Interruption of the Aortic Arch with Associated Cardiac Anomalies* 
Survival to Adulthood
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Interruption of the aortic arch is a rare and usually lethal cardiac malformation. We report a rare case of a patient with IAA distal to the left subclavian artery associated with double outlet right ventricle, subaortic VSD and patent DA who survived to adulthood. In this patient, the complete diagnosis was made by cardiac catheterization and angiocardiography. We discuss the probable mechanisms, diagnostic problems and therapeutic implications of such long survival.

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DA = ductus arteriosus; IAA = interruption of the aortic arch; NYHA = New York Heart Association; VSD = ventricular septal defect

Unless treated surgically, 75 percent of patients with IAA die within the first month of life and 90 percent within the first year.1 Patients with isolated IAA not accompanied by other cardiovascular anomalies occasionally may survive to advanced age.2 We report a patient who survived to adulthood without surgery. This patient had IAA and associated double outlet right ventricle, subaortic VSD and patent DA. To our knowledge, this is the first case report of an adult with IAA and associated cardiac anomalies documented correctly antemortem.

Case Report

A 21-year-old man presented to us with dyspnea on exertion (NYHA class 2) since 6 months of age. He was completely asymptomatic during infancy, childhood and adolescence. On examination, he had central cyanosis and grade 2 clubbing. There was no inequality of peripheral pulses or differential cyanosis and clubbing. Precordial examination revealed moderate cardiomegaly, left parasternal activity, pulsations in the second left interspace parasternally, single loud second sound and grade 3/6 ejection systolic murmur with grade 2/6 early diastolic murmur in the pulmonary area. The chest x-ray film showed scoliosis of the upper thoracic spine, cardiothoracic ratio of 0.6 and massive dilatation of the main pulmonary artery and its branches with peripheral pruning. Electrocardiography revealed coronary sinus rhythm and right ventricular strain. Cross-sectional echocardiography showed viscerocostal situs solitus, d-looped ventricles, a large malaligned subaortic VSD, more than 50 percent aortic override, mitral-aortic discontinuity and side-by-side great arteries with aorta to the right of the pulmonary artery. There was evidence of severe pulmonary hypertension. High parasternal and suprasternal views were suboptimal. At cardiac catheterization the diagnosis of IAA was considered when the retrograde femoral arterial catheter entered

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FIGURE 1. Main pulmonary artery angiogram in frontal view. The main pulmonary artery (MPA) is aneurysmally dilated and continues into the descending aorta (DAO) through the patent DA into the pulmonary artery via the DA and despite repeated attempts the aortic arch could not be entered. The main pulmonary artery angiogram in frontal (Fig 1) and lateral views showed aneurysmal dilatation of the pulmonary artery which continued via the DA into the descending aorta. There was mild pulmonary regurgitation. No arch vessel arose from the descending aorta. Therefore, a right axillary arterial puncture was made to enter the ascending aorta and left ventricle. An ascending aortic angiogram in the left anterior oblique view revealed its W-shaped trifurcation into the innominate, left common carotid and the left subclavian arteries followed by complete interruption of the aortic arch (Fig 2). There were no collaterals to the descending aorta. There was mild aortic regurgitation. The left ventricular angiogram in the left anterior oblique view showed a large subaortic VSD with a double outlet right ventricle. Hemodynamic and saturation data are summarized in Table 1. Thus, a diagnosis of IAA distal to the left subclavian artery, double outlet right ventricle, subaortic VSD, patent DA, mild aortic regurgitation and severe pulmonary hypertension with mild pulmonary regurgitation was made.

Discussion

Interruption of the aortic arch is a highly fatal anomaly; median age at death without surgery is ten days.1 Nearly all patients with IAA present in the neonatal period with severe congestive heart failure and with signs of inadequate systemic perfusion due to the combined effects of volume overload from left-to-right intracardiac shunt and the high afterload imposed by the closing ductus in the presence of complete aortic interruption. Why then did our patient survive unrecognized into adulthood? Presumably, the patient survived because the postnatal ductal closure and the
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To our knowledge, this is the first case report of an adult with IAA and associated anomalies in whom the complete diagnosis has been made angiographically. Two other cases of IAA with associated anomalies surviving to adulthood have been described earlier. However, Takashina et al. have made a diagnosis on the basis of right ventricular angiography, without performing an ascending aortic angiogram. Their case could very well have been a severe preductal coarctation of the aorta associated with VSD, patent DA and severe pulmonary hypertension. An ascending aortic angiogram is mandatory to establish a diagnosis of IAA and to differentiate it from severe preductal coarctation. The second case of Pierpont et al. was mistakenly diagnosed as a reversed DA when the patient was 16 years old with the use of a pulmonary angiogram. However, a correct diagnosis of IAA with associated anomalies could be established in adulthood only at autopsy. A similar problem could have been encountered in our case had we not done the ascending aortic angiogram.

To date, urgent surgery remains the only definitive means of treatment for neonates with IAA and other forms of left ventricular outflow tract obstructions. Surgery, however, is high-risk, technically difficult, expensive and has unencouraging results. Recently, a case has been made for pharmacologic treatment directed at keeping pulmonary vascular resistance high and the DA permanently open as a means of prolonging life in neonates with aortic atresia. This could also be extrapolated to IAA as exemplified by our case. Avoidance of thoracotomy in such neonates would improve the chances of successful heart-lung transplantation later in life.

What therapeutic options do we have for our patient? Advanced pulmonary vascular disease (Table 1) contraindicates corrective cardiac surgery. Since he has only NYHA class 2 symptoms, medical management is a rational option at present. But, like other patients with the Eisenmenger syndrome, he could be considered for heart-lung transplantation if he develops marked functional disability, repeated syncopal episodes, life-threatening complications, such as massive hemoptysis and symptoms of severe right ventricular failure. However, heart-lung transplantation in IAA could be technically more demanding since it would involve performing two procedures, viz., repair of the interruption with ductal division and heart-lung transplantation. These could be done at the same time through a median sternotomy using an interposed synthetic tube graft to establish continuity in the recipient’s descending thoracic aorta first, followed by heart-lung transplantation with the usual ascending aortic anastomosis. Alternatively, these could be done as staged procedures with the former being done through a lateral thoracotomy using direct end-to-end anastomosis, an interposed tube graft or endogenous vascular tissue as a conduit. The large nonrestrictive VSD would protect against postoperative RV failure that could result from duct closure. Heart-lung transplantation then could be performed later depending on donor availability.

**References**

2. Dische MR, Tsai M, Baltaxe HA. Solitary interruption of the arch of the aorta: clinicopathological review of eight cases. Am J Cardiol 1975; 35:271-77