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Diagnosis of Interrupted Aortic Arch in an Adult during Coronary Artery Evaluation

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The authors have no financial conflicts of interest.

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A 51-year-old male presented at our hospital with exertional chest pain. His blood pressure was 171/98 mmHg in the left arm and 109/51 mmHg in the left leg. Coronary computed tomography angiogram (CTA) showed 70%–90% stenosis in the mid-left anterior descending artery and discontinuity of the aortic arch between the left subclavian artery and descending aorta without patent ductus arteriosus (**Figure 1A**). Aorta CTA revealed interruption of aortic arch (IAA) with multiple collateral channels (**Figure 1B-D**). A ventricular septal

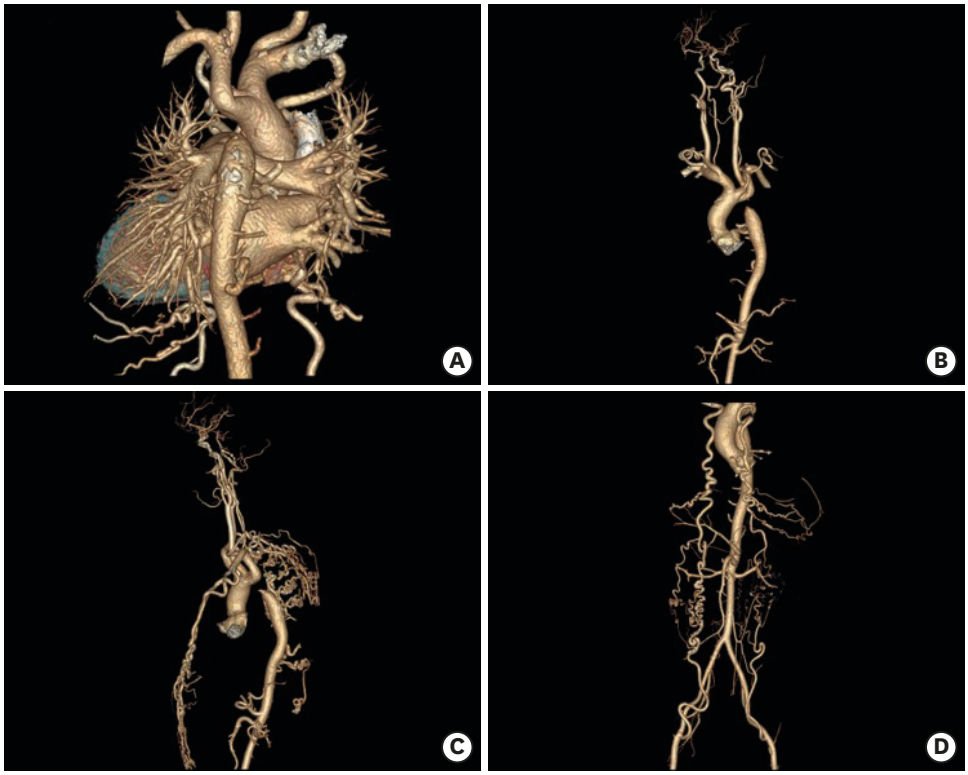

Figure 1. Cardiac CTA showed discontinuity of the aortic arch between the left subclavian artery and descending aorta without patent ductus arteriosus (A). Aorta CTA revealed interrupted aortic arch (B). Chest CTA showed multiple collateral vessels from the subclavian artery to costocervical trunk and to the thoracoacromial and descending scapular arteries (C). Aorta CTA showed multiple collateral channels and enlarged arteries from bilateral internal thoracic arteries to the superior epigastric artery, interior epigastric artery, and external iliac artery and from the subclavian artery to the vertebral artery, intercostal artery, and descending aorta (D).
CTA: computed tomography angiogram.

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defect was found on echocardiogram, left ventricular ejection fraction was 68%, and right ventricular size and function were within normal range without pulmonary hypertension (**Figure 2**). During coronary angiography, access was achieved through a right radial artery, but the guidewire could not pass though the right brachial artery. The angiogram showed occlusion of the brachial artery with good down-stream blood flow through a collateral vessel (**Figure 3A**). Percutaneous coronary intervention (PCI) was successfully performed though a left radial artery (**Figure 3B and C**). Aortogram confirmed IAA and collateral flow from intrathoracic arteries to the descending aorta (**Supplementary Video 1**). He was told that he needed surgery to correct a serious congenital heart disease. He denied surgery for financial reasons and did not experience further health problems. We closely followed the patient, provided optimal medical therapy for IAA, and performed PCI for coronary artery stenosis using a heart team approach.

IAA is a very rare congenital heart defect (affecting 3 per million live births)[1] in which the aorta is not completely developed. Defined as loss of luminal continuity between the ascending and descending portions of the aorta, this anomaly shows a very poor prognosis without surgical treatment.[2] IAA is associated with intracardiac malformation such as ventricular septal defect, patent ductus arteriosus, bicuspid aortic valve, left ventricular outflow tract obstruction, or aortopulmonary window. The first reported classification system, introduced by Celoria and Patton,[3] is still used almost universally. This system describes and classifies the site of aortic arch discontinuity, which can be distal to the left subclavian artery (type A), between the left carotid and left subclavian arteries (type B), or between the innominate and left carotid arteries (type C). In infants, clinical presentation

