described to repair structurally abnormal, functionally distorted, or iatrogenically damaged tricuspid valves in congenital heart disease.

Straddling

With partial or complete overriding of the chordae tendinae and/or a papillary muscle across the ventricular septum with left-sided attachments on the ventricular crest, otherwise described as biventricular insertion of the tricuspid valve tensor apparatus, tricuspid valve straddling exists.[1] A concomitant ventricular septal defect or ventriculoatrial malalignment is universally present,[1] such as the very rare cases of double-outlet right atrium with intact ventricular septum and a straddling tricuspid valve.[2] Septation to achieve biventricular correction may be extremely challenging or even impossible with important straddling, pending

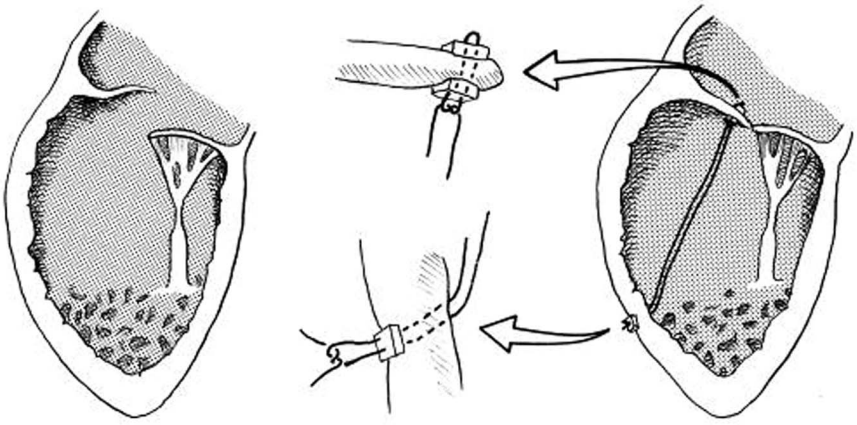

Figure 1.
Technique of chordal replacement. A single 4-0 expanded polytetrafluoroethylene suture is attached to the rim of the valve leaflet and the ventricular wall at the level corresponding approximately to the location of the papillary muscle origin, and reinforced at both points with Teflon felt pledgets. The insets indicated by arrows show the detail of leaflet and ventricular wall attachment. Reproduced with permission from Reddy et al. Repair of congenital tricuspid valve abnormalities with artificial chordae tendineae. Ann Thorac Surg 1998; 66: 172-176. Copyright © 1998, with permission from Elsevier.[3]

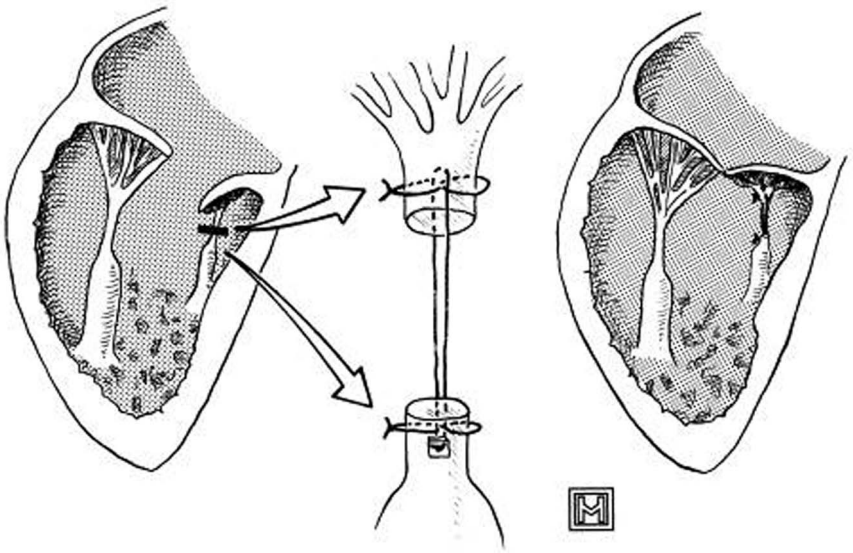

Figure 2.
Technique of chordal augmentation. The unrepaired valve, with short chordae, is shown on the left. The short chordae are transected and augmented, as indicated by the arrows, with one 2-0 expanded polytetrafluoroethylene suture, which is reinforced at its proximal and distal attachments with 4-0 polypropylene sutures. Reproduced with permission from Reddy et al. Repair of congenital tricuspid valve abnormalities with artificial chordae tendineae. Ann Thorac Surg 1998; 66: 172–176. Copyright © 1998, with permission from Elsevier.[3]

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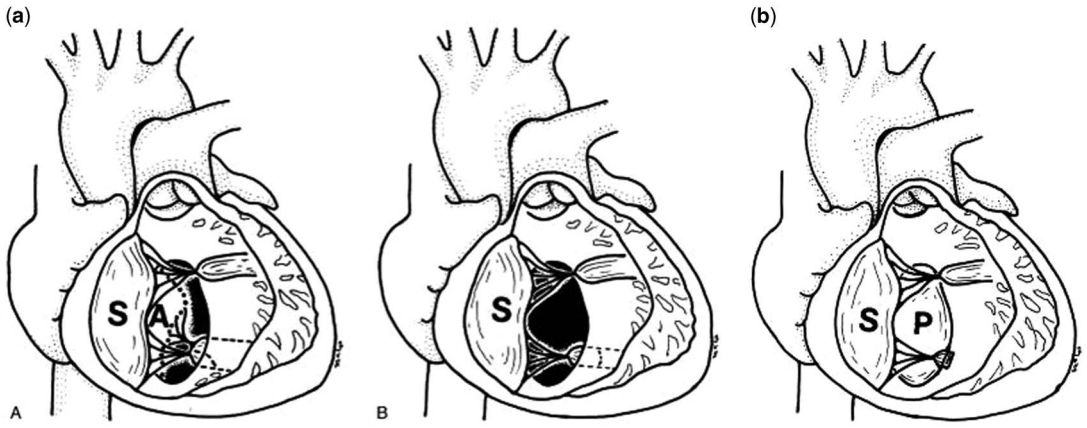

