Total Aortic Replacement for Chronic Aortic Dissection Occurring in Patients with and without Marfan's Syndrome

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Total aortic replacement including aortic valve was performed successfully in the two patients in whom this method of treatment was utilized to correct a chronic dissecting aortic aneurysm. Both patients had moderately severe aortic insufficiency producing increasing heart strain and progressive enlargement of the false lumen of aortic dissection involving the entire aorta despite ideal blood pressure control. In addition, one patient had Marfan's syndrome. The surgical treatment for both patients was performed in two stages. At the first operation, cardiopulmonary bypass, profound hypothermia, and circulatory arrest were employed while the aortic valve and the ascending and transverse aortic arch were replaced and the coronary and brachiocephalic vessels were reattached to the composite valve-graft used for replacement. At the second operation, the entire descending thoracic and abdominal aortic segments were replaced with a graft and the intercostal, lumbar, and visceral arteries reattached thereto. Left vocal cord paralysis occurred in both patients and transient mild paraparesis occurred in only one. Both patients are alive and well, one at 13 months and one at 6 weeks. This experience suggests an additional treatment modality for selected patients with complications of chronic aortic dissection.

THE SURVIVAL RATE of patients with acute aortic dissection in a collected series of 963 patients is 10% at 3 months and none at 3 years.[1] The survival rate of the 93% of the 527 patients in our collective departmental series of surgically treated patients with both acute and chronic disease from which updated follow-up was obtained for the period 1981 to 1982 was 57%, 32%, and 5% at 5, 10, and 20 years, respectively.[2] This long-term follow-up study revealed that the most common cause of late death is the rupture of fusiform aneurysms of segments of dissected aorta that were not removed at the operation performed at the time of and/or subsequent to the acute dissection. This fatal complication accounted for 29% of all late deaths. The incidence of late aneurysmal formation in these cases varied with the location, origin, and extent of dissection as well as the degree of control of hypertension. The incidence of late aneurysmal formation in Type I and IIIb was 30% and 38%, respectively, whereas in Type II it was 14% and in Type IIIa it was 16%. Regardless of location or extent of dissection, late aneurysms developed in 46% of patients with uncontrolled hypertension and in 17% of those in whom hypertension was controlled.

Thus, this study emphasizes the need for an annual or even more frequent follow-up of the patient in order to control the hypertension and detect the presence of late development of aneurysm. Detection of the latter is significant because these lesions can be surgically corrected by aortic reconstructive operation with survival in the majority of cases.[2-9] The usual approach to correct this problem consists of traditional segmental graft replacement of the particular segment of the aorta involved when such segment is located within the ascending aorta, the descending thoracic aorta, and/or the thoracoabdominal aortic segment.[2-4] Graft replacement of the transverse aortic arch for chronic dissecting aortic aneurysm rarely has been reported because such operations have not been safe until recently and therefore were not often performed. Recent developments in surgical techniques have now made it possible to safely replace aneurysms of the transverse aortic arch in over 90% of patients regardless of etiology.[5,6] Advances in technique of treatment of aneurysms involving the entire thoracic and abdominal aorta now permit treatment of lesions involving these segments at one operation with survival in nearly 90% of patients treated electively.[3,4] Combining these techniques in stages, the first on October 27, 1980 and the last on December 29, 1981, we first successfully replaced the entire aorta in a patient with mega-aorta resulting from medial degenerative disease.[6-9] This operation was performed subsequently in seven other patients with this disease and all eight patients survived without significant complica-

