An inoperable aortic arch interruption in a patient with differential cyanosis

Chengming Fan and Jinfu Yang*

Department of the Cardiothoracic Surgery, The Second Xiangya Hospital, Central South University, Middle Renmin Road 139, Changsha 410011, China

* Corresponding author. Tel: +86 731 85296107, Fax: +86 731 85296606, Email: yangjinfu0326@163.com

A 9-year-old Chinese boy with a 6-year history of heart murmur and progressive dyspnoea was referred to our institution for minimally invasive transthoracic patent ductus arteriosus (PDA) closure and ventricular septal defect (VSD) closure. Physical examination revealed differential cyanosis with an oxygen saturation in the upper and lower extremity of 95 and 82%, respectively. Blood pressures in the arm is 120/90 and 100/70 mmHg in the limb. A loud P2 with a grade 3/6 systolic murmur was audible at the second left intercostals space with radiation to the back. An echocardiogram from local county hospital revealed a non-restrictive VSD and a large PDA. A computed tomography angiography was performed and demonstrated an aortic arch interruption (IAA) with the absence of arch (Panel A, arrow), pulmonary arterial dilation (Panel B), and aortic discontinuity distal to the left subclavian artery (type A) (Panel C). A left ventriculography confirmed an IAA with a clear separation between three branches of aortic arch and the descending aorta (Panel D). A right heart catheterization revealed severe fixed pulmonary hypertension with a mean pulmonary artery pressure of 84 mmHg and pulmonary vascular resistance of 11 Woods units. Since the patient had fixed pulmonary hypertension, the surgical correction was unfortunately not performed. The patient was then referred to heart and lung transplantation.

This case highlights the essence of enhancing the level of healthcare and services in Chinese rural areas. Failure to identify IAA in this case of attempted PDA or VSD closure will lead to catastrophic lower-half ischaemia.

Panels A–D: Asc Ao, ascending aorta; CC, left common carotid artery; DAO, descending aorta; LA, left atrium; LPA, left pulmonary artery; LSC, left subclavian artery; mPA, main pulmonary artery; Inn, innominate artery; RA, right atrium; RPA, right pulmonary artery; SVC, superior vena cava.