

Safety and Efficacy of Endovascular Treatment of Middle Aortic Syndrome with Self-expanding Stent: A Single-center Experience

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ABSTRACT

Objective: To determine the immediate procedural outcome of self-expanding stent implantation in patients with the middle aortic syndrome.

Study design: Retrospective observational study.

Materials and methods: Five patients, aged 15 to 17 years (mean 26.2 years), with upper limb hypertension due to middle aortic syndrome and receiving endovascular treatment with self-expanding stent in the Department of Cardiology at a major tertiary center in Western India were analyzed.

Intervention: Self-expanding stents were implanted in the mid/lower thoracic/upper abdominal aorta.

Main outcome measure: Satisfactory deployment of stents with 50% or greater reduction in pressure gradient and incidence of periprocedural complications.

Results: In all patients, angiography showed long-segment stenosis in the mid or lower thoracic/upper abdominal aorta. The pressure gradient was between 51 and 140 mm Hg (mean 92.4 mm Hg). Five self-expanding stents were implanted. Immediately after implantation, the gradient decreased to between 16 and 55 mm Hg (mean 37.2 mm Hg). Angiography showed a satisfactory result with widely patent stents in all. There were no major periprocedural complications.

Conclusion: Endovascular self-expandable stent implantation is a safe and feasible modality for the treatment of patients with the middle aortic syndrome and merits further evaluation in larger number of patients with greater follow-up.

Keywords: Endovascular management, Middle aortic syndrome, Self-expanding stent, Takayasu's disease.

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INTRODUCTION

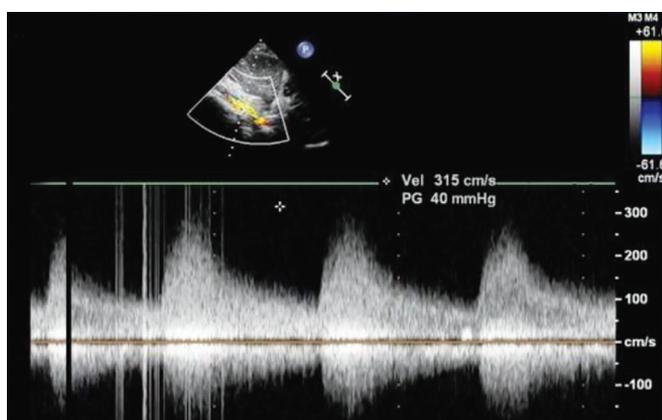
The middle aortic syndrome is an uncommon disease that may result from a congenital anomaly in the development of the aorta or from acquired causes, most commonly nonspecific aortoarteritis (Takayasu's disease). Surgical intervention is the gold standard for treating obstructive aortic lesions, yet postoperative morbidity and mortality have remained quite high. On the contrary, endoluminal stenting of aortic stenosis is less invasive compared with surgical options and more efficacious compared with balloon angioplasty alone, resulting in fewer procedural complications and lower mortality.

MATERIALS AND METHODS

This study is a retrospective observational study of all patients presenting with hemodynamically significant stenosis of the lower descending thoracic and/or supra-renal abdominal aorta treated with endovascular stenting over a 30-month period from October 2014 to May 2017 (Table 1). Patients were diagnosed clinically as isolated upper limb hypertension with echocardiographic evidence of narrowing and Doppler evidence of turbulence with pressure gradients at the "middle aorta" (Fig. 1). Serum C-reactive protein and erythrocyte sedimentation rate were normal in all patients at the time of procedure. Three of the four patients diagnosed as Takayasu's disease had received antituberculous therapy and two were on maintenance dose of corticosteroids. All patients underwent computed tomography aortogram, except the pregnant one, who underwent the magnetic resonance aortogram, followed by the invasive aortogram, to profile the aortic narrowing and involvement of other branches. The procedure was performed under local anesthesia and mild sedation in all patients, on the Siemens Artis Z cardiac catheterization system. The right common femoral artery was accessed with a 6F introducer sheath in all the patients. Unfractionated heparin 70 to 100 U/kg was administered. Additional radiation safety measures including an abdominal shield were used for the pregnant patient. A 6F Judkins Right (JR) catheter (Cordis, Johnson and Johnson Medical N V, Milpitas, CA) with a manually made side hole was used to cross the stenosis,

