Surgical Management of TAAA and AAA in Marfan's and Loeys-Dietz Syndrome

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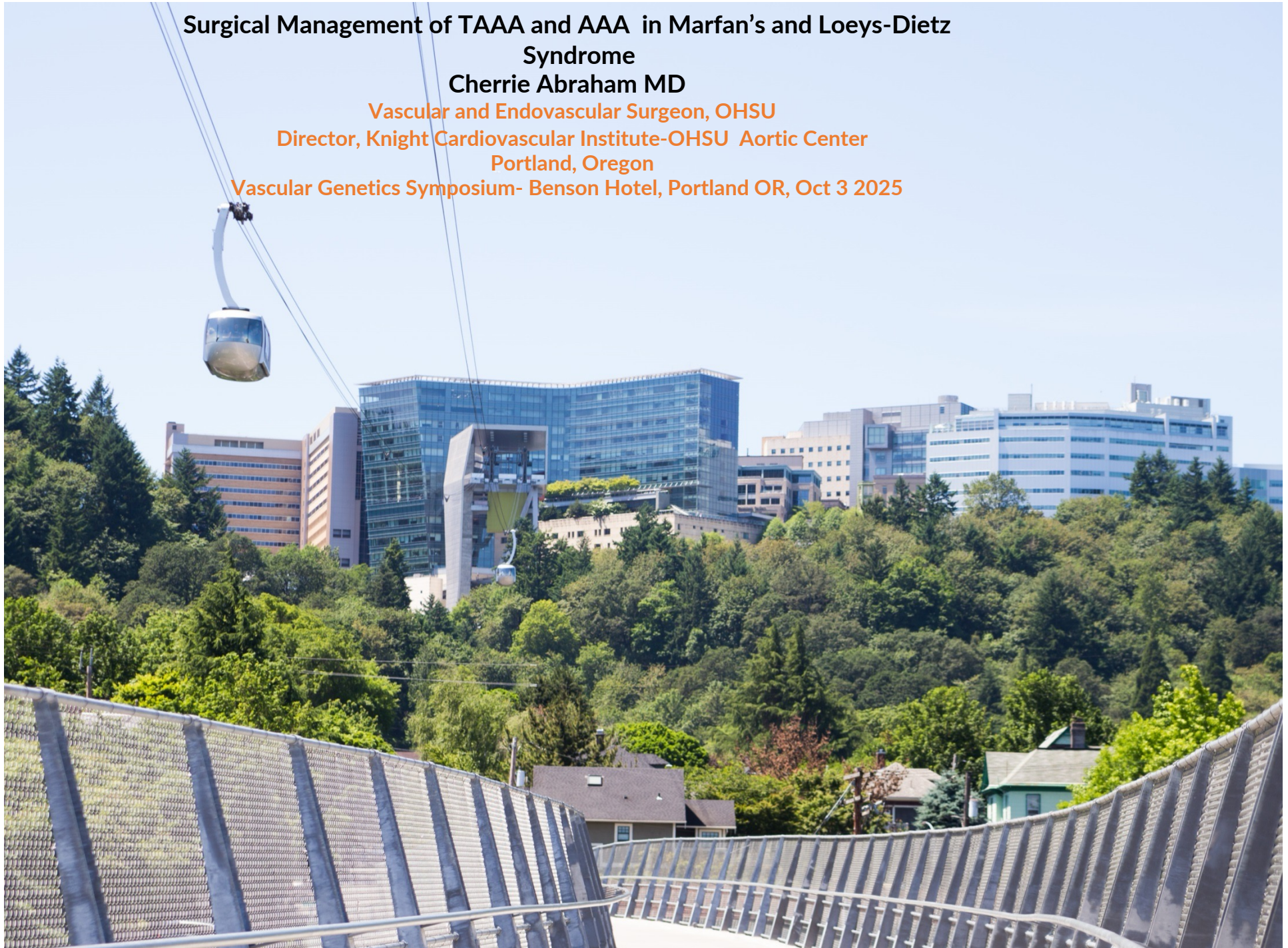


OHSU

Disclosures

- Educational and Research Grant Support – WL Gore, Cook Medical
- Clinical Trial Adjudicator- GORE Conformable Clinical Trial

Gold Standard- Open Repair

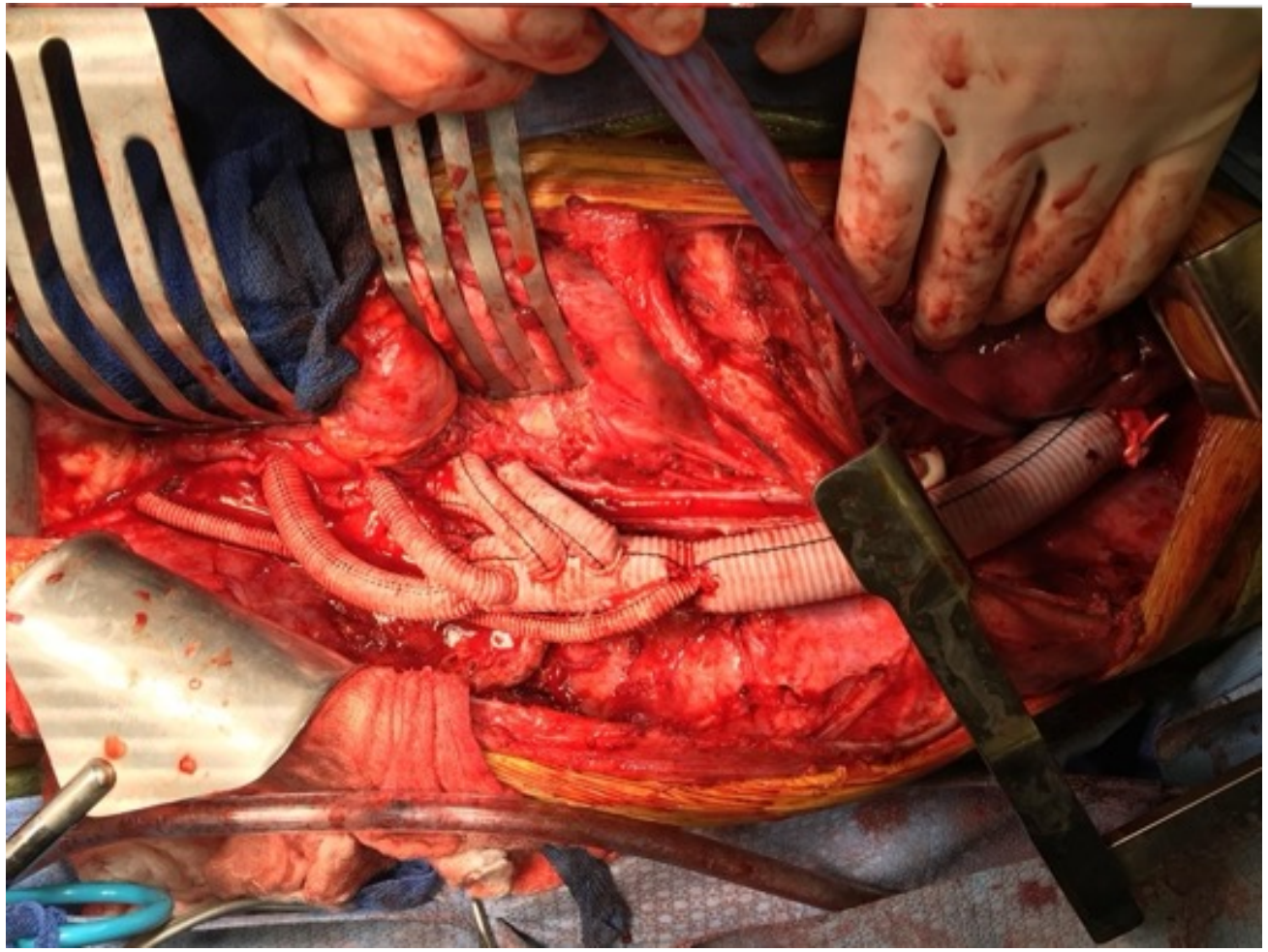
Marfan’s

Loeys-Dietz

Vascular Ehlers Danlos

Familial Aortopathy

Young patients with Aortic Pathology



Endografts in Genetic Aortpathy

- Rupture
- Aortoesophageal fistula
- Redo Thoracic or Redo Thoracoabdominal
- Co-morbidities precluding open surgery
- FET as a set up for distal open TAAA repair
- Proximal and Distal Landing Zones in Synthetic

Reintervention after TEVAR in Marfan's

- Le Huu et al 2022 35% at 3.6 years, 27 pts
- Kolbel et al 2021 17% at 42 months, 24 pts
- Eleshra et al 2021 19% at 40 months, 24 pts
- Jiang et al 2021 35% at 49 months, 26 pts
- Pellnec et al 2020 55% at 24 months, 18 pts

Steinmetz and Coselli, Annals of Surgery 2022

2024 Guidelines EACTS/STS

- LDS with TGFBR2 mutation, surgery to replace the intact aortic arch, descending aorta, or abdominal aorta at a diameter of >= 4.5 cm
- diameter of >= 4.0 cm in the presence of high-risk features, such as women with TGFBR2, small body size, severe extra aortic features, and family history of aortic dissection

Hopkins Experience- Sorber et al.

- J Vasc Surg . 2022 January ; 75(1): 90–98- **Connective tissue disease type mediates branch patency of grafts in open thoracoabdominal aortic reconstruction**

- 172 patients undergoing open thoracoabdominal replacement with a prosthetic branched graft for aortic aneurysm and/or dissection from 2006 to 2020 were included.
- All surgeries were performed by a single surgeon (J.H.B.), with standard approach for replacement of the renovisceral aorta is retroperitoneal with a four-branched Coselli-type graft

- Patients were classified into specific CTD subtypes based on the presence of genotypic documentation, including MFS (FBN1 mutation), LDS (TGFBR1 (2),TGFBR2 (2), SMAD3 (1)), or other CTD (includes vascular Ehlers Danlos syndrome (vEDS) (COL3A1, (1, haploinsufficient)), familial thoracic aortic aneurysm and dissection (FTAAD) (MYH11 (1), ACTA2 (1) , etc).
- Any patients carrying a diagnosis of CTD based on clinical features or family history but lacking a documented genotypic diagnosis were classified as other CTD.
- All patients without a diagnosis of CTD were classified as degenerative nonsyndromic
- Aneurysms, denoted non-CTD. The extent of thoracoabdominal aortic aneurysms was classified according to Crawford extent (I-V).

- 72 patients were included, of whom 45 (26.2%) had a CTD . The CTD cohort was predominantly made up of patients with MFS (32; 71.1%), with significantly, fewer LDS (n = 5), vEDS (n = 1), FTAAD (n = 2), and unspecified CTD (n = 5).
- The non-CTD (degenerative aneurysm) patients had a significantly higher mean age than the CTD pts - Patients with CTD (67.0 vs 44.9 years; P < .0001); among the CTD subtypes, there were no significant differences in age.

- The non-CTD patients had a significantly higher comorbidity burden, with higher rates of hypertension (92.1% vs 46.7%), chronic obstructive pulmonary disease (22.8% vs 4.4%), coronary artery disease (26.8% vs 4.4%), and positive smoking
- Patients with CTDs had a higher rate of preoperative systemic anticoagulation (40% vs 4.7%), beta-blockade (97.8% vs 66.9%), and ARB use (51.1% vs 19.7%), whereas non-CTD patients had higher rates of ACEis (33.9% vs 4.4%) and calcium channel blocker use (40.9% vs 13.3%; all $P < .01$).
- Patients with CTDs were more likely have had a prior aortic dissection (82.7% vs 7.3%), any prior cardiovascular surgery (60% vs 36.2%), and, specifically, an aortic root procedure (55.6% vs 5.5%; all $P < .001$). This was particularly significant for patients with MFS, 71.9% of whom had a previous aortic root replacement.

- There was no difference between patients with CTDs and non-CTD patients in terms of level of urgency, with equivalent proportions of each group requiring emergent intervention (17.3% vs 15.6%; P = .786)
- Patients with CTD had significantly more extensive aneurysms when compared with non-CTD patients (57.8% vs 26.7% extent II or III aneurysms; P < .001); most non-CTD patients (60.6%) had extent IV aneurysms.
- Patients with CTDs had significantly more reconstructed branches (3.5 vs 1.8; P < .0001), with a higher proportion of superior mesenteric artery (29.9% vs 86.7%), celiac (30.7% vs 80%), and right renal branches (30.7% vs 80.0%; all P < .0001).
- There was no significant difference in reconstruction branches or anatomical targets among the CTD subtypes.

- In terms of postoperative complications, there were no differences in in-hospital mortality or in-hospital overall complication rates between patients with and without CTDs.
- The in-hospital mortality rate was 7.6% in the perioperative period for all patients in the series (11.1% CTD vs 6.3% non-CTD; P = .294).

- There were no differences in subsequent dissection formation between the two groups; however, there was a significantly higher rate of new iliac aneurysm formation postoperatively in the CTD group (15.6% vs 3.1%; P = .003), which was demonstrated to be driven by the MFS and LDS groups
- Subgroup analysis - A significantly higher number of patients in the MFS group experienced aneurysm formation contiguous with their prior TAAA repair (18.8%; P = .016).
- There was no difference in late mortality between any of the groups.

- Overall branch patency was not different between the groups at 1 month or 6 month time points (Fig 1, B); however, at 1 year, the MFS group demonstrated significantly higher overall branch patency compared with the non-CTD group (98.8% vs 92.1%), a difference which remained significant at 5 years (98.8% vs 88.9%; both $P < .05$); this difference was not significant for the LDS (93.3%) or other CTD groups (89.9%), which demonstrated similar overall patency to the non-CTD group.

- Overall, these data recapitulate that patients with CTD as a whole are a younger population with fewer comorbidities and low rates of smoking. They are more likely to be on an ARB or beta blocker than ACEis or calcium channel blockers, which reflects the known therapeutic benefits of ARBs and beta blockade in MFS and LDS.15
- There is a high rate of systemic anticoagulation in the CTD group, which is likely coupled with the high rate of prior aortic root surgery and possibility for mechanical valve placement in the MFS and LDS groups.

Frankel et al. 2020- GenTAC Data

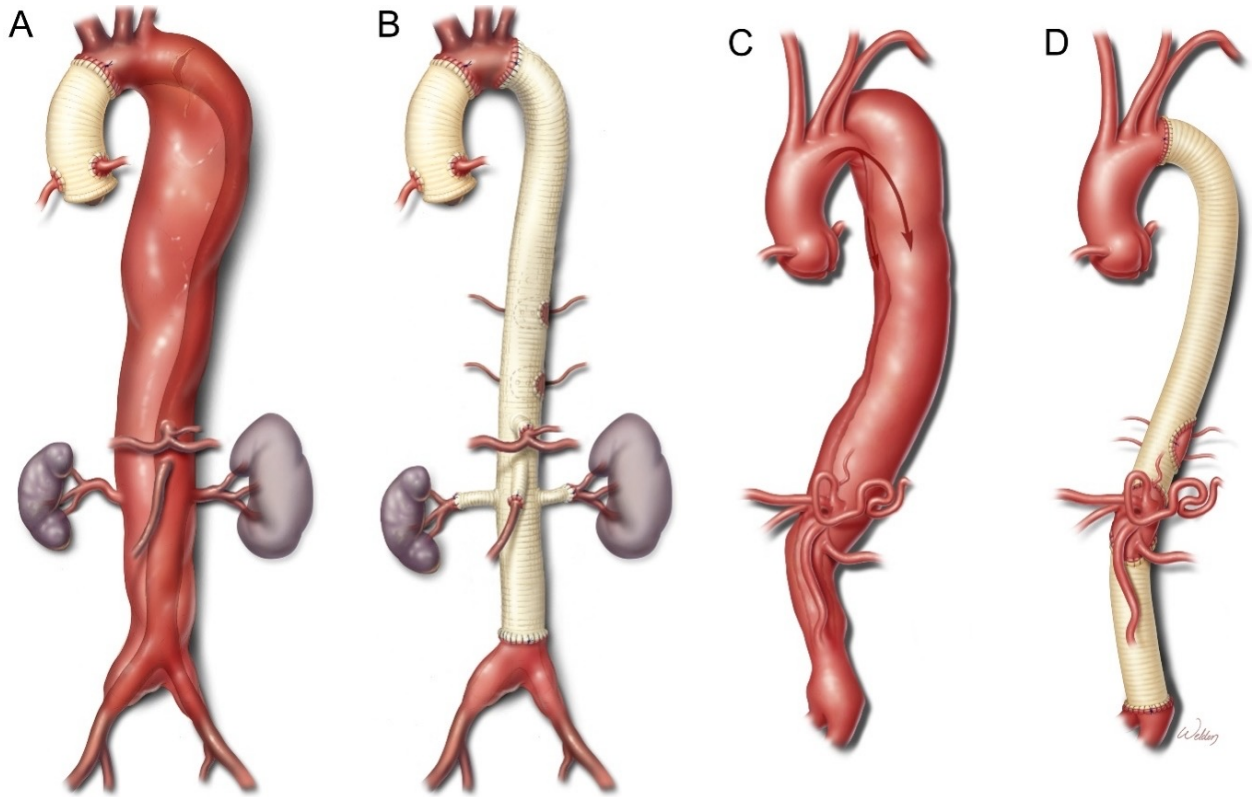
- Ann Thorac Surg 2020 May;109(5):1378-1384 , William C Frankel [1], Howard K Song [2], Rita K Milewski [3], Sherene Shalhub [4], Norma L Pugh [5], Kim A Eagle [6], Mary J Roman [7], Reed E Pyeritz [8], Cheryl L Maslen [9], William J Ravekes [10], Dianna M Milewicz [11], Joseph S Coselli [12], Scott A LeMaire [13]; GenTAC Investigators
- . 2020 May;109(5):1378-1384.
- 3699 patients enrolled at 8 participating centers in the Genetically Triggered Thoracic Aortic Aneurysms and Cardiovascular Conditions (GenTAC) Registry, and identified 155 open TAAA repairs in 142 unique patients.

