Primary Stenting in a Young Adult with Aortic Coarctation

VICTOR SALGADO, MD*†; JOSE MARTINEZ-TORO, MD*†; EDWIN RODRIGUEZ, MD†

ABSTRACT: A young male adult with significant aortic coarctation was initially referred to our clinics due to uncontrolled blood pressures. On evaluation the diagnosis of aortic coarctation was evident and confirmed with Magnetic Resonance Angiography (MRA). After discussing treatment options with the patient and his parents they opted for the least invasive procedure possible. Primary stenting of a simple discrete aortic coarctation was performed successfully. The following is a report on the methods used and immediate results.

Key words: Aortic coarctation. Percutaneous angioplasty. Stent. Adult congenital heart disease.

Aortic coarctation (CoA) is a congenital narrowing of the aorta, usually close to the ligamentum arteriosus or patent ductus arteriosus (PDA) that causes obstruction to blood flow to the lower extremities and abdominal organs with marked difference in blood pressures from the upper extremities to the lower extremities (1). It can be accompanied by other cardiac defects as bicuspid aortic valve, ventricular septal defects or William's Syndrome and is associated with early mortality and increased risk of hypertension in adult life even after adequate repair (2-7). The later the repair the higher the risk of developing chronic hypertension (8).

Surgical correction is plagued with residual problems such as recoarctation, aneurysm formation, paraplegia and need for reoperation (9-14). In the last twenty years non-surgical catheter directed balloon angioplasty has emerged as an alternative option for these patients. Initially balloon management seemed promising but long term follow-up uncovered a high incidence of several complications like aortic dissection (15-17), cerebrovascular accidents (18-19), aneurysm formation (19-29) and more importantly incomplete relief of the CoA gradient (17-18, 21-22), which at follow up could be significant (>20 mmHg) in as much as 7-36% of patients (18, 20, 21, 23).

More recently, stents have been used to treat recoarctation after surgery, angioplasty and less frequently, as primary stenting for CoA with very promising results. With stenting there is no need to over dilate the coarcted segment, the stent can be dilated to the desired diameter and dilated further with a larger balloon immediately or in a second procedure, if necessary (24). Stents prevent the acute re-modeling of aortic wall (25), and have been found in follow up of up to 3 years to have a much lower incidence of complications as recoarctation, aneurysm formation or dissection (24, 26-28). The newer larger stents may allow primary stenting to be performed in the pediatric population since they can be re-dilated to the size of the adult aorta as the patient grows (29). They also allow for re-dilation in a restenosed segment without causing strut fatigue (26). The unexpanded size of these newer stents is smaller which allow the use of smaller introducer sheaths and decrease the risks of complications related to vascular access as hematomas or pseudoaneurysms compared with older stents.

Due to the lack of randomized clinical trials, the role of primary stenting for CoA remains controversial. We report our initial results performing primary stenting in a young adult male with simple, discrete aortic coarctation.

Case History

An 18 year old male was referred to our clinics by his primary physician due to uncontrolled blood pressures despite treatment with enalapril 10 mg bid and metoprolol 50 mg bid for the last six months. The patient denied any cardiac symptoms or history of cardiovascular disease but complained of easy leg fatigability and cramps when performing exercise. On physical examination he was 69" inches tall and weighted 85 kg. His blood pressure was 160/90 mmHg on his right arm and 140/80 mmHg on his left arm. We were unable to measure blood pressures at the ankles. Pulses on his upper extremities were brisk and

*Section of Cardiology, Department of Medicine, University of Puerto Rico School of Medicine, San Juan, PR.
†Centro Cardiovascular de Puerto Rico y el Caribe, San Juan, PR.

Address for correspondence: Victor M. Salgado, Section of Cardiology, Department of Medicine, UPR School of Medicine, P.O. Box 365067, San Juan, PR 00936-5067

strong but barely palpable in his lower extremities. There was a harsh grade II systolic ejection murmur all over the precordium. EKG was suggestive of left ventricular hypertrophy (LVH). 2-D Echo examination was done and the diagnosis of LVH confirmed, the aortic valve could not be adequately visualized to rule out a bicuspid aortic valve but there was no significant gradient. The aortic arch was not adequately visualized. A magnetic resonance angiography (MRA) was ordered which showed aortic coarctation distal to the left subclavian artery with an estimated diameter of 5 mm. The patient was scheduled for an initial catheterization and angiogram to make appropriate measurements of aortic gradients and diameters prior to a definite procedure. Angiogram showed a mean gradient of 38 mmHg (60 mmHg peak gradient across the coarctation. The coarctation diameter was 4 mm and the diameter of the proximal aorta at the level of the origin of the left subclavian artery was 14 mm. There was no gradient across the aortic valve. Treatment options were discussed with the patient and his family, and after acquiring consents the patient was scheduled for stent dilatation of aortic coarctation.

