Catheterization and Cardiovascular Interventions 74:1085–1088 (2009)

Case Report

Relief of Severe Immediate Postoperative Superior Vena Cava Stenosis with Covered Stent: Case Report With Midterm Follow Up

Raul I. Rossi F.,* MD, MSc, FSCAI and João Luiz L. Manica, MD

Sinus venous atrial septal defects are commonly associated with abnormal pulmonary venous connection. Numerous surgical techniques have been proposed with excellent short- and long-term outcomes. Pulmonary and superior vena cava obstructions, as well as rhythm disturbances, are the most common problems seen during follow up. However, acute postoperative superior vena cava obstruction with successful percutaneous covered-stent implantation has not been reported in the literature. The objective of this study is to report a unique case of acute obstruction of the superior vena cava on the first postoperative day after sinus venous atrial septal defect repair in an infant who was successfully relieved by a percutaneous angioplasty with covered-stent implantation, and the midterm follow up after this intervention. © 2009 Wiley-Liss, Inc.

Key words: superior vena cava syndrome; pediatric interventions; complications adult cath/intervention

INTRODUCTION

Sinus venous atrial septal defects occur when there is an unroofing of the right upper pulmonary vein into the superior vena cava, and are commonly associated with an abnormal pulmonary venous connection, which precludes the possibility of closing this defect by percutaneous intervention [1]. Numerous surgical techniques have been proposed with excellent short- and long-term outcomes [2,3]. Pulmonary and superior vena cava obstructions, as well as rhythm disturbances, are the most common problems seen during follow up. However, acute postoperative superior vena cava obstruction with successful percutaneous stent implantation has not been previously reported in literature.

The objective of this study was to report a unique case of acute obstruction of the superior vena cava on the first postoperative day after sinus venous atrial septal defect repair, in an infant, who was successfully relieved by a percutaneous angioplasty with a covered-stent implantation, and the midterm follow up of this intervention.

CASE REPORT

An 18 month-old-boy diagnosed with a large sinus venous atrial septal defect was referred to our institution. An electrocardiogram showed sinus rhythm and right ventricular hypertrophy. The chest X-ray showed cardiomegaly with increased pulmonary vascular markings. On physical examination, there was a short ejection pulmonary outflow murmur and fixed splitting of the second sound.

The patient was submitted to surgical repair of the defect who demanded 65 min of extracorporeal circulation and 45 min of cross clamp time. During the surgical procedure, abnormal pulmonary venous drainage from the upper right veins into the lateral aspect of the superior vena cava was diagnosed. Closure of the defect was performed using a single patch technique,

Congenital Heart Disease Interventional Unit, Institute of Cardiology of Rio Grande do Sul-FUC, Porto Alegre, RS, Brazil

Conflict of interest: Nothing to report.

*Correspondence to: Raul I. Rossi F., MD, MSc, FSCAI, Unidade de Pesquisa do IC/FUC, Av. Princesa Isabel, 370 Santan, Porto Alegre, RS – 90620-000, Brazil. E-mail: rossi.pesquisa@cardiologia.org.br

Received 23 April 2009; Revision accepted 6 June 2009

DOI 10.1002/ccd.22189
Published online 7 August 2009 in Wiley InterScience (www.interscience.wiley.com)

