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MARCH 3-7, 2019

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DIAGNOSTIC AND THERAPEUTIC
APPROACHES TO VASCULAR DISEASE

Arterial Aneurysms in a Patient with Ehlers Danlos Syndrome

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DISCLOSURE

Molly Zaccardi, MHA, RVT

- No relevant financial relationship reported

Arterial Aneurysms and EDS

- Indication: 34 year old woman with known iliac artery aneurysms presents with new right calf pain
- Exams performed:
 - Ankle/Brachial Indices
 - Duplex of aorta-iliac and right lower extremity arteries
- Findings:
 - ABI on R=1.03, L=1.17
 - Duplex

Vessel	PSV	Diameter
Distal aorta	90 cm/s	2.5 cm - ectatic
Rt com iliac	82 cm/s	3.4 cm - aneurysmal
Rt ext iliac	303 cm/s	3.2 cm - aneurysmal w/ intraluminal defect

Arterial Aneurysms and EDS

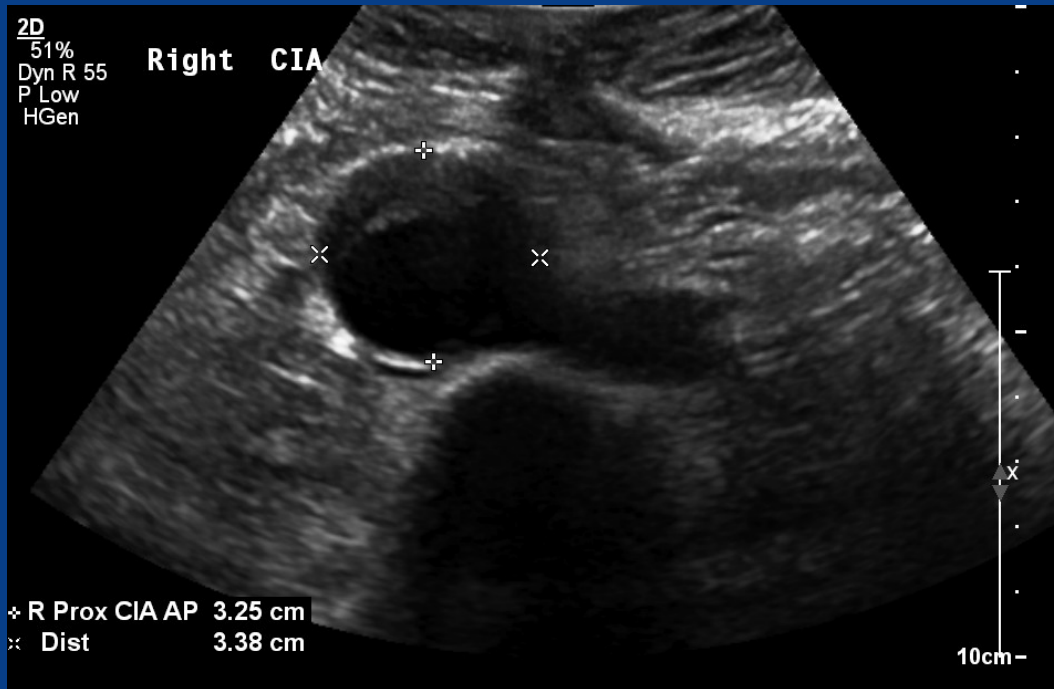
- Findings continued:
 - No evidence of stenosis in the right lower extremity arteries
 - Incidental Finding suggests dilatation of the following:

Vessel	Diameter
Superior mesenteric artery - ectatic	1.9 cm
Celiac axis - dilated	1.5 cm
Hepatic artery - aneurysmal and thrombosed	2.3 cm

