CIRA Case of the Week
March 2016

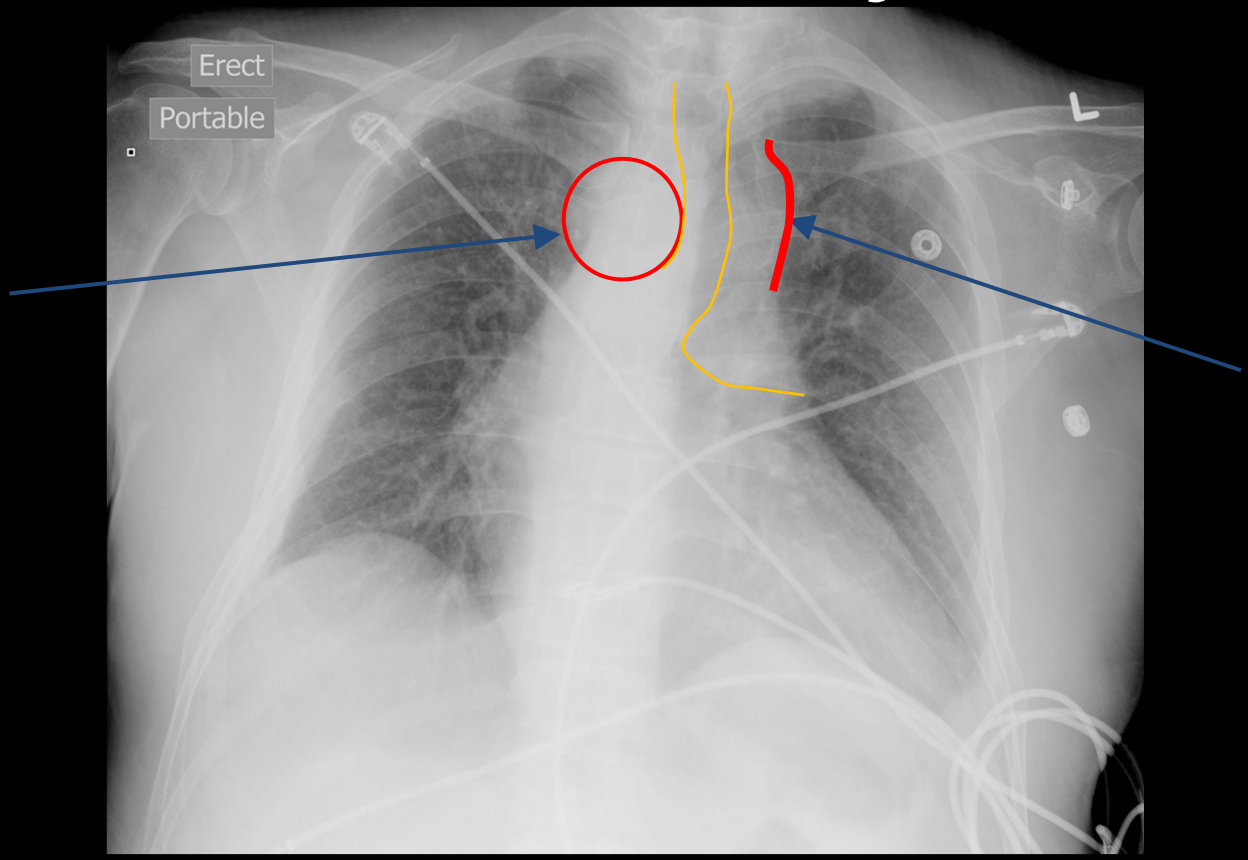
Case Courtesy of Drs. Sacha Oomah and Siuchan Sookhoo

University of Manitoba

Patient KD

- 56 year old male with new onset of shortness of breath
- Also had been complaining of progressive dysphagia
- PMHx: progressive muscular dystrophy NYD
- No infectious symptomatology, WBC normal

