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CASE REPORT

Hybrid Repair of a Thoraco-abdominal Aortic Aneurysm Associated with Loeys-Dietz Syndrome

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Introduction: Loeys-Dietz syndrome (LDS) is a genetic syndrome caused by mutations in transforming growth factor beta receptors (TGFBR) 1 and 2. It can manifest with craniofacial, musculoskeletal, cognitive abnormalities, and vascular pathologies including early onset aortic root aneurysms, extensive aortic dissections, and TAAA. Open repair is considered the gold standard treatment but carries morbidity risks, especially in patients with multiple previous aortic procedures. Endovascular treatment is associated with treatment failure when used in the native aorta, because of inherent wall weakness precluding seal. This case report adds to the available literature on hybrid treatment of LDS associated aortic pathologies.

Report: This is the report of staged hybrid TAAA treatment in a 24 year old male patient with multiple previous aortic procedures via sternotomy and thoracotomy. Retrograde infrarenal aortic visceral debranching was performed using 14 mm by 7 mm bifurcated Dacron grafts. These emerged from the limbs of an 18 mm by 9 mm bifurcated Dacron graft in an aortobi-iliac reconstruction. This was followed by staged thoracic endovascular aortic repair (TEVAR) seven days later using three endografts (26 mm–22 mm × 150 mm distal, 30 mm × 200 mm bridging, then 32 mm × 100 mm proximal). The endograft landed in an old thoracic aortic graft proximally and the new infrarenal aortic graft distally. Follow up at 11 months showed patency and no sac expansion.

Conclusion: Hybrid TAAA repair was a valid treatment option in this patient with LDS and multiple previous aortic procedures. It minimised the morbidity of revision surgery and mitigated potential treatment failure by achieving an endovascular seal in surgical grafts. Short term surveillance showed no complications. Limitations to making recommendations include lack of long term follow up.

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INTRODUCTION

Loeys-Dietz syndrome (LDS) is a genetic syndrome caused by mutations in transforming growth factor beta receptors (TGFBR) 1 and 2. It can manifest with craniofacial, musculoskeletal, cognitive abnormalities, and vascular pathologies.[1] These vascular pathologies include early onset aortic root aneurysms, extensive aortic dissections, and TAAA.[2,3] LDS patients present at a younger age than patients with Marfan syndrome, and aortic replacement is often necessary in childhood.[4]

REPORT

This is the case of a 24 year old male with LDS who was referred to an aortic clinic. He had a history of mitral valve replacement, valve sparing aortic root, and ascending aorta replacement. He also had two left thoracotomies for open descending thoracic aorta replacements at another institution, and multiple thoracolumbar spinal fusions. He had a residual chronic dissection of the entire native thoracoabdominal aortic segment, which progressed in diameter from 38 mm to 45 mm over six months (Fig. 1).

The patient was taken for an infrarenal aortobi-iliac bypass with total visceral debranching through a transperitoneal approach. An 18 mm by 9 mm bifurcated Dacron graft (Getinge, Gothenburg, Sweden) was used with presewn bifurcated 14 mm by 7 mm Dacron grafts (Getinge, Gothenburg, Sweden) to each graft limb. The main body of the graft was left long (Fig. 2) to accommodate the distal landing zone for a stent graft. The aorta and iliac arteries were dissected and then anastomosed in an end to end fashion and reinforced with felt pledgets. Internal iliac patency was preserved to minimise paraplegia risk. Retrograde bypasses were performed from the iliac grafts to the four visceral branches (coeliac bypass tunnelled retropancreatically) in end to side fashion and the origin of each

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vessel was ligated. The retroperitoneum was re-approximated over the graft.

The second stage of the repair was performed seven days later and was an endovascular repair of the entire thoraco-abdominal aorta extending from the descending thoracic aortic graft to the infrarenal abdominal aortic graft. Both the proximal and distal landing zones were longer than 5 cm. Three Medtronic Valiant endografts (Medtronic, Dublin, Ireland) were inserted. These were: 26 mm—22 mm × 150 mm distal, 30 mm × 200 mm bridging, then 32 mm × 100 mm proximal. Spinal cord protection measures included: pre-operative cerebrospinal fluid drain insertion and maintaining mean arterial pressure of 90—100 mmHg, haemoglobin >90 g/L, and oxygen saturation >95%. Completion angiograms showed no evidence of early or delayed endoleaks, and patency of the visceral branches. His post-operative stay was complicated by CSF leak managed conservatively and right femoral nerve paraesthesia that improved with physiotherapy. He was discharged on post-operative day 20.

Follow up CT angiograms at five days (Fig. 3) and 11 months post-treatment showed exclusion of the aneurysm with unchanged size and a small type II endoleak originating from a posterior intercostal branch. Bi-annual surveillance is planned with CT angiograms.

