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ORIGINAL ARTICLE

Aortic root replacement in young adults: disease characteristics and early outcome

Zile Singh Meherwal · Sathiakar Paul Collison · Ashok Gupta · Abhay Choudhary · Naveen Saraf

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Abstract
Introduction Aortic pathology requiring replacement of the aortic root is rare in young adults and maybe a group prone to atypical presentation and poorer outcomes. Here, we have studied the clinical features, pathological extent of aortic root disease and outcome of young adults undergoing aortic root replacement at this institution between 1995 and 2005.
Patients and methods Retrospective study of the patients who underwent aortic root replacement at this institution between 1995 and 2005 between the ages of 18 and 40 (n= 53). Preoperative, intra-operative and postoperative data were collected on a standardized proforma.
Results There were 48 males (90.5%) and five females (9.5%). Mean age was 32.3±0.7 years. 44% of patients had a preoperative diagnosis of either bicuspid or rheumatic aortic valve disease; 46 (87%) presented with chest pain and in 34 patients (64%) an aortic regurgitant murmur was audible. Most patients had aorto-annular ectasia, with 17 (32%) with aortic dissection. The dissecting flap arose at or near the sinotubular junction in 88% and terminated in the ascending aorta in 60%. 45 patients (84.9%) received modified Bentall's procedure; the rest underwent separate aortic valve and supra coronary aortic replacement. In hospital mortality was 1.8%. Follow up was 96% at mean follow up of 8.56 years after surgery.
Conclusion Aortic surgery may be performed in young adults with good results. The disease characteristics of aortic dissection in this age group are favorable. The use of the Bentall procedure or separate aortic valve and supra coronary ascending aortic replacement offers good early and late clinical outcomes.

Keywords Aortic root · Aortic dissection · Echocardiography

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Z. S. Meherwal (✉) · S. P. Collison · A. Gupta · A. Choudhary · N. Saraf
Escorts Heart Institute and Research Centre,
Okhla Road,
New Delhi 110025, India
e-mail: meherwal@hotmail.com

Introduction

Ascending aortic pathology requiring replacement of the aortic root, with or without concomitant aortic valve replacement is encountered rarely in young adults [1]. Recent literature indicates that similar aortic pathology may be seen, though rarely, even in the pediatric age group [2].

Arriving at a prompt diagnosis is one of the keys to obtaining a favorable outcome in diseases of the aortic root [3]. In older adults, numerous studies have defined the "typical" risk groups, clinical presentation, characteristics' of the pathological anatomy and outcomes [4]. However, such data is not available for the less commonly encountered, younger patients with these disorders. Anecdotal observations have suggested that younger patients represent a group prone to atypical presentations, delayed diagnosis and poorer outcomes than older groups of patients with similar pathologies. It is necessary to carefully describe the disease characteristics and clinical presentation of young adults with aortic pathology to clarify if these differences are actually present. Hence, in this report, we have studied the clinical features, pathological extent of aortic root disease and the surgical outcome of young adults undergoing aortic root replacement at this institution between 1995 and 2005.

Patients and methods

Patient selection and data collection

The medical records and computer database of this institution was searched to provide data regarding the patients who underwent aortic root replacement at this institution between 1995 and 2005 who were between the ages of 18 and 40 (n=53). Using a standardized data form incorporating standard definitions, data was collected retrospectively regarding preoperative and post-operative outcomes; operative data was obtained from our computerized operative record and echocardiographic follow-up data was obtained from the patients'outpatient records.

The consent of the Institutional Review Board was obtained and patient consent was waived in view of the retrospective nature of the study.

Preoperative evaluation

The preoperative evaluation of these patients consisted of a detailed clinical examination, standard poster anterior and lateral roentgenogram and transthoracic echocardiography during the early years of the study and additionally, computer tomography aided angiography of the thoracic aorta when it became available. Coronary angiography and transesophageal echocardiography were not performed. A duplex color doppler imaging of the femoral vessels was performed to assess the size and status of disease in the femoral arteries.

Surgical technique

The criteria for selecting patients to undergo separate aortic valve replacement and supra coronary ascending aortic replacement were those recommended by David et al, namely, in patients who had dilated (>50 mm) ascending aortas and normal or minimally dilated aortic sinuses [5].