Chest X-Ray



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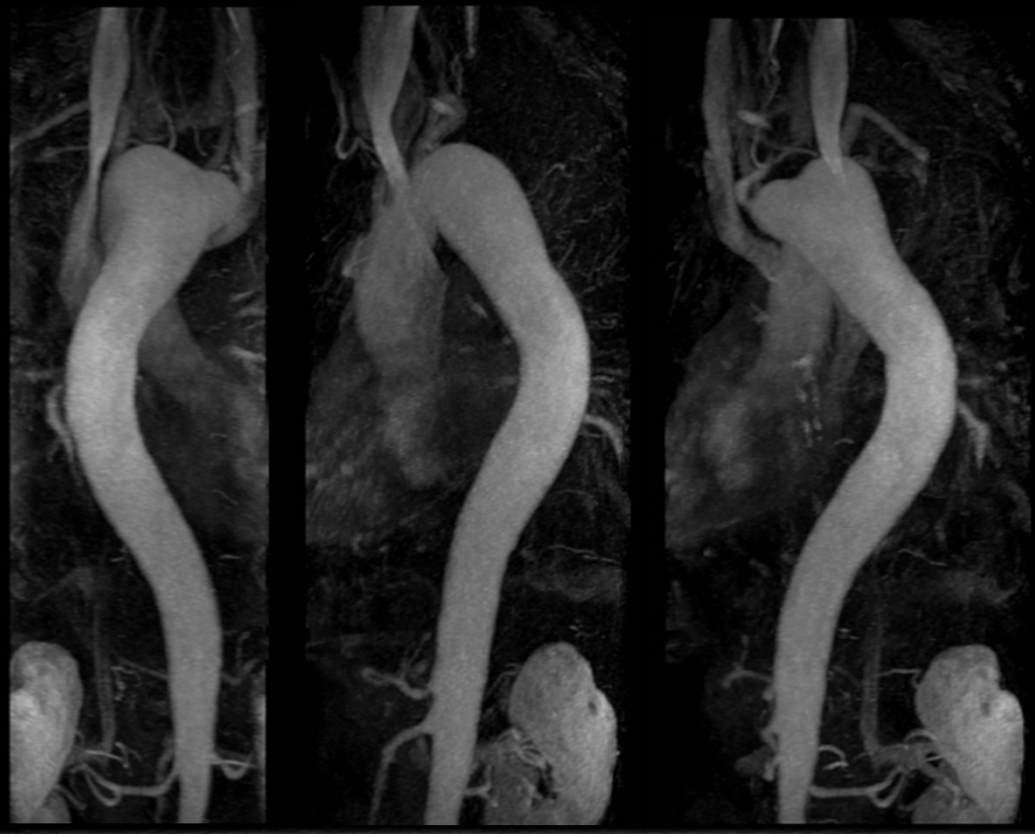
Chest X-Ray

- Findings:
  - Pulmonary edema
  - Right sided aortic arch
  - Added convexity to the left paratracheal stripe, in the expected location of a left aortic arch
  - No indentation on the left trachea

Cardiac MRI 2010



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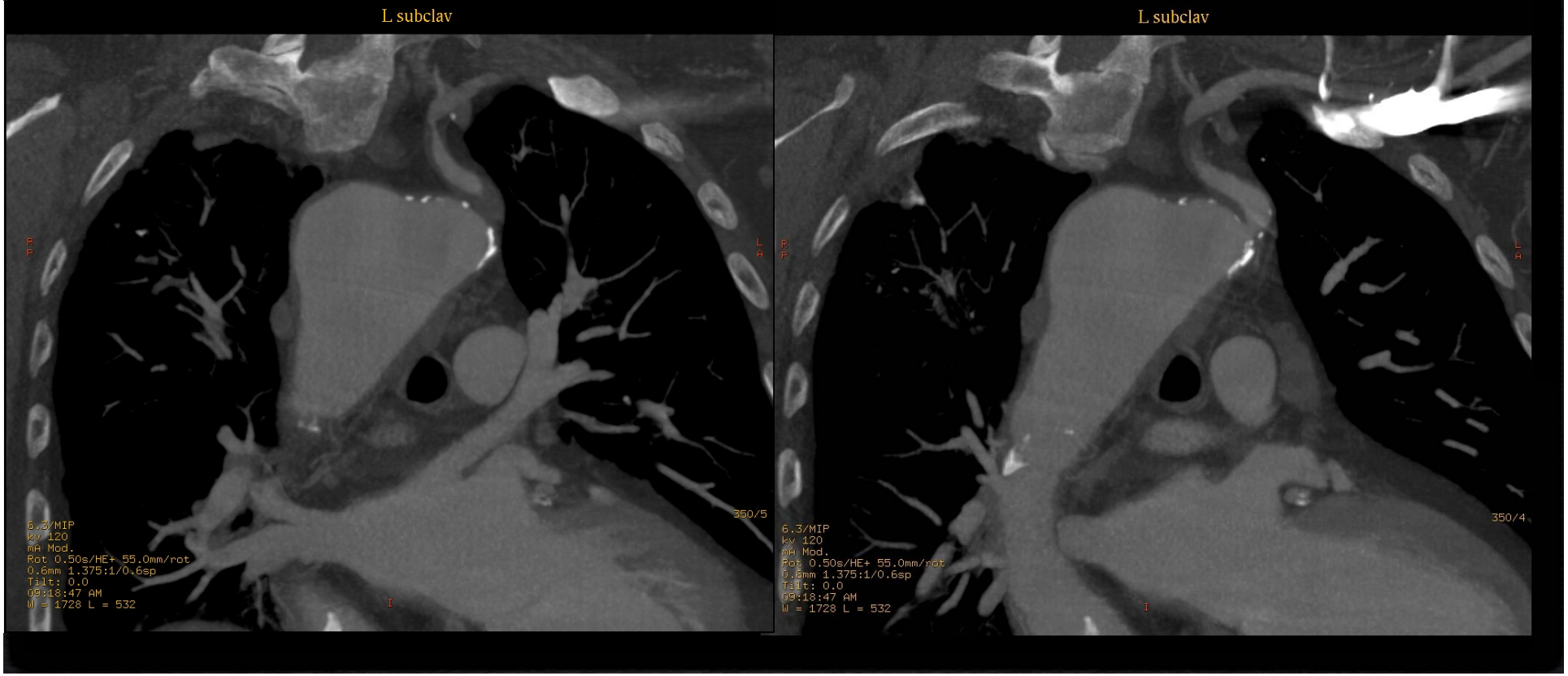
Initial Management

