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CASE REPORT

Single Branch Endovascular Aortic Repair Procedure for an Abdominal Aortic Aneurysm in a Patient With Horseshoe Kidney: A Case Report

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Introduction: Horseshoe kidney (HK) is an anatomical variant characterised by abnormalities in the position, rotation, and vascular supply of the kidney, with functioning renal masses on both sides of the vertebral column fused together at the isthmus. Due to the altered pattern of kidney vasculature, endovascular aortic repair for aortic abdominal aneurysm (AAA) in the presence of HK requires vascular anatomy specific planning.
Report: A 68 year old male, with multiple comorbidities, presented with an asymptomatic AAA and HK. The kidney vasculature was characterised by the presence of three arteries: two arising laterally at the same level and a third polar artery arising from below. The polar artery was 6 mm in diameter and larger than the other two; therefore, in order to preserve this artery, a custom-made device with a single side branch was implanted below the main renal arteries. A balloon expandable covered stent was used to complete the side branch into the polar renal artery. The follow-up computed tomography angiography revealed a successful outcome, with total aneurysm exclusion, branched graft patency, no endoleak, and unchanged renal function.
Discussion: This case report shows a possible surgical solution for a case of HK with AAA and the importance of accurate endovascular planning. Large polar arteries, if present, need to be preserved, and custom-made devices in the modern endovascular era permit that. This approach could represent the best option for complicated patients.

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INTRODUCTION

Horseshoe kidney (HK) is the most common fusion defect of the kidneys, with a prevalence of 0.25% in the normal population and a male preponderance of 2:1.[1,2] This alteration consists of a fusion of the two kidneys across the midline, merged into an isthmus of renal parenchyma (80%) or fibrous band (20%).[1] The vasculature of HKs frequently diverges from the ordinary, with additional arteries arising from the aorta or iliac arteries. Most HK cases described in the literature show vasculature with one artery for each side plus one artery, originating from the aorta, for the isthmus.[3] The rest of the cases are represented by various patterns, described by Eisendrath *et al*. in 1925.[3] The classification system currently used for patients with HK is derived from this work. It includes five types, depending on number and origin of the arteries supplying the HK parenchyma: I. one artery of each side of the HK; II. an auxiliary aortic branch to the renal isthmus in addition to type I; III. two renal arteries on each side; IV. two renal arteries on each side with one or more originating from the iliac arteries or the isthmus branch; and V. multiple renal arteries arising from the aorta, mesenteric, and iliac arteries.

In the case of aortic abdominal aneurysm (AAA), the concomitant presence of an HK with regular vasculature does not modify the treatment strategy and a standard endovascular aortic repair (EVAR) remains feasible, considering technical details and anatomical conditions. Problems arise when the vasculature is different with irregular vessels essential for renal perfusion.[4]

For this reason, more complex endovascular techniques, such as fenestrated or branched customised devices, are the current best solutions to exclude AAAs with a concomitant HK and complex renovascular anatomy that does not allow standard EVAR, so as not to occlude extra renal arteries arising from the aorta.[4] Open surgical repair (OSR) remains the gold standard for fit patients, but the invasive approach, extended post-operative period, and high risk of complications relegate this option to well defined cases.

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This case report aimed to describe a successful case of a custom made branched EVAR (B-EVAR) performed in a patient with AAA and concomitant HK but who was unfit for OSR.

REPORT

A 68 year old male patient was admitted to the institution with an asymptomatic AAA and concomitant HK. He had a previous history of arterial hypertension, dyslipidaemia, acute myocardial infarction, type 2 diabetes mellitus, and chronic obstructive pulmonary disease. The renal vascular supply pattern was represented by three arteries, two arising laterally (03:00 and 10:15 position on a clock) at the same height, and a third polar artery arising 75 mm below (Eisendrath classification type 2). The diameter of the polar artery was 6 mm, which was larger than the other two (Fig. 1). The aneurysm was about 64 mm in diameter with an adequate infrarenal neck regarding the two main renal arteries. If all three renal arteries are considered, the AAA was strictly pararenal because the polar one arose from the middle of the aneurysm (Fig. 1D).

