



# **BOTRYOID RHABDOMYOSARCOMA OF THE BLADDER**

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# Agenda

- **Definition / Incidence**
- **Types of Rhabdomyosarcoma**
- **Our Patient Presentation**
- **Differential Diagnosis based of Patient Presentation**
- **Radiographic Modalities Pre-op / Post-op**
- **Other locations of the tumor and subsequent radiographic films**
- **Prognosis**
- **Summary**



# Definition / Incidence

## Definition:

- Rhabdomyosarcoma is a type of malignant tumor that usually arises from primitive muscle cells.
- Idiopathic in nature
- Can also arise from other areas that lack skeletal muscles such as genitourinary tract, head and neck.

## Incidence:

- Very rare - 6 cases per 1 000 000 population.
- 250 cases diagnosed per year in the United States.



# Types of Rhabdomyosarcoma

## 3 Types of Rhabdomyosarcoma:

- ◎ Embryonal rhabdomyosarcoma – occurs in children
  - Botryoid rhabdomyosarcoma – Our Patient
  - Spindle cell rhabdomyosarcoma
  - Anaplastic rhabdomyosarcoma
- ◎ Alveolar rhabdomyosarcoma – occurs in adolescents
- ◎ Pleomorphic rhabdomyosarcoma



# Our Patient: Presentation

- Ms. S is a 2yr old girl with history of bleeding and mass coming out of the introitus.
- The mass was thought to be urethral prolapse and she was prescribed Premarin Cream (conjugated estrogens).
- After 3 weeks, she presents with increased bleeding and increase in the size of the mass.
- On physical exam, the mass was found to be sub-urethral.

Therefore, an ULTRASOUND was done followed by other modalities to confirm the diagnosis.



# Differential Diagnosis

## Based on the Patient Presentation:

- ⦿ **Rhabdomyosarcoma**
- ⦿ **Fibroepithelial Polyp**
- ⦿ **Metastasis**
- ⦿ **Mesenchymal neoplasm**



# Radiographic Modalities

- **Ultrasound**
- **MRI**
- **Voiding Cysto-Urethrogram (VCUG)**
- **CT Scan**
- **Bone Scan**
- **After Treatment - Loopogram**



# Ultrasound

One of the **first modalities** used to diagnose pathologies in the bladder.

## Benefits:

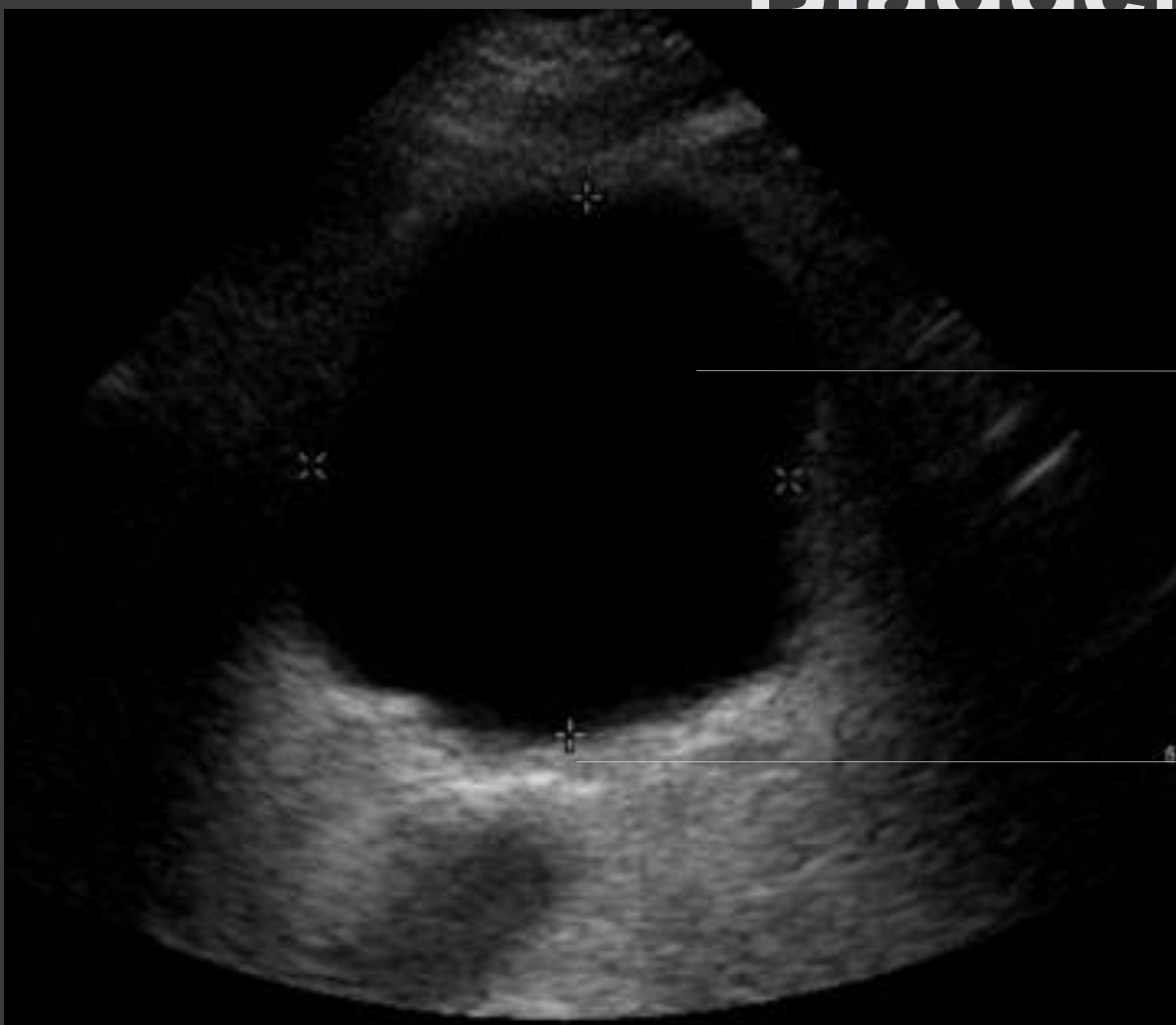
1. Easy to do and painless
2. Zero radiation
3. Perfect for identifying tumors in the pelvis compared to chest as the ribs obstruct the sound waves.





