Objective: To evaluate pediatric atrioventricular valve repair with artificial chordae. Methods: Between February 2001 and January 2006, artificial chords were used in 21 children with severe mitral or tricuspid valve regurgitation. Patients with AVSD were excluded. Median age was 84 (1—194) months. Five patients had isolated tricuspid valve anomalies, 16 had mitral valve anomalies (associated tricuspid annular dilatation in 4). Tricuspid neochordae were placed to anterior (three patients) and septal (two patients) leaflets. Mitral neochordae were placed to anterior (15 patients) and posterior (1 patient) leaflets. Additional ring annuloplasties were performed in 12 (mitral 11, tricuspid 1), as well as 2 de Vega tricuspid annuloplasties. Patch insertion was used in acute endocarditis (tricuspid one). All echocardiographic studies were reviewed and analyzed by a single cardiologist. Results: No mortality occurred. Follow-up was complete (mean 28 ± 18 months). Two patients were reoperated, one for mitral ring dehiscence and one for recurring mitral valve insufficiency. Both valves were replaced by mechanical valve prosthesis. At last follow-up tricuspid insufficiency was mild (three) or moderate (two). Moderate insufficiency occurred due to remaining restriction of the septal leaflet after repair in endocarditis (one) and remaining prolapse of the anterior leaflet (one). Mitral insufficiency was absent (five), mild (seven), or moderate (two). Moderate insufficiency was caused by recurrent anterior leaflet shortening after valve repair in rheumatic valve disease (two). Valve restriction caused by artificial chordae was not found. Conclusions: Mitral and tricuspid valve repair with artificial chordae in children demonstrated acceptable results. Despite patient growth, valvular restriction by the artificial chordae was not observed ad mid-term follow-up.

Keywords: Congenital; Mitral valve repair; Artificial tendinous chords

1. Introduction

Artificial ePTFE (Gore-Tex®, W.L. Gore and Associates, Flagstaff, AZ, USA) chordae for mitral valve repair in adults were first reported by David in 1989 and by Frater a year later [1,2]. Since then further studies have reported good mid- and long-term results of mitral valve repair in adults using this technique [3–5]. Goretex neochordae are now widely accepted for mitral valve surgery in the adult.

In children conventional valve repair techniques in mitral valve disease may have limitations related to the wide spectrum of abnormalities of the valvular structures, the small size of the valve, and associated cardiac malformations. Prosthetic valve replacement is preferably avoided, because of associated risks of thromboembolism, cumadin-related hemorrhage, and need for reoperation with growth.

Artificial chordae were therefore introduced as an additional technique for mitral valve repair in children with prolapse of the anterior mitral valve leaflet and abnormalities of the subvalvular tension apparatus, and have shown good short- and mid-term results [6–9]. Uncertainty remains about the long-term durability of the artificial chordae and the biological adaptation of the heart.

In congenital tricuspid valve repair artificial chordae have also been used, especially when standard techniques would not have resulted in optimal tricuspid valve function. A few cases have been reported, where artificial chords were inserted because of leaflet tethering due to short chordae or a total absence of chordae [10–14].

However, in the growing child, artificial chordae may eventually result in restriction of leaflet motion. To assess this potential problem, we evaluated our results of mitral valve and tricuspid valve repair with artificial chordae in 21 children operated between 2001 and 2006.

