AMSER Case of the Month December 2022

62 y.o. female with history of AAA found to have a congenital portosystemic shunt

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Patient Presentation

HPI: 62 y.o. female with history of AAA on surveillance with ultrasound since 2014

PMH: No other medical hx. Not on ASA/statin

FamHx: Hx AAA & repair in both parents

SurgHx: C-section in 1984 and 1986

SocHx: Currently smoking, 1 PPD for 40y

Physical Exam: Vitals: BP 122/69, Pulse 70

GI: Obvious pulsatile mass present predominately 2cm right of midline in epigastric/periumbilical area. No bruits on auscultation.

What Imaging Should We Order?

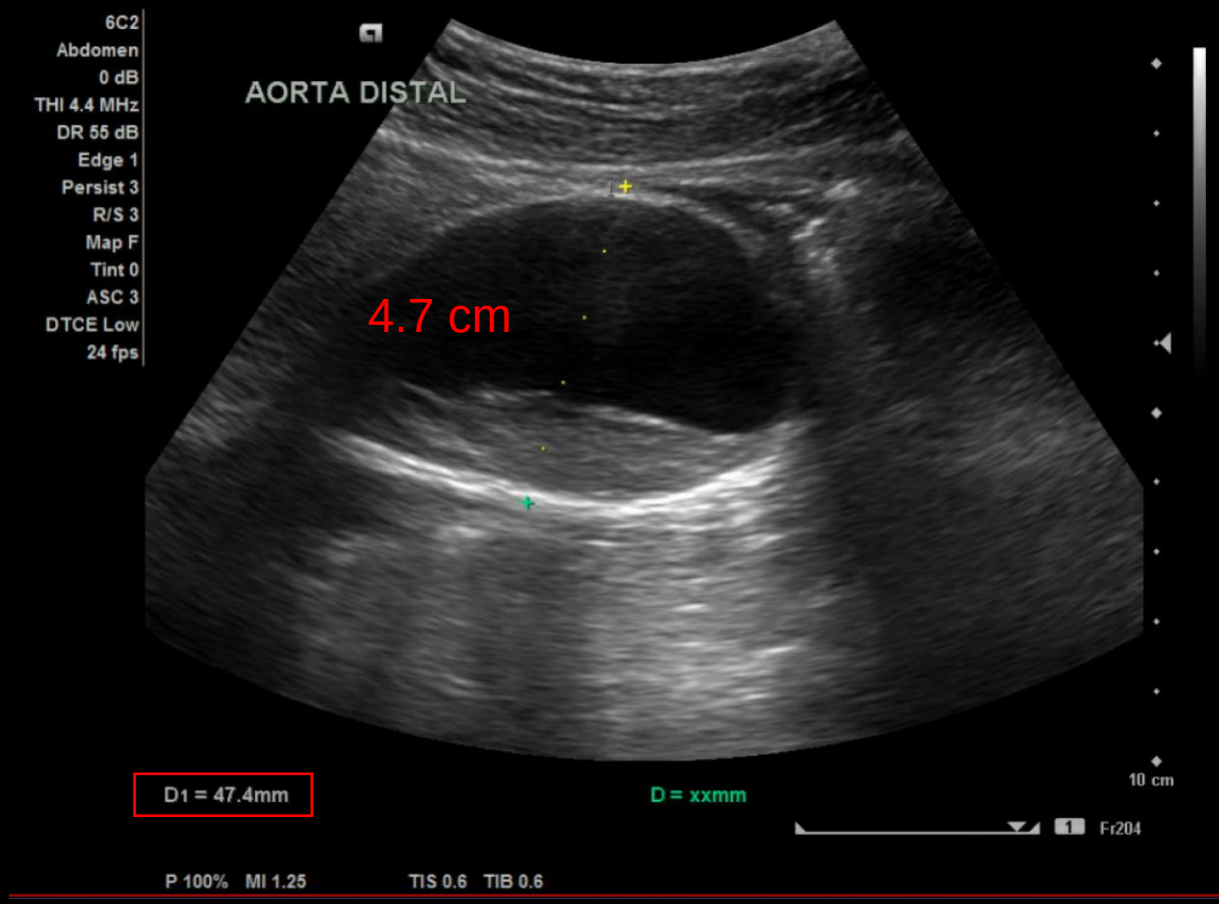
Suspected AAA – ACR appropriateness criteria

