



Coarctation of aorta with complete aortic occlusion

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Abstract

Survival to advanced age is exceptional in patients with unrepaired aortic coarctation. We report the case of an 81-year-old man with aortic coarctation and total occlusion who was otherwise asymptomatic. Coarctation was suspected when a femoral-radial pulse delay was noted during his routine physical examination. A 70-mmHg systolic blood pressure gradient between the upper and lower extremities was detected. Subsequent magnetic resonance angiography, aortography, and coronary angiography revealed severe coarctation of the aorta, well-developed collateral vessels, and severe coronary artery disease. A staged percutaneous coronary intervention procedure was performed and the coarctation was managed conservatively with antihypertensive medication.

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1 Introduction

Coarctation of the aorta (CoA) occurs in 1 in 2000 live births in the USA and is the fifth most common congenital cardiac defect. It is more common in males (M: F ratio: 1.7: 1) and affects Caucasians seven times more than other races.^[1] Seventy five percent of children with CoA have another cardiac anomaly, most commonly patent ductus arteriosus (PDA), bicuspid aortic valve, ventricular septal defect (VSD) and mitral valve anomalies. CoA is usually diagnosed in childhood and early adulthood, and there is a reduced life expectancy in patients who have not undergone correction. Survival to older age is rare, due to severe cardiovascular complications.^[1] There are only a few cases of elderly patients with uncorrected CoA,^[2–4] and management strategies in such patients are controversial. We describe the case of a man first diagnosed with CoA and total occlusion of the aorta at an advanced age.

2 Case report

An 81-year-old man was referred to our clinic for evaluation and treatment of typical angina during exercise. His

medical history included arterial hypertension for many years; there was no history of traumatic injury or Takayasu's arteriitis. He was otherwise asymptomatic. On physical examination, the blood pressure was 140/85 mmHg in both arms. The lower extremity pulses were diminished and delayed. The patient denied symptoms of claudication and weakness of his legs and reported to walk more than one kilometer without any problems. Results of routine blood chemistry and urine analysis were normal. Electrocardiography revealed left ventricular hypertrophy without evidence of prior myocardial infarction. Chest X-ray demonstrated typical rib notching (Figure 1).

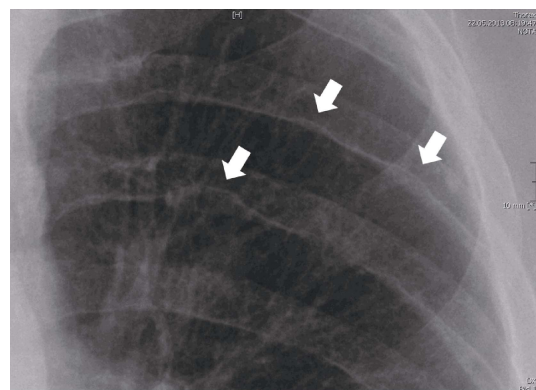


Figure 1. Chest radiography: Rib notching (white arrows).

Concentric left ventricular hypertrophy, a normal tricuspid aortic valve, a normal mitral valve, and normal left ventricular systolic function could be demonstrated on echocardiographic examination. However, the distal part of the aor-

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tic arch was not adequately visualizable. Coronary angiography (Figure 2) was performed via the radial route, and severe triple-vessel disease was detected (SYNTAX (SYNERgy between PCI with TAXUS™ and Cardiac Surgery) Score of 35 points).

Aortography (Figure 3) revealed a postductal CoA with total occlusion of the aortic arch. Simultaneous pressure tracings in the aortic arch and the descending aorta demonstrated a pressure gradient of 70 mmHg. There was no evidence of clot formation in the blind end of the aorta distal to the coarctation.

