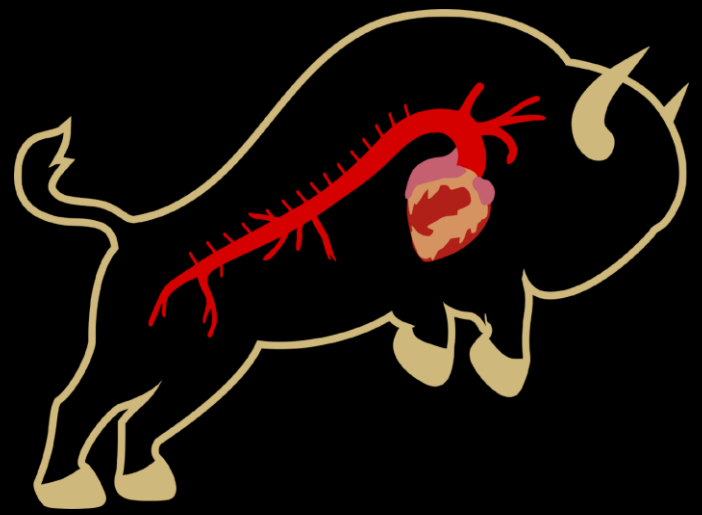
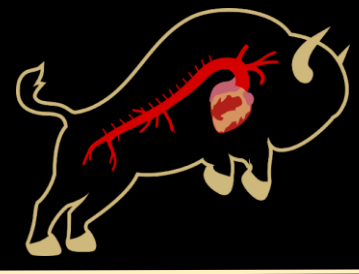
Subclavian Artery Aneurysms in Marfan Syndrome: A Clinical Dilemma

Lance Dzubinski (1), Adam Carroll (1), Rafael Malgor (1), Donald Jacobs (1), Muhammad Aftab (1), T. Brett Reece (1)
(1) University of Colorado Anschutz, Denver, CO

No disclosures



Introduction



- Cardiovascular presentations of Marfan syndrome typically present as aortopathy or valvular disease
- Any vessel can degenerate, can present with peripheral aneurysms
  - Subclavian artery aneurysms (SCAA) particularly rare, representing <1% of all peripheral aneurysms
  - Connective tissue disease associated with <10% of SCAA
- Management sparsely described given rarity of occurrence
- We describe a familial case of Marfan syndrome with SCAA

Methods

- We review two patients with Marfan syndrome and SCAA
  - 37-year-old-male and his 57-year-old-mother
- Describe presentation, operative management, post-operative course
- Re-constructed vascular pathology in 3D-slicer

Patient 1

- Presented at age 32, prior history of aortic root, mitral valve replacement
- Enlarging type B dissection, involving distal arch including left subclavian
  - Staged repair with left carotid-subclavian bypass, elephant trunk, extent 2 TAA repair
- Two years following, developed bilateral axillary aneurysms (R 4.5[3.3]cm, L 2.3[0.8]cm), and enlargement of the LSCA (2.3[1.5]cm)
- Underwent right SCA to brachial bypass
- Left subclavian included multiple branches, including vertebral
- Underwent coil embolization, appeared to have sac thrombosis

