

Rescue Circumflex Artery Stenting for Major Right Ventricular Dysfunction after Ross-Konno Operation

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ABSTRACT

A single coronary artery is an anomaly in which only 1 coronary artery arises from the aortic root, supplying the entire heart. We report the case of a 5-year-old girl in whom a single coronary artery ostium was discovered during a Ross-Konno operation. This patient required extracorporeal membrane oxygenation for postoperative right ventricular infarction due to proximal stenosis of a dominant left circumflex artery in combination with an occluded minor right coronary artery. Rescue stent implantation allowed for recovery of right ventricular function and weaning from extracorporeal membrane oxygenation.

INTRODUCTION

A single coronary artery is an unusual congenital anomaly in which only 1 coronary artery arises from the aortic root by a single coronary ostium, supplying the entire heart [Kang 2006; Hussain 2008]. We describe a rare case of a suspected single coronary artery, discovered during a Ross-Konno procedure, which led to inadequate myocardial ischemic protection of the right ventricle.

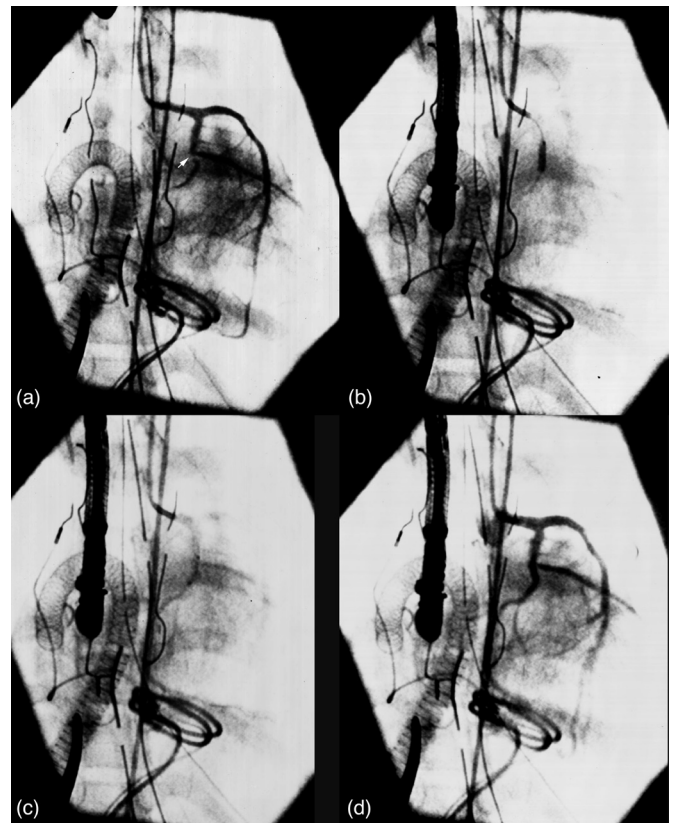
CASE REPORT

The case patient was a 5-year-old girl who had previously undergone patent arterial duct ligation at age 1 year and aortic, mitral, and tricuspid repair at age 4 years for congenital subvalvular aortic stenosis and functional mitral and tricuspid regurgitation at another institution. She was referred to our institution for recurrent severe subvalvular aortic stenosis with mild aortic regurgitation and severe mitral regurgitation. The child underwent a Ross-Konno operation and mitral valve repair. Cardiopulmonary bypass was established by femorofemoral cannulation before redo sternotomy.

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The aorta was cross-clamped and vented from the right superior pulmonary vein, and the aorta was transected. Antegrade crystalloid cardioplegia was infused selectively into the only visible coronary ostium. Careful examination revealed no other coronary ostium. Because the entire heart was arrested by selective cardioplegia into this ostium, the presence of a single coronary artery was suspected, and the epicardial coronary anatomy was not explored. The mitral valve was first repaired through a left atriotomy by implantation of



Left coronary angiography in 30° right anterior oblique. A, Critical stenosis of the proximal left circumflex artery (arrow), just distal to first marginal artery. B, Balloon dilatation of the left circumflex artery stenosis. C, Stent being deployed. D, Left coronary angiography showing resolution of the stenosis.

a 16-mm Bioring Kalangos annuloplasty ring, with no residual leak on water testing. Subaortic fibrous tissue was excised, and a Ross-Konno operation was performed, with reimplantation of the single coronary artery. An 18-mm cryopreserved aortic homograft was used to reconstruct the right ventricular outflow tract. The aortic cross-clamping time was 134 minutes.