Due to the patient's multiple comorbidities and the uncommon vascular pattern, a tailored endovascular approach was selected. A custom made, branched, abdominal endograft was planned, considering the non-aneurysmal aortic tract immediately below the two main renal arteries as the proximal sealing zone. One internal and external side branch 6 mm in diameter and 18 mm in length, arising nearly in the sagittal plane (12:15 position on a clock), was designed for the polar renal artery. The bifurcated configuration was planned to land distally at the level of the common iliac arteries. Two limb extensions completed the system, with distal landing in the common iliac arteries (Fig. 2).

Branched endovascular aneurysm repair (B-EVAR) was undertaken, under general anaesthesia, using bilateral femoral and left axillary open surgical access. A custom-made device (Cook Zenith G38846-AAA-Branch-Graft, Cook Medical, Bloomington, IN, USA) was deployed via the femoral approach (sheath diameter 20 F), the polar artery was cannulated via axillary access (sheath diameter 7 F), and the branch component was completed with a 7 x 57 mm balloon-expandable stent (BeGraft PLUS, Bentley InnoMed GmbH, Hechingen, Germany) to line the branch and ensure complete vascularisation of the HK (Fig. 3A and B). The B-EVAR was completed in a standard bifurcated

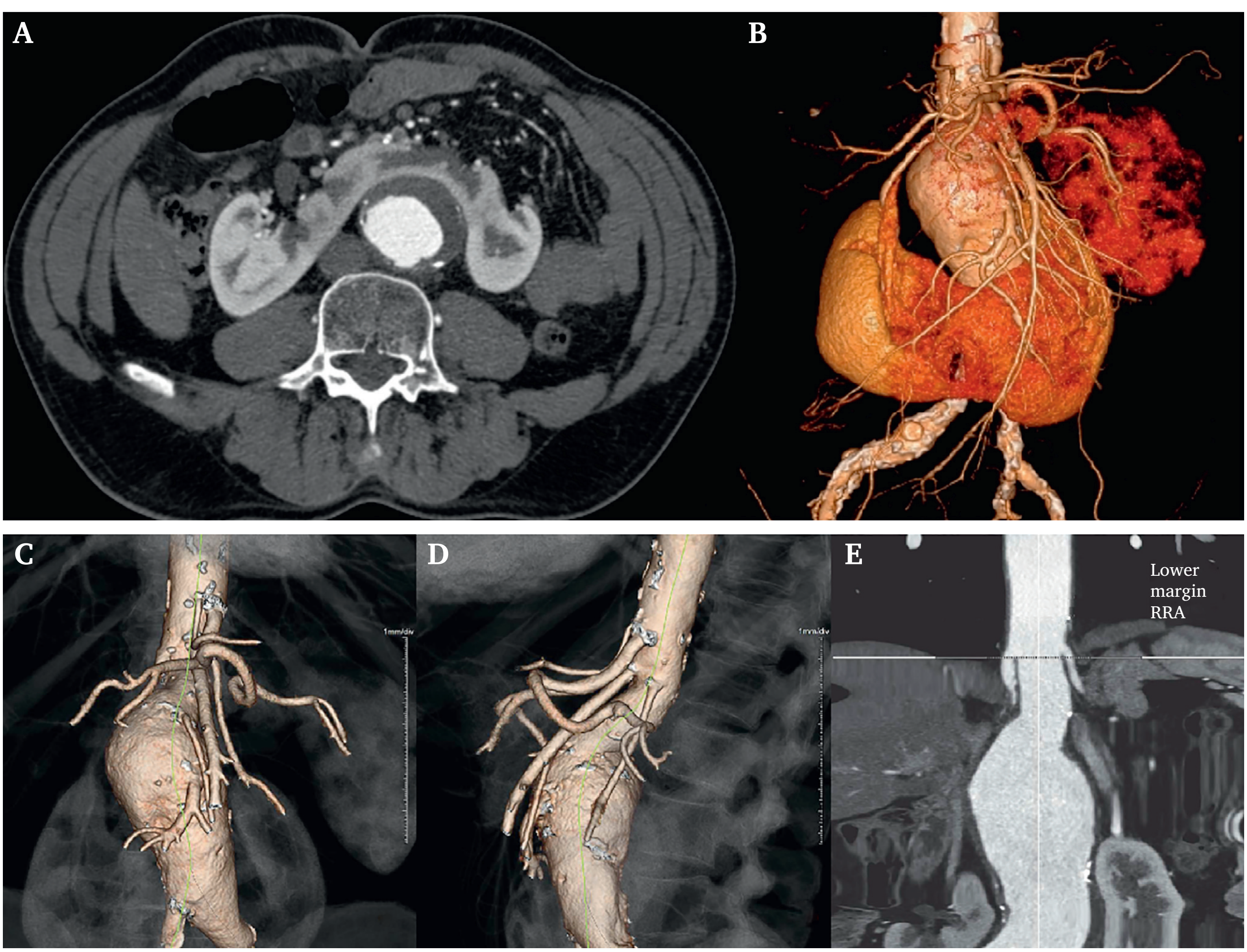

Figure 1. Pre-operative images of the aortic aneurysm and horseshoe kidney. A. Computed tomography angiography scan. B. 3D computed tomography angiography reconstruction. C and D. Renal vessel details. E. Coronal centre line adjusted computed tomography scan.

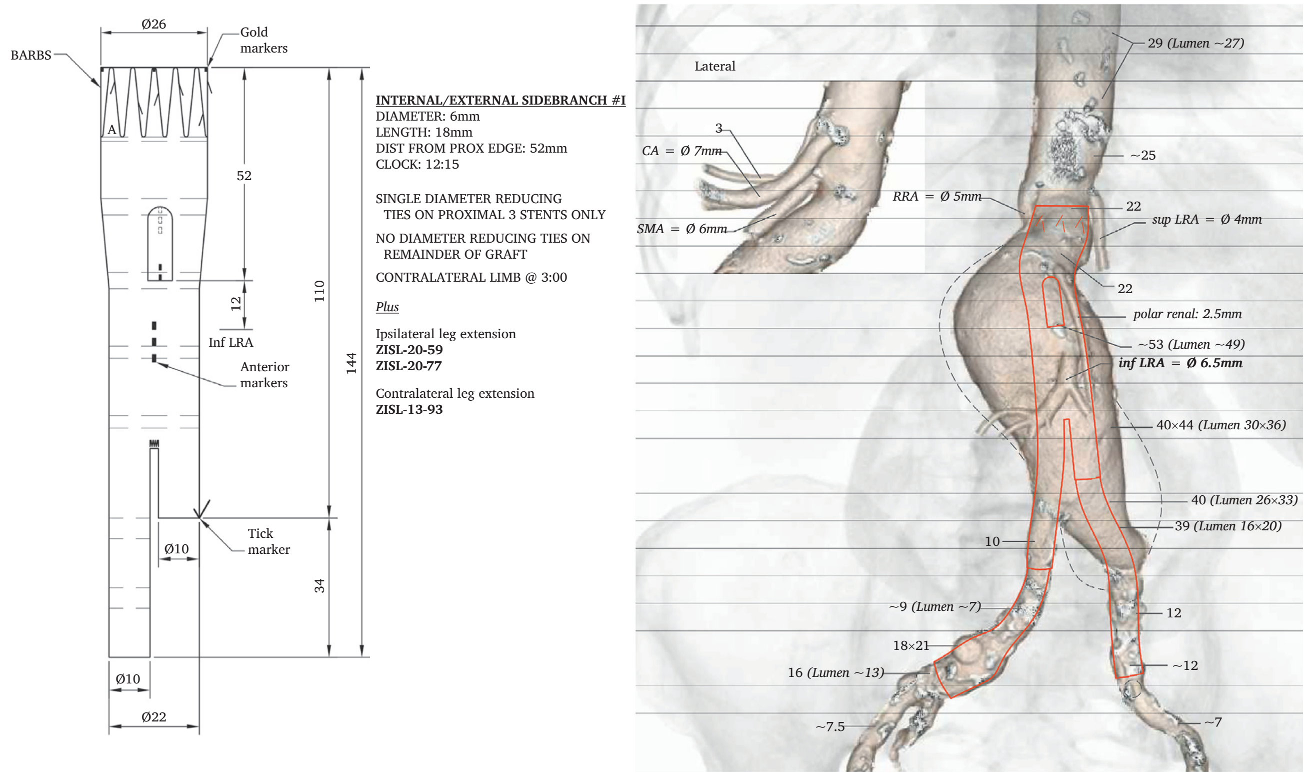

Figure 2. Endovascular graft work sheet with graft technical details.

configuration with distal seal achieved in the common iliac arteries (Cook ZISL-20-93 and ZISL-13-93, Cook Medical). After moulding the entire implant with a Coda Balloon Catheter (Cook Medical), the intra-operative completion angiogram showed adequate exclusion of the aneurysm, with patency of the renal branch, unaltered renal perfusion, and no evidence of endoleak (Fig. 3C).

