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Brief Report

Implantation of drug-eluting stents for relief of obstructed infra-cardiac totally anomalous pulmonary venous connection in isomerism of the right atrial appendages

Derek T. H. Wong,[1] Shi-Joon Yoo,[1,2] Kyong-Jin Lee[1]

[1]*Division of Cardiology, Department of Pediatrics;* [2]*Department of Diagnostic Imaging, The Labatt Family Heart Centre, The Hospital for Sick Children, University of Toronto School of Medicine, Toronto, Canada*

Abstract We describe an infant with severe obstruction of infra-cardiac totally anomalous pulmonary venous connection associated with right isomerism, atrioventricular septal defect, pulmonary atresia, and multiple aortopulmonary collateral arteries. Implantation of a stent into the obstructed descending vertical vein provided effective palliation, with a dramatic increase in saturations of oxygen obviating the need for urgent high-risk surgery.

Keywords: Heterotaxy; asplenia; congenital cardiac disease

Children with isomerism of the right atrial appendages frequently have a poor prognosis in consequence of the complex combinations of cardiac abnormalities known to exist in this setting.[1,2] For example, operative mortality after repair of totally anomalous pulmonary venous connection is known to be significantly increased when associated with functionally univentricular physiology, earlier surgical repair, and in the presence of obstruction,[3] features which often come together when there is right isomerism. Progressive pulmonary venous stenosis may also be relentless after repair, particularly when the anomalous pulmonary venous connection is infracardiac.[3] Evolving strategies to improve survival have included implantation of stents to provide temporary relief of severe obstruction in the effort to postpone surgery, and more aggressive surgical techniques.[1,4–7] Drug-eluting stents have been implanted in adults with coronary arterial disease to decrease neointimal proliferation.[8] With all these aspects in mind, we present here our experience with a critically ill neonate with right isomerism and obstructed infracardiac totally anomalous pulmonary venous connection whom we palliated using a drug-eluting stent.

Correspondence to: Dr Kyong-Jin Lee, The Hospital for Sick Children, 555 University Avenue, Toronto, Canada M5G 1X8. Tel: 416 813 7326; Fax: 416 813 5857; E-mail: kyong-jin.lee@sickkids.ca

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Case report

A male infant, born at term weighing 3.9 kilograms, presented with respiratory distress and a saturation of oxygen of 50% immediately after birth. With 100% oxygen, pre- and post-ductal saturations of oxygen were 72%. The infant was intubated and ventilated, commenced on prostaglandin, and transferred to our institution. Echocardiographic interrogation revealed right isomerism, a left-sided heart with the apex pointing to the left, left-sided aortic arch, an atrioventricular septal defect with a common atrioventricular valve appropriately shared between the ventricles, and pulmonary atresia, with the aorta arising exclusively from the right ventricle. There were bilateral superior caval veins, each connecting to the atrial roof, and an inferior caval vein draining to the left-sided atrium. Confluent but hypoplastic central pulmonary arteries were identified, but the sources of flow of blood to the lungs were not fully delineated. Only 2 pulmonary veins were visualized with certainty. They drained via a small confluence to

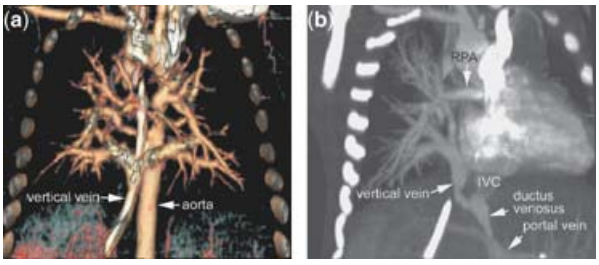

Figure 1.
*Computed tomographic angiograms. (**a**) Volume rendered three-dimensional image reconstructed in a coronal plane shows major aortopulmonary collateral arteries originating from the descending aorta. (**b**) Maximum intensity projection image reconstructed in an oblique coronal plane shows the pulmonary veins draining to the vertical vein, which in turn connects to the venous duct near its junction with the inferior caval vein. The vertical vein shows stenosis of its lower segment. Key: RPA = right pulmonary artery; IVC = inferior caval vein*

a descending vertical vein that was obstructed at its connection to the venous duct, with a mean gradient of 9 mmHg. A contrast-enhanced computed tomographic scan was performed further to delineate the anatomy (Fig. 1). It demonstrated 4 pulmonary veins draining via a confluence to a descending vertical vein, which inserted into the venous duct just adjacent to the inferior caval vein. There was complex pulmonary arterial anatomy supplied by 5 major aortopulmonary collateral arteries.

