Cardiol Young 2005; 15: 417–421

ISSN 1047-9511

Original Article

Prolapse of the antero-superior leaflet of the tricuspid valve secondary to congenital anomalies of the valvar and sub-valvar apparatus: a rare cause of severe tricuspid regurgitation

Dominic J. R. Abrams,[1] Philip Kilner,[2] Janice A. Till,[1] Darryl F. Shore,[3] Babulal Sethia,[3] Rodney C. G. Franklin,[1] Alan G. Magee[1]

Departments of [1]Paediatric Cardiology, [2]Cardiac Magnetic Resonance and [3]Cardiac Surgery, The Royal Brompton & Harefield NHS Trust, London, United Kingdom

Abstract Congenital anomalies of the tricuspid valve, and/or its supporting apparatus, leading to severe tricuspid regurgitation are rare. Although well tolerated in early childhood, long-standing and progressive volume loading of the right heart leads to symptoms of decreased exercise tolerance, and may predispose to arrhythmias in the long term. We report three cases of severe tricuspid regurgitation related to anomalies of the cords supporting the antero-superior leaflet of the tricuspid valve. Shortened cords leading to tethering of the leaflet were seen in two cases, and hypoplasia of the leaflet in the other. In all cases, the regurgitant jet was directed posteriorly towards the coronary sinus and atrial septum. Surgical repair was possible in one case, while it proved necessary to replace the valve in a second. The third child is asymptomatic and under regular review.

Keywords: Atrioventricular valve; tendinous cords; chordae tendineae; anomalies; regurgitation

TRICUSPID VALVAR REGURGITATION SECONDARY to congenital anomalies is rare in childhood in the absence of Ebstein's malformation. It may be due to abnormal development of the cords, papillary muscles, or the valvar leaflets.[1] Long-standing regurgitation leads to dilatation of the right atrium, predisposing to reduced exercise capacity and arrhythmias. Right atrial enlargement with secondary annular dilatation increases the regurgitant fraction, leading to progression of the disease. The nature of the anomalies of the leaflets or the sub-valvar tension apparatus may dictate the direction of the regurgitant jet, leading to specific anatomical changes. Here we describe three children with anomalies of the cords supporting the antero-superior leaflet of the tricuspid valve, permitting the leaflet to prolapse into the right atrium during ventricular systole. Additional lesions included hypoplasia of the antero-superior leaflet, and shortened cords to both the septal leaflet and the zone of apposition between the septal and antero-superior leaflets. In all cases, the constellation of lesions created a posteriorly directed jet of tricuspid regurgitation, producing structural distortion of right atrial anatomy in two cases.

Correspondence to: Dr Alan G. Magee, Department of Paediatric Cardiology, The Royal Brompton & Harefield NHS Trust, Sydney Street, London SW3 6NP, United Kingdom. Tel: +44 20 7352 8121; Fax: +44 20 7351 8547; E-mail: a.magee@rbh.nthames.nhs.uk

Accepted for publication 23 March 2005

Case 1

A 11-year-old girl presented with a 2-year history of increasing dyspnoea, recurrent palpitations, and presyncope. On examination, her jugular venous pressure was raised, and a pansystolic murmur was audible at the left lower sternal edge. Her electrocardiogram showed right atrial enlargement, right axis deviation, first-degree heart block, with the PR interval measured at 174 milliseconds, and partial right bundle branch block, the duration of the QRS complex being 114 milliseconds. Chest radiography showed gross right atrial enlargement and subsequent cardiomegaly.

Cardiac imaging was performed using transoesophageal echocardiography (Fig. 1) and cardiac