- The patient received a one year follow-up CT
- The CT demonstrated that the aorta at the level of the Kommerell diverticulum had increased from 6.2 cm to 6.7 cm.
- Intervention was planned

CT Angio

CT Angio

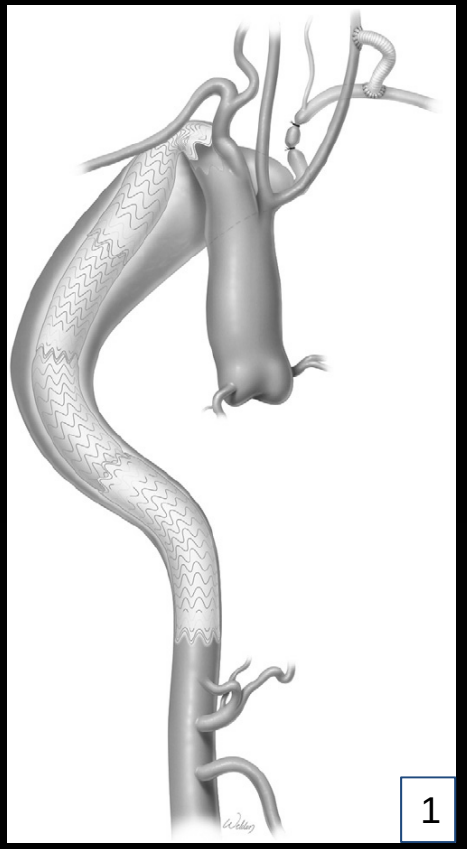

L subclav
L subclav
6.3/MIP
kv 120
mA Mod.
Rot 0.50s/HE+ 55.0mm/rot
0.6mm 1.375:1/0.6sp
Tilt: 0.0
09:18:47 AM
W = 1728 L = 532
350/5
6.3/MIP
kv 120
mA Mod.
Rot 0.50s/HE+ 55.0mm/rot
0.6mm 1.375:1/0.6sp
Tilt: 0.0
09:18:47 AM
W = 1728 L = 532
350/4

Intervention Options

- Non-Operative Conservative Management:
  - 19% of patients present with rupture
  - 53% of patients present with rupture/dissection
  - 100% associated mortality rate with rupture
  - Aggressive treatment suggested when > 3 cm
- Surgical Management:
  - Right, Left thoracotomy and/or median sternotomy
  - ± Left subclavian to carotid transposition
  - Aneurysmorrhaphy, with aortic cross-clamp and bypass
  - Occasionally, cardiopulmonary bypass required
  - 14% mortality rate

Intervention Options

- Endovascular Management
  - Only three cases reported in the literature
  - Okada et al. initially performed aortic endograft which sacrificed the left subclavian.
  - Klonaris et al. performed aortic endograft of a descending aortic aneurysm, that did not extend into the diverticulum of Komerrell
  - Naoum et al. performed aortic endograft with left carotid to subclavian bypass
  - Mortality rate of 0% in these 3 cases