- The primary diagnoses included Marfan syndrome (n = 76; 54%), familial thoracic aortic aneurysm and dissections (n = 31; 22%), and Loeys-Dietz syndrome (n = 10; 7%).
- Most repairs were performed for aneurysms associated with aortic dissection (n = 110; 71%). The most common repairs involved the entire descending thoracic aorta with distal extension (21% Crawford extent I and 36% extent II).
- The operative mortality rate was 1.3%. Other complications included paraplegia (4%), acute renal failure (5%), and vocal cord paralysis (21%). Reoperation after TAAA repair was required in a subset of cases for early bleeding (n = 15; 10%) and late repair failure (n = 7; 5%).

- Operative Details
- The mean age at the time of surgery was 42 years, and the mean maximum TAAA diameter was 6.0 +/- 1.6 cm (Table 3). Of the 110 repairs performed in patients with aortic dissection, only 7 (6%) were done during the acute phase.
- Eight operations were performed for failure of previous repair, including 6 for endoleaks after endo- vascular procedures. The most common repairs involved the entire descending thoracic aorta with extension into the suprarenal abdominal segment (extent I, 21%) or the entire abdominal aorta (extent II, 36%)
- TAAA repair technique varied, and several adjuncts were used within the study population. Of note, 41% of cases involved left heart bypass, 23% involved cardiopulmonary bypass, and 16% involved hypothermic circulatory arrest. Cerebrospinal fluid drainage was used in 56% of cases, cold renal perfusion in 45% of cases, selective visceral perfusion in 25% of cases, and evoked potential monitoring in 10% of cases. Bypass grafts were commonly placed to the visceral and renal arteries; branched grafts were used for this purpose in 10% of cases. As expected, the use of various adjuncts differed substantially based on the extent of repair.

- As expected, the incidence of complications was highest after the most extensive repairs (extent II). Early reoperation was required in 15 patients (10%) because of bleeding.

- Late Outcomes
- Seven patients (5%) necessitated late reoperation to treat repair failures (5%), including 3 patients who developed patch aneurysm, 2 who developed pseudoaneurysm, and 1 who developed acute bleeding at the proximal anastomosis (the mode of failure was unclear in 1 patient).
- There were 24 late deaths recorded during the study period (2006 to 2016). These occurred after a median of 1.2 years (range, 38 days to 11.5 years) after TAAA repair.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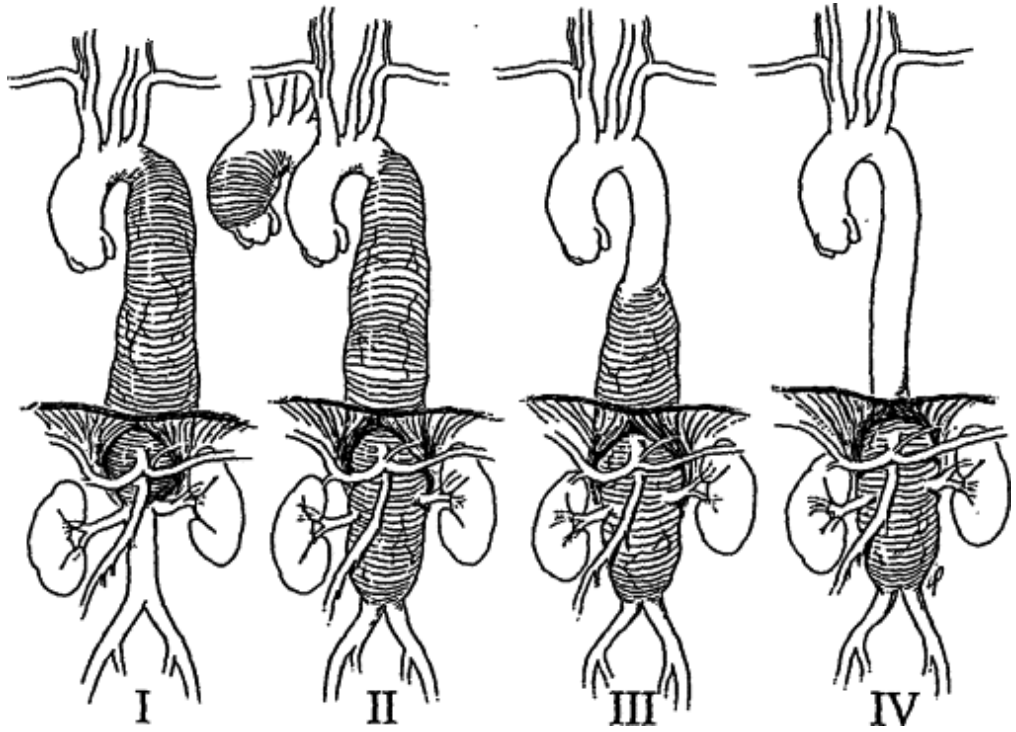
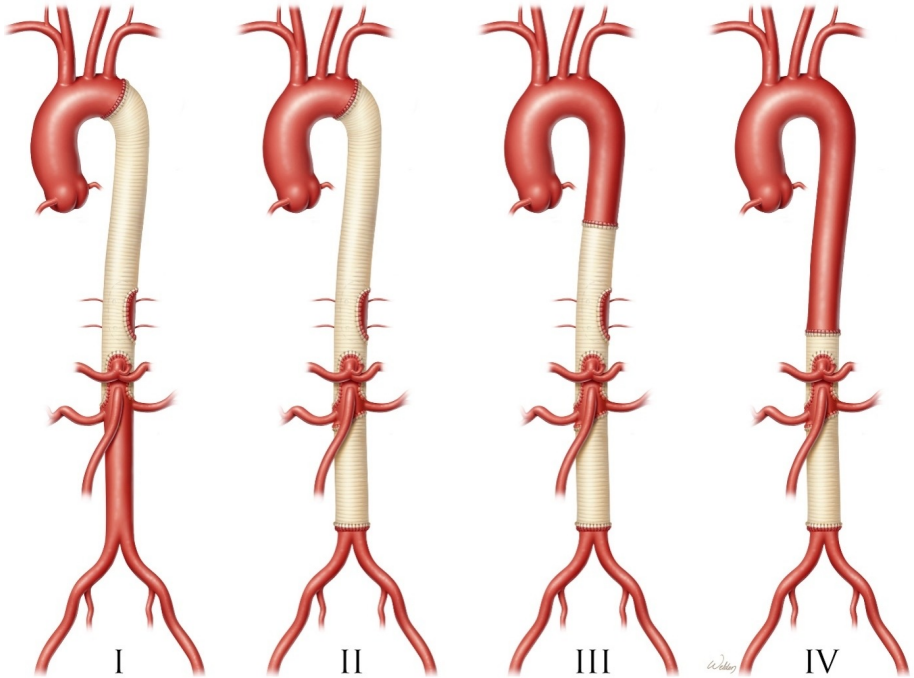
C
D