Weaning from cardiopulmonary bypass was impossible. Transesophageal echocardiography showed biventricular systolic dysfunction and a dyskinetic interventricular septum due to the ventricular septal patch. Cardiopulmonary bypass was converted to venoarterial extracorporeal membrane oxygenation. Troponins were initially at 20.4 µg/L, a finding attributed to the ventricular-septal incision, but increased to 139.61 µg/L during the next 48 hours. Despite the worsening cardiac enzymes, the patient's hemodynamic status improved under extracorporeal membrane oxygenation, allowing progressive weaning to 25% of the predicted cardiac output at 48 hours. Transesophageal echocardiography at that time showed an akinetic right ventricular free wall, with normal left ventricular systolic function and discrete residual mitral regurgitation. A coronary angiogram was performed to assess the coronary anatomy in the operating room. The single coronary ostium was engaged with an ART5 F Judkins 3.5 guiding catheter. Angiography showed a left coronary anatomy with a normal left main coronary artery, nonsignificant irregularities of the midportion of the left anterior descending artery, and a significant stenosis of the circumflex artery distal to the first marginal branch (Figure, part a). The lesion was crossed with a 0.014 BMW floppy guidewire without difficulty. An Invatec Avion 2 × 10 mm angioplasty balloon (Invatec S.p.A., Roncadelle, Italy) was inflated to 2 atmospheres (Figure, part b), with a residual stenosis greater than 50%. A Biotronik PRO-kinetic 2 × 8 mm stent (Biotronik, Berlin, Germany) was then deployed with 16 atmospheres ballooning to obtain optimal luminal diameter (Figure, part c). The poststent angiography showed a well-expanded stent with restoration of normal left coronary arterial flow (Figure, part d). The right coronary artery was not visualized, however. Loading doses of clopidogrel and acetylsalicylic acid were administered immediately after stent implantation.

Troponins progressively decreased to 52.278, and extracorporeal membrane oxygenation was weaned successfully under levosimendan and adrenaline 5 days after stenting. Transesophageal echocardiography showed recovery of contractility of the right ventricular free wall. Hemodynamics improved during the following 24 hours; however, the child presented with septic shock, with subsequent positive blood cultures for *Staphylococcus hominis*. Despite empirical antibiotherapy (vancomycin and meropenem), fluid resuscitation, increase in adrenaline, and introduction of noradrenaline, the patient died 24 hours later.

DISCUSSION

Myocardial protection during aortic cross-clamping is of primary importance, particularly during long ischemic times such as are common in complex biventricular outflow tract reconstruction. Coronary anomalies and stenoses

can complicate optimal myocardial protection and induce myocardial infarction [Gaudino 1997]. On weaning from cardiopulmonary bypass, our patient presented biventricular systolic dysfunction, which was attributed to the long cross-clamp time and dyskinetic ventricular septal patch of the Konno left ventricular outflow tract enlargement. Isolated right ventricular dysfunction was evident only after 48 hours of extracorporeal membrane oxygenation support, and was likely secondary to stenosis of a dominant circumflex artery and chronically occluded minor right coronary artery. A congenital anomaly of the right coronary artery cannot be excluded; however, previous reports have demonstrated complete opacification of the right coronary from the circumflex artery [Kang 2006], which was not the case in our patient. Circumflex artery lesion caused by sutures is a known complication of mitral annuloplasty [Virmani 1982]. The middle portion of the circumflex artery, however not the proximal, is at risk, because that is the location closest to the mitral annulus. Furthermore, the Bioring Kalangos (Lonay, Switzerland) biodegradable annuloplasty ring is implanted subendocardially into the mitral annulus and does not require suture placement. Focal coronary stenosis is a known late complication of Kawasaki disease [Newburger 2004]; however, our patient had no previous relevant history. The association of multiple focal coronary lesions makes this hypothesis the most probable. Screening for congenital lipid disorders was not performed, because the clinical presentation and lesions were not characteristic [Tissot 2007].

At the time of catheterization, the patient was under full inotropic and extracorporeal membrane oxygenation support. Surgical revascularization was not considered an option, because the coronary anastomosis of a bypass graft would require cardioplegia, which had previously proven inadequate. The choice of revascularization was thus initially for a percutaneous angioplasty, performed by using a bare-metal device rather than a drug-eluting stent, because there are no evidence-based data regarding the advantages of the drug-eluting stent in the pediatric population [Morice 2002; Moses 2003; Schofer 2003; Tissot 2007]. After stenting, the patient's troponins rapidly decreased and right ventricular systolic function recovered, allowing extracorporeal membrane oxygenation weaning.

CONCLUSION

Percutaneous coronary artery stent implantation may be considered a valid rescue method to ensure patency of significant coronary artery stenosis in children in a critical state after complex congenital cardiac surgery. Further studies are required to determine the long-term benefits and disadvantages of this technique.

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