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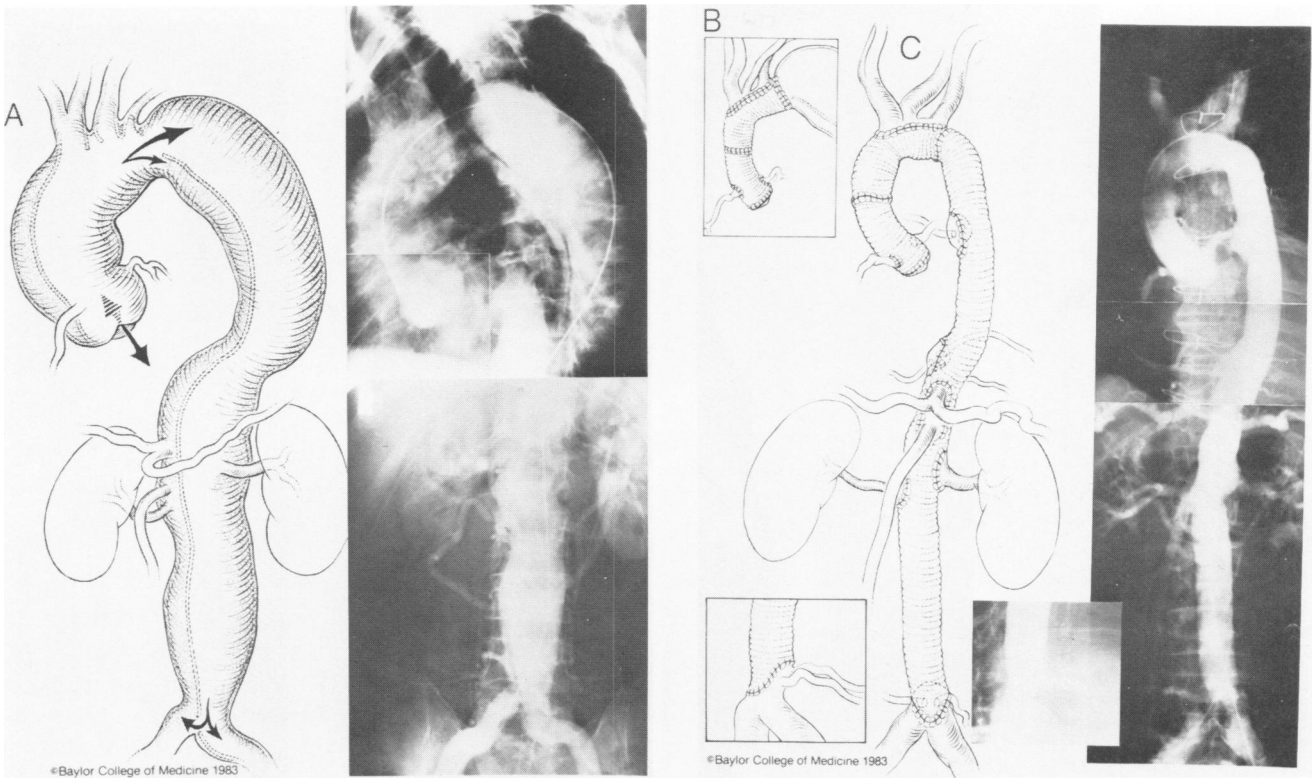

FIG. 1. Illustrations of a patient with chronic Type I aortic dissection complicated by aortic valvular insufficiency and fusiform aneurysm involving the entire aorta. A. Drawing and aortogram made before operation showing massive aneurysmal dilatation of false lumen throughout the aorta extending along the greater curvature of the ascending and transverse arch and the left posterolateral wall of descending and abdominal aorta, compressing the true lumen on the right (the route of dissection in 90% of cases). B. Drawing showing method of graft replacement used to replace aortic valve, ascending, and transverse aortic arch in the first operation and C. drawing and aortogram made after second operation showing total replacement of the aorta with preservation of intercostal, visceral, and lumbar circulation.

tions.[6-9] These cases form the basis of another report regarding the unique pathologic features of mega-aorta, its clinical manifestations, and special requirements for its management.

The experience gained from the performance of this operation in the treatment of these eight patients suggested its application in the treatment of certain patients with aortic dissections. Total aortic replacement was therefore employed in the treatment of two patients with chronic Type I aortic dissection complicated by aortic valvular insufficiency and fusiform aneurysm involving the entire aorta, one of whom was briefly mentioned in a previous communication.[8] This report, however, is primarily concerned with a detailed presentation of these two cases and a discussion of the role that total aortic replacement may have in the future management of patients with aortic dissection.