I
II
III
IV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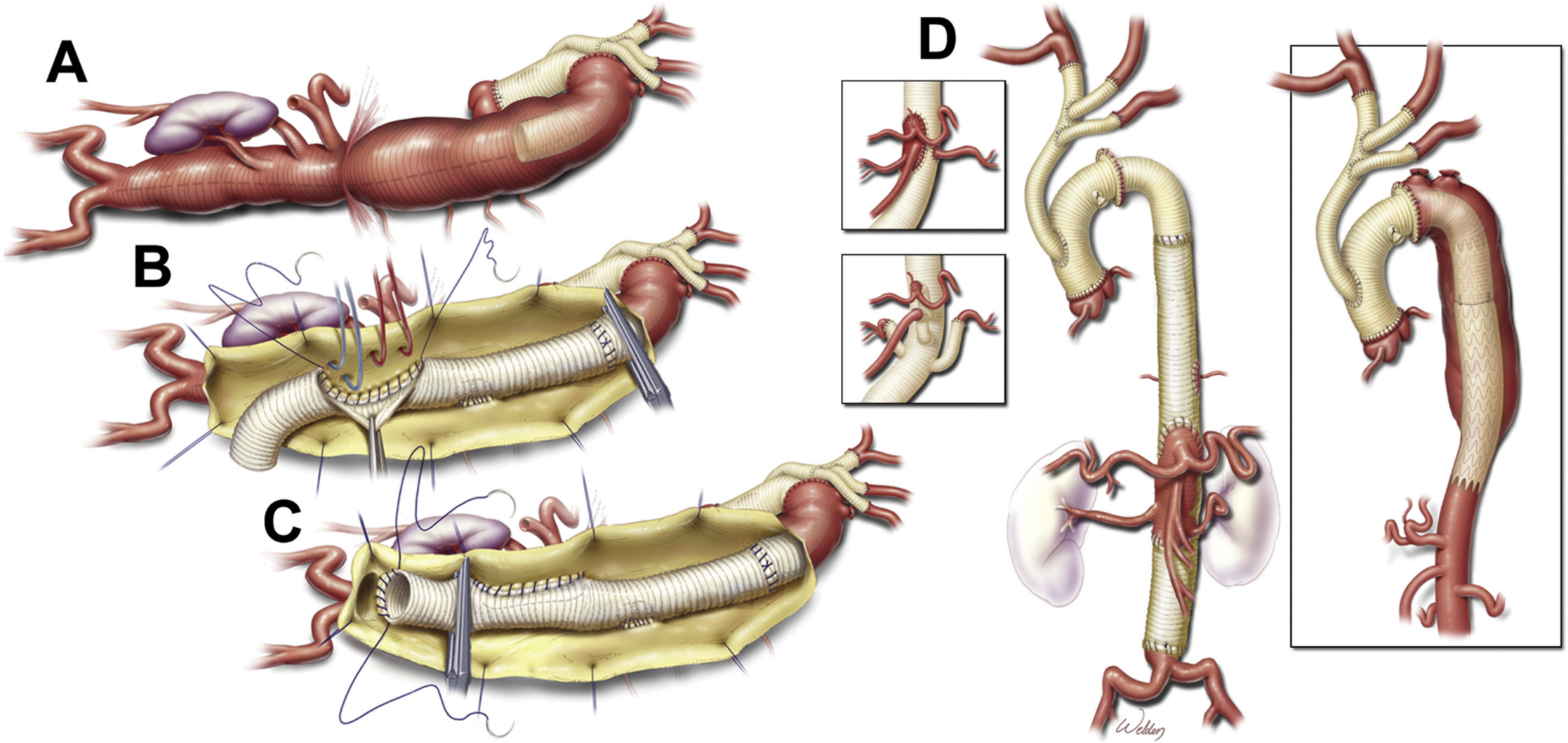
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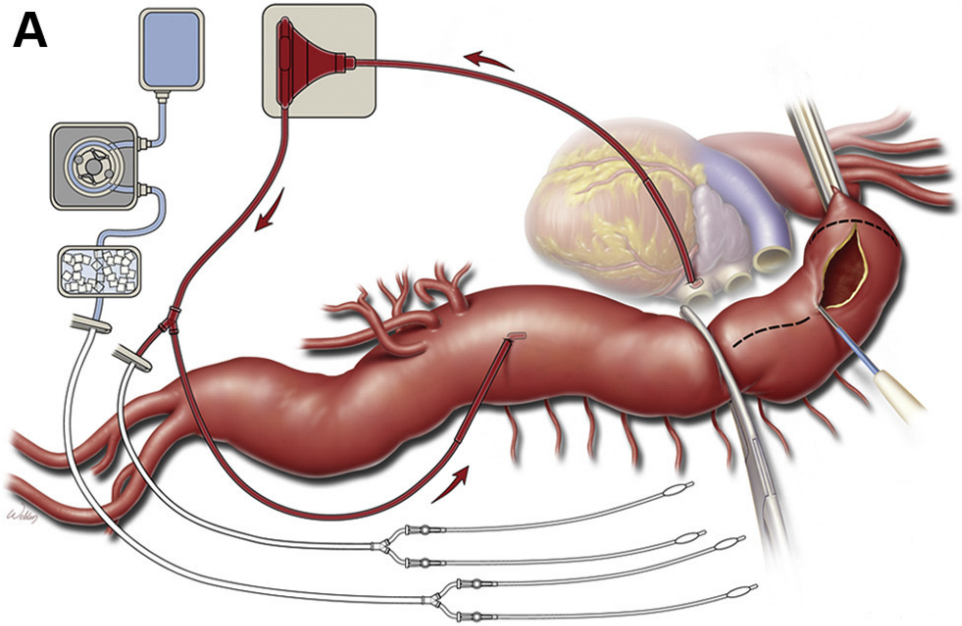
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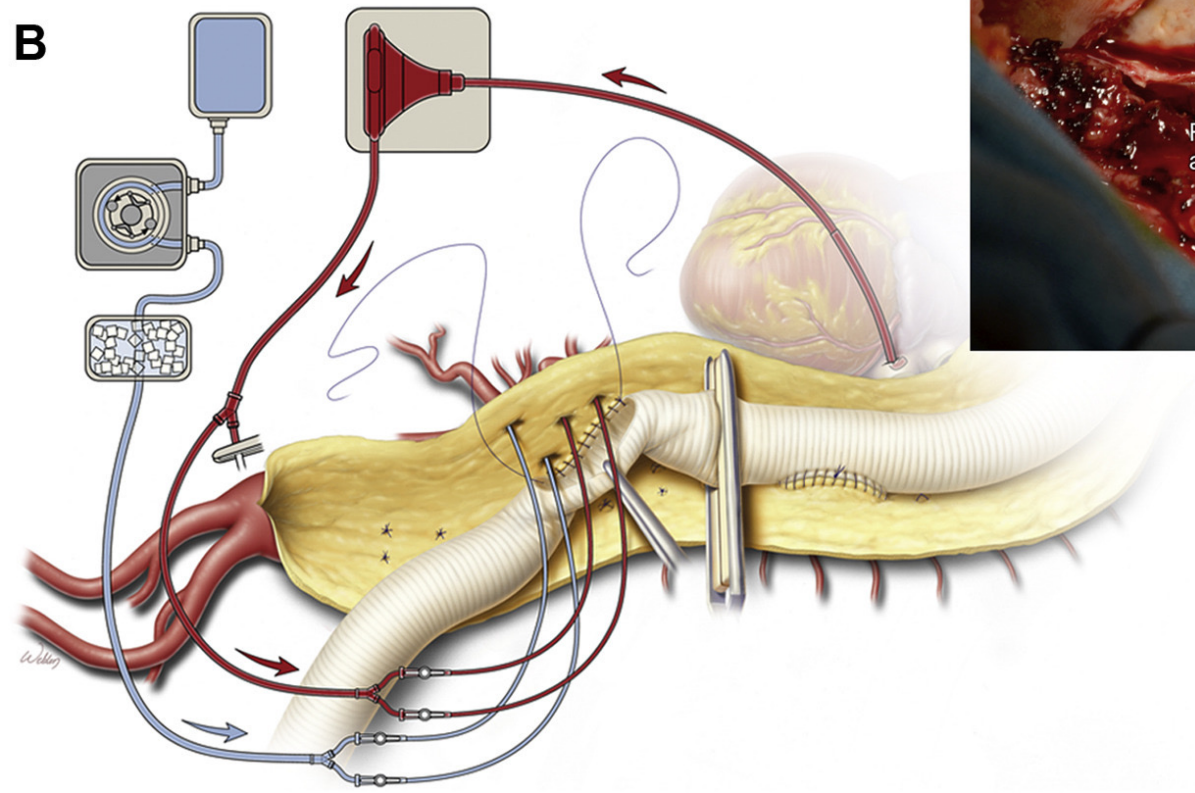
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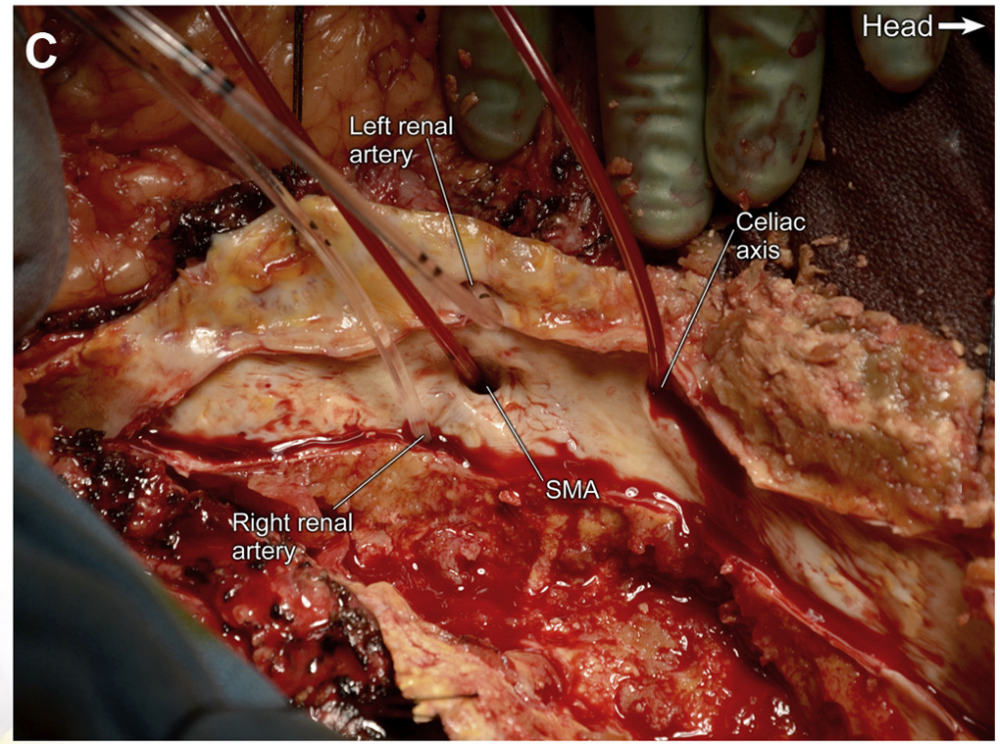
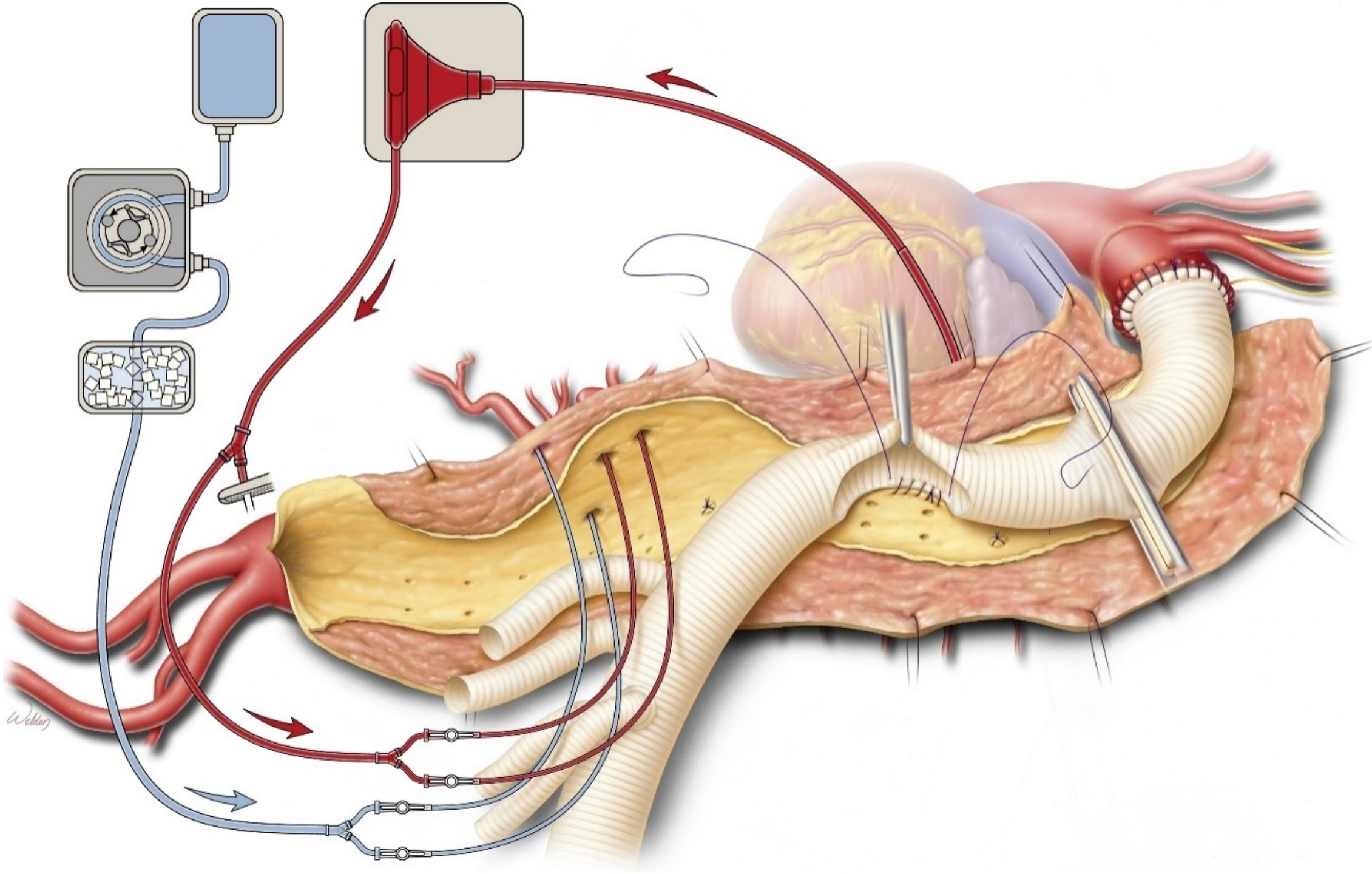
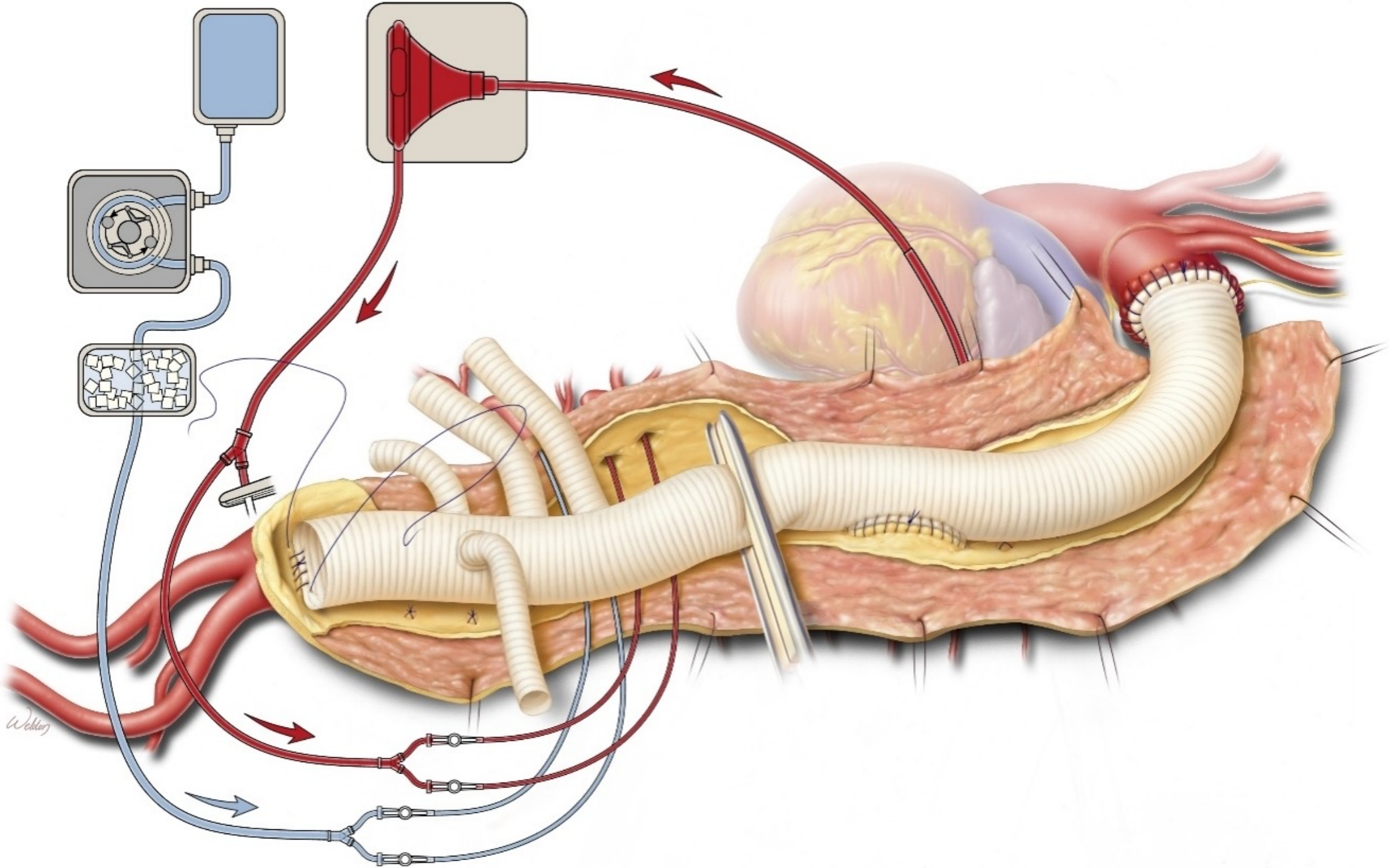
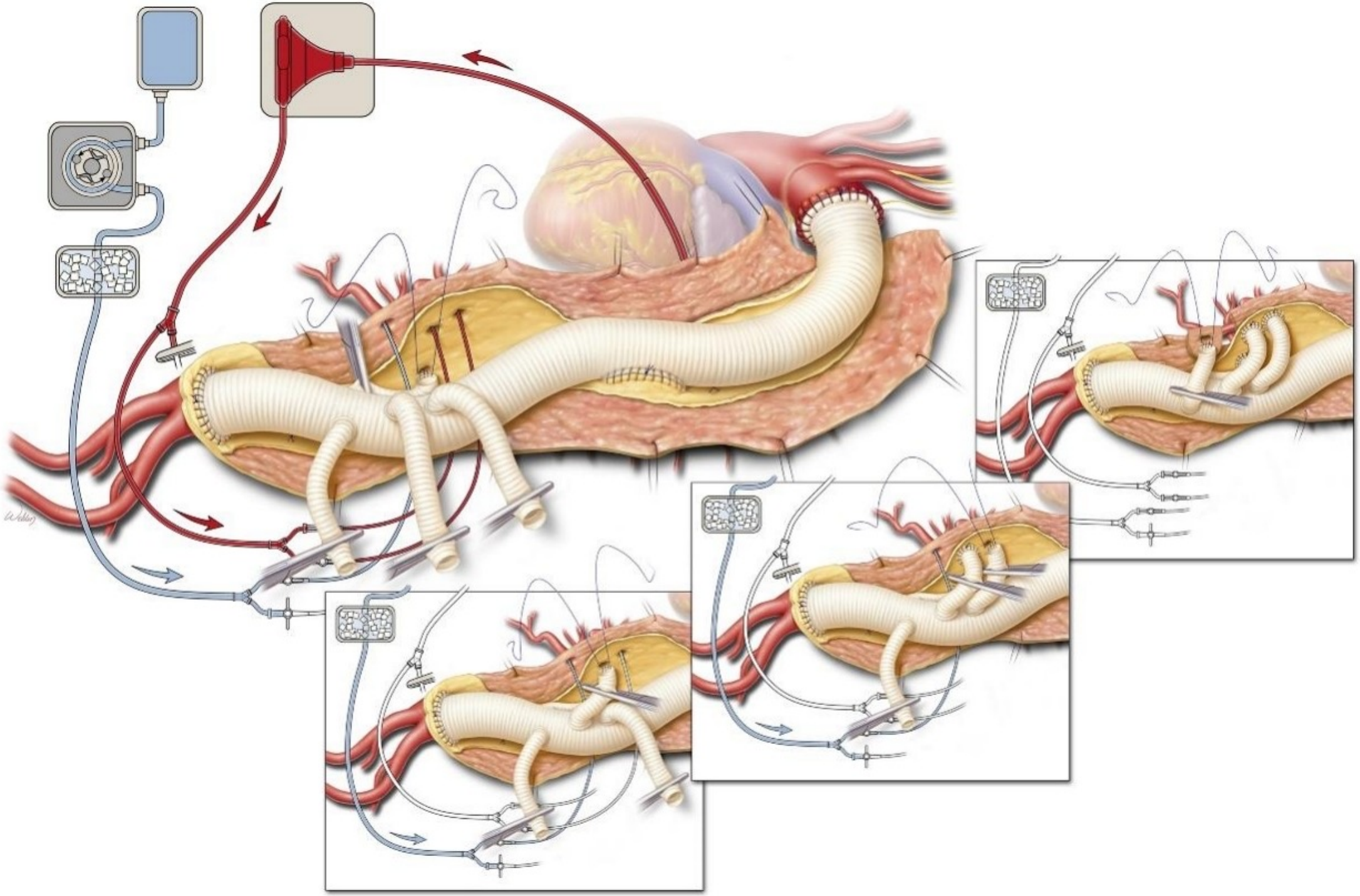
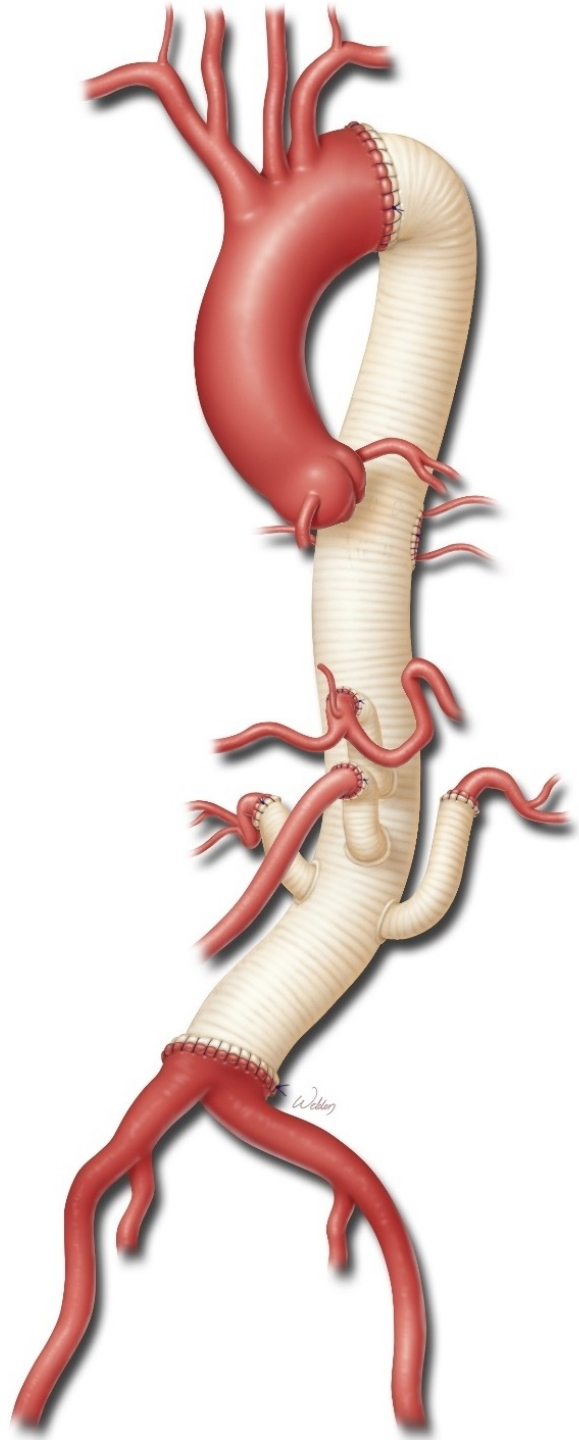

