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Research Article

Balloon Angioplasty with Stent Implantation in Superior Vena Cava Stenosis in Pediatric Patients

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ABSTRACT

Introduction and Objective: Superior vena cava (SVC) obstruction can be a complication after congenital heart disease surgery. In the current review we examined our experience and practice in stent implantation after balloon angioplasty in superior vena cava (SVC) obstruction. The objective was to evaluate safety and effectiveness results in patients with SVC stenosis treated by balloon angioplasty with stent implantation.

Methods: Our patient cohort included 11 pediatric patients followed at our institution after surgery for congenital heart disease (CHD) correction, who developed SVC stenosis as a complication of CHD surgery, and 1 patient with severe renal failure and dialysis catheter-related SVC obstruction, all of them requiring endovascular SVC stent placement after balloon angioplasty. Our data acquisition was performed retrospectively and it comprised age, gender, heart diagnosis, cardiovascular surgeries received, age at surgery, weight at surgery, echocardiographic data during follow-up, date of SVC stent placement, weight at intervention, symptoms presented at the time of the endovascular intervention, and clinical follow up data. Final result was evaluated by assessing the re-establishment of normal flow in SVC, the disappearance of collateral venous circulation, and the presence of normal pressure in systemic superior venous system.

Results: In a period comprised between January 2011 and June 2019, 11 patients required SVC endovascular stent placement in our center. Gender distribution was 5 males and 6 females, and median age was 6,5 years (6-11). Average weight at stent placement was 24 kg. Amongst our patients we had 6 sinus venous type atrial septal defect with partial anomalous pulmonary venous return, 2 total anomalous pulmonary venous return, 2 Transposition of great arteries with Mustard correction, and 1 patient with SVC obstruction secondary to chronic dialysis catheter placement. All stents were placed in biventricular circulation hearts. At the time of stent placement, only 2 out of 8 patients had clear SVC syndrome related symptoms. Median time from surgery to SVC stenting was 26 months (10-72 months) with longer times related to Mustard surgeries. Average mean pressure gradient in SVC territory was 8 mmHg (12-3 mmHg) before stenting, and no gradient was found after stenting. Only 1 patient required re-dilation due to neointimal proliferation, more than 7 years after stent implantation, which was a small cell one. At the time of data acquisition, no other patients had required re-dilatation or re-stenting of SVC. Minimum and maximum follow-up time after endovascular intervention was 4 months and 6 years (mean follow up of 18.5 months), respectively. The overall outcome of the intervention was good in all the cases, with re-establishment of normal circulation and pressure balance throughout the superior venous territory. There were no major complications.

Conclusion: SVC obstruction is a non-frequent but important complication of CHD surgery and catheter placement. Balloon angioplasty with stent implantation performed by an experienced team is a safe, effective method to improve patients' situation and reestablish normal venous return.

Keywords: Balloon Angioplasty . Stent Implantation . SVC Stenosis

Introduction

Main causes of superior vena cava (SVC) obstruction in children and adolescents in recent years are more frequently iatrogenic, being those secondary to surgery for congenital heart disease the most common (interatrial septum defects, especially superior venous sinus, cavo-pulmonary anastomosis –bidirectional Glenn-, atrial switch for the repair of d-transposition of the great arteries), pacemaker implantation through the superior vena cava, ventriculoatrial shunts in patients with hydrocephalus, patients who underwent ECMO or ventricular assistance as a bridge to cardiac transplantation, carriers of central venous catheter for either parenteral nutrition, treatment of hematological tumors, or hemofiltration-hemodialysis techniques [1-3].

Treatment options for SVC stenosis include surgical relief or catheter-based interventions, including balloon dilation or endovascular stent implantation. Since first description by Fontaine y Nijiar in 1997 [4], endovascular procedures for superior vena cava stenosis have become the elective treatment due to its low complication rate and good results in short and long term. While both percutaneous techniques have been proved to be safe and effective, some studies comparing the effectiveness have reported better outcomes when combining angioplasty with stent implantation [5,6].

