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# Clinical cases referring to diagnosis and management of patients with thoracic aortic pathologies involving the aortic arch: a companion document of the 2018 European Association for Cardio-Thoracic Surgery (EACTS) and the European Society for Vascular Surgery (ESVS) expert consensus document addressing current options and recommendations for the treatment of thoracic aortic pathologies involving the aortic arch

Martin Czerny[a,*,†], Jürg Schmidli[b,‡], Luca Bertoglio[c,‡], Thierry Carrel[b,†], Roberto Chiesa[c,‡], Rachel E. Clough[d,‡], Martin Grabenwöger[e,†], Fabian A. Kari[a,†], Carlos A. Mestres[f,†], Bartosz Rylski[a,†], Florian Schönhoff[b,†], Konstantinos Tsagakis[g,†] and Thomas R. Wyss[b,‡]

[a] Department of Cardiovascular Surgery, University Heart Center Freiburg-Bad Krozingen, Freiburg, Germany
[b] Department of Cardiovascular Surgery, Inselspital, Bern University Hospital, University of Bern, Bern, Switzerland
[c] Division of Vascular Surgery, "Vita salute" University, Ospedale San Raffaele, Milan, Italy
[d] Division of Imaging Sciences and Biomedical Engineering, St Thomas' Hospital, King's College London, London, UK
[e] Heart Center Hiertzing, Vienna, Austria
[f] University Clinic for Cardiovascular Surgery, University Hospital Zurich, Zurich, Switzerland
[g] West German Heart Center, Essen, Germany

* Corresponding author. University Heart Center Freiburg-Bad Krozingen, Albert Ludwigs University Freiburg, Faculty of Medicine, Hugstetterstrasse 55, 79106 Freiburg, Germany. Tel: +49-761-27028180; fax: +49-761-27025500; e-mail martin.czerny@universitaets-herzzentrum.de (M. Czerny).

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This compilation of clinical cases is meant as a companion to the 2018 European Association for Cardio-Thoracic Surgery (EACTS) and the European Society for Vascular Surgery (ESVS) expert consensus document addressing current options and recommendations for the treatment of thoracic aortic pathologies involving the aortic arch [1]. This document will help translate the recommendations given in the main manuscript into clinical practice with a particular emphasis on the small details that often guide the decision in the direction of open surgery or endovascular therapy or sometimes towards conservative treatment when general conditions put the likelihood of survival or a quality-of-life benefit of aortic treatment into perspective.

†Representing the European Association for Cardio-Thoracic Surgery (EACTS).
‡Representing the European Society for Vascular Surgery (ESVS).
On behalf of the writing committee for the 2018 EACTS/ESVS expert consensus document addressing current options and recommendations for the treatment of thoracic aortic pathologies involving the aortic arch.

The main document introduced a modified terminology for better describing the extent of repair by using the Ishimaru attachment zones not only as a reporting standard for endovascular procedures but also for replacement/exclusion from circulation for open procedures [2] as shown in Fig. 1.

## CASE 1: AORTIC ARCH ANEURYSM

### Clinical scenario

A 73-year-old man undergoes annual computed tomography angiography (CTA) surveillance of an atherosclerotic aortic arch aneurysm that was detected by chance when a chest radiograph was performed for unspecific pain 5 years ago. Within the last year, a diameter increase of 6 mm that now equals a maximum diameter of 57 mm was detected. The patient is clinically asymptomatic. Due to size and progression, therapy is recommended.

REPORT

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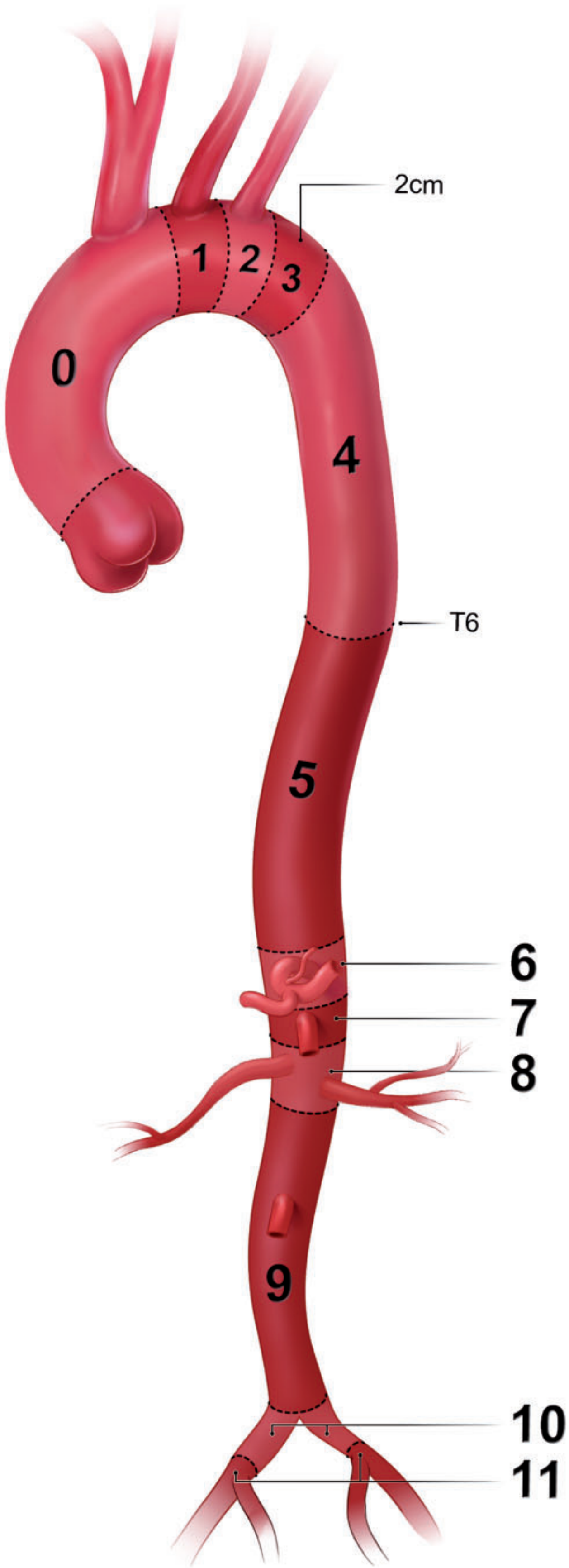


