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Endovascular Repair of Ruptured Type B Aortic Dissection in a 26 Week Pregnant Female with Marfan Syndrome

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Case

- 26 year-old female with history of Marfan syndrome
  - Presented to OSH c/o severe back pain
  - CTA showed a descending aortic dissection with contained rupture of a 9-cm thoracic aortic aneurysm
  - Emergent air transport to Mayo
- Past Medical History:
  - G2P1001 currently 25 2/7 weeks pregnant
  - Acute ascending dissection 2 years prior with placement of 25mm bioprosthetic AVR & ascending aortic homograft

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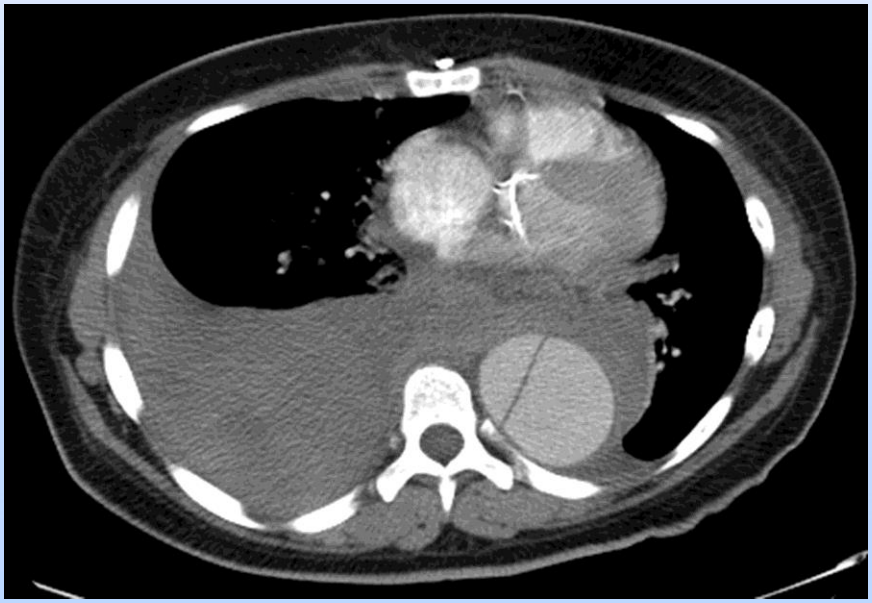
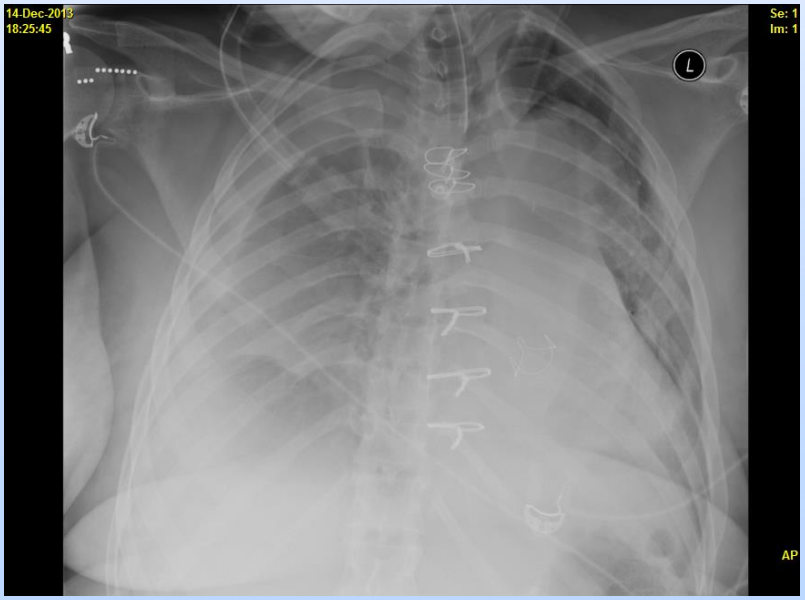
Clinical Course

- En Route – decompensated requiring intubation
  - Transfused 2 units PRBCs
- Review of outside CT confirmed rupture of Type B dissection with right hemothorax
- Repeat chest X-ray at Mayo Clinic showed temporal evolution of dissection





