**CASE REPORT**

**Single stage treatment of complex aortic pathology (mega-aortic syndrome) using frozen elephant trunk technique**

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**SUMMARY**

This is a case report of single-stage total thoracic aortic repair by the frozen elephant trunk technique for a 75-year-old female with a complex aortic pathology of mega-aorta extending from the ascending aorta to the descending aorta. We used a Thoraflex™ Hybrid device, a frozen elephant trunk device with four branched grafts and the distal stent graft. The distal stent graft was inserted into the downstream descending aorta via an aortic arch and positioned 15 cm beyond the left subclavian artery after total arch replacement had been performed using a four-branch graft. The postoperative course was unremarkable with no complications. A post procedural computed tomography scan demonstrated complete exclusion of the descending thoracic aeurysm without endoleak. Therefore, fixing the whole mega-aorta in a single stage using the frozen elephant trunk was effective and safe.

**INTRODUCTION**

Treating a complex aortic aneurysm extending from the ascending aorta to the descending aorta (mega-aorta) is controversial and poses technical challenge. It is an area of ongoing development and innovation. We report here the surgical treatment of a case of mega-aorta in one-stage, using the frozen elephant trunk (FET) procedure.

**CASE REPORT**

A 75-year-old lady, with the history of hypertension, hyperlipidemia and hyperthyroidism was admitted for surgery on a mega aorta extending from the ascending aorta to the descending thoracic aorta. She presented with one-year history of recurrent chest and inter-scapular pain. The blood investigations were within normal limits and coronary angiogram did not reveal any significant disease. Transthoracic echocardiography showed preserved left ventricular function and normal valves.

The preoperative computed tomography (CT)-scan (Figure 1A) identified an atherosclerotic degenerative aneurysm of the ascending aorta (5.0cm), the proximal arch (5.6cm), the distal aortic arch and proximal descending aorta of 6.6 cm. The diameter of the aorta was 5.0cm at the level of the 5th thoracic vertebrae, 4.0cm at the level of the 8th thoracic vertebra, 3.0cm at the level of the 10th, and 2.5cm at the level of the 12th. We selected one-stage operation using the frozen elephant trunk technique for this patient with mega-aorta. CT was used to calculate the appropriate hybrid stent-graft size.

After median sternotomy, cardiopulmonary bypass was established (195 minutes) via the distal ascending aortic cannulation, single two-stage venous cannula in the right atrium and left ventricular venting. Under severe hypothermia of \(22^\circ\)C of nasopharyngeal temperature, circulatory arrest (65 minutes) introduced and selective antegrade cerebral perfusion initiated (total of 75 minutes). A Thoraflex Hybrid Plexus stent-graft (32/38/150mm) (Figure 2A) was deployed into the descending thoracic aorta after the aortic arch was laid open and all the neck vessels were transected from the arch. Total arch replacement was performed using a four branch graft of 32mm in diameter by suturing the collar onto the distal arch. All the neck vessels were anastomosed separately using the superior three branches of the graft. The plication to the sinotubular junction was performed using a 32mm graft. The completed surgery is illustrated in a diagram (Figure 2B). We verified the position of the stent-graft by transesophageal echocardiography.

The patient recovered well and there was no stroke, paraplegia or other complications. A postoperative CT-scan demonstrated complete exclusion of the descending thoracic aneurysm without endoleak (Figure 1B).

**DISCUSSION**

The treatment of mega-aortic syndrome (MAS), defined as an aneurysmal dilatation involving the ascending aorta, the arch and the descending aorta still remains challenging.\(^1\) An open surgical approach is invasive and complex for this sort of lesion. Treating in one stage is associated with significant mortality, whereas if performed in two different stages, the mortality during the waiting period between the two procedures and the rate of patients refusing the second stage are considerable issues.\(^1\) Since the introduction of endovascular repair for thoracic aortic aneurysms, it has been considered as an alternative treatment modality that may be associated with lower mortality and morbidity.\(^2\)\(^4\) Hybrid procedure involving endovascular stenting of the total aortic arch with bypass of the supra-aortic vessels has recently become a popular and less invasive technique for arch aneurysm.\(^7\) However, this technique is not suitable for a mega-aorta that does not have an appropriate proximal
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Fig. 1a: Preoperative computed tomography scan. The maximum diameters of the thoracic aortic aneurysm were 3.3 cm at the sinus of Valsalva, 5.0 cm for the ascending aorta, 5.6 cm for the proximal aortic arch, and 6.6 cm for distal arch and proximal descending thoracic aorta. The diameter was 5.0 cm at the level of the 6th thoracic vertebra, 4.0 cm at the 8th, 3.0 cm at the 10th, and 2.5 cm at the 12th.

Fig. 1b: Postoperative computed tomography demonstrated complete exclusion of the descending thoracic aneurysm without endoleak.

Fig. 2a: The picture of the Thoraflex Hybrid Plexus Stent-Graft

Fig. 2b: The diagram of the completed surgery.
landing zone and stent sizes on the ascending aorta. The frozen elephant trunk technique is performed via a median sternotomy, and an endovascular stent-graft is placed into the descending aorta in an antegrade fashion through the opened aortic arch. The ascending aorta and aortic arch are then replaced conventionally.

This technique has a combination of advantages of surgical and interventional approaches, and therefore, appears to promising. The frozen elephant trunk technique has fewer anatomical limitations in terms of the proximal and distal aorta compared with an endovascular stent-graft.

The current indications for the FET procedure are MAS, chronic dissection after acute Type A aortic dissection repair or Type B dissection associated with ascending or arch aneurysms. The application of this technique in acute dissections is still controversial.

Good results of the FET procedures can be obtained if good strategies of myocardial, cerebral and visceral protection adopted. One of the distinctive complication of this technique is spinal cord injury (SCI). The incidence of paraplegia after implantation of the frozen elephant trunk is 3.3% according to a review report by Karck and Kamiya. SCI after the frozen elephant trunk procedure was associated with a range of factors, such as occlusion of vital intercostal arteries along the thoracic vertebral level, where the distal end of the frozen elephant trunk was deployed, the history of abdominal aortic operation and perioperative hypotension. Cerebrospinal fluid drainage was performed before the operation in elective cases with a history of aortic repair to the thoraco-abdominal or abdominal aorta or when the pathological condition required stent-graft delivery lower than the ninth thoracic vertebra or length of coverage of the descending aorta more than 15 cm.

Our patient presented with MAS and was contemplated with different strategies. The two-stage operation would carry a risk of aortic rupture occurring between the procedures and a hybrid technique was not suitable as there was no appropriate landing zone and stent size. Therefore, we performed one-stage total thoracic aortic repair of the mega-aorta using the frozen elephant trunk technique. This was indeed the first successful operation of this nature done in our institution and Malaysia.

In conclusion, the FET technique represents a new surgical paradigm for the effective treatment as a single-stage operation for mega-aorta.

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**CONFLICTS OF INTEREST**
None declared

**REFERENCES**