Intervention Options

- Endovascular Management
  - The decision was made to place a more proximal situated endograft due to diffuse aortic ectasia
  - An initial CT angiogram of the carotids and circle of Willis was performed for preoperative planning
  - Bilateral carotid to subclavian bypass was performed prior to endovascular therapy

Carotid to Subclavian Bypass

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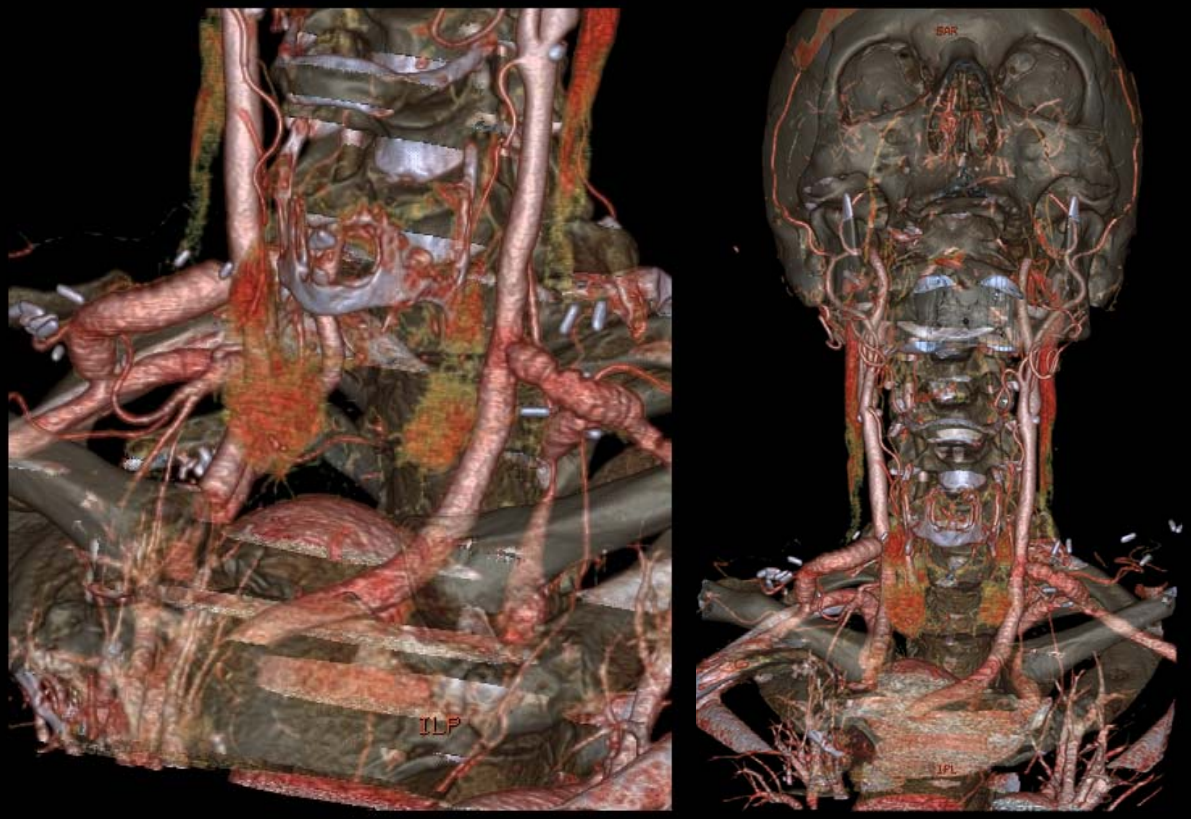
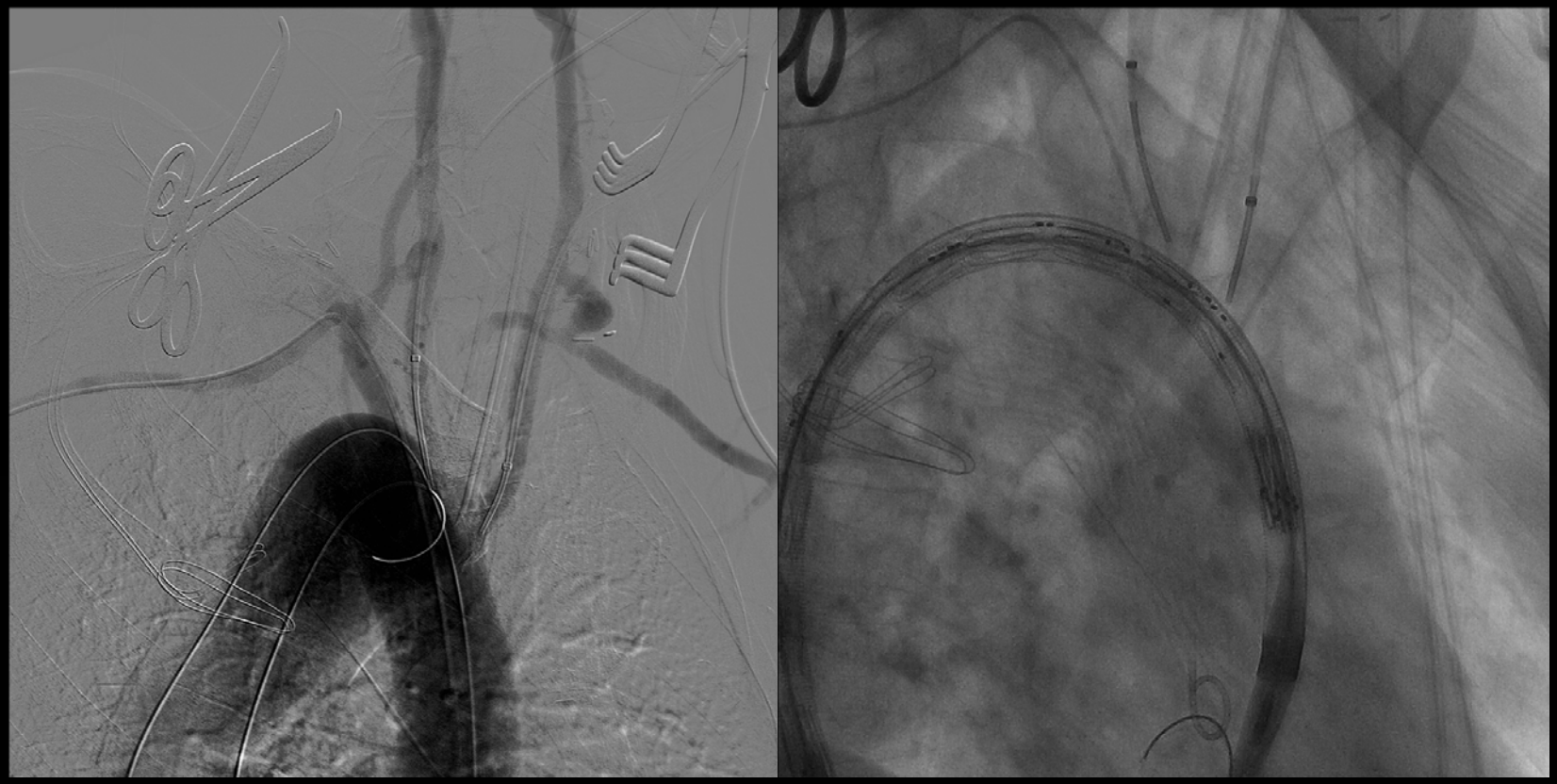
- 8 mm polyester graft on the right
- 7 mm polyester graft on the left