Figure 3.
*Straddling chordae tendineae of the posterior segment of the septal leaflet (S) of the tricuspid valve. (**a**) The straddling chordae insert into the posteromedial papillary muscle to the right of the chordae to the inferior segments of the mitral valve leaflets. The dashed line indicates the wedge resection of the right side of the posteromedial papillary muscle with its adherent straddling chordae. (**b**) The straddling chordae insert into an accessory papillary muscle. The dashed line indicates the line of resection of the papillary muscle. Reproduced with permission from van Son et al. Suspension of straddling tricuspid valve chordae into the appropriate ventricle. Ann Thorac Surg 1998; 65: 850–852. Copyright © 1998, with permission from Elsevier.*[4] *A = anterior leaflet of the mitral valve; P = posterior leaflet of tricuspid valve.*

iatrogenic injury, and severe tricuspid valve insufficiency (Figs 1–3).[3–5] Techniques to allow subsequent septation by closure of the ventricular septal defect include leaving the overriding chordae intact and incorporating them through the patch with or without a shortening plication-plasty, chordal and/or papillary muscle transfer, or wedge resection of part of the interventricular septum and transfer to the ipsilateral ventricle.[3,4,6] Results are good, notwithstanding an increased risk for complete heart block when papillary muscle transfer is attempted.[3] Although the various techniques almost always allow the surgeon to handle straddling and proceed with septation, curtain-like tricuspid chordae may present a contraindication to attempting biventricular repair in double-outlet right ventricle.[4,6] When specific anatomic lesions with severe straddling and/or borderline right ventricular dimensions exist, converting the heart to univentricular physiology may be safer in select cases.[4,7]

Uhl's anomaly

Uhl's anomaly is a very rare congenital abnormality whereby the right ventricular myocardial tissue is replaced by the fibroelastic tissue, and hence contractility is severely diminished.[8,9] The right ventricle is dilated, resulting in right heart failure and tricuspid valve insufficiency, and the majority of patients present in infancy, as survival into adulthood is rare without intervention (Fig 4).[5,9–14] Antegrade flow to the pulmonary arteries depends on right atrial contraction and paradoxical septal motion.[12] Surgical options include conversion to single-ventricular physiology and/or cardiac transplantation if the

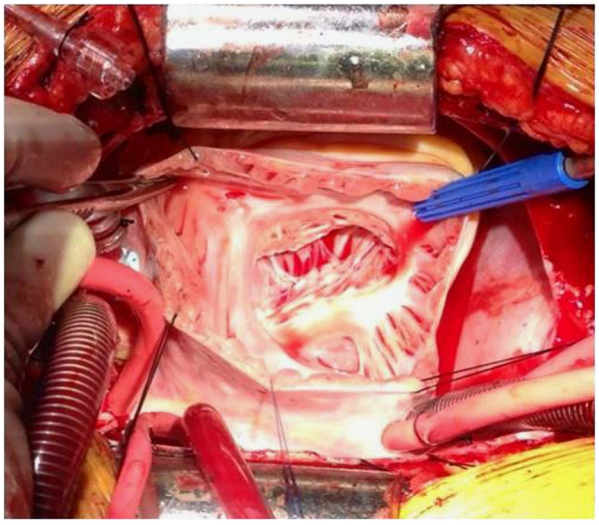

Figure 4.
Photograph showing a transatrial view of the tricuspid valve and thinned right ventricular cavity in a patient with Uhl's Anomaly in an 18-year-old female patient. There was significant tricuspid regurgitation with developed pressure in the right ventricle, which allowed tricuspid annuloplasty and pulsatile pulmonary artery flow.[5]

single-ventricle strategy fails.[9–11] In select patients, this initially involves atrial septectomy, tricuspid valve exclusion, and a bidirectional Glenn anastomosis, with or without right ventricular plication.[10,12–14] These variants of one-and-a-half repair may subsequently allow completion of total cavopulmonary anastomosis.[11,12] The results of surgery have been disappointing in the past, but more recent reports based on earlier diagnosis and a more aggressive approach towards single-ventricle palliation and right

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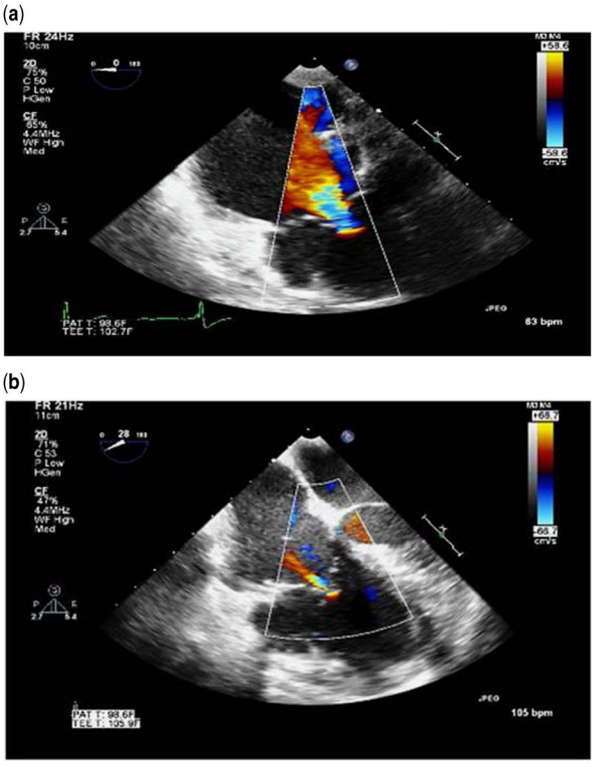

Figure 5.
*Colour flow Doppler echocardiograms showing preoperative severe tricuspid regurgitation (**a**) in a patient with Uhl's anomaly who underwent ringed annuloplasty with a # 28 ring. The postoperative result is shown (**b**) with significantly improved tricuspid valve function.*[5]

ventricular exclusion have resulted in patient survival, albeit still ending in eventual cardiac transplant candidacy.[9,12] Rarely, some degree of right ventricular function may be recruited for cardiac output, justifying an attempt at tricuspid annuloplasty to provide antegrade pulmonary flow (Fig 5),[5] before reverting to Fontan physiology, as has successfully been achieved by the senior author.

Double-orifice tricuspid valve

Double-orifice tricuspid valve, also known as atrioventricular valve duplication, is a very rare congenital malformation,[15,16] with <30 cases described in the literature,[16,17] either as an isolated lesion, or associated with other congenital anomalies such as mitral and pulmonary valve anomalies, atrial or ventricular septal defects, transposition of the great arteries, Ebstein's anomaly, or tetralogy of Fallot.[15,17,18] It was first described by Greenfild in 1876 for mitral and tricuspid valves.[19] There are three different types described: Type 1, with a simple leaflet fenestration; Type 2, with fusion of the leaflet tissue bridging the orifice and dividing into two orifices but with normal atrioventricular valve papillary muscles;

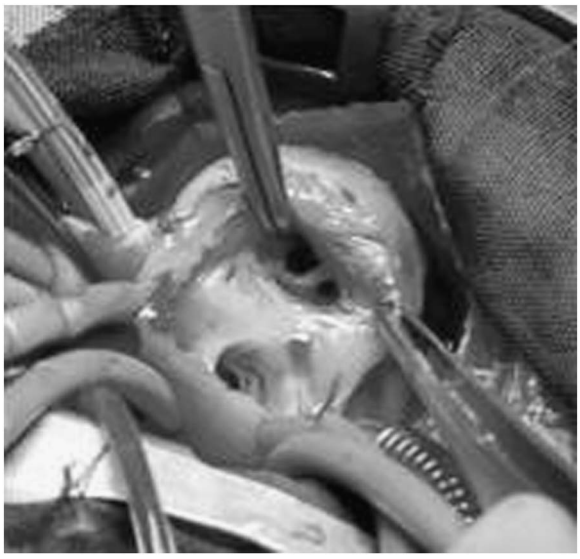

Figure 6.
Double-orifice tricuspid valve in an infant with tetralogy of Fallot. Intraoperative view with surgical orientation. Double-orifice tricuspid valve visualised through the right atriotomy. Reproduced with permission from Oppido et al. Double orifice tricuspid valve in an infant with tetralogy of Fallot. Ann Thorac Surg 2006; 81: 1121–1123. Copyright © 2006, with permission from Elsevier.[18]

and Type 3, with each orifice being supported by its own tension apparatus.[15,20] Wang et al[17] state that the accessory orifice of the tricuspid valve has its own independent chordae tendinae and mastoid muscle and should be treated surgically when discovered.

Patients may present with cyanosis or cardiac insufficiency, depending on the associated lesions and/or the functional status of the valve. Rarely, the valve is competent, although regurgitation and/or stenosis are more common.[16] Surgery, if at all indicated, will address stenosis or insufficiency accordingly. Associated congenital heart defects are of course addressed concomitantly, although it is often necessary to detach the anterior and septal leaflets for enhanced visualisation of other defects, followed by reattachment at the annular level.[18] The indication for surgery is usually dictated by correction of concomitant congenital cardiac anomalies, whose quality of repair will determine the outcome (Figs 6, 7).[5,18,20,21] The results of repair for the tricuspid valve anomaly are satisfactory in all reported series.[17,20,21]

Right atrioventricular valve in patients with atrioventricular valve septal defect

With unrepaired atrioventricular septal defect of any Rastelli classification, either with balanced or unbalanced ventricles, right-sided atrioventricular valve regurgitation may exist owing to clefts, from straddling/overriding, or from right ventricular and

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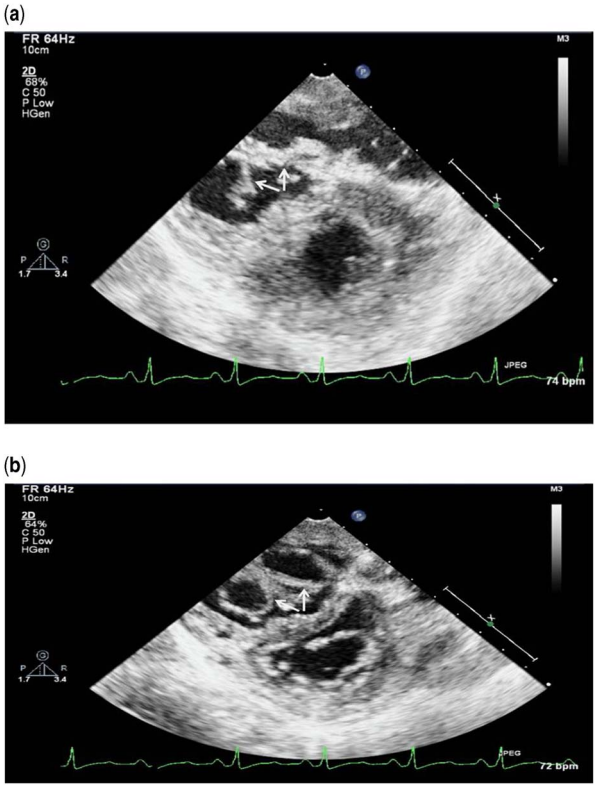

Figure 7.
*Epicardial echocardiograms show a double-orifice tricuspid valve in systole (**a**, valve closed) and diastole (**b**, valve open) in a 35-year-old patient who underwent resection of subvalvar right ventricular outflow tract obstruction. The double-orifice tricuspid valve forced a ventricular rather than an atrial approach for adequate resection because of the small orifices of the tricuspid valve.*[5]

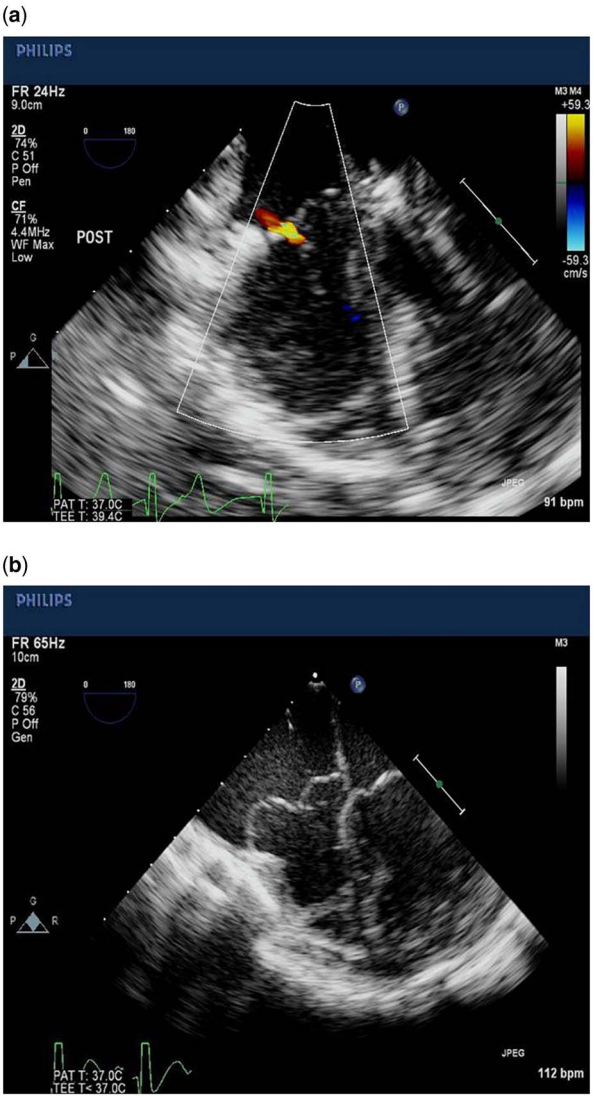

Figure 8.
*Echocardiogram shows an intact tricuspid prosthetic ring in a 21-month-old female with Marfan syndrome who presented with severe mitral and tricuspid regurgitation. The mitral and tricuspid annular dimensions allowed for adult-sized rings to be placed in both the tricuspid (28 mm) and mitral (30 mm) valves. (**a**) Upper segment colour flow Doppler echocardiogram shows the trivial amount of tricuspid regurgitation after ring annuloplasty. (**b**) The lower segment shows the anatomic configuration with excellent coaptation. The significant prolapse owing to elongated chordae remains, however.*[5]

annular dilatation owing to the ventricular septal defect shunt, poor ventricular function, or from pulmonary hypertension and backflow.[22–29] In patients after complete correction of atrioventricular valve septal defect, postoperative right-sided insufficiency may be an iatrogenic sequel from technical error at the time of repair and/or insufficient availability of the tissue to achieve adequate coaptation of the leaflets.