2. Patients and methods

Between February 2001 and January 2006, 21 consecutive patients under 18 years of age were operated for mitral
Table 1
Patient characteristics with mitral valve regurgitation

<table>
<thead>
<tr>
<th>Pat.</th>
<th>Age (months)</th>
<th>Sex</th>
<th>Grade</th>
<th>MS</th>
<th>X-clamp (min)</th>
<th>Etiology</th>
<th>Chordae</th>
<th>Morphology</th>
<th>Annular dilatation</th>
<th>Associated TI</th>
<th>Previous operations</th>
<th>Associated cardiac lesions</th>
<th>NYHA class preop.</th>
<th># neochordae/location</th>
<th>Ring annuloplasty</th>
<th>Other mitral valve procedures</th>
<th>Associated cardiac procedures</th>
</tr>
</thead>
<tbody>
<tr>
<td>2</td>
<td>180</td>
<td>F</td>
<td>Severe</td>
<td>–</td>
<td>203</td>
<td>Marfan</td>
<td>Elongation</td>
<td>Prolapse AL A2</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>II</td>
<td>2 A2</td>
<td>Ring 34</td>
<td>–</td>
<td>David-operation</td>
</tr>
<tr>
<td>4</td>
<td>76</td>
<td>F</td>
<td>Severe</td>
<td>–</td>
<td>99</td>
<td>Rheumatic</td>
<td>Elongation</td>
<td>Prolapse AL A2-A3</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>4 A2-A3</td>
<td>Ring 26</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>5</td>
<td>135</td>
<td>F</td>
<td>Severe</td>
<td>–</td>
<td>123</td>
<td>Rheumatic</td>
<td>Short</td>
<td>Relative prolapse AL; short posterior leaflet</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>IV</td>
<td>2 A2</td>
<td>Ring 26</td>
<td>–</td>
<td>Aortic valve repair</td>
</tr>
<tr>
<td>6</td>
<td>72</td>
<td>M</td>
<td>Severe</td>
<td>–</td>
<td>47</td>
<td>Congenital</td>
<td>Elongation</td>
<td>Prolapse AL A1</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>II</td>
<td>4 A2-A3</td>
<td>Ring 26</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>7</td>
<td>180</td>
<td>F</td>
<td>Severe</td>
<td>+</td>
<td>152</td>
<td>Congenital</td>
<td>Normal</td>
<td>Prolapse AL; cleft AL</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>IV</td>
<td>6 A1-A2-A3</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>8</td>
<td>168</td>
<td>M</td>
<td>Severe</td>
<td>–</td>
<td>96</td>
<td>Rheumatic</td>
<td>Rupture</td>
<td>Prolapse AL A2-P2-P3</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>I</td>
<td>2 A2</td>
<td>Ring 28</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>9</td>
<td>144</td>
<td>F</td>
<td>Severe</td>
<td>–</td>
<td>115</td>
<td>Rheumatic</td>
<td>Elongation</td>
<td>Prolapse AL A2-A3</td>
<td>+</td>
<td>+</td>
<td>Coarctectomy</td>
<td>Coarctation</td>
<td>IV</td>
<td>4 A2-A3</td>
<td>Ring 30</td>
<td>de Vega plasty</td>
<td>–</td>
</tr>
<tr>
<td>10</td>
<td>1</td>
<td>M</td>
<td>Severe</td>
<td>–</td>
<td>97</td>
<td>Congenital</td>
<td>Short</td>
<td>Prolapse AL A2</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>III</td>
<td>4 A1</td>
<td>–</td>
<td>de Vega plasty</td>
<td>–</td>
</tr>
<tr>
<td>11</td>
<td>194</td>
<td>F</td>
<td>Severe</td>
<td>–</td>
<td>44</td>
<td>Congenital</td>
<td>Normal</td>
<td>Prolapse AL A1</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>II</td>
<td>4 A1</td>
<td>Ring 26</td>
<td>Bilateral Wooler Closure cleft AMVL</td>
<td>–</td>
</tr>
<tr>
<td>12</td>
<td>10</td>
<td>F</td>
<td>Severe</td>
<td>–</td>
<td>200</td>
<td>Congenital</td>
<td>Rupture</td>
<td>Prolapse AL A2</td>
<td>+</td>
<td>–</td>
<td>Coarctectomy</td>
<td>AOS</td>
<td>IV</td>
<td>6 PL P1-P2-P3</td>
<td>–</td>
<td>Bilateral Wooler Expanded Ross procedure</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>84</td>
<td>M</td>
<td>Severe</td>
<td>–</td>
<td>80</td>
<td>Myocarditis</td>
<td>Elongation</td>
<td>Prolapse AL A2</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>IV</td>
<td>6 A1-A2-A2</td>
<td>Ring 26</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>15</td>
<td>48</td>
<td>F</td>
<td>Severe</td>
<td>–</td>
<td>65</td>
<td>Ischemic</td>
<td>Elongation</td>
<td>Prolapse AL A2</td>
<td>+</td>
<td>–</td>
<td>Takeuchi correction (ALCAPA)</td>
<td>ALCAPA</td>
<td>IV</td>
<td>6 A2</td>
<td>–</td>
<td>Bilateral Wooler Bilateral Wooler</td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>184</td>
<td>F</td>
<td>Severe</td>
<td>–</td>
<td>74</td>
<td>Congenital</td>
<td>Elongation</td>
<td>Prolapse AL A2</td>
<td>+</td>
<td>–</td>
<td>Switch/AP banding coarctectomy</td>
<td>Taussig Bing/ coarctation</td>
<td>III</td>
<td>2 A2</td>
<td>–</td>
<td>PV commissurotomy</td>
<td>–</td>
</tr>
</tbody>
</table>

Pat., patient; MS, mitral valve stenosis; X-clamp, cross-clamping time; MI, mitral valve insufficiency; TI, tricuspid valve insufficiency (due to annular dilatation); NYHA, New York Heart Association Class; preop., preoperative; AL, anterior mitral valve leaflet; PL, posterior mitral valve leaflet; A1-A2-A3: segments anterior leaflet; P1-P2-P3, segments posterior leaflet; DORV, double outlet right ventricle; AOI, aortic valve insufficiency; AOS, aortic valve stenosis; PS, pulmonary valve stenosis; ASD, atrium septal defect; ALCAPA, anomalous origin of the left coronary artery from the pulmonary artery; switch arterial switch operation; AP, pulmonary artery; TVP, tricuspid valve plasty; PV, pulmonary valve.
and/or tricuspid valve anomalies. There were 8 males and 13 females. Median age at operation was 84 (1–194) months and mean weight was 26.3 ± 24.1 kg. Five patients were younger than 1 year at the time of operation. All patients had concordant atrioventricular and ventriculoarterial connections. Patients with AVSD were excluded from this study.

Isolated mitral valve anomalies were present in 16 patients (associated tricuspid annular dilatation in 4); all patients had severe mitral insufficiency. Mitral valve insufficiency was caused by prolapse of the anterior leaflet in 14 patients, by posterior leaflet prolapse in 1 patient, and by prolapse of both leaflets in another patient. Prolapse of the anterior leaflet was caused by congenital mitral valve anomalies with elongation of the chordae in eight patients, and by chordal rupture in one patient. One patient had normal chordae of the anterior leaflet, but a restricted posterior leaflet caused insufficiency (pseudo prolapse anterior mitral valve leaflet). Five patients had a history of acute rheumatic fever, with elongation of the chordae in three patients to the anterior leaflet, chordal rupture in one patient on both the anterior and posterior leaflet, and shortened chordae in another patient. Prolapse of the posterior leaflet in one patient, with congenital abnormal mitral valve, was caused by rupture of two chordae to the free edge of the posterior leaflet. Associated cardiac anomalies were observed in eight patients (Table 1).

Isolated tricuspid valve anomalies were found in five patients, all patients had severe tricuspid insufficiency. In four patients tricuspid valve insufficiency was caused by congenital valve anomalies associated with prolapse of the anterior leaflet in two patients, restrictive anterior and septal leaflet motion in one patient, and restrictive chordae of the septal leaflet in another patient. Acquired tricuspid valve regurgitation was present in one patient, with rupture of the chordae of the septal leaflet and a defect of the septal leaflet due to endocarditis. Associated cardiac anomalies were observed in two patients (Table 2).

