

# Urachal Rhabdomyosarcoma

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# Urachal RMS

- The importance of primary site in prognosis of RMS

# Prognosis by Site

**Most Favorable**

Orbit/Head and Neck

GU, Non-bladder/prostate (Think, Para-testis and GYN)

GU, Bladder/Prostate

Parameningeal

Other

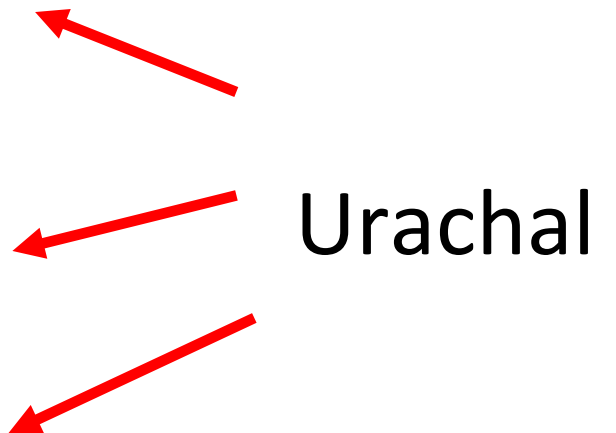
Extremity

**Least favorable**

**Urachal**

# Stage

	Stage	Sites	T	Size	N	M
<b>Favorable</b>	1	Orbit Head and neck (excluding parameningeal) GU – non-bladder/ non-prostate Biliary Tract/Liver	T <sub>1</sub> or T <sub>2</sub>	a or b	N <sub>0</sub> or N <sub>1</sub> or N <sub>x</sub>	M <sub>0</sub>
<b>Unfavorable, Small</b>	2	Bladder/Prostate Extremity, Cranial Parameningeal, Other (includes trunk, retroperitoneum, etc.) Except Biliary tract/Liver	T <sub>1</sub> or T <sub>2</sub>	a	N <sub>0</sub> or N <sub>x</sub>	M <sub>0</sub>
<b>Unfavorable, Large, or Nodes</b>	3	Bladder/Prostate Extremity Cranial Parameningeal, Other (includes trunk, retroperitoneum, etc.) Except Biliary tract/Liver	T <sub>1</sub> or T <sub>2</sub>	a b	N <sub>1</sub> N <sub>0</sub> or N <sub>1</sub> or N <sub>x</sub>	M <sub>0</sub> M <sub>0</sub>
<b>Metastatic</b>	4	All	T <sub>1</sub> or T <sub>2</sub>	a or b	N <sub>0</sub> or N <sub>1</sub>	M <sub>1</sub>



Site → T1= confined to anatomic site of origin, T2= extension to surrounding tissue

Size → a= ≤5 cm, b= >5 cm

Nodes → N0= no regional nodes, N1= regional nodes clinically involved, Nx= status of nodes not known

Mets → M0= no distant mets, M1= + mets

# Group

**Group 1:** Localized disease, completely resected

**Group 2:** Gross total resection with evidence of regional spread

- a. Grossly resected tumor with microscopic residual
- b. Regional disease with involved nodes, completely resected without residual disease
- c. Regional disease with involved nodes, grossly resected but with microscopic residual and/or involvement of the most distal node in the dissection

**Group 3:** Incomplete resection with gross residual disease

- a. After biopsy only

**Group 4:** Distant mets

- Includes lung, liver, bones, BM, brain, distant muscle, nodes, + CSF, or implants on pleural or peritoneal surface

# Urachal RMS

- Rarely reported entity, though it is probably under-reported
  - Commonly described as “other” in series