The infant continued to remain severely desaturated at 60%, with a partial pressure of oxygen of 35 mmHg when breathing 100% inspired oxygen. The critical condition of the infant necessitated prompt relief of the pulmonary venous obstruction. On the other hand, the complexity of the pulmonary vascular bed, with collateral arteries of good size, favoured delayed surgical intervention. Cardiac catheterization was performed at 22 hours of age. A 4 French catheter (Super Torque, Cordis, Cordis Corporation, Miami, FL) was positioned in the inferior caval vein and the insertion of the descending vertical vein to the venous duct was crossed with a 0.014 inch Wizdom™ wire (Cordis Corporation, Miami, FL) (Fig. 2). Two overlapping paclitaxel drug-eluting stents (TAXUS®, Boston Scientific Corporation, Natwick, MA, of 4.5 mm diameter and 12 and 16 mm long, were implanted into the descending vertical vein (Figs 2c, d). The indication for the second stent was to push intimal tissue protruding through the struts of the first stent against the vessel wall. Saturations of oxygen immediately increased to 80% in room air. The time needed for fluoroscopy was 15 minutes, and the overall procedural time was 82 minutes. The infant was extubated 48 hours after implantation of

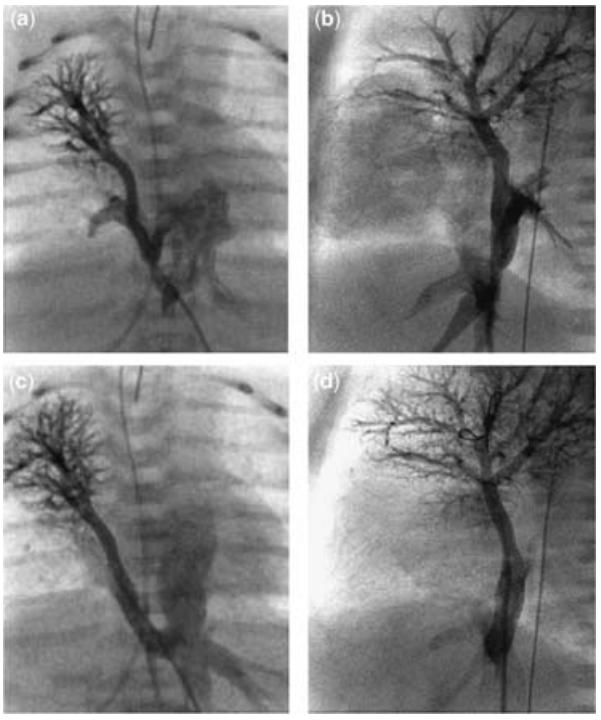

Figure 2.
*Angiograms showing anterior-posterior and lateral views of the obstructed totally anomalous pulmonary venous connection before [(**a**) and (**b**)], and after [(**c**) and (**d**)] implantation of the stents.*

the stent. He was discharged home on the 13th day of life, with saturations of oxygen of 80% in room air, medicated with clopidogrel and low molecular weight heparin, chosen at the preference of the physician. At 7 weeks of age, the infant was thriving, with saturations of oxygen of 85%. Cardiac catheterization was electively performed to re-examine the stents and the pulmonary arterial anatomy. There was no evidence of in-stent stenosis, but a narrowing had developed at the junction of the stents with the inferior caval vein. This was effectively relieved by implantation of an additional paclitaxel drug-eluting stent, of 4.5 mm diameter and 16 mm long. Angiography of the aortopulmonary collateral vessels confirmed the previous findings from computerised tomography. The infant continued to do well, with maintenance of saturations of oxygen in the mid 80's. At 6 months of age, the infant underwent unifocalization of the aortopulmonary vessels, placement of a 5 mm modified right Blalock-Taussig shunt, division of the vertical vein, and extraction of the stents. Intra-operative exploration demonstrated small orifices of both right and the left lower pulmonary veins necessitating repair which was performed utilizing the sutureless technique.[1] Echocardiography 24 days post-operatively demonstrated unobstructed pulmonary venous flow. The infant was discharged on the

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48th post-operative day. Future candidacy for further functionally single or biventricular surgery remains to be determined.

Discussion

To the best of our knowledge, ours is the first reported case of stenting of obstructed infracardiac pulmonary venous return. Right isomerism is generally accompanied by complex cardiac anatomy. Of necessity, the pulmonary venous return is anatomically anomalous, with up to half of cases being obstructed.[1,2] A 26-year review estimated overall rates of survival for patients with right isomerism of 71% at 1 month, 49% at 1 year, and 35% at 5 years.[2] Mortality in infants requiring neonatal cardiovascular surgery was 75%, versus 51% for those having a later first operation.[2] Surgical mortality with pulmonary venous repair was 95%. Evolving therapeutic strategies, including sutureless repair of totally anomalous pulmonary venous drainage, has resulted in improved survival, but obstruction of the infracardiac type remains a significant risk factor for mortality.[1]

Bare metal stenting has been reported temporarily to relieve obstruction of the ascending vertical vein in the setting of totally anomalous pulmonary venous connection.[4–6] The potential benefit of drug-eluting stents is attenuation of neointimal hyperplasia associated with implantation of the stents, as well as the progressive pulmonary venous obstruction of right isomerism.[8] There is large experience showing benefit with such implantation of drug-eluting stents in adults with coronary arterial disease.[8] For other blood vessels, implantation of drug-eluting stents have been reported into the arterial duct feeding an isolated left pulmonary artery,[9] a conduit in the right ventricular outflow tract of an infant,[10] and the vertical vein of a supracardiac totally anomalous pulmonary venous connection.[7] It remains to be determined whether superior protection is offered by using drug-eluting over bare-metal stents in the prevention of neointimal proliferation associated with congenital cardiac disease. In this regard, the palliative stenting provided prompt and effective relief of critical obstruction, and obviated the need for urgent cardiac surgery. Such postponement of cardiac surgery may improve the clinical outcomes in this population, with a known poor prognosis when early surgery is required.

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