

Right-sided aortic arch with Kommerell's diverticulum: a case report of a rare cause of type B dissection

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Background

A right-sided aortic arch (RAArch) is present in approximately 0.1% of the population. A Kommerell's diverticulum (KD), a remnant of the dorsal aortic arch usually refers to an aneurysmal aortic enlargement at the origin of an aberrant left subclavian artery (ALSA) and is associated with an increased risk of aortic dissection.

Case summary

A 59-year-old female smoker with a history of hypertension and hypercholesterolaemia presented with a 24-hour history of sudden-onset and severe stabbing chest pain radiating to the interscapular region. Physical examination was normal except for bilateral basal crepitations. Computed tomography angiography (CTA) showed a type B aortic dissection in a RAArch with an ALSA arising from KD with a peri-aortic haematoma and haemothorax without any active contrast extravasation. After medical stabilization, a semi-urgent hybrid repair was performed with a right carotid-subclavian bypass, thoracic endovascular aortic repair (TEVAR), a plug in the left subclavian artery, and left carotid-subclavian bypass due to severe ischaemia of the left arm. The post-operative CTA showed patent bypasses, aortic remodelling, and a minimal type IIa endoleak at the level of the ALSA.

Discussion

In patients with a type B dissection and KD, hybrid repair including TEVAR is feasible after careful pre-operative assessment of the patient's unique anatomy and may reduce post-surgical morbidity and mortality compared to open surgery. Prophylactic repair may be considered in patients with an asymptomatic RAArch and KD.

Keywords

Right-sided aortic arch • Congenital aortic anomaly • Aortic dissection • Acute aortic syndrome • Kommerell's diverticulum • Case report

ESC Curriculum

9.1 Aortic disease • 2.1 Imaging modalities

Learning points

- A right-sided aortic arch with an aberrant left subclavian artery and a Kommerell's diverticulum is associated with an increased risk of type B acute aortic syndrome.
- Hybrid treatment including endovascular repair is technically feasible in patients with a right-sided aortic arch and Kommerell's diverticulum who develop a type B dissection after careful assessment of the patient's unique anatomy.
- A prophylactic surgical repair may be considered in asymptomatic patients with a right-sided aortic arch and Kommerell's diverticulum.

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Introduction

A right-sided aortic arch (RAArch) is a congenital anomaly of the aortic arch with an incidence of approximately 0.1%. It results from a disordered embryogenesis of the branchial arches with either abnormal persistence or involution of embryonic vascular segments.¹ Edwards et al.² have divided RAArch into three major types. Type I presents with mirror-image branching of the major arteries ascending from the aortic arch. It is mostly associated with cyanotic congenital heart disease such as tetralogy of Fallot, truncus arteriosus, and transposition of the great arteries. Type II presents with an aberrant left subclavian artery (ALSA) typically running behind the oesophagus through the mediastinum and is rarely associated with congenital heart disease. In 20–60% of cases, the ALSA originates from Kommerell's diverticulum (KD), a remnant of the left dorsal aortic arch which presents as an aneurysmal aortic enlargement at the origin of the ALSA. These patients with a KD have an increased risk of aortic dissection and rupture.³ Type III presents with an isolated left subclavian artery and is often associated with congenital heart disease. Type I and II each account for roughly half of the cases, whereas type III is extremely rare.^{1,4} In this case report, a type B aortic dissection in a patient with a type II RAArch with an ALSA arising from KD is presented.

Timeline

Day 1	The patient presented to the ED with a 24-hour history of sudden-onset and severe stabbing chest pain. CTA showed a type B aortic dissection in a RAArch with an ALSA arising from Kommerell's diverticulum.
Day 1–3	Optimal medical management was given in the ICU.
Day 3	Semi-urgent hybrid repair with a right carotid-subclavian bypass, TEVAR, plug in the left subclavian artery, and left carotid-subclavian bypass due to ischaemia of the left arm.
Day 6	Postoperative CTA showed a minimal type IIa endoleak at the level of the ALSA.
Day 3–16	The patient was diagnosed with a respiratory infection requiring antibiotics, intubation, and prolonged ICU stay.
Day 16	The patient was transferred to vascular ward.
Day 22	The patient was discharged in good cardiovascular and neurological condition.
1-month follow-up after discharge	The patient was in good postoperative state with bilateral radial pulses, no recurrent pain, well-controlled blood pressure, and successful smoking cessation.
3-month follow-up after discharge	The patient remained in good general condition with a well-controlled blood

pressure and persistent smoking cessation. Follow-up CTA showed a good position of the stent, patent carotid-subclavian bypasses, a diminished endoleak, and a decrease in size of the false lumen and Kommerell's diverticulum.