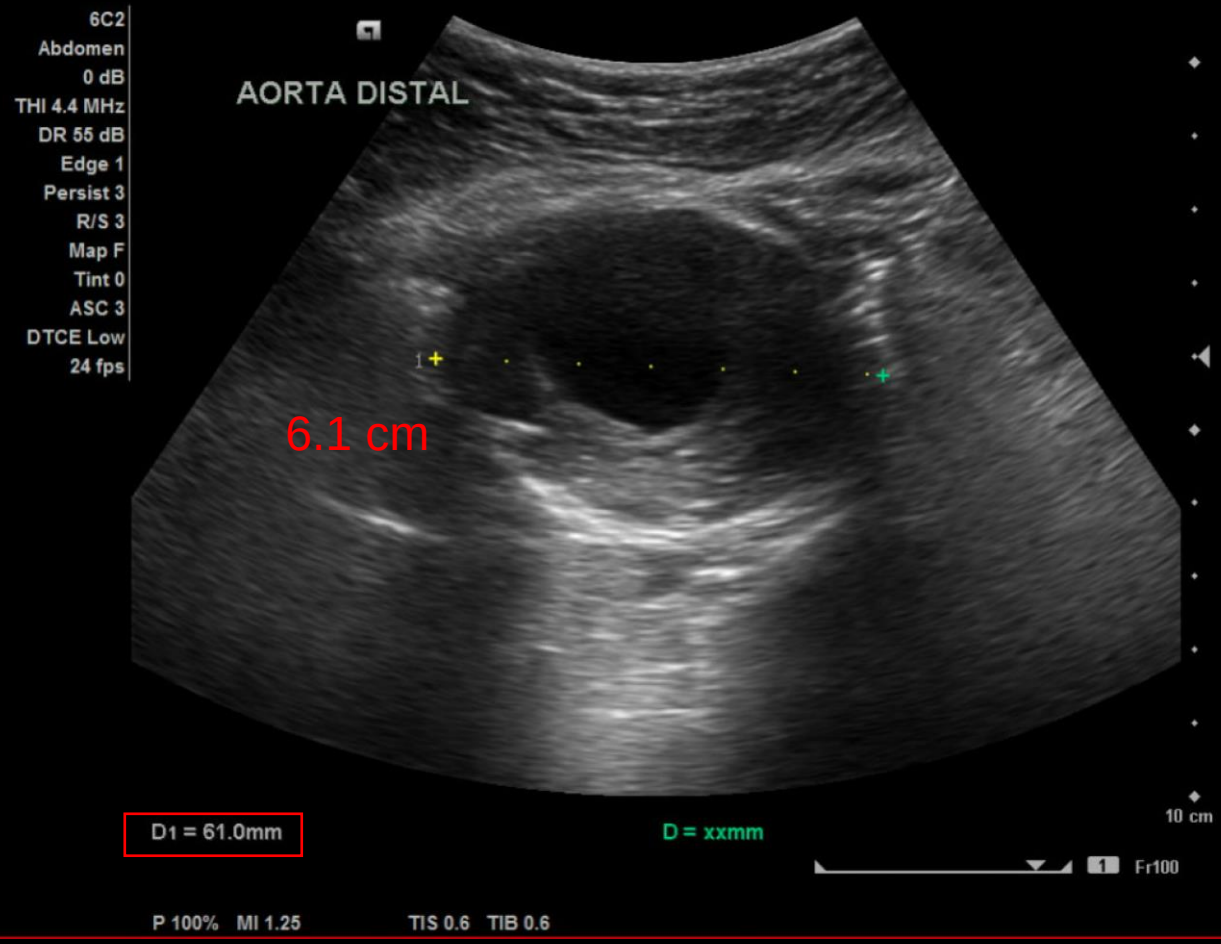
Variant 1: **Pulsatile abdominal mass, suspected abdominal aortic aneurysm.**

Radiologic Procedure	Rating	Comments	RRL*
US aorta abdomen	9		O
CTA abdomen with IV contrast	8		☢☢☢
MRA abdomen without and with IV contrast	8		O
CT abdomen without IV contrast	7		☢☢☢
CT abdomen with IV contrast	7		☢☢☢
CT abdomen without and with IV contrast	7		☢☢☢☢
MRA abdomen without IV contrast	7		O
Aortography abdomen	4		☢☢☢
FDG-PET/CT abdomen	2		☢☢☢☢

Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate

*Relative Radiation Level


6C2
Abdomen
0 dB
THI 4.4 MHz
DR 55 dB
Edge 1
Persist 3
R/S 3
Map F
Tint 0
ASC 3
DTCE Low
24 fps
AORTA DISTAL
4.7 cm
D1 = 47.4mm
D = xxmm
10 cm
Fr204
P 100% MI 1.25
TIS 0.6 TIB 0.6

