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:**

<table>
<thead>
<tr>
<th>Radiologic Procedure</th>
<th>Rating</th>
<th>Comments</th>
<th>RRL*</th>
</tr>
</thead>
<tbody>
<tr>
<td>US aorta abdomen</td>
<td>9</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>CTA abdomen with IV contrast</td>
<td>8</td>
<td></td>
<td>★★★☆</td>
</tr>
<tr>
<td>MRA abdomen without and with IV contrast</td>
<td>8</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>CT abdomen without IV contrast</td>
<td>7</td>
<td></td>
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<td>7</td>
<td></td>
<td>★★★☆</td>
</tr>
<tr>
<td>MRA abdomen without IV contrast</td>
<td>7</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>Aortography abdomen</td>
<td>4</td>
<td></td>
<td>★★☆☆</td>
</tr>
<tr>
<td>FDG-PET/CT abdomen</td>
<td>2</td>
<td></td>
<td>★★☆☆☆</td>
</tr>
</tbody>
</table>

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

*Relative Radiation Level*
# Interventional Planning for AAA

## Variant 1:

Planning for pre-endovascular repair (EVAR) or open repair of AAA.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>CTA abdomen and pelvis with IV contrast</td>
<td>Usually Appropriate</td>
<td>★★★★★</td>
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<tr>
<td>MRA abdomen and pelvis without and with IV contrast</td>
<td>Usually Appropriate</td>
<td>O</td>
</tr>
<tr>
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<td>May Be Appropriate</td>
<td>O</td>
</tr>
<tr>
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<tr>
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<td>May Be Appropriate</td>
<td>★★★</td>
</tr>
<tr>
<td>CT abdomen and pelvis without and with IV contrast</td>
<td>May Be Appropriate</td>
<td>★★★★</td>
</tr>
<tr>
<td>US aorta abdomen with duplex Doppler</td>
<td>Usually Not Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>X-ray abdomen and pelvis</td>
<td>Usually Not Appropriate</td>
<td>★★★★</td>
</tr>
<tr>
<td>CT abdomen and pelvis without IV contrast and US aorta abdomen with duplex Doppler</td>
<td>Usually Not Appropriate</td>
<td>★★★★</td>
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</table>
Findings (unlabeled)
Findings: (labeled)

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

- L gastric vein (portal circulation)
- L renal vein (systemic circulation)
- Portosystemic Shunt
Findings (unlabeled)

From post-op CT with better venous evaluation
Findings (labeled)

From post-op CT with better venous evaluation

Intact Portal Vein
3D Model (created using Horos)

- Hepatic Portal Venous System
- Portal v.
- R renal v. (systemic)
- L. gastric v.
- Portosystemic Shunt
- Splenic v.
- L renal v.
Follow up imaging after AAA repair

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<td>CTA abdomen and pelvis with IV contrast</td>
<td>Usually Appropriate</td>
<td>💣💣💣💣</td>
</tr>
<tr>
<td>MRA abdomen and pelvis without and with IV contrast</td>
<td>Usually Appropriate</td>
<td>0</td>
</tr>
<tr>
<td>Aortography abdomen</td>
<td>May Be Appropriate</td>
<td>💣💣💣</td>
</tr>
<tr>
<td>CT abdomen and pelvis without and with IV contrast</td>
<td>May Be Appropriate</td>
<td>💣💣💣</td>
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</tr>
<tr>
<td>MRA abdomen and pelvis without IV contrast</td>
<td>May Be Appropriate</td>
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</tr>
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</tr>
<tr>
<td>CT abdomen and pelvis without IV contrast</td>
<td>May Be Appropriate</td>
<td>💣💣💣</td>
</tr>
<tr>
<td>CT abdomen and pelvis with IV contrast</td>
<td>May Be Appropriate (Disagreement)</td>
<td>💣💣💣</td>
</tr>
<tr>
<td>X-ray abdomen and pelvis</td>
<td>May Be Appropriate</td>
<td>💣💣💣</td>
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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)
• 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\(^4\)
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:


