Reoperations and late adverse outcome in Marfan patients following cardiovascular surgery

Thierry Carrel*, Lukas Beyeler, Aurelia Schnyder, Pia Zurmühle, Pascal Berdat, Jürg Schmidli, Friedrich S. Eckstein

Clinic for Cardiovascular Surgery, University Hospital, CH-3010 Berne, Switzerland

Received 29 October 2003; received in revised form 15 January 2004; accepted 21 January 2004

Abstract

Objectives: Anulo-aortic ectasia represents the most common cardiovascular manifestation requiring surgery in Marfan patients. Aim of this report was to analyze the type of presentation and the incidence of cardiovascular lesions and the clinical follow-up after initial surgery, mainly aortic root repair or replacement. Methods: Between 1/1990 and 6/2003 a total of 71 patients (mean age 29 ± 17 years, 8–65 years) received first surgical treatment at our institution. 69 received root repair or replacement. 22 patients presented with acute aortic dissection (31%), out of them, 3 pregnant females and 1 just after delivery. Composite graft replacement was performed in the majority of patients (61/71, 85%). Aortic valve sparing root repair was performed in 7 patients, supra-coronary graft with refixation of the aortic valve in 2 and replacement of the descending aorta in 1 patient. All patients underwent close clinical and imaging follow-up in a specialized outpatient consultation. Results: During a mean follow-up interval of 5.2 ± 1.8 years, 14 patients (20.5%) underwent a total of 27 aortic reoperations. Seven patients had one and 6 patients had up to 4 reoperations; 13/14 patients had chronic aortic dissection. There was no hospital mortality and no major cardiac or neurologic morbidity following reoperation. During follow-up, 2 patients suffered from acute type B dissection following aortic root surgery and 3 patients surprisingly died: 2 from a rupture of a normal-sized descending aorta and one from intracranial hemorrhage. These 5 patients had had uncomplicated primary aortic root operation. Conclusions: The incidence of reoperations is significantly higher in patients who presented initially with acute type A aortic dissection than in those with dilatation only. In addition, this survey demonstrates that unexpected fatal outcome may appear in the remaining native aorta following uncomplicated elective aortic root surgery, even if the aorta is normal-sized. A close follow-up of all Marfan patients is necessary to detect asymptomatic changes requiring surgery because complex elective redo-operations can be performed with a very low perioperative risk.

© 2004 Elsevier B.V. All rights reserved.

Keywords: Marfan syndrome; Aortic root; Reoperations; Outcome

1. Introduction

Dilated aortic root represents the most common cardiovascular manifestation requiring surgery in Marfan patients. Elective repair or replacement of the aortic root is the treatment of choice for these patients. Following successful aortic root surgery, a substantial number of patients may present various additional cardiovascular pathologies, e.g. aneurysms in the residual native aorta and less frequently mitral valve regurgitation, dilatative cardiomyopathy and more rarely coronary artery disease that requires operative treatment.

Residual or recurrent disease after initial repair, expanding aneurysm in a previously dissected but not resected aortic segment, pseudoaneurysms at anastomotic suture lines as well as ongoing degeneration of the aortic valve leaflets following a valve-sparing root repair represent potential reasons leading to reoperation at the level of the thoracic aorta [1–6]. There is a considerable amount of literature dealing with aortic surgery, but few reports have addressed the special problems of redo-surgery, especially in the context of Marfan syndrome [7–12].

Aim of this report was to analyze the type of presentation and the incidence of cardiovascular lesions and the clinical
follow-up after successful initial surgery, mainly aortic root repair or replacement.

2. Patients and methods

Between 1/1990 and 6/2003 a total 71 patients with Marfan syndrome (mean age 29 ± 17 years, 8–65 years) underwent primary surgery at our institution. 22 patients (31%) presented with acute aortic dissection (21 type A and 1 type B). Three female patients were pregnant at the time of type A acute aortic dissection and one suffered acute type B dissection immediately after delivery. All patients with aortic dissection were operated on an emergency basis [13]. The remaining 49 patients received elective surgery.

Table 1 summarizes the main patients’ demographic factors. The diagnosis of Marfan syndrome was established using the Gent nosology and a mutation analysis in occasional cases.