In unoperated atrioventricular valve septal defect, it is extremely uncommon to perform surgery for isolated right-sided atrioventricular valve insufficiency. Rather, the future tricuspid valve is repaired "en passant" while completing the atrioventricular septal defect correction. Furthermore, associated pulmonary valve and right ventricular pathology are often confounding, so that structural versus functional right-sided valve pathology may only be discovered during surgery. After complete correction of atrioventricular septal defect, when the septal defects, pulmonary hypertension, and ventricular function aspects may reasonably be excluded from the equation, indications to address isolated tricuspid valve insufficiency may include but are not limited to symptoms, reduced exercise intolerance, cyanosis, right-ventricle dilatation, right ventricle dysfunction, and/or the onset/progression of arrhythmias.[25] Techniques may be adapted to achieve valvar competence, including the De Vega annuloplasty, the Kay–Wooler plasty, ring annuloplasty, surgical creation of a double-orifice valve (Alfieri stitch), and leaflet patch enlargement plasty, depending on the underlying mechanism. With unbalanced ventricles,

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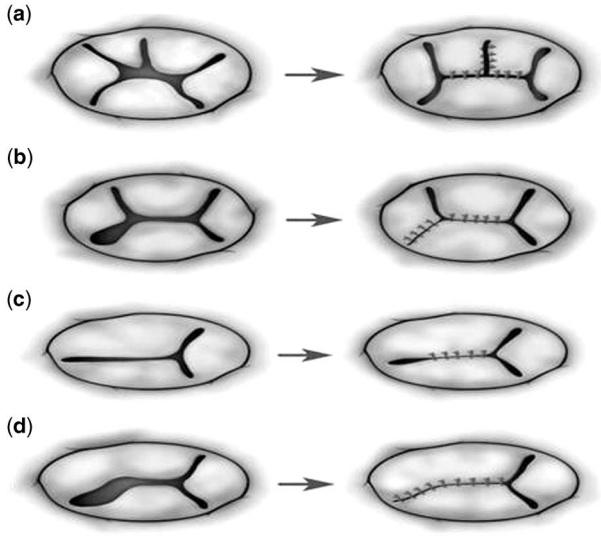

Figure 9.
*Two common types of edge-to-edge repair for repairing the morphologically common atrioventricular valve (**a**, **b**) and the tricuspid valve (**c**, **d**). (**a**) The superior and inferior bridging leaflets are sutured together, and the cleft is directly closed. (**b**) A non-coapting commissure is entirely closed in addition to edge-to-edge suture on the superior and inferior bridging leaflets. (**c**) The anterior and septal leaflets are sutured together making two effective orifices. (**d**) A non-coapting anteroseptal commissure is totally obliterated in addition to the edge-to-edge suture.[36] Reproduced with permission from Honjo et al. Atrioventricular valve repair in patients with single-ventricle physiology: mechanisms, techniques of repair, and clinical outcomes. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2011; 14: 75–84. Copyright © 2011, with permission from Elsevier.[38]*

with or without straddling or other concerns precluding septation, a one-and-a-half repair with a bidirectional cavopulmonary anastomosis or other variants of single-ventricle physiology should be considered.[22–31]

Marfan's disease

Marfan's disease is an autosomal dominantly inherited connective tissue disorder, of which the most common cardiovascular manifestations include aortic aneurysms and aortic valve insufficiency. However, all four valves may be affected, more often the mitral valve in order of importance, followed by the tricuspid valve with or without tetralogy of Fallot.[32,33] Mechanisms of tricuspid regurgitation include floppy leaflet movement, poor coaptation, prolapse, or elongated chordae.[32,33] Valve replacement or valve-sparing techniques, concomitant to mitral and aortic valve surgery, such as a combination of edge-to-edge Alfieri repair with chordal plasty[32] have led to satisfactory results continuing through mid-term follow-up (Fig 8).[5,34,35] To the best of our knowledge, isolated surgery for tricuspid valve disease in

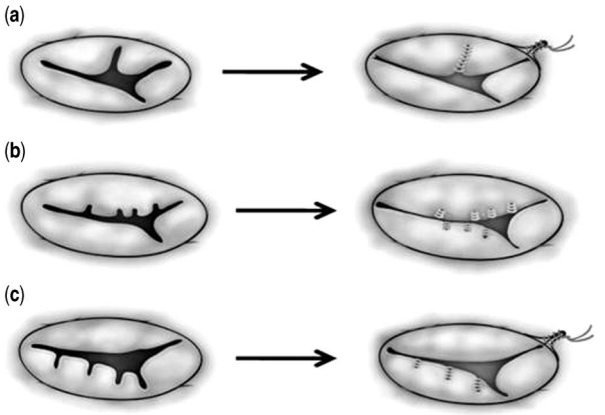

Figure 10.
*Surgical strategies to repair the dysplastic or dysmorphic tricuspid valve in hypoplastic left heart syndrome hearts. (**a**) Standard, direct cleft closure along with annuloplasty (functional commissuroplasty) on the anteroinferior commissure. (**b**) Repair of the irregular dysplastic leaflets. (**c**) Repair of dysmorphic leaflets by means of the edge-to-edge type repair. Reproduced with permission from Honjo et al. Atrioventricular valve repair in patients with single-ventricle physiology: mechanisms, techniques of repair, and clinical outcomes. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2011; 14: 75–84. Copyright © 2011, with permission from Elsevier.[38]*

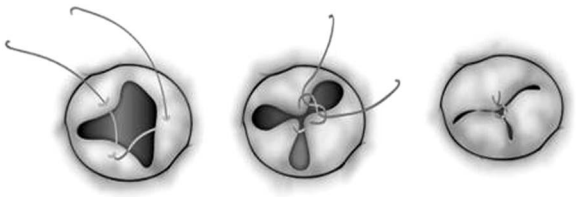

Figure 11.
The clover technique described by Alfieri et al[35] for repairing traumatic tricuspid valve insufficiency. The edges of all three leaflets are sutured together making three effective orifices. Reproduced with permission from Honjo et al. Atrioventricular valve repair in patients with single-ventricle physiology: mechanisms, techniques of repair, and clinical outcomes. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2011; 14: 75–84. Copyright © 2011, with permission from Elsevier.[38]

Marfan's disease has not yet been described in the literature, and indication remains uncertain.