Preoperatively, two (10%) patients were in New York Heart Association Class I, four (19%) in Class II, seven (33%) in Class III, and eight (38%) in Class IV. Class IV patients were dependent on mechanical ventilation and inotropic support.

2.1. Previous and associated procedures

Eight patients had undergone previous procedures for palliation or correction of associated cardiac anomalies: coarctectomy (four), arterial switch operation (one), pulmonary valvotomy (one), banding of the pulmonary artery (one), Takeuchi correction for ALCAPA (one), and DORV repair (two). Some patients had more than one previous procedure.

Repair of concomitant cardiac lesions at the time of mitral or tricuspid valve repair was performed seven times: David valve sparing root replacement (one), aortic valve plasty (one), extended Ross procedure (one), pulmonary valve repair (one), Glenn-shunt (one), modified Blalock-shunt (one), and placement of a Contegra* conduit (Medtronic Inc., Minneapolis, Minnesota, USA) between the right ventricle and the pulmonary artery (one).
2.2. Surgical technique

In all patients with mitral valve regurgitation an annuloplasty was performed: ring annuloplasty (11) or Wooler—Kay bilateral commissural plication annuloplasty (5). Prolapse of the anterior leaflet was corrected with insertion of artificial chordae (15 patients) and prolapse of the posterior leaflet was treated with quadrangular resection of the leaflet's midportion (P2) in one patient and with insertion of artificial chordae in another. The mean number of artificial chordae used for the anterior leaflet was 4.4 ± 2.4.

A total of six neochordae were used in one patient with prolapse of the posterior leaflet.

The associated tricuspid annular dilatation was repaired by a de Vega plasty (two patients), a ring annuloplasty (one patient), or closure and plication of the posteroseptal commissure (one patient).

In the patients with isolated tricuspid valve insufficiency, prolapse of the anterior leaflet (three patients) and prolapse of the septal leaflet (two patients) were corrected with the use of artificial chordae. The mean number of neochordae used for repair of the anterior leaflet was 4.6 ± 1.6 and for repair of the septal leaflet 4 ± 0. Additional techniques to correct prolapsing leaflets were closure of anteroseptal and/or posteroseptal commissures (two patients). In one patient with endocarditis of the tricuspid valve the septal leaflet defect was repaired with an autologous pericardial patch.

2.3. Technique of insertion of artificial chordae

Expanded PTFE sutures were used (Gore-Tex®, W.L. Gore and Associates, Flagstaff, AZ, USA). CV-5 sutures were used for older children, while CV-7 was typically used in neonates and small infants.

The ePTFE suture is first tied to the fibrous top of the papillary muscle and then the two ends are fixed to the free edge of the valve leaflet in a V-shape as described by El Khoury et al. [15]. For the anterior leaflet the length of the new chord is measured by bringing the free edge of the valve to the level of the anterior annulus. Alternatively, the length of the new chord can be compared to that of healthy, non-elongated (attached to a non-prolapsing part of the valve) native chords in the adjacent area. After correct length measurement both ends of the sutures are again passed through the free edge and tied on the ventricular side of the leaflet, to prevent the knot from interfering with the coaptation zone. Because the sutures are placed in a V-shape, one suture accounts for two new artificial chords.

2.4. Echocardiography

All patients had an echocardiographic evaluation pre-, intra-, and postoperatively. Intra-operative TEE echocardiography was used in all patients. Postoperative echostudies were performed at an interval of 3—6 months by the referring cardiologist. Restricted leaflet motion was defined as a lack of adequate coaptation of the leaflets in end-systole due to impaired movement of the leaflets. Different echocardiographic views were reviewed for each patient to look for signs of restriction. Restriction caused by artificial chordae was specifically sought after. All echo-Doppler studies were in retrospect analyzed by one pediatric cardiologist. Last follow-up echo studies were all performed within 3 months before closure of the study (February 1, 2006).