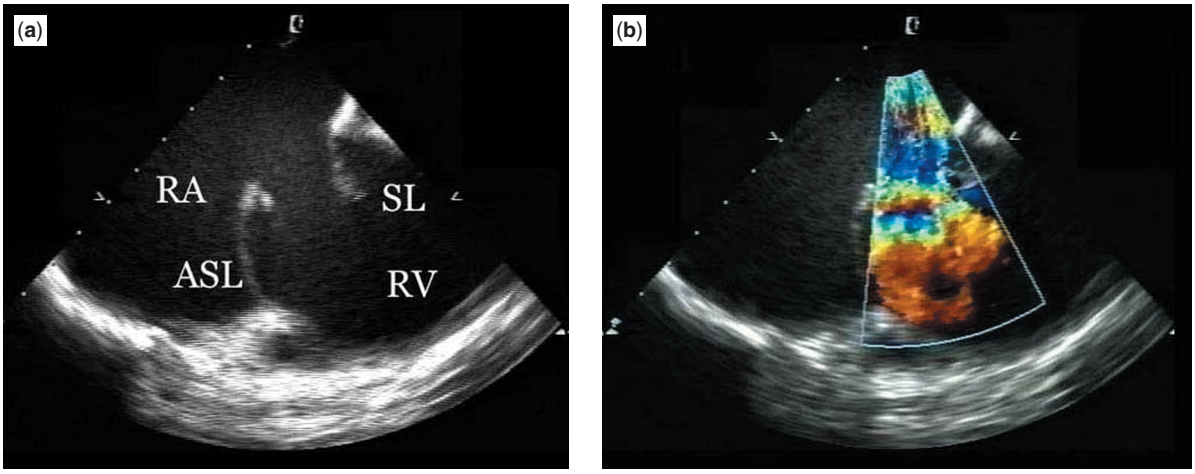

Figure 1.
*Cross-sectional (**a**) and colour flow Doppler (**b**) transoesophageal echocardiographic images seen in the transverse plane during ventricular systole. The right atrium (RA) and right ventricle (RV) can be seen separated by the antero-superior leaflet (ASL) and septal leaflet (SL) of the tricuspid valve. Marked prolapse of the ASL can be seen leading to a broad, posteriorly directed regurgitant jet depicted (**b**).*

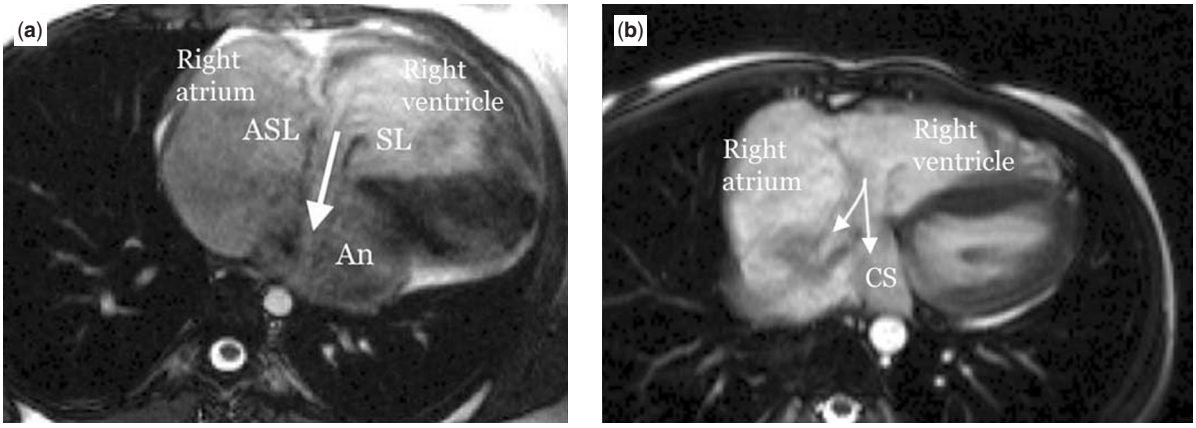

Figure 2.
*Cardiac magnetic resonance images seen in the mid-transverse plane from Case 1 (**a**) and Case 2 (**b**). In (**a**) prolapse of the antero-superior leaflet (ASL) gives rise to a broad posteriorly directed regurgitant jet (white arrow) entering a large aneurysm (An) extending beneath the inferior surface of the heart. In (**b**) prolapse of the ASL produces a Y-shaped regurgitant jet (white arrow) entering the right atrium and coronary sinus (CS). SL: septal leaflet.*

magnetic resonance (Fig. 2). The right atrium was markedly dilated, with the tricuspid valvar annulus measuring 48 millimetres. The cords of the septal leaflet were foreshortened, tethering the leaflet during ventricular systole, while the cords of the antero-superior leaflet appeared to be absent, allowing the leaflet to prolapse into the atrium. The net effect was failure of coaptation, allowing a jet of tricuspid regurgitation 17 millimetres wide to pass posteriorly towards the orifice of the coronary sinus and the atrial septum. The proximal coronary sinus was dilated to 16 millimetres, and appeared to give rise to a large diverticulum extending towards the left lower pulmonary vein.

