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# Kinking of frozen elephant trunk: reality versus myth—a case report and literature reported

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**Background:** Kinking or iatrogenic stenosis in the frozen elephant trunk (FET) is a possible complication, however, there have been few cases reported.

**Case Description:** A 43-year-old male with acute type A aortic dissection (ATAAD) underwent total arch replacement and FET installation. After weaning from the cardiopulmonary bypass, both femoral pulses were absent. A right axillo-bifemoral bypass using 8 mm graft was performed. Postoperative aortography showed a 100 mmHg-stenosis at the FET and 28 mm stent-graft was inserted to relieve stenosis. Eleven cases of postoperative FET stenosis have been reported from 2007 to 2019. The ages ranged from 30 to 72 years and 6 patients had ATAAD, 4 had chronic type A dissection, and 1 had non-dissection. They all underwent total arch replacement. To correct the iatrogenic stenosis of the FET, additional TEVAR was done in 8 patients, 2 had axillo-femoral bypass, 1 had a bare stent graft, and one required re-anastomosis.

**Conclusions:** To prevent the FET kinking, surgeons should place the stented portion of the FET in the aortic arch angle. Also, we should make the non-stented portion as short as possible at the distal anastomosis. The DANE (distal anastomosis new entry) should be avoided by the secure anastomosis. Over-sizing or under-sizing of the FET should be minimized.

**Keywords:** Total arch replacement; frozen elephant trunk; acute aortic dissection; distal arch aneurysm; kinking of the frozen elephant trunk

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#### Introduction

The Japanese made frozen elephant trunk (Frozenix®, Japan Lifeline, Tokyo, *Figure 1*) was first launched in the Japanese market in 2014. The case number of the Frozenix in 2014 was only 600 in Japan, however, the number has increased dramatically to 3,600 in 2020. Over 17,000 products have been used by the end of 2020. In the of 2014 or in 2015, the Frozenix was used for non-dissection aneurysm (NDTAA) in 50% and for acute type A aortic dissection (ATAAD) in 33% of patients. In 2020, this ratio was reversed to 51% in ATAAD and 41% in non-dissection TAA. As for ATAAD surgery in Japan, 55% of patients had replacement of ascending aorta or hemiarch, 30% had conventional total

arch replacement (TAR), and 15% had TAR plus FET among 5,250 patients in 2016. In 2020, 7,000 patients had surgery for ATAAD and number of patients who had replacement of the ascending aorta or hemiarch decreased to 40% and that of TAR plus FET increased to 28%. Dominant sizes of the Frozenix used in ATAAD have been 23 to 29 mm in diameter and 9 or 12 cm in length (1).

Kinking or iatrogenic stenosis in the frozen elephant trunk (FET) is a possible complication and have been sporadically reported. In this report, our experience of this complication is presented, and possible mechanisms are discussed. The author presents the following case in accordance with CARE reporting checklist (available at https://cdt.amegroups.com/article/view/10.21037/cdt-22-28/rc).

546 Okita. Kinking of the FET

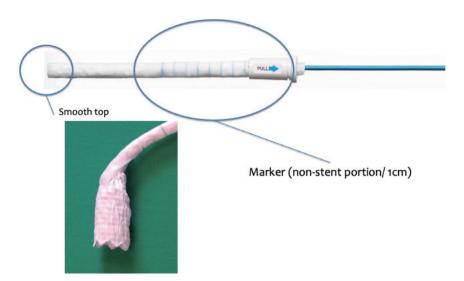


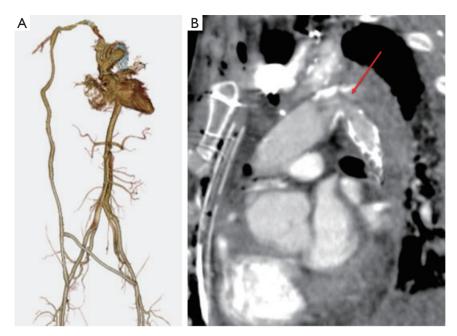
Figure 1 Frozenix.