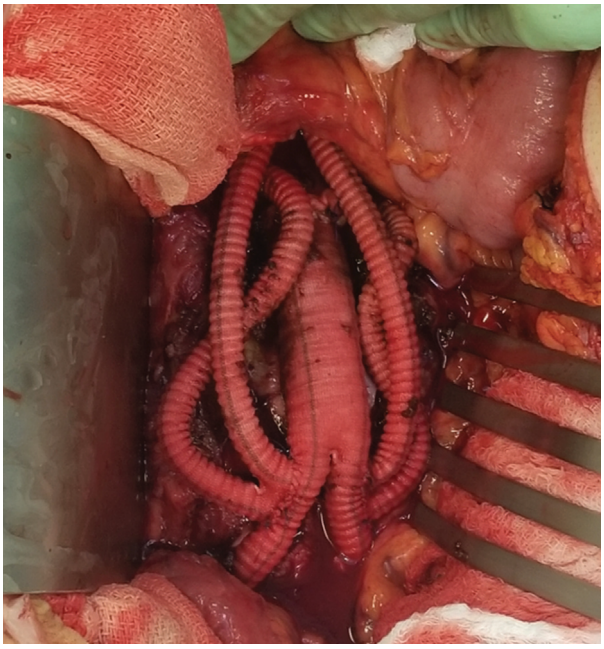

Figure 2. Intra-operative image of the first stage repair showing an infrarenal aortobiliac graft with retrograde visceral bypasses.

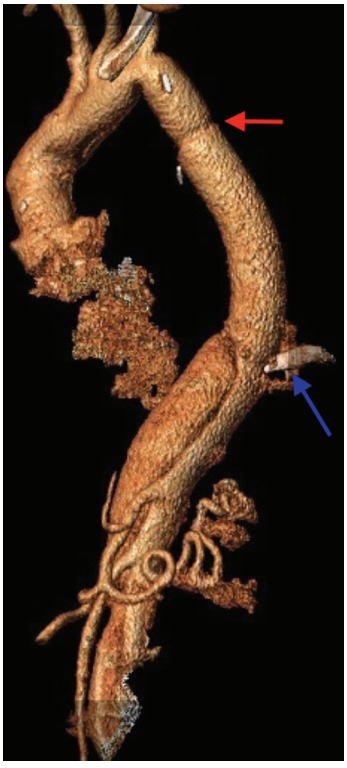

Figure 1. Three dimensional reconstruction of the thoraco-abdominal aorta showing the aneurysm and previous thoracic aortic repair locations. Red solid arrow denotes the distal extent of previous thoracic repair. Blue solid arrow denotes level of diaphragm.

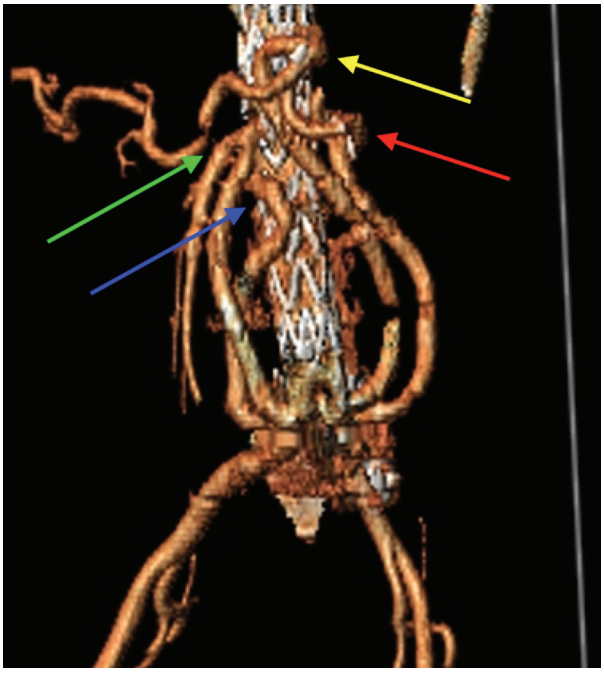

Figure 3. Three dimensional reconstruction showing infrarenal aortic repair with visceral debranching, and endovascular stenting across the thoraco-abdominal aorta. Yellow arrow: coeliac trunk bypass; red arrow: left renal artery bypass; green arrow: superior mesenteric artery bypass; blue arrow: right renal artery bypass.

Table 1. Hybrid treatment of LDS associated thoracic/thoraco-abdominal aortic pathologies in the literature.