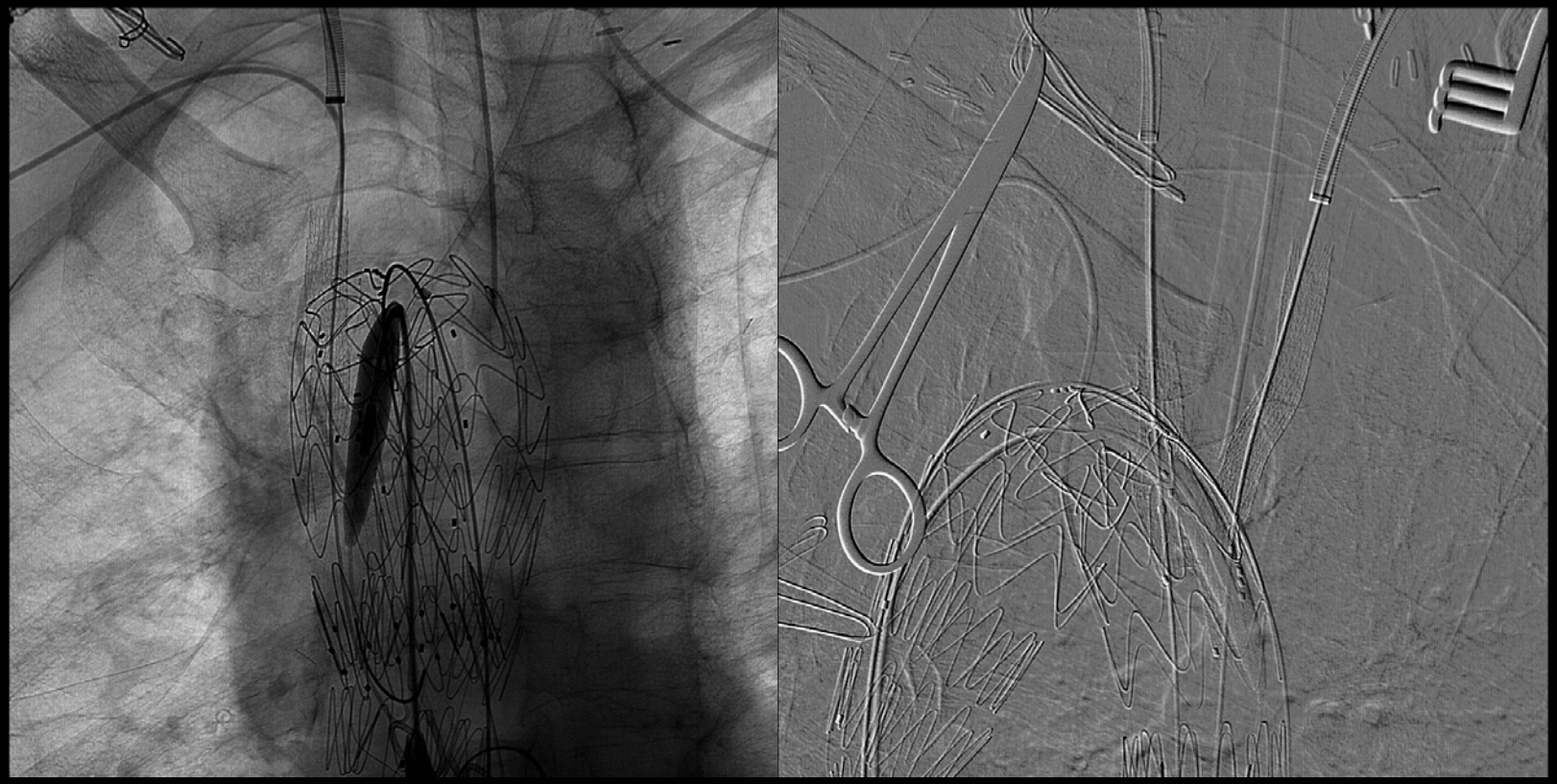
Endovascular Stent Grafting

- Intubated with General Anaesthesia
- Cardiology was involved – rapid ventricular pacing
- Bilateral femoral cutdowns were performed
- Bilateral carotid cutdowns were performed
- Bilateral brachial cutdowns were performed