Case Reports

Case 1

This 51-year-old hypertensive salesman developed acute Type I aortic dissection on February 15, 1981. He was successfully treated medically at this time according to the principles originally described by Wheat.[10] Despite careful and frequent follow-up and ideal control of blood pressure, the patient developed aortic insufficiency and progressive enlargement of the entire aorta, which was evident by plain roentgenograms of the chest, abdominal palpation, and computerized tomography (CT) scans. The patient's activities became increasingly limited because of shortness of breath and pain in both the chest and low back. He developed recurrent severe substernal and back pain on April 6, 1982 despite blood pressure maintenance at 120/80 mmHg level. Myocardial infarction was excluded by appropriate studies, causing his physicians to conclude that the pain represented progression of his aortic disease and therefore he was referred for surgical treatment on April 7, 1982.

At the time of admission, the patient was essentially asymptomatic with the exception of moderate to severe aortic valvular insufficiency and diffuse dilatation of the false lumen of a chronic dissecuon involving the entire aorta (Fig. 1A). On April 12, 1982, employing cardiopulmonary bypass, profound hypothermia (16°C, rectal), and circulatory arrest, the ascending and transverse aortic arch were replaced with a composite valve graft and the coronary and brachiocephalic arteries reattached directly to openings made in the graft using the inclusion technique described elsewhere in detail (Fig. 1B).[5,9] The patient did well and was discharged in excellent condition on April 27, 1982.

After a complete recovery, the patient was readmitted and the remaining aorta was replaced on July 12, 1982 with a graft utilizing a thoracoabdominal incision and the inclusion technique as previously described (Fig. 1C).[3,4] Two intercostal arteries were reattached to the graft at midthoracic level and three intercostal arteries were reattached