Our surgical technique is standardized [6]. Aortic root replacement was performed by the classic or modified Bentall technique or by the Cabrol technique [7]. Cardiopulmonary bypass was instituted with femoral-atrial or aortic-atrial cannulation. In patients with dissection, arterial cannulation was always via the femoral artery. Moderate systemic hypothermia was used in those who had distal anastomosis during aortic cross-clamping. Patients having open distal anastomosis were cooled to 16°C with deep-hypothermic circulatory arrest. Retrograde cerebral perfusion was used during hypothermic circulatory arrest. The left ventricle was vented through the right superior pulmonary vein in all cases. Myocardial protection was achieved with antegrade and retrograde cold blood cardioplegia. Direct coronary implantation was performed when the coronary ostia were lifted well above the annulus. When the coronary ostia were close to the annulus, they were mobilized as buttons and implanted into the graft. The Cabrol technique was applied in some early cases, using separate 6-mm conduits to implant the coronary ostia into the conduit. In cases of aortic dissection, the conduit was cannulated after distal anastomosis, and cardiopulmonary bypass was re-instituted through this cannula. A composite graft with a Bjork-Shiley valve (Shiley, Inc., Irvine, CA, USA) was used in 10 (18.8%) patients, and a Carbomedics bileaflet valve (Carbomedics Inc, Austin, TX, USA) in 2 (3.7%) in the early period of the study. More recently, we used a composite Dacron graft with a St. Jude bileaflet valve (St. Jude Medical, Inc., St. Paul, MN, USA) in 41 (78%) patients. Gelatin-resorcinol-formaldehyde glue (GRF-glue; Fii, Saint-Just-Malmont, France) was used in all of the 17 patients with dissection to restore continuity between the separated layers of the aorta. The anastomosis was buttressed with Teflon strips to reinforce the fragile suture line. All patients were preoperatively, on the basis of the investigations, planned to undergo modified Bentall's procedure. However, in eight cases, intra operatively, the aortic sinuses of Valsalva were found to be only mildly dilated and in these cases aortic valve replacement and separate supra coronary ascending aortic replacement was performed.

Follow up

Clinical follow-up consisted of six monthly transthoracic echocardiography and yearly thoracic computer tomography. Follow-up was 96% at a mean of 8.56 years after surgery.

Results

Fifty three patients between the ages of 18 and 40 years were identified. This group of patients represents 35.8% (53 of 148 patients) who have undergone aortic root replacement at this institution between 1995 and 2005. In this group of patients, there were 48 males (90.5%) and 5 females (9.5%). The mean age was 32.3±0.7 years (range 18–40 years). As shown in (Fig. 1), only a minority of patients had been previously diagnosed to have a condition known to predispose to aortic root disease. Around 44% of patients had been diagnosed before presentation with either rheumatic heart disease or bicuspid aortic valve; no patient in the study had a preoperative diagnosis of Marfan's syndrome. Figure 2 illustrates the clinical presentation of the studied patients and highlights that in 46 patients (87%), the initial symptom was of chest pain (either mild or severe in nature) and in 34 patients, (64%) the murmur of aortic

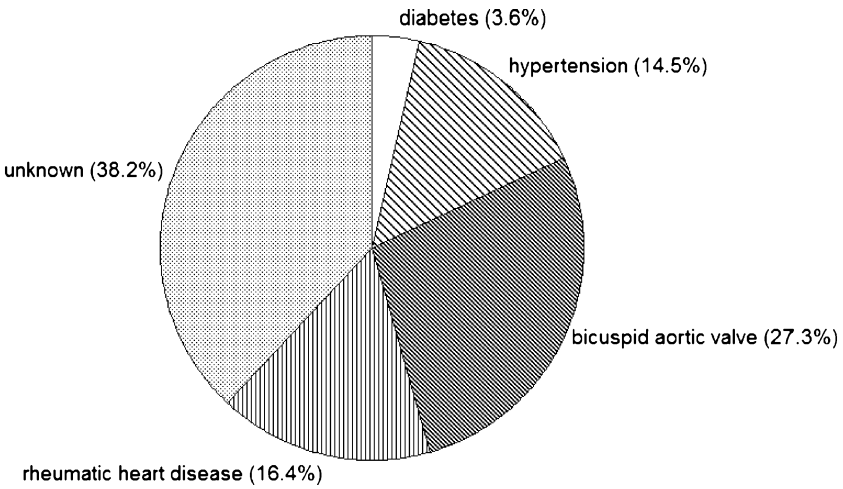

Fig. 1 Predisposing factors for the aortic root disease

ension, syncope and palpitations are uncommon, even in patients with aortic dissection.

Table 1 presents the aortic pathology found in these patients. Around 44% of patients had either bicuspid aortic valve or rheumatic aortic disease. The vast majority was found to have aorto-annular ectasia, the forme fruste of Marfan's syndrome, and of these roughly half presented with aortic dissection. None of the patients with bicuspid or rheumatic heart disease had aortic dissection (Table 2).

Table 3 details the anatomic extent of the aortic disease in the 17 patients with aortic dissection. Of note, the site of origin of the flap was at the sinotubular junction or just

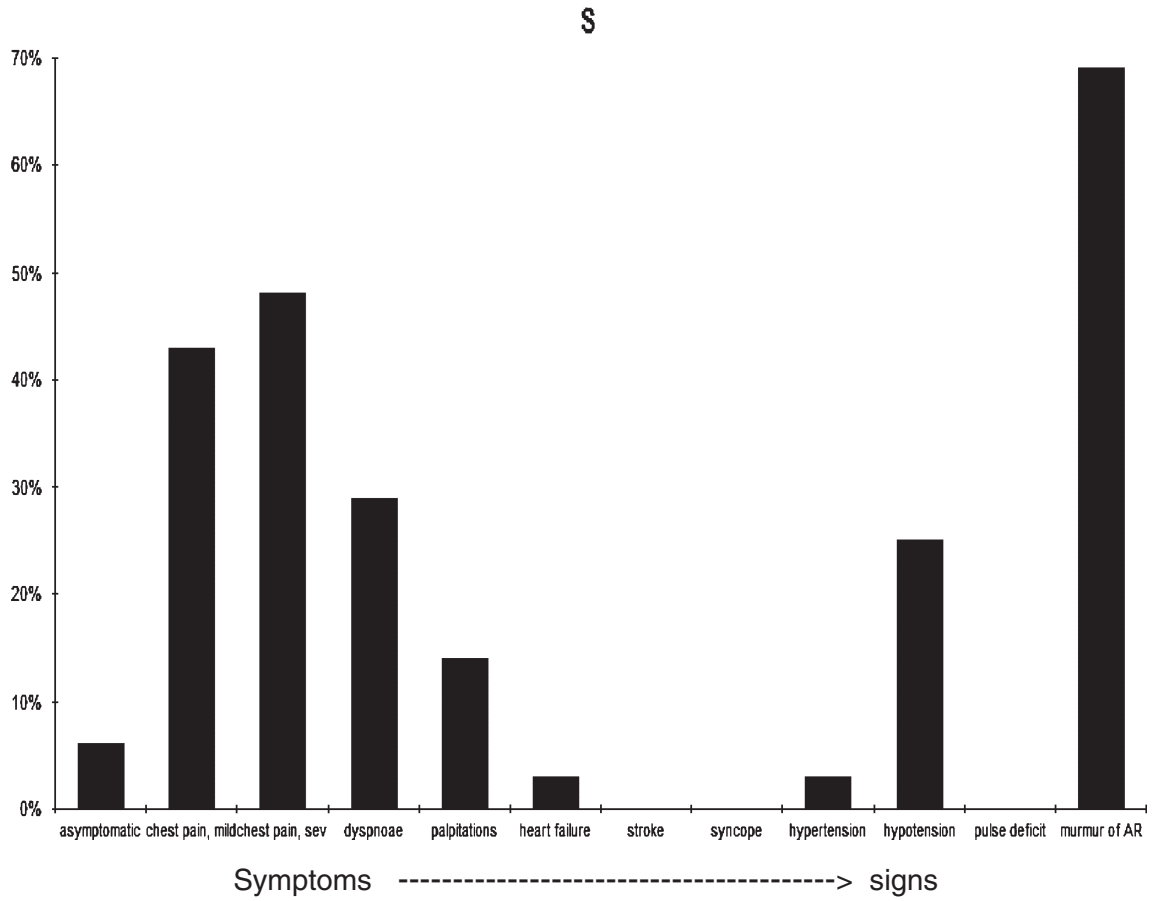

Fig. 2 Distribution of the initial presenting complaints

Table 1 Characteristics of the Aortic pathology

Rheumatic heart disease with root enlargement		12 (22.6%)
Aorto-annular ectasia	Without dissection	17 (32%)
	With aortic dissection	17 (32%)
Bicuspid aortic valve with root enlargement		7 (13.2%)

above in the ascending aorta in the vast majority (88%). In 60% of patients the distal extent of the flap ended in the ascending aorta itself. The aortic dimensions of these patients were fairly large-mean aortic root diameter of 5.63 cm, mean sinotubular diameter of 5.54 cm and mean ascending aortic diameter of 5.24 cm.

Table 2 presents the intra-operative data of our patients. The majority (84.9%, 45 of 53 patients) received composite valved conduit replacement of the ascending aorta with re-implantation of the coronary arteries (modified Bentall procedure). This was performed with need for deep circulatory arrest in only seven patients (13%). In three patients (5.6%) the coronary arteries could not be re-implanted successfully due to technical issues; in these patients, the coronary arteries were bypassed. Table 4 lists the postoperative complications seen. Mortality was 1.8% with low rates of morbidity.

Discussion

Despite numerous advances over the last 35 years, aneurysms and dissections of the ascending aorta remain a major challenge for cardiac surgeons [8]. Many pathological conditions that involve dilatation of the aorta with aortic regurgitation require replacement of the aortic valve and ascending aorta, either separately or as aortic root replacement with re-implantation of the coronary ostia into the graft. Dissection of the thoracic aorta remains the most lethal condition involving the aorta. Studies have emphasized "typical" risk factors and presentation of aortic dissection in adults [9]. However, it has been suggested that younger patients may be more likely to have an atypical presentation of acute aortic disease and dissection [2]. In this study, we have tried to describe the disease profile, possible etiologies, operative techniques and outcome of younger adults requiring aortic root surgery.

Table 2 Intra-operative data

Procedure	Modified Bentall's procedure	45
	AVR + supracoronary aortic replacement	8
	Cabrol fistula	13
	CABG[a]	3
Composite valved conduit size	25.94±2.5 mm	
Route of cardioplegia	Antegrade alone	15
	Combine ante + retrograde	38
Bypass time	160.38±35.66 mins	
Cross-clamp time	93.51±22.86 mins	
Circulatory arrest time (n=6)	23.3±12.3 mins	

[a] Coronary artery bypass graft

Disease profile and outcome

We found that the vast majority of patients of patients had aorto-annular ectasia which is the forme fruste of Marfan's syndrome [10] and other similar connective tissue disorders but none of these patients had been diagnosed to be even marfanoid prior to presentation. This reflects the lack of awareness among the public and the general physician community regarding the potential lethality of Marfan's syndrome. Indeed, even at this institution, accurate measurements to define the syndrome as per the criteria of Ghent [11] were not done, not to mention molecular markers to prove the existence of Marfan's syndrome.

Importantly, we found that the clinical presentation of aortic disease in this group of young adults is similar to older adults- most patients presented with chest pain and/or a murmur of aortic regurgitation. Thus it merits emphasis that even in this age group of patients a complaint of chest pain warrants a clinical suspicion of aortic disease.

Interestingly, we found a difference among our patients' and that of the International Registry for Aortic Dissection (IRAD) [12] patients' in terms of disease extent with aortic dissection (Table 3). Our patients' appear to have more localized disease—flap which arises at the sinotubular junction and most often ends in the ascending aorta itself, which is amenable to surgical correction easier and may be the factor responsible for the very low mortality and morbidity seen in our patients; in only 13% of patients was circulatory arrest required, implying the presence of healthy distal ascending aorta amenable to cross clamping.

There are many options for managing patients' diseases of the aortic root: separate aortic valve replacement and supracoronary ascending aortic replacement (AVR-SUP), modified Bentall's procedure using mechanical prostheses or bioprostheses (which may be stented or stentless), allograft replacement, pulmonary autograft replacement (Ross procedure) and newer investigational endovascular therapies [13].