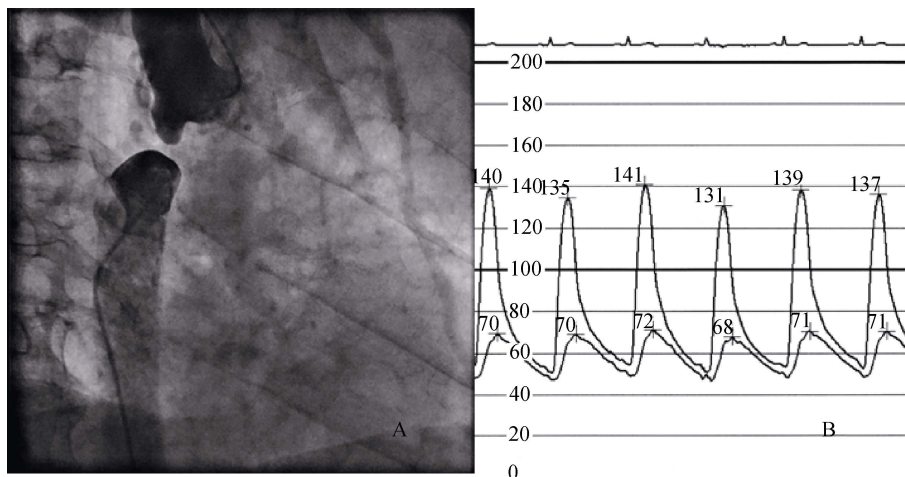


Figure 3. Antegrade and retrograde aortography. (A): Aortic coarctation with total occlusion of the descending aorta; (B): Simultaneous pressure tracings obtained proximal and distal of the aortic occlusion demonstrating a pressure gradient of about 70 mmHg.

Magnetic resonance (MR) angiography was performed and confirmed the aortic occlusion just below the origin of the left subclavian artery, showed slight post-stenotic dilatation, and distinct collateral circulation (Figure 4).

Cranial MR was without evidence of intracerebral aneurysm. The patient refused operative treatment of his triple-vessel coronary artery disease, and underwent finally a staged percutaneous coronary intervention procedure. Surgical correction of the CoA was not performed because the patient was asymptomatic with a good exercise tolerance and his hypertension was well controlled with a combination of three antihypertensive agents (β -blocker, diuretic, and angiotensin-receptor-blocker). At clinical follow-up after 8 months the patient remained free of symptoms.

3 Discussion

Coarctation of the aorta is a relatively common defect that accounts for 5%–8% of all congenital heart defects,^[5] and should be diagnosed and corrected early in life. Among 2192 CoA patients reported to the Pediatric Cardiac Care Consortium from 1985-1995, 1337 were infants, 824 were

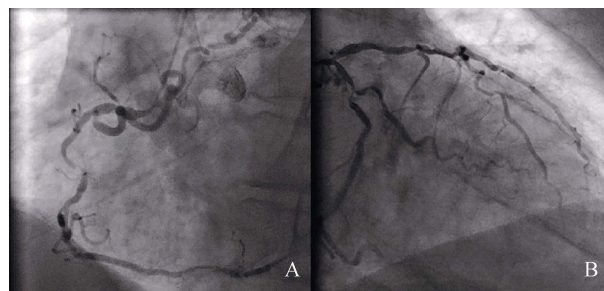


Figure 2. Coronary angiography: Triple vessel disease of the right coronary artery (Panel A) and left coronary artery (Panel B).



Figure 4. Magnetic resonance imaging: Aortic coarctation with total occlusion of the aorta and collateral circulation.

children, and 31 were adults.^[6] Associated problems that may contribute to death or morbidity include hypertension, cardiovascular disease, intracranial hemorrhage, aortic rupture or dissection, endocarditis, and congestive heart failure. Early repair of CoA in juvenile patients improves both quality of life and long-term survival.^[7] Therapeutic goals in old

patients with uncorrected CoA are symptomatic improvement and control of hypertension whereas the presence of any survival benefit from CoA repair in the age group older than 70 years is unclear. There are few reports of patients initially diagnosed with uncorrected coarctation at very late age (> 70 years), and there is no consensus on how to manage them. Historical studies suggest that the mortality rate in patients in whom coarctation of the aorta is not surgically repaired is 90% by age 50 years, with a mean age of 35 years.^[1] Two case reports of a 72 years^[2] and a 76 years^[3] old patients without surgical repair reported follow-up for several years with both patients dying for non-cardiac reasons. In contrast, some surgeons have performed successful correction in these older patients.^[8-10] Aris, *et al.*^[9] and Bauer, *et al.*^[10] reported good surgical outcomes for CoA repair in 8 and 15 patients, respectively, older than 50 years; however, only 2 and 3 patients, respectively, were older than 60 years. These studies demonstrated that symptomatic improvement occurred, and hypertension was well controlled after correction in these older patient groups although most of the patients still needed antihypertensive medication at late follow-up. Survival of a patient to 81 years of age is exceptional in cases of uncorrected coarctation. Our patient had severe triple-vessel disease but refused coronary artery bypass grafting and underwent finally a staged percutaneous coronary intervention procedure. Surgical repair of the CoA^[6,11] or balloon angioplasty/stentimplantation^[12,13] was not indicated in our case, because of his good exercise tolerance and his well-controlled arterial hypertension.

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