# Normal Ultrasound of Bladder

## Normal Ultrasound of the Bladder

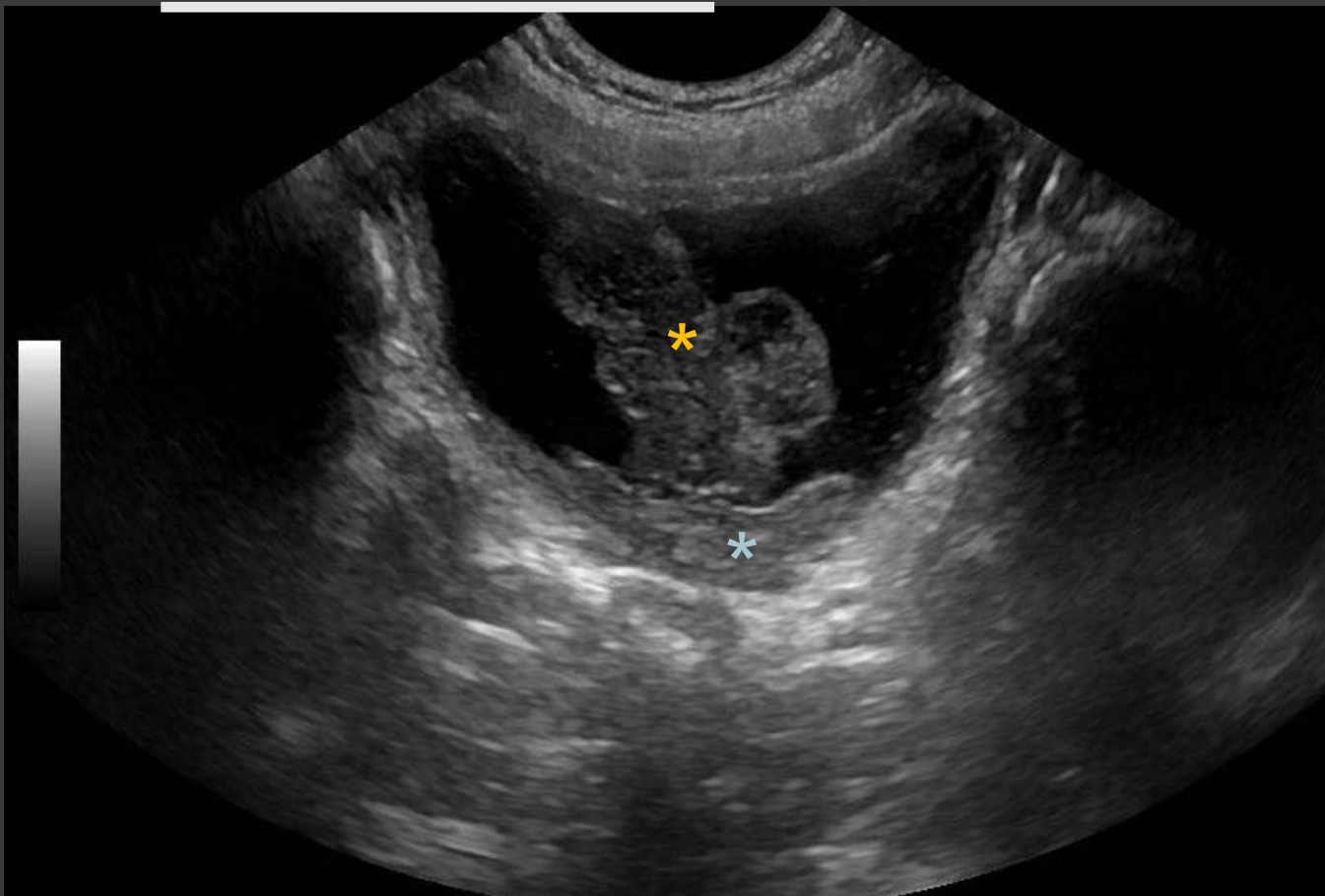


- **Normal fluid-filled bladder**
  - Anechoic structure
- **Posterior Bladder Wall**
  - Hyperechoic in nature
  - Normal wall thickness

Image Source : <http://www.med-ed.virginia.edu/courses/rad/gu/anatomy/bladder.html>



# Our Patient – Mass on Ultrasound



- Mass at the base of the bladder

- Attached to posterior wall

- Lobulated Appearance

- Heterogenous in echogenecity

- Posterior Bladder wall thickness

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**After the ultrasound, biopsy was done which confirmed a Embryonal Rhabdomyosarcoma.**

**This was followed by an MRI.**



# Magnetic Resonance Imaging (MRI)

## Advantages:

1. Better suited than the CT scan for imaging tumours
2. Superior to CT scan for imaging soft tissues
3. Good soft tissue differentiation
4. No Radiation

## Disadvantages:

1. Takes a long time
2. Less detail of bony structures



# Our Patient: Tumor Mass on MRI

*Fat-Saturated Post-Contrast T1 Weighted  
Image*



• **Multi-lobulated  
mass in the lumen**

• **Classic Grape-like  
appearance:**

- **Boytroid type of  
Embryonal RMS**

**Axial**

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# Our Patient: Tumor Mass on MRI