# Urachal RMS

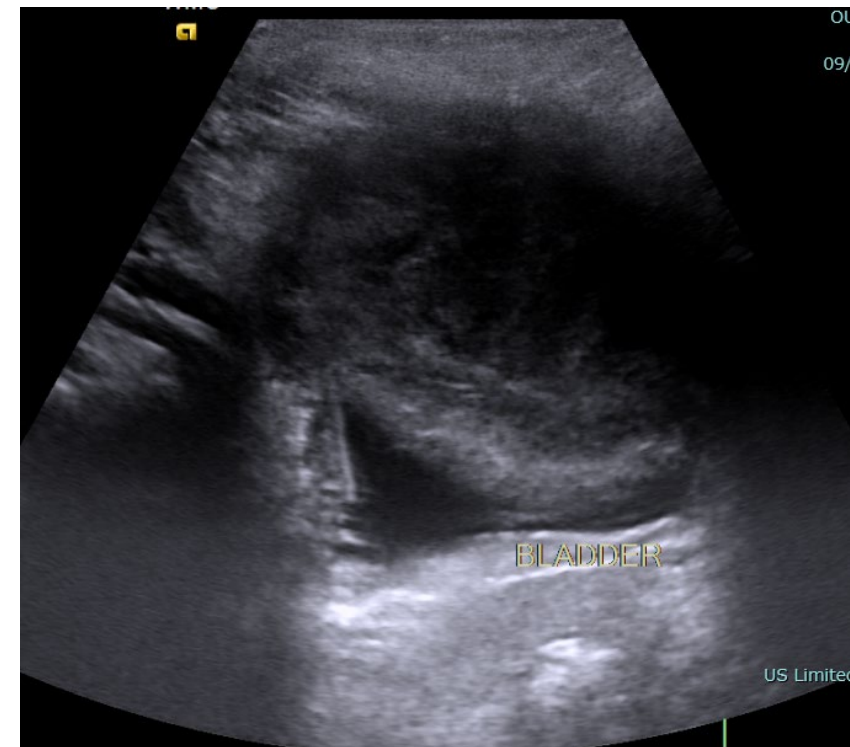
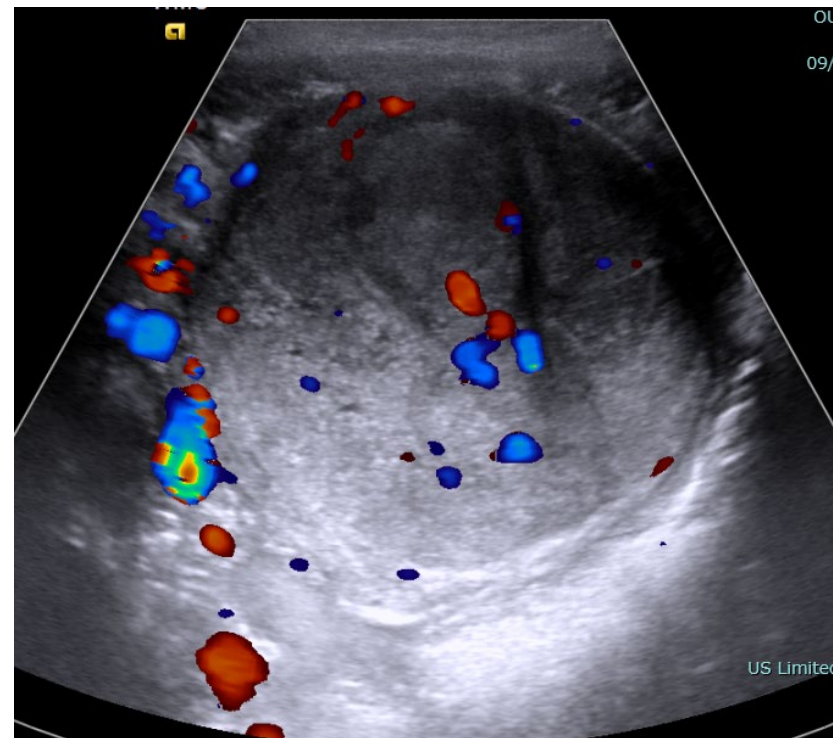
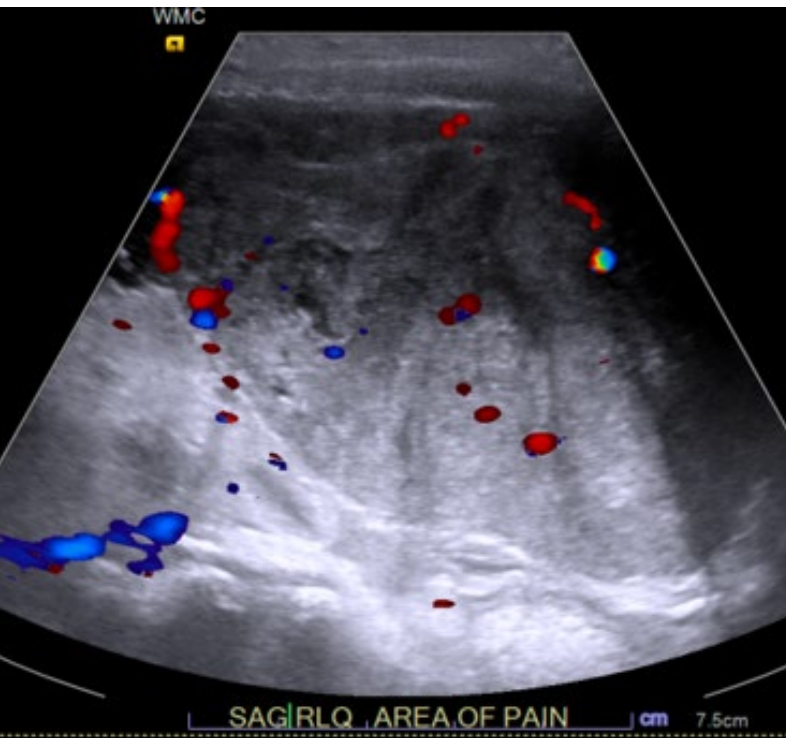
- Case based discussion
  - Huge thank you to Dr. Halstead!