Methods

The procedure was performed by an experienced interventional cardiologist with the assistance of a pediatric interventional cardiologist. An anesthesiologist was present to provide conscious sedation.

Vascular access was obtained from the right femoral artery with an 8 French 10 cm introducer sheath and 5,000 units of heparin were administered intravenously immediately after insertion of the introducer. A 0.35 cm x 260 cm Amplatz guide wire (Cordis Corporation, Miami, Florida, USA) was inserted across the lesion to the proximal aorta. A 6 French pigtail catheter was advanced over the wire across the stenosis and the guide wire removed. Anteroposteriol (AP) and leftanterior oblique (LAO) angiographic views were done to confirm previously measured diameters. The guide wire was re-inserted and the pigtail catheter removed. The 23 mm x 21mm Genesis stent (Cordis Europa N.V., Roden, The Nederlands) was mounted on a 14 mm x 21mm TYSHAK II balloon catheter (Nu Med, Inc., Hopkinton, NY, USA). A Fast Cath open ended introducer catheter (St. Jude Medical, Minnetonka, MN, USA) was advanced across the lesion and the stent and balloon advanced inside the introducer catheter up to the site of the coarctation. While holding the balloon and stent in place, the introducer catheter was removed just distal to the coarctation, exposing the stent to the stenosis site.

Using a controlled pressure device the balloon was inflated at 6 atm for 30 seconds (expected balloon diameter 14 mm) and the stent deployed. Figure 1 illustrates deployment of the stent in the coaretation of the aorta. The balloon was deflated and removed and the pigtail catheter was inserted for angiographic views. These post dilatation views showed persistence of a small waist on the stent. The pigtail catheter was removed and a larger TYSHAK II 16 mm x 4 cm balloon (Nu Med, Inc., Hopkinton, NY, USA) was advanced to the stent and inflated at 4 atm for 30 seconds (expected balloon diameter 14 mm). AP and LAO angiographic views were repeated and showed adequate stent deployment and disappearance of the stenotic waist.

The immediate post dilatation gradient was 0 mmHg and a diameter of 13 mm was achieved. The patient developed paradoxical hypertension with blood pressures 181/61 (119) mmHg. Propanolol 1mg IV was administered. Final blood pressure was 142/90 (70) mmHg. Patient was

