Background. The aorta is considered pathologically dilated if the diameters of the ascending aorta and the aortic root exceed the norms for a given age and body size. A 50% increase over the normal diameter is considered aneurysmal dilatation. Such dilatation of the ascending aorta frequently leads to significant aortic valvular insufficiency, even in the presence of an otherwise normal valve. The dilated or aneurysmal ascending aorta is at risk for spontaneous rupture or dissection. The magnitude of this risk is closely related to the size of the aorta and the underlying pathology of the aortic wall. The occurrence of rupture or dissection adversely alters natural history and survival even after successful emergency surgical treatment.

Methods. In recommending elective surgery for the dilated ascending aorta, the patient's age, the relative size of the aorta, the structure and function of the aortic valve, and the pathology of the aortic wall have to be considered. The indications for replacement of the ascending aorta in patients with Marfan's syndrome, acute dissection, intramural hematoma, and endocarditis with annular destruction are supported by solid clinical information. Surgical guidelines for intervening in degenerative dilatation of the ascending aorta, however, especially when its discovery is incidental to other cardiac operations, remain mostly empiric because of lack of natural history studies. The association of a bicuspid aortic valve with ascending aortic dilatation requires special attention.

Results. There are a number of current techniques for surgical restoration of the functional and anatomical integrity of the aortic root. The choice of procedure is influenced by careful consideration of multiple factors, such as the patient's age and anticipated survival time; underlying aortic pathology; anatomical considerations related to the aortic valve leaflets, annulus, sinuses, and the sino-tubular ridge; the condition of the distal aorta; the likelihood of future distal operation; the risk of anticoagulation; and, of course, the surgeon’s experience with the technique. Currently, elective root replacement with an appropriately chosen technique should not carry an operative risk much higher than that of routine aortic valve replacement. Composite replacement of the aortic valve and the ascending aorta, as originally described by Bentall, DeBono and Edwards (classic Bentall), or modified by Kouchoukos (button Bentall), remains the most versatile and widely applied method. Since 1989, the button modification of the Bentall procedure has been used in 250 patients at Mount Sinai Medical Center, with a hospital mortality of 4% and excellent long-term survival. In this group, age was the only predictor of operative risk (age > 60 years, mortality 7.3% [9/124] compared with age < 60, mortality 0.8% [1/126], p = 0.02).

Conclusions. This modification of the Bentall procedure has set a standard for evaluating the more recently introduced methods of aortic root repair.

Replacement of the ascending aorta is the most frequently performed procedure for thoracic aortic pathology [1]. Current indications for replacement of the ascending aorta and aortic root may be divided into two broad categories.

Mandatory Indications

These are usually urgent situations involving acute dissection of the ascending aorta and related pathology, spontaneous rupture [2], and intramural hematoma [3]. The value of replacing the entire aortic root for severe destruction of the aortic annulus due to bacterial endocarditis is also well established [4]. Solid clinical experience and information support these indications.

Elective Indications

These are relative indications, since these operations are generally prophylactic in nature, aiming to prevent progression of aortic insufficiency and rupture or dissection of the aorta: in Marfan's syndrome related pathology, in the presence of degenerative dilatation of the ascending aorta [6] with or without a bicuspid aortic valve [7], and in chronic dissection of the ascending aorta. Severe athero-
sclerosis of the ascending aorta (with mobile plaque causing embolization, or discovered during unrelated cardiac surgery) is an emerging indication for elective replacement of the ascending aorta [8, 9]. The decision to operate in these cases is frequently not simple: there is a substantial gray area that changes with the times, greatly influenced by the introduction of new, safe, and effective surgical techniques. The following is a brief review of the current indications for replacement of the ascending aorta, the timing of the operation, and the choice of surgical technique.