Table 1: Baseline patient characteristics

	<i>Patient 1</i>	<i>Patient 2</i>	<i>Patient 3</i>	<i>Patient 4</i>	<i>Patient 5</i>
Age (years)	25	24	42	15	25
Sex	Female	Female	Male	Female	Female
Site of stenosis in the aorta	Abdominal (suprarenal)	Abdominal (suprarenal)	Lower descending thoracic and upper abdominal	Abdominal (suprarenal)	Abdominal (suprarenal)
Hypertension	Yes	Yes	Yes	Yes	Yes
Heart failure	No	No	Yes	Yes	No
Etiology	Takayasu's disease	Takayasu's disease	Atherosclerotic vascular disease	Takayasu's disease	Takayasu's disease

**Fig. 1:** Color Doppler echocardiography demonstrating narrowing and turbulence in the aorta at the diaphragm with gradients and diastolic tailing on pulsed wave doppler interrogation

with the help of an angled tip guidewire (TERUMO corp, Japan) and the pressure gradient measured. Over a 260-cm 0.035" angled tip exchange length guidewire (Cordis, Johnson and Johnson Medical N V, Miami lakes, FL), the JR was then exchanged with a 5 or 6 F marker pigtail (Merit medical systems, Inc. UT), which was positioned just proximal to the narrowed segment. Angiographic study through the pigtail catheter revealed the stenotic segment in the thoracoabdominal aorta. The length from the proximal relatively unaffected segment to the distal such segment and the average diameter of the healthy segments were taken to decide on the stent sizing, keeping in mind their tendency to elongate after deployment. The premounted self-expanding stent

systems from Boston Scientific (Wallstent or Epic) were used to deploy the stent. Single stent was sufficient in all patients. Angiographic and hemodynamic results were determined at immediate postprocedure (Table 2). Procedural success was defined as fall in the transaortic gradient by 50% or more. Patients were shifted to the recovery unit. The femoral sheath was removed with manual compression when the activated clotting time was below 200 seconds. All patients were discharged by day 3 postprocedure. Acetylsalicylic acid (aspirin) at a dose of 75 mg was administered to all the patients for 6 weeks.

RESULTS

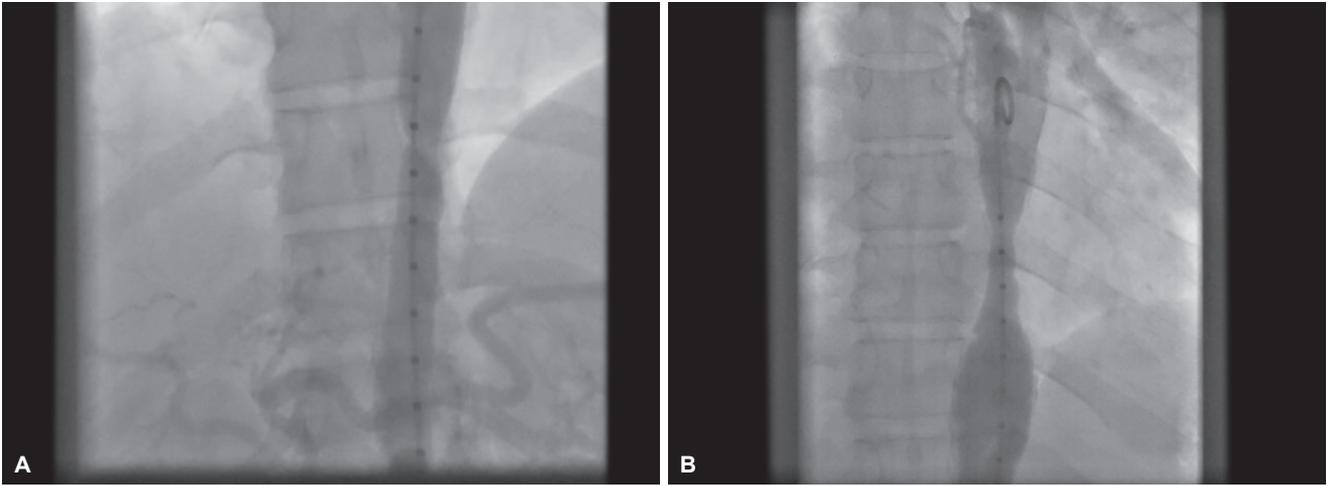
Data from five patients of middle aortic syndrome, who underwent endovascular treatment, were analyzed. There were four females (80%) and one male. The mean age was 26.2 years (15–42 years). Uncontrolled upper limb hypertension was the most common presentation. Frank heart failure symptoms were present in two patients.

