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 Prête, 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.

A degree of tricuspid valve regurgitation is found on Doppler echocardiography in the vast majority of otherwise healthy adults. It can be classified as physiological, primary, or secondary. Primary tricuspid regurgitation results from congenital or acquired morphological anomalies of the tricuspid valve apparatus, while secondary regurgitation is caused by dilatation of the annulus because of right ventricular volume or pressure overload with otherwise normal tricuspid valve morphology. Tricuspid regurgitation may be classified as physiological in the absence of any primary or secondary form of tricuspid regurgitation (fig 1A).

Various congenital tricuspid valve malformations can cause tricuspid regurgitation. These include tricuspid valve prolapse, Ebstein’s anomaly, isolated dysplasia of the tricuspid valve, cleft tricuspid leaflet, unguarded tricuspid valve orifice, dysplasia of the left sided tricuspid valve in congenitally corrected transposition of the great arteries (atrioventricular and ventriculo-arterial discordance), atrioventricular septal defect (common atrioventricular junction), or pulmonary atresia with intact ventricular septum. Asymmetrically short tendinous chords to the septal leaflet have also been reported to cause congenital tricuspid regurgitation.

Here we report 10 patients in whom a diagnosis of severe tricuspid regurgitation caused by tethering by aberrant tendinous chords was made by echocardiography. Although tethering of the tricuspid valve has been found in tricuspid regurgitation from other causes, we are unaware of any previous reports describing this malformation as the sole mechanism of regurgitation. We will define the diagnosis of tricuspid valve tethering in the context of the normal and pathological anatomy of the tendinous chord apparatus of the tricuspid valve, and distinguish it from other malformations involving tethering.

METHODS
All 13,500 echocardiographic studies performed at our laboratory between January 1999 and March 2002 were reviewed to identify patients with unexplained severe tricuspid regurgitation. Videotape recordings of these examinations were re-evaluated to identify the mechanism of tricuspid regurgitation. This was possible because in all patients with unexplained findings, multiple and partially atypical imaging planes are routinely recorded by the same cardiologist (RJ) in our laboratory. All three tricuspid valve leaflets were thoroughly inspected for the presence of aberrant tendinous chords and short non-aberrant tendinous chords. The normal anatomical distribution of tendinous chords in the tricuspid valve is summarised in table 1.

Tethering was defined by the following anatomical and functional criteria:

- The presence of aberrant tendinous chords to one or more tricuspid valve leaflets (fig 1B). The tricuspid valve leaflets are divided into four zones: the free edge, the rough zone, the clear zone, and the basal zone. The rough zone describes the area between a leaflet’s free edge and its line of closure. It is termed “rough” because the majority of tendinous chords insert into it. In contrast, the clear zone is thin and translucent, stretching from the line of closure to the basal zone of the leaflets, where the basal chords insert. Aberrant tendinous chords were defined as...
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.

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 mm/m² body surface area and of an elongated and redundant anterior leaflet.⁷

**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 1** Classification of tendinous chords of the tricuspid valve (adapted from Silver and colleagues⁸)

<table>
<thead>
<tr>
<th>Chord</th>
<th>Site of insertion</th>
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<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>Commissures between the leaflets</td>
</tr>
<tr>
<td>Leaflets with tethering from aberrant chords</td>
<td></td>
</tr>
<tr>
<td>Ant</td>
<td>–</td>
</tr>
<tr>
<td>Sept</td>
<td>–</td>
</tr>
<tr>
<td>Post</td>
<td>–</td>
</tr>
<tr>
<td>Leaflets with short primary chords</td>
<td></td>
</tr>
<tr>
<td>Ant</td>
<td>–</td>
</tr>
<tr>
<td>Sept</td>
<td>+</td>
</tr>
<tr>
<td>Post</td>
<td>–</td>
</tr>
</tbody>
</table>

**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>LVEDD (mm)</th>
<th>LVEF (%)</th>
<th>Origin of 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>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>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>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>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>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>21</td>
<td>27.1</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>52</td>
<td>40.9</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>32.0</td>
<td>Apical</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>43</td>
<td>Pulmonary stenosis, ASD II</td>
<td>45</td>
<td>35</td>
<td>Apical</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>10</td>
<td>F</td>
<td>20</td>
<td>Pulmonary stenosis</td>
<td>30</td>
<td>30</td>
<td>Apical</td>
<td>–</td>
<td>+</td>
</tr>
</tbody>
</table>
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 atrioventricular...
concordance and ventriculo-arterial discordance (d-transposi-
tion of the great arteries) and underwent a Blalock-Hanlon
atrioventriculectomy at four days and a Mustard procedure at
eight years; one had a situs inversus with mesocardia and
atrioventricular and ventriculo-arterial discordance (with
d-transposition of the great arteries); and two patients had
pulmonary valvar stenosis and had undergone commissuro-
tomy 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). 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 dis-
placement 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
cords, which also arise from the endocardium, but insert
into the leaflet only in close proximity to the annular region
(maximum 2 mm). 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, 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
anterioleafflet. 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. Tethering can also be distinguished from short,
non-aberrant tendinous chords as a cause of tricuspid
regurgitation. Short non-aberrant chords connect the papil-
lar 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
coapation, 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.12 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>Category</th>
<th>Etiology</th>
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<tbody>
<tr>
<td>1. Primary tricuspid regurgitation</td>
<td></td>
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<tr>
<td>(A) Congenital</td>
<td></td>
</tr>
<tr>
<td>Tricuspid valve prolapse</td>
<td></td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
<td></td>
</tr>
<tr>
<td>Tricuspid valve dysplasia (grades I–III)</td>
<td></td>
</tr>
<tr>
<td>Abnormal number of leaflets</td>
<td></td>
</tr>
<tr>
<td>Atrioventricular channel</td>
<td></td>
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<tr>
<td>Cleft of a tricuspid leaflet</td>
<td></td>
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<tr>
<td>Pulmonary atresia with intact ventricular septum</td>
<td></td>
</tr>
<tr>
<td>Short tendinous chords</td>
<td></td>
</tr>
<tr>
<td>(B) Acquired</td>
<td></td>
</tr>
<tr>
<td>Aberrant tendinous chords with tethering of the tricuspid leaflets</td>
<td></td>
</tr>
<tr>
<td>Endocarditis</td>
<td></td>
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<tr>
<td>Rheumatic valve disease</td>
<td></td>
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<tr>
<td>Right ventricular infarction</td>
<td></td>
</tr>
<tr>
<td>Heart transplantation</td>
<td></td>
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<tr>
<td>Carcinoid (or other tumours)</td>
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<tr>
<td>Trauma</td>
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<tr>
<td>Rheumatic arthritis</td>
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<td>Radiation therapy</td>
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<td>Papillary muscle dysfunction</td>
<td></td>
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<tr>
<td>Hypereosinophilic syndrome</td>
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<td>Thyrotoxicosis</td>
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<tr>
<td>Anorectic drugs</td>
<td></td>
</tr>
<tr>
<td>2. Secondary tricuspid regurgitation</td>
<td></td>
</tr>
<tr>
<td>3. Physiological tricuspid regurgitation</td>
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</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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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, dyslipidaemia, 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.

REFERENCES