#### **Case presentation**

The patient was a 43-year-old male who was diagnosed to have an acute type A aortic dissection (ATAAD) in a local hospital and transferred to us 7 hours after onset of chest pain. His consciousness was clear and all arterial pulses were palpable. There was a small pericardial effusion and aortic regurgitation was minimum by echocardiography. The CT scan showed that he had DeBakey type I dissection and that the false lumen was not thrombosed. There was a big entry tear at the level of the left subclavian artery and the true lumen was narrowed at the aortic bifurcation.

The cardiopulmonary bypass was started using the femoral cannulation and he was cooled to 23 °C. The patient was positioned to the Trendelenburg position, and the ascending aorta and aortic arch was incised after circulatory arrest and the heart was arrest using the retrograde cardioplegia. The antegrade cerebral perfusion was started using three cannulas inserted to the arch vessels from inside the arch. The aorta was transected distal to the intimal tear at the level of the left subclavian artery and a frozen elephant trunk (FET; Frozenix<sup>®</sup> 23 mm × 6 cm, Japan Lifeline, Tokyo) was inserted in the true lumen of the descending aorta. The FET was fixed with an outer Teflon felt strips with three 5-0 monofilament mattress sutures. A four branched Dacron graft (J graft® 26 mm, Japan Lifeline, Tokyo) was anastomosed using a continuous 4-0 monofilament suture to the proximal stump of the descending aorta including the FET. Then, distal perfusion was resumed and the patient was rewarmed. Proximal anastomosis was done at the level

of the ST junction of the ascending aorta. Three aortic valve commissures were resuspended and an inner strip of the Dacron graft and an outer Teflon felt were used to reinforce the anastomosis. Then, coronary perfusion was re-started, and the heart was defibrillated. Three arch vessels were anastomosis using the button technique to the graft branches. Duration of the CPB was 237 minutes, cardiac ischemic time 117 minutes, duration of the circulatory arrest of the lower body 62 minutes, duration of the ACP 181 minutes. The minimum temperature of the tympanic membrane and rectum was 22.1 and 28.2 °C, respectively. The patient weaned the bypass without any difficulty, however, the femoral pulses were not palpable and there was no urine production. A right axillary artery to bilateral femoral extraanatomical bypass was made using a 8 mm ringed Gore-Tex graft. On the 3<sup>rd</sup> postoperative day, an aortography and CT scan showed a severe stenosis at the non-stented portion of the FET (Figure 2). The pressure gradient at the stenotic portion was 100 mmHg and there was a leakage to the false lumen. An endovascular stent-graft (TX-D® 28 mm. Zenith Cook) was used to relieve the stenosis (Figure 3). He was discharged and back to the normal life. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.



**Figure 2** (A) postoperative CT scan of the present case. Total arch replacement with additional right axillo-bifemoral artery bypass. (B) kinking FET (arrow). FET, frozen elephant trunk.

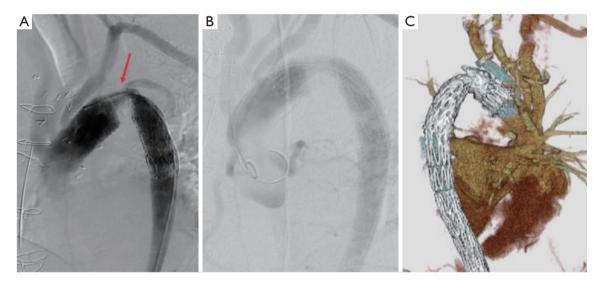


Figure 3 (A) postoperative angiography, kinking FET (arrow) with distal anastomotic leakage, (B) a stent-graft was inserted, (C) postoperative CT scan. FET, frozen elephant trunk.

#### **Discussion**

In 1983, Borst *et al.* introduced the "free" elephant trunk principle into the surgical strategy for an extensive thoracic aortic disease (2). In 1996, Kato *et al.* (3) first reported 10 patients who had undergone home-made stent-graft insertion in the descending aorta, and they

named this method "open stent grafting". In 2003, Karck et al. (4) reported 4 patients who had an open aortic arch replacement with stent-graft insertion on the descending aorta. This was the first report where this method was called "frozen elephant trunk (FET)". Since then, the frozen elephant trunk procedure has been widely spread mainly in

548 Okita. Kinking of the FET

Europe and many clinical investigations have been reported using commercial-made products (5-10).