One patient was pregnant, in the 24th gestational week, and presented with prior bad obstetric history, retrospectively attributed to uncontrolled hypertension secondary to middle aortic syndrome.

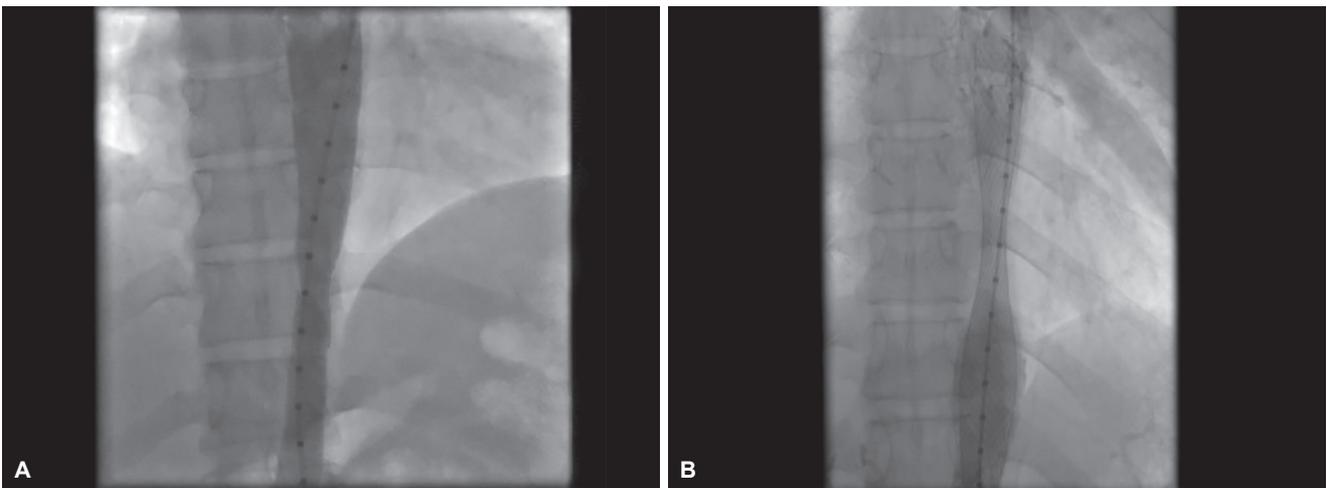
The clinical and hemodynamic data are presented in Tables 1 and 2. The pressure gradient across the stenosis was between 51 and 140 mm Hg (mean 92.4 mm Hg). Angiography confirmed long-segment stenosis in the thoracoabdominal aorta (Fig. 2). The minimum diameter of the stenosis was between 3.8 and 6.8 mm (mean 5 mm)