Arterial Aneurysms and EDS

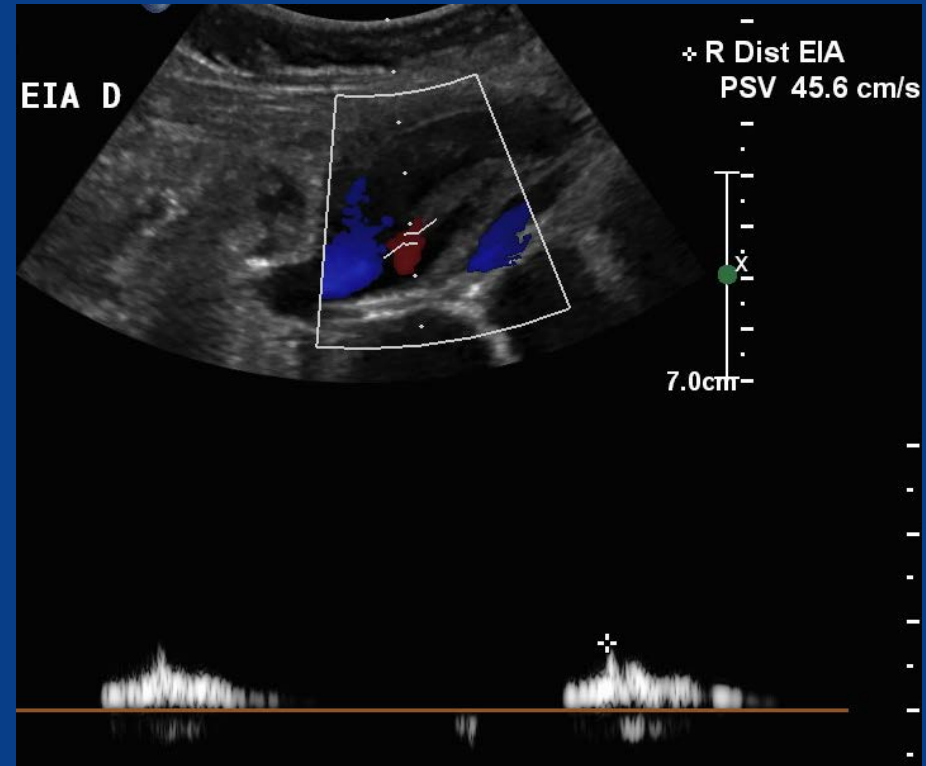
CT Angiogram

Rt Common iliac artery



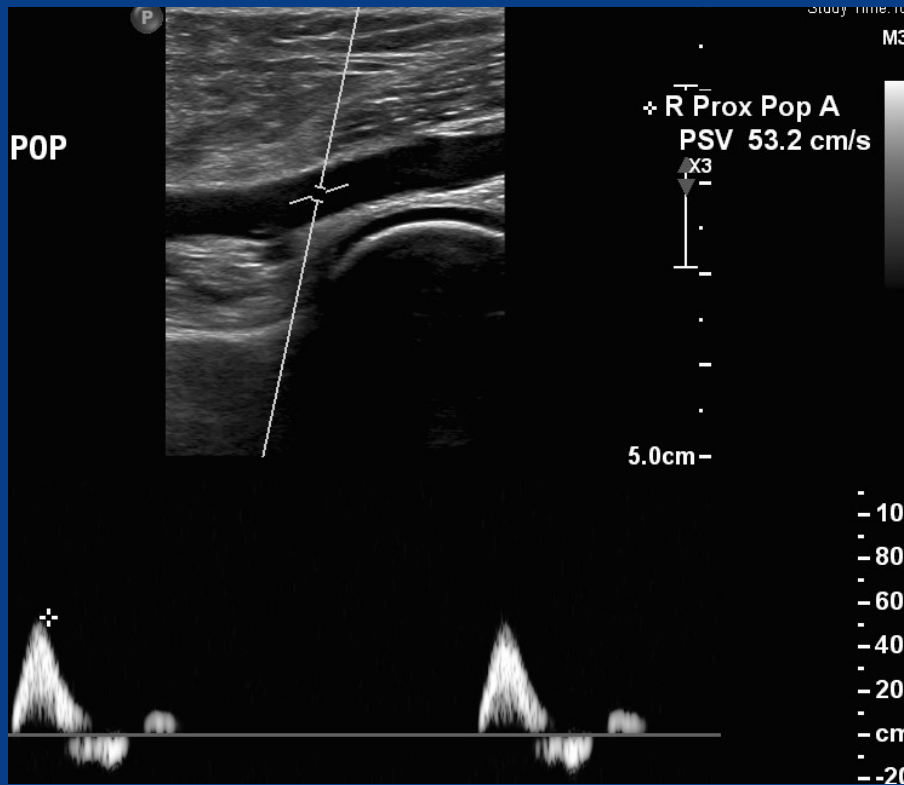
Arterial Aneurysms and EDS

Right external iliac artery

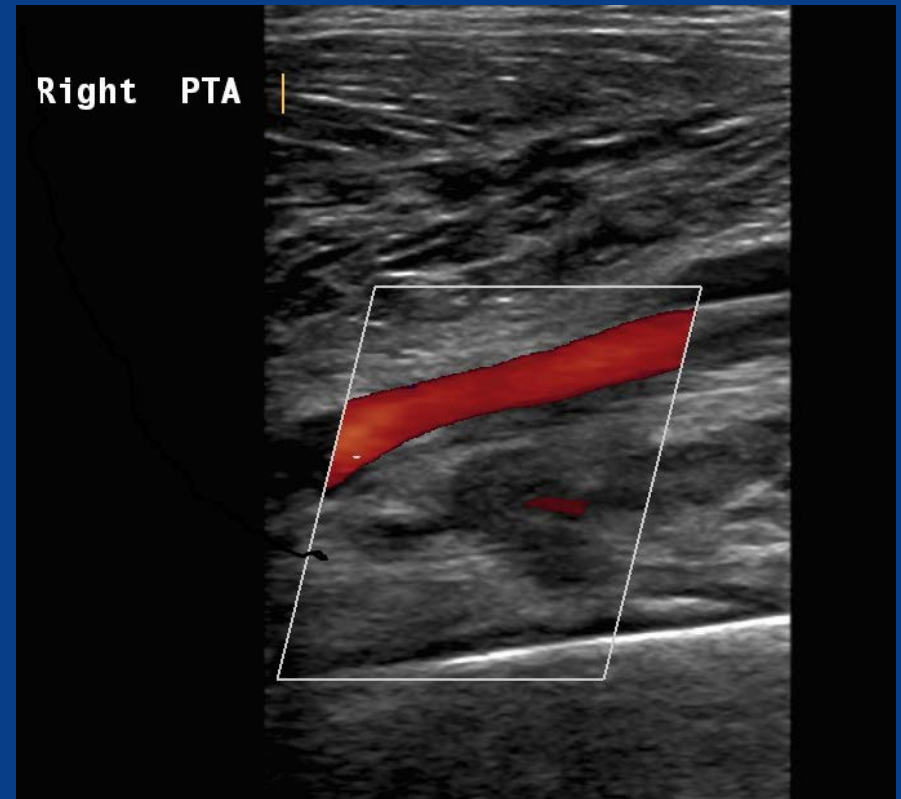


Arterial Aneurysms and EDS

