Aberrant tendinous chords with tethering of the tricuspid leaflets: a congenital anomaly causing severe tricuspid regurgitation

R Kobza, D J Kurz, E N Oechslin, R Prêtre, M Zuber, P Vogt, R Jenni

Objective: To define the entity of tricuspid regurgitation caused by tethering of the tricuspid valve leaflets by aberrant tendinous chords.

Design: Retrospective study.

Setting: Tertiary care centre (university teaching hospital).

Patients: 10 patients with unexplained severe tricuspid regurgitation.

Methods: The last 13,500 echocardiographic studies from our facility were reviewed to identify patients with severe unexplained tricuspid regurgitation. Tethering was defined by the presence of aberrant tendinous chords to the tricuspid valve leaflets limiting the mobility of the tricuspid leaflet and resulting in incomplete coaptation and apical displacement of the regurgitant jet origin. Aberrant tendinous chords were defined as those inserting at the clear zone of the tricuspid leaflet and not originating from the papillary muscle. Patients fulfilling the diagnostic criteria for Ebstein’s anomaly were excluded.

Results: 10 patients with aberrant tendinous chords tethering one or more tricuspid valve leaflets were identified. There were short non-aberrant tendinous chords in seven patients, five of whom also had right ventricular or tricuspid annulus dilatation.

Conclusions: Tethering of the tricuspid valve leaflets by aberrant tendinous chords can be the sole mechanism of congenital tricuspid regurgitation. It is often associated with short non-aberrant tendinous chords, which may develop secondary to right ventricular or tricuspid annulus dilatation. Awareness of tethering as a cause of tricuspid regurgitation may be important in planning reconstructive surgery.
inserting at the clear zone of the tricuspid leaflet and originating from the ventricular wall instead of the papillary muscle.

- Incomplete coaptation and impaired mobility of the tricuspid leaflets with tricuspid regurgitation resulting from tethering of the tricuspid valve leaflets.

- Apical displacement of the origin of the regurgitant jet (fig 1A, B).

Short non-aberrant tendinous chords have been defined previously as short tendinous chords inserting at their normal location at the tricuspid leaflet free edge, connecting to the papillary muscle by a tendinous chord that is too short, resulting in incomplete closure of the tricuspid leaflets (fig 1C). In this situation the tricuspid leaflet tips close apically from the annulus.

### Table 1 Classification of tendinous chords of the tricuspid valve (adapted from Silver and colleagues)

<table>
<thead>
<tr>
<th>Chord</th>
<th>Site of insertion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leaflet chords</td>
<td></td>
</tr>
<tr>
<td>1. Rough zone chords</td>
<td>Rough zone and free margin</td>
</tr>
<tr>
<td>2. Free edge chords</td>
<td>Free edge</td>
</tr>
<tr>
<td>3. Deep chords</td>
<td>Rough zone</td>
</tr>
<tr>
<td>4. Basal chords</td>
<td>Basal zone (2 mm perimeter of the annular region)</td>
</tr>
<tr>
<td>Interleaflet chords</td>
<td></td>
</tr>
<tr>
<td>1. Fan shaped chords</td>
<td>Commissures between the leaflets</td>
</tr>
</tbody>
</table>

Patients with Ebstein's anomaly were excluded. Ebstein's anomaly was defined by the presence of apical displacement of the septal tricuspid valve leaflet by ≥ 8 mm²/m² body surface area and of an elongated and redundant anterior leaflet.

### RESULTS

Among the 13 500 echocardiographic studies analysed, severe tricuspid regurgitation was present in 229 cases (1.7%). Of these, 219 had recognised and defined primary or secondary aetiologies. The 10 patients (five male, five female, mean age 38 years, range 12–73 years) with initially unexplained severe tricuspid regurgitation all had aberrant tendinous chords tethering one or more tricuspid valve leaflets (table 2, figs 2, 3, and 4). Tethering with impaired leaflet mobility resulting in incomplete coaptation was the only mechanism of tricuspid regurgitation in three patients, whereas it was associated with short non-aberrant tendinous chords in seven (table 2). Among these 10 patients, secondary dilatation of the right ventricle (apical four chamber view) was present in four, and of the tricuspid annulus in seven. Five of the seven patients with short non-aberrant chords in addition to tethering had either right