C
Head
Left renal artery
Celiac axis
SMA
Right renal artery

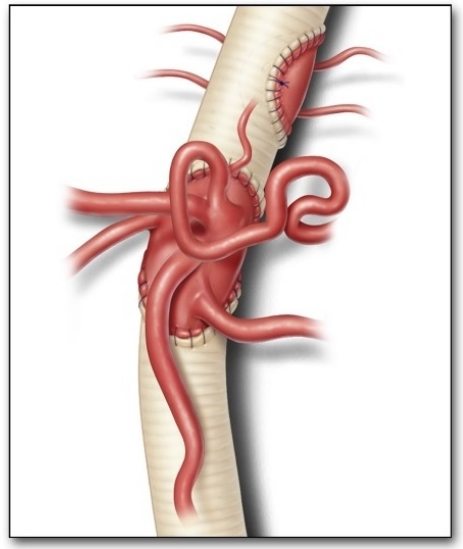


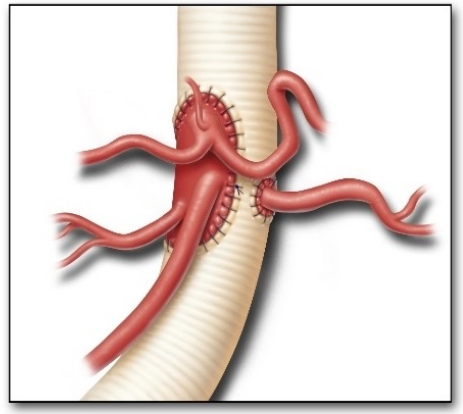


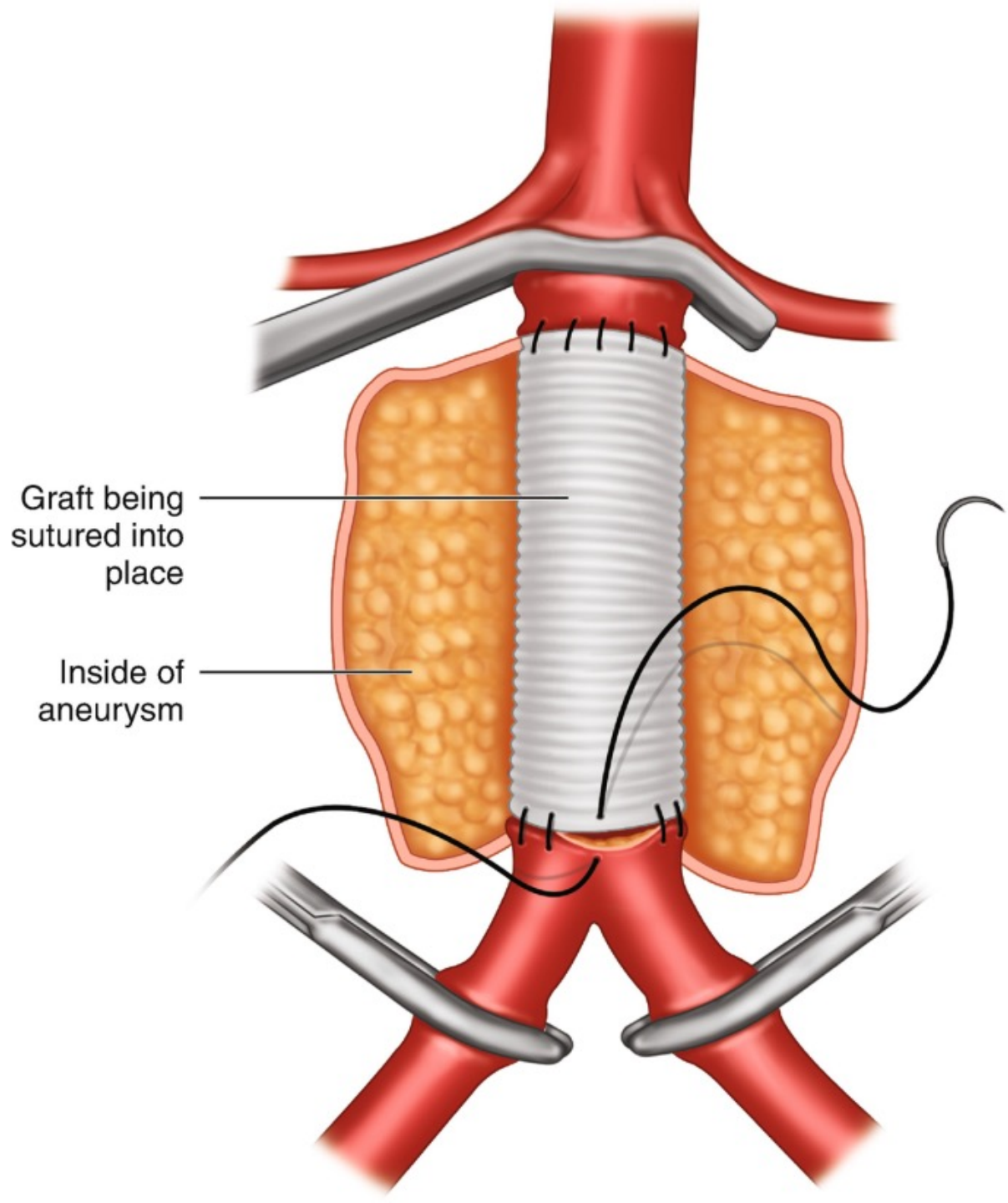


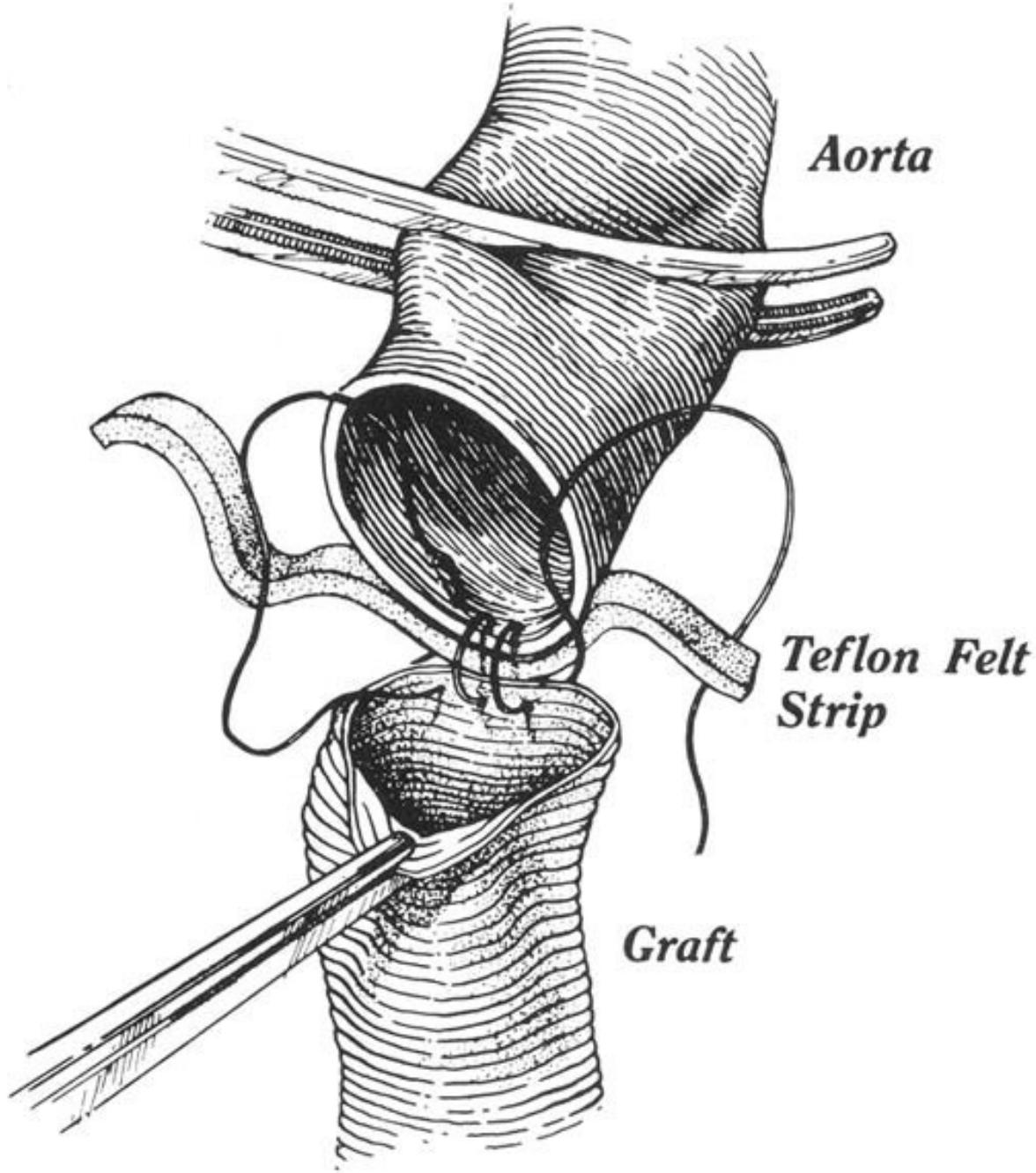

Graft-graft anastomosis of the lumber artery

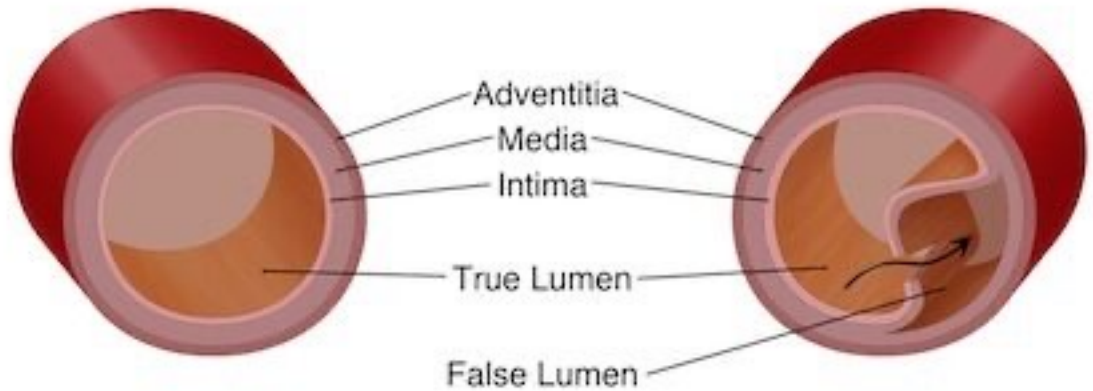







Graft being sutured into place
Inside of aneurysm


Aorta
Teflon Felt
Strip
Graft

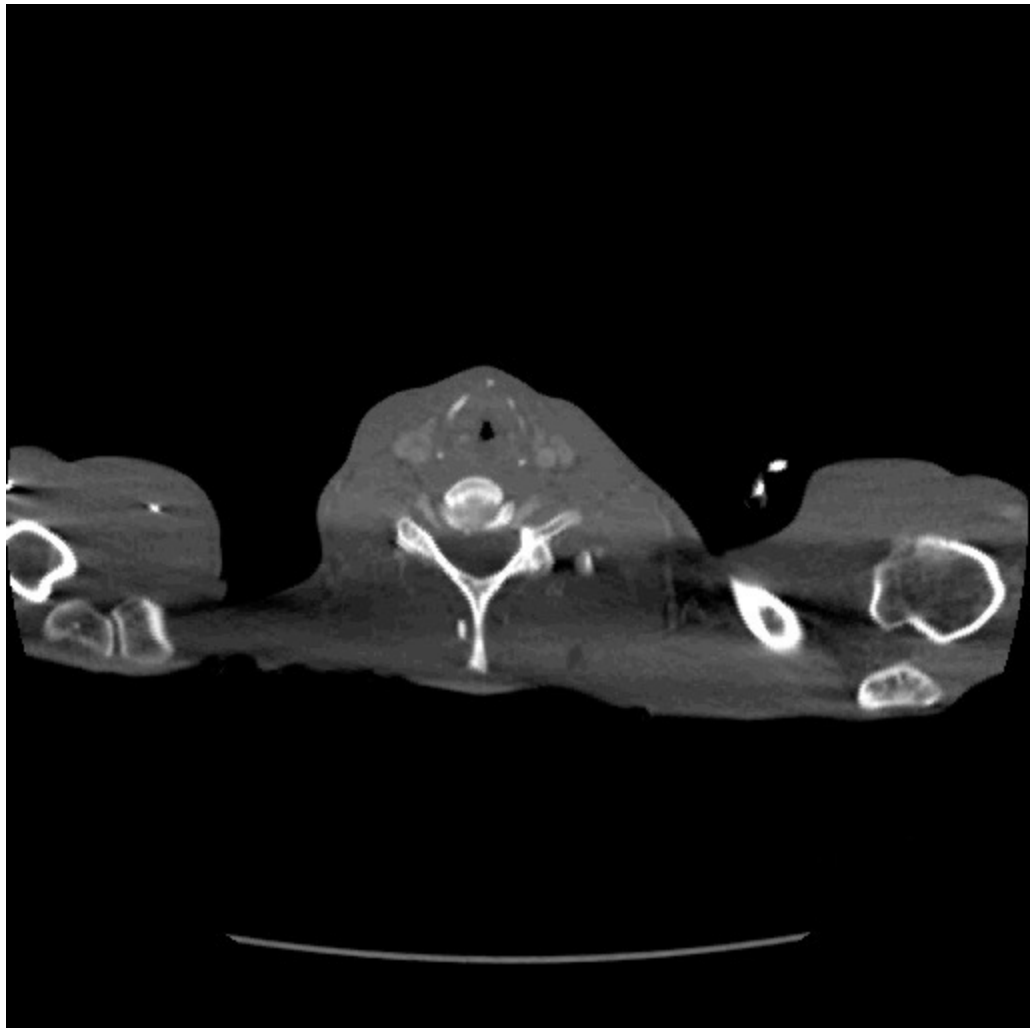