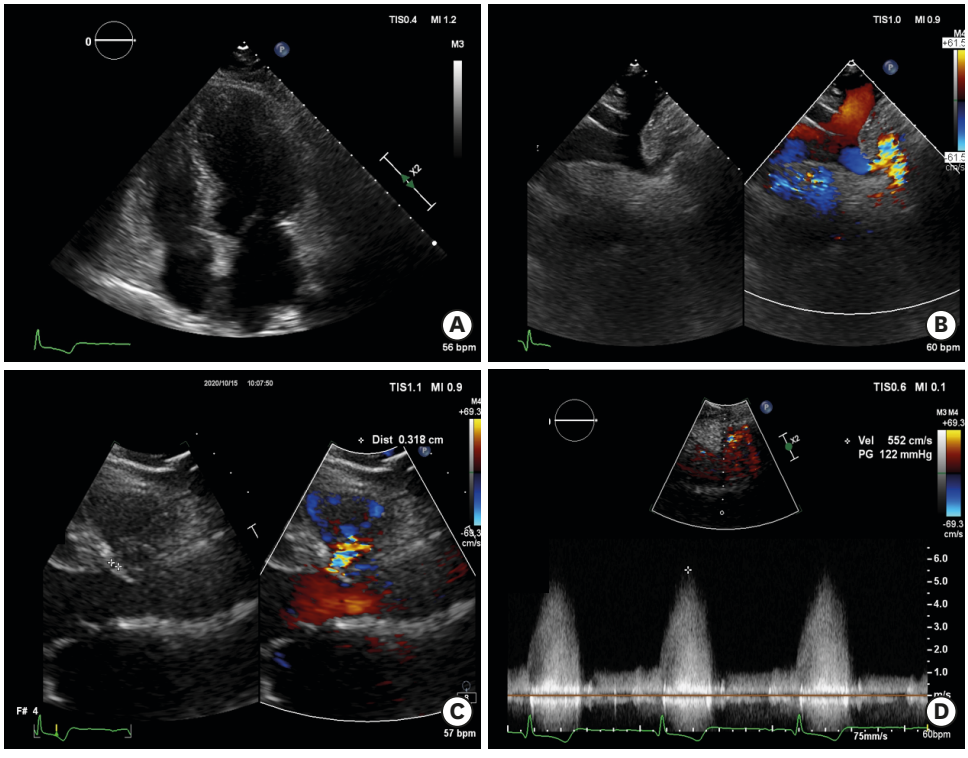

Figure 2. Echocardiogram showed normal left and right ventricular size and systolic function (A) and discontinuity of the aortic arch beyond the left subclavian artery (B). Subarterial type VSD was found by echocardiogram and showed 3.2 mm maximal diameter (C) and 122 mmHg of pressure gradient through the VSD (D).
VSD: ventricular septal defect.

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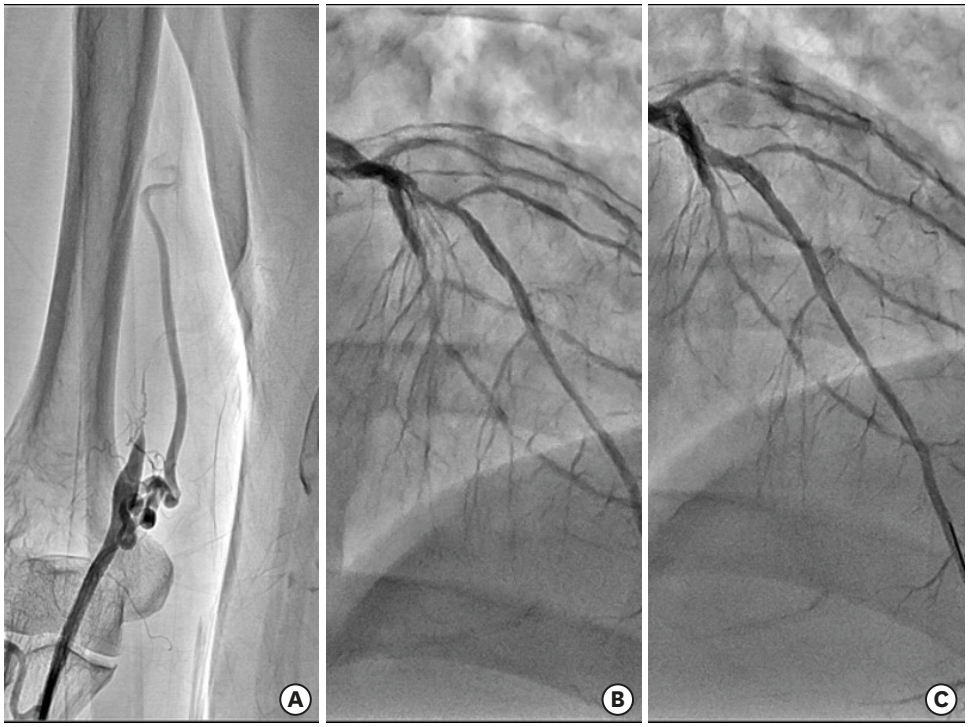

Figure 3. The right brachial artery was totally occluded, but there was good flow through a vigorous collateral vessel (A). PCI for mid-left anterior descending artery was successfully performed though a left radial artery. Pre-PCI (B). Post-PCI (C).
PCI: percutaneous coronary intervention.

involves severe congestive heart failure; if the condition is left untreated, 90% of affected infants die at a median age of 4 days.[4] We presented a rare case of IAA type A with substantial collateral circulation to maintain flow and enable survival without surgery until 50 years. However, collateral vessels are subject to atrophy and atherosclerosis, which can lead to other challenging problems.

SUPPLEMENTARY MATERIAL

Supplementary Video 1

Aortogram confirmed interruption of aortic arch and collateral flow from intrathoracic arteries to the descending aorta

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