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**Table 2 Morphological characteristics of patients with severe tricuspid regurgitation caused by tethering**

<table>
<thead>
<tr>
<th>Patient number</th>
<th>Sex</th>
<th>Age (years)</th>
<th>Other congenital cardiac abnormalities</th>
<th>A P syst / RA syst (mm Hg)</th>
<th>Triicuspid annulus diameter (mm, normal &lt; 31 mm)</th>
<th>Area-D (cm²)</th>
<th>Fac (%)</th>
<th>Origin of the TR jet displaced</th>
<th>Leaflets with tethering from aberrant chords</th>
<th>Leaflets with short primary chords</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>12</td>
<td>–</td>
<td>25</td>
<td>45</td>
<td>32.0</td>
<td>42</td>
<td>Apical</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>56</td>
<td>–</td>
<td>28</td>
<td>25</td>
<td>18.7</td>
<td>49</td>
<td>Apical</td>
<td>+</td>
<td>–</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>73</td>
<td>–</td>
<td>39</td>
<td>35</td>
<td>14.0</td>
<td>46</td>
<td>Apical</td>
<td>+</td>
<td>–</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>18</td>
<td>–</td>
<td>21</td>
<td>44</td>
<td>24.0</td>
<td>20</td>
<td>Apical</td>
<td>–</td>
<td>+</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>17</td>
<td>Superior sinus venous septal defect</td>
<td>15</td>
<td>39</td>
<td>20.1</td>
<td>48</td>
<td>Apical</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>41</td>
<td>AV/VA discordance (d-TGA)</td>
<td></td>
<td>21</td>
<td>27.1</td>
<td>12</td>
<td>Apical</td>
<td>–</td>
<td>+</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>35</td>
<td>AV concordance/VA discordance (d-TGA)</td>
<td></td>
<td>52</td>
<td>40.9</td>
<td>48</td>
<td>Apical</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>8</td>
<td>M</td>
<td>62</td>
<td>Situs inversus with mesocardia; AV/VA discordance (d-TGA)</td>
<td>48</td>
<td>33.0</td>
<td>69</td>
<td>Apical</td>
<td>–</td>
<td>–</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>43</td>
<td>Pulmonary stenosis, ASD II</td>
<td></td>
<td>45</td>
<td>35</td>
<td>14.2</td>
<td>Apical</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>10</td>
<td>F</td>
<td>20</td>
<td>Pulmonary stenosis</td>
<td></td>
<td>30</td>
<td>13.5</td>
<td>44</td>
<td>Apical</td>
<td>–</td>
<td>+</td>
</tr>
</tbody>
</table>

Ant, anterior; Area-D, end diastolic area of the right ventricle obtained in the apical four chamber view (normal < 25 cm²); ASD, atrial septal defect; AV/VA discordance, atrioventricular and ventriculo-arterial discordance; AV, atrioventricular; Fac, fractional area change (normal > 25 cm²) defined as end diastolic area minus end systolic area divided by end diastolic area of the right ventricle; Post, posterior; RA, right atrium; RV, right ventricle; Sept, septal; syst, systolic; TGA, transposition of the great arteries; TR, tricuspid regurgitation; ΔP, difference in pressure; VA, ventriculo-arterial.
ventricular or tricuspid annulus dilatation, or both, while two had neither. Interestingly, short non-aberrant chords were not found in the patient with the largest right ventricle and tricuspid annulus (patient 7).

Tethering was associated with a congenital cardiac abnormality in six patients (table 2): one patient had a superior sinus venosus atrial defect; one had atrioventricular and ventriculo-arterial discordance (congenitally corrected l-transposition of the great arteries); one had atroventricular discordance; and three had tricuspid valve tethering (table 2). One patient had both tricuspid valve tethering and tricuspid regurgitation (patient 7).
concordance and ventriculo-arterial discordance (d-transposition of the great arteries) and underwent a Blalock-Hanlon atrioseptectomy at four days and a Mustard procedure at eight years; one had a situs inversus with mesocardia and atroventricular and ventriculo-arterial discordance (with d-transposition of the great arteries); and two patients had pulmonary valvar stenosis and had undergone commissurotomy at the ages of 5 months (patient 10) and 4 years (patient 9).