*Fat-Saturated Post-Contrast T1 Weighted Image*



- Multi-lobulated mass at the base of bladder

- In the Trigonal Area

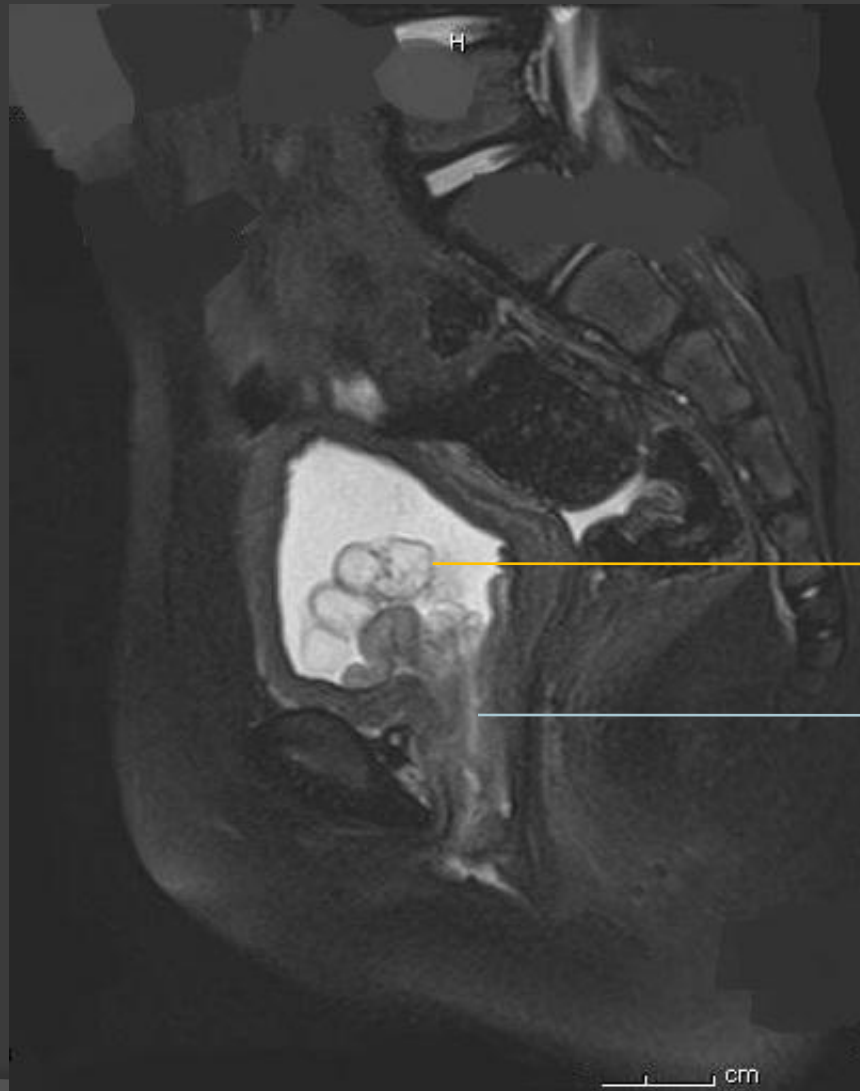
- Grape-like configuration

- Urethra dilated



# Our Patient: Tumor Mass on MRI

*Fat-Saturated T2 Weighted Image*



- Multi-cystic mass extending peripherally in the lumen

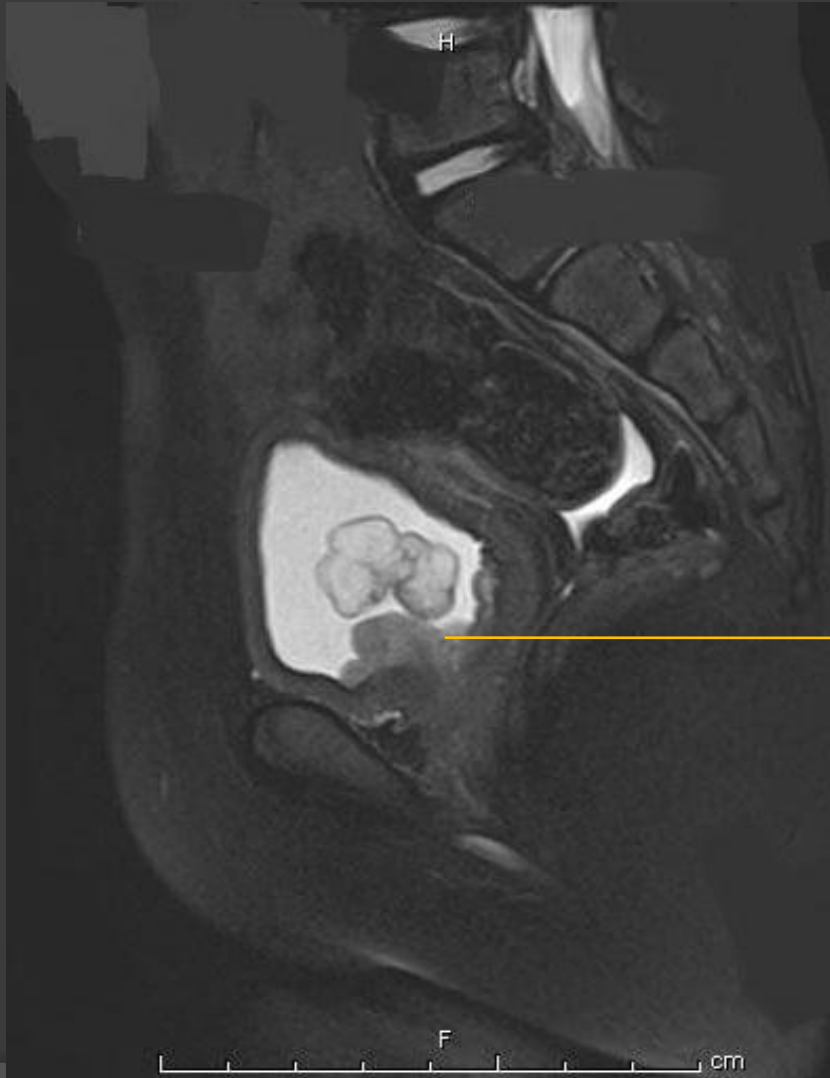
- Grape-like appearance

- Urethra dilated



# Our Patient: Tumor Mass on MRI

*Fat-Saturated T2 Weighted Image*



• Plate-like portion in the bladder base

- Extending in the urethra





# Summary of MRI findings

- Multi-lobulated mass seen in the bladder base
- Present in Trigonal Area.
- Urethra Dilated.
- Mass measured 2 x 3.5 x 3.6 cm  
(less than 5 cm)



**This was followed by a Voiding  
Cystourethrogram (VCUG)**



# Voiding Cystourethrogram (VCUG)

One of the best modalities for assessing the structure of the genito-urinary system.

## Advantages:

1. Provides detailed information about the conditions of the genito-urinary system like
  - Tumors
  - Bladder Obstruction
  - Vesicoureteral Reflux
  - Stricture of the urethra
  - Stones

## Disadvantages:

1. Discomfort after the procedure.



# VCUG Procedure

- ① Urethral catheterization is done
- ① The bladder is filled with approximately 250cc of contrast material (CystoConray)
- ① Fluoroscopic images are used to determine any anatomical variants or presence of tumors, stones or a reflux.