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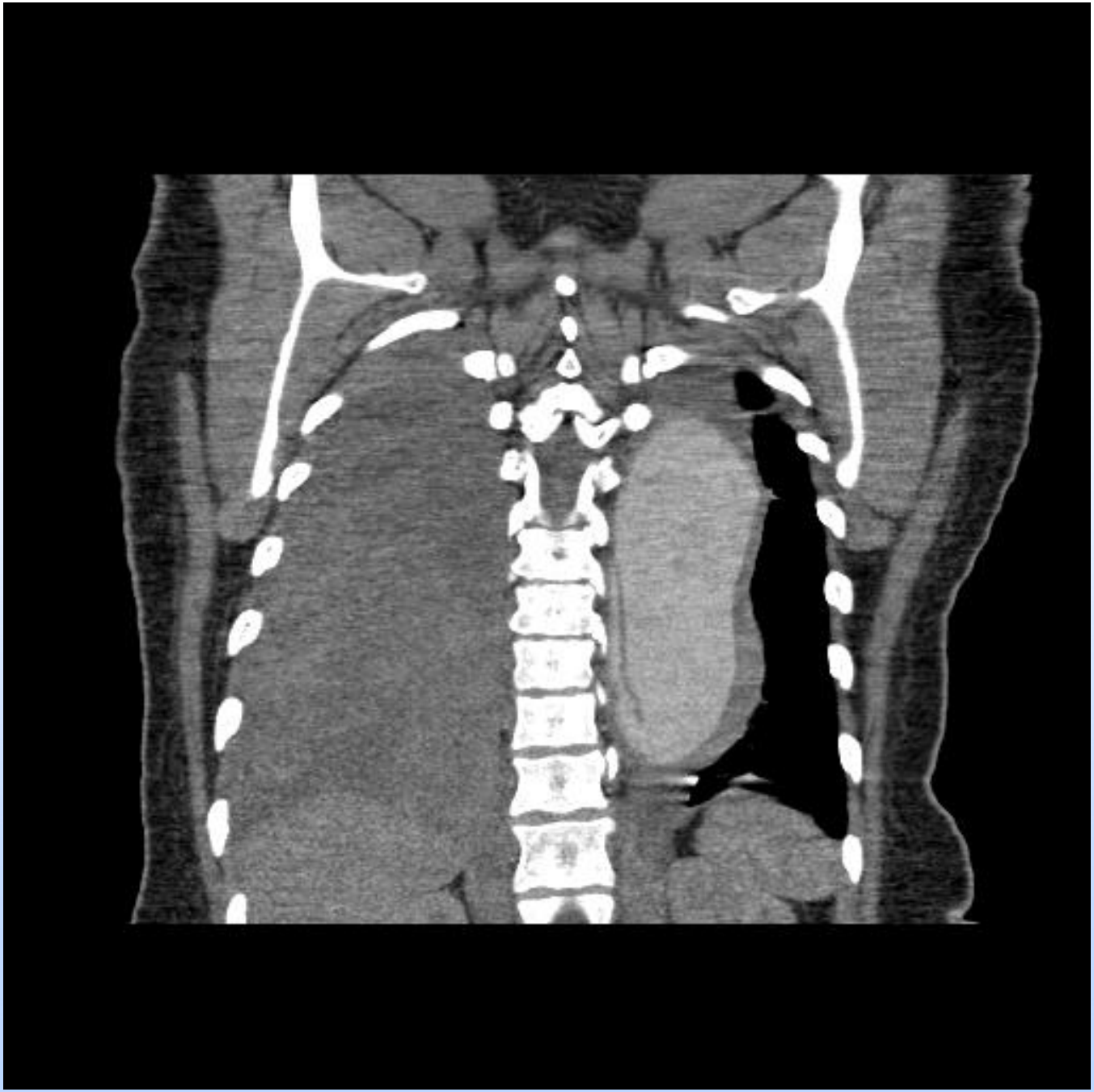

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Clinical Course (cont'd)

- Initial vitals in ED: HR 130's, right radial art line showed initial BP 130s/80s.
- Started on an Esmolol
- Emergent MFM consultation – fetal HR in the 150s good fetal motion.
- Prophylactic antenatal steroid given
- Social work involvement, discussion with father, MFM, Vascular, and CV surgery regarding fetal monitoring
- CV surgery/Vascular surgery discuss options
- Hemodynamics decompensated – 70's/60's – Esmolol discontinued
- Transported to OR for emergent repair

