Endovascular treatment of thoracoabdominal aortic aneurysm in Loeys-Dietz syndrome

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A 37-year-old woman with Loeys-Dietz syndrome (LDS) was referred to our centre due to diameter progression (to 52 mm) of a dissecting thoracoabdominal aortic aneurysm (TAAA) (Fig. 1). Medical history revealed previous acute thoracic aortic dissection with entry on the level of the left subclavian artery (LSA) five days after caesarean section in 2008. She underwent urgent thoracic endovascular aortic repair (TEVAR) with stent-graft deployment intentionally occluding LSA and additional stent placement in the dissected superior mesenteric artery (SMA). Six years later, due to ascending aortic dissection, the patient underwent valve-sparing aortic root replacement with transposition of brachiocephalic trunk and left common carotid artery, with distal prosthetic anastomosis to the previously implanted stent-graft. Each procedure was performed in a different centre. Moreover, the patient suffered from poorly controlled asthma and obesity, and she did not agree to extensive open surgery. We had two major problems: the genetic origin of the dissection and difficulty of vascular access due to previous interventions. TEVAR resulted in complete LSA orifice closure, which precluded left brachial access. Brachiocephalic trunk was extremely tortuous after its transposition, thus hindering the possibility of right brachial access (Fig. 2). Nonetheless, the patient was scheduled for endovascular treatment using a custom-made device (CMD). Based on computed tomography (CT), a three-fenestration stent-graft with one upward branch was designed and manufactured by Cook Medical (Bloomington, IN, USA) (Fig. 3). Despite the fact that the left renal artery was directed downwards, an upward branch was designed due to the lack of upper vascular access. The surgery started with placement of a thoracic endograft through femoral access ending below the previously implanted stent-graft. Then, the CMD was introduced, and visceral arteries were identified in angiography. Afterwards, the stent-graft was partly opened to cannulate fenestrations. Balloon-expandable Bentley covered stents (Innomed, Hechingen, Germany) were introduced to the celiac trunk, SMA, and right renal artery. Then, the CMD was fully opened, overlapping with the previously implanted stent-graft, and stents were deployed. Next, the left renal artery was cannulated via the branch. The vessel was bridged with a stent-graft using a self-expandable Fluency covered stent (Bard Peripheral Vascular, Tempe, AZ, USA) and reinforced with a Zilver bare stent (Cook Medical, Bloomington, IN, USA). Subsequently, a bifurcated stent-graft was deployed down to the common iliac arteries. Control angiography showed full patency of the stent-graft and visceral arteries. Postoperative stay and three-month follow-up were uneventful. Control CT showed full patency of the stent-graft, branches, and bridged vessels, and aneurysm sac shrinkage with insignificant endoleak type II left for further observation (Fig. 4). In conclusion, endovascular treatment of TAAA in LDS is a feasible alternative for open surgery, even if it requires patient-tailored solutions. Long-term follow-up is necessary to determine the outcome of the procedure.

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Conflict of interest: Piotr Hammer — owner of Hammermed.

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