**Figure 1:** Definition of attachment zones, also known as Ishimaru zones. Printed with permission from © Campbell Medical Illustration.

## Additional examinations guiding how to proceed

The patient undergoes a full cardiovascular workup including a new electrocardiogram (ECG)-triggered CTA starting cranially including the circle of Willis and extending below to the level of the femoral heads. Furthermore, coronary angiography, transthoracic echocardiography (TTE) and duplex scanning of the supra-aortic branches are done.

## Constellations favouring open aortic arch replacement

The aortic arch aneurysm is accompanied by a proximal ascending aortic dilatation up to 47 mm in the mid ascending aorta and 55 mm at the level of the offspring of the brachiocephalic trunk (BCT). At the transition between zone 2 and zone 3, the diameter is 38 mm.

The morphology of the arch is a type III aortic arch, and the distance between the sternal notch and the left subclavian artery (LSA) is 5 cm, which should enable sufficient exposure of the entire aortic arch pathology via a median sternotomy. The maximum diameter of the aortic root is 38 mm. There is moderate to heavily calcified aortic valve stenosis and a 90% complex stenosis of the left anterior descending coronary artery (LAD). Biventricular function is normal and the remaining valvular status shows morphologically and functionally regular findings. The supra-aortic vessels are free from haemodynamically significant lesions.

***Treatment plan.*** It is recommended that the patient undergoes an open ascending and aortic arch replacement and a left internal mammary artery bypass (LIMA) to the LAD. Finally, the aortic valve is replaced with a biological prosthesis. The aortic arch is replaced with a branched graft to eliminate the largest possible amount of native diseased aortic arch tissue and to reduce the risk of any kind of suture aneurysm caused by a trade-off in leaving native aortic arch tissue in place.

## Constellations favouring open arch replacement using the frozen elephant trunk technique

CTA shows that zone 3 is still diseased with a maximum diameter of 55 mm, and regular diameters are reached again in proximal zone 4. The aortic root diameter is 37 mm; the mid ascending diameter is 45 mm. The valvular status is regular and there is no coronary heart disease.

***Treatment plan.*** It is recommended that the patient undergoes total aortic arch replacement using the frozen elephant trunk (FET) technique including supracoronary replacement of the ascending aorta. The descending anastomosis of the FET prosthesis cuff (at the transition between the surgical prosthesis and the stent graft portion) can be performed proximally in the aortic arch in zone 2 to reduce the risk of palsy of the left laryngeal nerve and to have better exposure due to the gained proximity, which also eases haemostasis. The stent graft portion of the FET prosthesis should be chosen to treat the entire pathological area in one step or, if any doubt arises regarding the elevated risk of the occurrence of symptomatic spinal cord injury, to treat a short segment with a secondary distal thoracic endovascular aortic repair (TEVAR) extension. Intraoperative aortoscopy during hypothermic circulatory arrest is not mandatory; however, it might be helpful to control the distal landing zone and the stent graft portion during the deployment process.

### Constellations favouring hybrid aortic arch replacement

The ascending aortic diameter is 37 mm and the aortic arch aneurysm starts in zone 2 and extends to the transition of zone 4 to zone 5. Revascularization of both the left common carotid artery (LCCA) and the LSA would provide a 2.5-cm long proximal landing zone.