2.5. Data analysis

Actuarial freedom from reoperation was calculated by the Kaplan—Meier method and expressed as mean values ± SD. Statistical differences in weight were calculated by the paired Student t-test. Results of the echocardiographic studies were not statistically compared, because of the small numbers of patients and underlying valve anomalies.

3. Results

3.1. Mortality

There was no early or late mortality.

3.2. Morbidity

Median duration of ICU stay was 4 (1—14) days. One patient was reoperated for postoperative bleeding and one patient received a pericardial drain for pericardial effusion 4 days after valve repair.

No intra-operative complications related to the use of artificial chordae were observed (endocarditis, thromboembolism). None of the patients required a pacemaker implantation. One patient had a cerebrovascular accident after replacement of the mitral valve.

3.3. Follow-up

Follow-up data were complete in all patients. The mean observation period was 28 ± 18 months. Mean weight at follow-up was 33 ± 23.5 kg. Growth of the patients was established by a paired Student t-test and was found to be significant (p < 0.001).

Actuarial freedom from reoperation at 3 years is 90.5%. Twenty patients are now in New York Heart Association Class I, while one is in Class II.

3.4. Reoperation

Two patients (9%) required reoperation, 20 and 31 days after their initial operation.

One patient with a congenital valve anomaly (patient 10) had undergone mitral valve repair at the age of 11 days with placement of two chordae to the anterior leaflet and commissural annuloplication. There was no mitral regurgitation immediately after the operation. This patient was reoperated 20 days later for recurring mitral insufficiency due to failure of the commissural annuloplasty. A St. Jude® mechanical prosthesis (St. Jude Medical, Inc., St. Paul, Minnesota, USA), size 17 mm, was used to replace the mitral valve.

The other patient (patient 1) was a 14-year-old boy with a history of coarctectomy, DORV repair, and resection of a subvalvular aortic stenosis. Two months after correction of anterior mitral valve prolapse with artificial chordae and a
ring annuloplasty, severe mitral valve insufficiency recurred, due to dehiscence of the annuloplasty ring and tearing of the artificial chordae from the tip of to the papillary muscle. The anterior mitral valve leaflet and the ring were removed and replaced by a St. Jude mechanical valve prosthesis, size 33 (St. Jude Medical, Inc., St. Paul, Minnesota, USA).

3.5. Echocardiography

Echocardiography at last follow-up showed mitral valve insufficiency to be absent in five, mild in seven, and moderate in two (Fig. 1). Both patients with moderate mitral valve insufficiency had a history of rheumatic valve disease. Mitral valve insufficiency was absent or mild postoperatively but ongoing shortening of the anterior leaflet led to recurrent moderate valve insufficiency. Restriction due to the use of artificial chords was not found. In the four patients who had concomitant tricuspid annuloplasty, tricuspid insufficiency was absent or trivial at last follow-up.

In the five patients who had tricuspid valve repair, last echocardiography showed insufficiency to be mild in three and moderate in two (Fig. 2). Moderate insufficiency was caused by remaining restriction of the septal leaflet after repair for endocarditis and by remaining prolapse of the anterior leaflet. In these patients there was no evidence for restriction caused by the use of artificial chordae.

4. Discussion

Several techniques for mitral or tricuspid valve repair in children are known, and they are used to prevent replacement of the valve. Replacement of native valves in children is associated with high mortality rates, particularly in the neonates and young infants. Mortality and morbidity is associated with required coagulation, valvular dysfunction, and high risk of reoperation due to patient somatic growth. Mitral valve repair in children have shown to be feasible compared to mitral valve replacement. Since the first publication of Carpentier in 1976, there has been a lot of research toward mitral valve repair in children. Several authors have shown that conventional techniques are not sufficient for repair of the mitral valve and artificial chordal (re)placement adds to the actuarial freedom of reoperation [7,8]. There might only be one problem, which is the risk of developing restriction, due to patient growth, as the artificial lack the potential of growth.

