A 43-year-old man presented with dyspnea of 1.5-year history. He was a smoker and had hypertension as a risk factor. Upper extremity blood pressure was 160/90 mmHg, lower extremity blood pressure was 120/50 mmHg, and heart rate was 97 bpm. A grade 2-3/6 diastolic murmur was heard on the mesocardiac area. All pulses were palpable, but pulses of the lower extremities were weaker. Other physical findings were normal. Transthoracic echocardiography showed left ventricular hypertrophy, calcific aortic valve leaflets, grade 1-2 aortic insufficiency, and a peak gradient of 25 mmHg. Transesophageal echocardiography showed a bicuspid aortic valve and grade 2-3 aortic insufficiency. The diameter of the ascending aorta was 54 mm. Cardiac catheterization showed grade 2-3 aortic insufficiency and normal coronary anatomy. The descending aorta was totally occluded (Fig. A). Computed tomography demonstrated dilatation of the ascending aorta to approximately 55 mm, extending up to the proximal aortic arch and total occlusion of the descending aorta in the proximal region (Fig. B, C). A two-stage operation was planned. First, a Dacron graft (14 mm x 8 cm) was implanted to the proximal region of the descending aorta. Then, a Bentall procedure (composite graft replacement of the aortic valve, aortic root, and ascending aorta, with re-implantation of the coronary arteries into the graft and mechanical aortic valve replacement) was performed (Fig. D, E). The postoperative course was uneventful and the patient was discharged without any complication. During a two-year follow-up, he continued to enjoy an excellent result.

**Figures.** (A) Descending aortography shows a type A interrupted aortic arch. (B, C) Computed tomography images showing an ascending aortic aneurysm and interrupted aortic arch at the isthmic region of the aorta. (D, E) Postoperative computed tomography images following a two-stage operation involving Dacron grafting of the interrupted aorta and Bentall procedure for the ascending aortic aneurysm, respectively.