# Our Patient: Tumor Mass on VCUG

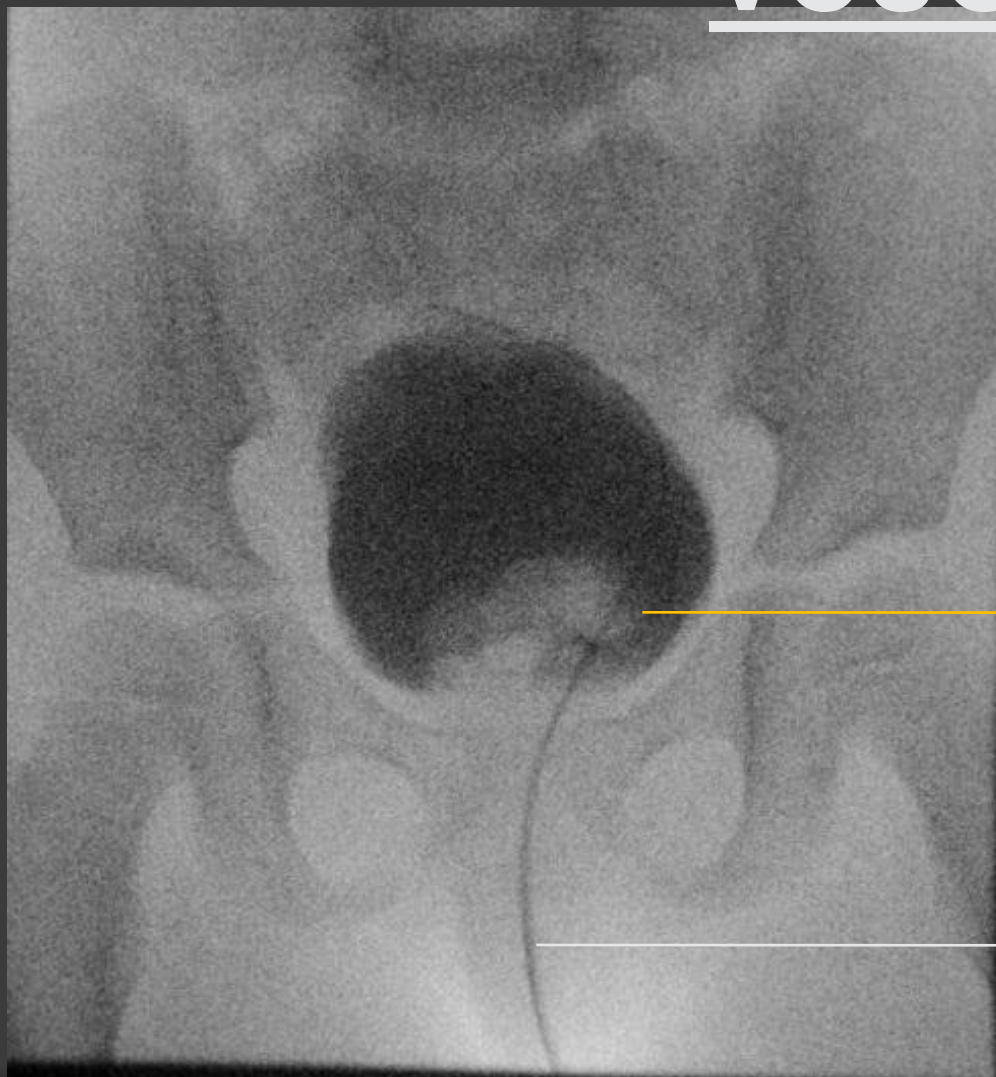


- Lobulated filling defect seen in the inferior part of bladder
  - Present in trigonal area
- No Vesicoureteral reflux seen

Urinary catheter



# Our Patient: Tumor Mass on VCUG



- Lobulated filling defect seen in the inferior part of bladder
- Present in trigonal area
- No Vesicoureteral reflux seen

Urinary catheter



Next, a CT Scan was done



# CT Scan

## Advantages:

- Provides good detail of all internal structures like soft tissue, bone and the blood vessels
- Takes a short time compared to MRI

## Disadvantages:

- Exposure to ionizing radiation





# Our Patient: Tumor Mass on CT

## Scan

*CT Abdomen*



\* Thickening of the superior bladder wall

→ Tumor Mass seen at the bladder base

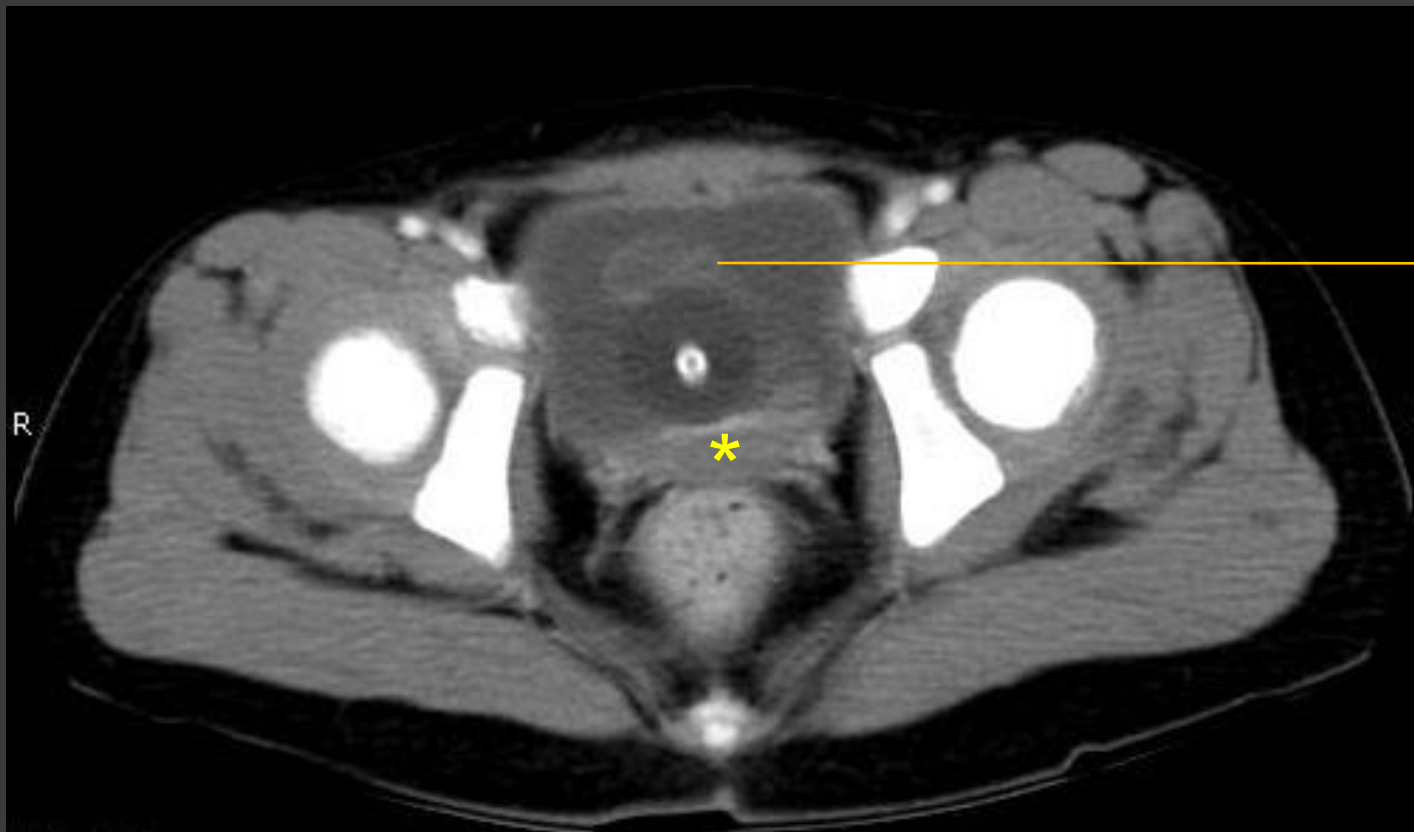
→ Foley's Catheter

Coronal

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# Our Patient: Tumor Mass on CT Scan

*CT Abdomen*



**Tumor Mass  
seen in the  
anterior aspect**

**\* Thickening of  
posterior wall  
seen**

**Axial**

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# Our Patient: Tumor Mass on CT

## Scan

CT Abdomen



\* Thickening of the superior and posterior bladder wall

→ Tumor Mass seen anteriorly



# LETS LOOK AT WHAT WE KNOW SO FAR



# 4 Stages of Rhabdomyosarcoma

- ◎ **Stage I**: N0 , M0
  - Orbit
  - Eyelid
  - Head and neck (excluding parameningeal),
  - Genitourinary (non-bladder, non-prostate)
  
- ◎ **Stage II** : **< 5 cm**, N0, M0
  - **Bladder**
  - Prostate
  - Extremity
  - Parameningeal
  
- ◎ **Stage III** : > 5 cm, N0 or 1, M0
  - Bladder, prostate, extremity, trunk, parameningeal
  
- ◎ **Stage IV** : all others, any N, M1



This tumor is **Stage II** as it's **less than 5 cm** and is localized to the **bladder**.