Rt Popliteal artery



Rt Posterior tibial artery



Incidental Findings

Superior mesenteric artery



Celiac and hepatic arteries



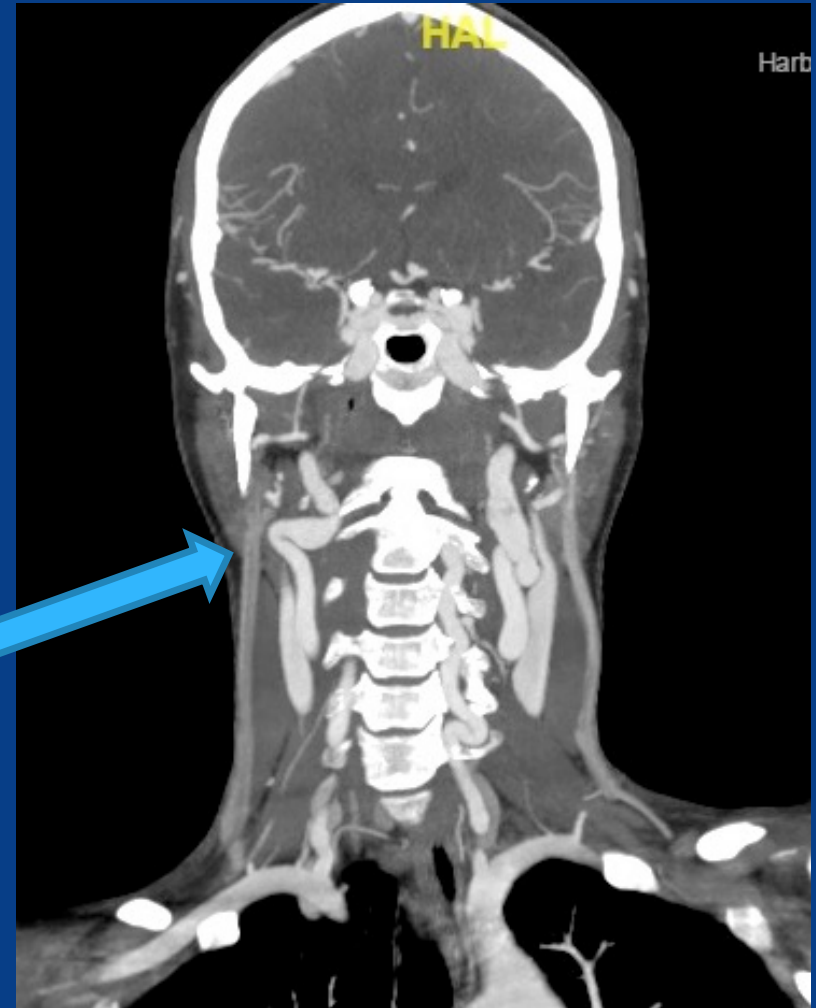
Patient history

- Headaches since age 8
- Father died at 40 - abdominal aneurysm rupture
- Uncle had EDS