Japan-made FET, Frozenix, was launched 2014 and since then, more than 17,000 prostheses have been implanted. The Nitinol stent was hand-knitted not to change length as the diameter change. All stents were fully covered with the Dacron prosthesis. The proximal portion of the Frozenix stent graft was composed of a simple Dacron graft. The Dacron graft had external velour with a thickness of 300 micro mm and its water porosity was 150 mL/cm<sup>2</sup>. The delivery system consisted with a malleable rod (10 Fr) and could be advanced into the descending aorta over a 0.035-inch flexible guide wire the stent-graft. The system was wrapped by a smooth-surfaced polyester mesh. The sheath had markers of 1 cm-interval in the non-stented portion and the last marker should be pointed at the edge of distal aorta. Because the stent did not change its length throughout, the distal end of the stent graft can be fixed as expected. Withdrawal of the outer sheath while the inner rod was held steady released the stented portion of the Frozenix stent graft. The proximal Dacron tube could then be released by simply pulling back both the sheaths and rod. The products had a range of diameter from 21 to 39 mm (2 mm step) and the length of the stented portion was 60 mm, 90 mm, 120 mm, and 150 mm. The total length of the stent graft was 200 mm in all. The length of the total system was 57 cm and 12 mm in diameter (Figure 1).

Main advantages of usage of the FET in patients with ATAAD is to facilitate distal anastomosis in zone II or in zone III. In addition, bleeding from the distal anastomosis is seldom seen, and there is less incidence of leakage to the distal false lumen occurs, resulting in positive remodeling of the distal aorta. Some of the patients who had FET at the first stage had the subsequent endovascular stent-graft insertion in the descending aorta. We conducted a multiinstitutional study dealing with FET usage from 2016 to 2018, and there were 154 patients with ATAAD underwent total arch replacement plus FET insertion. Mean ages were 61.1 years and hospital death occurred in 3 patients (1.9%), permanent stroke in 12 (7.8%), and paraplegia in 4 (2.6%) (11,12). The annual survey of Japanese Association of Thoracic Surgery found a steep increase of usage of the FET in surgical strategy for the ATAAD. In 2016, patients with ATAAD who had total arch replacement plus FET consisted of 15% in whole 5,250 patients who had surgery for the ATAAD and that became 28% of 7,000 patients with ATAAD in 2020 (1). Subsequently, ratio of the total arch replacement in total ATAAD patients in Japan increased

from 49% in 2016 to 60% in 2020.

Several complications regarding with the FET usage have been recognized. The main concern is a relatively higher incidence of development of spinal cord ischemia. The incidence was not negligible and has remained constant. Deliberate deployment of the distal stent-graft in the descending aorta above the level of the aortic valve should be warranted to prevent spinal cord complications (11). Also, exclusion of the patients with shaggy descending aorta is recommended. Distal stent-induced new entry (DSINE) has been reported to occur in 5% to 7% in FET usage and DSINE has occurred more frequently in routine TEVAR (thoracic endovascular aortic repair) procedures in patients with type B aortic dissection (13). Placing the FET in the vertical position in the true lumen of descending aorta and avoidance of over-sizing the FET relative to the diameter of the descending aorta was effective.

Kinking of the FET or stenosis have been reported on several occasions, resulting in the low perfusion status of the lower body and proximal hypertension. The reports have been published in series of case reports and there was no collective reviews focusing on this complication (14-22) (Table 1). Majority of the patients who had the FET stenosis showed that the junction of the stented portion and non-stented portion of the FET was placed at the steep angle of the aortic arch. Taguchi et al. (20) warned a risk of kinking of the FET when it was used in patients with right aortic arch because that there is steep angle in the distal arch in this setting. Most reports warned that the length of the non-stented portion was too long. Especially the FET was used in non-dissection aneurysm cases who had zone II distal anastomosis where a large space was remained around the FET. Sometimes, they had anastomotic leakage (distal anastomotic new entry; DANE) to the false lumen of the descending aorta (Figure 3), which may cause a pressurization of false lumen of the descending aorta. In most cases, this stenosis was corrected by an additional stent-graft insertion from the femoral artery. Deschka et al. (22) used a bare stent. Alternatively, Uchida et al. (15) re-anastomosed the proximal portion of the FET and distal portion. Taguchi et al., and we performed an extra-anatomical bypass to the lower body.