Definition
Echocardiography in general, and especially transesophageal echocardiography, remains the main source of information for definition of a norm for ascending aortic size. The normal diameter of the ascending aorta, aortic sinuses, and the aortic annulus correlates with body size and age in both men and women [10]. Body size is the predominant determinant of aortic annular and sinus of Valsalva dimensions, whereas age is a more important determinant of the size of the sino-tubular junction and the ascending aorta proper [10]. The effect of age on the dimension of the ascending aorta is thought to be the result of age-related fragmentation and loss of elastin in the media, leading to overall weakening of the aortic wall [11]. Smoking accelerates the elastin depletion of the aging aorta by increasing the levels of circulating elastolytic enzymes [12]. The dimension of the aortic root in the normal population shows substantial variability. Predicted dimensions at the level of the sinuses can be calculated by the use of the regression formula described by Roman and associates [10]: for an individual 18–40 years of age, average sinus dimension (cm) = 0.97 + (1.12 × BSA [m²]). Similar formulas exist for children and for adults over the age of 40. The use of nomograms for individuals increases the sensitivity of determination of the presence of aortic dilatation by enabling comparison of predicted values with actual measured dimensions. Specificity of 98% is attained by the use of an upper normal limit of 2.1 cm/m² for the aortic diameter at the sinuses [10]. The aorta is pathologically dilated if the diameter exceeds the norm for a given age and body size. An aneurysm is defined as a 50% increase over the normal diameter [13]. Use of an adjusted nomogram for Marfan patients has been suggested in order to compensate for their relatively tall stature [14].

Pathologic and Clinical Consequences of a Dilated Ascending Aorta
Since there are several important physiologic and pathologic consequences of a dilated ascending aorta, the size of the ascending aorta remains the most important component of the equation that leads to a decision to replace the ascending aorta on an elective basis. Dilatation of the ascending aorta is currently the most common cause of isolated aortic valvular regurgitation [15]. A normal aortic valve becomes incompetent as a result of the passive stretching of its leaflets and commissures due to dilatation of the sino-tubular ridge, the ascending aorta, or the sinuses, although the aortic annulus often remains normal in size.

There is a clear relationship between a dilated ascending aorta and a bicuspid aortic valve, even in the absence of significant hemodynamic dysfunction of the valve. This association has been linked to related abnormalities of the aortic wall [16]. A familial clustering of bicuspid valves was described recently, suggesting a possible genetic connection [17], which in turn raises the question of routine screening for siblings of patients with bicuspid aortic valves. The use of adrenergic beta-blocker therapy to retard continued expansion in such individuals with borderline dilatation of the ascending aorta remains an open question [7]. A growing knowledge of the close relationship between a bicuspid valve and a dilated ascending aorta sometimes represents a particular dilemma for the operating surgeon. The most important consequence of an enlarged ascending aortic dimension is the proportional increase in its incidence of rupture, dissection, and reoperation, the latter especially after valve replacement for a bicuspid valve.

There remains a considerable void in our knowledge of the natural history of ascending aortic dilatation. Although there is solid information available in Marfan's syndrome, such data are lacking for other pathologies. Therefore, in the absence of a better measure, some data are routinely transposed from the far better documented natural history of the descending aorta. With this caveat in mind, there are three clues that connect increasing size of the ascending aorta to the incidence of rupture or dissection:

1. An ascending aortic diameter of 6 cm emerges as the mean or the median diameter quite consistently in all reliable contemporary natural history studies. Coady and associates found that a diameter of 6 cm is the “hinge point,” beyond which there is a 30% increase in the probability of rupture [18].

2. The measured diameter of the aorta at the time of acute dissection in all series is significantly larger than the norm. Epperlein and colleagues reported a mean aortic root diameter of 3.2 cm/m² in DeBakey type I dissection [19]. Similarly, in our experience with more than 140 acute type A dissections, the ascending aorta was dilated at the time of presentation in 73%, with a mean diameter of 4.8 cm.

3. The incidence of postoperative dissection is significantly higher if the aorta is 5 cm or larger at the time of aortic valve replacement. Prenger and associates [32] reported an incidence of 27% if the aorta was 5 cm or larger, as opposed to a 0.6% incidence of postoperative dissection if aortic size was normal. This is a strong argument in favor of dealing with a dilated aorta at the time of valve replacement in order to prevent postoperative dissection.

