

# An Adult with Aortic Arch Interruption Associated with Sinus Venosus Atrial Septal Defect and Partial Anomalous Pulmonary Venous Connection

Yu-Cheng Hsieh<sup>1,2</sup>, Tsu-Juey Wu<sup>1,2\*</sup>, Kuo-Yang Wang<sup>1</sup>, Kae-Woei Liang<sup>1,2</sup>,  
Wei-Wen Lin<sup>1</sup>, Ying-Tsung Chen<sup>1</sup>, Chih-Tai Ting<sup>1,2</sup>

<sup>1</sup>Cardiovascular Center, Taichung Veterans General Hospital, Taichung, and <sup>2</sup>Institute of Clinical Medicine and Cardiovascular Research Center, National Yang-Ming University School of Medicine, Taipei, Taiwan, R.O.C.

Interruption of the aortic arch is a rare and usually lethal congenital anomaly that is often associated with multiple cardiac malformations. Most neonates with aortic arch interruption perish once the ductus arteriosus closes after birth. However, sporadic cases have been reported to survive into adulthood uneventfully. Here, we report a 19-year-old male with a 3-month history of exertional dyspnea. A series of cardiovascular studies confirmed the presence of aortic arch interruption in conjunction with sinus venosus atrial septal defect and partial anomalous pulmonary venous connection. To the best of our knowledge, such an association has not been previously reported in adults. [*J Chin Med Assoc* 2007;70(1):30–32]

**Key Words:** atrial septal defect, interrupted aortic arch, partial anomalous pulmonary venous connection

## Introduction

Interruption of the aortic arch (IAA) is characterized by complete anatomic discontinuity between the ascending and descending aorta.<sup>1</sup> This congenital anomaly rarely occurs as an isolated lesion and is often associated with other intracardiac malformations, most commonly ventricular septal defect and patent ductus arteriosus (PDA).<sup>2,3</sup> Although 90% of affected infants die of circulatory failure within the first year of life, sporadic reports have documented survival into adulthood, which is rare but possible.<sup>4</sup> Here, we report a 19-year-old male with IAA associated with sinus venosus atrial septal defect (SVD) and partial anomalous pulmonary venous connection (PAPVC).

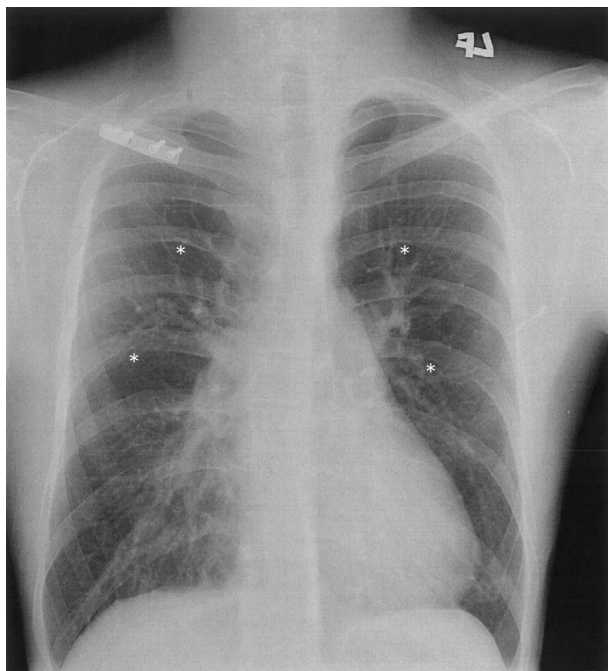
## Case Report

A 19-year-old male presented to our hospital with a 3-month history of exertional dyspnea and leg weakness,