Table 2: Procedural characteristics and outcomes

	<i>Patient 1</i>	<i>Patient 2</i>	<i>Patient 3</i>	<i>Patient 4</i>	<i>Patient 5</i>
Minimum diameter of stenosis (mm)	5.5	4.3	6.3	3.8	5.0
Length of stenosis (mm)	38.5	146.5	103.0	72.4	48.0
Pressure gradient across stenosis (mm Hg)	78	108	85	51	140
Stent (diameter in mm × length in mm)	Boston Scientific Epic stent 12 × 60	Boston Scientific Wallstent 14 × 90	Boston Scientific Wallstent 24 × 70	Boston Scientific Wallstent 14 × 90	Boston Scientific Epic stent 14 × 70
Sheath compatibility (F)	6	10	12	10	6
Postdilatation	Not done	10 × 60 mm TYSHAK balloon	12 × 40 mm TYSHAK	Not done	Not done
Poststenting residual pressure gradient (mm Hg)	37	45	33	16	55



Figs 2A and B: Angiographic demonstration of narrowed aortic segments in: (A) Patient with Takayasu's disease; and (B) atherosclerotic vascular disease



Figs 3A and B: Angiogram after stent deployment in above patients

and the length of the stenosis ranged between 38 and 147 mm (mean 81.7 mm).

Self-expanding stent systems were used in all patients. Cobalt–chromium–nickel alloy (Elgiloy) Wallstent (Boston Scientific, Inc.) was used in three patients, while the nitinol-based epic stent was used in two patients (Boston Scientific, Inc.; mean stent length 76 mm). Immediately after stent implantation, the residual gradient across the stented segment ranged from 16 to 55 mm Hg (mean 37.2 mmHg). Angiography showed a satisfactory result with patent and correctly positioned stents in all patients (Fig. 3). Postdilatation with noncompliant TYSHAK balloon was performed in two patients due to partial stent recoil (Fig. 4).

No major immediate periprocedural complication occurred. The average fluoroscopy time was 7.3 minutes. Prior to discharge, blood pressure was well controlled with the previous treatment in all the patients. Antihypertensive treatment could be fully withdrawn in one patient after stent implantation, while it was



Fig. 4: Postdilatation of self-expanding stent

significantly decreased in the others. The femoral arterial pulses were well palpable in all patients. The pregnant patient underwent uneventful cesarean section (for obstetric indications) at-term and delivered a healthy child.

DISCUSSION

The middle aortic syndrome is an uncommon disease that may result from a congenital anomaly in the development of the aorta or from acquired causes.^{1,2} The acquired causes consist of retroperitoneal fibrosis, mucopolysaccharidosis, neurofibromatosis, fibromuscular dysplasia, and more commonly nonspecific aortoarteritis (Takayasu's disease), an inflammatory disease of unknown etiology that affects the aorta and its branches. In other words, an autoimmune process causes an inflammatory process that destroys the elastic media of the aorta and its main branches. The syndrome was originally described from our institute in the seminal article by Sen et al.³ In their own words, which hold true over half a century later, "The typical patient is a young woman below the age of 25 years who complains of symptoms of hypertensive disease or, less frequently, of lower limb claudication or abdominal angina. Most of them superficially resemble cases of coarctation of aorta and are diagnosed as such, while the remainder are diagnosed as suffering from essential hypertension. The unusual nature of the block in the aorta is indicated by absent or weak pulse in one or both superior extremities, and a bruit heard over the abdomen or in the interscapular region lower down than is usually heard in congenital coarctation. The pathology of this block seems to be some sort of non-specific allergic panarteritis of a segmental nature involving the middle part of the aorta—more often supradiaphragmatic but not infrequently infradiaphragmatic, and occasionally both (Fig. 5).³ Involvement of the renal arteries is an additional factor responsible for hypertension in some cases. An active tuberculous lesion elsewhere in the body is found in most cases, and this may possibly be an allergen. Clinical evidence of