After size, pathology is the second most important determinant of the risk of rupture or dissection in a...
Elective Resection

Current Recommendations: The Rationale for Elective Resection

There is very little doubt that the occurrence of rupture or dissection is a catastrophic event that changes the natural history of a dilated ascending aorta dramatically. Rupture uniformly, and dissection in the vast majority of the patients, is fatal without urgent surgical treatment, which carries a substantially higher risk than elective surgery. Even if the patient survives the acute incident as the result of a successful operation or proceeds into the chronic phase of the dissection, he or she will remain at a higher risk for distal aorta-related complications, which are the most important determinants of long-term survival in most cases.

Current recommendations for elective resection of the ascending aorta are based on the fact that the mean diameter of the aorta at the time of dissection or rupture is around 6 cm, but this actually means that half of the patients will already have experienced one of these highly lethal complications by the age of 60 years, and at the other extreme, by patients of the same aged 50% to 70% of the patients with aortic valve insufficiency are present, as opposed to marked dilatation and severe aortic regurgitation. These two considerations bring to mind arguments for earlier repair of mitral regurgitation. Why should one wait until long-term survival is substantially reduced in 50% of the patients due to an irreparably damaged ventricle if a repair can be done with minimal risk to the patient? Similarly, why wait to operate on the aortic root until half the patients are exposed to the risk of a lethal complication, given that the ascending aorta can be replaced with only a small surgical risk as well as a better chance of sparing the aortic valve if operation is undertaken earlier?

The data emerging on Marfan patients also point out the fallacy of applying an absolute size criterion to all patients. One should be thinking more in terms of ratios or aortic indices rather than absolute sizes. A ratio of 1.3 in a 2-m2 adult of age < 40 years translates to a diameter of 4.2 cm, a dimension much smaller than the commonly recommended size of 5 cm for elective resection in Marfan’s. These data show us that the risk of rupture or dissection is far from negligible at much smaller aortic sizes than those traditionally used as indications for operation [21].

Based on these considerations, the best current recommendations for timely intervention in a dilated ascending aorta can be summarized. There is a spectrum of conditions, represented at one extreme by the Marfan patient with a positive family history of premature rupture or dissection (ratio 1.3, or a diameter of 4.3 cm for an average 2-m2 adult < 40 years of age), and at the other extreme, by a patient of the same size with a dilated aorta due to medial degeneration without significant aortic regurgitation, or one whose dilatation is discovered during unrelated routine cardiac surgery (ratio 1.5, or diameter 4.8–5 cm). Indications for chronic dissections should be considered akin to those for Marfan syndrome because of the shared factor of a weakened aortic wall. Patients with a bicuspid aortic valve fall between these two extremes, especially if operation is indicated primarily for a dysfunctional valve: it is probably prudent to proceed with definitive treatment of the ascending aorta if the ratio exceeds 1.4 (diameter 4.5 cm) at the time of valve replacement. Again, the factor that prompts earlier resection in these cases is the associated inherent weakness of the aortic wall.

In patients with medial degeneration and secondary aortic regurgitation, the degree of insufficiency may dictate earlier operation regardless of aortic size. Delaying definitive treatment in these cases not only will jeopardize long-term outcome due to deterioration of left ventricular function, but also will reduce the probability of being able to spare the aortic valve. Of course, the
Surgeons’ experience has to be taken into account in recommending relatively early operation to an otherwise asymptomatic patient. We therefore would suggest adding 0.15 to the ratio or 0.5 cm to the diameter to account for this factor in different hands. This would bring these recommendations more in line with some of the more aggressive traditional criteria (Table 1). Adherence to these guidelines will not eliminate the occurrence of rupture or dissection, but would be expected to halve the incidence of lethal complications and prevent substantial numbers of emergency operations.

**Choice of Procedure**

Currently, there are a variety of operative techniques available for surgical treatment of a dilated ascending aorta. These represent a large spectrum, ranging from separate replacement of the aortic valve and ascending aorta to the most widely applied and versatile technique of composite root replacement with one of the three principal modifications of the technique originally described by Bentall and DeBono [24], and also include the more recently introduced techniques of pulmonary autograft [25] and valve-sparing root replacement [26]. Among these alternatives, the often neglected but time-tested method of aortic root wrapping is worth mentioning. It has the advantage of simplicity, and more importantly, preserves the endothelial lining of the ascending aorta. There is evidence to suggest that, when done properly, wrapping is a good compromise in older patients with a borderline dilated aorta, especially during operations for other cardiac pathology [27]. Choosing the technique appropriate for the particular patient and pathology requires careful consideration of many factors, including the surgeon’s experience with the particular method. Among these factors are:

1. **Age and expected survival.** In an older, high-risk patient, simple valve replacement and wrapping of the aorta may be a good compromise solution [27]. Similarly, separate valve and ascending aortic replacement may be appropriate therapy if life expectancy is limited [28].