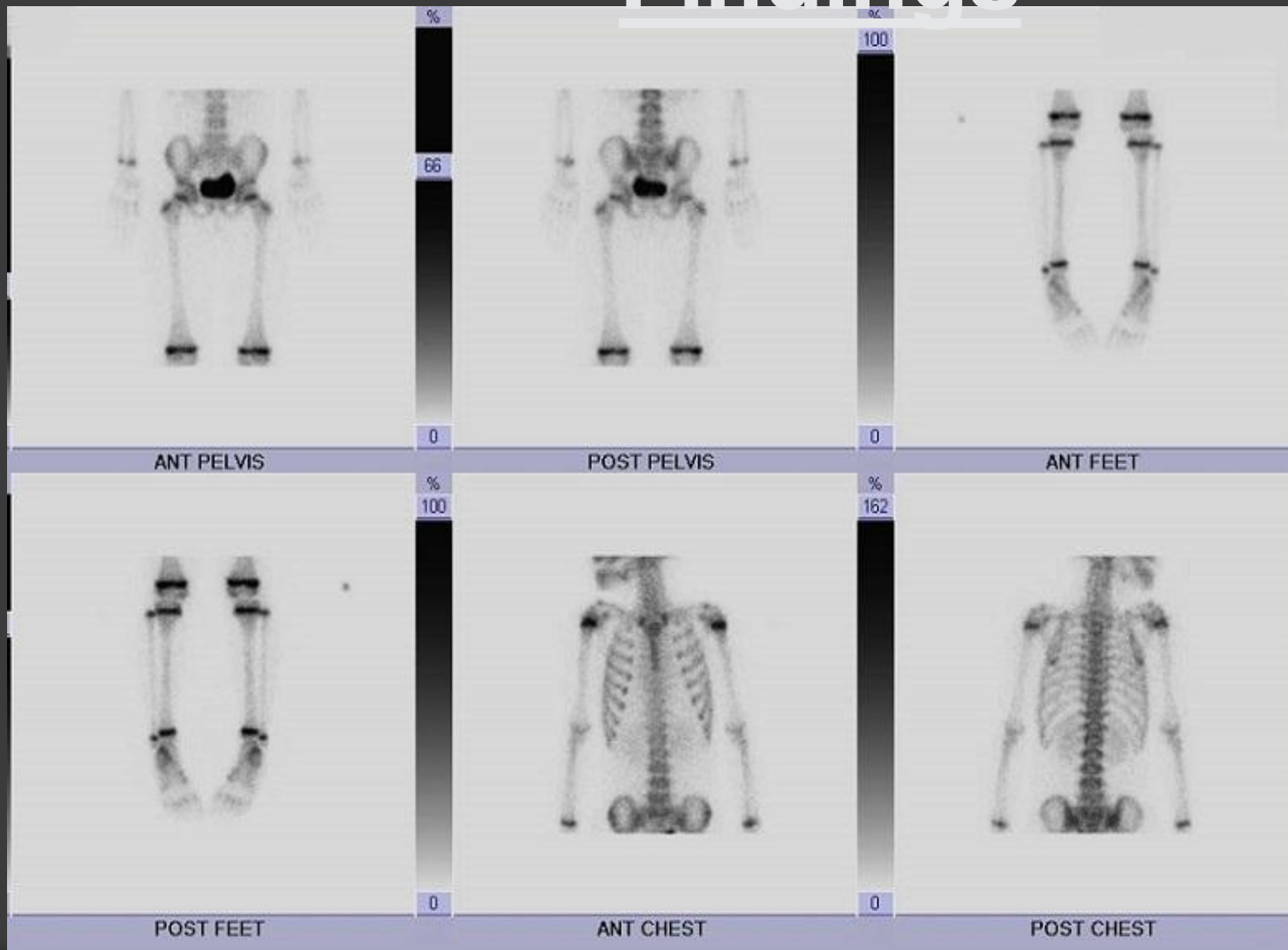
To rule out metastasis, Bone Scan was the next investigation done.



# Bone Scan

- ⦿ Patient is injected a small amount of **radiographic material (tracer)**.
- ⦿ About half of the tracer localizes in the bones
- ⦿ The rest is excreted via the kidneys and bladder.
- ⦿ Useful for identifying **bone lesions / metastasis** in the bones.

# Our Patient: Bone Scan Findings



**No metastasis  
seen**





# Treatment

- ⦿ **Complete resection of the bladder and removal of tumor**
- ⦿ **Sigmoid conduit made as a urinary diversion.**
- ⦿ **Ureters attached to the conduit in a uretero-sigmoid anastomosis.**
- ⦿ **Stoma present in the patient's left lower quadrant attached to a urostomy bag**
- ⦿ **Patient stays on urostomy bag and oral antibiotics for life.**
- ⦿ **Regular follow-ups.**