Surgeons should be aware of the arch topology, such as the size, direction of the blood stream, and morphology preoperatively and should try to place the stented portion of the FET in the aortic arch angle. Also, we should make the non-stented portion as short as possible at the distal anastomosis to prevent FET kinking. The DANE may cause the pressurization of the distal false lumen and should

Table 1 Reported cases of kinking or stenosis of the frozen elephant trunk

Authors	City	Year	Case	Lesions	Procedure	Distal anastomosis	DANE	FET	Secondary procedure	Outcomes
Easo	Oldenburg	2007	53 male	CTAAD s/p Hemiarch	TAR			Chavan- Haverich	TEVAR	Alive
Pacini	Bologna	2008	52 male	CTAAD Marfan, s/p MVR, Scoliosis	TAR, Bentall	Zone III		Evita	TEVAR	Alive
Nakao	Tokyo	2013	49 male	CTBAD, Kommerell div	TAR	Zone II		Gore-Tag	TEVAR	Alive
Deschka	Munster	2014	51 male	ATAAD	TAR, Freestyle root	Zone III		Evita	Palmaz stent	Alive
Present case	Kobe	2015	43 male	ATAAD	TAR	Zone III	Yes	Frozenix	Ax-FA bypass, TEVAR	Alive
Motomatsu	Fukuoka	2016	72 male	TAA, distal arch	TAR	Zone II		Frozenix	TEVAR	Alive
Wakiyama	Kobe	2017	76 male	ATAAD retrograde	TAR	Zone II		Frozenix	TEVAR	Alive
Taguchi	Hirosaki	2018	67 male	ATAAD, RAA, Kommerell div	TAR	Zone III		Frozenix	Ax-FA bypass	Alive
Morizaki	Osaka	2019	44 male	ATAAD	TAR, VSRR	Zone II		Frozenix	TEVAR	Alive
			30 male	ATAAD, AAE, Marfan	TAR, VSRR	Zone II		Frozenix	TEVAR	Alive
Uchida	Yamagata	2019	66 male	CTAAD, TAA distal arch	TAR	Zone II		Frozenix	Re-anastomosis	Alive

s/p, status post-operation; DANE, distal anastomosis new entry; FET, frozen elephant trunk; ATAAD, acute type A aortic dissection; CTAAD, chronic type B aortic dissection; TAA, non-dissection thoracic aorta aneurysm; MVR, mitral valve replacement; RAA, right aortic arch; div, diverticulum; AAE, annuloaortic ectasia; TAR, total arch replacement; VSRR, valve sparing aortic root replacement; TEVAR, thoracic endovascular aneurysm repair; Ax-FA, axillo-femoral.

be avoided by the secure and meticulous techniques. Oversizing or under-sizing of the FET should be minimized because the FET may cause DSINE and migrated distally (23). Alternatively, Yamamoto et al recommended "zone 0 arch repair" to facilitate distal anastomosis in patients with ATAAD (24).

#### **Conclusions**

To prevent the FET kinking, surgeons should place the stented portion of the FET in the aortic arch angle. Also, we should make the non-stented portion as short as possible at the distal anastomosis. The DANE (distal anastomosis new entry) should be avoided by the secure anastomosis. Oversizing or under-sizing of the FET should be minimized.

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#### **Footnote**

Provenance and Peer Review: This article was commissioned by the Guest Editors (Mohamad Bashir, Mohammed Idhrees and Edward P. Chen) for the series "Frozen Elephant Trunk" published in Cardiovascular Diagnosis and Therapy. The article has undergone external peer review.

Reporting Checklist: The author has completed the CARE reporting checklist. Available at https://cdt.amegroups.com/article/view/10.21037/cdt-22-28/rc

550 Okita. Kinking of the FET

Conflicts of Interest: The author has completed the ICMJE uniform disclosure form (available at https://cdt.amegroups.com/article/view/10.21037/cdt-22-28//coif). The series "Frozen Elephant Trunk" was commissioned by the editorial office without any funding or sponsorship. The author has no other conflicts of interest to declare.

Ethical Statement: The author is accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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