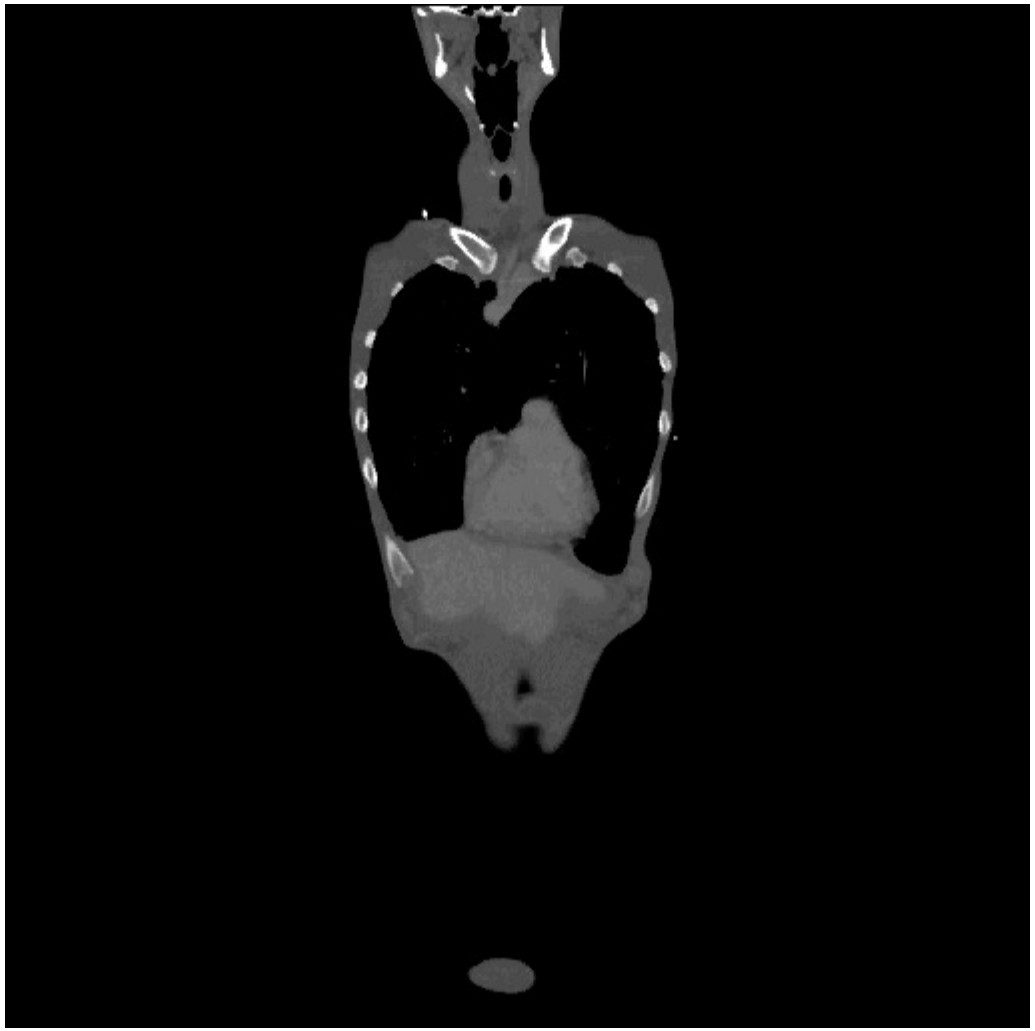
Normal Dissected

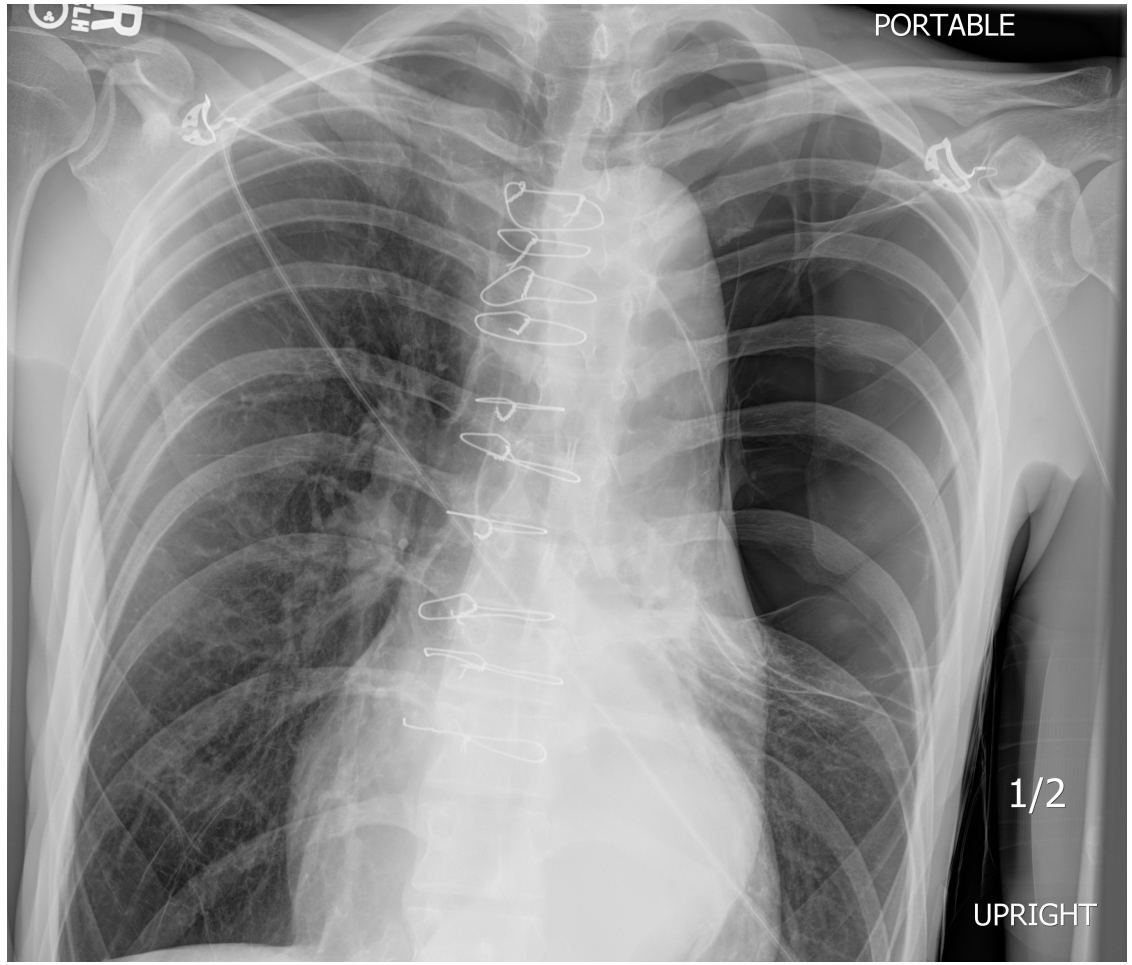

BrownMed
1
2

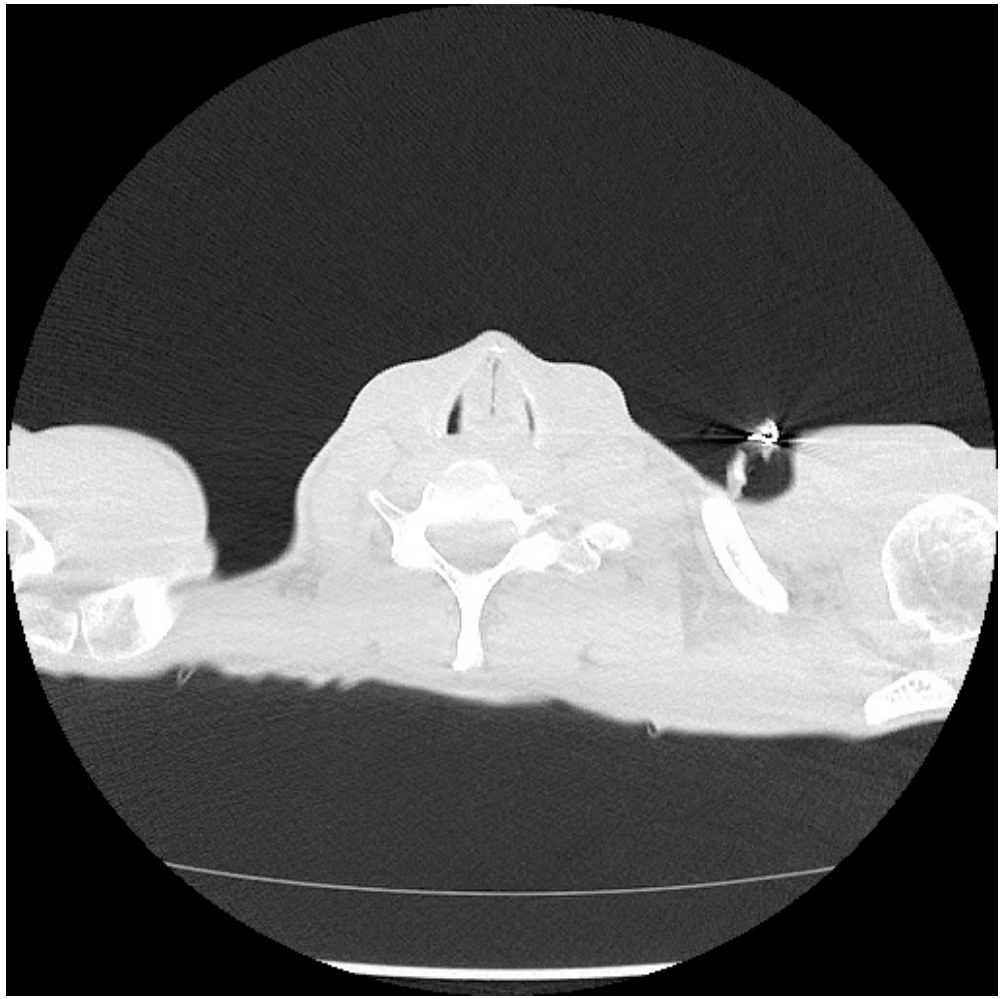
Modern Day Elective Aortic Intervention in Aortopathy Patients

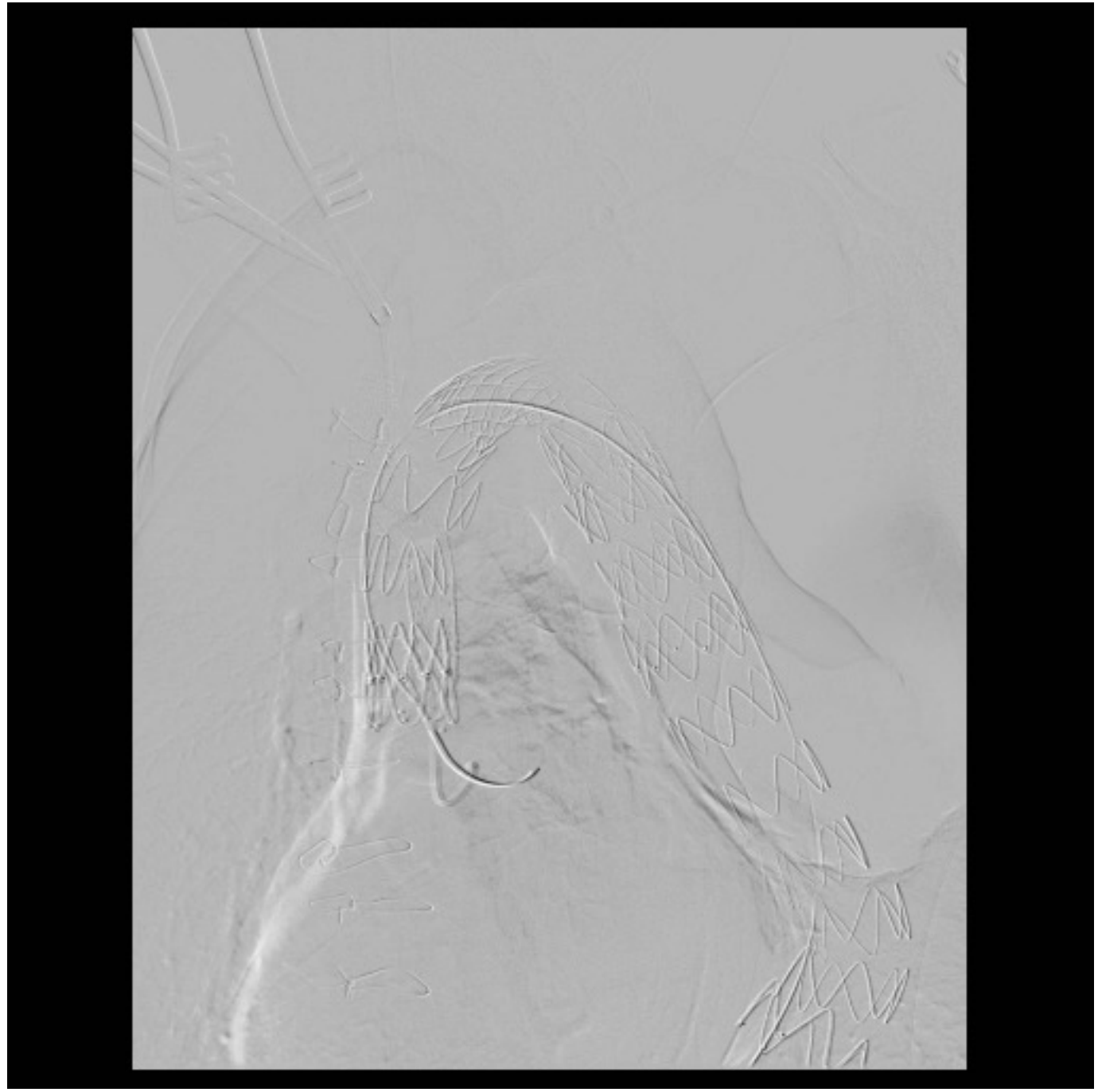
- 34 yo LD patient
- Hx of Type A Dissection repair at Standford
- Presented with rapidly enlarging thoracic region of thoracoabdominal aortic aneurysm

