



## Hybrid Treatment of Mega Aortic Syndrome With Device Closure of Pseudoaneurysm

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### ABSTRACT

A young patient with mega aortic syndrome, aortic and mitral incompetence and a pseudoaneurysm of the abdominal aorta at the left renal artery origin was admitted with dyspnea. We describe a complete treatment of all her problems in two stages along with a device closure of the pseudoaneurysm. The patient recovered well and had no complications. We advocate this method of treatment of such extensive aortic disease with much lower morbidity and mortality than traditional surgery.

### KEYWORDS

Aneurysm, Hybrid, Aorta, Device

### CASE REPORT:

A 24-year-old female patient with complaints of dyspnea on exertion for 5 years (New York Heart Association class III) was diagnosed to have severe aortic and mitral incompetence with a left ventricular ejection fraction of 35%. Her blood investigations were normal. CT Aortogram showed multiple fusiform aneurysms of the ascending aorta, arch and descending aorta with a large sacular pseudoaneurysm of the abdominal aorta at the level of the left renal artery origin. Genetic testing done revealed that she was a case of Marfan syndrome.

She underwent Bentall-de Bono procedure, classical elephant trunk technique with reimplantation of the arch vessels and mitral valve repair. Cardiopulmonary bypass was established with right femoral arterial and bicaval venous cannulation and the patient was cooled to 18 degrees celcius. The circulation was arrested at 18 degrees celcius and selective antegrade cerebral protection was initiated through the left innominate and left carotid arteries. The elephant trunk anastomosis to the descending thoracic aorta was done with a 22 mm Datascope graft. The patch containing the arch vessels was anastomosed to the elephant trunk graft, completing the elephant trunk procedure. Following this, Bentall's procedure was done with a 21 mm St. Jude composite tube graft and mitral valve was addressed with a 30 mm Physio II ring. The duration of

circulatory arrest was 22 minutes, aortic cross clamp time was 95 minutes and cardiopulmonary bypass time was 220 minutes. Postoperatively her neurological status and renal function were normal. Biopsy report of the diseased aortic wall showed cystic medial necrosis suggestive of Marfan's syndrome.

Endovascular repair of the remaining diseased aorta was carried out in the same admission through the left femoral artery. Self-expanding stent grafts were deployed (Zenith Thoracic graft 30mm x 200mm) in the thoracic aorta and the aortic pseudoaneurysm was excluded using an Amplatzer device. (Figure 1) There was no focal neurological deficit observed following the procedure.

The patient recovered well with no complications. Her renal function tests along with all the other postoperative investigations were normal. Postoperative computed tomography scan was normal. (Figure 2) Total ICU stay was 5 days and hospital stay was 12 days. One year follow up revealed that the patient was doing well with good exercise tolerance.

**Figure 1 :**  
**PREOPERATIVE CT AORTOGRAM SHOWING ANEURYSMAL DILATATION OF THE ASCENDING AORTA, ARCH AND THORACIC DESCENDING AORTA. THE PSEUDOANEURYSM AT**

**THE LEVEL OF LEFT RENAL ARTERY IS WELL APPRECIATED**



**FIGURE 2: POSTOPERATIVE IMAGES SHOWING REPLACEMENT OF THE ENTIRE THORACIC AORTA AND AMPLATZER DEVICE OCCLUDING PSEUDOANEURYSM OF THE ABDOMINAL AORTA**



**DISCUSSION:**

Mega aortic syndrome is defined as diffuse aneurysmal involvement of the ascending aorta, arch and the thoracoabdominal aorta. Aggressive replacement of an extensive portion of the aorta, if not the entire aorta, is considered to be the ideal treatment for several subsets of patients such as mega aortic syndrome, Marfan syndrome and chronic aortic dissection. Diffuse aneurysmal disease and the tendency for the disease to rapidly progress are two important factors, which necessitates extensive operations in these patients. (1) The 5-year mortality of mega aortic syndrome is 100%, if left untreated. The most common etiologies of the mega aortic syndrome are cystic medial necrosis and myxomatous degeneration of the aortic wall. (2)

Extensive disease of the aorta was traditionally treated exclusively with surgery. However, endovascular treatment has emerged as a promising option with lesser complications and is here to stay. The evolution of treatment for extensive thoracic aneurysms appears to have moved away from the extensive initial single-stage to two-stage procedures (elephant trunk procedure). In the first stage an “elephant trunk” graft is placed that hangs in the descending aorta. During the second-stage, an endovascular approach is then used to place a stent graft that connects the elephant trunk to the descending aorta. The elephant trunk graft provides a good landing zone for the stent-graft, and endovascular completion is a useful alternative to conventional second-stage surgery. This method has very few complications, and a postoperative paraplegia rate of 1.1 %. (1,2,3)

Pseudoaneurysm of the aorta is a potentially fatal condition. Symptoms are noticed usually only after an asymptomatic period of several years. However, the incidence of rupture of a pseudoaneurysm is significant with noteworthy mortality rates. Hence, treatment is recommended irrespective of the presence of symptoms. (4)

In our patient, the pseudoaneurysm had an opening of 10mm and was opposite the left renal artery. A hybrid thoracoabdominal procedure was required surgical debranching of the visceral vessels adding to the morbidity. Complete endovascular replacement of the thoracoabdominal aorta is technically demanding. Considering the anatomy and technical feasibility, the septal occlude device was thought to be a simpler and viable option.

Our patient suffered from mega aortic syndrome with aneurysmal involvement of the entire aorta with a pseudoaneurysm at the base of the left renal artery. We used the Amplatzer septal occluder to successfully close off the pseudoaneurysm.

This device has proven to be versatile in reference to the numerous case reports on its use in other intracardiac defects, paravalvar leaks and occlusion of the left atrial appendage. (4,5,6)

There have been a few case reports which have described this technique of occluding pseudoaneurysms, however none of them involve treatment of mega aortic syndrome using the elephant trunk technique as well. We believe that this technique of occluding a pseudoaneurysm and endovascular stent grafting is the first of its kind. (4,5,6)

In conclusion, hybrid aortic repair is emerging as an easier and safer option for the treatment of extensive aortic diseases. Pseudoaneurysm of the aorta, can simply be occluded by the Amplatzer device, in addition to the elephant trunk technique and endovascular stent grafting used to treat mega aortic syndrome.

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