- At 16-fainting spells prompted Neurology clinic visit
- Declined genetic testing

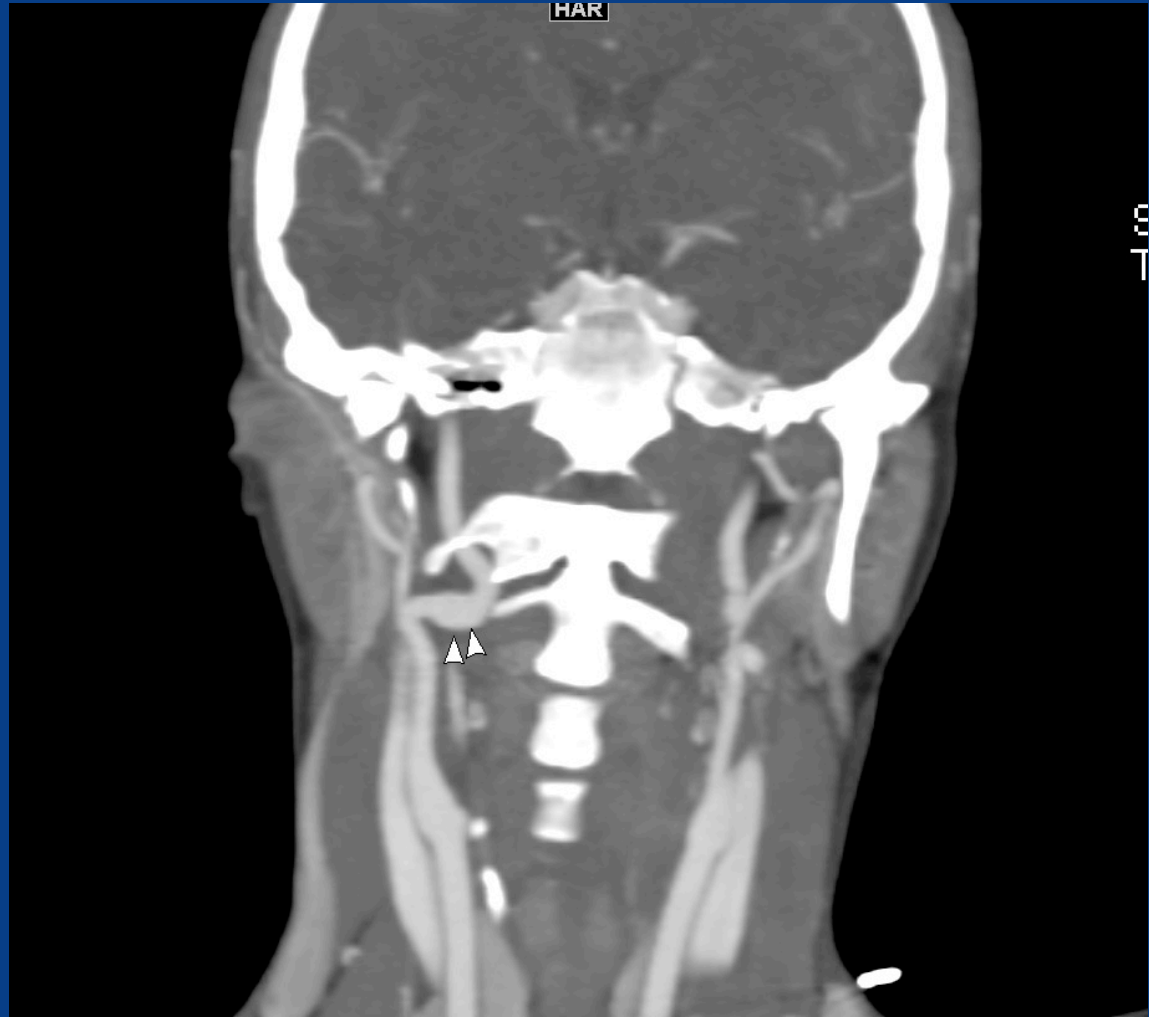
Ehlers-Danlos Syndrome

- At 22 yo, more fainting episodes prompted a cerebrovascular workup revealing bilateral internal carotid artery aneurysms suggesting Ehlers - Danlos Syndrome.