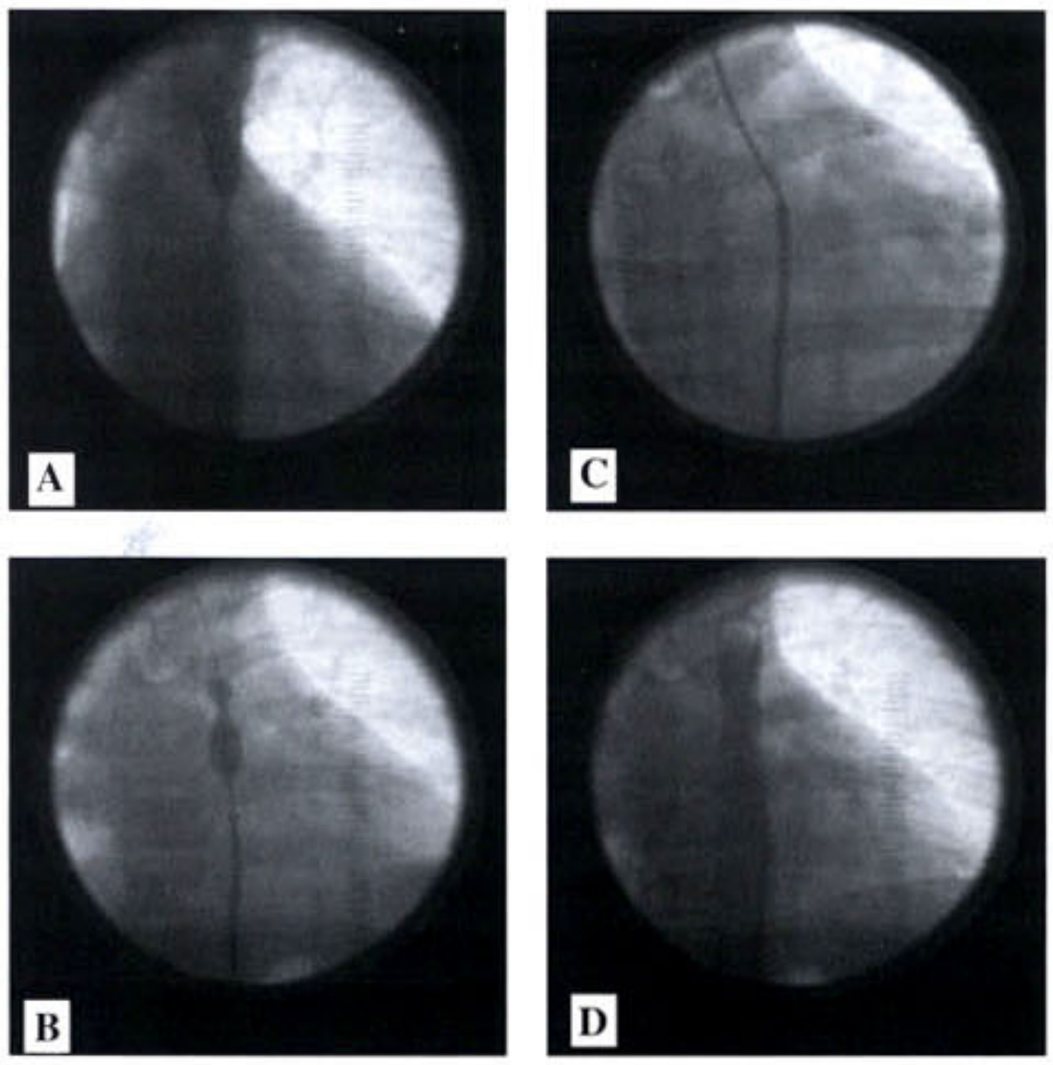

Figure 1. (A) Predilatation aortogram. (B) Balloon inflation. (C) Deployed stent. (D) Post dilatation aortogram.

discharged home the next day with the recommendation to continue his treatment for hypertension for at least six months.

Unfortunately, the patient has failed to show for follow up visits despite our attempts to contact him.

Discussion

Aortic coarctation should be suspected in any young individual with new onset or refractory hypertension. The

diagnosis can be easily made by physical examination and confirmed with very sensitive non invasive tests as 2-D Echo, Trans-esophageal echocardiography (TEE), magnetic resonance (MR) or computer tomography (CT) angiography (30). Once the diagnosis is made treatment should be sought as early as possible due to the increased cardiovascular mortality associated with chronic hypertension (1-2).

The type of coarctation (discrete, tubular or isthmic hypoplasia) and the presence of associated complex cardiovascular disease (ventricular septal defects, bicuspid aortic valve) should be taken into consideration when evaluating treatment options.

Most studies done up to date have shown that stenting of all types of aortic coarctation is safe and effective with immediate gradients below 10 mmHg, with few complications and a low rate of stenosis in mid-long term follow up (24-29, 31). Patients with discrete aortic coarctation had higher rates of success and lower rates of complications and re-stenosis than patients with tubular coarctation or isthmic hypoplasia (31).

To the best of our knowledge, this is only the second case in Puerto Rico in which percutaneous stenting has been used as primary treatment for coarctation of the aorta in an adult patient. Both were performed at the Cardovascular Center of Puerto Rico and the Caribbean. The methods and techniques used in both cases were very similar to the ones reported on previous studies and publications. The first case was performed in 2002 by Dr. Pedro Colon and Dr. Edwin Rodriguez; the patient was a 45 year old male with diabetes and end-stage-renal-disease on hemodialysis who arrived at the emergency room with a hypertensive crisis and was diagnoses with 2-D ECHO with coarctation of the aorta. His initial gradient was 50 mmHg. After stenting the gradient was reduced to 0 mmHg and his systolic blood pressure dropped from 180 mmHg to 130 mmHg. Our results were similar to the ones reported in previous studies with the immediate post dilatation gradient dropping to 0 mmHg. However our patient developed paradoxical hypertension after the procedure. The occurrence of this complication was very rare in previously reported series but has been well documented. The mechanism by which this occurs is not understood but a higher incidence was found in patients who suffered prolonged hypertension (25-26, 31). Based on these previous findings it is not altogether surprising that our patient developed this complication, which resolved quickly with administration of 1mg of propanolol. It is generally recommended that patients who have suffered prolonged hypertension should continue medical treatment for several months and doses tapered down during follow up visits.