Endovascular Stent Grafting



Carotid Stenting

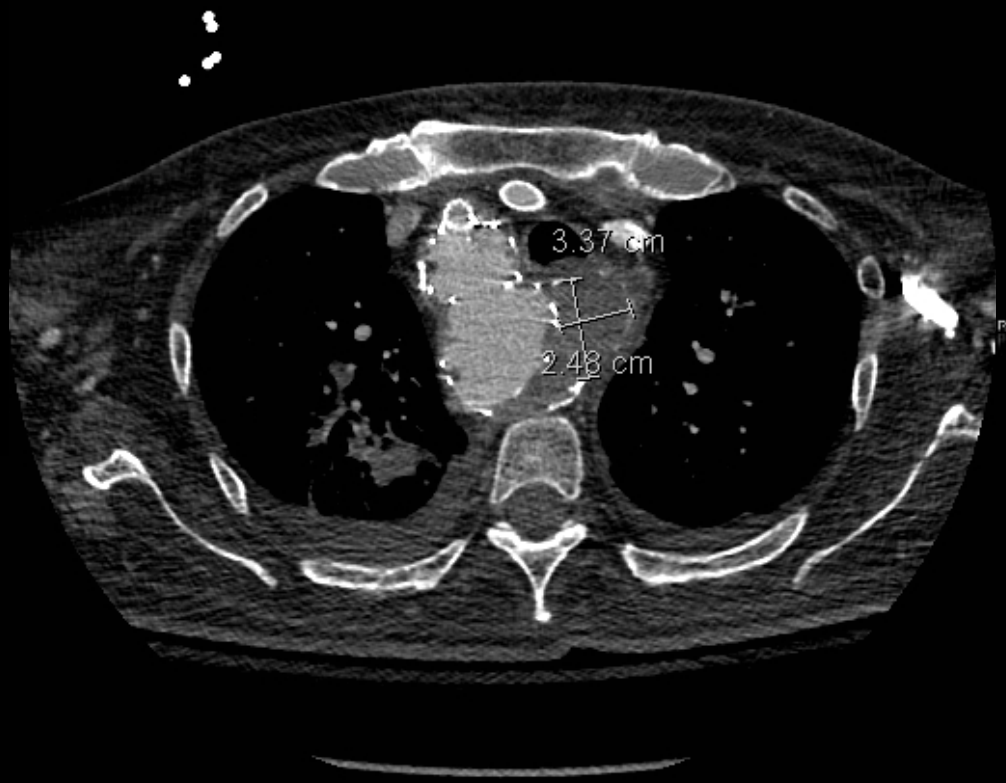


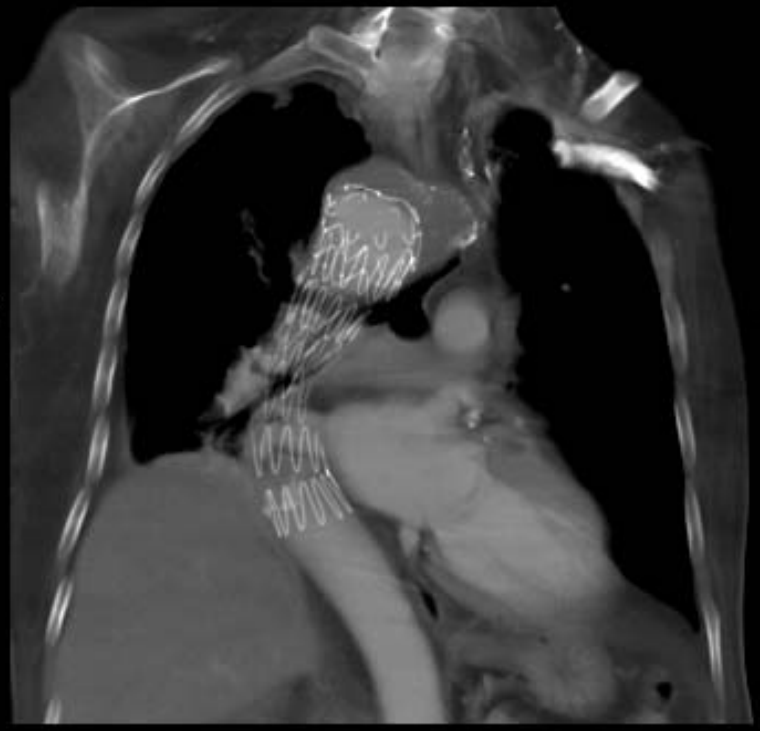
Subclavian Occlusion

Post Procedure

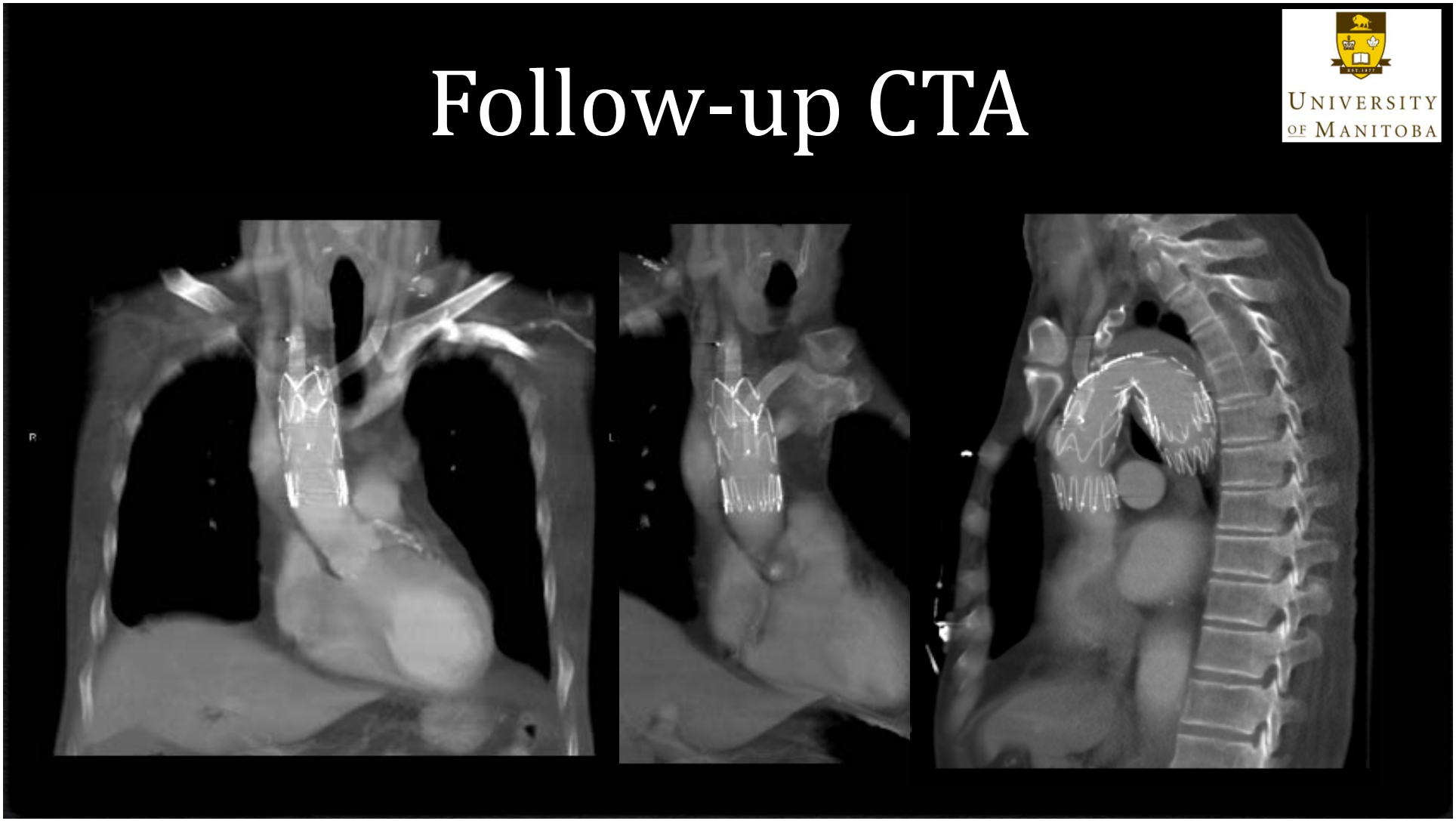
- Recovered in ICU
- Extubated neurologically intact with weakness due to his muscular dystrophy

Follow-up CTA





Follow-up CTA



Follow-up

- Symptomatically, dysphagia slowly improved
- No endoleak
- No documented complication as of 3 years

Discussion

- Right sided aortic arch – 0.05-0.1% of radiology and autopsy cases
- 50% of RAA have an aberrant left subclavian
- 60% of RAA with aberrant subclavian have a Kommerell's diverticulum

Discussion

- Clinically associated with lusoria, a combination of dysphagia and left arm paraesthesias
- Less common symptoms include dyspnea, stridor, wheezing, choking, coughing spells, obstructive emphysema, and recurrent pneumonias related to airway compression. Also may have diploplia, nystagmus, syncope, nausea, vertigo, or ataxia with homonymous hemianopsia due to subclavian steal syndrome
- Endovascular therapy presents a new treatment modality for RAA associated with lusoria or aneurysm, with potentially decreased mortality

References

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- Kimura-Hayama ET, Melendez G, Mendizabal AL et al. *Uncommon Congenital and Acquired Aortic Diseases: Role of Multidetector CT Angiography.* Radiographics (2010) 30:79-98. **[2]**
- Klonaris C, Avgerinos ED, Katsargyris A et al. *Endovascular Repair of a Right-Sided Descending Thoracic Aortic Aneurysm Associated with a Right Aortic Arch and a Left Subclavian Artery Arising from a Kommerell's Diverticulum.* Cardiovasc Intervent Radiol (2009) 32: 758-761.
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- Okada K, Sueda T, Orihashi K et al . *Endovascular stent-graft repair for thoracic aortic aneurysm associated with right-sided aortic arch.* J Thorac Cardiovasc Surg (2001) 122:185-6.
- Turkvatan A, Buyukbayraktar FG, Olcer T et al. *Congenital Anomalies of the Aortic Arch: Evaluation with the Use of Multidetector Computed Tomography.* Korean J Radiol (2009) 10: 178-184. **[3]**
- van Son JAM, Konstantinov IE. *Burckhard F. Kommerell and Kommerell's diverticulum.* Tex Heart Inst J (2002) 29: 109-12. **[4]**