# HPI

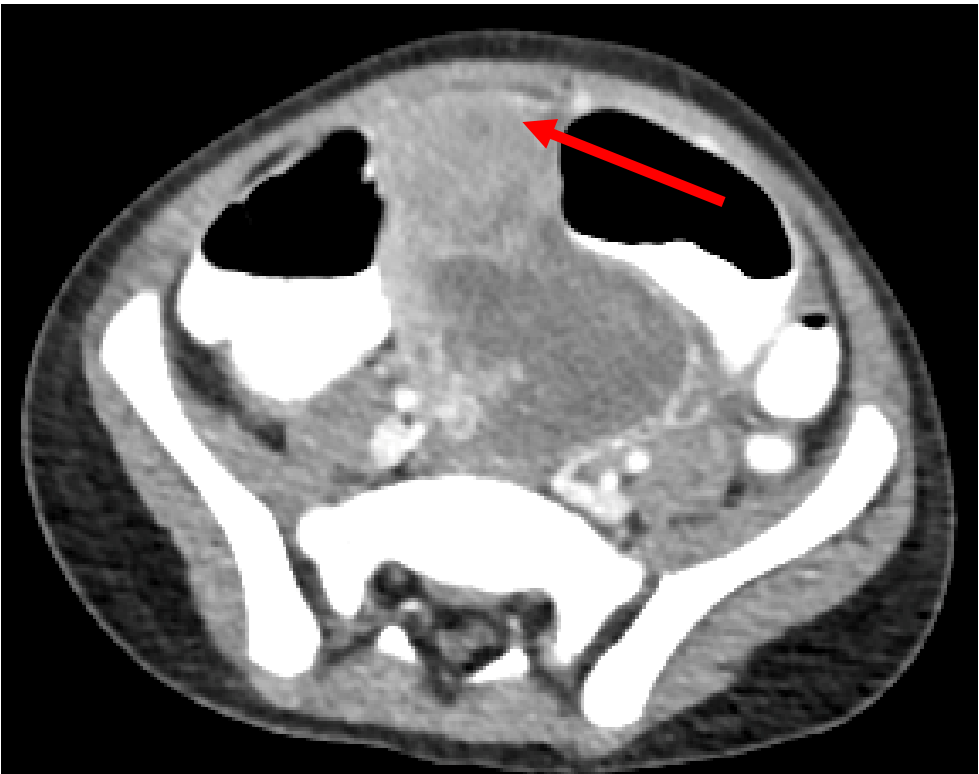
- Presented to OSH with several week h/o **abdominal pain**
  - *Endorses (+)*: Fatigue, anorexia, urinary frequency
  - *Denies (-)*: Fever, weight loss, hematuria
  - Enlarged spleen palpated
- Spiked fever on arrival. Blood cultures drawn.
- Labs:
  - WBC: 11.22
  - Uric acid: 5.7
  - UA: blood present
  - WNL: CMP, AFP, bHCG, LDH, HMA, VMA