2. **Underlying pathology and quality of the aortic wall.** A weakened aortic wall in Marfan syndrome or dissection will require complete excision of the dilated portion of the aorta and the root. The button modification of the Bentall procedure has proven in our hands the most versatile and durable reconstruction [29]. Sparing the aortic valve in Marfan syndrome is controversial [30].

3. **Anatomic condition of the aortic valve, the sinuses, and the sino-tubular ridge.** The anatomic condition of the important elements of aortic valvular integrity usually dictate whether the valve can be spared or whether a separate valve-ascending aortic replacement may be feasible. In our experience, the ideal candidate for a valve-sparing root replacement is a patient with a normal valve and annulus in whom a dilated sino-tubular ridge or sinuses lead to aortic insufficiency. A separate valve and ascending aorta replacement that leaves behind portions of dilated sinuses is a compromise that should be avoided in patients with a relatively long life expectancy [28].

4. **Condition of the distal aorta.** If the condition of the distal aorta mandates future operation for associated distal arch or descending aneurysm or dissection, then a fail-safe initial repair at the root is of paramount importance. The presence of even modest degrees of aortic regurgitation may substantially complicate an operation on the distal aorta, which frequently requires the utilization of hypothermic circulatory arrest. In these cases, we would prefer a composite replacement rather than a valve-sparing procedure.

5. **The risk of anticoagulation.** Considerations of long-term anticoagulation risk weigh heavily in favor of a valve-sparing operation if feasible, a pulmonary autograft, or use of a composite graft with a tissue valve.

6. **Presence of active annular infection.** Although there is little evidence in the literature to support it, many surgeons believe an allograft may be preferable for root replacement in this situation.

Table 2 summarizes our current preferences for various kinds of pathology.

**Clinical Experience**

Between January 1988 and February 1998, a total of 497 patients underwent operations at Mount Sinai Medical Center for aortic root pathology. Between 1988 and 1992, 286 patients had surgery for a susceptible aortic annulus or sinus, and 201 underwent surgery for extensive aortic root disease. Of the latter group, 199 underwent surgery for aortic root dissection, 111 for aortic root dilatation, and 33 for Marfan’s syndrome. The remodeling of the aortic root was significantly lower in patients with a normal valve and annulus in whom a dilated sino-tubular ridge or sinuses led to aortic insufficiency. A separate valve and ascending aorta replacement that leaves behind portions of dilated sinuses is a compromise that should be avoided in patients with a relatively long life expectancy [28].

### Table 1. Current Guidelines for Surgery

<table>
<thead>
<tr>
<th>Adult Age &lt; 40 years BSA 2 m²</th>
<th>Diameter</th>
<th>Ratio</th>
</tr>
</thead>
<tbody>
<tr>
<td>Marfan’s (+family history)</td>
<td>&gt; 4.3</td>
<td>1.3</td>
</tr>
<tr>
<td>Chronic dissections</td>
<td>&gt; 4.3</td>
<td>1.3</td>
</tr>
<tr>
<td>Degenerative without AI</td>
<td>&gt; 4.8</td>
<td>1.5</td>
</tr>
<tr>
<td>Degenerative with AI (degree!)</td>
<td>&gt; 4.8</td>
<td>1.5</td>
</tr>
<tr>
<td>Bicuspid valve with dysfunction</td>
<td>&gt; 4.5</td>
<td>1.4</td>
</tr>
<tr>
<td>Other cardiac surgery</td>
<td>&gt; 4.8</td>
<td>1.5</td>
</tr>
<tr>
<td>Surgeons’ experience</td>
<td>+0.5</td>
<td>0.15</td>
</tr>
</tbody>
</table>