Invasive electrophysiological testing demonstrated normal sinus and atrioventricular nodal function. Anterograde and retrograde atrioventricular nodal conduction was decremental, with no accessory pathway or dual atrioventricular nodal physiology evident. Tachycardia could not be initiated with programmed atrial stimulation in the presence of isoprenaline. At cardiac catheterisation, the mean right atrial pressure was 7 millimetres of mercury, and the right ventricular end-diastolic pressure was 6 millimetres of mercury. Injection of contrast showed that drainage from the left lower pulmonary vein was hindered by compression by the right atrial diverticulum.

At surgery, the right atrium was grossly enlarged, with a large diverticulum extending beneath the inferior surface of the heart, originating close to the mouth of the coronary sinus. On inspection of the valve, there was annular dilatation, but no evidence of dysplasia

https://doi.org/10.1017/S1047951105000867 Published online by Cambridge University Press

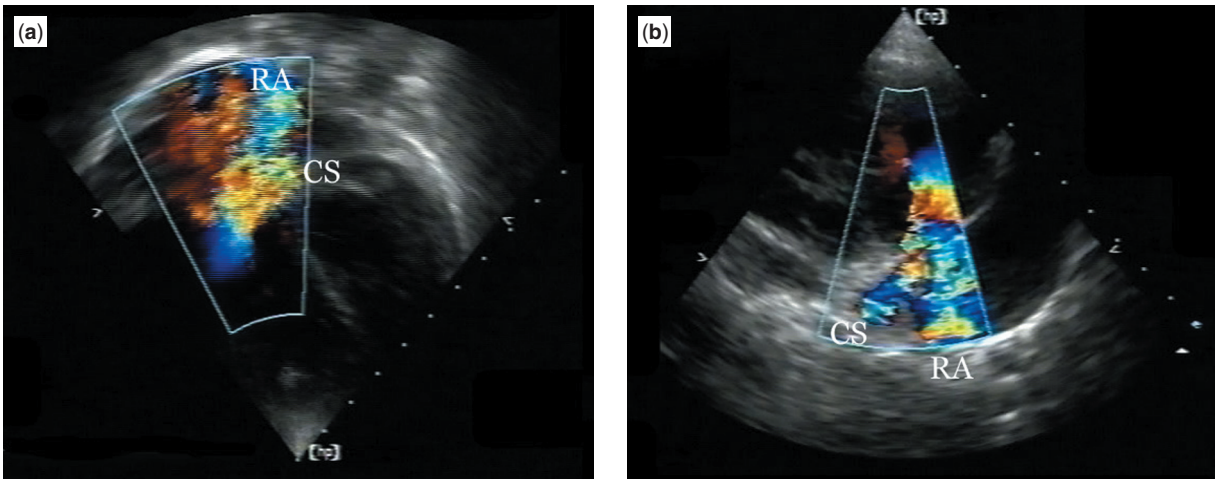

Figure 3.
*Apical (**a**) and short-axis (**b**) transthoracic echocardiographic images from Case 2 seen during ventricular systole. There is a broad Y-shaped, posteriorly directed jet of regurgitation into the right atrium (RA) and coronary sinus (CS).*

of the leaflets. The attachments of the septal leaflet appeared normal, although there was clearly deficient cordal attachment of the antero-superior leaflet. The cord close to the zone of apposition between the antero-superior and septal leaflets was short, and attached directly to the septum without a papillary muscle. A cord supporting the leaflet close to the zone of apposition with the inferior leaflet was clearly elongated, and the mid-section of the antero-superior leaflet was unsupported. A De Vega annuloplasty was performed, with shortening of the elongated cord to the antero-superior leaflet and plication of the unsupported mid-section of the leaflet. The right atrial diverticulum, and a section of the right atrial free wall, was excised. Residual tricuspid regurgitation on intra-operative transoesophageal echo was mild. The patient remains well 2 years later, with complete resolution of symptoms.