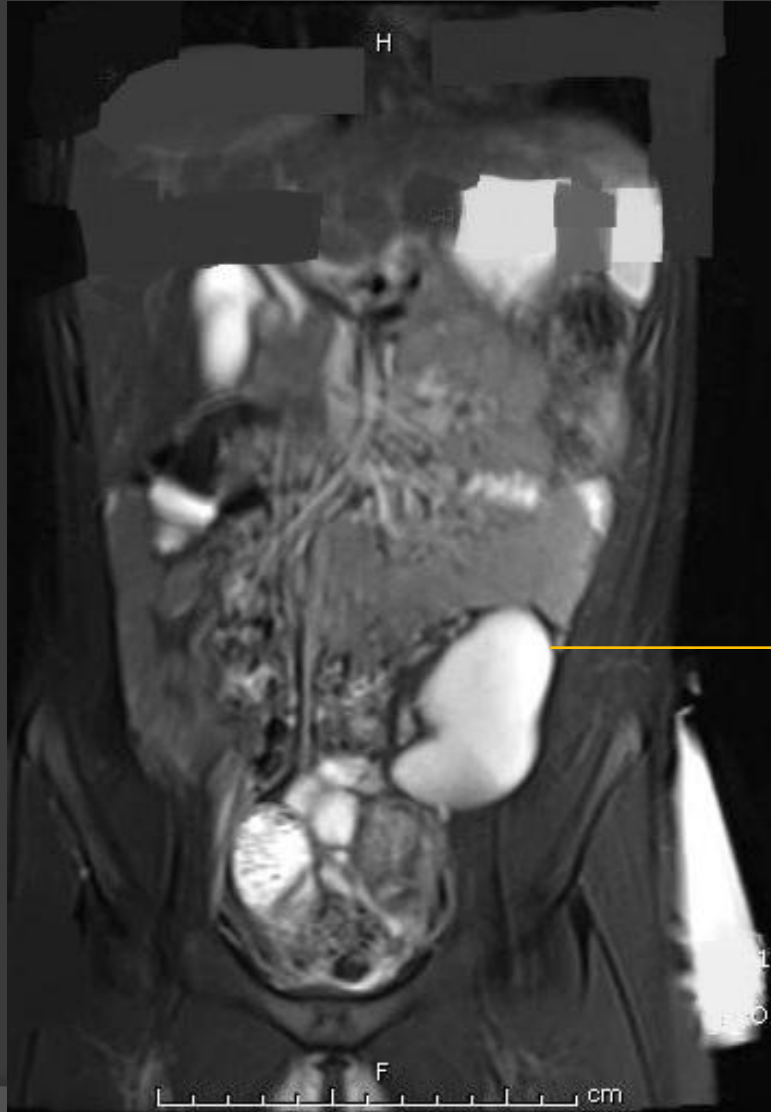


**After treatment, the patient was followed with regular follow-ups including MRI and Loopogram**



# Our Patient: Post – Op MRI

*Fat-Saturated Post-Contrast T2 Weighted Image*



Coronal

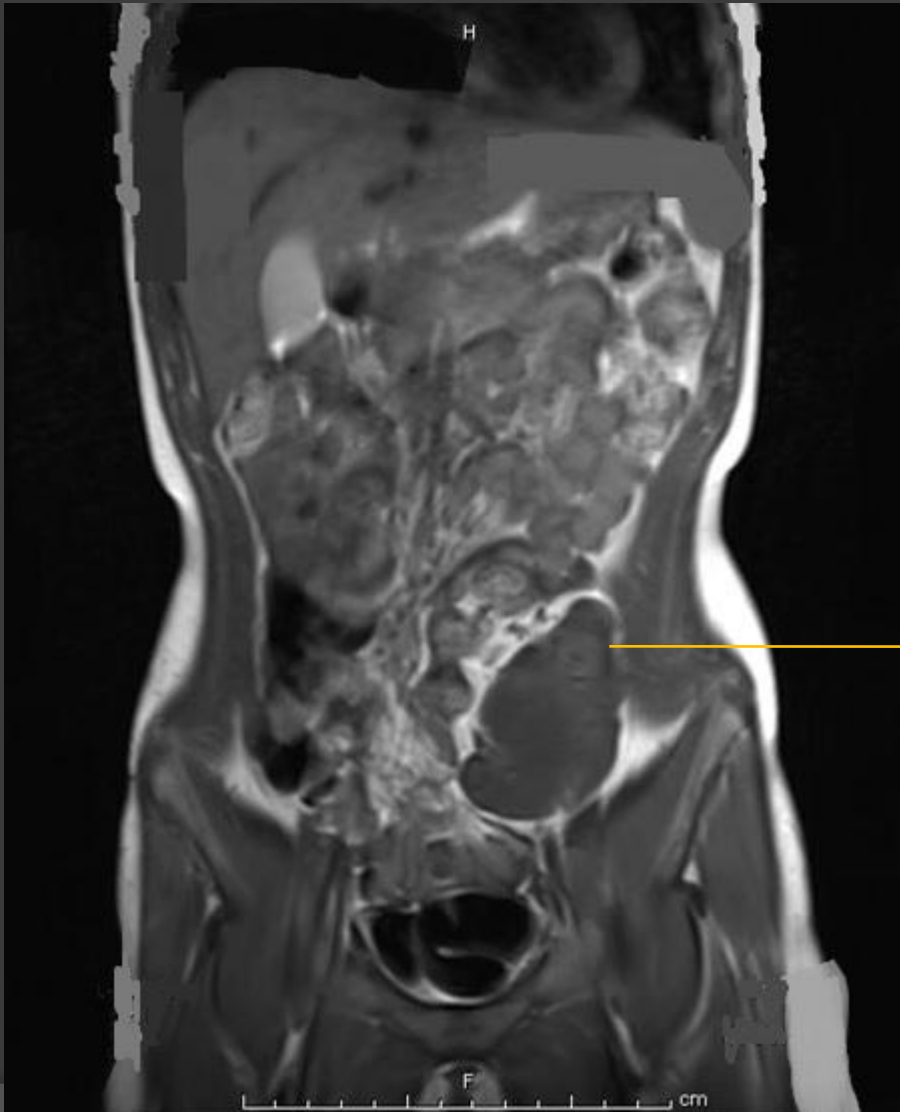
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- Sigmoid conduit present in the left lower quadrant
- Contains urine
- No lesions or masses identified in the neobladder