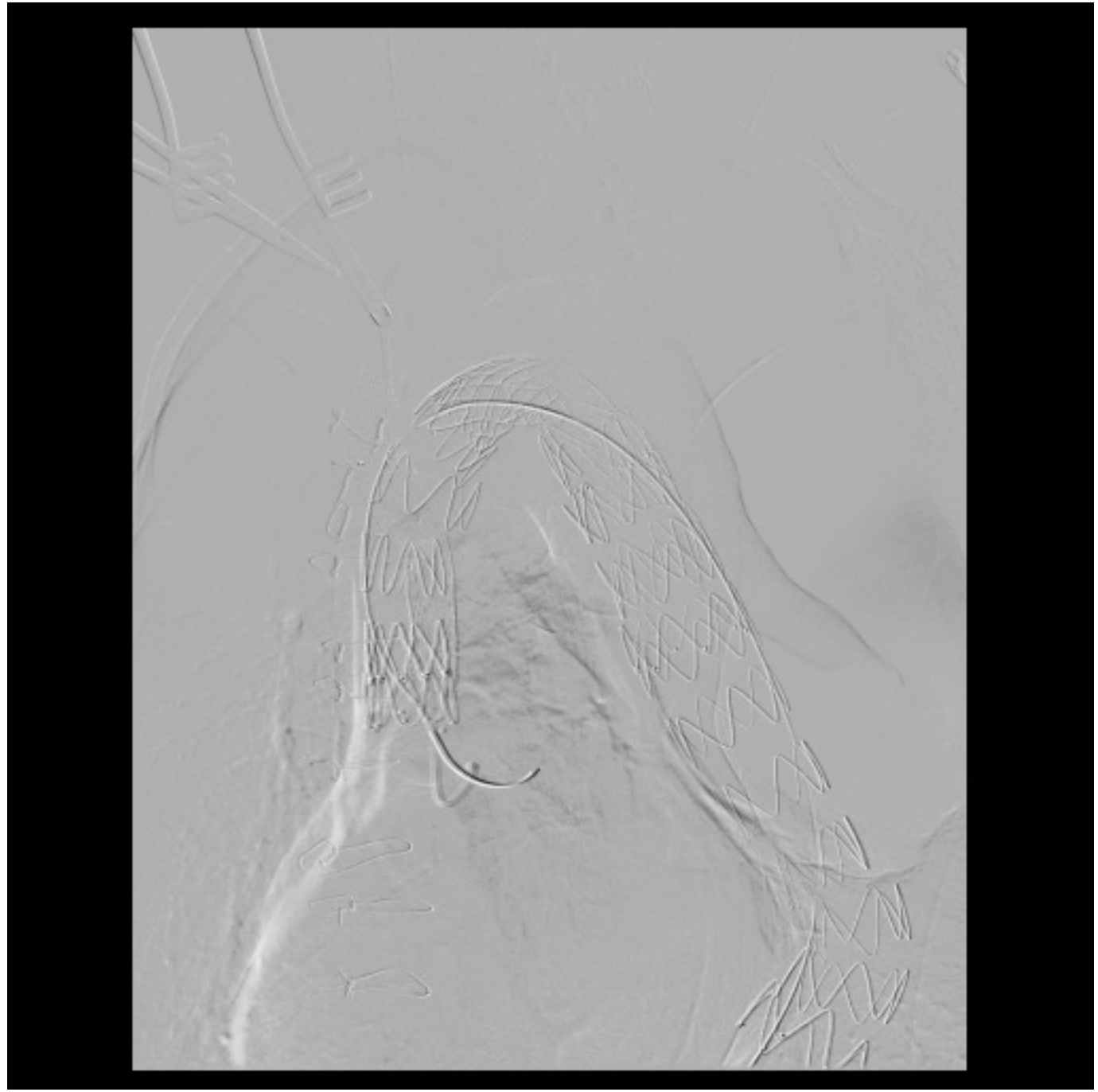





PORTABLE
1/2
UPRIGHT

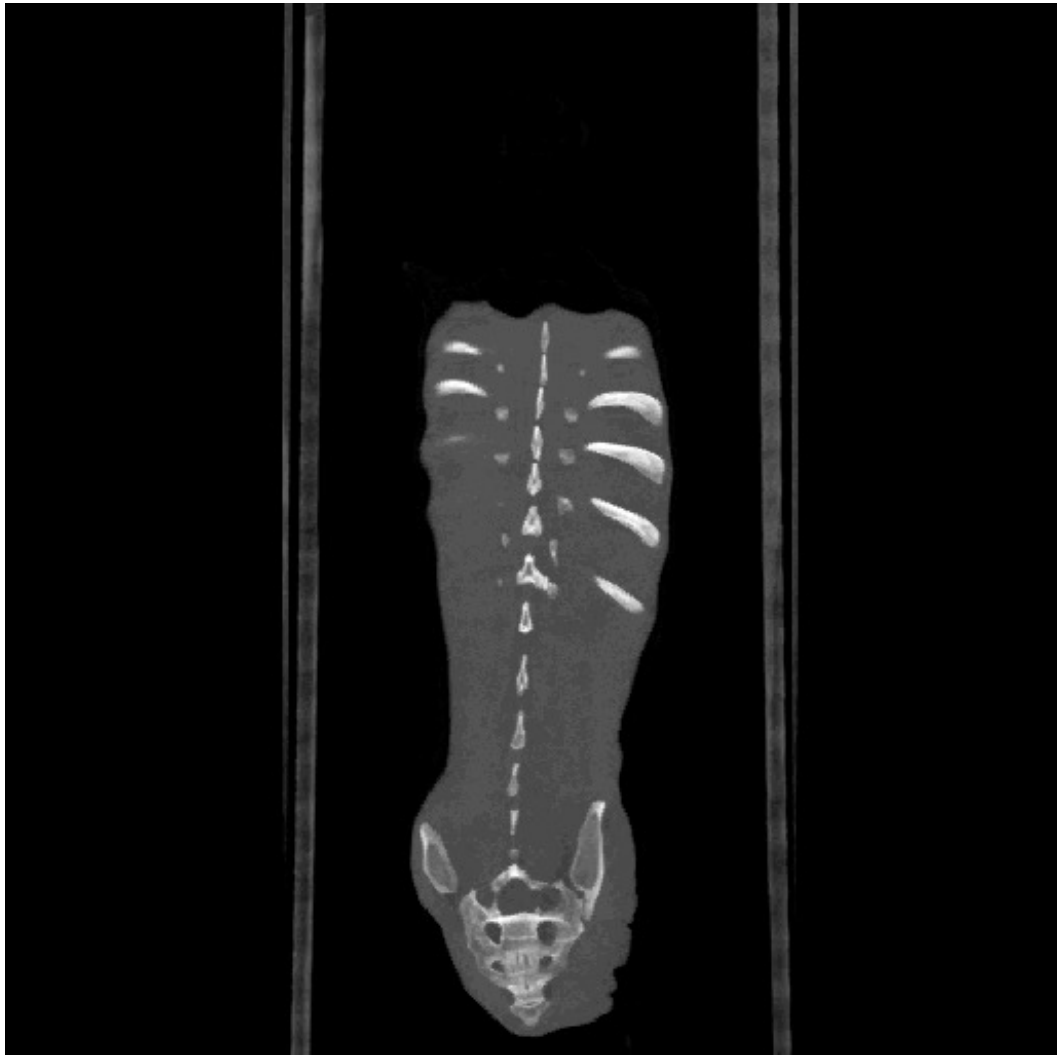


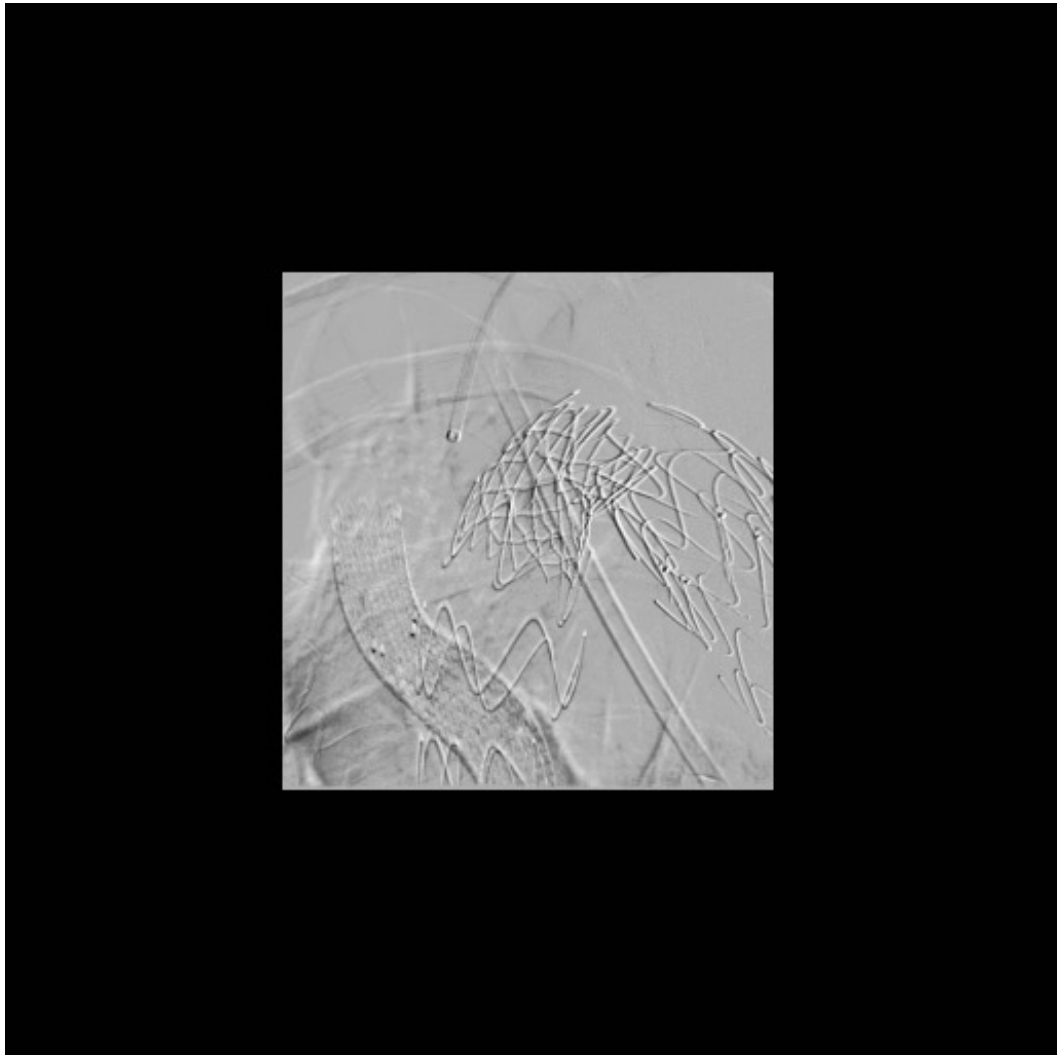


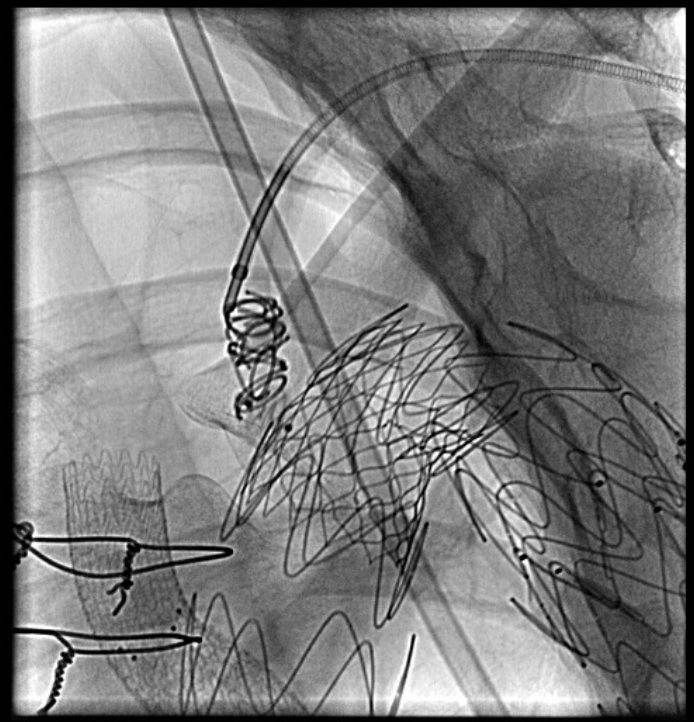


Stage II – a few days later

- Aorto-biiliac graft with infrarenal clamp
  - to CIA bifurcation bilaterally
- Separate bypass to pair of lumbar arteries sewn to aortic graft
- Retrograde debranching to hepatic artery, SMA, and bilateral renals using two 14 x 7 mm bifurcated grafts anastomosed to aorto-biiliac graft