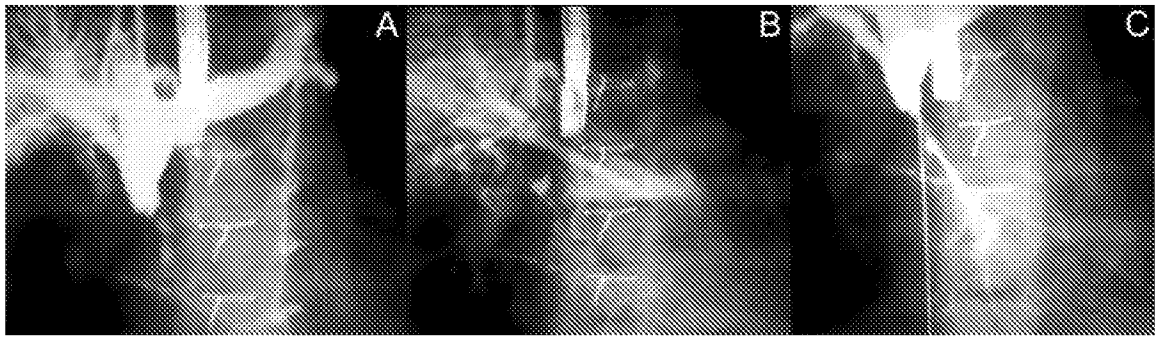

Fig. 1. A: Hand injection angiography through the jugular sheath is documented near the occlusion of the superior vena cava (SVC) and anomalous drainage of the right upper pulmonary vein. B: Reverse jet of flow in the pulmonary trunk clearly demonstrates severe intravenous pressure and anomalous pulmonary venous drainage. C: After dilation of the lesion with a 3 mm PTCA balloon (WorldPass, Boston Scientific, Co.), a super-stiff exchange guide wire (Amplatz, Boston Scientific, Co.) was again snared, thus establishing a venous rail.

and the abnormal pulmonary venous drainage was difficult to correct and was left in place.

The transoperative period proceeded normally and the patient arrived at the intensive care unit in need of inotropic support and showed differential cyanosis as part of the superior vena cava syndrome. The surgical team suspected that the cause of the symptoms was an eventual extrinsic compression due to an inflammatory reaction and local edema. An electrocardiogram at that time showed low-frequency junctional rhythm and pacing was required. During the initial hours of postoperative period, the patient developed significant edema and cyanosis of the trunk, head, and neck. A transthoracic echocardiogram showed an obstruction of the superior vena cava, but could not rule out the possibility of a thrombus as the cause of the problem. The patient was then submitted to transesophageal echocardiography which confirmed the subtotal superior vena cava obstruction at the junction with the right atrium.

The patient was sent to the catheterization laboratory in an attempt to relieve the obstruction in <24 hr after the surgical procedure. Venous access was obtained through the right-internal jugular and right-femoral veins. A hand injection angiography was made through the jugular sheath and documented the near-occlusion of the superior vena cava. Also, the anomalous drainage of the right-upper pulmonary vein was clearly demonstrated. Because of the severe intravenous pressure, there was reverse flow to the pulmonary trunk (Fig. 1).

The obstruction was crossed with a flexible 0.014″ hydrophilic-coated guide wire (Choice PT2, Boston Scientific Corporation, Natick, MA), which was snared with a nitinol gooseneck snare catheter (Amplatz Snare; Microvena, White Bear Lake, MN) from below, allowing support to dilate the obstruction with a 3 mm PTCA balloon (WorldPass, Boston Scientific, Co.).

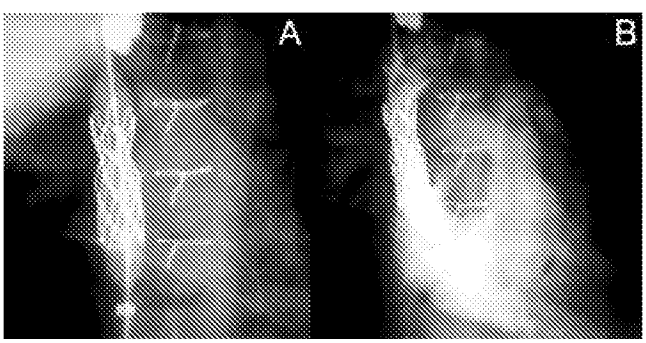

Fig. 2. A: Implantation of a 34 mm covered stent (CP-stent, NuMed, Inc., Laboratories, Hopkinton, NY) mounted on a 10 mm PowerFlex balloon (Cordis, Inc.). B: Control angiography demonstrates a residual obstruction remaining in the midportion of the stent.

After enlarging the small hole, a 4 F right coronary Judkins (Cordis, Inc., Miami, FL) was put across the obstruction, and a super-stiff exchange guide wire (Amplatz, Boston Scientific, Co.) was again snared, thus establishing a venous rail. Afterward, a 6 mm high-pressure balloon was used to further enlarge the obstruction, and finally, a 13 F Mullins sheath (Cook Medical, Inc., Bloomington, IN) introduced from the femoral vein allowed implantation of a 34 mm covered stent (CP-stent, NuMed, Inc., Laboratories, Hopkinton, NY) mounted on a 10 mm PowerFlex balloon (Cordis, Inc.). The last inflation pressure was 12 atm. Despite this procedure, a residual obstruction remained in the middle of the stent (Fig. 2). As there was marked angiographic and also clinical improvement, we believed that the result was acceptable and completed the procedure. The pullback gradient across the residual lesion was not measured at that time. The stent did not interfere with the abnormal venous drainage from the right superior pulmonary vein, and it was not considered a reason for concern due to the benign course