ALSA, aberrant left subclavian artery; CTA, computed tomography angiography; ED, emergency department; ICU, intensive care unit; RAArch: right-sided aortic arch; TEVAR, thoracic endovascular aortic repair.

Case presentation

A 59-year-old female smoker with a history of hypertension and hypercholesterolaemia was admitted to the emergency department (ED) with a 24-hour history of sudden-onset and severe stabbing chest pain radiating to the interscapular region. Hypertension and hypercholesterolaemia were treated by her primary care physician with a beta-blocker, a thiazide, and a statin. Her right brachial blood pressure upon admission was 151/97 mmHg without significant right/left difference. A regular heart rate of 87 bpm, an oxygen saturation of 91%, and a temperature of 36.0°C were noted. Intravenous opioids were administered to control her pain. Physical examination was normal except for bilateral basal crepitations. Arterial blood gas showed a respiratory alkalosis (pH 7.46) with a decreased pO₂ (64 mmHg), a decreased pCO₂ (32 mmHg), and a normal HCO₃ (22 mmol/L). Electrocardiogram showed signs of left ventricular hypertrophy with a Sokolow index > 35 mm without signs of cardiac ischaemia or ST elevation. An urgent computed tomography angiography (CTA) revealed a RAArch with an ALSA arising from KD with a type B aortic dissection extending from the ALSA to the level of the diaphragm together with a peri-aortic hematoma and a right-sided haemothorax without any active contrast extravasation (Figures 1 and 2). The patient was admitted to the intensive care unit (ICU) for aggressive blood pressure control using intravenous beta-blockade. Echocardiography showed no structural nor valvular heart disease with a normal cardiac function and no pericardial effusion.

The patient was discussed within the local multidisciplinary aortic team, and there was decided to perform a semi-urgent repair to treat this acute type B dissection with a peri-aortic haematoma and right-sided haemothorax. Due to the semi-urgent need for repair, there was opted for a hybrid repair, which was also influenced by the patient's choice, who preferred not to have open aortic surgery despite her young age, and the fact that she was a physically unfit active smoker. Pre-operatively, a spinal drain was placed. In the hybrid room, bilateral radial arterial lines, a central venous catheter, and a rapid pacing electrode were introduced through the left femoral vein. An arch angiogram demonstrated a dominant right vertebral artery. Hence, a right-sided carotid-subclavian bypass was performed (Gelsoft plus 6 mm, Terumo, Vascutek Ltd, Scotland, UK) followed by stent-grafting (31-31 200), the entire descending thoracic aorta covering the ostia of both subclavian arteries to land 3 cm above the celiac trunk (GORE® TAG® Conformable Thoracic Stent Graft with ACTIVE CONTROL, W.L. Gore & Associates, Flagstaff, USA). The ALSA was occluded with an Amplatzer plug 12 mm (Abbott vascular, Diegem, Belgium) via the left brachial artery just distal of the KD. Completion angiogram showed bilateral patent