**DISCUSSION**

The normal anatomy of the tricuspid valve chord apparatus is complex and has been described in detail previously (summarised in table 1).7 8 10–12 Chords are classified by their morphology and site of insertion into the tricuspid leaflet. We have defined tethering anatomically and functionally by the presence of aberrant tendinous chords, which results in impaired mobility, incomplete coaptation, and apical displacement of the tricuspid leaflet tips during systole (fig 1B). As a result, the origin of the regurgitant jet is displaced towards the apex compared with other mechanisms of tricuspid regurgitation. Aberrant tendinous chords insert at the clear zone of the tricuspid leaflet—which is usually free of chord insertions—instead of the usual site of insertion at the free edge or rough zone, and connect to the endocardium instead of the papillary muscle. They are distinct from basal chords, which also arise from the endocardium, but insert into the leaflet only in close proximity to the annular region (maximum 2 mm).7 One or more of all three tricuspid valve leaflets may be affected by tethering. In fig 5 normal and aberrant tendinous chords are shown in pathology specimens.

Tethering of the tricuspid valve is a prominent feature of Ebstein’s anomaly. Among 41 patients with this disorder described in detail by Shiina and colleagues,7 35 had tethering of the septal and anterior tricuspid leaflets. However, diagnostic criteria for Ebstein’s anomaly set up by the same group rely on the apical displacement of the septal leaflet by at least 8 mm/m² and the presence of an elongated anterior leaflet.7 Tethering was not included in the diagnostic criteria, and the presence of tricuspid valve tethering has been a frequent source of mistaken diagnosis of Ebstein’s anomaly.7 Tethering can also be distinguished from short, non-aberrant tendinous chords as a cause of tricuspid regurgitation.7 Short non-aberrant chords connect the papillary muscle to the leaflet free edge, but are too short, resulting in incomplete closure of the tricuspid leaflets (fig 1C) and apical displacement of the leaflet tips from the annulus plane during systole. Thus, this pathology results in a functional defect similar to tethering, but by chords in an anatomically correct location. This architectural difference has implications for surgical repair; while aberrant tethering chords may simply be severed, thus restoring normal leaflet coaptation, resection of short non-aberrant chords would result in a flail tricuspid leaflet. Aberrant tendinous chords with tethering may occur alone or in combination with short tendinous chords. These two malformations were found to be associated in 70% of our cases. Both entities may affect each of the three tricuspid leaflets. However, distinguishing short chords as a primary congenital defect leading to tricuspid regurgitation from chords which have become too short in the course of right ventricular or tricuspid annulus dilatation from other causes may be difficult. This was apparent in our series, in which five of seven patients with short chords had associated dilatation of the right ventricle or tricuspid annulus. Thus in some cases a possible sequence of events might be the development of right ventricular or tricuspid annulus dilatation caused by severe tricuspid regurgitation from tethering as a primary defect, which then leads to short non-aberrant chords.

Many congenital tricuspid valve anomalies can cause tricuspid regurgitation. Independent of classification, which in addition may share some overlap owing to imprecise diagnostic criteria, awareness of tethering as a mechanism of tricuspid regurgitation is of clinical relevance and should be considered in cases of regurgitation with impaired leaflet mobility and apical displacement of the regurgitant jet origin. An updated classification of tricuspid malformations causing regurgitation is proposed in table 3.

Surgical repair of a regurgitant tricuspid valve is carried out according to the principles established for mitral valve repair.13 14 Reduction of the valvar annulus is often the only necessary measure to correct the great majority of secondary tricuspid valve insufficiency. In patients with tethering by aberrant chords, however, the reduction of the annulus alone may not reduce the regurgitation sufficiently. Although to date we have no experience of this, we believe that resection of the tethering chords could restore full mobility to the