Material and Methods

A retrospective analysis of the diagnoses of superior vena cava stenosis was performed in the center's database since 2010 (transfer to the new Hospital La Fe), patients with a diagnosis of univentricular physiology (Glenn or Fontan) or those who developed stenosis after ECMO procedures, and heart transplantation (including both techniques bi-atrial and bi-cava) were excluded because the program started recently in our institution, leaving a total of 11 patients with a diagnosis of SVC stenosis and who have received percutaneous treatment at some point in their evolution. Data were collected from the patients regarding age, sex, cause and diagnosis and type of surgery, as well as the date of the same, in postoperative cardiac patients, the presence of symptoms prior to percutaneous treatment, echocardiographic data and data of catheterization: date of performance, vascular access used, hemodynamic and angiographic data of the obstruction and type of treatment used (balloon angioplasty or angioplasty with stent implantation, with the type of balloon and / or stent used, if applicable). Sodium heparine was used, and LMWH were used within the first 24 h after the procedure. Double anti-aggregation was given during the first 3 months after the procedure, subsequently maintaining treatment with AAS for six more months.

Results

In a period comprised between January 2011 and June 2020 a total of 11 patients required SVC endovascular stent placement in our center (Table 1). Gender distribution was 5 males and 6 females, and median age was 6,5 years (6-11). Average weight at stent placement was 22 kg. Amongst our patients we had 6 sinus venous type atrial septal defect with partial anomalous pulmonary venous return, 2 total anomalous pulmonary venous return, 2 Transposition of great arteries with Mustard correction, and 1 patient with SVC obstruction secondary to chronic dialysis catheter placement. All stents were placed in biventricular circulation hearts. At the time of stent placement, only 2 out of 8 patients had clear SVC syndrome related symptoms. Median time from surgery to SVC stenting was 26 months (10-72 months) with longer times related to Mustard surgeries. Average mean pressure gradient in SVC territory was 8 mmHg (12-3 mmHg) before stenting, and no gradient was found after stenting. Only 1 patient required re-dilation due to neointimal proliferation, more than 7 years after stent implantation, which was a small cell one (Andrastent®). Most patients were treated with CP® stents (7/11, 63%) (Figure 1). Main vascular access was femoral vein (exception of 2 patients with very severe stenosis in whom a jugular access was needed too, to perform veno-venous loop) (Figure 2). At the time of data acquisition, no other patients had required re-dilatation or re-stenting of SVC. Minimum and maximum follow-up time after endovascular intervention was 4 months and 6 years (mean follow up of 18.5 months), respectively. The overall result of the intervention was good in all the cases, with reestablishment of normal superior venous territory circulation and pressure balance. There were no major complications in this series such as balloon rupture, device embolization, excessive blood loss, vascular rupture, and vascular damage.