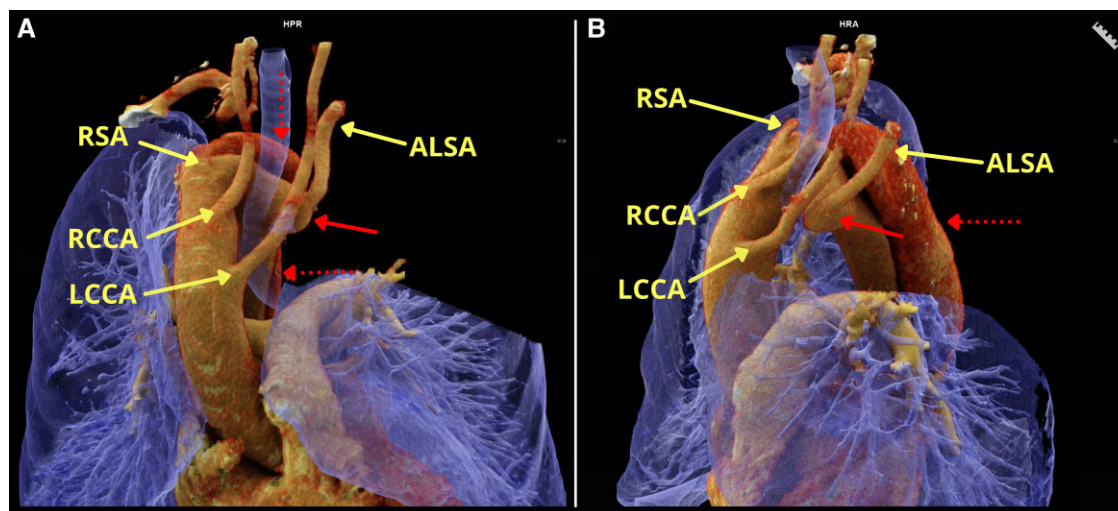


Figure 1 Three-dimensional virtual rendering technique reconstructions of the pre-operative computed tomography angiography in an antero-posterior (A) and a left lateral (B) view, left lung apex is cut away. Solid arrow: Kommerell diverticulum; dotted arrow: false lumen of the dissection. ALSA, aberrant left subclavian artery ascending from Kommerell's diverticulum; RSA, right subclavian artery; RCCA, right common carotid artery; LCCA, left common carotid artery. Images: D. Devos, MD, PhD, cardiovascular radiologist.

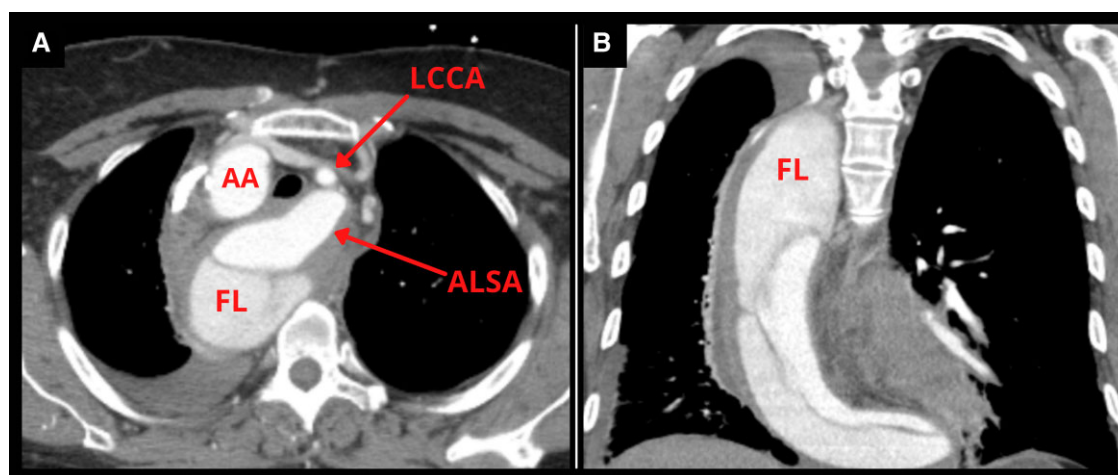
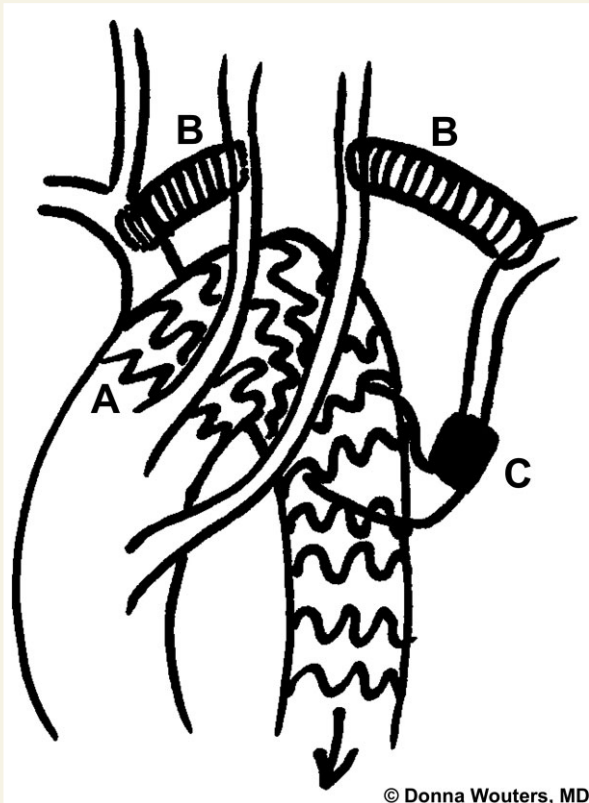