***Treatment plan.*** It is recommended that the patient undergoes a double transposition of the supra-aortic vessels via an upper median hemisternotomy. This operation includes transposition of the LCCA into the BCT followed by transposition of the LSA into the already transposed LCCA. If there is an isolated offspring of the left vertebral artery (LVA) from the aortic arch, it can be re-implanted to the LCCA or LSA with or without (depending on the local anatomical situation) interposition of the great saphenous vein. Due to the extent to the transition of zone 4 to zone 5, a cerebrospinal fluid (CSF) drain is inserted prior to the procedure to protect the spinal cord. Potentially, an extrathoracic right common carotid-to-LCCA-to-LSA artery bypass could be done, but this type of extra-anatomic rerouting is more prone to occlusion and to erosion into adjacent structures such as the oesophagus and the trachea. Finally, a tracheostomy, if needed, is challenging.

### Constellations favouring branched endovascular aortic arch repair

The patient has regular ascending aortic dimensions without any signs of atherosclerosis. The pathology ends at the transition between zone 3 and zone 4. CTA shows that the distance between the BCT and the LSA is short, and a double transposition might provide a proximal landing zone of only 1.5 cm in zone 1. There is no coronary heart disease and the valvular status is normal; however, the left ventricular ejection fraction is severely reduced to 25%. The anatomy of the aortic arch, including the diameter of the supra-aortic vessel, is suitable for implantation of a double-branched endograft; distal access vessels are suitable for retrograde delivery of the main portion of the stent graft and proximal access vessels are suitable for bridging stent grafts into the BCT and into the LCCA. Because commercially available prostheses do have 2 branches, the LSA needs revascularization either in advance or, preferably to avoid a second opening of the incision, simultaneously.

***Treatment plan.*** It is recommended that the patient undergoes a simultaneous LSA-to-LCCA bypass and implantation of a double-branched endograft by retrograde delivery of the main body via the femoral artery and by delivery of the supra-aortic bridging grafts into the BCT and the LCCA via surgical cutdowns over both common carotid arteries.

Because the distal landing zone of the stent graft component is at the transition between zone 3 and zone 4, CSF drainage is not deemed necessary for additional spinal cord protection.

## CASE 2: DESCENDING AORTIC ANEURYSM INVOLVING THE DISTAL AORTIC ARCH

### Clinical scenario

A 77-year-old man presents with a 65-mm thoracic aortic aneurysm starting immediately distal to the origin of the LSA in zone 3. Five centimetres cranial to the coeliac trunk (CT) in zone 5, the diameter becomes regular again. The diagnosis was made by chance because the patient had a chest radiograph prior to a urological procedure.

### Additional examinations guiding how to proceed

The patient undergoes a full cardiovascular workup including a new ECG-triggered CTA starting cranially, including the circle of Willis and extending below to the level of the femoral heads. Furthermore, coronary angiography, TTE and duplex scanning of the supra-aortic branches are done.

### Constellations favouring a limited combined vascular and endovascular procedure

Coronary angiography reveals regular findings. TTE shows good biventricular function, morphological and functional regular valvular status, and supra-aortic vessels that are free from haemodynamically relevant lesions. The proximal landing zone without any kind of transposition is 8 mm from the distal end of the LSA offspring to the beginning of the lesion measured on the inner curvature of the aortic arch. The diameters of the ascending aorta and the aortic arch at the level of the intended proximal landing zone are 3.8 cm and 3.6 cm, respectively. The LSA-to-LCCA bypass/transposition would provide a proximal landing zone of 2.5 cm. The access vessels for retrograde delivery are of sufficient size and morphology.

***Treatment plan.*** It is recommended that the patient undergoes an LSA-to-LCCA bypass/transposition with simultaneous TEVAR. A CSF drain is inserted prior to the procedure to protect the spinal cord.

### Constellations favouring a more extensive combined vascular and endovascular procedure

Coronary angiography reveals regular findings. TTE shows a good biventricular function and a morphological and functional regular valvular status, and the supra-aortic vessels are free from haemodynamically relevant lesions. Diameters of the ascending aorta and the aortic arch at the level of the intended proximal landing zone are 3.8 cm and 3.6 cm, respectively. The proximal landing zone without any kind of transposition is 8 mm from the distal end of the LSA offspring to the beginning of the lesion measured on the inner curvature of the aortic arch, but the LSA-to-LCCA transposition would merely extend the proximal landing zone to 14 mm. Additionally, the patient has an isolated offspring of the LVA from the aortic arch. A more extensive form of transposition (double transposition—in this case, actually triple transposition due to the isolated LVA offspring) would extend the proximal landing zone to 2.7 cm. The access vessels for retrograde delivery have sufficient size and morphology.

***Treatment plan.*** It is recommended that the patient undergoes a double transposition (LCCA to BCT and LSA to LCCA transposition with additional transposition of the LVA) with a simultaneous TEVAR from zone 1 to zone 5. A CSF drain is inserted prior to the procedure to protect the spinal cord.

## Constellations favouring a total endovascular approach

Coronary angiography reveals 2-vessel disease with lesions that are good targets for percutaneous coronary intervention (PCI). TTE shows severely reduced left ventricular function, the valvular status is morphologically and functionally regular and the supra-aortic vessels are free from haemodynamically relevant lesions. The proximal landing zone without any kind of transposition is 8 mm from the distal end of the LSA offspring to the beginning of the lesion measured on the inner curvature of the aortic arch. The diameters of the ascending aorta and the aortic arch at the level of the intended proximal landing zone are 3.8 cm and 3.6 cm, respectively. Neither LSA to LCCA nor double transposition would create a sufficient proximal landing zone, but due to the regular ascending aortic diameters, total endovascular aortic arch repair (together with LSA revascularization) is possible. Access vessels for retrograde delivery do have a sufficient size and morphology.