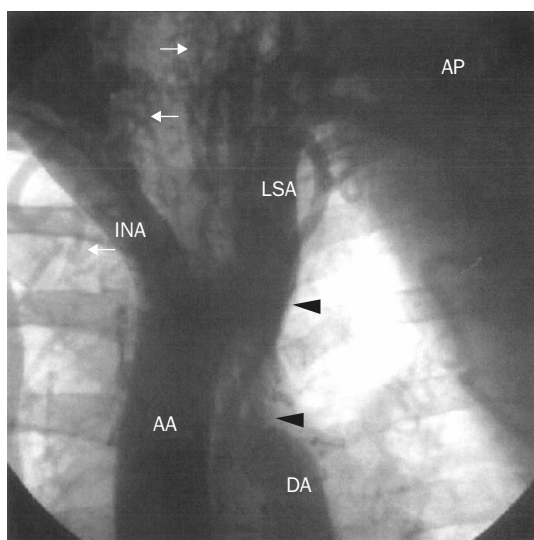
but no claudication. His past history was unremarkable except for trauma with right clavicle fracture sustained in a traffic accident 3 years previously. Blood pressure was 128/75 mmHg in the upper limbs, and 92/60 mmHg in the lower limbs. Lower-limb pulses were equal but weak, with a marked radial-femoral delay. Auscultatory findings were a grade 2/6 systolic ejection murmur over the left sternal border and a widely fixed split S<sub>2</sub>. Electrocardiography revealed right axis deviation, incomplete right bundle branch block and low atrial pacemaker rhythm. Transesophageal echocardiography showed SVD that was not visualized by transthoracic approach. Chest radiography disclosed a diminished aortic knob, increased pulmonary vascularity and bilateral rib notching (Figure 1). The patient was preliminarily diagnosed with coarctation of aorta with SVD and underwent cardiac catheterization for complete assessment.

Cardiac catheterization was performed via the right radial artery for the ascending aorta, right femoral artery for the descending aorta and right femoral vein

\*Correspondence to: Dr Tsu-Juey Wu, Cardiovascular Center, Taichung Veterans General Hospital, 160, Section 3, Chung-Kang Road, Taichung 407, Taiwan, R.O.C.  
E-mail: tjwu@vghtc.gov.tw • Received: April 12, 2006 • Accepted: August 18, 2006

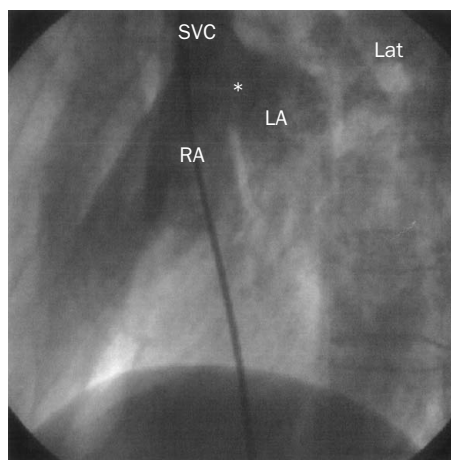


**Figure 1.** Chest radiography shows a diminished aortic knob, increased pulmonary vascularity and bilateral rib notching (asterisks).



**Figure 2.** Ascending aortogram (anteroposterior view) shows complete aortic interruption immediately distal to the origin of the left subclavian artery (arrowheads). Note that the descending aorta is being filled by extensive collateral vessels (arrows). AP = anteroposterior view; INA = innominate artery; LSA = left subclavian artery; AA = ascending aorta; DA = descending aorta.

for the right heart. Ascending aortogram showed a complete interruption of the aorta immediately distal to the origin of the left subclavian artery (Figure 2). Delayed filling of the descending aorta was constructed via extensive collateral vessels. The descending aortogram demonstrated the interrupted end and



**Figure 3.** Superior vena cava (SVC) angiogram (lateral view) shows an anomalous connection (asterisk) in the high interatrial septum, the sinus venosus atrial septal defect. Note that the pulmonary catheter was advanced to the anomalous right superior pulmonary vein through the right atrium and SVC, proving the presence of partial anomalous pulmonary venous connection to the SVC. Lat = lateral view; RA = right atrium; LA = left atrium.

excluded the existence of PDA. During right heart catheterization, the anomalous right superior pulmonary vein (RSPV) could be directly approached from the superior vena cava (SVC), with a simultaneous oxygen saturation of 94.6% at the SVC, verifying the presence of PAPVC. With the tip of the diagnostic catheter located at the junction of the SVC and anomalous RSPV, SVC angiogram showed the presence of SVD in the high interatrial septum (Figure 3).

After catheterization study, the diagnosis of IAA with SVD and PAPVC was confirmed. Due to the progression of clinical symptoms and the complexity of this congenital anomaly, surgical treatment was suggested. However, the patient was reluctant to receive surgery and was regularly followed up instead.