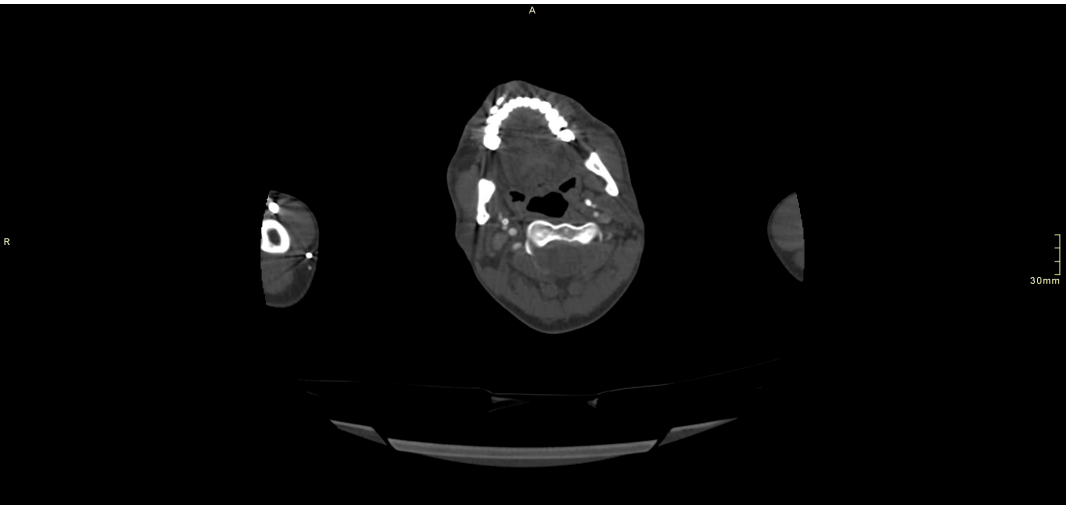






Success Short Term

- No Endoleak
- Discharged
- Followup??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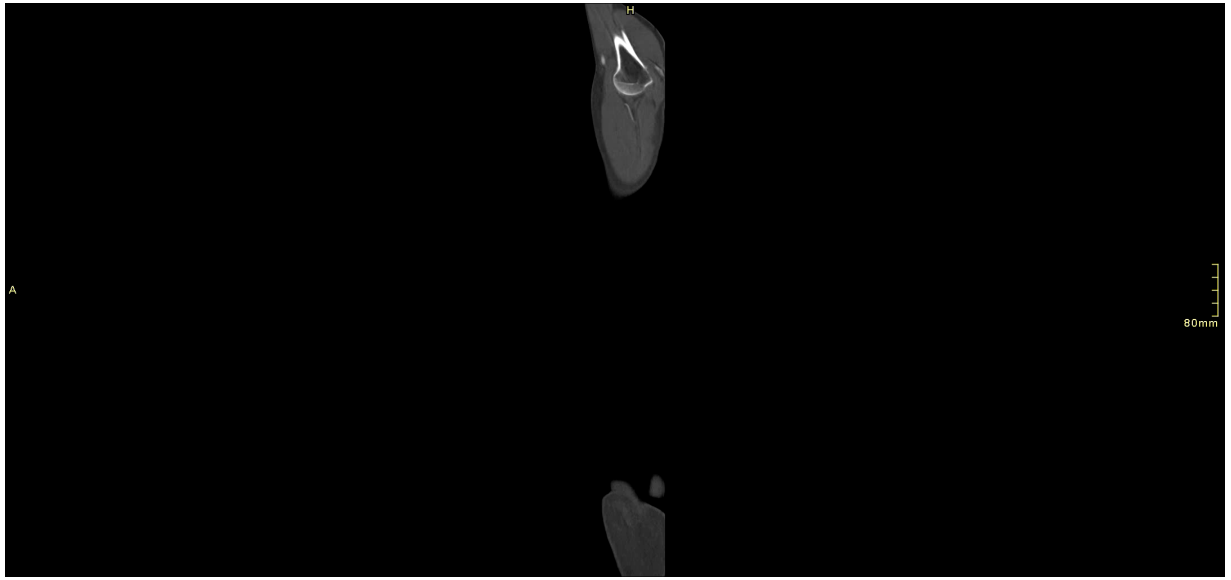
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A
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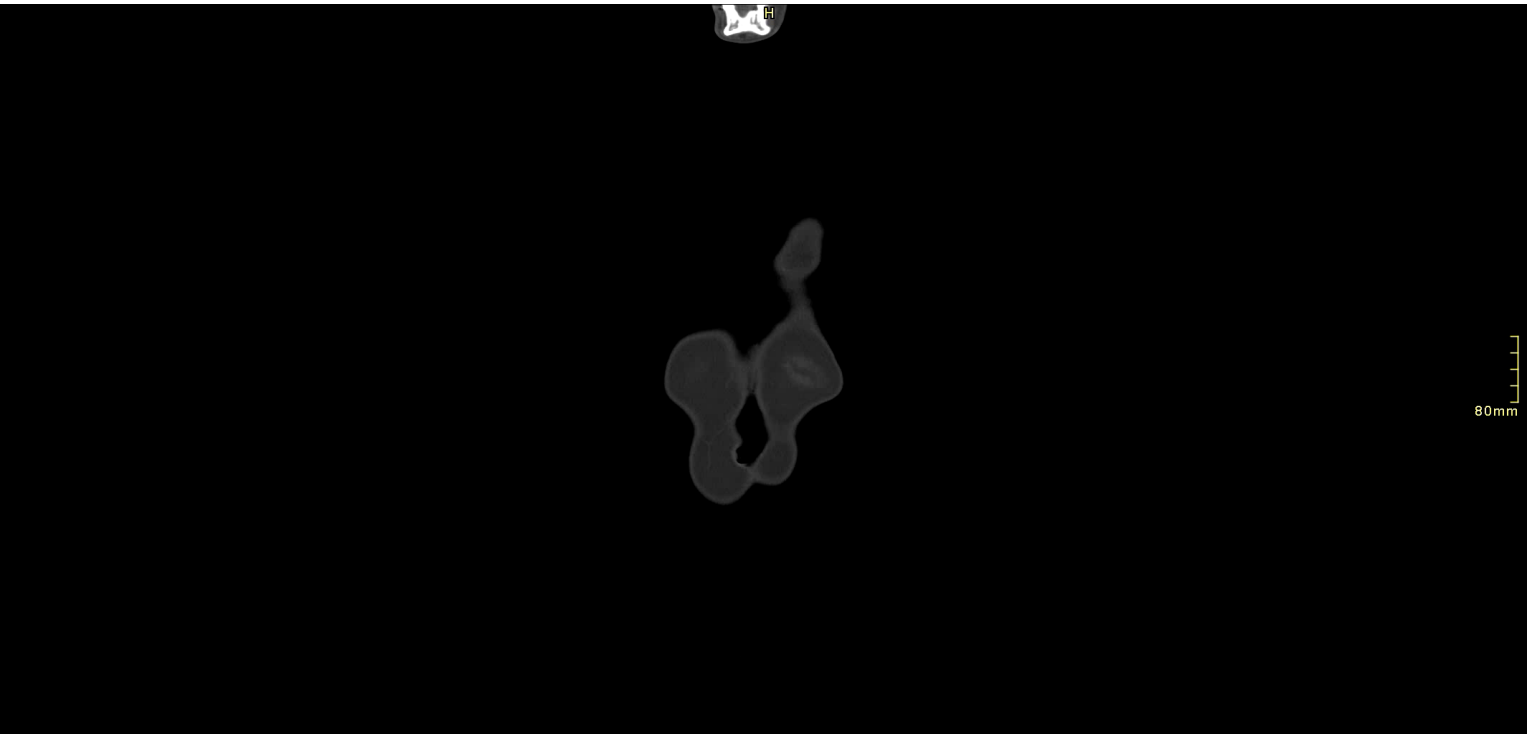
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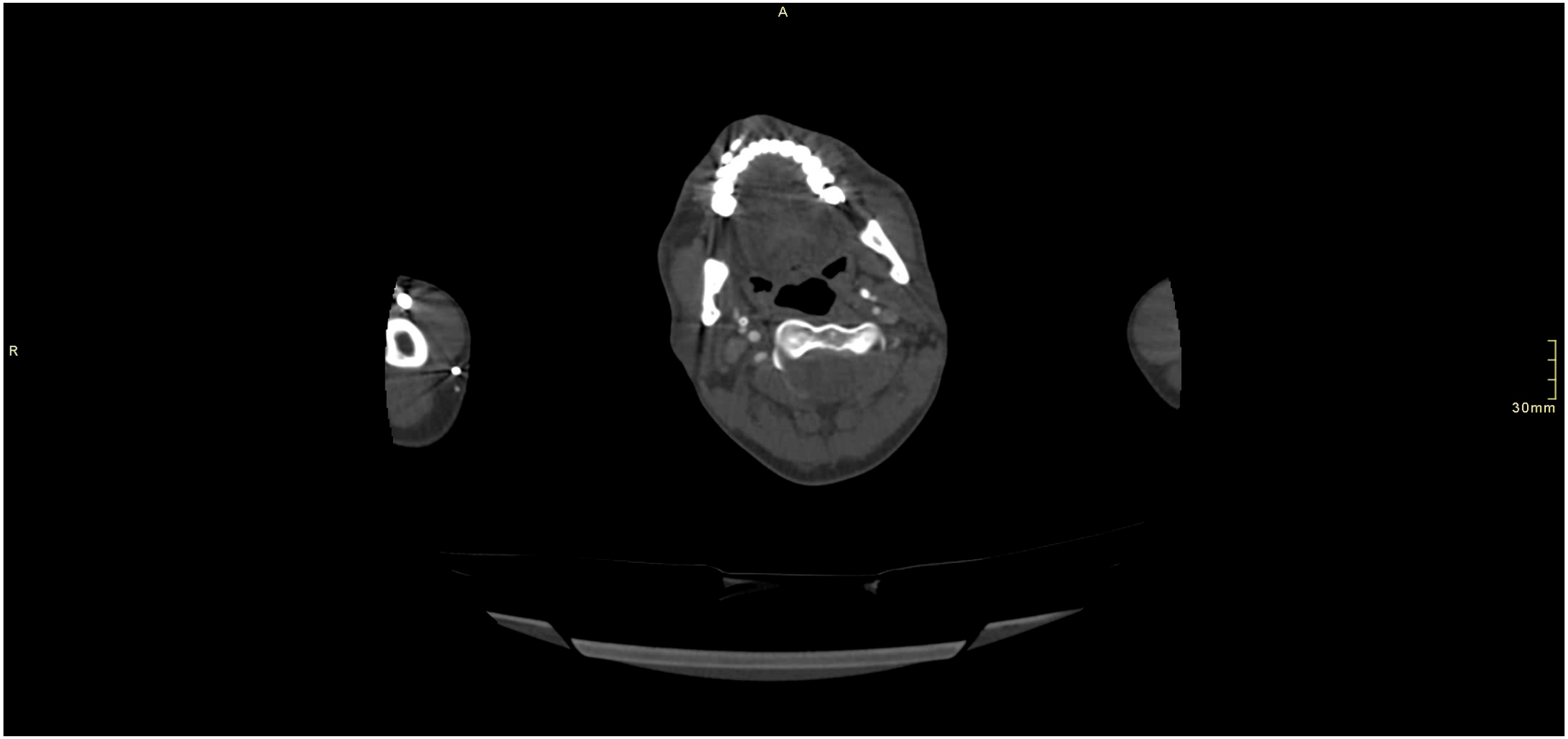
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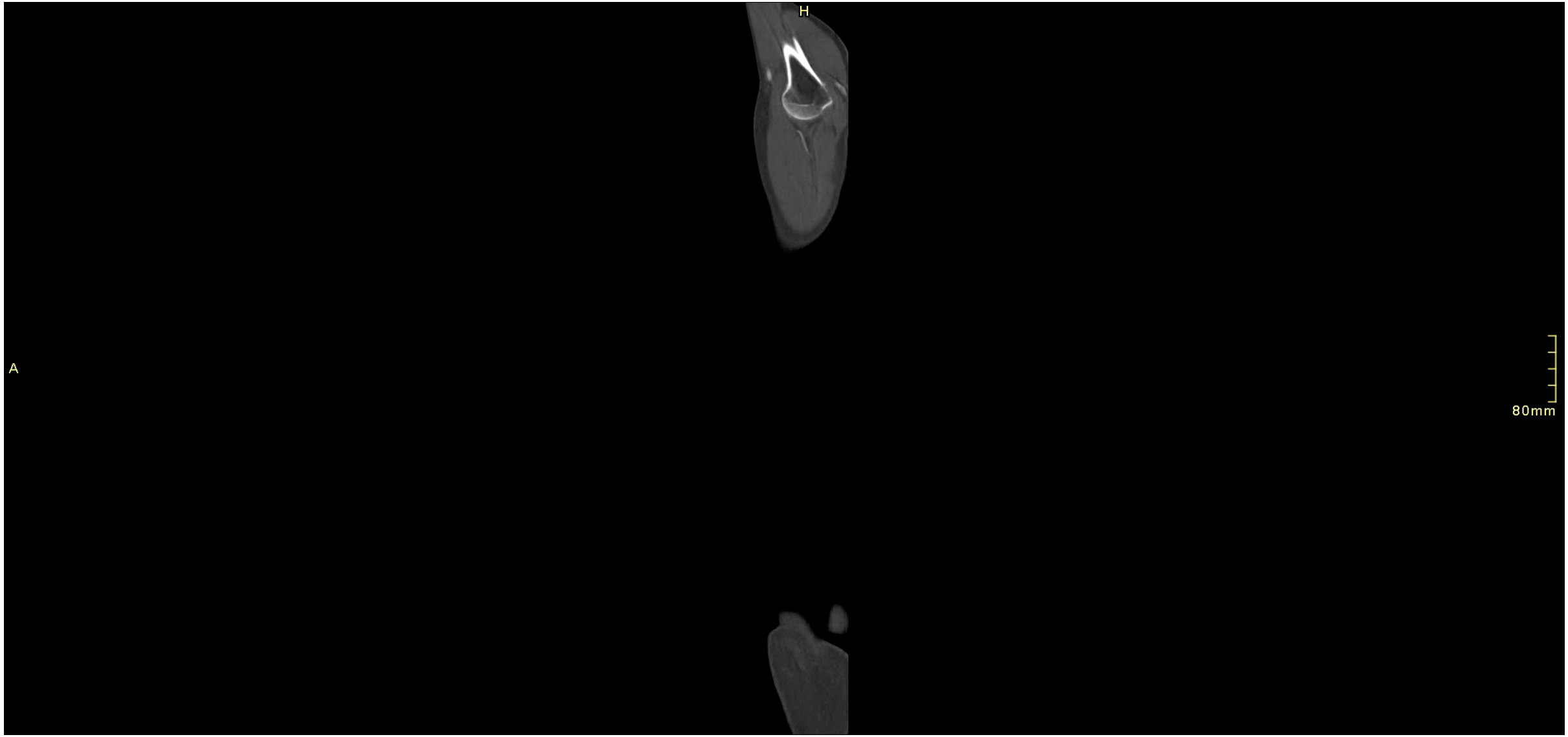
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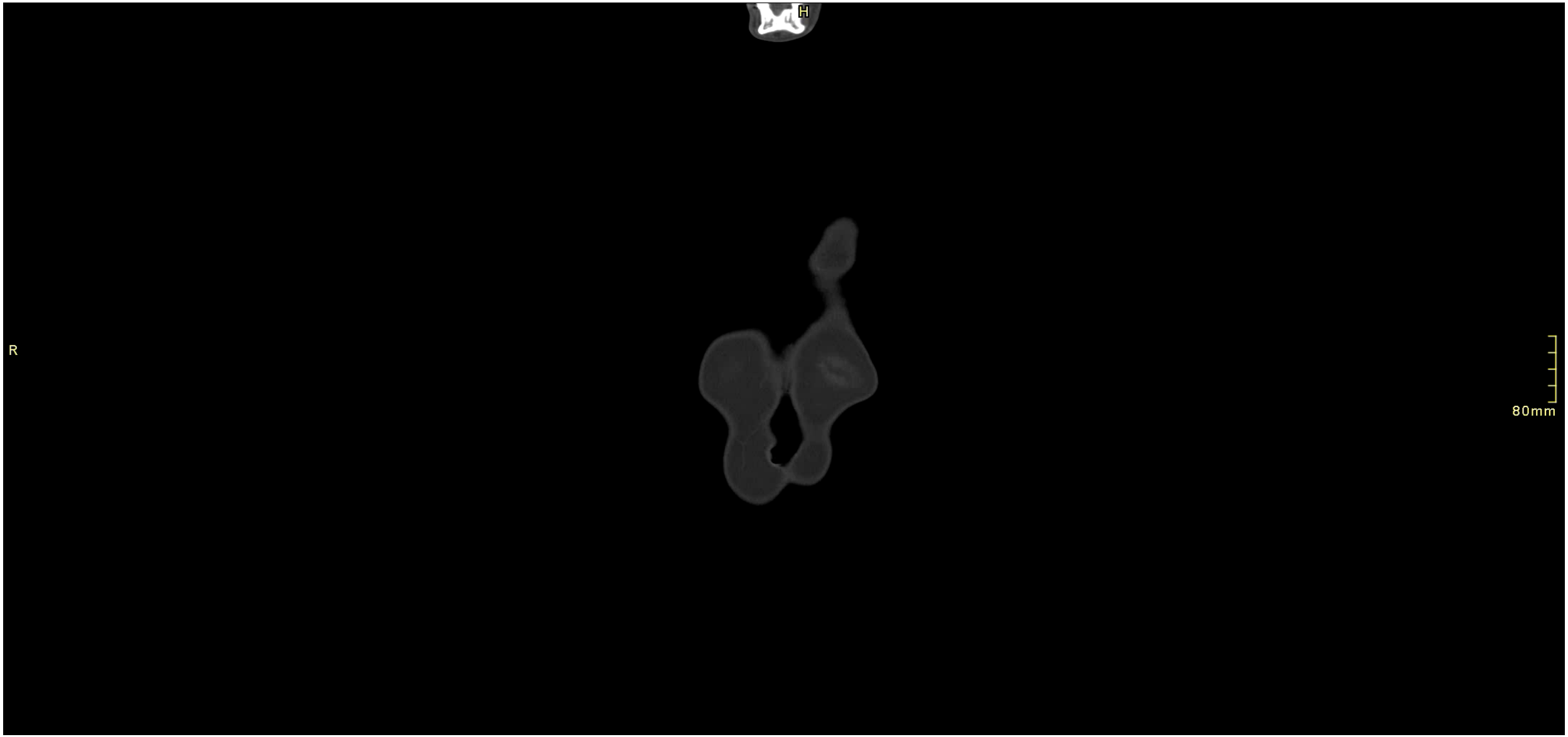
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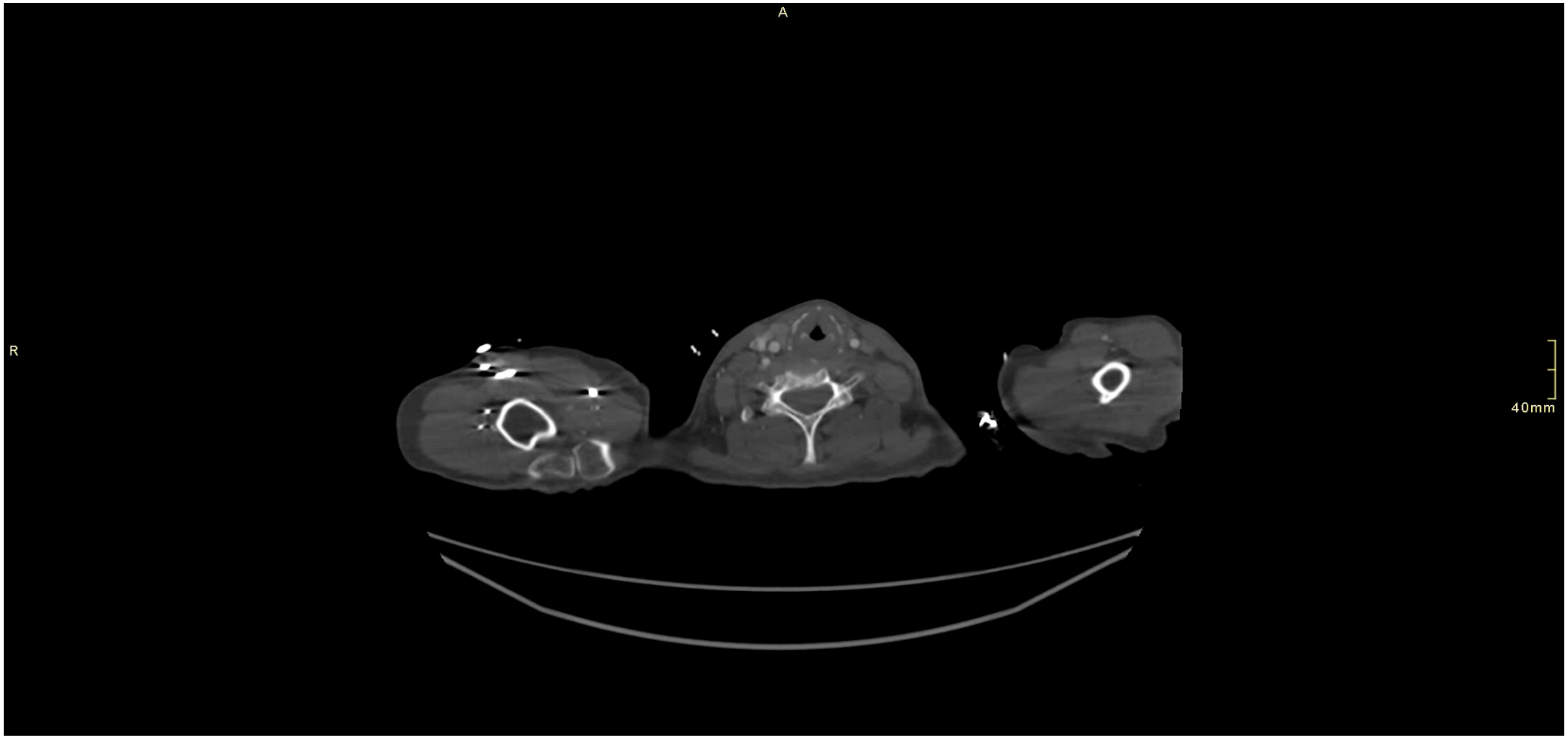
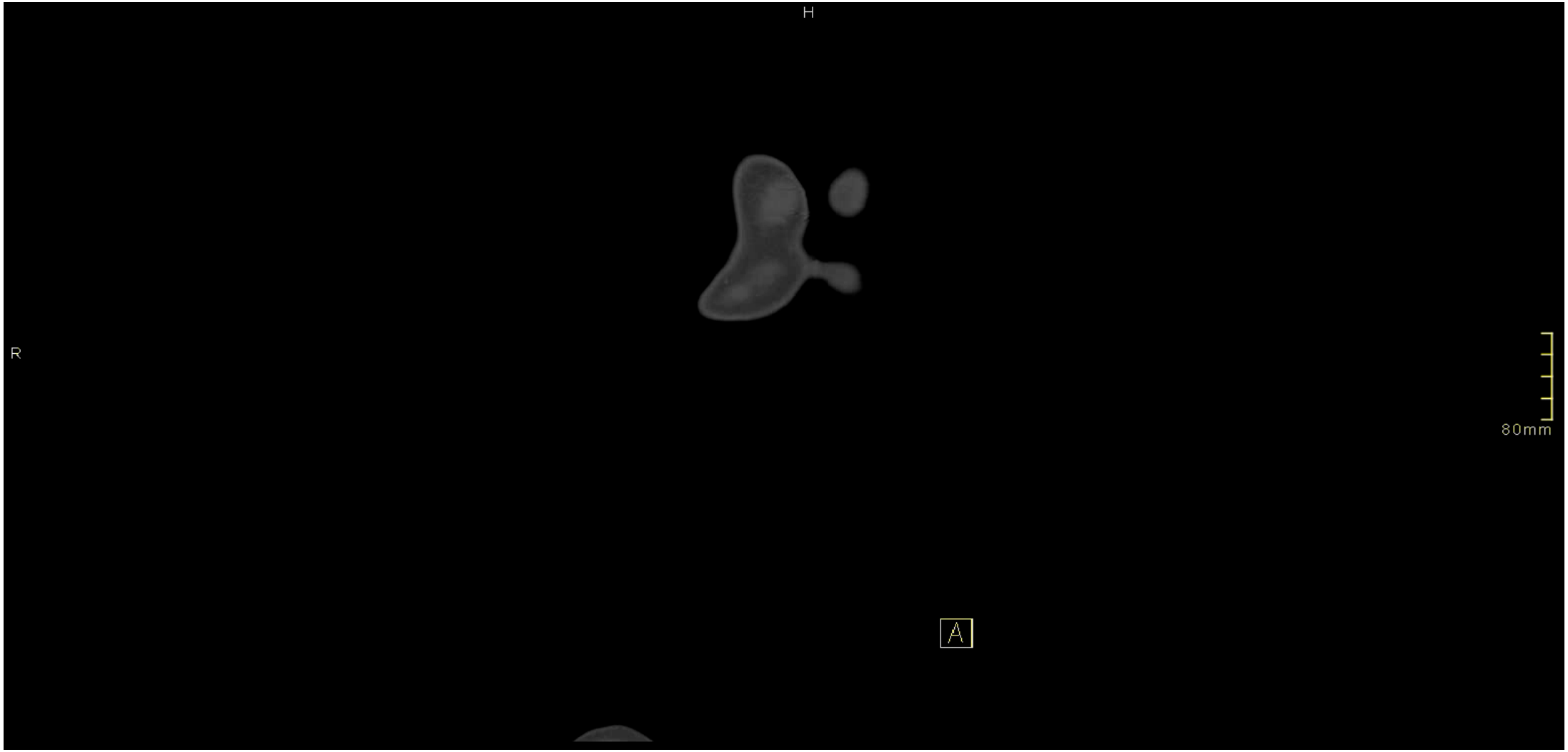
30mm
[proj]