***Treatment plan.*** Due to the new onset of symptoms (hoarseness due to left laryngeal nerve palsy), the pressure to accomplish treatment increases. The initial plan to perform PCI and to wait until it was time to start dual antiplatelet therapy is too risky (insertion of a CSF drain requires refraining from antiplatelet therapy to avoid bleeding complications during the insertion process). The patient undergoes simultaneous LSA-to-LCCA revascularization and branched endovascular aortic arch repair with distal TEVAR extension to exclude the entire lesion from zone 0 down to zone 5. On day 2, the CSF drain was removed; on day 7, a 2-vessel PCI was performed successfully.

## Constellations favouring open surgery using the frozen elephant trunk approach with secondary distal thoracic endovascular aortic repair extension

Coronary angiography reveals 3-vessel disease with lesions that are suboptimal targets for PCI. TTE shows a regular biventricular function but also haemodynamically significant aortic stenosis with a calcific distribution pattern that is suboptimal for transcatheter aortic valve implantation. The proximal landing zone without any kind of transposition is 8 mm from the distal end of the LSA offspring to the beginning of the lesion measured on the inner curvature of the aortic arch. Neither LSA to LCCA nor double transposition would create a sufficient proximal landing zone. The diameter of the ascending aorta is 4.5 cm, which obviates total aortic arch debranching because of the high risk of retrograde type A aortic dissection. Access vessels for secondary retrograde delivery do have sufficient size and morphology.

***Treatment plan.*** It is recommended that the patient undergoes triple coronary artery bypass grafting using the LIMA-LAD, saphenous vein grafts to the circumflex artery, and the right coronary artery. Total arterial revascularization is not preferred because of the potential need for vasopressors after total aortic arch replacement and the risk for vasospasm, biological aortic valve replacement and aortic arch replacement using the FET technique with a short stent graft component ending in proximal zone 4. After a recovery phase of 6 weeks, the patient undergoes TEVAR extension down to zone 5, having had a CSF fluid drain inserted in advance after temporary cessation of antiplatelet therapy.

## Constellations favouring open surgery through a left-sided thoracotomy using left heart bypass

The patient had prior coronary artery bypass grafting with an LIMA-LAD bypass. The aortic arch is not diseased and has a maximum diameter of 3 cm. The offspring of the LVA derives directly from the aortic arch. There are no remaining cardiovascular or pulmonary issues and the clinical condition is excellent.

***Treatment plan.*** It is recommended that the patient undergoes a left-sided muscle-sparing thoracotomy in the 4th intercostal space with single-lung ventilation. Motor-evoked potentials are monitored. A CSF drain is inserted before the procedure to protect the spinal cord. A left-heart bypass is installed (left lower pulmonary vein/left external iliac artery or descending thoracic aorta). The aortic arch is accessed through the pericardium and clamped between the LCCA and the LSA. The descending aorta is clamped below the aneurysm. The aneurysm is opened and the LSA is blocked with a soft Fogarty balloon. The patency of the circle of Willis is confirmed in advance to secure the potential for the intended steal for persistent antegrade flow into the LIMA during the clamping phase.

# CASE 3: REMAINING DISSECTION AFTER TYPE A REPAIR

## Clinical scenario

A 76-year-old male patient presents with a 67-mm proximal descending aortic aneurysm on the basis of a remaining dissection 5 years after type A repair. At that time, the patient received a supracoronary ascending aortic replacement with a 28-mm tube graft. At the index surgery, a hemiarch replacement was performed. The patient was lost to follow-up due to a change of residence and now had a chest radiograph due to planned orthopaedic surgery that showed a large mediastinal mass. Secondarily, a non-ECG-triggered thoracic CTA was performed that established the diagnosis. The mechanism for aneurysm formation is thought to be a large communication between the true and the false lumen in zone 2 that is functionally acting as a new primary entry tear enhancing continuing growth.

## Additional examinations guiding the decision of how to proceed

The patient undergoes a full cardiovascular workup including a new ECG-triggered CTA starting cranially, including the circle of Willis and extending below to the level of the femoral heads. Furthermore, we performed coronary angiography, TTE and duplex scanning of the supra-aortic branches.

## Constellations favouring open surgery using the frozen elephant trunk technique

CTA shows the remaining dissection of all 3 supra-aortic vessels. The descending aorta reaches a regular size at the transition between zones 4 and 5; the size of the true lumen is 32 × 20 mm; there are no communications between the lumina at the thoracic level; the CT, the superior mesenteric artery (SMA) and the right renal artery (RRA) originate from the true lumen; and the left renal artery (LRA) originates from the false lumen with a communication between the lumina at that level. TTE shows good biventricular function but moderate aortic regurgitation with a vena contracta of 4 mm based on a remaining/recurring prolapse of the non/right coronary commissure. The remaining valvular status is morphologically and functionally normal. Coronary angiography and duplex scanning of the supra-aortic branches show regular findings.