In this series with short- to mid-term follow-up we have found no evidence of restriction of mitral or tricuspid leaflets caused by the use of artificial chordae in the growing child, even though a significant increase in body weight was observed. This is in correspondence with previous research [6—8,11].

We have operated 21 patients for mitral and tricuspid valve regurgitation, with different kinds of etiologies: congenital (elongated chordae, hypoplastic and short to very short chordae), rheumatic valve disease, endocarditis, and ischemic mitral valve disease. We aimed to demonstrate a range of pathologies can be successfully corrected using artificial chordae as pointed out by Minami et al. [7], resulting in a majority (81%) of our patients having no or only mild regurgitation of the mitral or tricuspid valve at the moment of last follow-up. Moderate regurgitation at last follow-up had several causes: recurrent shortening of the anterior leaflet in rheumatic mitral valve disease, prolapse of the anterior tricuspid valve leaflet, and remaining restriction of the septal leaflet of the tricuspid valve after endocarditis.

Two patients required reoperation because of recurring regurgitation of the mitral valve. In these patients mitral valve replacement was performed mainly because of failure of the annuloplasty.

Our actuarial freedom from reoperation of 90.5%, at 3 years has been similar as reported by Minami et al. [7].

In summary, ePTFE artificial chordae is a useful tool for pediatric mitral and tricuspid valve repair. Leaflet prolapse caused by elongated or ruptured chords can be corrected by inserting artificial chords. Sometimes in these cases chordal shortening has been performed in other studies, which have shown high rates of recurrent elongation or rupture of shortened chordae [16,17]. Therefore, we chose to insert artificial chords. Absent, hypoplastic, and short chordae can also be replaced by ePTFE chordae as they can also be used to replace destructed or absent chords in acute or healed endocarditis.
Adequate leaflet area is needed for successful outcome of this maneuver, although pericardial patch insertion may in some instances be used to augment the leaflet surface. Theoretically, the use of artificial chords in growing children may be limited by the progressive development of leaflet restriction. So far we have not found evidence of this potential complication, but the follow-up is limited and the number of neonates and young infants (in which the risk of restricted leaflet motion is probably greatest) is small.

References


Appendix A. Conference discussion

Dr A. Takriti (Damascus, Syria): I would like to ask you the type of rings you use in these children. Is it a resorbable ring?

Dr Boon: We use the physiomitral valve ring in these patients. The smallest we used was 26 and the largest we used was 34.

Dr D. Metras (Marseille, France): This is an exceptionally important experience. Within 5 years you have operated upon 21 patients with a prolapse, in many cases of congenital. And considering that five of them in particular were below 1 year, do you consider you can answer the good question you have initially given: Will this lead to restriction? Don’t you think you have to wait more in the growth of the child to really answer this question?

Dr Boon: I think that the patients under the age of 1, they are the most important children of this whole group. Because we have one child who we have followed for 4 years, and that’s the patient who had prolapse of the posterior leaflet. And now, 4 years after that, there is still mild regurgitation in the mitral valve leaflet when the patient has his weight three-fourths increased. So you might say that there is a significant growth and we have a good result with that.

Dr Metras: Probably the short follow-up. The youngest were operated, then best is to prove that there is no restriction.

Dr Boon: Yes, I guess so. But there was one patient who had the mitral valve replacement.

Dr H. Safranen (Helsinki, Finland): Did you ever estimate from preoperative echo what is the need for shortening of the chordae, how long the chordae should be?

Dr Boon: We did not measure the chordae or the length of the chordae preoperatively. We only looked at the mechanism of the insufficiency. So if there was a prolapse of the anterior leaflet, or if not a leaflet, we wanted to insert the artificial cords.