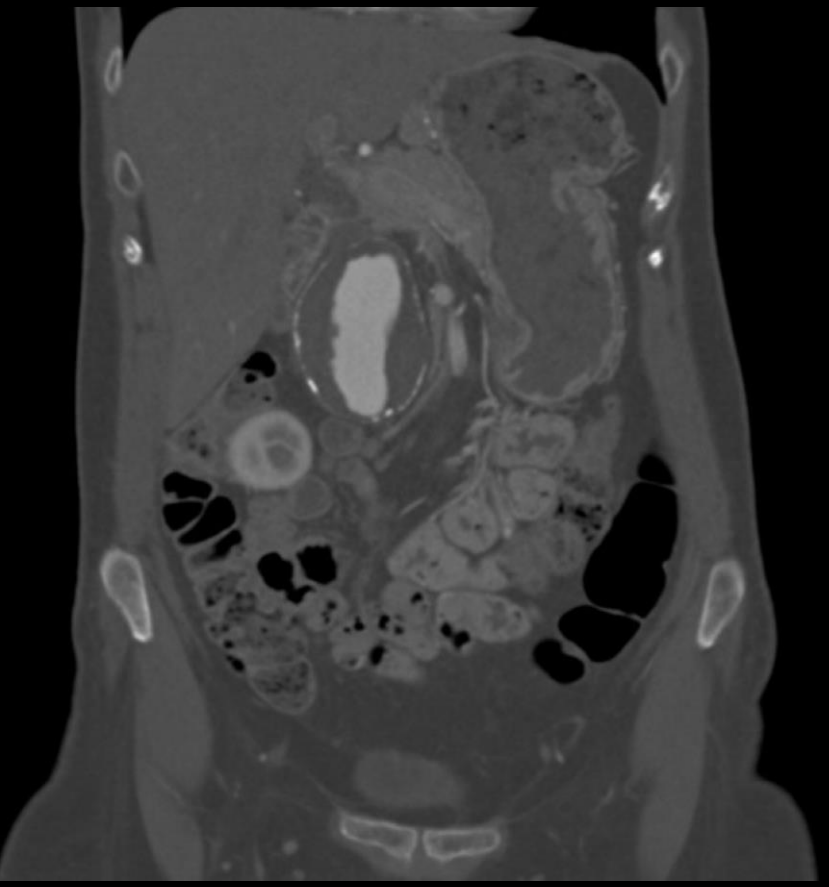

6C2
Abdomen
0 dB
THI 4.4 MHz
DR 55 dB
Edge 1
Persist 3
R/S 3
Map F
Tint 0
ASC 3
DTCE Low
24 fps
AORTA DISTAL
6.1 cm
D1 = 61.0mm
D = xxmm
10 cm
Fr100
P 100% MI 1.25
TIS 0.6 TIB 0.6

Interventional Planning for AAA

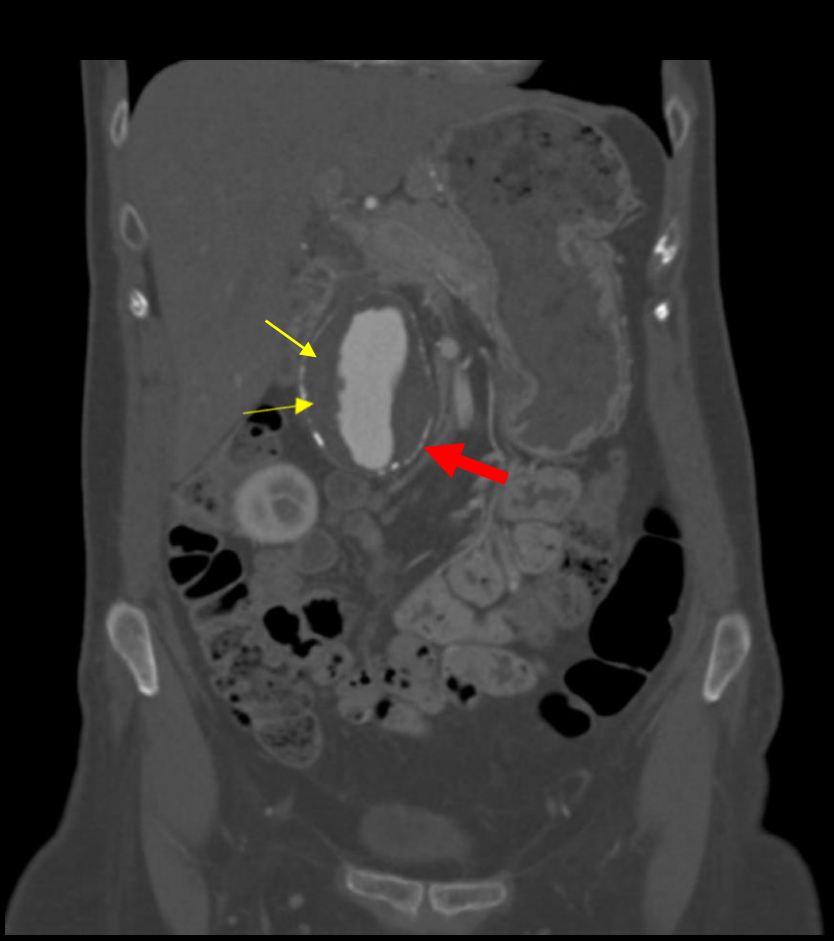
Variant 1: **Planning for pre-endovascular repair (EVAR) or open repair of AAA.**

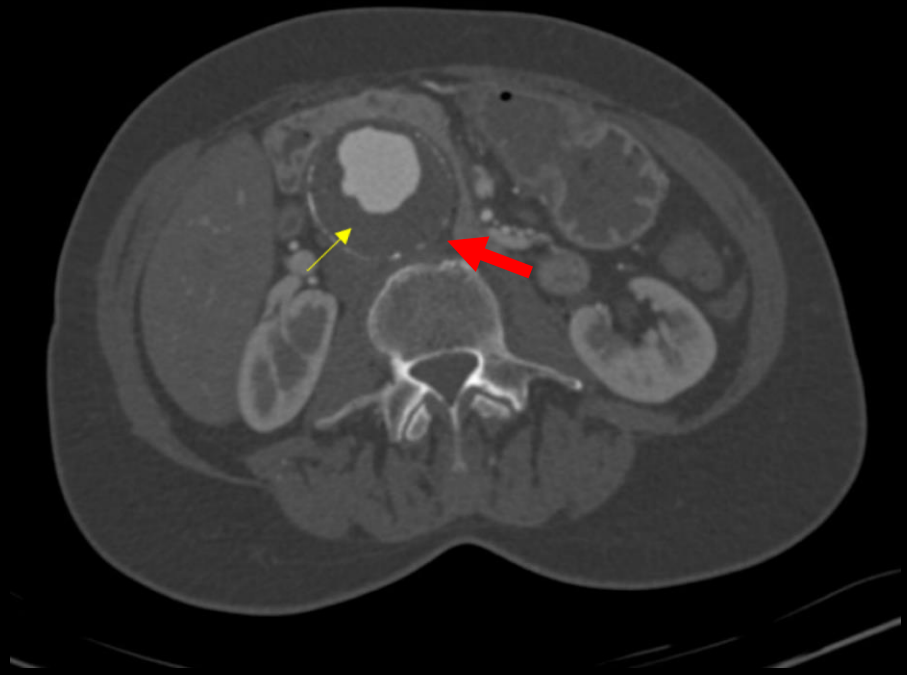
Procedure	Appropriateness Category	Relative Radiation Level
CTA abdomen and pelvis with IV contrast	Usually Appropriate	☢☢☢☢
MRA abdomen and pelvis without and with IV contrast	Usually Appropriate	O
MRA abdomen and pelvis without IV contrast	May Be Appropriate	O
CT abdomen and pelvis with IV contrast	May Be Appropriate	☢☢☢
CT abdomen and pelvis without IV contrast	May Be Appropriate	☢☢☢
Aortography abdomen	May Be Appropriate	☢☢☢
CT abdomen and pelvis without and with IV contrast	May Be Appropriate	☢☢☢☢
US aorta abdomen with duplex Doppler	Usually Not Appropriate	O
X-ray abdomen and pelvis	Usually Not Appropriate	☢☢☢
CT abdomen and pelvis without IV contrast and US aorta abdomen with duplex Doppler	Usually Not Appropriate	☢☢☢