**AI** = aortic insufficiency.

### Table 2. Surgical Options

<table>
<thead>
<tr>
<th>Valve</th>
<th>Annulus</th>
<th>Sin/ST-Rdg</th>
<th>Procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>+++ (old)</td>
<td>N</td>
<td>N</td>
<td>Aorta + valve, wrap (?)</td>
</tr>
<tr>
<td>+++ (young)</td>
<td>N</td>
<td>N</td>
<td>Bentall, pulmonary autograft (?)</td>
</tr>
<tr>
<td>N</td>
<td>N</td>
<td>+++</td>
<td>Valve-sparing</td>
</tr>
<tr>
<td>N</td>
<td>+++</td>
<td>+++</td>
<td>Bentall, valve-sparing (?)</td>
</tr>
<tr>
<td>N (Marfan)</td>
<td>+++</td>
<td>+++</td>
<td>Bentall</td>
</tr>
<tr>
<td>N (Marfan)</td>
<td>N</td>
<td>+++</td>
<td>Bentall, valve-sparing (?)</td>
</tr>
<tr>
<td>Infected</td>
<td>N</td>
<td>N</td>
<td>Bentall with–without allograft</td>
</tr>
</tbody>
</table>

* (?) = secondary choice.

N = normal; +++ = severe pathology autograft.
Center on the aortic root, the ascending aorta, and the proximal portion of the aortic arch exclusive of total arch replacement. The indications for surgery in these cases were in accord with the clinical philosophy outlined above. Dissections, both acute and chronic, and degenerative dilatation of the ascending aorta accounted for about 60% of the cases.

The overall hospital mortality was 8% (40/497). Mortality was significantly higher in urgent operations (12.3%, 23/187) than in elective surgery (5.5%, 17/310); $p = 0.05$ [chi square]) and in patients older than 60 years (12.1%, 34/281) compared with those younger than 60 (2.8%, 6/216; $p = 0.01$ [chi square]). Interestingly enough, the presence of acute dissection did not significantly influence operative mortality (12.2%, 15/123, vs 9.9%, 10/101).

Composite replacement of the aortic valve and the ascending aorta, with one of the three principal modifications of the Bentall procedure, was the most common surgical method, used in 311 patients. The proportions of the various surgical options and their associated mortality with respect to age are shown in Figure 1. The button modification of the Bentall procedure was used in 250 patients, with a 4% overall hospital mortality (10/250). Again, age was the only significant factor influencing operative risk. Mortality in patients older than 60 years was 7.3% (9/124) compared with 0.8% (1/126) in patients younger than 60 ($p = 0.02$ [chi square]). Urgency of operation and/or presence of acute dissection did not increase mortality significantly. A hybrid technique in which the coronary orifices are dissected out as buttons surrounded by aortic tissue before end-to-end anastomosis to the Cabrol graft [2] combines the principles of the
button [29] and Cabrol modifications [31] of the original Bentall procedure [24], and ensures the safe repair and reinforcement of even the most seriously dissected coronary orifices. It has been very helpful in achieving a strikingly low operative mortality with radical root replacement in acute dissections. The overall results of the button modification of the Bentall procedure in this series of patients compares very favorably with results obtained with alternative techniques, including valve-sparing replacement and pulmonary autografts, with durable long-term results. Event-free survival was 79.1% ± 3% at 5 years and 62.3% ± 7% at 8 years. There were significantly fewer late reoperations and deaths related to the aorta or the repair in this group compared with patients who had repairs other than the button modifications of the Bentall procedure. Cardiac and distal aorta-related events account for most cases of long-term attrition (Figure 2).

Conclusions
Currently available surgical techniques yield good immediate and long-term results with minimal risk in elective operations on the ascending aorta and aortic root. The mortality of emergency operation still remains high. The occurrence of dissection adversely affects long-term outlook. In light of surgical advances, the previously accepted indications for elective replacement of the dilated ascending aorta may be too conservative, and a revision is needed toward more liberal indications in order to prevent lethal complications and emergency operations. New data from detailed natural history studies will undoubtedly help in refining operative guidelines. Appropriate choice of surgical procedure results in excellent long-term results, but cardiac and distal aorta-related events ultimately determine survival.

References