It is important to remember that patients with CoA are never cured of their disease and should be followed closely for development of complications as hypertension, recoarctation or formation of aneurysms. Patients should have blood pressure measurements on arms and legs in each visit. A difference of more that 20 mmHg in systolic blood pressure should prompt radiological evaluation to rule out recoarctation. Although exercise testing was previously advocated as an essential part of follow up to diagnose exercise induced hypertension, the validity of this recommendation has recently come into question with various studies finding no difference in the prevalence of exercise induced hypertension between patients with corrected coarctation and the general population (32). Echocardiograms are usually done yearly with Doppler evaluation of the aortic arch. In the case of our patient a TEE should be considered during follow up to effectively rule out a bicuspid aortic valve which accompanies CoA in up to 30% of cases. CT and MR angiography are similarly useful for the noninvasive evaluation in patients with coarctation to rule out aneurysm formation and are usually done six months after the procedure and then every 2-3 years if no complications were found (33). Some authors prefer CT since stents may cause artifacts during MRI (24).

We believe the results obtained in our case add to the increasing evidence that stenting of aortic coarctation is safe and effective, and should be considered as the initial therapeutic option in adult patients with native coarctation of the aorta, especially when dealing with simple, discrete lesions.

Resumen

Se presenta un paciente varón de 19 años de edad. Fue referido por su médico primario debido a hipertensión sin control a pesar de tratamiento con enalapril y metoprolol. Con angiografía de resonancia magnética se diagnosticó coartación de la aorta distal a la arteria subclavia izquierda con un diámetro de 5 mm y un gradiente de 38 mmHg. El paciente recibió dilatación con malla como primera opción de tratamiento. Desarrolló un episodio de hipertensión paradójica, el cual fue controlado con propanolol intravenoso. Constituye el segundo caso donde se hizo este procedimiento en un adulto en el Centro Cardiovascular de Puerto Rico y el Caribe.

References

1 Webb G, Smallhorn F, Therrien J, Redington A. Chapter 56: Congenital Heart Disease, *Braunwald's Heart Disease 7th Edition,* Philadelphia, Elsevier 2005;p1532-1535.