Figure 2 Conventional slices of the pre-operative computed tomography angiography. Axial slice showing the aberrant left subclavian artery ascending from Kommerell's diverticulum and the start of the aortic dissection, the peri-aortic hematoma and the right-sided haemothorax (A). Coronal slice showing the distal part of the aortic dissection in the descending aorta and the peri-aortic hematoma (B). AA, ascending aorta; ALSA, aberrant left subclavian artery ascending from Kommerell's diverticulum; FL, false lumen; LCCA, left common carotid artery.

common carotid arteries and right-sided carotid-subclavian bypass, and aortic remodelling without any flow in the KD. Because of severe ischaemia of the left arm and hand, a left sided carotid-subclavian bypass was performed (Gelsoft plus 6 mm) during the same procedure (Figure 3). The postoperative course in ICU was complicated with a respiratory infection requiring intravenous antibiotics, ventilatory support, and prolonged ICU-stay. The postoperative CTA showed patent bypasses, aortic remodelling, and minimal type IIa endoleak at the level of the ALSA.

The patient was discharged in good cardiovascular and neurological condition after 3 weeks with a well-controlled blood

pressure. Her medication consisted of olmesartan 40 mg q.d., amlodipine 5 mg b.i.d., bisoprolol 10 mg q.d., atorvastatin 40 mg q.d., and 80 mg of aspirin q.d. Smoking cessation was strongly encouraged. At 1- and 3-month follow-up, the patient had quit smoking and was still fatigued but in good shape. Clinical examination showed normal wound healing, bilateral normal brachial and femoral pulses, and good blood pressure control. At 3 months follow-up, a follow-up CTA showed a good position of the stent-graft, patent carotid-subclavian bypasses, a diminished type IIa endoleak, and a decrease in size of the false lumen and the KD (Figure 4).



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Figure 3 Illustration of the vascular anatomy after hybrid repair with an endovascular stent-graft covering the entire descending thoracic aorta including the ostia of both subclavian arteries (A), a bilateral carotid-subclavian bypass (B), and an Amplatzer plug in the aberrant left subclavian artery just distal of Kommerell's diverticulum (C). Copyright Donna Wouters, MD, general surgery resident.

Discussion

A RAArch is mostly asymptomatic. In children, symptoms are related to associated congenital heart disease or to compression of the oesophagus or trachea by an ALSA. In adults with a type II RAArch, approximately 5% of patients present with symptoms, mostly due to compression of surrounding structures causing e.g. dysphagia, dyspnoea, or atypical chest pain.^{1,3,5} Rarely, as in this case, the RAArch is identified due to an acute type B aortic dissection starting at the origin of the KD. This diverticulum is associated with a higher risk of dissection and rupture; however, due to its rareness, the true incidence of these complications is unknown since only small case series have been described.^{3,6} The largest cohort of patients with KD included 212 cases of which 11% and 4% presented with aortic dissection and rupture, respectively.³ Likewise, since nearly all cases had surgical repair, the natural course of an aortic dissection with KD is unknown. The 2014 ESC guidelines on the diagnosis and treatment of aortic diseases recommend aortic repair only in complicated type B dissection cases, defined as having persistent or recurrent pain, uncontrolled hypertension despite medical treatment, evidence of early aortic expansion, malperfusion, or signs of rupture.⁷ In this case, the entry site of the type B dissection was associated with KD and a peri-aortic haematoma and haemothorax; hence, surgical treatment was warranted.³

Several case reports have been published highlighting the technical challenges of open surgical, endovascular, and hybrid treatments associated with the anatomical complexity of this pathology. Given the rarity of RAArch dissections, no evidence-based treatment guidelines are available.^{3,6,8–10} The type of intervention should be discussed in a multidisciplinary aortic team and will be influenced by the patient's anatomy and comorbidities as well as the available surgical expertise. Endovascular repair and hybrid surgery are technically feasible in patients with a type B dissection and KD with careful pre-operative assessment of the patient's unique anatomy and may reduce post-surgical morbidity and mortality compared to open surgery.^{3,6,11}