The post-operative period was uneventful and the patient was discharged on the third post-operative day on dual antiplatelet therapy. The follow-up computed tomography angiography scan showed a successful outcome, with total aneurysm exclusion, branched graft patency, and no sign of endoleak (Fig. 3D). Nine months post-operation, the last duplex ultrasonography confirmed aneurysm exclusion and graft patency.

DISCUSSION

With their particular morphology, horseshoe kidneys differ from ordinary kidneys in three main ways: location, orientation, and vasculature. With a HK, the ascent to the final site is held back by the inferior mesenteric artery at L3 level or lower. During the second month of development, the ascent is coupled with 90° medial rotation. Due to the isthmus, HKs experience malrotation and the ureters need to either pass over the isthmus or down the anterior kidney surface, causing urinary stasis and urinary tract infections.[5] No single genetic cause has been described, although several aetiological factors may contribute to the development of HK, such as teratogenic drugs like thalidomide, alcohol consumption, and structural factors such as flexion or rotation of the caudal spine.[2] They are mainly asymptomatic with an incidental identification. A small percentage of patients present with non-specific clinical symptoms such as abdominal pain or urinary tract infection.

Despite the high frequency of abnormalities in the extraparenchymal vessels, due to the poor collateral arterial supply, the intrarenal vascular segmental pattern remains almost unaltered. Surgically, this means that ligation of any of these arteries could result in localised segmental renal ischaemia.[6]

While EVAR has progressively become the mainstay in the treatment of infrarenal AAA, the simultaneous presence of a HK requires more advanced solutions to ensure satisfactory technical success and optimal clinical outcome. Open surgical repair has been the first choice for years, with isthmus preservation and conservation of the renal vasculature (especially with vessels $>$3 mm in diameter); however, in the endovascular era, this approach is becoming less common, due to extensive dissection, trauma to structures, and blood loss, which are inevitable in OSR. Conventional EVAR allows preservation of the isthmus with minimal surgical access, avoiding the forementioned complications. This means a decrease in hospital length of stay and lower peri-operative mortality and morbidity rates. A standard EVAR can be an option in truly Eisendrath type I or II HK,[4] or in the presence of small diameter collateral branches. In a few case reports, analysed by Sachsamanis *et al*.,[7] low grade acute renal deterioration was reported, although these changes were