Findings (unlabeled)



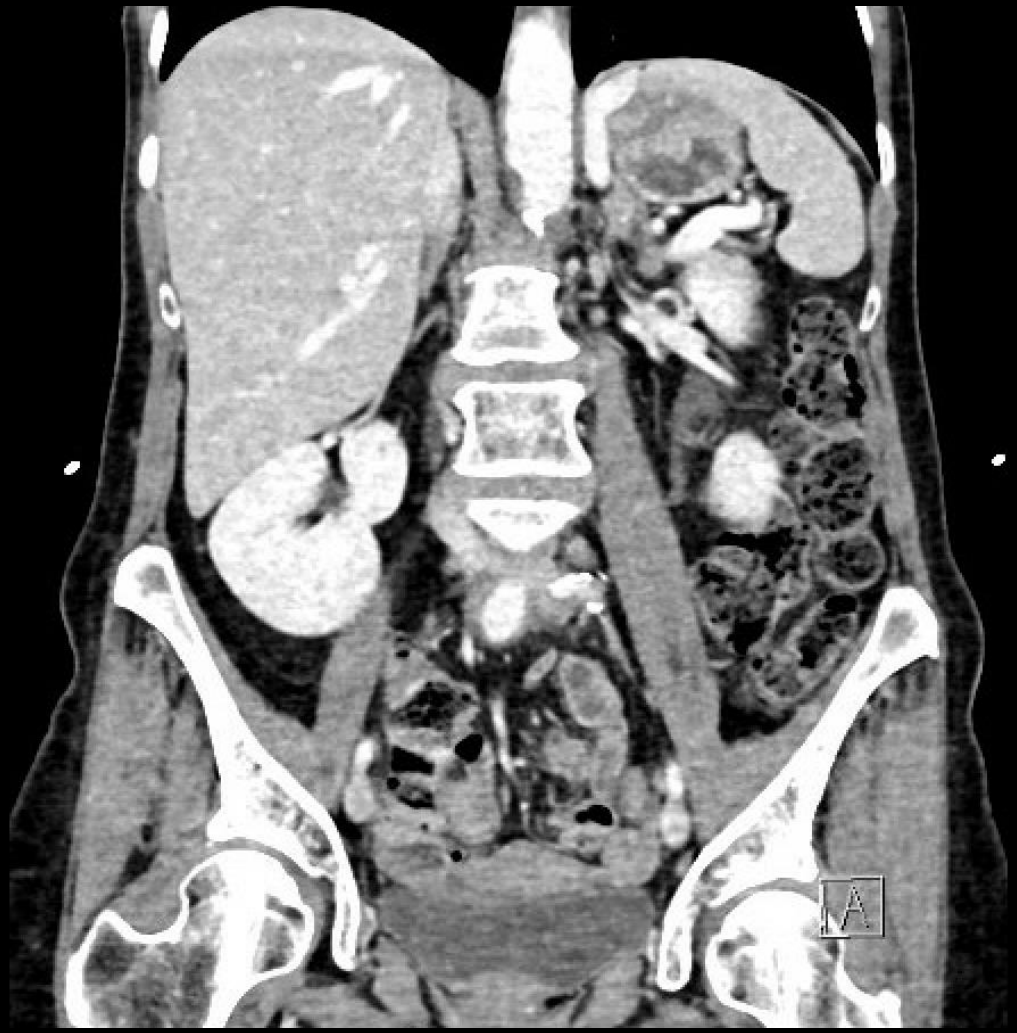
Findings: (labled)