A
R
40mm

FET Set up for Distal Aortic Intervention


H
R
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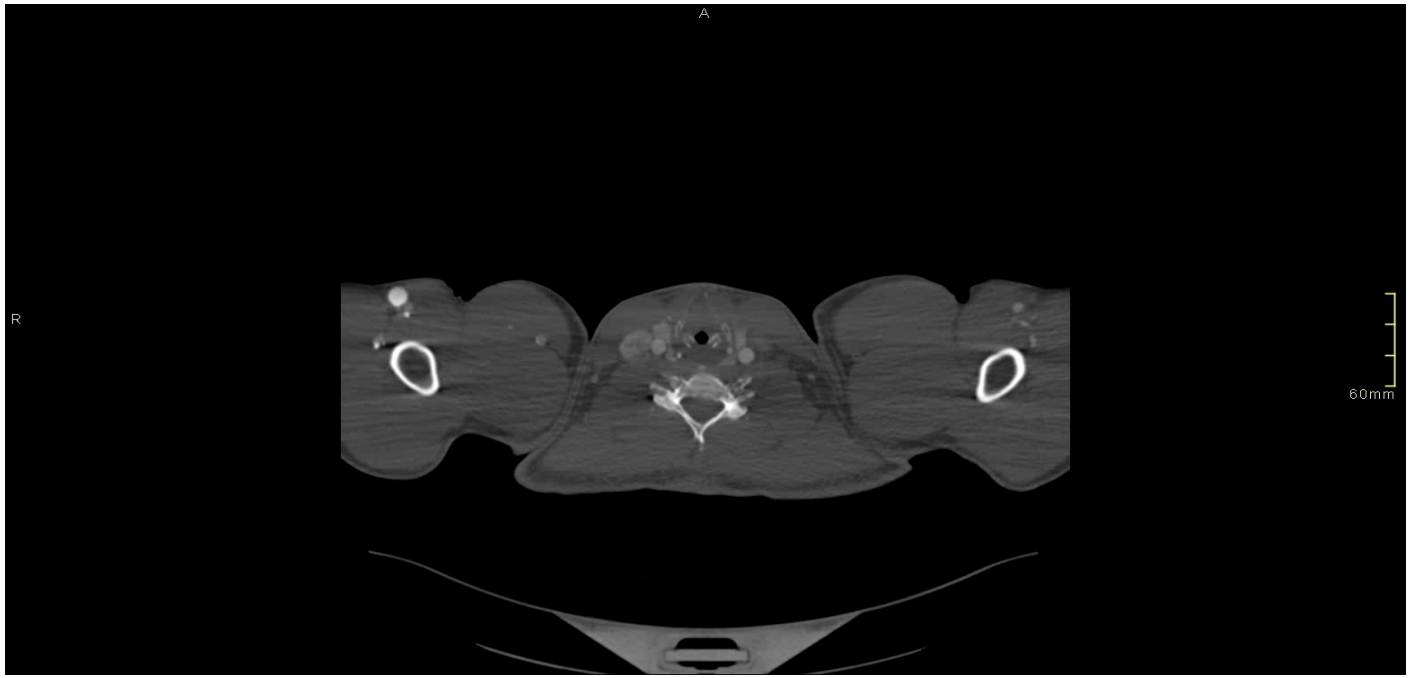
A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60mm
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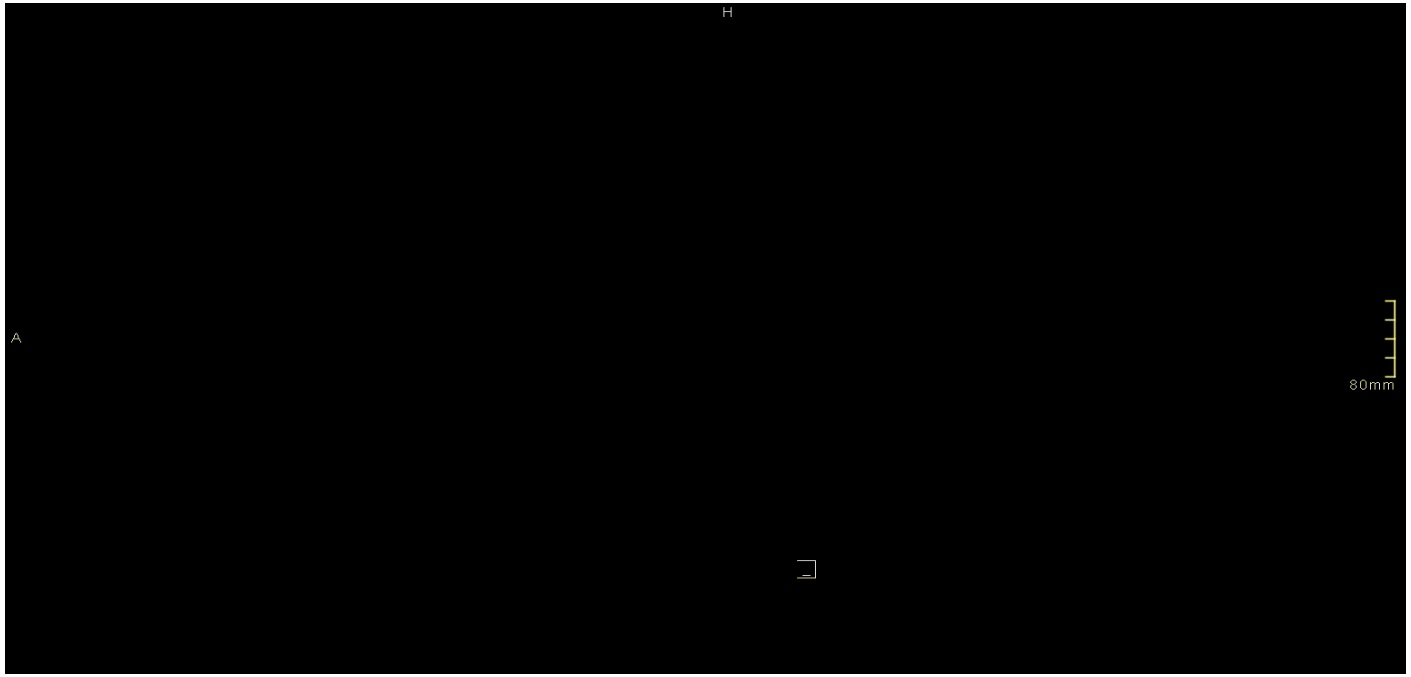
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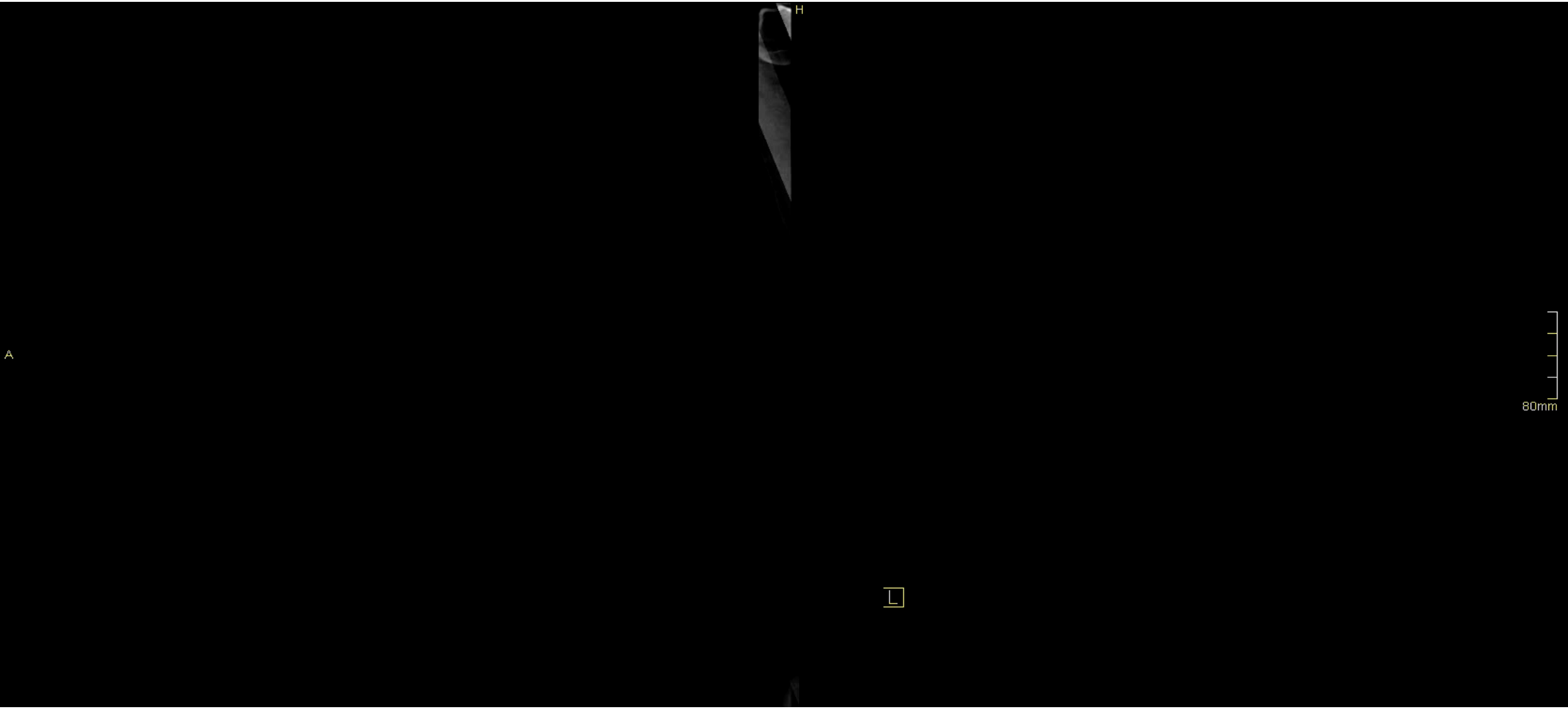
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A
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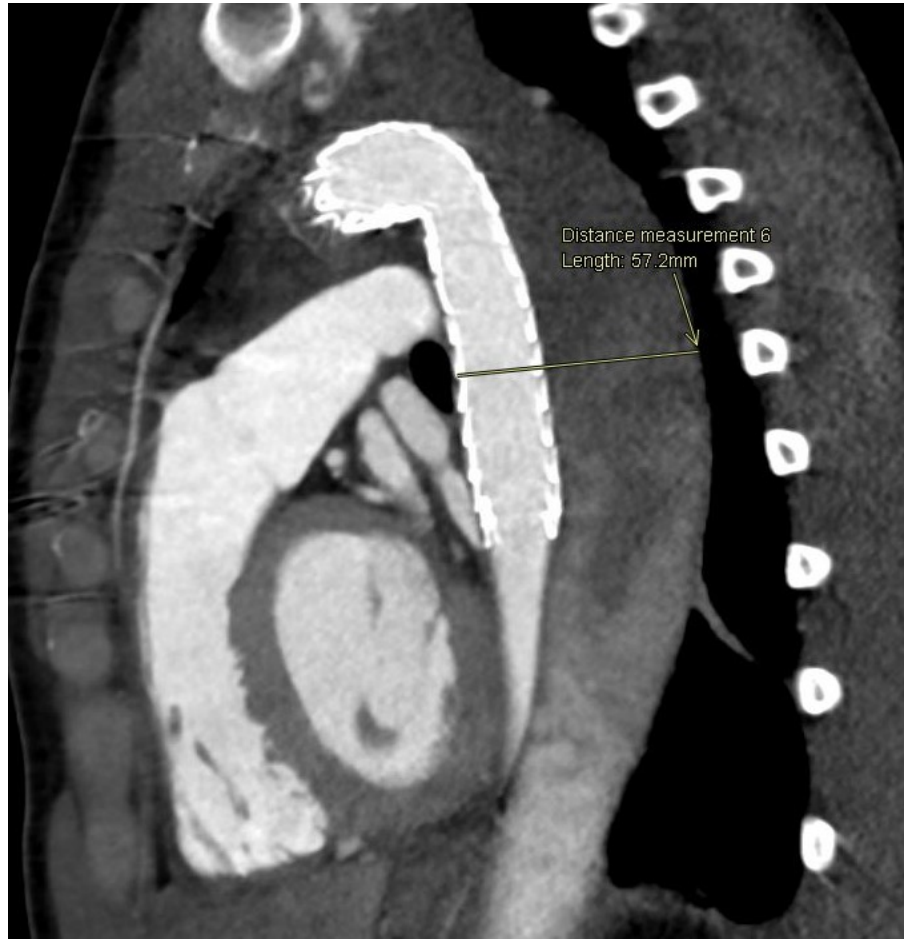
80mm


Distance measurement 6
Length: 57.2mm

Thank You
Dr Shalhub
Industry Sponsorship