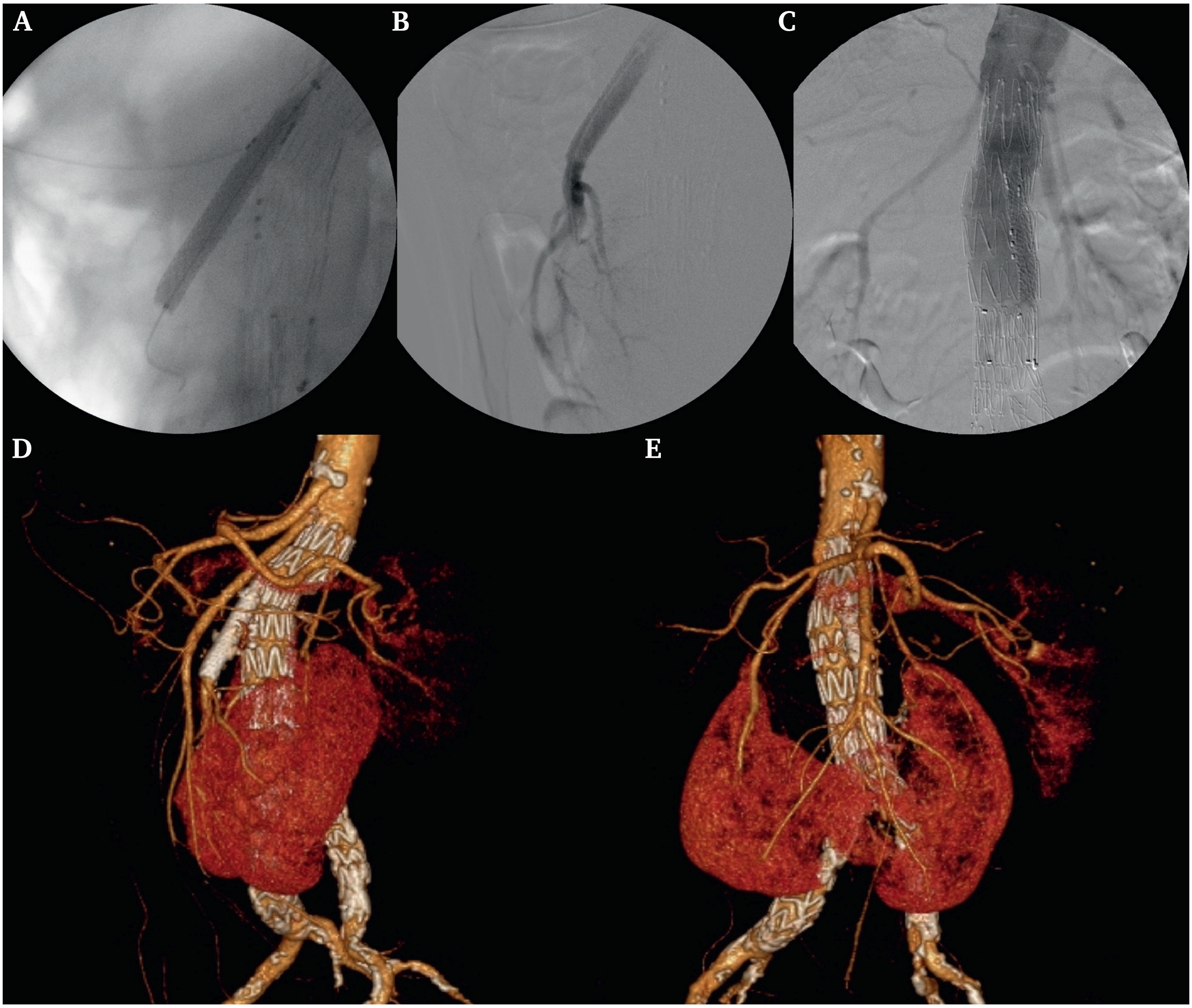

Figure 3. Procedure and results. A. Polar artery branch stent deployment. B. Branch angiography. C. Intra-operative angiography control. D and E. Post-operative 3D computed tomography angiography reconstruction.

almost all transient. However, in cases of more complex renal vessel anatomy (or with arteries >3 mm in diameter), the elective use of custom-made devices or physician-modified and in situ laser fenestration solutions for urgent settings are of great relevance and enable more surgical indications, especially for patients who are unsuitable for OSR. Recent case reports on the use of fenestrated endografts have shown promising short term results, while longer term outcomes are needed.[8,9]

Unlike these cases, this case report chose a branched endograft because of position and orientation of the polar renal artery. Arising from the middle of the aneurysm, there was no neck to use as a sealing zone in the area adjacent to this artery. Moreover, there was a gap between the ostium and the expected position of the endograft >5 mm.[10] It was also preferred to use a branched graft because length and morphology of the polar artery (with no bifurcations or collateral vessels) enabled sufficient distal overlap. The orientation of the artery (downwards <30° with the aorta in a sagittal plane) permitted a final anatomical geometry close to the natural one. Lastly, it was decided to use a BeGraft PLUS (Bentley InnoMed GmbH) for the polar artery because of its high radial force and its behaviour pattern in the renal arteries.

Conclusion

This case report shows that appropriate planning tailored to the expected aberrant renovascular anomalies enabled a successful B-EVAR in a multimorbid patient. In these rare anatomical conditions, the preservation of all the large renal vessels with custom made endovascular devices may represent the best option and should be considered both in elective and urgent settings.

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CONFLICTS OF INTEREST

None.

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