Composite graft replacement was performed initially for the majority of patients (61/71, 85%). Aortic valve sparing root repair was performed in 7, supra-coronary graft with refixation of the aortic valve in 2 and replacement of the descending aorta in 1 patient. In one patient, composite graft was combined to mitral valve repair. All patients underwent close clinical and imaging follow-up in a specialized outpatient consultation.

3. Results

Fourteen patients underwent a total of 27 reoperations. There were 7 male and 7 female patients with a mean age of 37 ± 16 years. Three redo-procedures were performed on the proximal aorta and 22 on the downstream aorta. One patient received mitral valve replacement and one cardiac transplantation. Mean interval between initial surgery and first redo was 5.2 ± 1.8 years but decreased to 2.8 ± 1.1 years when subsequent surgery following first redo was required. Seven patients had one and six patients had up to 4 reoperations; 13/14 patients who required a redo-procedure had chronic aortic dissection. The majority of these (12/14) could be performed on an elective basis. Table 2 presents some details of redo-operations. Interesting findings are demonstrated in Figs. 1 and 2.

There was no hospital mortality and no major perioperative cardiac or neurologic (stroke, paraparesis or paraplegia) morbidity in any of these 14 patients. These results compared favourably with those observed following primary elective surgery in Marfan patients: 3.9% mortality (2/49) following elective aortic root or ascending aortic arch surgery and 13.6% mortality (3/22) following repair of aortic dissection. Postoperative follow-up was complete. CT scan or MRI was performed before hospital discharge in patients following repair of aortic dissection. Every patient was seen 3 and 6 months postoperatively and subsequently every year in the specialized Marfan and outpatient aortic consultation. Two patients required additional percutaneous interventions. One patient had a pseudoaneurysm of the left coronary artery button 13 years following composite graft which was successfully coiled with a device currently used in patients with chronic atrial
fibrillation to occlude the left atrial appendage. The second patient underwent successful endovascular stenting of a pseudoaneurysm at the distal suture line following replacement of the descending aorta.

### 3.1. Late adverse outcome

During follow-up interval, 2 patients suffered from acute type B dissection 3 and 5 years following uneventful aortic root repair. The intimal tear was classically located below the left subclavian artery and was not considered to have a relationship with the previous surgery. Both patients were treated conservatively. Surprisingly, 2 younger patients (13 and 14 years old) died from rupture of a normal-sized, non-dissected descending aorta, 2 and 3 years following uncomplicated aortic root operation. Both had underwent minor surgery some days before rupture (one had inguinal hernia repair, the other partial excision and laser of a facial naevus flammeus). Arterial hypertension was not present in any of these two patients. A 49-year-old patient died from intracranial hemorrhage 18 months following uncomplicated aortic root replacement with a mechanical composite-graft. At that time he was on warfarin but INR value was in the therapeutic range.

### 4. Discussion

Recent reports have demonstrated an incidence of ongoing pathology of the aorta and/or appearance of late surgical complications requiring redo-surgery between 7 and 25%, depending on the type of prior operation, the nature of the disease and the extent of follow-up [1–6,13,14]. In the specific context of Marfan patients, the same incidence varies between 5 and 35% [9–12,15,16].

In the most historical and largest series, Crawford reported a series of more than 700 patients operated on the ascending aorta and aortic arch. Of this, 150 patients had previous cardiac or aortic operation (21%), the causes of
reoperations being progression of residual aneurysm, aortic dissection, graft stenosis or infection, false aneurysm, valvular insufficiency and coronary artery disease [4]. Kouchoukos reported the results with 172 aortic root replacement, including 16% of patients who had undergone previous aortic operations. In contrast to the findings of this series dealing with Marfan patients, Kouchoukos observed a rather long interval between first and second operation [17].

It is expected that patients with Marfan syndrome will require multiple interventions on the remaining native aorta. In a long-term observational study, Finkbohner found that the most common pattern of aneurysm repair was proximal ascending aeurysm repair followed by descending aortic surgery. The following variables predicted patients requiring second surgery: acute or chronic dissection at the time of initial surgery, hypertension and a history of smoking. Patients who received prophylactic aortic root repair were less likely to require subsequent aortic surgery [15].