# Our Patient: Post – Op MRI

*T1 Weighted Image*



→ • Sigmoid conduit present in the left lower quadrant

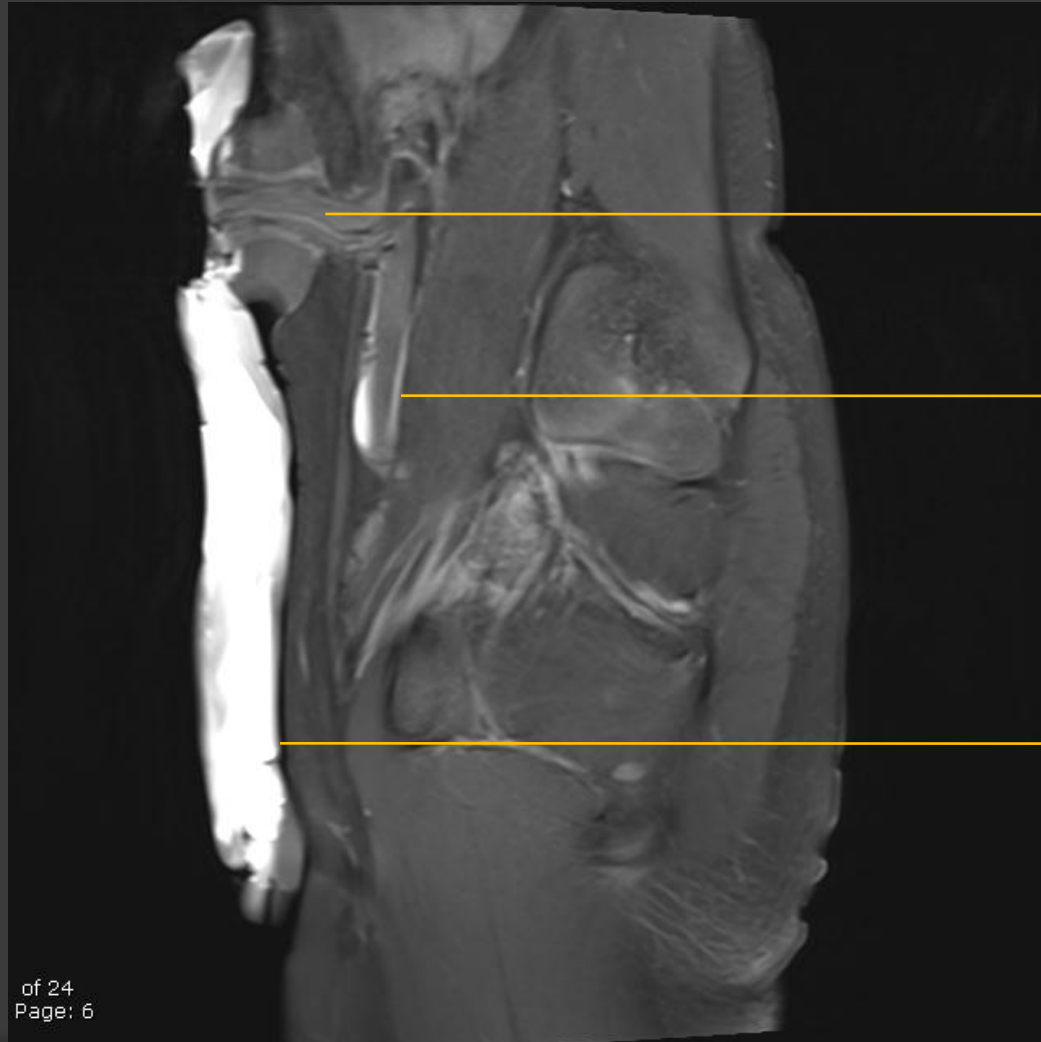
- Contains urine

• No lesions or masses identified in the neobladder



# Our Patient: Post – Op MRI

*Post contrast T1 Image*



**Conduit connected  
to Urostomy bag**

**Sigmoid conduit**

**Urostomy bag**



# Loopogram

**Fluoroscopic procedure** to determine the anatomy and functionality of the sigmoid conduit.

## Indications for Loopogram:

- Follow-up after surgery
- Difficulty emptying the conduit
- Vesicoureteral reflux

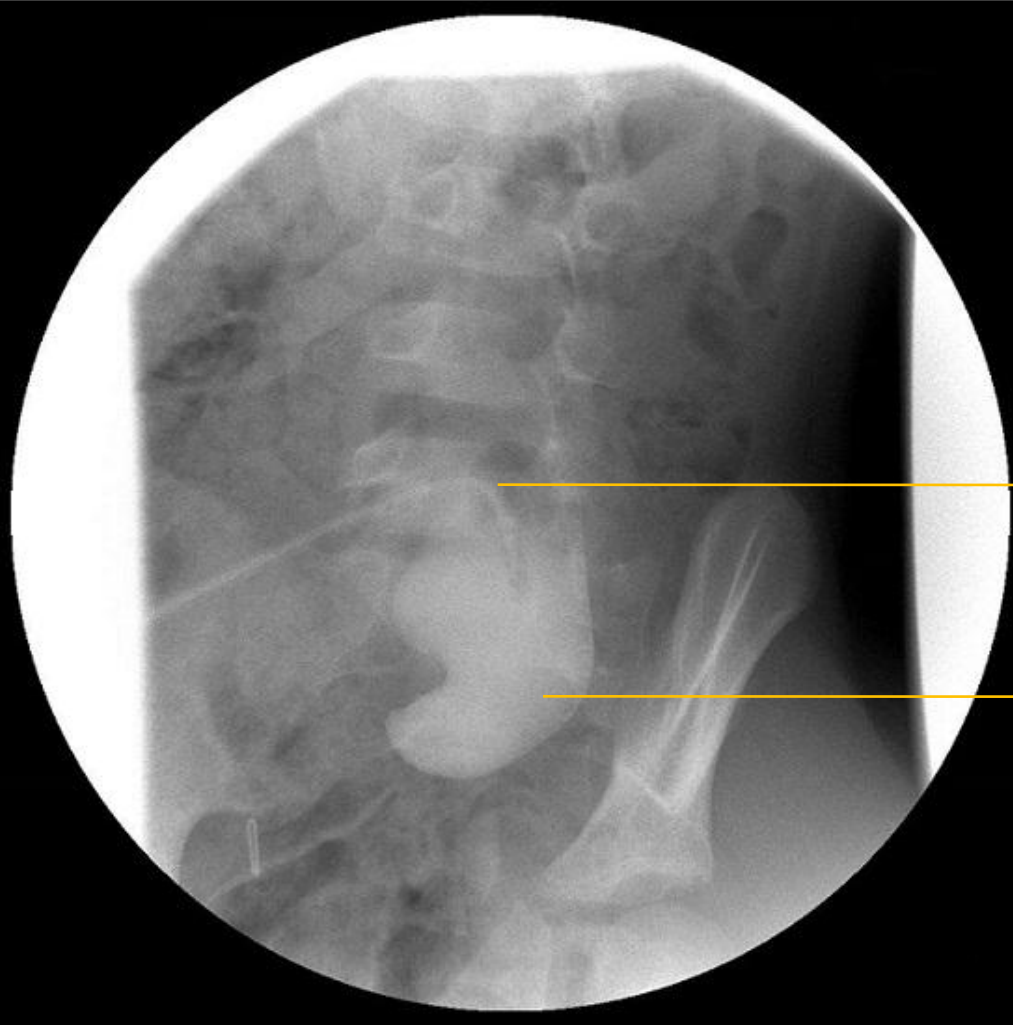


# Procedure for Loopogram

- ① 12 French Foley Catheter inserted into the stoma.
- ① Balloon inflated and mild traction is given.
- ① Contrast material (CystoConray) instilled into the conduit.
- ① Fluoroscopic images are taken to visualise the anatomy and functionality of the conduit.