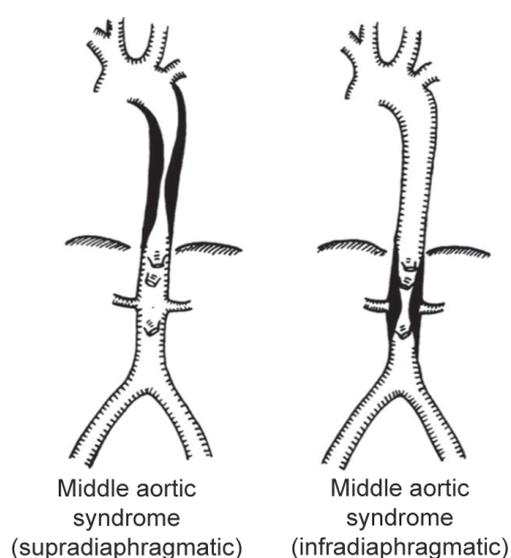


Fig. 5: Diagrammatic representation of middle aortic syndrome

such a focus strengthens the diagnosis of middle aortic syndrome."

Surgical intervention is the gold standard for treating obstructive aortic lesions, yet postoperative morbidity and mortality have remained quite high.⁴ In Taketani's study,⁵ 33 patients with abdominal aortic coarctation resulting from Takayasu arteritis over a 44-year period underwent surgical treatment (aorto-aortic bypass). Of 29 initial survivors (four hospital deaths due to bleeding, pneumonia, and renal failure), 22 patients had complications during follow-up, including anastomotic aneurysm (10 patients), heart failure (3 patients), cerebrovascular accident (3 patients), abdominal aortic aneurysm (2 patients), graft deterioration (2 patients), anastomotic stenosis, and renal failure (2 patients).

On the contrary, endoluminal stenting of aortic stenosis is less invasive compared with surgical options and more efficacious as compared with balloon angioplasty alone, resulting in fewer procedural complications and lower mortality.^{2,6} Nevertheless, open surgical repair seems to be the first choice in the treatment of abdominal aortic coarctation when associated with aneurysmal degeneration, unfavorable anatomy (such as severe occlusion of the aorta or extension to the aortic bifurcation), and severe hypertension.⁷ Some authors believe that endovascular treatment of the abdominal aortic stenosis is not effective or safe for lesions located near the renal arteries and may not have long-term benefits.

Our study, to the best of our knowledge, represents the largest cohort of adult patients with the middle aortic syndrome to undergo endovascular treatment with self-expanding stents. Upper limb hypertension and related symptoms, such as headache and dyspnea were common presentations. Four of the five patients, all females, had a clinical diagnosis of Takayasu's disease as the etiology. Of these, one of the patients, reported elsewhere,⁸ was diagnosed for the first time in her late second trimester of pregnancy and had previous bad obstetric history and uncontrolled hypertension. Endovascular stenting was followed by uneventful delivery of a healthy baby at-term. The third patient, and the only male in our series, presented with resuscitated sudden cardiac death, co-existing hypertensive heart disease, and diabetes mellitus. He demonstrated severe generalized calcification of the aorta, and the coronary lesions on angiography deemed to be of atherosclerotic etiology. An automated implantable cardioverter-defibrillator device was implanted a month after endovascular therapy.

The decision to use self-expanding stents was made on the grounds that such lesions are densely fibrotic and deployment of balloon-expandable stents would pose greater risk of perforation, edge dissection, and subsequent aneurysm formation. Although their radial strength is significantly lower than balloon-mounted

stents, the hemodynamic criterion of success—of a 50% or greater reduction in the peak systolic pressure gradient—could be achieved in all patients with post-dilatation performed only in selected cases. Excessive postdilatation should be avoided. Fall in gradients is the only hemodynamic end point.

CONCLUSION

Endovascular treatment with self-expanding stent systems is a safe and effective therapeutic modality for the middle aortic syndrome with good immediate results. No major procedural complications were observed. Larger studies with longer follow-up are required for universal validation of this modality.

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