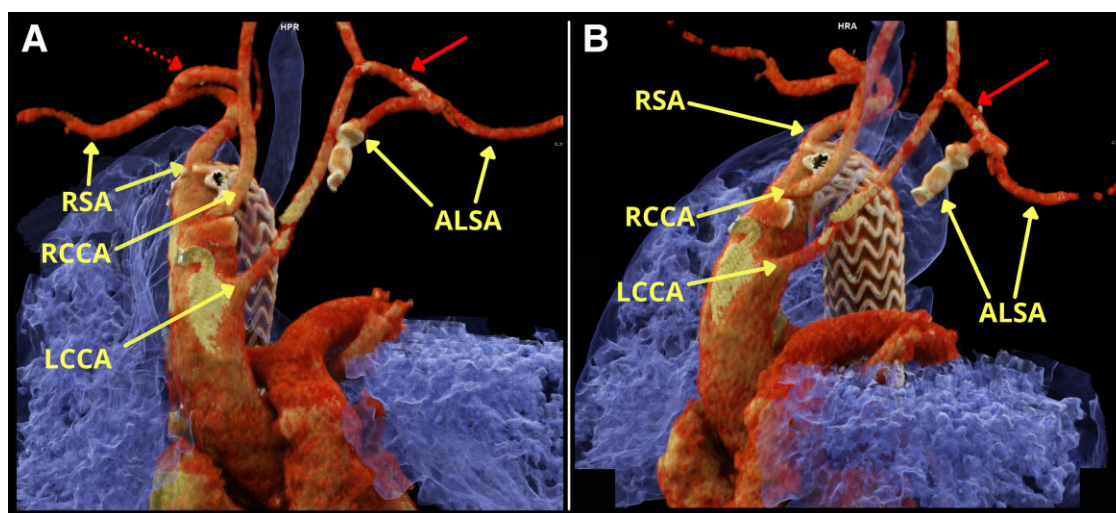


Figure 4 Three-dimensional virtual rendering technique reconstructions of the follow-up computed tomography angiography in an anteroposterior (A) and a left lateral (B) view, left lung apex is cut away. Solid arrow: left carotid-subclavian bypass; dotted arrow: right carotid-subclavian bypass. ALSA: aberrant left subclavian; LCCA: left common carotid artery; RSA, right subclavian artery; RCCA, right common carotid artery. Images: D. Devos, MD, PhD, cardiovascular radiologist.

Increasingly, type II RAArch is incidentally noted on imaging studies. In symptomatic patients, especially with severe oesophageal or tracheal compression, the surgical indication is clear. However, in asymptomatic patients, the indication for preventive treatment is less straightforward.^{3,11} All patients with a RAArch should have a CTA or magnetic resonance angiography to evaluate the anatomy and to identify a KD.¹² Some authors have suggested to intervene based on the aortic aneurysm size or the size of the origin of the ALSA; however, rupture and dissection have been described in KD diameters as small as 20–25 mm. Hence, given the increased risk of complications, early surgical intervention may be considered with lower threshold in size of the aneurysm (i.e. the KD) compared to a classical descending aortic aneurysm. The indication and type of intervention should be discussed in a multidisciplinary aortic team taking into account the risks associated with open, endovascular, and hybrid repair.^{3,11} Optimal medical management should be initiated promptly in these patients including strict blood pressure control.⁷

Conclusion

A RAArch with an ALSA arising from KD presenting as an acute type B aortic dissection is an extremely rare condition. In these patients, hybrid treatment including endovascular repair is technically feasible after careful pre-operative assessment of the patient's unique anatomy and may reduce post-surgical morbidity and mortality compared to open surgery. Prophylactic repair may be considered in patients with an asymptomatic RAArch and KD.

Lead author biography



Dr. Sami Marzouki is a recently graduated medical doctor from Ghent University, Belgium. Currently, he is in his first year of Radiology residency at AZ Sint-Jan in Bruges, Belgium. He has a genuine interest in cardiovascular imaging and academia.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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