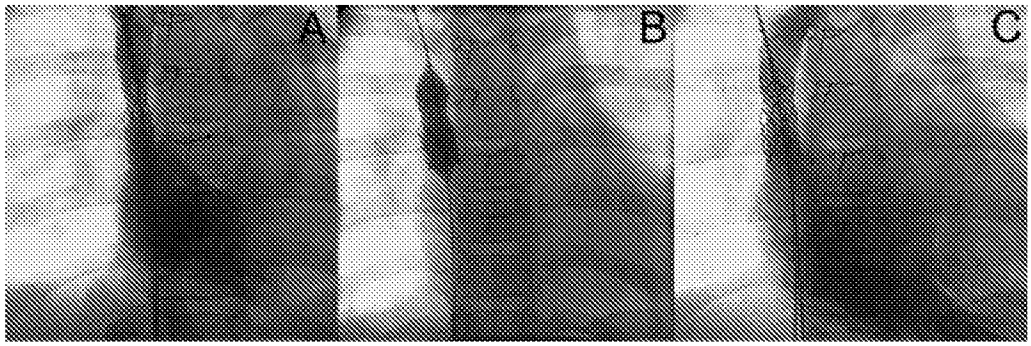

Fig. 3. A: The previously implanted stent shows mild neointimal hyperplasia with a residual stricture at the medium third causing a mild pressure gradient in the superior vena cava and right atrium. B: The lesion was dilated with a14 mm Z Med II balloon (NuMed, Inc., Laboratories, Hopkinton, NY). C: Angiographic result and evidence of relief of the residual obstruction.

of this situation. There was no vascular complication attributed to the use of the 13 F sheath in this case.

The intraprocedural transesophageal echocardiography also documented an immediate relief of the obstruction with the presence of triphasic flow through the superior vena cava. The patient developed pulmonary congestion and bilateral pleural effusion and was initially treated with intravenous diuretics. On postintervention day 2, the patient was extubated and inotropic agents were suspended. The pleural effusion improved and oral diuretics were stopped on postintervention day 6. On postintervention day 7, the patient was delivered from hospital with sinus rhythm and no signs of superior vena cava syndrome.

Routine outpatient follow up was performed with an annual electrocardiogram, and roentgenogram, and echocardiogram. The EKG showed sinus rhythm with periods of junctional rhythm. Frequent Holter monitoring showed stable sinus rhythm with a mean heart rate ranging from 57 to 72 beats/min over the next 7 years. Chest X-rays showed diminishment of the pulmonary vascular markings and right ventricular area compared with the preoperative period, and a waist inside the stent that did not change during the follow up. Serial echocardiograms showed increasing velocities through the previously stented superior vena cava, beginning with 1.28 m/s soon after the procedure which increased to 1.83 m/s 6 years later.

The patient was submitted for a new catheterization, and the previously implanted stent showed mild neointimal hyperplasia with a residual stricture at the midportion, causing a mild pressure gradient (2 mm Hg) between the superior vena cava and right atrium. This lesion was dilated with a 14 mm Z Med II balloon (NuMed, Inc., Laboratories, Hopkinton, NY) with an excellent angiographic result and evidence of relief of the residual obstruction (Fig. 3). There was a residual pullback gradient of 1 mm Hg. Currently, the patient is asymptomatic and a control echocardiogram showed 1.0 m/s velocity through the pulmonary vein.

DISCUSSION

Partial anomalous pulmonary venous connection (PAPVC) to the superior vena cava with or without an atrial septal defect ASD is a challenge for the surgeon because it was first described in the early 1960s [4,5]. Numerous techniques have been described over the past 40 years in attempts to minimize the complications related to sinoatrial disease and pulmonary and cavoatrial connection [6,7]. Single and double patch baffling approaches have been widely performed with good early- and long-term results, and are the techniques of choice at many centers around the world [2]. However, the high incidence of sick sinus node with these strategies and the difficulties involved in repairing the connection of pulmonary veins to the high-superior vena cava near the innominate vein suggest a medical need for new techniques [6]. The Warden procedure and its variants with caval division, right atrial appendage-superior vena cava anastomosis, and the intra-atrial baffle through the sinus venous defect strongly diminish the incidence of sinoatrial disease with little enhancement of cases of pulmonary and systemic venous obstruction. Reviewing the recent literature, we found few cases of superior vena cava obstruction following PAPVC to superior vena cava repair, which always presents late. Alsoufi, in a series of 175 patients, described one case of late superior vena cava obstruction following the Warden Procedure, but it was mild enough to be clinically treated [8]. In a series of 27 patients, Stewart et al. reported five cases of late superior vena cava obstruction. Among these, only one, presenting 2 years after the surgical procedure, was severe enough to require balloon angioplasty [9]. Iyer et al. described eight patients with a mild residual gradient at