Case 2

An asymptomatic 8-year-old girl was referred for cardiac evaluation having been previously seen at another centre. She had suffered from persistent pulmonary hypertension as a neonate, requiring a period of cardiorespiratory support. Echocardiography at the time had demonstrated marked tricuspid regurgitation, which had largely resolved concomitant with resolution of her pulmonary hypertension. Subsequent to this, she was lost to follow-up for several years, representing following the detection of a murmur.

On examination she was normally saturated, with a right ventricular heave and a pansystolic murmur at the left lower sternal edge. Her electrocardiogram demonstrated a PR interval of 166 milliseconds, right atrial enlargement, and partial right bundle branch block. Cardiac magnetic resonance and cross-sectional echocardiography showed the right atrium and ventricle to be structurally normal but enlarged, with the tricuspid annulus measuring 44 millimetres. While the septal leaflet and its cords appeared normal, there was marked prolapse of the antero-superior leaflet, leading to complete failure of coaptation. A Y-shaped regurgitant jet was directed posteriorly into the right atrium and coronary sinus. This was markedly dilated, measuring 8 millimetres in diastole and 16 millimetres in systole (Figs 2 and 3). The patient remains asymptomatic, and is being followed conservatively at present.

Case 3

A patient was referred for evaluation of a murmur and cyanosis on the first day of life, at which time moderate tricuspid regurgitation was documented on cross-sectional echocardiography. She was managed conservatively, and by the age of 3 months, she was pink and asymptomatic. She remained well, with normal exercise capacity on regular testing, until the age of 12 years, when she reported reduced exercise tolerance, which could be demonstrated objectively with formal exercise testing. There were no symptoms of arrhythmia. At this time, echocardiography showed the right atrium and ventricle to be grossly enlarged, with a dilated tricuspid annulus measuring 50 millimetres. The left atrium and ventricle were functionally normal, but were compressed by the enlarged right heart. The septal leaflet of the tricuspid valve was tethered by shortened cords, while the cords of the antero-superior leaflet appeared to insert proximally, leaving a large section of flail leaflet, and permitting a broad jet of tricuspid regurgitation

https://doi.org/10.1017/S1047951105000867 Published online by Cambridge University Press

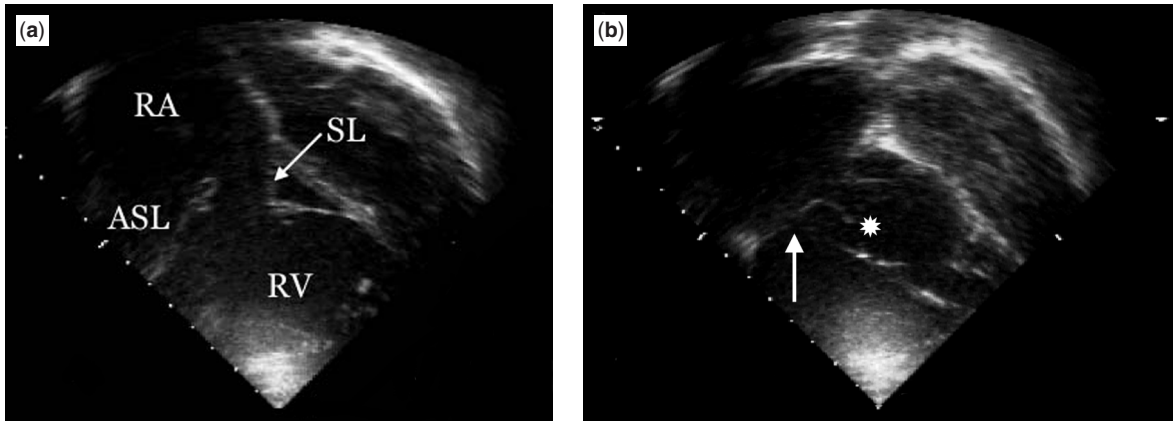

Figure 4.
*Cross-sectional echocardiographic images seen from the sub-costal right oblique view in Case 3. In (**a**), the septal leaflet (SL) appears tethered by shortened cords attached directly to the ventricular septum, while the antero-superior leaflet (ASL) prolapses into the right atrium (RA). (**b**) A close-up of the tricuspid valve, demonstrates the proximal insertion of the cords to the ASL (white arrow), allowing prolapse of leaflet tissue (*) beyond this point. RV: right ventricle.*