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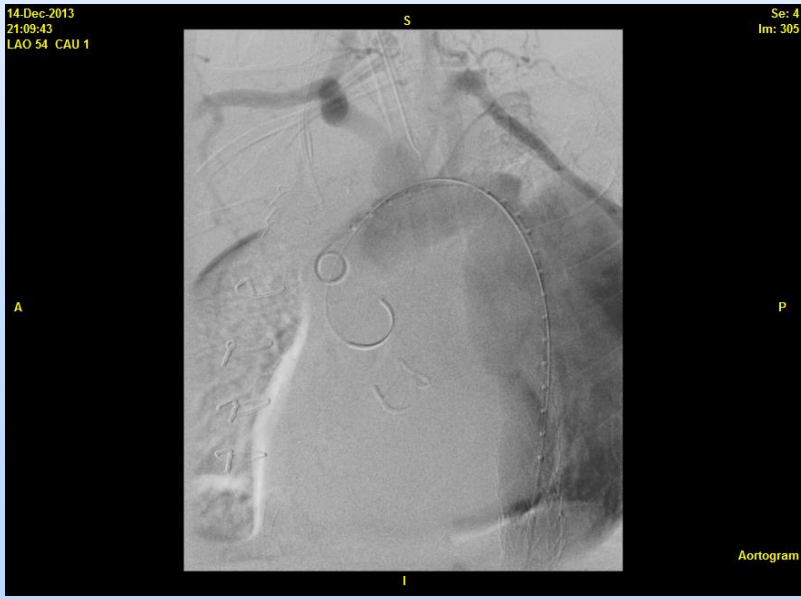
Clinical Course

- Dissection extended from the subclavian to just proximal to the celiac
- Received total of 11 units PRBC, 1 unit Platelets, 6 units of FFP, & 1 unit of cryoprecipitate
- Required vasopressor support: Norepinephrine, Epinephrine, Vasopressin
- 4 cTAG endovascular stent grafts (31x150cm, 37x100Cm and two 40x100 cm)
- Chest tube (1.9L out) and lumbar drain placed postoperatively



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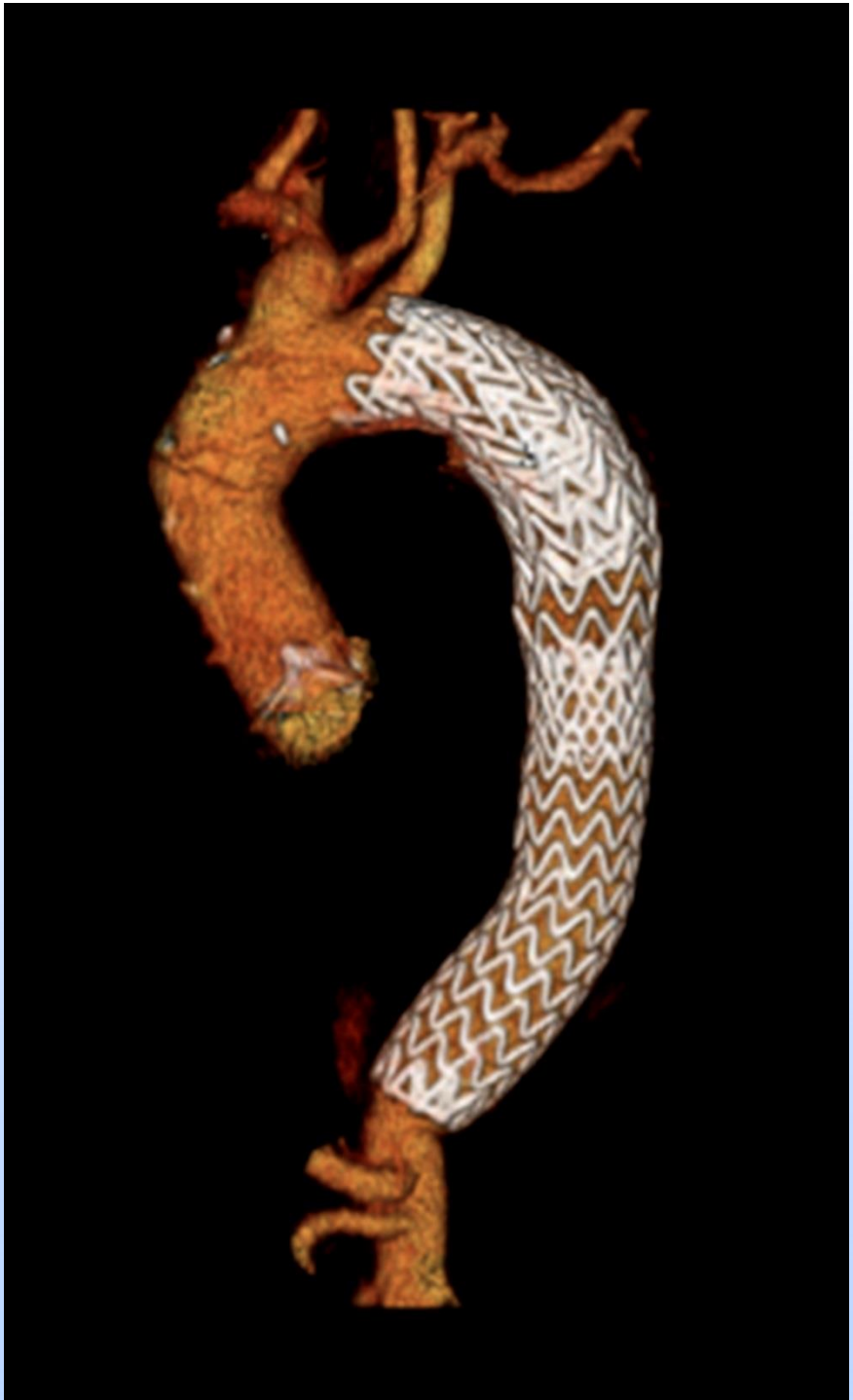