***Treatment plan.*** It is recommended that the patient undergoes total aortic arch replacement using the FET technique using the short stent graft component for spinal cord protection extending into the transition between zones 3 and 4 with the potential to induce complete remodelling of the entire thoracic aorta simultaneously with the biological aortic valve replacement to fix the aortic regurgitation component. If a distal extension is needed, secondary TEVAR can be performed at a later stage on the basis of the completion CTA findings. The LRA originating from the false lumen is not at risk after thoracic false lumen thrombosis because the communication between both lumina will provide continuous blood supply across the membrane.

## Constellations favouring a classical elephant trunk implant

CTA shows the remaining dissection of all 3 supra-aortic vessels. The descending aorta reaches a regular size at the transition between zones 4 and 5. However, the true lumen shows a pseudocoarctation-like narrowing at the level of zone 4. The CT, the SMA and the RRA originate from the true lumen. The LRA originates from the false lumen with a communication between the lumina at that level.

***Treatment plan.*** A true lumen FET implant is highly likely to induce immediate lower body malperfusion. Therefore, a 2-step approach to fix the entire disease process is needed. It is recommended that the patient undergoes total aortic arch replacement using the classical elephant trunk technique with excision of the dissection membrane during lower body hypothermic circulatory arrest as far distally as possible to maintain regular distal perfusion and to be able to access the elephant trunk component in zone 4 at the second stage. After recovery, distal open surgical extension to zone 5 is done.

## Constellations favouring transposition of supra-aortic vessels and thoracic endovascular aortic repair

CTA shows no remaining dissection of the supra-aortic vessels, and the aortic arch is free from remaining dissection up to zone 2. The descending aorta reaches a regular size at the transition between zones 4 and 5; the size of the true lumen is 32 × 20 mm; and there are no communications between the lumina at the thoracic level. The CT, the SMA and the RRA originate from the true lumen. The LRA originates from the false lumen with a communication between the lumina at that level. An LSA-to-LCCA bypass/transposition would create a 2.5-cm landing zone for TEVAR. The CTA showed an interrupted circle of Willis with the rare constellation of a stand-alone LVA supplying the entire posterior circulation with no posterior communicant arteries.

***Treatment plan.*** It is recommended that the patient undergoes an LSA-to-LCCA bypass (because a transposition needs longer clamping times, thereby creating unnecessary risk in this stand-alone LVA constellation) with TEVAR extending into zone 5 where the aortic diameters are regular. The procedure is performed using a CSF drain to protect the spinal cord. The LRA originating from the false lumen is not at risk after thoracic false lumen thrombosis because the communication between both lumina will provide continuing blood supply across the membrane.

## Constellations favouring total endovascular aortic arch repair

CTA shows that the indwelling ascending aortic prosthesis has a sufficient length of 8 cm, and there is no remaining dissection of the supra-aortic branches. The LCCA has a diameter of 8 mm. The descending aorta reaches a regular size at the transition between zones 4 and 5; the size of the true lumen is 32 × 20 mm; there are no communications between the lumina at the thoracic level; the CT, the SMA and the RRA originate from the true lumen; and the LRA originates from the false lumen with a communication between lumina at that level. Regarding the proximal landing zone, neither the LSA-to-LCCA bypass/transposition or even a double transposition would gain a landing zone of sufficient length. The valvular status is regular. However, the left ventricular ejection fraction is severely reduced and the patient has poor pulmonary function.

***Treatment plan.*** A total endovascular approach is recommended due to an ideal proximal landing zone consisting of a long straight ascending graft and a sufficiently large LCCA for placement of the supra-aortic extension (vessels smaller than 7 mm are not recommend because the risk for graft occlusion is high). Simultaneous LSA-to-LCCA bypass grafting for maintaining posterior cerebellar circulation and for maintaining inflow to the spinal cord is performed. Additionally, distal TEVAR extension into zone 5 should be considered. The procedure is performed using a CSF drain to protect the spinal cord. The LRA originating from the false lumen is not at risk after thoracic false lumen thrombosis because the communication between both lumina will provide continuing blood supply across the membrane.

## CASE 4: TYPE B AORTIC DISSECTION

### Clinical scenario

A 58-year-old female patient with a chronic type B aortic dissection presents at the outpatient clinic for an expert opinion. For 6 years, the type B dissection has been asymptomatic under the best medical treatment. The patient reports no physical restrictions. The ECG-triggered CTA confirms the location of the primary entry tear immediately distal to the LSA offspring. The dissection extends downstream along the entire thoraco-abdominal aorta and ends in zone 10. The CT, the SMA and the RRA arise from the true lumen, and the LRA is perfused from both the true and the false lumens. All thoracic segmental arteries arise from the false lumen. The diameter of the proximal descending aorta is 58 mm and demonstrates an 8-mm increase within the last year.