Ehlers-Danlos Syndrome

- Gene mutation
- Connective tissue disorder



Patient history

At 23 yo, more fainting spells returns to Neurology clinic and reported interest in sky diving

Recommendation against vigorous exercise.

Repeat imaging

- Aortogram – ectatic aorta
- Carotid arteries – aneurysmal and dissecting
- Internal carotid artery pseudoaneurysm
- Vertebral may have stenosis, pseudoaneurysm or dissection

At 24 yo, returned for follow up in Genetics clinic.

- Brother diagnosed with EDS and suffered a colonic rupture and aortic rupture.
- Patient is “doing well from a physical standpoint but not quite as well from a psychological standpoint”
- Decided against having children
- Was encouraged to stop smoking
- Recommend to monitor blood pressure

Patient history

- At 25 yo, repeat imaging following a MVA
 - carotid and iliac aneurysms are progressing in diameter
 - right external iliac has dissected
- At 32 yo, hospitalized for a renal artery dissection and renal infarct
- At 33 yo, embolization of a carotid cavernous fistula

Vascular History-Summary

- Embolization of left carotid cavernous fistula
- Right renal infarct
- Bilateral internal carotid artery ectasia
- Tortuosity and ectasia of the bilateral vertebral arteries
- Left subclavian artery aneurysm
- Celiac artery aneurysm
- Splenic artery thrombosis with splenic infarct
- Splenic artery branch aneurysm
- Common hepatic artery aneurysm-now thrombosed
- Superior mesenteric artery dissection with stenosis
- Superior mesenteric artery branch aneurysm
- Right renal artery aneurysm with dissection
- Left renal artery stenosis and aneurysm
- Ectasia of the distal aorta
- Dissection of the distal aorta into the right common iliac artery
- Right common and external iliac artery aneurysms with dissection
- Left external iliac artery aneurysm and dissection

Ehlers Danlos Syndrome

Name	Type	
Hypermobility EDS	3	Can suffer from osteoarthritis and pain from frequent joint dislocations
Classical EDS	1	Similar to type 3 but with skin involvement
Vascular EDS	4	Affects 1 in 50K-200K 25% develop significant health problems by age 20 Life threatening complications by age 40
Kyphoscoliosis EDS	6	Curvature of the spine and muscle weakness
Arthrocholasia	7A	Hip dislocation
Dermatosparaxis	7C	Fragile sagging skin

Recommendations for our patient with EDS

- Protect blood vessels
- **Monitor blood vessels**
- Monitor blood pressure
- Avoid smoking

Thank you for your attention



Pacific Northwest Endovascular Conference 2018 - Vascular Ultrasound

- 4:35 *Portal Duplex – A Case Study* by *Bonnie Brown RVT, Pacific Vascular, Inc*
- 4:40 *Interesting Case from the V.A.* by *Janelle Barnes, RVT Veterans Administration*
- 4:45 *EVAR – Duplex Follow up* by *Jim Brown RVT, Pacific Vascular, Inc*
- 4:50 *Comparison of Reflux Testing Methods* by *Mariah Elliott RVT, University of Washington Medical Center*
- 4:55 *Usefulness of Ultrasound in Stroke* by *Anne Moore RVT, Harborview Medical Center*
- 5:00 *Everything but the Kitchen Sink: The Utility of Extravascular Ultrasound* by *Jill Sommerset RVT / Yolanda Vea, MD, PeaceHealth*
- 5:10 *Arterial Aneurysms in a patient with Vascular Ehlers-Danlos Syndrome* by *Watson Smith RVT, University of Washington Medical Center*
- 5:15 *TCD and EC-IC Bypass* by *Ahmad Siyar, Bellevue College Student*
- 5:20 *Cystic Adventitial Disease* by *Sara Van de Roovaart, Seattle University Student*
- 5:25 *Arteriovenous Fistula Aneurysm Assessment* by *Sherene Shalhub, MD, University of Washington Medical Center*
- 5:35 *Two Point Compression for DVT* by *Susanna Shinn, MD – University of Washington at Valley Hospital*
- 5:45 *Panel Discussion*