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Clinical Course

- Transferred intubated to ICU postoperatively
- Lumbar Drain removed postoperative day (POD) 1 with intact lower extremity motor function
- Fetal monitoring was carried out during the perioperative course
- Extubated on POD#3
- Magnetic resonance imaging (MRI) performed on POD#7 showed no evidence of fetal intracranial hemorrhage
- Discharged on POD#10 on a beta-blocker, aspirin, and perinatal vitamins

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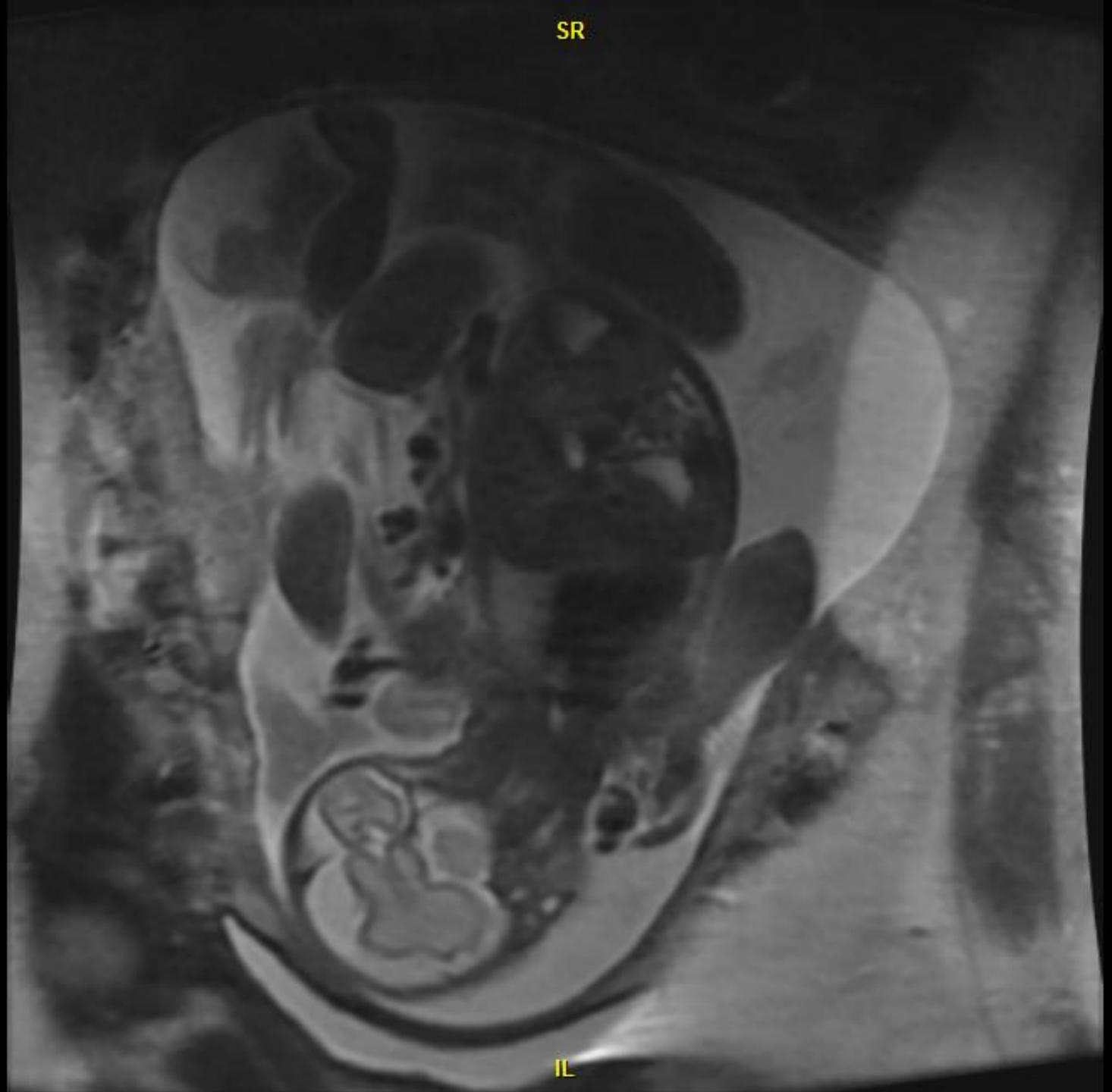
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Clinical Course