### Additional examinations guiding the decision of how to proceed

The patient undergoes a full cardiovascular workup including a new ECG-triggered CTA starting cranially including the circle of Willis and extending below to the level of the femoral heads. Furthermore, coronary angiography, TTE and duplex scanning of the supra-aortic branches are done.

### Constellations favouring a combined vascular and endovascular approach

The true lumen, seen with CTA in zones 4 and 5, has not collapsed. Multiplanar reformation of the thoracic aorta from 1.5-mm CTA slices demonstrates the following:

- Diameter of the ascending aorta: 40 mm
- Diameter in zone 2: 35 mm
- Sealing zone between the distal rim of the left carotid artery orifice and the entry tear: 23 mm
- Diameter of the entire aorta in zones 4 and 5: 42 mm
- Maximal and minimal diameter of the true lumen in zones 4 and 5: 28 mm and 11 mm, respectively.

The diameter of the LCCA is 8 mm. In CTA and duplex scanning of the supra-aortic vessels, neither calcification nor stenosis is observed. In cerebral CTA, the circle of Willis is patent.

***Treatment plan.*** A 2-staged approach is recommended: First, an LSA-to-LCCA bypass followed by TEVAR starting in zone 2 extending into zone 5 is planned. Occlusion of the LSA origin proximal to the LVA will be performed during the first surgical procedure or endovascularly during the second procedure. Due to the perfusion of all intercostal arteries from the false lumen and the risk of spinal cord injury, stent graft deployment above level TH8 is planned. Extension of the treatment downstream will follow thereafter if remodelling is insufficient. Prior to TEVAR, the patient receives a CSF drain to protect the spine.

### Constellation favouring an open surgery frozen elephant trunk procedure

CTA scans show that the true lumen in zones 4 and 5 has not collapsed. The maximum diameter of the potential proximal landing zone in zone 2 is greater than 40 mm and the potential length is less than 20 mm. Another constellation could be a concomitant aneurysm of the ascending aorta extending more than 50 mm into the arch.

***Treatment plan.*** An FET is planned. CSF drainage is not routinely required but should be considered depending on the intended length of the TEVAR component of the FET prosthesis. The basic strategy should be closure of the primary entry tear and, according to the consecutive extent of remodelling, secondary TEVAR should extend into distal zone 5 if needed.

### Constellation favouring open distal arch and descending aortic repair

The true lumen seen in the CTA scans of zones 4 and 5 is collapsed. Two channels of false lumen are embracing the true lumen in the middle. The diameter of the aortic arch is 35 mm.

***Treatment plan.*** Open surgical repair of the distal arch and the descending aorta via a left posterolateral thoracotomy is considered. Monitoring of CSF pressure and somatosensory and motor evoked potentials will be planned intraoperatively. A left-heart or partial bypass will be used to avoid hypothermic circulatory arrest. The aortic arch is accessed for clamping from a lateral direction after mobilizing the vagal and left laryngeal nerve or through the pericardium and clamped between the LCCA and LSA. The aorta is replaced and thoracic segmental arteries are reimplanted selectively.

## CASE 5: NON-A-NON-B AORTIC DISSECTION

### Clinical scenario

A 65-year-old man is admitted to the emergency department because of acute, severe chest pain and compensated ischaemia of the left leg. The patient presents with uncontrolled hypertension. The ECG-triggered CTA shows a non-A-non-B aortic dissection extending into the aortic arch with collapse of the descending aortic true lumen. The patient presents a normal neurological status.

### Additional findings guiding the decision of how to proceed

The patient undergoes a full cardiovascular workup, including a new ECG-triggered CTA starting cranially, including the circle of Willis and extending below to the level of the femoral heads. Furthermore, coronary angiography, TTE and duplex scanning of the supra-aortic branches are done.

## Constellations favouring open surgery using the frozen elephant trunk technique

CTA shows entry in the aortic arch at the outer curvature between the left common carotid and the LSA. Dissection extends in the arch up to the distal edge of the BCT and into the LCCA. The maximum diameter of the ascending aorta is 47 mm. In the TTE scans, moderate aortic valve regurgitation is observed. The remaining valvular status is normal. The biventricular function is good.

***Treatment plan.*** It is recommended that the patient undergoes total aortic arch replacement using the FET technique including replacement of the ascending aorta and of the biological aortic valve. The anastomosis of the FET prosthesis cuff (at the transition between the surgical prosthesis and the stent graft portion) can be performed in zone 2 to reduce the risk of recurrent nerve injury. The stent graft portion of the FET prosthesis should be short (the intention is to close the primary entry tear) to reduce to a minimum the remaining risk of symptomatic spinal cord injury. Because the entry is close to the LSA, the short stent graft portion will still allow closure of the primary entry tear, effective redirection of blood flow into the true lumen and true lumen expansion, probably eliminating lower extremity malperfusion and, in the mid-term follow-up, remodelling of the thoracic aorta.