to be directed posteriorly (Fig. 4). In view of her symptoms, and the progressive dilatation of the right heart and tricuspid annulus, she was referred for elective surgery. At operation, the antero-superior leaflet of the valve was found to be hypoplastic, such that the valve could not be adequately repaired. It was replaced with a 33-millimetre bovine pericardial bioprosthesis. At follow-up 10 months later, the patient had recovered full exercise capacity, and was symptom free, with good function of the prosthetic valve.

Discussion

Isolated tricuspid regurgitation secondary to congenital anomalies of the tricuspid valve or its supporting tension apparatus is rare[1–6] outside the setting of Ebstein's malformation. In the first accurate account of this lesion, Becker et al. described a range of valvar lesions, including focal or diffuse valvar thickening, deficient development of cords or papillary muscles, improper separation of valvar components from the ventricular wall, and focal agenesis of valvar tissue.[1]

Cordal anomalies leading to significant regurgitation may relate to absent or lengthened cords, which allow prolapse of the associated leaflet into the right atrium, or shortened cords which tether the leaflet, preventing it from reaching the level of the atrioventricular junction so as to ensure valvar coaptation during ventricular systole. The direction of the regurgitant jet, and hence the distortion of right atrial anatomy, is dictated by the tricuspid valvar leaflet or leaflets involved. In one case, the cords appeared to insert proximally into the antero-superior leaflet, allowing the distal leaflet to prolapse into the right atrium. On direct inspection, the leaflet was found to be hypoplastic, thus preventing acceptable surgical repair.

In all three cases, the anatomical nature of the anomalies was such that the regurgitant jet was directed posteriorly towards the atrial septum and the mouth of the coronary sinus. This created significant dilatation of the coronary sinus in two cases, and the formation of a large diverticulum originating close to the mouth of the coronary sinus, extending beneath the inferior surface of the heart towards the left inferior pulmonary vein, in one. Such dilatation of the coronary sinus may be caused by overall enlargement of the right atrium due to volume loading, may be a direct effect of the regurgitant jet, or could be secondary to the presence of a persistent left superior caval vein. We were able to demonstrate retrograde flow in the coronary sinus in ventricular systole using both cardiac magnetic resonance imaging and echocardiography, a feature described only once previously, to the best of our knowledge, as an acquired phenomenon in an adult.[7] Whether this results in increased pressures in the coronary sinus, with an impact on coronary venous drainage, is unclear, albeit that none of our patients had evidence of myocardial ischaemia.

The pathophysiology of severe tricuspid regurgitation is marked and progressive dilatation of the right atrium, right ventricle, and tricuspid valvar annulus. Two of our three patients were symptomatic, and congestive heart failure has been reported by other authors.[2–4] As in our two patients who were symptomatic, surgical correction results in a significant improvement in symptoms and features of cardiac failure in all cases of congenital valve dysplasia reported to date. A recent series of adults with predominantly traumatic tricuspid regurgitation caused by flail

https://doi.org/10.1017/S1047951105000867 Published online by Cambridge University Press

leaflets reported a high associated morbidity and mortality, with a significant number of patients asymptomatic at presentation experiencing heart failure and arrhythmia. Surgical correction with low operative risk was associated with symptomatic improvement.[8]

Tricuspid regurgitation secondary to valvar or subvalvar anomalies may be more severe in the neonatal and infantile period. Elevated pulmonary vascular resistance will increase right ventricular afterload, worsening regurgitation across a dysplastic tricuspid valve. This may be especially severe in the neonatal period in conjunction with persistent pulmonary hypertension of the newborn[2,4] or with severe regurgitation, necessitating early surgical intervention. Symptoms of right-sided heart failure appear to resolve in the first few months of life as pulmonary vascular resistance falls to adult levels.[4] Persistence of an oval foramen or an atrial septal defect may lead to cyanosis due to right-to-left atrial shunting.[4] Tricuspid valvar dysplasia may also be seen in the fetal period, where it carries a poor prognosis secondary to gross right heart enlargement and pulmonary hypoplasia.[9]