- Follow-up CTA showed type II endoleak
- Planned elective cesarean section at 31 weeks
  - Cardiopulmonary bypass on standby
  - Viable female delivered (1,650 grams, APGAR scores 2 and 8)
  - Buccal misoprostol and oxytocin for uterine atony
- 1 week post delivery - Open descending thoracic aortic repair with stent graft explantation
  - Reconstruction with 26-mm Dacron graft from the left subclavian origin to the celiac artery
  - Deep hypothermic circulatory arrest without intercostal reimplantation.

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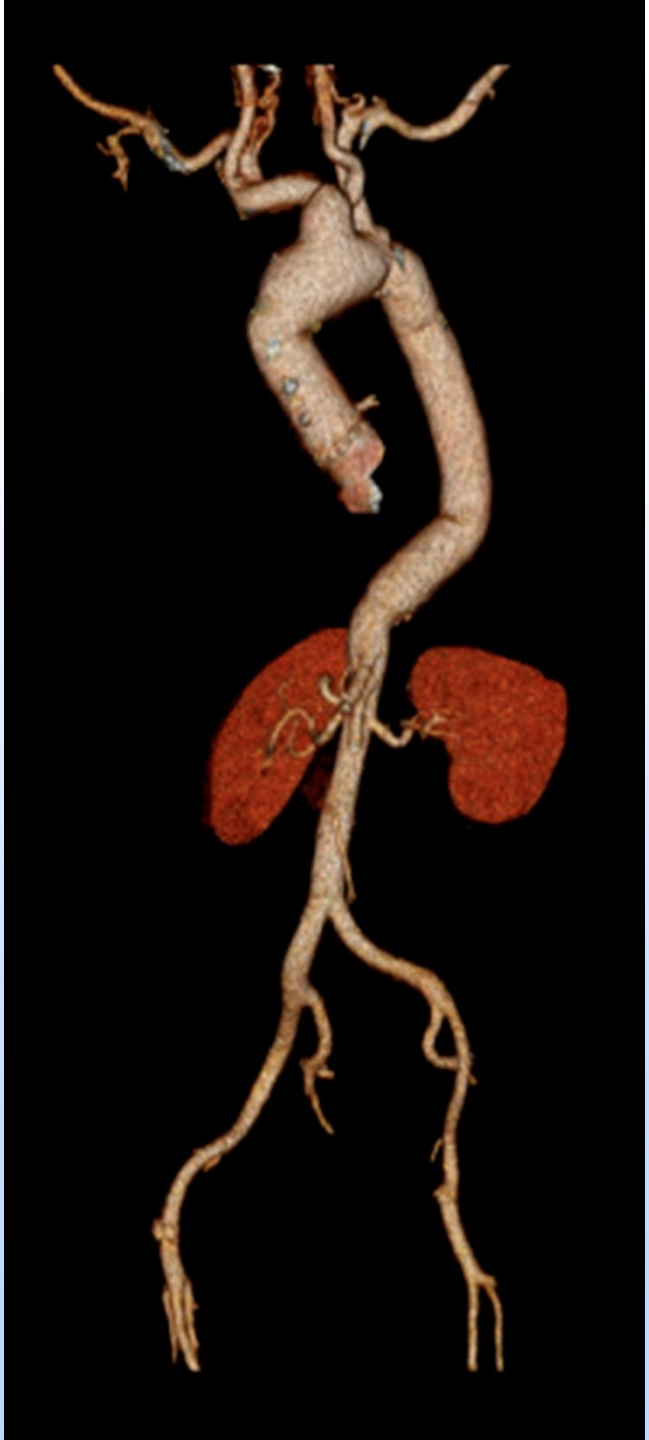