the cavoatrial junction who did not require intervention [2]. Nakahira et al. reported three cases of percutaneous interventions at late follow up for systemic venous obstruction among patients undergoing the Warden procedure with patch augmentation of the superior vena cava [10]. To the best of our knowledge, there is no report in the English literature of symptomatic acute obstruction of the superior vena cava following PAPVC repair. Moreover, there is also no case of immediate postoperative covered stent placement to treat this kind of postsurgical complication.

Our patient developed a life-threatening situation after surgery in the initial hours. The need to deal with the local inflammatory reaction, edema, friable tissue, and suture lines to solve the problem must be taken into consideration, but the only course of care for the infant was to relieve the superior vena cava obstruction. Extra-corporeal circulation and cardio respiratory arrest to perform a new surgical intervention would be dangerous and technically difficult, and stent implantation can be a high-risk procedure because of tissue friability and the possibility of vessel rupture. Therefore, the choice of a covered stent was, in our opinion, the safest option at the time, and proved to be effective in the long-term follow up.

New technologies with low-profile sheaths, catheters, and covered stents to manage pediatric cases in the catheterization laboratory have profoundly increased the number of therapeutic procedures performed by the interventional cardiologist, with no contra-indications concerning moment of intervention or kind of lesion. Throughout the years, the CP-covered stent has proved to be an excellent tool when dealing with high risk vascular angioplasty.

REFERENCES

1. Allen HD, Driscoll DJ, Shaddy RE, Feltes TF, editors. Moss and Adams' Heart Disease in Infants, Children, and Adolescents. Philadelphia: Lippincott Williams & Wilkins; 2008.
2. Iyer AP, Somanrema K, Pathak S, Manjunath PY, Pradhan S, Krishnan S. Comparative study of single- and double-patch techniques for sinus venosus atrial septal defect with partial anomalous pulmonary venous connection. J Thorac Cardiovasc Surg 2007;133:656–659.
3. Shahriari A, Rodefeld MD, Turrentine MW, Brown JW. Caval division technique for sinus venosus atrial septal defect with partial anomalous pulmonary venous connection. Ann Thorac Surg 2006;81:224–229; discussion 229-230.
4. Cooley DA, Ellis PR, Bellizi ME. Atrial septal defects of the sinus venosus type: Surgical considerations. Dis Chest 1961;39: 158–160.
5. Schuster SR, Gross RE, Colodny AH. Surgical management of anomalous right pulmonary venous drainage to the superior vena cava, associated with superior marginal defect of the atrial septum. Surgery 1962;51:805–808.
6. Warden HE, Gustafson RA, Tarnay TJ, Neal WA. An alternative method for repair of partial anomalous pulmonary venous connection to the superior vena cava. Ann Thorac Surg 1984; 38:601–605.
7. DeLeon SY, Freeman JE, Ilbawi MN, Husayni TS, Quinones JA, Ow EP, Bell TJ, Pifarré R. Surgical techniques in partial anomalous pulmonary veins to the superior vena cava. Ann Thorac Surg 1993;55:1222–1226.
8. Alsoufi B, Cai S, Van Arsdell GS, Williams WG, Caldarone CA, Coles JG. Outcomes after surgical treatment of children with partial anomalous pulmonary venous connection. Ann Thorac Surg 2007;84:2020–2026; discussion 2020–2026.
9. Stewart RD, Bailliard F, Kelle AM, Backer CL, Young L, Mavroudis C. Evolving surgical strategy for sinus venosus atrial septal defect: Effect on sinus node function and late venous obstruction. Ann Thorac Surg 2007;84:1651–1655; discussion 1655.
10. Nakahira A, Yagihara T, Kagisaki K, Hagino I, Ishizaka T, Koh M, Uemura H, Kitamura S. Partial anomalous pulmonary venous connection to the superior vena cava. Ann Thorac Surg 2006;82:978–982.