## Discussion

IAA is a rare and usually lethal congenital anomaly that occurs in 5.8 per 100,000 live births.<sup>2</sup> This congenital anomaly is frequently associated with multiple cardiovascular malformations, such as PDA, ventricular septal defect, atrial septal defect, subaortic stenosis, truncus arteriosus, aortopulmonary window, double-outlet right ventricle, and DiGeorge syndrome.<sup>2,5</sup> Celoria and Patton classified IAA into 3 types based on the site of interruption.<sup>1</sup> This patient was classified as having type A IAA as the interruption was distal to the origin of the left subclavian artery. In addition, the pulmonary catheter directly entered the anomalous

RSPV via the SVC with a simultaneous oxygen saturation step-up at the SVC during catheterization, proving the concomitant PAPVC. SVC angiography further confirmed the anomalous communication between the right atrium and left atrium in the high interatrial septum, the SVD. Consequently, this patient was diagnosed with type A IAA with SVD and PAPVC, which has not been reported in an adult before. In this patient, the definite diagnosis was completed by conventional cardiac catheterization. However, magnetic resonance angiography or multislice computed tomography can provide noninvasive visualization of the SVD as well as the pulmonary veins and definitely help to establish the diagnosis.<sup>6,7</sup>

In neonates with IAA and PDA, the ascending aortic flow normally comes from the left ventricle, while the descending blood is supplied from the right ventricle through the ductus arteriosus. However, constriction of the ductus arteriosus after birth would abolish the flow distal to the interruption. The grave sequelae include cardiogenic shock, pulmonary edema, acute renal failure and finally death. There are 2 possible mechanisms to explain how this patient survived uneventfully into adulthood. First, Higgins et al<sup>8</sup> have postulated that some cases of type A IAA might contribute to the progression of severe aortic coarctation to complete occlusion. It is possible that the IAA in this patient had followed severe aortic coarctation. During the period of progression to complete aortic interruption, the collaterals gradually developed to compensate for the decreased descending flow. Secondly, the extensive collateral vessels may have already developed *in utero* so that the obliteration of the ductus arteriosus after birth did not completely cut off the descending flow.<sup>9</sup>

The clinical prognosis in patients with IAA depends largely on the associated congenital anomalies. In this patient, although the descending blood flow is being maintained by collaterals at present, the collaterals are liable to atherosclerosis and atrophy, which would result in other challenging problems.<sup>10</sup> Furthermore, the associated SVD and PAPVC would eventually lead to congestive heart failure requiring surgical intervention.

Surgical intervention by reconstructing aortic continuity and correcting the associated SVD and PAPVC is the preferred treatment for this patient.

In conclusion, IAA associated with SVD and PAPVC is an extremely rare congenital anomaly, especially in adult patients. Once the diagnosis is established, surgical intervention is the definitive treatment.

## Acknowledgments

This study was supported in part by the Yen Tjing Ling Medical Foundation and by a grant from Taichung Veterans General Hospital (TCVGH-943105C, to Tsu-Juey Wu), Taiwan, R.O.C.

## References

1. Celoria GC, Patton RB. Congenital absence of the aortic arch. *Am Heart J* 1959;58:407-13.
2. Loffredo CA, Ferencz C, Wilson PD, Lurie IW. Interrupted aortic arch: an epidemiologic study. *Teratology* 2000;61:368-75.
3. Collins-Nakai RL, Dick M, Parisi-Buckley L, Fyler DC, Castaneda AR. Interrupted aortic arch in infancy. *J Pediatr* 1976;88:959-62.
4. Messner G, Reul GJ, Flamm SD, Gregoric ID, Opfermann UT. Interrupted aortic arch in an adult: single-stage extra-anatomic repair. *Tex Heart Inst J* 2002;29:118-21.
5. Lee ML, Wang JK, Wu MH, Lue HC. Morphologic features of the interrupted aortic arch in Chinese patients. *Am J Cardiol* 1995;76:982-5.
6. Otsuka M, Itoh A, Haze K. Sinus venosus type of atrial septal defect with partial anomalous pulmonary venous return evaluated by multislice CT. *Heart* 2004;90:901.
7. Varghese A, Gatzoulis M, Mohiaddin RH. Magnetic resonance angiography of a congenitally interrupted aortic arch. *Circulation* 2002;106:9-10.
8. Higgins CB, French JW, Silverman JF, Wexler L. Interruption of the aortic arch: preoperative and postoperative clinical, hemodynamic and angiographic features. *Am J Cardiol* 1977;39:563-71.
9. Bapat VN, Tendolkar AG. Aortic arch interruption with rheumatic mitral stenosis and Ebstein's anomaly. Report of a case with successful surgical outcome. *Tex Heart Inst J* 1997;24:362-5.
10. Akdemir R, Ozhan H, Erbilin E, Yazici M, Gunduz H, Uyan C. Isolated interrupted aortic arch: a case report and review of the literature. *Int J Cardiovasc Imaging* 2004;5:389-92.