Aneurysm Reconstruction in 3D-Slicer

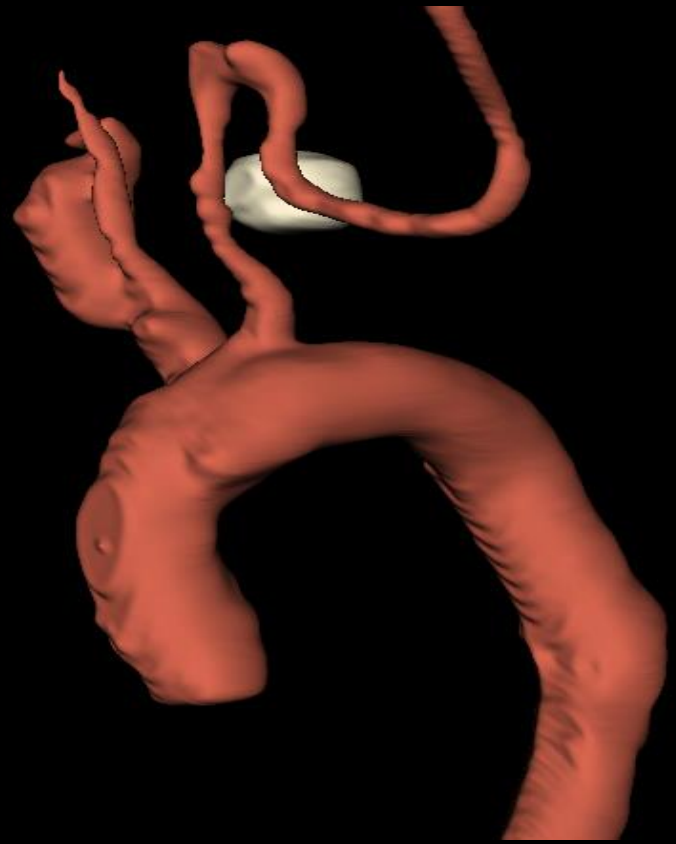
Patient 1

- Post-operative imaging at three months demonstrated ongoing flow to the aneurysm
- Had also developed left arm pain in setting of new flow-inhibiting left axillary and brachial aneurysm

Aneurysm Reconstruction in 3D-Slicer

Patient 1:

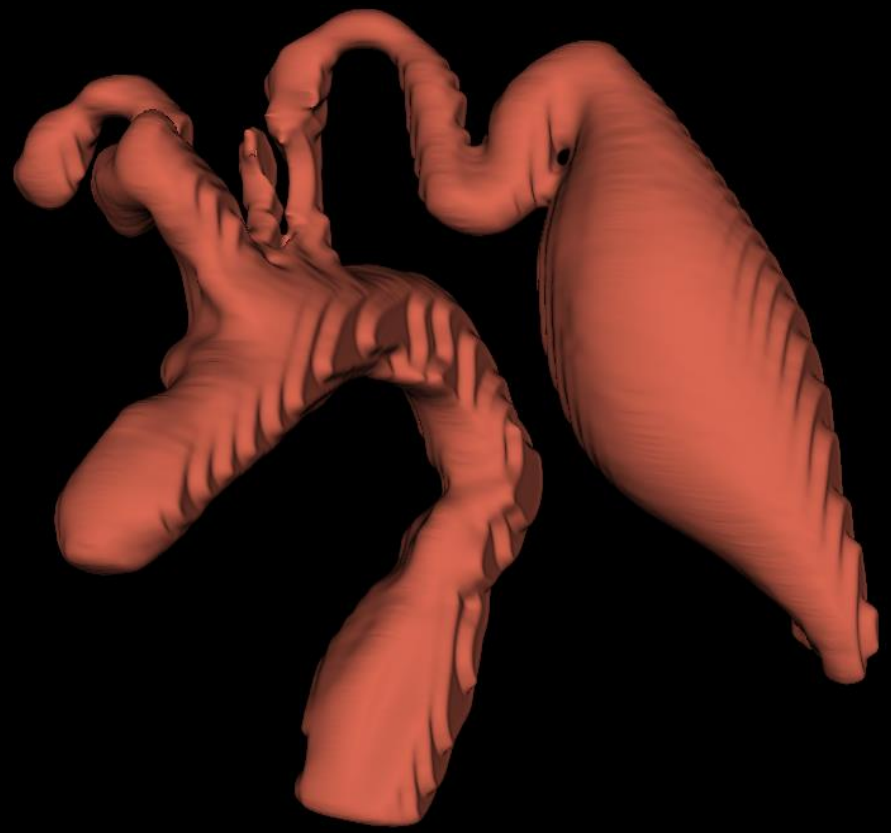
- Underwent open ligation of L SCAA, excision and bypass of AA aneurysm
- LSCAA was isolated, however, significant bleeding occurred from top of aneurysm
  - Divided off bypass, suture closed
  - Residual L SCA unsuitable for bypass, ligated
  - Residual carotid-SCA bypass sutured to axillary-brachial graft
- Following procedure, symptoms of arm ischemia had improved, despite complex operation was discharged on post-operative day 2
- Despite extensive interventions, remains with residual filling in LSCAA stump from internal mammary and thyrocervical trunk
- Pending approval of new embolic plugs, will undergo further intervention



Aneurysm Reconstruction in 3D-Slicer
(White: Residual Aneurysm Sac)