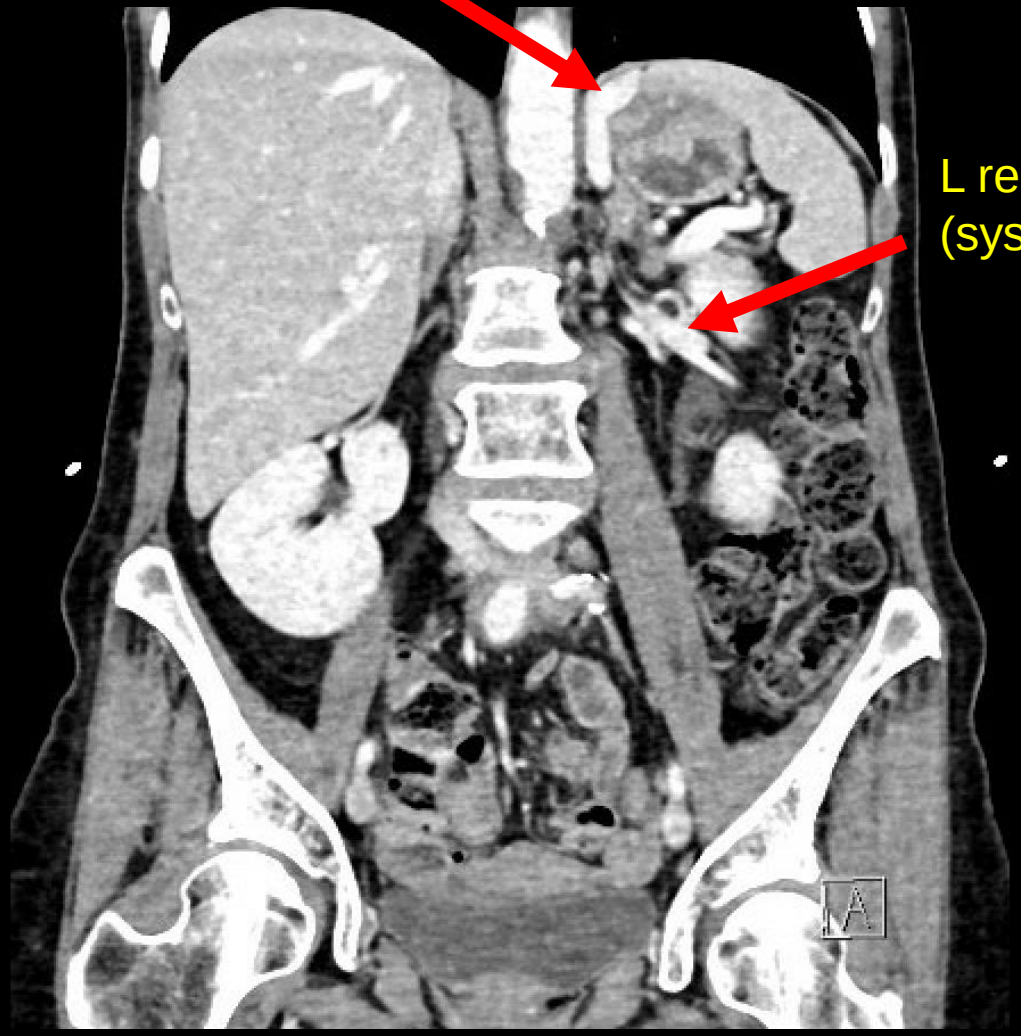


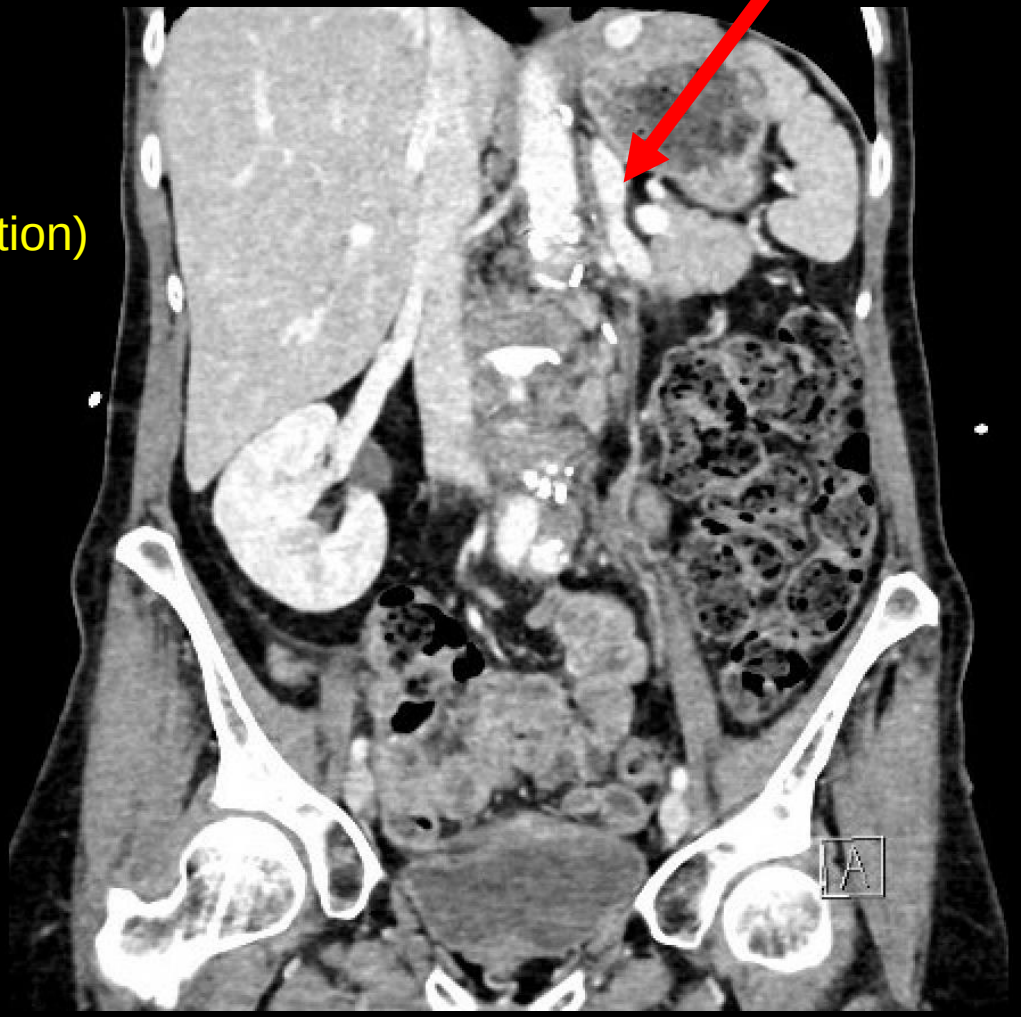
5.0 cm x 4.6 cm Juxtarenal saccular AAA (↑) w/ extensive mural thrombus (↑)