Kazui recently reported on a series of 14 Marfan patients who required late reoperation at a mean interval of 8 years following composite graft replacement of the aortic root. Main problems were aneurysm at the site of coronary artery reimplantation and suture line aneurysms following repair of aortic dissection using the inclusion technique. They concluded that the open coronary artery button technique was the most appropriate way to reattach the coronary arteries to avoid late complications [8]. Similarly to our findings, acute type A aortic dissection was a frequent initial event in patients requiring reoperation. Distal aortic lesions (e.g. at the level of the aortic arch and/or descending aorta) were also frequently observed. To diminish some of the complications encountered, Kazui recommends a separate reattachment of the supra-aortic branches to exclude the maximum of pathologic aortic tissue. We fully agree with this recommendation in Marfan patients but not as a general rule when complete aortic arch repair is required in atherosclerotic aneurysms and aortic dissection.

Pseudoaneurysm at anastomotic suture lines is a rare but serious complication which can occur at any suture line following graft to aorta anastomoses or reattachment of coronary, supra-aortic, intercostal and visceral arteries and branches. This complication was well known following the inclusion technique during composite graft repair [17] and has decreased since open repair has been established. The mechanism of pseudoaneurysm includes most probably excessive tension on the suture line and persistent oozing or bleeding in the perigraft space when the aorta is wrapped around the graft. In the present series, both patients who developed pseudoaneurysms could be treated successfully with endovascular interventional techniques. Long-term observation will show if this less invasive option provides a durable repair.

Aortic valve sparing root repair has provided excellent clinical outcome and relatively few valve-related complications [16,18–20]. The function of the reconstructed aortic root remains unchanged in the majority of patients during early and mid-term follow-up. Reoperation rate is around 4% during a follow-up of approximately 3 years [21]. In Yacoub’s series, results were slightly better in the absence of dissection [18,19]. In the largest series on aortic root replacement reported by Gott, 30 day mortality was 1.5% for elective, 2.7% for urgent repair and 11.7 for emergency surgery [22]: composite graft was performed in the majority of patients in this series.

One of the most interesting question of this study is: can the necessity of redo-surgery be influenced by initial surgical behaviour? We could not eliminate the incidence of redo although we have adopted the following rules: (1) most complete eradication of the pathologic tissue during initial surgery, especially at the level of the aortic root, (2) extensive use of separate reattachment of the aortic branches during aortic arch and of the visceral arteries during abdominal aortic replacement [12], (3) open distal anastomosis and hemi-arch repair [23] as minimal requisite during repair of acute aortic dissection, (4) extensive use of the conventional and retrograde elephant trunk to facilitate subsequent operations [24], (5) restrictive use of the graft inclusion technique and (6) aggressive treatment of concomitant pathology, e.g. mitral valve regurgitation during initial.

5. Conclusions

Results obtained after elective primary surgery in Marfan patients are comparable to those obtained in non-Marfan patients; in the last 100 elective composite graft procedures, mortality was 0% compared to 3.2% (2/61) in Marfan patients. The necessity for reoperations is significantly higher in those patients who presented initially with acute type A aortic dissection. Unexpected fatal outcome may happen in the downstream aorta following uncomplicated elective aortic root surgery despite normal sized aorta. This type of events have not been described before and may be addressed very carefully. Marfan patients who require non-cardiac surgery associated with postoperative pains seem to be at an unforeseeable risk. A close follow-up of all Marfan patients is necessary to detect all potential asymptomatic changes requiring surgery. Complex redooperations can be performed with a low perioperative risk.

References

References

Appendix A. Conference discussion

Mr J. Pepper (London, UK): It is a very wide experience you have. I am interested in the individual reattachment of the head vessels. If you have problems that extend into the bifurcation, for example, of the right carotid and the right subclavian, do you prefer to approach that surgically or with a combination of stent and surgery, assuming this is an elective situation?

Dr Eckstein: We would perform a conventional operation with replacement of the diseased vessel. We don't have any experience in endovascular stenting in Marfan patients, so I can't comment on that. Do you have some recommendation or do you have an experience with stents?

Mr Pepper: A very small experience. When you replace more and more of the aorta with Dacron, the impedance, of course, is enormous. So the energy transfer from the left ventricle goes to the branch vessels off the aorta. So one is likely to have continuing problems. And as it gets more peripheral, we are beginning to use stenting, but we have a very limited experience.