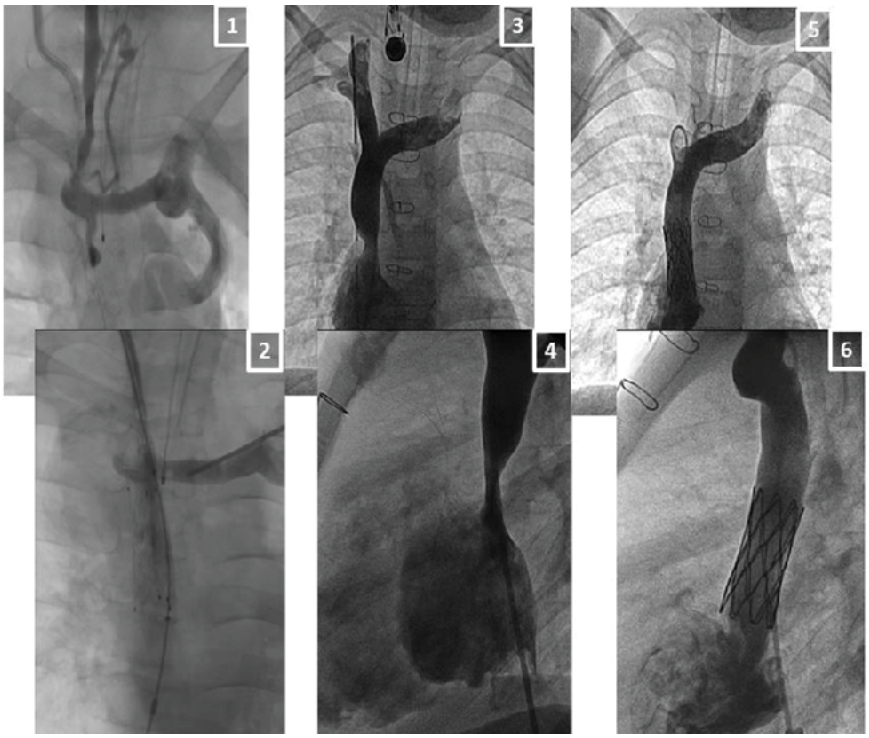

Figure 1: 1 and 2: Patient under dialysis with a RIJV central line and thrombosis of SVC, recanalization through the CVL and after balloon dilation, deployment of an autoexpandable stent with nice result; 3 and 4: AP and lateral view of a severe SVC stenosis in a patient after Warden surgery; 5 and 6: dilation with CP stent with a very nice result.

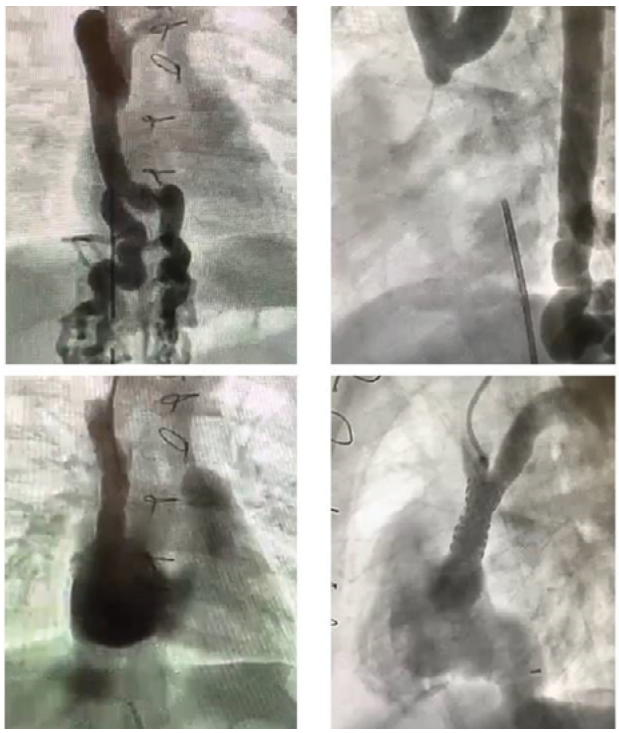

Figure 2: Superior line: Patient after supradiaphragmatic TAPVD surgery, with a critical SVC stenosis and patent collateral veins in AP and lateral angiographic views; Inferior line: same patient after balloon dilation with a Formula® stent implantation with normal antegrade flow and collateral vein disappearance.