Findings (unlabeled)



Findings (labeled)





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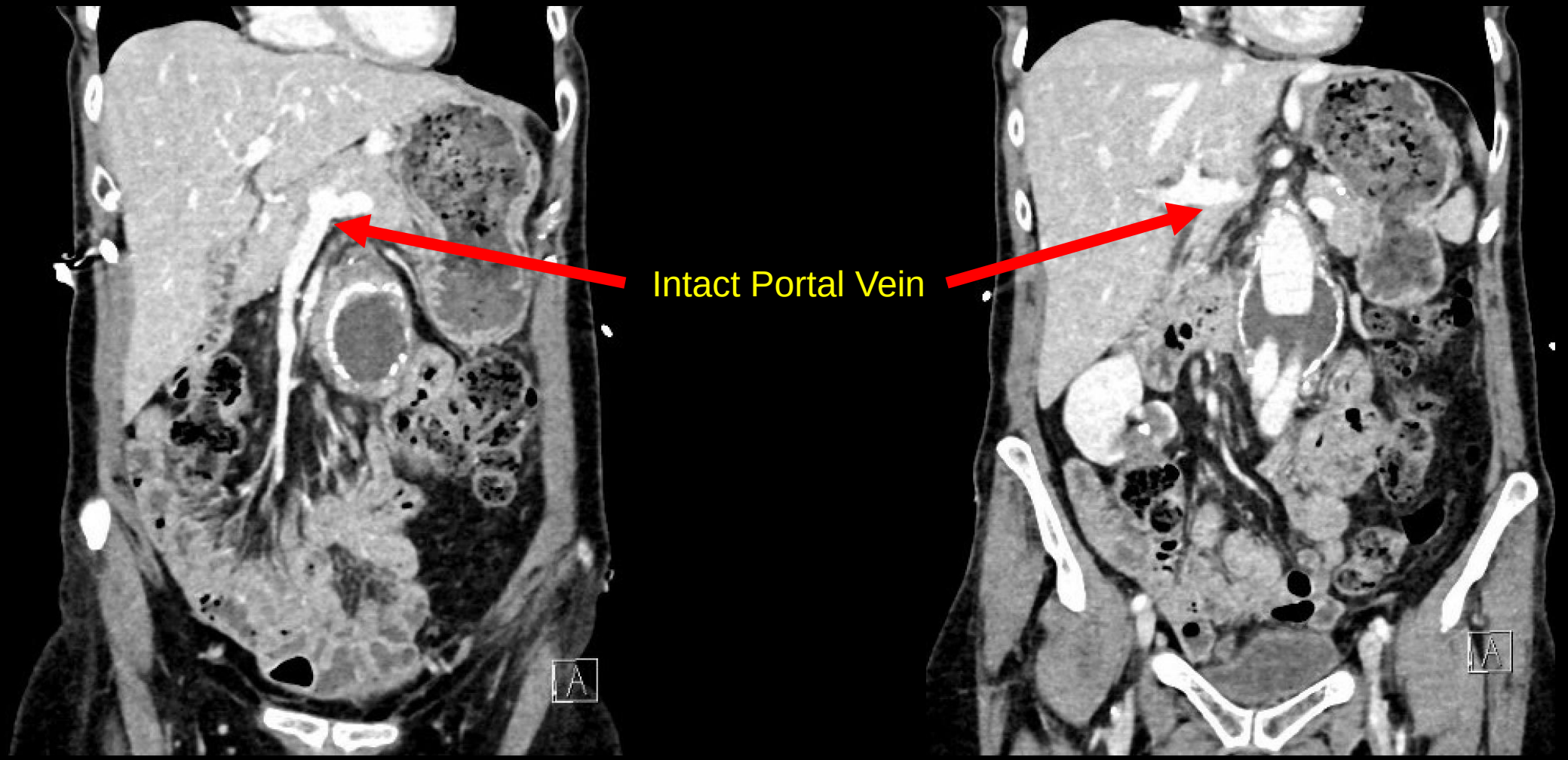
Findings (unlabeled)