# Imaging: US



# Imaging: CT

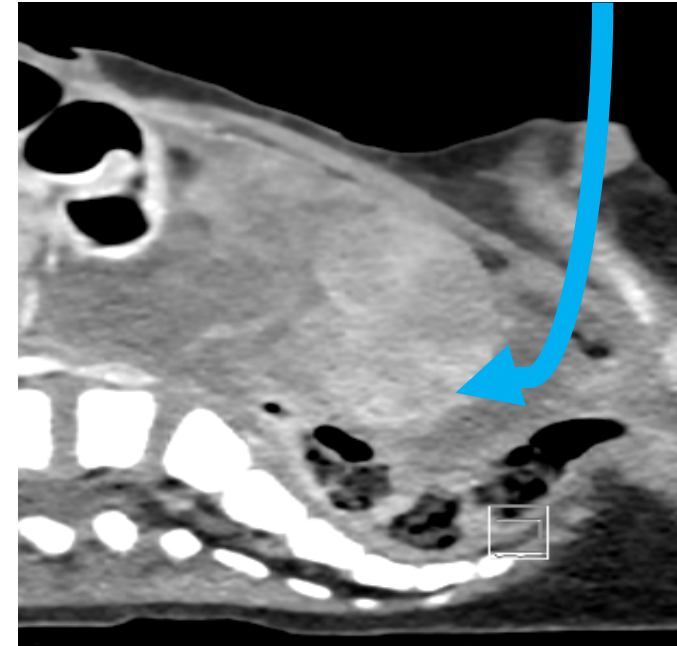


# Diagnostic Biopsy

## Transvesical mass biopsy

### Mediport placement

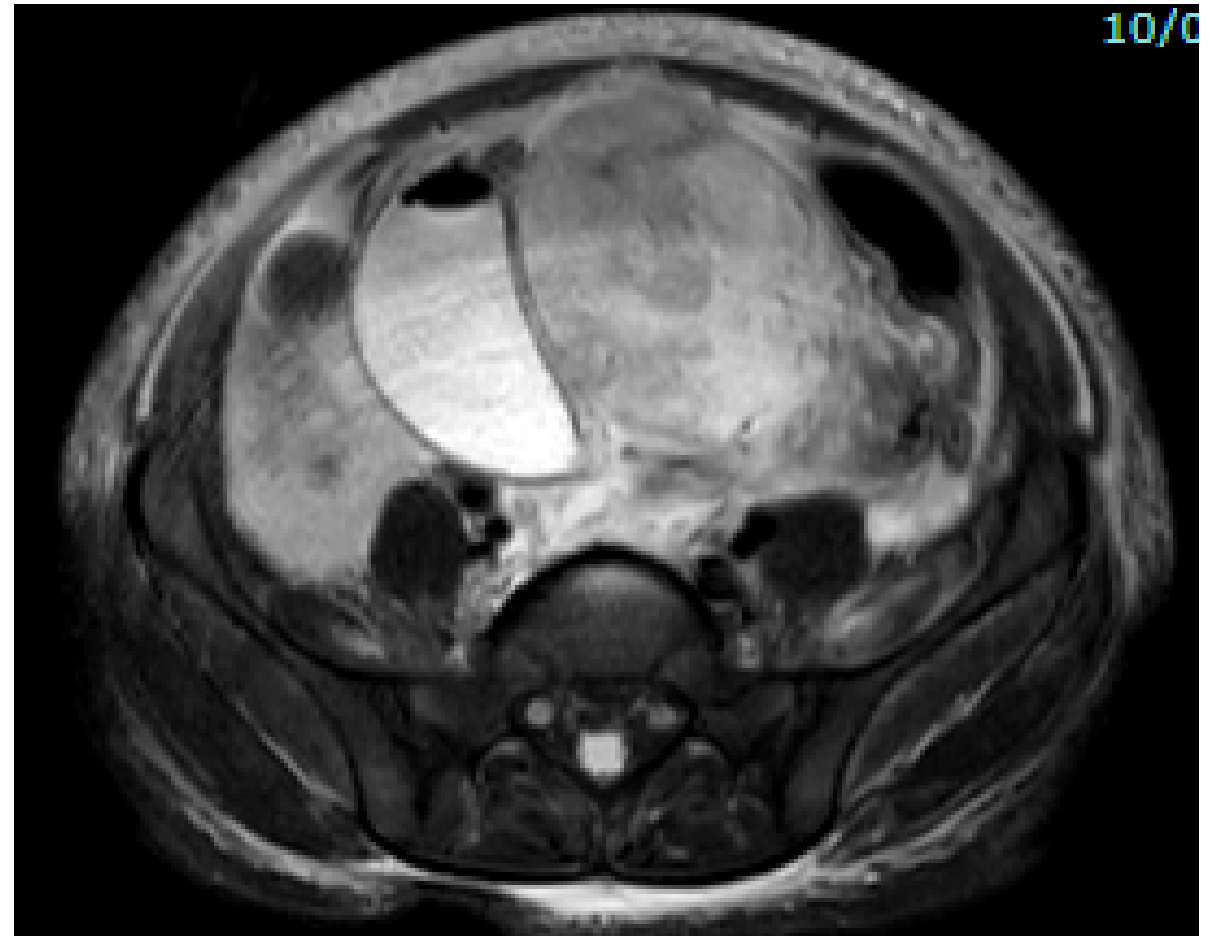
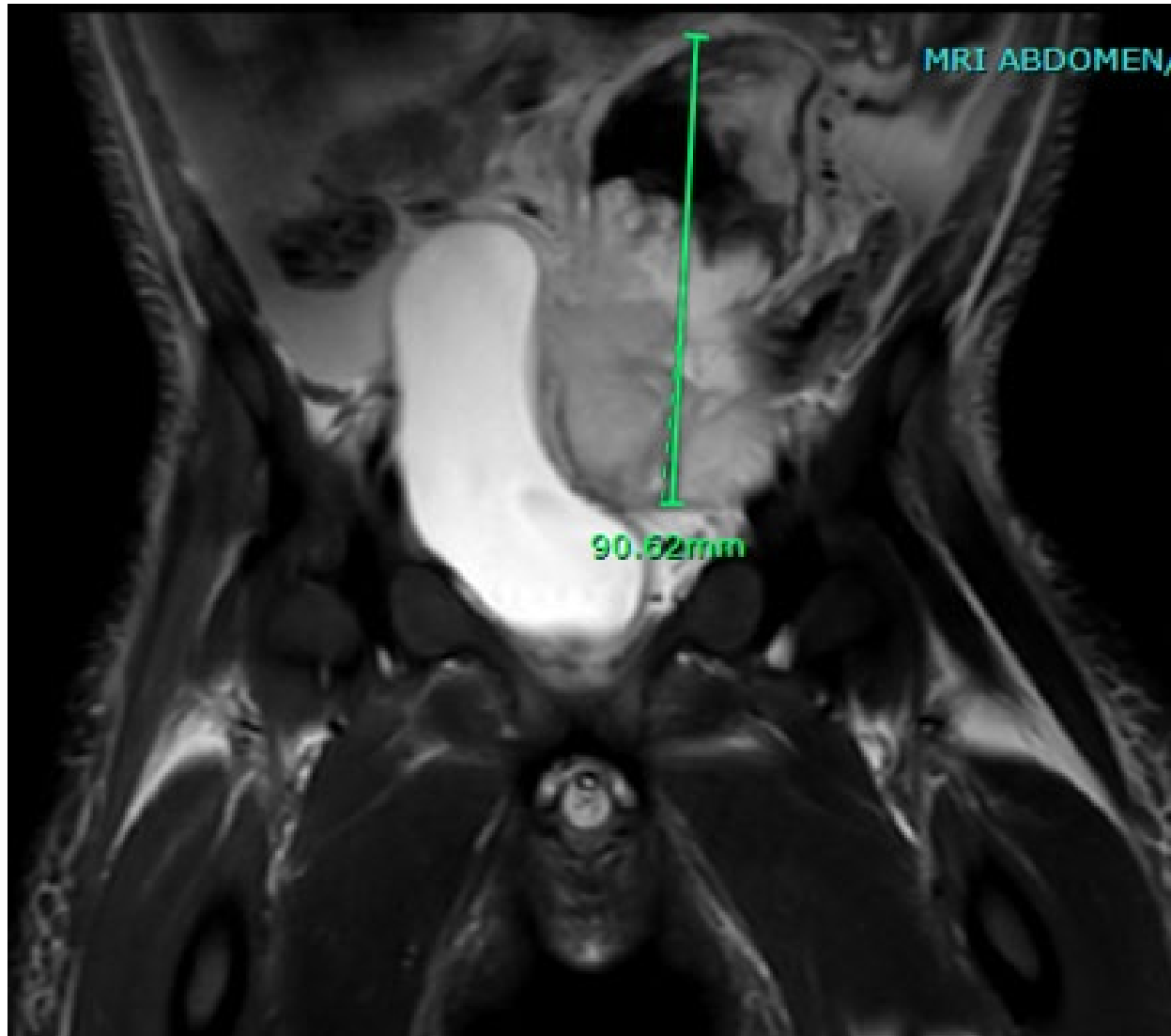
- Mass effect from tumor noted at anterior bladder wall
- Transvesical 18 gauge core needle biopsy through cystoscope working channel
- Preliminary frozen = **rhabdomyosarcoma**
- Due to frozen, proceeded with port placement