## Constellations favouring transposition of supra-aortic vessels and thoracic endovascular aortic repair

CTA shows no dissection of the supra-aortic vessels, and the entry is 2 cm distally to the LSA. There is a retrograde intramural haematoma extending into the arch up to zone 2. The ascending aortic diameter is 38 mm, and on the TTE scan, the aortic valve is morphologically and functionally normal. The LSA-to-LCCA bypass/transposition would provide a 3.5-cm TEVAR landing zone free of dissection entry.

***Treatment plan.*** LSA to LCCA bypass/transposition and a zone 2 TEVAR are recommended to the patient. Because the ascending aorta is normal, the risk of retrograde dissection is low. TEVAR, using a 3.5-cm free of the entry landing zone, will redirect the blood flow into the true lumen, close the primary entry tear and probably eliminate lower extremity malperfusion.

# CASE 6: AORTIC ARCH INTRAMURAL HAEMATOMA

## Clinical scenario

A 72-year-old woman with a history of hypertension, smoking and diabetes presents with acute onset piercing chest pain. Upon examination, she has high blood pressure (210/100 mmHg). Emergency CTA reveals an intramural haematoma extending from zone 2 to zone 5. There is no visible intimal disruption or penetrating aortic ulcer-like projection. The maximum diameter of the intramural haematoma is 4.5 cm and that of the proximal descending aorta, 3.0 cm.

## Additional findings guiding how to proceed

Strict blood pressure and pain management was initiated immediately, which quickly rendered the patient symptomless. After 7 days, the patient was discharged home after having had a completion CTA with stable findings and was scheduled for a follow-up visit 3 months later.

***Treatment plan.*** Best medical treatment.

## Outpatient examination after 3 months

At the 3-month follow-up visit, the patient reports an uneventful clinical course. CTA shows a maximum diameter of the distal aortic arch at the transition of zone 2 to zone 3 of 5.5 cm with a 1.2-cm haematoma component. In addition, the primary entry tear has demasked in zone 2, immediately opposite the LSA offspring. The descending thoracic aorta remains unchanged. Formally, rapid expansion has occurred and the diameter threshold of 5.5 cm has been reached.

## Additional findings guiding the decision of how to proceed

The patient undergoes a full cardiovascular workup including a new ECG-triggered CTA starting cranially, including the circle of Willis and extending below to the level of the femoral heads. Furthermore, coronary angiography, TTE and duplex scanning of the supra-aortic branches are done.

## Constellations favouring a combined vascular and endovascular procedure

Despite the extension of the intramural haematoma into zone 2, an LSA-to-LCCA bypass creates a 2.5-cm extension from the primary entry tear to the distal LCCA offspring, thereby creating a proximal landing zone sufficient for TEVAR. The diameter of the ascending aorta is 36 mm and that of the aortic arch at the level of the proximal landing zone is 34 mm. The diameter of the access vessel for retrograde delivery is 8 mm and therefore adequate. Biventricular function is good; the valvular status is morphologically and functionally normal; the coronary angiogram shows regular findings, and the supra-aortic vessels are free from haemodynamically relevant lesions.

***Treatment plan.*** An LSA-to-LCCA bypass/transposition and TEVAR extending from zone 2 to zone 4 are performed, whereby the primary entry tear is fully closed. A CSF drain is inserted prior to the procedure to protect the spinal cord.

## Constellations favouring open surgery with primary use of the frozen elephant trunk technique

CTA shows a maximum diameter of the distal aortic arch at the transition from zone 2 to zone 3 of 5.5 cm; additionally, the primary entry tear is demasked in zone 2 immediately opposite the LSA offspring. The maximum diameter of the descending aorta is now 5.0 cm, reaching regular diameters in zone 5. From the now-visible primary entry tear downstream, a classical type B dissection component has developed extending into zone 10. TTE confirms normal valve function and a good biventricular function. Coronary angiography shows a significant stenosis of the right coronary artery, and duplex scanning of the supra-aortic branches shows regular findings. Additionally, the ascending aorta has a maximum diameter of 45 mm, and transposition of all 3 arch vessels would be needed to create a sufficient proximal landing zone for TEVAR.

***Treatment plan.*** The patient is scheduled for coronary artery bypass surgery and an FET procedure using the short stent graft component as the primary entry tear is thereby covered. No CSF drain is inserted because, with the strategy chosen, the remaining risk of symptomatic spinal cord injury is deemed low. The option for a secondary TEVAR extension remains if aortic remodelling turns out to be insufficient. Total aortic arch rerouting is not favoured because the combination of total aortic arch rerouting and TEVAR in patients with acute aortic syndromes (in addition to a large ascending aortic diameter) has a substantial risk for retrograde type A aortic dissection.

## CASE 7: PENETRATING ATHEROSCLEROTIC ULCER OF THE AORTIC ARCH

### Clinical scenario

A 62-year-old man with prostate carcinoma undergoes CTA for carcinoma staging. CTA shows a large penetrating atherosclerotic ulcer (PAU) with diameters of 25–30 mm in the distal aortic arch. The patient is asymptomatic and has no history of previous aortic repair. There are no metastases of the prostate carcinoma.