One of our three patients reported recurrent symptoms of palpitation and pre-syncope, although the electrophysiology study was negative. Arrhythmias, specifically atrial fibrillation, have been described in conjunction with tricuspid regurgitation.[8] Paroxysms of atrial fibrillation not reproducible during electrophysiological testing may have been responsible for symptoms in this patient. Volume loading of the right atrium secondary to an atrial septal defect, in the absence of arrhythmia-induced remodelling, leads to prolongation of regional refractory periods, prolonged recovery times for the sinus node, and conduction block at the terminal crest, which may persist following closure of the atrial septal defect.[10] This may explain the very high incidence of atrial fibrillation reported in adults after repair of tricuspid regurgitation,[8] and stresses the importance of early repair before electrophysiological remodelling becomes irreversible.

Due to the small number of cases reported to date, there are no clear guidelines dictating the need for surgical intervention. The onset of symptoms, and/or demonstrable evidence of reduced exercise capacity or arrhythmias, would appear to be a good indication for surgery. In a series of adults with acquired tricuspid regurgitation, severe dilatation of the right heart was found to be predictive of a poor outcome,[8] and while it is difficult to directly extrapolate experience in adults with acquired tricuspid regurgitation to children, surgery for prognostic reasons in selected cases with progressive right heart enlargement on serial investigation may be warranted.

Surgical repair, rather than replacement, is possible in the vast majority of cases of tricuspid valve and cordal anomalies reported to date.[2–4] This was not possible in one of our patients due to hypoplasia of the antero-superior leaflet, although the use of a bioprosthesis ensured anti-coagulation was only necessary in the short term. In the setting of absent or shortened cords, implantation or cordal lengthening using polytetrafluoroethylene, coupled with annuloplasty and commissural valvoplasty, has produced excellent results in the immediate and medium term.[3,4] If severe right atrial enlargement is present atrial reduction surgery may also be required, as in one of our cases.

References

1. Becker AE, Becker MJ, Edwards JE. Pathological spectrum of dysplasia of the tricuspid valve. Arch Pathol 1971; 91: 167–178.
2. McElhinney DB, Silverman NH, Brook MM, Hanley FL, Stanger P. Asymmetrically short tendinous cords causing congenital tricuspid regurgitation: improved understanding of tricuspid valve dysplasia in the era of color flow echocardiography. Cardiol Young 1999; 9: 300–304.
3. Reddy VM, McElhinney DB, Brook MM, Silverman NH, Stanger P, Hanley FL. Repair of congenital tricuspid valve abnormalities with artificial chordae tendinae. Ann Thorac Surg 1998; 66: 172–176.
4. Katogi T, Aeba R, Ito T, et al. Surgical management of isolated congenital tricuspid regurgitation. Ann Thorac Surg 1998; 66: 1571–1574.
5. Kobza R, Kurz DJ, Oechslin EN, et al. Aberrant tendinous cords with tethering of the tricuspid leaflets: a congenital anomaly causing severe tricuspid regurgitation. Heart 2004; 90: 319–323.
6. Dearani J, Danielson GK. Congenital heart surgery nomenclature and database project: Ebstein's anomaly and tricuspid valve disease. Ann Thorac Surg 2000; 69: S106–S117.
7. D'Cruz IA, Taylor J, Kaiser J. Retrograde systolic flow in the coronary sinus due to selective tricuspid regurgitant jet entry. Echocardiography 2000; 17: 341–343.
8. Messika-Zeitoun D, Thomson H, Bellamy M, et al. Medical and surgical outcome of tricuspid regurgitation caused by flail leaflets. J Thorac Cardiovasc Surg 2004; 128: 296–302.
9. Hornberger LA, Sahn DJ, Kleinman CS, Copel JA, Reed KL. Tricuspid valve disease with significant tricuspid insufficiency in the fetus: diagnosis and outcome. J Am Coll Cardiol 1991; 17: 167–173.
10. Morton JB, Sanders P, Vohra JK, et al. Effect of chronic atrial stretch on atrial electrical remodeling in patients with an atrial septal defect. Circulation 2003; 107: 1775–1782.