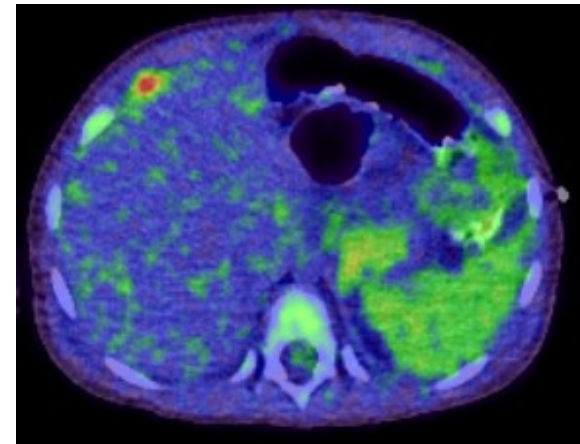
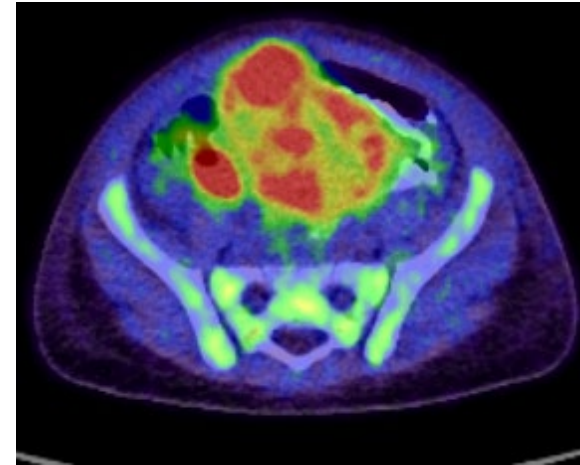
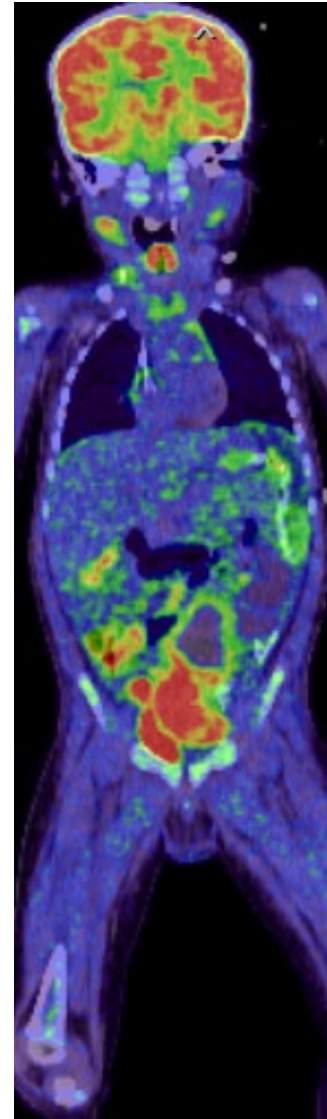
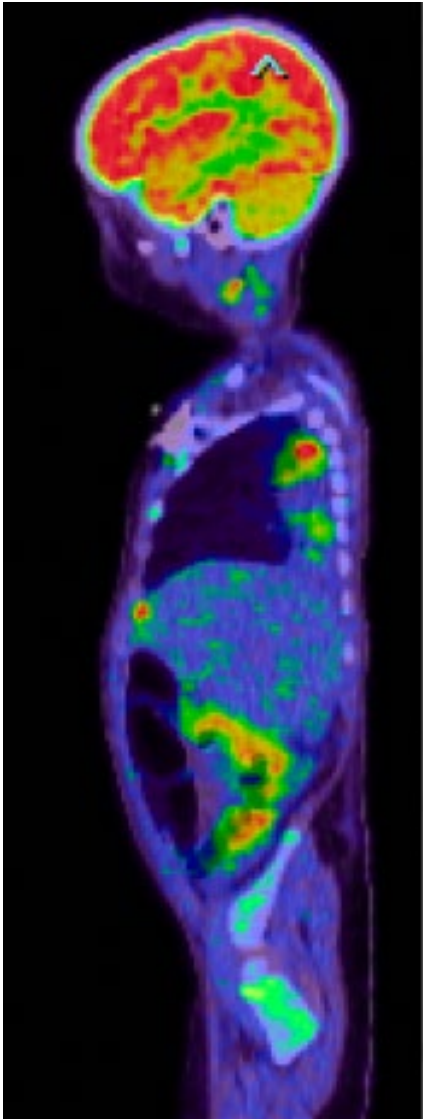
#### FINAL PATHOLOGY:

Embryonal rhabdomyosarcoma.  
FOXO1 fusion negative.