2 O'Sullivan J, Derrick G, Darnell R. Prevalence of hypertension in children after early repair of coarctation of the aorta: a cohort study using casual and 24 hour blood pressure measurement. *Heart*; Aug 2002;88:163-166.
3 Cohen M, Fuster V, Steele PM, et al. Coarctation of the aorta: long term follow up and prediction of outcome after surgical correction. *Am Heart J*; 1989;80:840-5.
4 Maron BJ, Humphries JO, Rowe RD, et al. Prognosis of surgically corrected coarctation of the aorta: a 20 year postoperative appraisal. *Circulation*; 1973;47:119-26.
5 Prasad M, Moodle D, Biechman G, et al. Long term follow-up of aortic coarctation in infants, children and adults. *Cardiol Young* 1993;3:20-6.
6 Presbitero P, Demarie D, Villani M, et al. Long term results (15-30 years) of surgical repair of aortic coarctation. *Br Heart J*; 1987;57:462-7.
7 Koller M, Rothlin M, Senning A. Coarctation of the aorta, review of 362 operated patients. Long term follow-up and assessment of prognostic variables. *Eur Heart J* 1987;8:670-9.
8 Bergadahl L, Bjork VO, Jonasson R. Surgical correction of coarctation of aorta: influence of age on late results. *J Thorac Cardiovasc Surg*; 1983;85:532-6.
9 Bobby JJ, Emami JM, Farmer RDT, et al. Operative survival and 40 year follow up of surgical repair of aortic coarctation. *Br Heart J*; 1991;65:271-6
10 Kappetein PA, Guit GL, Bogers, JJC, et al. Noninvasive long-term follow-up after coarctation repair. *Br Heart J* 1993;55:1153-9.
11 Malan JE, Benatar A, Levin SE. Long-term follow-up of coarctation of the aorta repaired by patch angioplasty. l *Int J Cardiol* 1991;30:23-32.
12 Stewart AB, Ahmed R, Travill CM, et al. Coarctation of the aorta life and health 20-40 years after surgical repair. *Br Heart J* 1993;69:65-70.
13 Kimball TR, Reynolds JM, Mays WA, et al. Persistent hyperdynamic cardiovascular state at rest and during exercise in children after successful repair of coarctation of the aorta. *J Am Coll Cardiol* 1994;24:194-200.
14 Zehr KJ, Gilivov AM, Redmond JM, et al. Repair of coarctation of the aorta in neonates and infants: a thirty-year experience. *Ann Thorac Surg* 1995;59:33-41.
15 Kulick DI, Kotlewski A, Hurvitz RJ, et al. Aortic rupture following percutaneous catheter balloon coarctoplasty in an adult. *Am Heart J* 1990;119:190-3.
16 Rico J, Sobrino N, Calvo L, et al. Angioplastia transluminar percutánea en la coartación aórtica del adulto joven. *Rev Esp Cardiol* 1989;42:415-17.
17 Biswas PK, Mitra K, De S, et al. Follow up results of balloon angioplasty for native coarctation of aorta. *Indian Heart J* 1996;48:673-6.
18 Fletcher SE, Nihill MR, Grifka RG, et al. Balloon angioplasty of native coarctation of the aorta: midterm follow-up and prognostic factors. *J Am Coll Cardiol* 1995;25:730-4.
19 Schrader R, Bussmann WD, Jacobi V, et al. Long-term effects of balloon coarctation angioplasty on arterial blood pressure in adolescent and adult patients. *Cathet Cardiovasc Diagn* 1995;36:220-5.
20 Fawzy ME, Dunn B, Galal O, et al. Balloon coarctation angioplasty in adolescents and adults: early and intermediate results. *Am Heart J* 1992;124:167-71.
21 Beekman RH, Rocchini AP, Dick M, et al. Percutaneous balloon angioplasty for native coarctation of the aorta. *J Am Coll Cardiol* 1987;10:1078-84.
22 Morrow WR, Vick GW, Nihill MR, et al. Balloon dilation of unoperated coarctation of the aorta: short and intermediate-term results. *J Am Coll Cardiol* 1988;11:133-8.
23 Syamasundar P, Thapar MK, Kutayli F, et al. Causes of recoarctation after balloon angioplasty of unoperated aortic coarctation. *J Am Coll Cardiol 1989;13:109-15.*
24 Chessa M, Carrozza M, Butera G, et al. Results and mid-long-term follow-up of stent implantation for native and recurrent coarctation of the aorta. *Euro Heart J* 2005;26:2728-32.
25 Mullins CE, O'Laughlin MP, Vick GW, Palmaz JC, et al. Implantation of balloon-expandable intravascular grafts by catheterization in pulmonary arteries and systemic veins. *Circulation* 1988;77:188-99.
26 Harrison DA, McLaughlin PR, Lazzam C, et al. Endovascular stents in the management of carctation of the aorta in the adolescent and the adult: one year follow up. *Heart* 2001;85:561-566.
27 Thanopoulos BD, Hadjinikolaou L, Konstadopoulou GN, et al. Stent treatment for coarctation of the aorta: intermediate term follow up and technical considerations. *Heart* 2000;84:65-70.
28 Del Cerro MJ, Fernández-Ruiz A, Benito F, et al. Balloon Angioplasty for Native Coarctation in Children: Immediate and Medium-Term Results. *Rev Esp Cardiol.* 2005;58(9):1054-61.
29 Duke C, Rosenthal E, Qureshi SA. The efficacy and safety of stent redilatation in congenital heart disease. *Heart* 2003;89:905-912.
30 Stern HC, Locher D, Wallnofer K, et al. Noninvasive assessment of coarctation of the aorta: comparative measurements by two-dimensional echocardiography, magnetic resonance, and angiography. *Pediatr Cardiol* 1991;12:1-5.
31 Zabal C, Attie F, Rosas M, et al. The adult patient with native coarctation of the aorta: balloon angioplasty or primary stenting?
32 Swan L, Goyal S, Hsia C, Hechter S, et al. Exercise systolic blood pressures are of questionable value in the assessment of the adult with a previous coarctation repair. *Heart* 2003;89:189-192.
33 Hager A, Kaemmerer H, Leppert A, et al. Follow-up of Adults With Coarctation of the Aorta: Comparison of Helical CT an MRI, and Impact on Assessing Diameter Changes. *Chest* 2004;126:1169-76.