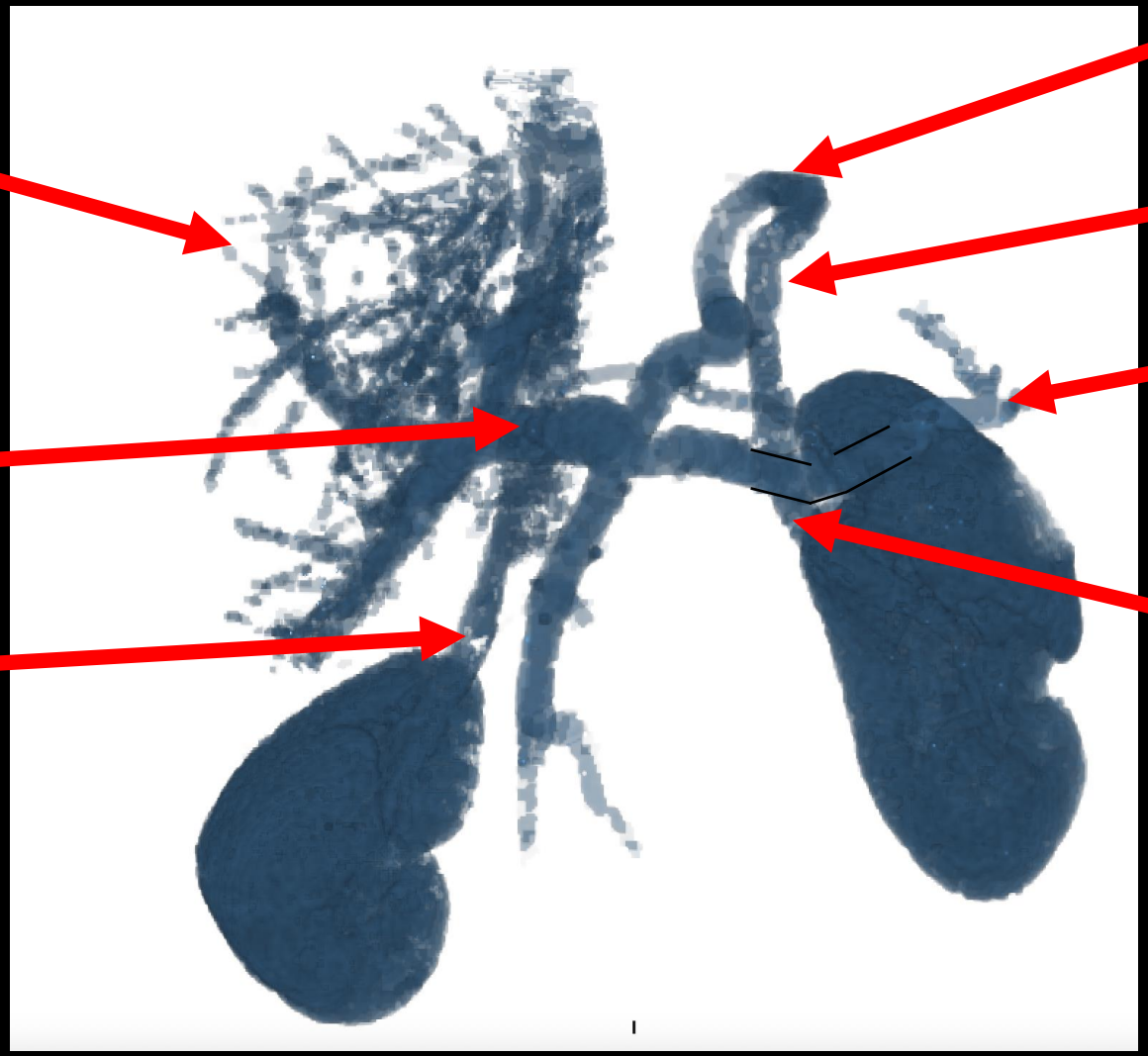
From post-op CT with better venous evaluation

Findings (labeled)



From post-op CT with better venous evaluation

3D Model (created using Horos)



Follow up imaging after AAA repair

Variant 2: **Follow-up for postendovascular repair (EVAR) or open repair of AAA.**

Procedure	Appropriateness Category	Relative Radiation Level
CTA abdomen and pelvis with IV contrast	Usually Appropriate	☢☢☢☢
MRA abdomen and pelvis without and with IV contrast	Usually Appropriate	O
Aortography abdomen	May Be Appropriate	☢☢☢
CT abdomen and pelvis without and with IV contrast	May Be Appropriate	☢☢☢☢
CT abdomen and pelvis without IV contrast and US aorta abdomen with duplex Doppler	May Be Appropriate	☢☢☢
MRA abdomen and pelvis without IV contrast	May Be Appropriate	O
US aorta abdomen with duplex Doppler	May Be Appropriate	O
CT abdomen and pelvis without IV contrast	May Be Appropriate	☢☢☢
CT abdomen and pelvis with IV contrast	May Be Appropriate (Disagreement)	☢☢☢
X-ray abdomen and pelvis	May Be Appropriate	☢☢☢

Aortobiiliac bypass graft with decreased size of AAA measuring 4.5cm x 3.5 cm

Final Dx:

Extrahepatic Congenital Portosystemic Shunt
(Left gastric vein to Left Renal Vein)

Case Discussion

- Congenital Portosystemic Shunts (CPSS) are rare vascular malformations between portal (intestinal) veins and systemic veins[2]
  - Results from incomplete involution of fetal/embryonic vessels[2]
- Estimated prevalence 1:30,000 at birth with 1:50,000 being permanent shunts[3]
  - Intrahepatic shunts are more likely to spontaneously close[4]
- Can be further evaluated by Ultrasound with Doppler, CT, MRI, and angiography with occlusion test[2]