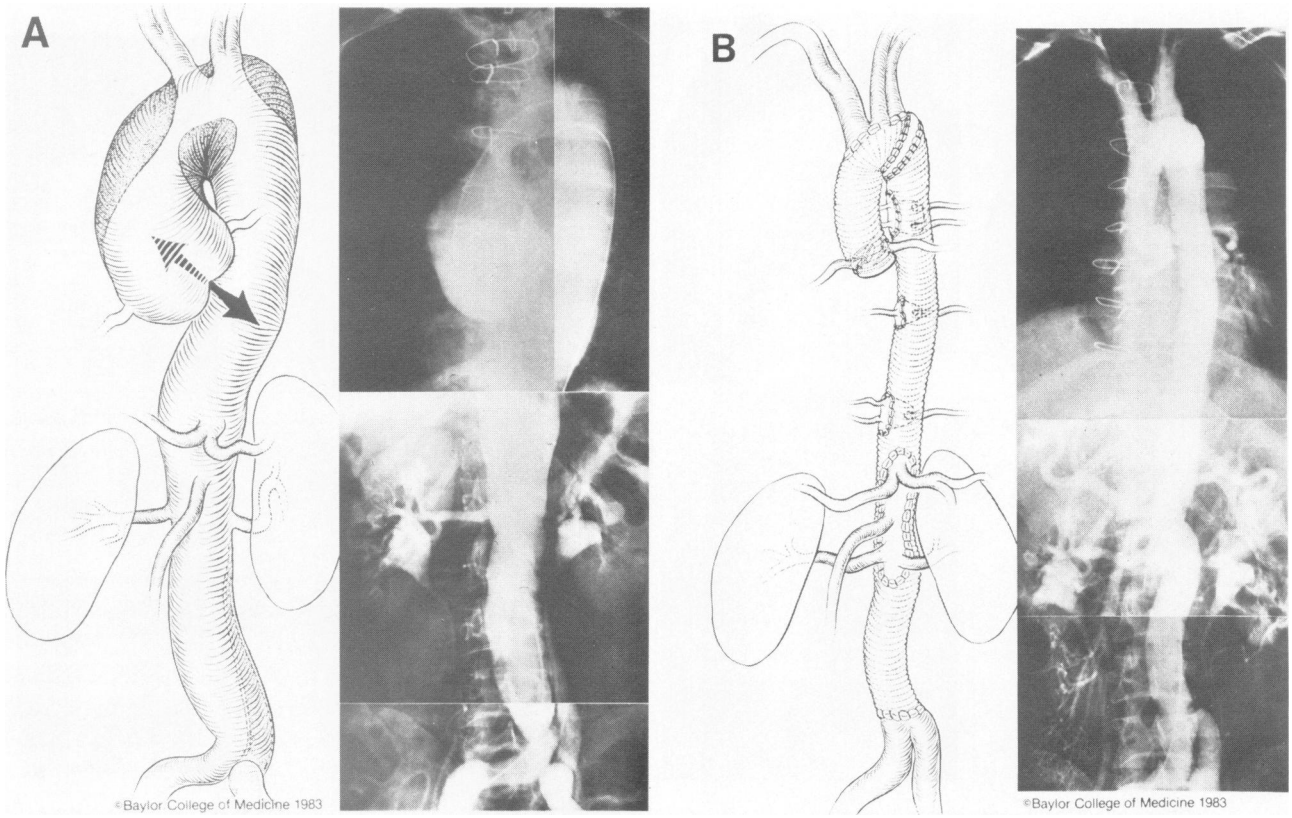

FIG. 2. Illustration of patient with Marfan's syndrome who had chronic Type I aortic dissection superimposed on fusiform aneurysm of the ascending aorta complicated by aortic insufficiency and fusiform aneurysm involving the entire aorta. A. Drawing and aortogram made before operation showing aortic valvular insufficiency and massive dilatation of the false lumen of dissection extending throughout the aorta on the right in the lower thoracic and abdominal aorta compressing the true lumen located in the left side of aorta (the route of dissection in 10% of cases). B. Drawing and aortogram made after the second operation showing replacement of the aortic valve and aorta with preservation of intercostal, visceral, and lumbar circulation.

just above the diaphragm. The last set of lumbar arteries were spared by the technique shown at the bottom of Figure 1C. The patient did well following the operation with the exception of left recurrent nerve paralysis, weakness of the right hip flexors and adductors, and an neurogenic bladder. These problems improved with physical therapy and at the time of discharge on August 23, 1982, the patient was walking with a walker and voiding by self catheterization. Subsequent follow-up indicated that by January 1983, normal bladder function returned and the patient could walk unaided. At the last follow-up on August 8, 1983, the patient had completely recovered with the exception of left vocal cord paralysis that did not impair satisfactory phonation.

Case 2

A 35-year-old social worker with Marfan's syndrome manifested by occular, skeletal, and genetic manifestations developed acute Type I aortic dissection in February 1980 complicated by acute aortic valvular insufficiency. Within hours, he was submitted to operation in his hometown where he was found to have a dissection superimposed upon a fusiform aneurysm of the ascending aorta. He was treated by proximal ascending aortic transection, resuspension of aortic valvular apparatus by oversewing the inner and outer layers of dissection proximally and primary end-to-end anastomosis to the similarly oversewn distal ascending aortic segment. The dilated proximal ascending aorta was corsetted by completely enclosing it within a Dacron® wrap using Dacron fabric. The patient recovered uneventfully and returned to his normal activity. He was followed frequently after discharge and his blood pressure was controlled at the 120/80 mmHg level and his pulse rate maintained at 60 bpm with appropriate medication. At the time of his January 4, 1983 visit to his doctor for evaluation, he was found to have aortic valvular insufficiency and "heart strain," a significant increase in size of the ascending aorta as shown by echocardiography, and a large palpable abdominal aortic aneurysm.