The Ross procedure [14] is intuitively a good option in these younger patients in an effort to avoid long-term

Table 3 Comparison of the sub-group of patients with aortic dissection to the data from the International Registry for Aortic Dissection

Parameter	Current study (n=17)	IRAD <40 yr (n=)	IRAD >40 yr (n-)
Site of origin of flap			
Aortic root	1 (6%)	20 (29%)	206 (23%)
Sinotubular junction	8 (47%)	14 (21%)	63 (7%)
Ascending aorta	7 (41%)	12 (18)	248 (28%)
Other	1 (6%)	22 (32%)	–
Flap extends to			
Ascending aorta	10 (59%)	13 (19%)	80 (9%)
Arch of aorta	1 (6%)	9 (13%)	90 (10%)
Further distal	6 (35%)	46 (73%)	800 (90%)
Widest aortic dimension			
Aortic annulus	2.71±0.46 cm	3.4±0.9 cm	3.1±0.9 cm
Aortic root	5.63±0.66 cm	4.9±17 cm	4.1±0.88 cm
Sinotubular junction	5.54±0.94 cm	4.6±0.57 cm	3.9±1.1 cm
Ascending aorta	6.24±0.97 cm	5.4±1.8 cm	4.8±1.3 cm
Left ventricular function			
Ejection fraction	47%	NA	NA
End-diastolic dimension	6.58±0.96 mm	NA	NA
End-systolic dimension	5.03±0.5 mm	NA	NA

The data from the IRAD includes patients with both Type A and Type B aortic dissection

anticoagulation and requirement of re-operation for prostheses dysfunction with other forms of therapy. However it is clear that pulmonary autografts dilate when the Ross procedure is performed as aortic root replacement. Thus the role of the Ross procedure for aortic root replacement has been questioned [15]. The limited availability of allograft is one of the limiting factors in its usage.

AVR-SUP is an option especially in those patients with mild dilatation of the sinuses. David et al. [5], compared 133 who underwent AVR-SUP with 452 who had Bentall's procedure and documented good results with both techniques at a mean follow up of 4.6 years, concluding that "AVR-SUP and the Bentall operation provide comparable long-term results. The Bentall operation is more appropriate for patients with aortic root abnormality and a dilated ascending aorta, whereas aortic valve replacement and supracoronary replacement of the ascending aorta is a perfectly acceptable operation for patients with aortic valve disease, normal or mildly dilated aortic sinuses, and a dilated ascending aorta'".

Table 4 Postoperative data

Parameter	Number (%)
Mortality	1 (1.8%)
Re-exploration for bleeding	4 (7%)
CVA	2 (3.6%)
Renal failure	3 (5.4%)
Prolonged ventilation	3 (5.4%)
Complete heart block	1 (1.8%)
Duration of ICU stay	3.2±0.87 days
Length of hospital stay	9.4±2.3 days

CVA cerebrovasular accident, *ICU* intensive care unit

The gold standard of care for diseases of the aortic root remains the modified Bentall procedure which with today's standard of care may be performed with low risk and good long term outcome. For non-dissecting aortic aneurysmal dilatation, the early mortality is around 2–4% [9]. In general, the outcome of patients for aortic dissection carries a worse outcome, in view of the heterogeneity of the pathological anatomy and the fact that most of these operations are life-saving emergencies [12]. Our excellent results that are comparable to any published series serves to emphasis that for the majority of our patients, the pathology was limited to the proximal ascending aorta leaving the distal ascending aorta intact and hence amenable to cross-clamping thus reducing the complexity of the procedure

In the patient with non-dissected but disease aortic root (bicuspid or rheumatic disease, aorto-annular ectasia without dissection), the choice between AVT-SUP and a modified Bentall is often not easy; AVR-SUP carries with it the risk of development of aortic root aneurysm. This risk, however, should be weighed against the hazards of coronary artery re-implantation and other problems related

to aortic root replacement-like intra operative bleeding and kinking that is difficult to manage, and late problems, such as false or true aneurysm of the arterial button, coronary artery, or both, as well as peri-aortic fistulas that may occur [16]

Conclusion

Aortic root pathology, between the ages of 18 and 40 years although rarer than in adults, is mainly due to bicuspid and rheumatic aortic valve disease or aorto-annular ectasia. These patients present with sudden onset of typical chest pain and have a murmur of aortic regurgitation. The disease in our patients appears to be localized to the aortic root and ascending aorta. Using either the Bentall procedure or separate aortic valve and supra coronary ascending aortic replacement offers good early and late clinical outcomes.

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