Study	Age	Previous aortic repair	Indication	TAA/TAAA treatment	Follow up — mo	Aorta related complications (Yes/No)	Mortality at last follow up (Yes/No)	Notes
Neri et al. 2010[11]	25	Valve-sparing root replacement, TEVAR	Chronic type B AD	Open TAAA repair	0.6	No	No	Novel device used to crimp endograft then re-deploy it in sewn graft
Preventza et al. 2014[8]	N/A	Elephant trunk, TEVAR	Post TEVAR endoleak	TEVAR explant, open TAA	N/A	Yes — post TEVAR endoleak required explant at 20 mo	No	
Wipper et al. 2015[12]	44	Open ruptured AAA repair	Retrograde subacute type B AD	Open TAAA with Gore hybrid branch grafts	3	No	No	Suture-less distal visceral anastomoses
Kalra et al. 2015[13]	30	Composite ascending aortic replacement	Contained rupture TAA	TEVAR	23	No	No	
	27	Ascending, hemiarch replacement, elephant trunk	Contained rupture TAA	TEVAR	48	No	No	
Williams et al. 2015[3]	29	Open extent II TAAA repair	Type A AD	TEVAR with root replacement and arch debranching	54	No	No	
	51	Open extent III TAAA repair	Type A AD	TEVAR with total arch replacement	16	No	Yes — sepsis	Severe scoliosis, descending aorta crossing into right thorax
Hashizume et al. 2017[14]	41	Ascending aortic replacement, aortic valve replacement	Aortic sinus, arch and TAAA aneurysm	Stage 1: Bentall procedure, arch replacement Stage 2: TEVAR distal arch Stage 3: TEVAR distal descending TAA Stage 4: EVAR Stage 5: FEVAR with PMEG	24	No	No	
Shalhub et al. 2018[2]	40	TEVAR	Chronic type A AD with aneurysmal degeneration	TEVAR explant with open TAAA I repair	N/A	Yes — post TEVAR false lumen expansion at 17 mo	No	
Kölbel et al. 2018[15]	19	Open AAA, frozen elephant trunk	Acute type B AD	TEVAR with PMEG, open TAAA	1	No	Yes — ruptured vertebral artery aneurysm	

AD = Aortic dissection; N/A = Not available; TAA = thoracic aortic aneurysm; TAAA = Thoraco-abdominal aortic aneurysm; PMEG = Physician modified endograft.

DISCUSSION

Open repair is the gold standard repair for patients with heritable aortopathies.[5] Various series have established acceptable outcomes for open TAAA treatment with connective tissue diseases.[6] A recent review of TAAA treatments from the Genetically Triggered Thoracic Aortic Aneurysms and Cardiovascular Conditions (GenTAC) registry also confirmed that open TAAA treatment is associated with low peri-operative mortality and morbidity. Late treatment failure was also low at 5%.[7] Annual surveillance with CT or MRI is mandated in LDS patients,[5] with up to 88% survival at 10 years.[4]

Patients with LDS undergo multiple aortic procedures during their lifetime, which renders them high risk for re-interventions.[2,7] LDS is less frequent than Marfan syndrome and other hereditary aortopathies. In addition, patients tend to present younger.[2,7,8] An endovascular only approach is possible when surgical grafts form both the proximal and distal landing zones but is associated with an

up to 30% rate of retrograde dissection and 40% requiring stent graft explant.[2] A hybrid approach (reported literature summarised in Table 1) can be employed to reduce perioperative morbidities especially those involving redo thoracotomy.[9] In LDS patients, there are concerns with use of endografts in native aortas as radial forces would result in an already fragile aorta undergoing further dilation. This probably explains the high re-intervention rates and morbidity.[2] Preventza et al. reported a 17% re-intervention rate in a cohort which contained 90% of endografts landing in native aorta.[8]

It was decided against an open repair to avoid redo thoracotomy in a patient with two previous sternotomies, and two left thoracotomies. In addition, he was at high risk of spinal cord ischaemia given planned long aortic coverage. There were difficulties anticipated in positioning for open repair because of thoracolumbar spinal fusions. The decision was made to perform a two stage hybrid repair, which would potentially reduce the risk of paraplegia.[10]

Previous experiences with hybrid repair mainly constitute case series with limited follow up. These include a patient who initially underwent staged elephant trunk repair followed by TEVAR but required explant and an open repair for persistent endoleak.[8] The Duke University experience also included two patients with LDS who underwent aortic arch debranching with TEVAR. At three month follow up, there was one non-aortic mortality.[3]

Conclusions

Hybrid TAAA repair was a valid treatment option in this patient with LDS and multiple previous aortic procedures. It minimised the morbidity of revision surgery and mitigated potential treatment failure by achieving endovascular seal in surgical grafts. Short term surveillance showed no complications. Limitations to making recommendations include lack of long term follow up.

CONFLICT OF INTEREST

None.

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APPENDIX A. SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.ejvsvf.2021.04.004.

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