Single ventricles and the tricuspid valve

Tricuspid valve insufficiency may be a complicating factor for patients with single-ventricle physiology and is not uncommon with hypoplastic left heart syndrome or unbalanced atrioventricular canal with a hypoplastic right ventricle. Functionally, chronic volume overload, annular dilatation and lack of leaflet coaptation, myocardial damage from subendocardial ischaemia resulting in papillary muscle dysfunction, or regional ventricular wall motion abnormality may

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all lead to tricuspid insufficiency. However, the vast majority of the insufficient tricuspid valves in the single ventricles have structural anomalies, including the bileaflet or quadrileaflet valves, clefts, accessory orifices, and prolapsed leaflets. Techniques of repair include the Alfieri stitch that creates a double-orifice valve or the Alfieri clover technique if multiple stitches are placed,[24,35] the Carpentier ring plasty, edge-to-edge repair,[36] suture annuloplasty, cleft closure, patch augmentation of a leaflet, chordal repair, and bridging annuloplasty,[37,38] among others (Figs 9–11).[3,5,35,36,38]

Fortunately, in neonates with single-ventricle physiology, tricuspid valve insufficiency is rarely severe at birth but progresses with time as an acquired lesion. When severe valvar regurgitation is present at birth, cardiac transplantation seems like a more reasonable option, when institutionally available.

With progressive worsening of tricuspid insufficiency, the indication and timing for an operation is difficult and ill-defined, as it presents a formidable potential insult to an already struggling single ventricle, not to mention the technical surgical difficulties in performing a reoperation and cross-clamping the aorta on a patient after a Damus–Kaye–Stansel anastomosis with arch reconstruction, as is the case in patients with hypoplastic left heart syndrome.[11] When tricuspid insufficiency is severe enough after Stage I palliation for hypoplastic left heart syndrome, interstage surgery before stage II may become necessary, but is a high-risk procedure.[39] If the regurgitation is mild-to-moderate, repair is preferably performed concomitant to cavopulmonary anastomosis at Stage II. As mentioned earlier, tricuspid valve insufficiency is rarely only a functional problem owing to volume overload. Therefore, at Stage II palliation for hypoplastic left heart syndrome, every effort should be made to correct structural deficiencies and achieve valve competence, rather to expect that the insufficiency will resolve by simple volume unloading with cavopulmonary anastomosis. If a patient presents to stage III Fontan completion with moderate-to-severe valve insufficiency, repair is warranted if it may be achieved without unreasonably long cardiopulmonary bypass or cross-clamp times,[11] as morbidity and surgical mortality may otherwise reach prohibitive levels. In cases where repair seems difficult or questionable, valve replacement could be a more reasonable and expeditious alternative. In patients with older atriopulmonary Fontan connections requiring takedown and Fontan conversion, repair may be indicated and has successfully been performed for mild-to-moderate insufficiency.[40]

Valve repair or replacement in patients with single-ventricle physiology remains a high-risk procedure,[39] with a reported surgical mortality of 17% and an ongoing potential for late death after hospital discharge. In a multi-institutional study from Australia including single-ventricle patients requiring not only tricuspid but also mitral valve surgery, independent predictors of mortality included a common atrioventricular valve, the need for postoperative mechanical support, and requirement for atrioventricular valve repair between palliative stages I and II.[39] Freedom from valve reoperation or embolism at 10 years was 56 and 70%, respectively. During follow-up, 11 of 48 surviving patients had moderate-to-severe regurgitation, and only 34 reached Fontan completion.[39]

Tricuspid valve repair in single-ventricle patients remains a challenge, but results are improving. Results are highly dependent on three-dimensional stereophysiological understanding of parachute valve anatomy and established newer techniques such as edge-to-edge valvuloplasty, central valvar stabilisation for atrioventricular canal, localised suture annuloplasty, selective use of pericardial augmentation patch valvuloplasty, and selective off-label use of extant prosthetic rings. Effort should be made to achieve durable repair in earlier surgical palliative stages, as repair or replacement become increasingly hazardous with subsequent redo surgical-site scarring, varying challenging physiology of single-ventricle staging, and superimposed diastolic ventricular dysfunction and myocardial scarring.[39] Although more judicious surgical timing may be part of the success, long-term results remain to be seen and will remain skewed by many valve-unrelated morbidities as is inherent to many older patients with single-ventricle physiology.