	AGE	SEX	DIAGNOSYS		ECHO		CARDIAC CATH							CLINIC
			SURGERY	AGE	Δ P (MMHG)	Æ MÍN. (MM)	AGE	ACCESS	Δ P (MMHG)	Æ MÍN. (MM)	LENGHT (MM)	STENT	BALLOON	
1	15 yo	M	SVS ASD + PAPVD		1,6	5	7,5 yo	RFV	3	6	24	Andra 35	ZMed2 16 × 50	NO
			WARDEN	6 yo	1,2	9	15 yo	RFV	2	10	24	NO	Atlas 16 × 40	NO
2	12 yo	F	d-TGA + SI		2,2	4	8 yo	RFV	8	3,5	20	Andra 26	ZMed2 12 × 40	NO
			MUSTARD	1 mo										
3	8 yo	M	SVS ASD + PAPVD		1,6	5,5	7 yo	RFV	4	6	18	CP 22	BIB 14 × 30	NO
			WARDEN	6 yo										
4	13 yo	F	SUPRA TAPVD		1,8	5	11,5 yo	RFV	6	6	22	CP 28	BIB 16 × 35	NO
			REPAIR	1 mo										
5	10 yo	M	TCRF + DIALISIS		Crit.	Crit.	9 yo	RIJV + RFV	Crit.	2	35	Autoexp.	Ø	SVCS
			CVL	5 yo										
6	9 yo	F	SVS ASD + PAPVD		1,6	6	7 yo	RFV	5	6,5	22	CP 28	BIB 16 × 35	NO
			WARDEN	5 yo										
7	12 yo	M	d-TGA + IS		1,9	5,3	10 yo	RFV	6	4,5	14	CP 22	BIB 14 × 30	NO
			MUSTARD	1,5 mo										
8	8 yo	F	SVS ASD + PAPVD		1,4	5	7 yo	RFV	4	6	20	CP 28	BIB 14 × 35	NO
			WARDEN	5 yo										
9	7 yo	F	SVS ASD + PAPVD		1,5	5	6,5 yo	RFV	4	6	20	CP 28	BIB 14 × 35	NO
			WARDEN	5 yo										
10	8 yo	F	SVS ASD + PAPVD		1,4	5,5	7 yo	RFV	4	6,5	28	CP 34	BIB 16 × 50	SVCS
			WARDEN	6 yo										
11	9 mo	F	SUPRA TAPVD		--	--	9 m	RIJV + RFV	12	1	14	Formula 8×16	8 × 16	SVCS
			REPAIR	3 mo										

Note: Yo: Years Old; Mo: Months Old; M: Male; F: Female; SVS ASD: Superior Sinus Venous Atrial Septal Defect; PAPVD: Partial anomalous pulmonary vein drainage; TAPVD: Total anomalous pulmonary vein drainage; d-TGA: d-transposition of great arteries; TCRF: Terminal cronic renal failure; CVL: Central venous line; ΔP: Mean Pressure gradient; ⌀ MÍN: Minor diameter; RFV: Right femoral vein; RIJV: Right internal jugular vein; Crit.: Critical stenosis; Autoexp.: Autoexpandable stent; SVCS: Superior vena cava syndrome

Table 1: Patients with severe SVC stenosis and percutaneous treatment with stent (only biventricular hearts) in our centre between January 2011 and June 2020.

Discussion and Conclusion

Iatrogenic obstruction of the SVC after corrective surgery for congenital heart disease is a rare but serious complication. Particularly in infants, the surgeon should avoid causing stenosis of the SVC by the cannulation sutures or by additional trauma to the vascular wall, using the smallest size of cannula possible that allows adequate systemic venous flow during cardio-pulmonary bypass. Following cardiac surgery for congenital heart disease, the SVC can become obstructed secondary to thrombosis or direct manipulation. Cannulation of the SVC is a risk factor for causing an endothelial injury that can become a nest for thrombosis. The turbulence created by flow through a stenotic vessel represents a significant risk factor for the formation of a thrombus and this poses an even greater risk in a vessel with laminar (non-pulsatile) flow such as SVC. Sharoni et al. [8] in a retrospective review of 1,853 cardiac surgical procedures, reported a 0.5% rate of iatrogenic SVC stenosis (all of them in neonates), mainly due to thrombosis at the cannulation site. Jayakumar et al. [9] contributed the causes of SVC obstruction at sites of surgical anastomosis to several factors including mismatch in vessel size, intimal hyperplasia, constriction by sutures, thrombosis, and strictures formation.