Patient 2

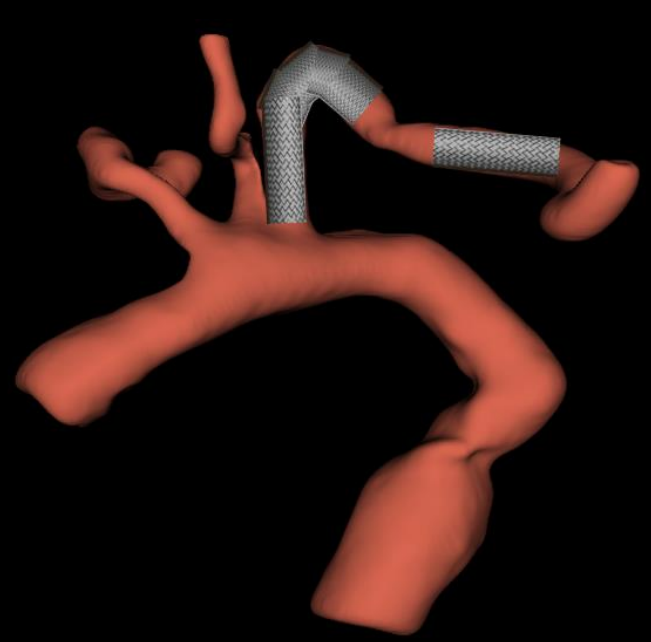
- Mother of patient 1
- Prior aortic valve, ascending aortic, hemiarch, abdominal aorta replacement
- Presented with worsening nerve compression due to expanding LSCA and massive axillary artery aneurysm measuring 4.9 x 7.8 cm



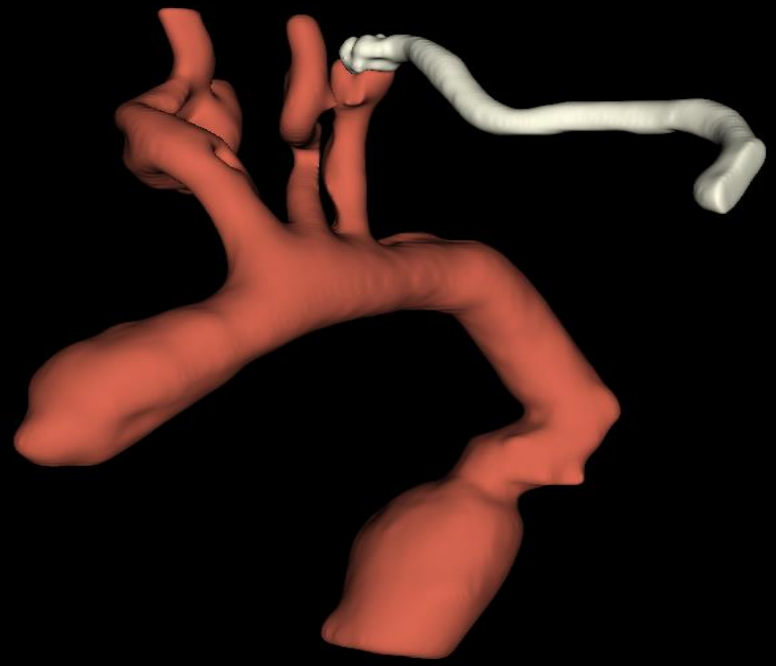
Aneurysm Reconstruction in 3D-Slicer

Patient 2

- Underwent L SCA and axillary stenting, convalescence uncomplicated
- A year later, presented with acute limb ischemia related to complete stent occlusion
- Attempted failed endovascular thrombectomy and thrombolytic therapy
- Required open thrombectomy, new graft placement with resolution

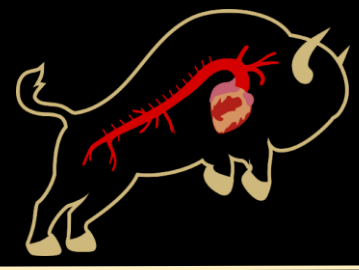


A) Post initial stenting



B) Post open thrombectomy, graft replacement

Conclusions



- Marfan patients with severe vasculopathy and peripheral aneurysms present a challenging clinical dilemma
- Often recur despite extensive operative intervention
- Require close surveillance and aggressive intervention given the potential for rapid expansion of pathology

Questions???