He was referred and admitted on May 15, 1983 for consideration of extensive aortic reconstruction. His doctor's findings were confirmed and total aortography showed aortic valvular insufficiency and diffuse dilatation of the false lumen of a chronic dissecting process involving the entire aorta located on the greater curvature of the ascending aorta and transverse aortic arch, the left posterolateral aspect of the descending thoracic aorta, and the right posterolateral aspect of the abdominal aorta extending into the right iliac artery (Fig. 2A). The aortic valve and the ascending and transverse aortic arch were replaced on May 19, 1983 with a composite valve graft in the same manner described. After a complete recovery at home, he was readmitted and on July 8, 1983, the remaining aorta was replaced as in the previous case (Fig. 2B). Four intercostal arteries were reattached to the graft in the upper chest, two in the midchest, and two just above the diaphragm. Two lumbar arteries were reattached at midabdominal level. The patient recovered unevent-

fully without complications and was discharged July 20, 1983 and has subsequently regained full activity.

Discussion

After having successfully replaced the ascending and transverse aortic arch in 12 patients with both acute and chronic aortic dissection and having replaced the entire descending thoracic and abdominal aorta for diffuse chronic aortic dissection in 58 other patients, ten of whom also had prior ascending aortic replacement with survival in 50 (86%), it was obvious that the first and second procedures could be successfully performed in stages to completely replace the aorta for dissecting aneurysms involving the entire aorta. The two cases presented here demonstrate the technical feasibility of this concept and both cases suggest that multiple intercostal and lumbar arterial reattachment minimizes the incidence and extent of paraplegia. Thus, a new mode of treatment is available for patients with dissection diffusely involving the entire aorta. This development raises a number of considerations including indications for its application, the incidence of these indications, the effectiveness of the procedure in prolonging useful life, and how the relationship of the latter compares to other methods of treatment, some of which include less extensive operation or medical treatment. The answers to these questions are unavailable at the present time due to limitations of experience in the application of this approach and its long-term results. Certain observations obtained from personal experience in the treatment of 216 patients with aortic dissection and a review of extensive pathologic and clinical studies of the disease by DeBakey, Johns, Hirst, and Roberts permit certain speculations that may or may not prove erroneous with increasing experience and these are considered separately.[2,11-13]

Indications

Total aortic replacement or reconstruction is considered only in patients with dissection involving the entire aorta which is associated with chronic diffuse aneurysmal formation of the outer wall of false lumen. We feel this is justified at the present time based upon the fact that death in the late follow-up period resulted from rupture of those dilated segments, regardless of location, which remain after one or more segmental replacements.[2] It is emphasized, however, that prophylactic replacement of undilated segments is not recommended unless other complications such as aortic valvular insufficiency and symptomatic branch vessel obstruction are present because such segments may never dilate.

Total aortic replacement in the patient with acute diffuse Type I dissection, although ideal in concept because of its curative implications, would rarely be considered in our hands. By definition, this would require total replacement of the aorta within the first 2 weeks of illness. Such an operation performed either in one or two stages at this time probably could not be tolerated because of the severity of the disease and its complications, the incidence of associated problems, and the magnitude of operation. For example, of our 11 patients submitted to extensive thoraco-abdominal aortic replacement for acute dissection associated with aortic rupture, three (27%) developed paraparesis in part due to the fact that intercostal and lumbar arteries could not be reattached and five (45%) of these cases did not survive. Total replacement would therefore be considered only in the patient with rare and unusual manifestations.

Incidence

Pathologic studies (autopsy) indicate that the tear from which dissection arises is located in the ascending aorta in 68% and the transverse aortic arch in 10% of patients dying from the disease.[11-13] Dissection in the majority of these cases involved the entire aorta. The incidence of this type of dissection in patients who survived to have the diagnosis made and who were submitted to operative treatment was 26.2% in our departmental series of cases.[2] The incidence of persistent or late aneurysmal formation in this surgically treated group of patients was 30% regardless of the number and extent of individual aortic segments replaced and actual rupture of the aneurysm accounted for 29% of late deaths.[2] These observations suggest a relatively frequent need for total aortic replacement in stages either during one treatment period or during the lifetime of the patient.

Results of Total Aortic Replacement

All patients submitted to total aortic replacement to date by the authors have survived operation, eight with medial degeneration and two with chronic dissection. Paraplegia occurred in only one and it was relatively mild and ultimately cleared. This experience is small and long-term results are not yet available to compare with lesser procedures or with medical treatment but are sufficiently good to justify its continued application in selected cases.

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