### Additional examinations guiding the decision of how to proceed

The patient undergoes a full cardiovascular workup including a new ECG-triggered CTA, starting cranially including the circle of Willis and extending below to the level of the femoral heads. Furthermore, coronary angiography, TTE and duplex scanning of the supra-aortic branches are done.

### Constellations favouring transposition of supra-aortic vessels and thoracic endovascular aortic repair

CTA shows PAU in the distal aortic arch at the lesser curvature at the level of the LSA at the transition from zone 2 to zone 3. The maximum diameters of the ascending aorta, the aortic arch and the descending aorta are 38, 35 and 32 mm, respectively. In TTE scans, the valvular status is normal and in the coronary angiographic images, there is no significant coronary heart disease.

***Treatment plan.*** Since revascularization of the LSA will provide a sufficient proximal landing zone of 28 mm, both LSA-to-LCCA bypass/transposition and zone 2 TEVAR are recommended. In the case of a too-short distance between the LSA and the LCCA, a double transposition and zone 1 TEVAR might be recommended. To minimize the risk of symptomatic spinal cord injury, a short stent graft (10–15 cm) is recommended. Because the distal landing zone is in proximal zone 4, CSF drainage might be omitted if there are no large thoracic segmental artery offspring at that level.

### Constellations favouring branched arch endograft implantation

CTA shows that the distance between the BCT and the LSA is short and that double transposition might provide a proximal landing zone of only 1.3 cm. The dimensions of the ascending aorta are regular, showing a 37-mm maximum diameter. There is a 50% stenosis of the LAD, and the valvular status and left ventricular ejection fraction are normal. The patient has been taking cortisone for 18 years for severe polyarthritis, and the healing process after any skin incision is challenging. The anatomy of the aortic arch including the supra-aortic vessels is suitable for implantation of a double-branched endograft. Access vessels show regular diameters for retrograde stent graft delivery.

***Treatment plan.*** It is recommended that the patient has simultaneous LSA-to-LCCA bypass and implantation of a double-branched endograft by retrograde delivery of the main body via the femoral artery and by delivery of the supra-aortic bridging grafts into the BCT and the LCCA via surgical cutdowns over both common carotid arteries.

### Constellations favouring open surgery using the frozen elephant trunk technique

CTA shows that the maximal diameter of the ascending aorta is 49 mm and that the patient has an aortic valve with moderate stenosis and moderate insufficiency. The left ventricle is dilated. Furthermore, there is severe coronary heart disease with significant stenosis in the left and right coronary systems. The PAU is situated at the level of the LSA at the transition from zone 2 to zone 3.

***Treatment plan.*** It is recommended that the patient has total aortic arch replacement using the FET technique including replacement of the ascending aorta, replacement with a biological aortic valve and revascularization of the coronary arteries. The anastomosis of the FET prosthesis cuff (at the transition between the surgical prosthesis and the stent graft portion) can be performed proximally to the aortic arch PAU in zone 1 or 2 to ensure the circular aortic wall for the cuff-aortic wall anastomosis and to reduce the risk of recurrent nerve injury. The length of the stent graft portion of the FET prosthesis should be short in order reduce the remaining risk of spinal cord injury to a minimum.

## CASE 8: GRAFT INFECTION INVOLVING THE AORTIC ARCH

### Clinical scenario

A 68-year-old man presents with fever and elevated inflammatory parameters 4 months after aortic root replacement with a biological valve-carrying conduit and after simultaneous ascending and hemiarch replacement due to aneurysmal formation. Due to diffuse bleeding during the index procedure, the sternum remained open, and chest sponge tamponade was performed for 48 h. Afterwards, the sternum was closed and the remaining clinical course was uneventful. Standard perioperative antibiotic therapy was applied using the institutional protocol.

## Additional examinations guiding how to proceed

The patient undergoes a full cardiovascular workup including a new ECG-triggered CTA starting cranially including the circle of Willis and extending below to the level of the femoral heads. Compared to the completion scan at discharge, air can be seen surrounding the prosthesis. At the level of the hemiarch anastomosis, a pseudoaneurysm can be seen due to partial leakage of the distal anastomosis. The neurocranium is free from detectable embolic events. A TTE shows vegetations on the biological valve. Blood cultures show sepsis due to *Staphylococcus aureus*. Therefore, and also because the coronary angiogram shows regular findings at the index procedure, repetition was omitted.

## Constellations favouring a conservative approach

None, if the clinical conditions are good.

***Treatment plan.*** It is recommended that the patient undergoes redo surgery with complete explantation of the alloplastic material; radical debridement and orthotopic reconstruction using a neoaortic root formed from a bovine xenopericardial tube and a biological prosthesis; and ascending and hemiarch replacement using an additional neoaortic xenopericardial tube graft. Alternatively, a homograft for the aortic root can be used. Also, re-alloplastic reconstruction with the option of additional omental flap plasty can be considered, but the remaining risk of reinfection then seems to be higher. In addition, life-long antibiotic therapy is warranted.

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