# Imaging: MRI



# Imaging: PET/CT



# Surgical Resection

1. Cystoscopy
  2. Temporary left ureteral stent placement
  3. **Resection of urachal mass**
    - i. Resection of mesenteric lymph node
    - ii. Evaluated liver intra-operatively
- Cystic area of tumor ruptured preoperatively
    - Encountered **bloody ascites**
  - Tumor involving **root of small bowel mesentery, sigmoid colon, and ileum**

# Final Pathology

- **Stage III, group IIIb** urachal embryonal rhabdomyosarcoma.
  - Focal necrosis.
  - Fusion negative.
  - Vascular margin negative.
  - Small foci of anaplasia.
- Lymph nodes negative for RMS.
- Negative for Li-Fraumeni, DICER-1, TP53, NF1

<b>Stage 3</b>	Unfavorable	T1 or T2	< 5 cm	N1	M0
		OR	≥ 5 cm	N0, N1, NX	M0

<b>Group III</b>	<b>Gross residual disease</b>
Group IIIa	Localized or regional disease, Biopsy
Group IIIb	Localized or regional disease, Resection (debulking of more than 50% of tumor)

Embryonal	III	2,3	Intermediate risk
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# Treatment

- **COG protocol D9803**
  - 42 weeks of Vincristine, Actinomycin, Cyclophosphamide
- XRT beginning at week 12
  - 24 Gy in 16 fractions to whole abdomen/pelvis
  - Then boost to sites of unresected disease for **total of 50.4 Gy**

*Currently: 2 years disease free*

Wolden SL, Lyden ER, Arndt CA, et al. Local Control for Intermediate-Risk Rhabdomyosarcoma: Results From D9803 According to Histology, Group, Site, and Size: A Report From the Children's Oncology Group. *Int J Radiat Oncol Biol Phys*. 2015;93(5):1071-1076. doi:10.1016/j.ijrobp.2015.08.040

Arndt CA, Stoner JA, Hawkins DS, Rodeberg DA, Hayes-Jordan AA, Paidas CN, Parham DM, Teot LA, Wharam MD, Breneman JC, Donaldson SS, Anderson JR, Meyer WH. Vincristine, actinomycin, and cyclophosphamide compared with vincristine, actinomycin, and cyclophosphamide alternating with vincristine, topotecan, and cyclophosphamide for intermediate-risk rhabdomyosarcoma: children's oncology group study D9803. *J Clin Oncol*. 2009 Nov 1;27(31):5182-8. doi:10.1200/JCO.2009.22.3768. Epub 2009 Sep 21. PMID: 19770373; PMCID: PMC2773476.





# Urachal RMS

- Largest series reported: 8 patients from 1984 to 2013
  - Age 1 yo – 8 yo
- Relatively **rare** entity
  - Typically embryonal (6/8)
  - **Ascites** is quite common (series showed 8/8 patients)
  - Median size: 10.3 cm
  - Typically stage III (2/8) or stage IV (6/8)

Cheikhelard A, Irtan S, Orbach D, Minard-Colin V, Rod J, Martelli H, Sarnacki S. Urachal rhabdomyosarcoma in childhood: a rare entity with a poor outcome. J Pediatr Surg. 2015 Aug;50(8):1329-33. doi: 10.1016/j.jpedsurg.2014.12.023. Epub 2015 Jan 7. PMID: 25913896.

# Urachal RMS

- **Poor prognosis** and can be associated with peritoneal metastasis
- Outcomes:
  - 7/8 in complete remission at end of treatment
  - 4/8 patients relapsed within median 25 months after treatment
  - **4/8 patients died 18-57 months after diagnosis**

# Conclusions

- **Rare** primary site for GU RMS
- **Poor prognosis** compared to other GU sites
  - Even worse than bladder/prostate RMS
- Aggressive, multimodality therapy likely warranted