Functional tricuspid stenosis from annular hypoplasia

In patients with certain forms of tetralogy of Fallot and pulmonary atresia with intact ventricular septum, with or without a small right ventricle and a diminutive tricuspid valve annulus, biventricular strategy may not be indicated or may be hazardous, as antegrade flow to the pulmonary arteries cannot be secured.[29] In tetralogy of Fallot with a diminutive tricuspid valve annulus, the actual valve is often morphologically normal,[41] but pulmonary valve insufficiency or pulmonary hypertension lead to right ventricular failure. In patients with pulmonary atresia with intact ventricular septum, the tricuspid valve is often dysplastic.[25] In both entities, functional tricuspid valve stenosis occurs with diminished antegrade flow to the pulmonary arteries. In selected patients, this set-up represents a good indication for a one-and-a-half repair with a bidirectional cavopulmonary anastomosis, closed interatrial septum, and pulsatile antegrade pulmonary flow.[29,42] Compared with a true univentricular physiology as with

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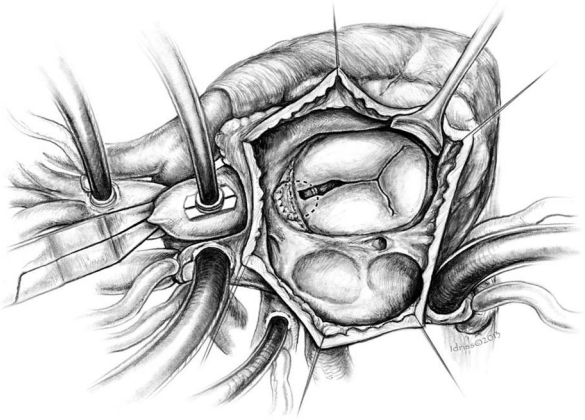

Figure 12.
In cases of septal and anterior leaflet tethering owing to adherences or impingement of the ventricular septal defect patch, poor coaptation between the septal and anterior leaflets can occur as shown in this drawing. The dotted line represents the approximate area of the previously placed ventricular septal defect. Reproduced with permission from Dodge-Khatami et al. Surgical techniques of tricuspid valve repair in patients without Ebstein malformation. In Giamberti A, Chessa M (eds): The Tricuspid Valve in Congenital Heart Disease. *Copyright © 2014, with permission from Springer.*[5]

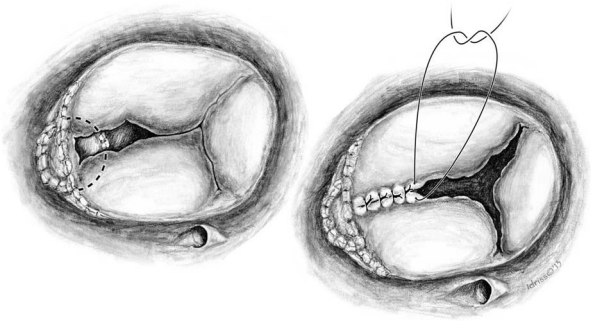

Figure 13.
On the left, the poor coaptation of the anterior and septal leaflets are noted and evaluated for the purpose of suture repair by leaflet apposition. On the right, sutures are being placed to coapt the anterior and septal leaflets, which will treat the commissural moiety and lend some structure to the valve apparatus. Individual sutures are placed towards the centre of the valve to achieve a competent valve, which can be assessed by a saline-filled bulb syringe. Aortobicaval cardiopulmonary bypass with cardioplegic arrest is performed to afford more accurate suturing and assessment of resultant valve function. Reproduced with permission from Dodge-Khatami et al. Surgical techniques of tricuspid valve repair in patients without Ebstein malformation. In Giamberti A, Chessa M (eds): The Tricuspid Valve in Congenital Heart Disease. *Copyright © 2014, with permission from Springer.*[5]

Fontan completion, this approach leaves all future options open, reduces the risk for an eventual future biventricular repair, while maintaining pulsatile pulmonary blood flow, normal oxygen saturations, and low right atrial pressures, although long-term follow-up data are lacking.[29,42,43]

Iatrogenic lesions after repaired tetralogy of Fallot

The incidence of moderate-to-severe tricuspid regurgitation after repaired tetralogy of Fallot varies between 11 and 19%.[44,45] Performing surgery solely to address a problematic tricuspid valve after repaired tetralogy of Fallot is extremely rare, although right ventricular failure and hepatic dysfunction would be clear indications.[46] More classical indications to intervene in repaired tetralogy of Fallot patients involve right ventricular dilatation from an insufficient and/or stenotic right outflow (pulmonary valve) or a residual ventricular septal defect, leading secondarily to functional tricuspid valve regurgitation from annular dilatation and lack of central coaptation in an otherwise morphologically normal valve, which may or may not need concomitant surgical attention. Structural tricuspid insufficiency may be an iatrogenic lesion during complete correction of tetralogy of Fallot, through inadvertent intraoperative injury to the valve and its apparatus, or distortion of the valve by anchoring stitches of the ventricular septal defect patch to the septal leaflet. For both functional and structural problems with the tricuspid valve, annuloplasty techniques are often required, with or without a stabilising prosthetic ring depending on growth considerations, and of course detachment of any tethered structures of the tricuspid valve apparatus has been caught and injured during ventricular septal defect closure.[30,31,41,46,47] In extreme cases, it may be necessary to detach part of the ventricular septal defect patch or recreate a ventricular septal defect, repair the tricuspid valve accordingly, and close the ventricular septal defect with a new patch. When insufficient tissue exists from prior iatrogenic damage to the valve, bicuspidalisation is a useful, reproducible, technically easy, and inexpensive method to restore valve competence.[41,48] The anterior and septal leaflets are brought together by placing a partial annuloplasty purse string suture, thereby compressing the posterior leaflet and forcing leaflet coaptation of a neo-bicuspidised valve (Figs 12, 13).[5,48]

Depending on the general condition of the patient and eventual superimposed haemodynamic and structural burdens on the heart after repaired tetralogy of Fallot, such as right ventricular dilatation and/or right outflow pathology, the risk for reoperation and the result of tricuspid valve repair will vary widely, ranging from a very safe procedure with an excellent result, or a very high-risk procedure (up to 16.7% reported mortality) and unstable valve

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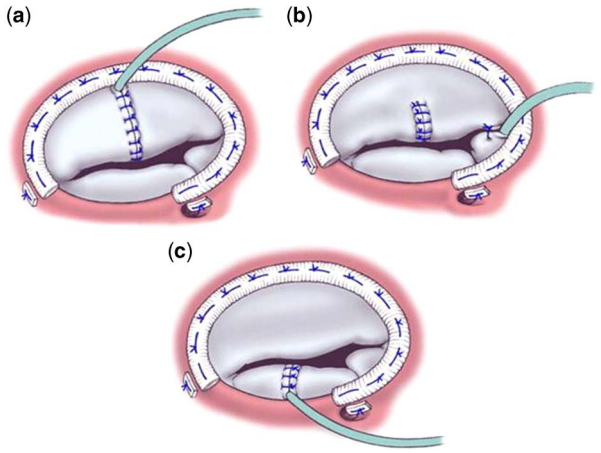

Figure 14.
*(**a–c**) In the presence of pacemaker or cardioverter defibrillator leads, repair techniques vary and depend on the degree of damage of leaflet(s). In the absence of extensive leaflet damage, valve repair is preferred and usually involves removing (incising) the lead away from the damaged leaflet, suture repair of the leaflet defect if present, or repositioning the lead by suture fixation in the recess of either the inferoseptal or anteroinferior commissure, and ringed annuloplasty is performed. Reproduced with permission from Said et al. Surgical management of congenital (non-Ebstein) tricuspid valve regurgitation. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2012; 15: 46–60. Copyright © 2012, with permission from Elsevier.*[25]

repair.[30,31,41,46,47] Follow-up data on the results and durability of tricuspid valve repair after complete correction of tetralogy of Fallot are scarce,[30,31,47] and it is difficult to evaluate whether the indication is for isolated surgery on the tricuspid valve or other cardiac structures. Namely, confounding factors related to right ventricular dimensions and geometry, right outflow functional status with stenosis and/or insufficiency, pulmonary hypertension, and eventual concomitant right bundle branch block or QRS widening may all impact the efficiency and results of surgery on the tricuspid valve. Indications to do anything to the tricuspid valve after tetralogy of Fallot repair are lacking and need to be defined, both in symptomatic and asymptomatic patients. Specifically, the general indications to intervene on patients after tetralogy of Fallot repair for right ventricular outflow tract procedures, in both symptomatic and asymptomatic patients, may not necessarily pertain to the tricuspid valve. Tricuspid valve insufficiency may resolve after insertion of a competent pulmonary valve if no anatomic tricuspid valve defects are present.[49] As overly conservative management eventually leads to irreversible valvar insufficiency and ventricular failure and secondarily to an increased risk for sudden death, it seems intuitive to intervene on the tricuspid valve concomitant to other structures of the right heart during redo surgery for repaired tetralogy of Fallot, although there are no data to support this contention. When relatively straightforward and reproducible techniques can restore valve competence and improve right-sided haemodynamics without taking unreasonable surgical risks with regard to bleeding, excessive bypass, and/or cross-clamp times, the authors support this holistic approach to right heart restoration.[41]

Iatrogenic lesions from intravenous pacemaker and defibrillator leads

Although epicardial pacing is increasingly being used and advocated for arrhythmia control in patients with congenital heart disease in whom steady somatic growth is a paramount issue, transvenous pacing and defibrillator leads have historically been placed and are increasingly encountered during follow-up, some of which will lead to iatrogenic lesions requiring therapy. Most transvenous leads cross the tricuspid valve and create insufficiency through mechanisms of traction, perforation, tethering, entanglement, fusion, and/or fibrosis.[25] If tricuspid valve function becomes severely compromised, repair of the valve and removal of the culprit pacing/defibrillator lead is indicated, with or without concomitant epicardial lead insertion if needed (Fig 14).[25] When repair is no longer possible, as reported in more than half of the surgical cases in the large series described by the Mayo Clinic, then tricuspid valve replacement is the only option.[25]

To conclude, the tricuspid valve is an important gateway to and safeguard of the heart, whose competence cannot be underestimated. It will either help preserve global cardiac function when functioning properly, or take the lead and escalate a cascade of deteriorating functional and structural anomalies. As with most structures in congenital heart disease with potential for growth, repair is always preferred to replacement and should be attempted, as congenital heart patients will invariably undergo somatic growth and should avoid unnecessary reoperations whenever possible. Indications to intervene solely on the tricuspid valve are lacking, as concomitant functional and structural cardiac lesions will often be the primary lesions to address, and whose direct or indirect consequences on tricuspid valve function and repair remain unclear. Accordingly, it is difficult to interpret or obtain any results of isolated tricuspid valve repair in congenital heart disease other than in patients with Ebstein's anomaly. Increased awareness by the congenital cardiac community regarding the importance of this previously "forgotten valve" and its crucial safeguard role[50] is leading more and more surgeons to perform earlier and more aggressive repair. Rather than waiting and watching, improving or fixing structural and/or functional pathology of

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the tricuspid valve earlier in life may hopefully lead to durable symptomatic improvement for the patient and save right heart function from irreversible failure.

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Conflict of Interest

None.

Ethical Standards

This is a review article and no research involved human or animal experimentation, so according to the guidelines of Cambridge University Press, this statement is omitted.

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