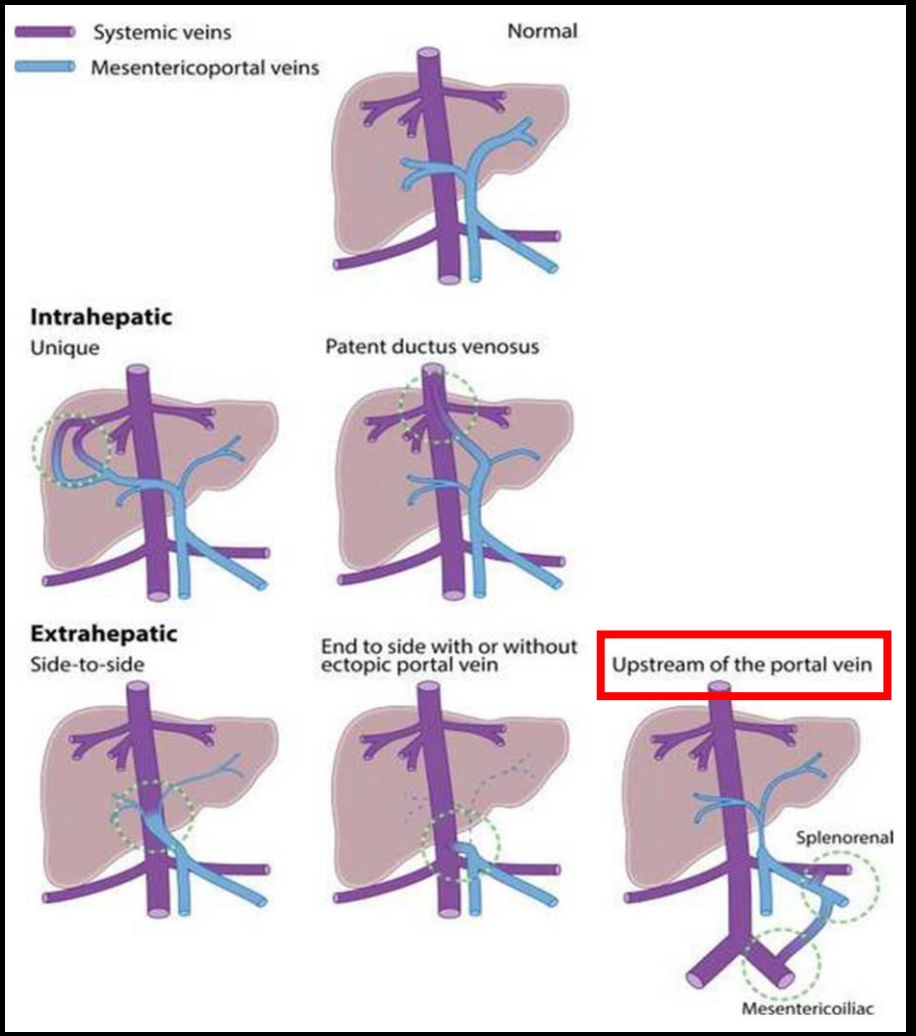
Complications of CPSS

- Potential complications of CPSS:[2]
  - Liver abnormalities
    - liver atrophy
    - biologic disorders (indirect hyperbilirubinemia, increased bile acids, hyperammonemia, etc)
    - benign/malignant tumors (focal nodular hyperplasia, hepatocellular adenoma, nodular regenerative hyperplasia, hepatocellular carcinoma, hepatoblastoma etc)
  - Neurologic
    - portosystemic encephalopathy
  - Cardiopulmonary
    - cardiac malformations,
    - portopulmonary HTN
    - hepatopulmonary syndrome
  - Other systems (renal, GU, GI, endocrine abnormalities)

Case Discussion

- There are different surgical and anatomical classification systems[4]
  - Historically divided into intrahepatic and extrahepatic shunts
  - Kanazawa added descriptor of severity of hypoplasia
  - Bicêtre surgical classification accounted for caval ending of shunt

- Combination of classifications used to approach clinical management[4]



Intrahepatic vs Extrahepatic CPSS
Guérin et al.

Case Discussion

- Treatment is indicated for cases with serious complications[2] (i.e. encephalopathy)
  - Treatment Options:
    - IR embolization (if possible)
    - Surgery
  - Preventative closure is controversial.
  - In the absence of encephalopathy, this patient did not require treatment.

References:

1. American College of Radiology. ACR Appropriateness Criteria®. Available at https://acsearch.acr.org/list . Accessed Sept 9 2022.

2. Franchi-Abella, S., Gonzales, E., Ackermann, O., Branchereau, S., Pariente, D. and Guérin, F., 2018. Congenital portosystemic shunts: diagnosis and treatment. *Abdominal Radiology*, *43*(8), pp.2023-2036.

3. Bernard, O., Franchi-Abella, S., Branchereau, S., Pariente, D., Gauthier, F. and Jacquemin, E., 2012, November. Congenital portosystemic shunts in children: recognition, evaluation, and management. In *Seminars in Liver Disease* (Vol. 32, No. 04, pp. 273-287). Thieme Medical Publishers.

4. Guérin, F., Abella, S.F., McLin, V., Ackermann, O., Girard, M., Cervoni, J.P., Savale, L., Hernandez-Gea, V., Valla, D., Hillaire, S. and Dutheil, D., 2020. Congenital portosystemic shunts: Vascular liver diseases: Position papers from the francophone network for vascular liver diseases, the French Association for the Study of the Liver (AFEF), and ERN-rare liver. *Clinics and Research in Hepatology and Gastroenterology*, *44*(4), pp.452-459