**Table 3 Aetiology of tricuspid regurgitation**

<table>
<thead>
<tr>
<th>1. Primary tricuspid regurgitation</th>
<th>2. Secondary tricuspid regurgitation</th>
<th>3. Physiological tricuspid regurgitation</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>(A) Congenital</strong></td>
<td><strong>(B) Acquired</strong></td>
<td><strong>(C) Physiological</strong></td>
</tr>
<tr>
<td>Tricuspid valve prolapse</td>
<td>Endocarditis</td>
<td>Rheumatic valvular disease</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
<td>Rheumatic valve disease</td>
<td>Right ventricular infarction</td>
</tr>
<tr>
<td>Tricuspid valve dysplasia (grades I-III)</td>
<td>Right ventricular infarction</td>
<td>Right ventricular infarction</td>
</tr>
<tr>
<td>Abnormal number of leaflets</td>
<td>Heart transplantation</td>
<td>Heart transplantation</td>
</tr>
<tr>
<td>Atrioventricular channel</td>
<td>Carcinoid (or other tumours)</td>
<td>Carcinoid (or other tumours)</td>
</tr>
<tr>
<td>Cleft of a tricuspid leaflet</td>
<td>Trauma</td>
<td>Trauma</td>
</tr>
<tr>
<td>Left sided tricuspid valve in congenitally</td>
<td>Rheumatoid arthritis</td>
<td>Rheumatoid arthritis</td>
</tr>
<tr>
<td>corrected transposition of the great arteries</td>
<td>Papillary muscle dysfunction</td>
<td>Papillary muscle dysfunction</td>
</tr>
<tr>
<td>Pulmonary atresia with intact ventricular septum</td>
<td>Hypereosinophilic syndrome</td>
<td>Hypereosinophilic syndrome</td>
</tr>
<tr>
<td>Short tendinous chords</td>
<td>Thryoxinosis</td>
<td>Thryoxinosis</td>
</tr>
<tr>
<td>Aberrant tendinous chords with tethering of the tricuspid leaflets</td>
<td>Anorectic drugs</td>
<td>Anorectic drugs</td>
</tr>
<tr>
<td>RA, right atrium; RV, right ventricle.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
involved leaflet, which could then reach the coaptation plane. Although it may be possible in some cases to remove the tethering chords by pulling them out between primary chords with a nerve hook, the best way to access them may be by detaching the involved leaflet from the annulus, resecting the restricting chords, and then reinserting the leaflet to the annulus. Recognition of this entity and a precise definition of the tricuspid lesion are therefore essential to tailor a specific and appropriate surgical repair.

CONCLUSIONS
Tethering of tricuspid valve leaflets by aberrant tendinous chords may, in rare cases, be the sole mechanism of severe tricuspid regurgitation. Awareness of this mechanism is of clinical relevance and should be taken into consideration when planning reconstructive surgery of the tricuspid valve for tricuspid regurgitation.

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D J Kurz and E N Oechslin contributed equally to this work

REFERENCES

FROM BMJ JOURNALS

Myocardial perfusion scintigraphy and coronary disease risk factors in systemic lupus erythematosus
E M C Sella, E I Sato, W A Leite, J A Oliveira Filho, A Barbieri

Objective: To evaluate the prevalence of myocardial perfusion abnormalities and the possible association between myocardial perfusion defects and traditional coronary artery disease (CAD) risk factors as well as systemic lupus erythematosus (SLE) related risk factors.

Patients and methods: Female patients with SLE, disease duration >5 years, age 18–55 years, who had used steroids for at least one year were enrolled. Traditional CAD risk factors evaluated were arterial hypertension, diabetes mellitus, dyslipidemia, postmenopausal status, smoking, obesity, and premature family CAD profile. Myocardial perfusion scintigraphy was evaluated by single photon emission computed tomography with technetium 99m-sestamibi at rest and after dipyridamole induced stress.

Results: Eighty two female patients with SLE without angina pectoris with mean (SD) age 37 (10) years, disease duration 127 (57) months, SLE Disease Activity Index (SLEDAI) score 6 (5), and SLICC/ACR-DI score 2 (2) were evaluated. Myocardial perfusion abnormalities were found in 23 patients (28%). The mean (SD) number of CAD risk factors was 2.2 (1.6). There was a significant positive correlation between age and number of CAD risk factors. Lower high density lipoprotein (HDL) cholesterol level showed a significant association with abnormal scintigraphy. Logistic regression analysis showed that lower HDL cholesterol level and diabetes mellitus were associated with myocardial perfusion abnormalities. Current vasculitis was also associated with abnormal scintigraphy.

Conclusions: Lower HDL cholesterol level and diabetes mellitus have a significant influence on abnormal myocardial perfusion results found in asymptomatic patients with SLE. Current vasculitis was associated with abnormal myocardial scintigraphy. These data suggest that abnormal myocardial scintigraphy may be related to subclinical atherosclerosis.
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