Presence of symptoms are mainly related to 2 factors: the rate of progression of the SVC stenosis and its extension and the flow ratio through the obstructed segment and the retrograde flow through the azygous system. Symptomatic SVC obstruction may cause swelling and cyanosis of head and upper limbs, headache, cerebral venous hypertension, syncope, cough, and airway obstruction [8]. Occasionally it can result in protein loosing enteropathy, pleural, and pericardial effusion from the retrograde congestion on the thoracic duct. Chylothorax can occur by direct trauma to the lymphatics, which frequently occurs during thoracic procedures or as a consequence of

high venous pressures that impair lymph flow due to "functional" outflow tract obstruction [10].

Therapeutic strategies in the presence of a SVC syndrome depend fundamentally on the causes and may include prompt thrombolysis when it's detected early, long-term anticoagulation, isolated angioplasty or more frequently combined with stent implantation or surgery (bypass, reconstructive surgery of the SVC or the implantation of a spiral vein graft) [3,11]. While the option of thrombolysis and anticoagulation in a postoperative seems somewhat risky, the performance of a new surgery to reconstruct the SVC does not seem a good option either. Balloon angioplasty combined in almost all cases with stent implantation has become the most widely used therapeutic option nowadays. Angioplasty alone does not reduce the degree of obstruction in the majority of cases, due in large part to the composition of the vascular wall of the SVC, which is why it seems useful to perform it initially as a carving and to assess the response, face to face. to the implantation in a second stage of a vascular stent [12].

The use of stents within the SVC has been reported with other etiologies (generally external compressions due to malignant lesions), but its use in postsurgical stenosis in the pediatric population is more recent [13]. Stents within the systemic veins are rapidly covered by neointimal, acquiring excellent long-term patency in some reports. Percutaneous stent treatment of SVC stenosis is associated with few complications. Tzifa et al. 6 reviewed their 22-year experience with stent in 63 patients. Acute complications after stent placement occurred in 12 (19%) patients and included an SVC tear (9.5%); right atrial perforations, 3%; and malposition or embolization of the stent, 6.5%. During the 22-year late follow-up, 28% of patients developed restenosis of the SVC caused mainly by exaggerated in-stent neointimal proliferation. Smayra et al. [14] in a series of 30 patients with SVC syndrome who underwent 49 stent implants reported only 7% complications with an acceptable patency rate of 46.7% (from 87.5% to 11.1%). Aldoss et al. [7], in their study concluded that SVC stent implantation is more effective than SVC balloon dilation in treating SVC stenosis with significantly better 6-month freedom from re-intervention.

In our experience, percutaneous angioplasty with stent implantation over an obstruction at the level of the SVC proved to be a viable option for the treatment of this patient with SVC syndrome after cardiac surgery. However, some technical points need to be mentioned. Main access is feasible trough the femoral vein, but in very severe stenosis maybe is needed a second access through the internal jugular vein to advance catheters more safely, and to have a good angiography control. In patients with repaired superior venous ASD, we followed the whole procedure with transesophageal echocardiography (TEE), to make sure we were not compressing any pulmonary vein close to the SVC. A rapidly performed venogram can help identify the exact location of the stenotic lesion and guide the selection of an appropriately sized stent. In our case, the stent was selected in order to allow a slight oversizing. The choice of a stent of a longer length may carry the risk of occlusion of some of the veins tributary to the SVC, such as the azygos or the subclavian. In our series we found great results with bared CP stent®, because it is the one who developed less re-stenosis or neo-intimal proliferation [15,16], probably due to its bigger cells; also it's a good stent for children up to 18-20 kgs, because it can be re-dilated to bigger diameters. For small children, best available options to dilate a severe stenosis nowadays are vascular stents such as Formula® or Valeo® Stents.

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