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Clinical Course

- Mother discharged on POD 7 with no complications.
- Baby discharged from neonatal intensive care unit after 41 days.
- Genetic testing on both mother and daughter showed FNB1 [fibrillin-1] frameshift gene mutations of exon 34 (c.4255dupC, p.Gln1419ProFsX12), consistent with MFS.

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Questions and Discussion

References

1. De Martino, et al. Ann Thorac Surg. 2015; 100:304–7
2. Elkayam U, Ann Intern Med. 1995;123(2):117
3. Jondeau G, Circulation. 2012; 125: 226-232
4. Miller's Anesthesia, 7th edition
5. Parodi JC, Ann Vasc Surg 1991; 5: 491-499
6. Dake MD N Engl J Med 1994; 331: 1729-1734;
7. Findeiss LK, Semin Intervent Radiol. 2011 March; 28(1): 107–117.

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Marfan Syndrome

- Inherited disorder of connective tissue
  - Incidence: 1 in 3,000-5,000
  - Autosomal dominant
  - 25–30% of cases represent new mutations
- Broad range of clinical severity
  - Various mutations involving the Fibrillin-1 (FBN1) gene located on chromosome 15
- Involves many systems but most prominent manifestations are of skeletal, ocular and cardiovascular origin

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Pregnancy in Marfan

- Pregnancy and the postpartum period is high-risk for aortic dissection and rupture in women with MFS
- Prophylactic aortic root and ascending graft repair recommended by AHA for women desiring pregnancy with diameters greater than 4 to 4.5 cm
- Cause for increased rates of dissection are unclear
  - Possible association between hyperdynamic and hypervolemic cardiocirculatory state of pregnancy and dissection.
  - Estrogen inhibits collagen and elastin deposition in the aorta
  - Progestogens shown to accelerate deposition of noncollagenous proteins in the aortas of rats.
  - Oxytocin thought to contribute to dissection risk

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Endovascular vs Open Aortic Repair

- Endovascular provides many advantages if technically feasible
- Endovascular technique first developed for AAA repair
- Use of endoluminal grafts to treat PAD began experimentally in the late 1960s
- Not until 1991, the first clinical use of the technique in five patients with AAA in Argentina
- Explosive use since those first case reports:
  - 29,542 of 61,598 of elective AAA repairs from 2001-2004 in Medicare patients

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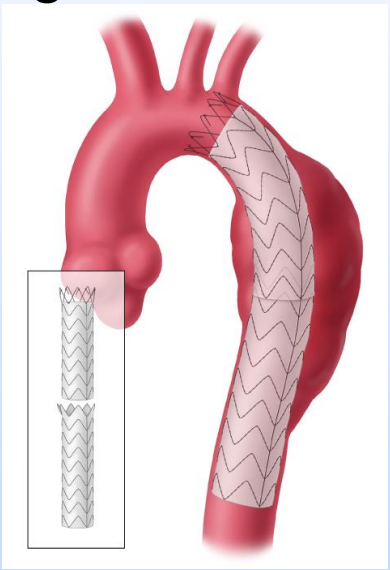
Endovascular Repair for Thoracic Dissection

- 1994 – first case report of endovascular repair of descending thoracic aneurysm
- 2005 – First FDA approved thoracic stent-graft device
- Advantages over open repair:
  - Smaller incisions
  - Avoid extensive abdominal dissections
  - Less blood loss and fluid shifts
  - Avoid aortic cross-clamp/CPB
- Severe complications:
  - Paraplegia, aortic rupture, stroke, renal failure, and respiratory failure, device failure



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