# Our Patient: Post-Op Loopogram



## Findings :

- Smooth shape of sigmoid conduit
- No filling defect seen

→ Catheter

→ Sigmoid conduit filling  
with contrast agent





**Next, we will look at a major  
complication that can occur with this  
treatment**



# Complications

## Vesicoureteral Reflux

### ● Grades:

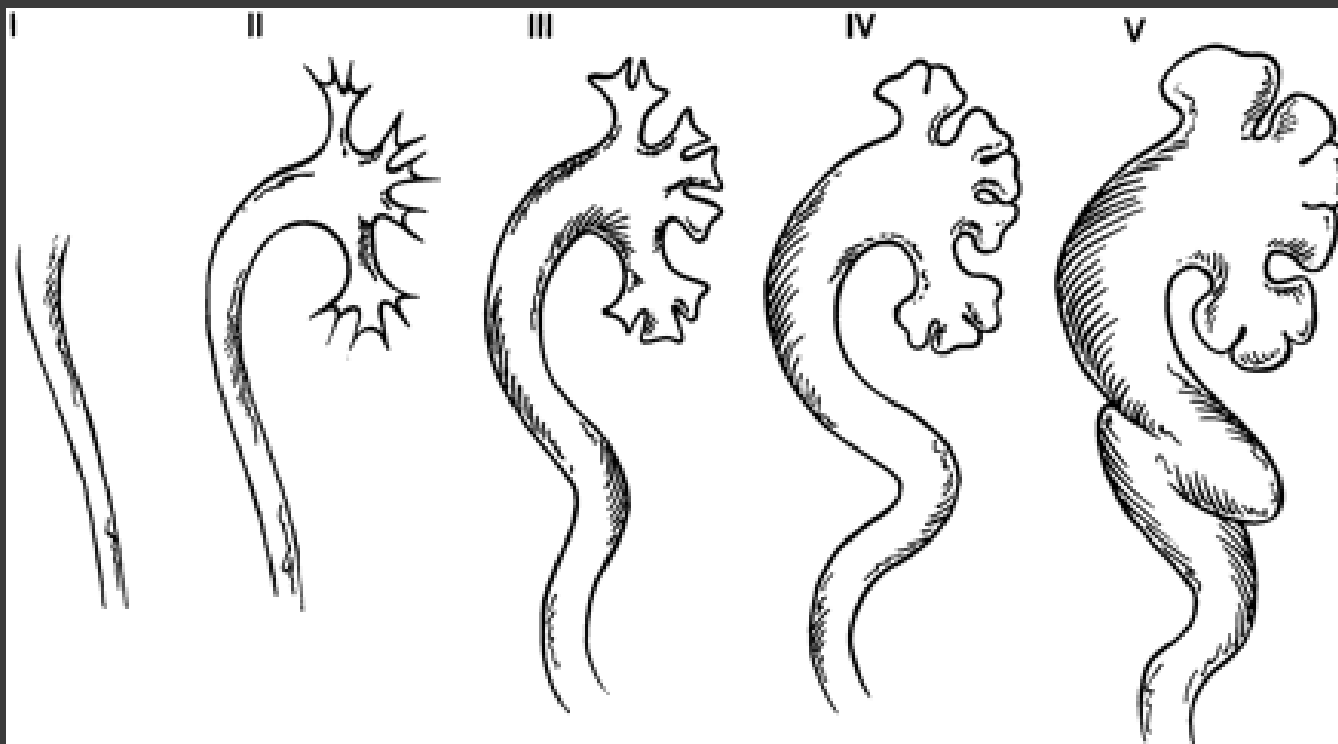


Image Source: <http://radiographics.rsna.org/content/20/1/155/F10.expansion.html>



**Now, we come to the other locations  
where this tumor can occur**



# Other Locations

Can occur almost anywhere in the body

## ◎ Usual

- Testicular
- Orbital
- Parameningeal
- Extremities

## ◎ Unusual

- Pancreas



# Companion Patient #1

## Testicular Ultrasound

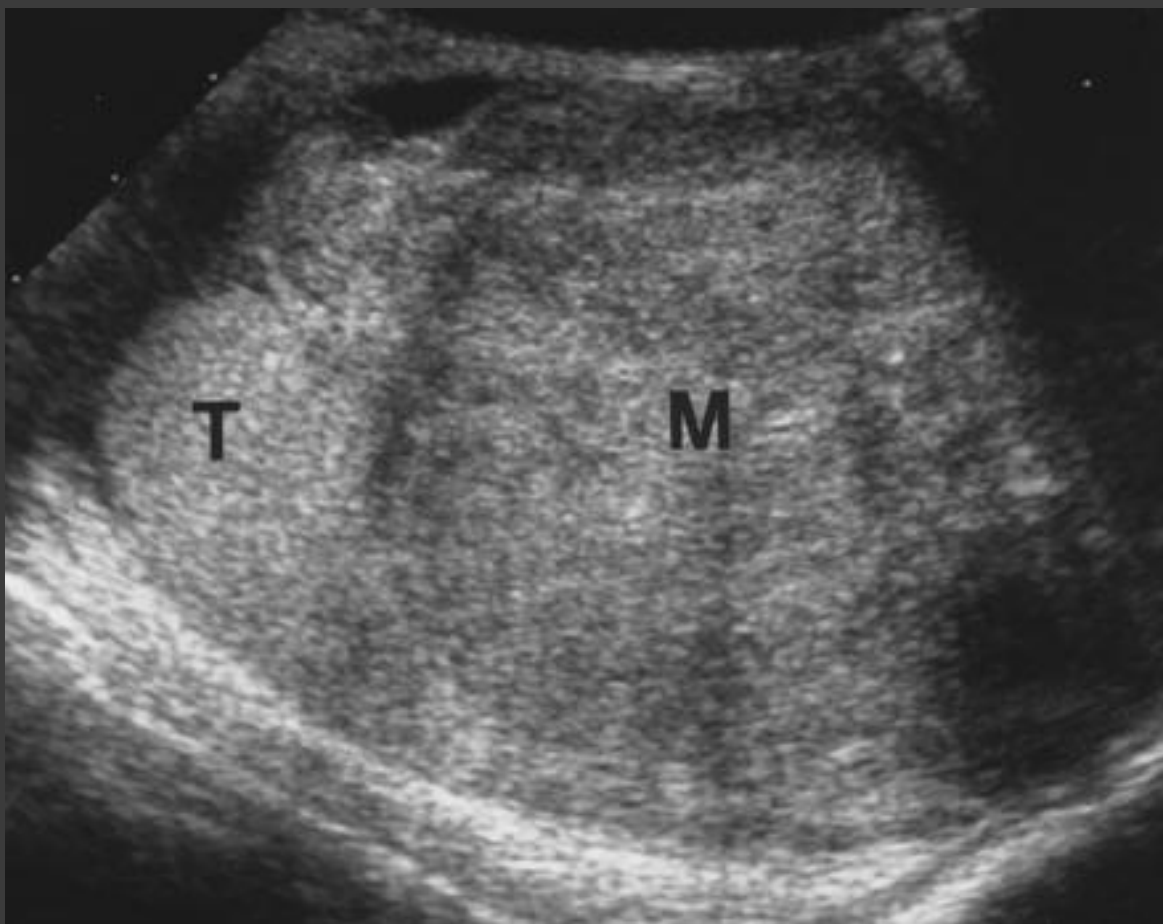


Image Source: <http://www.ajronline.org/content/176/6/1563.full>

In this patient, the mass was seen in the testicle as seen in the ultrasound.

## Ultrasound

T – Testicle      M – Mass

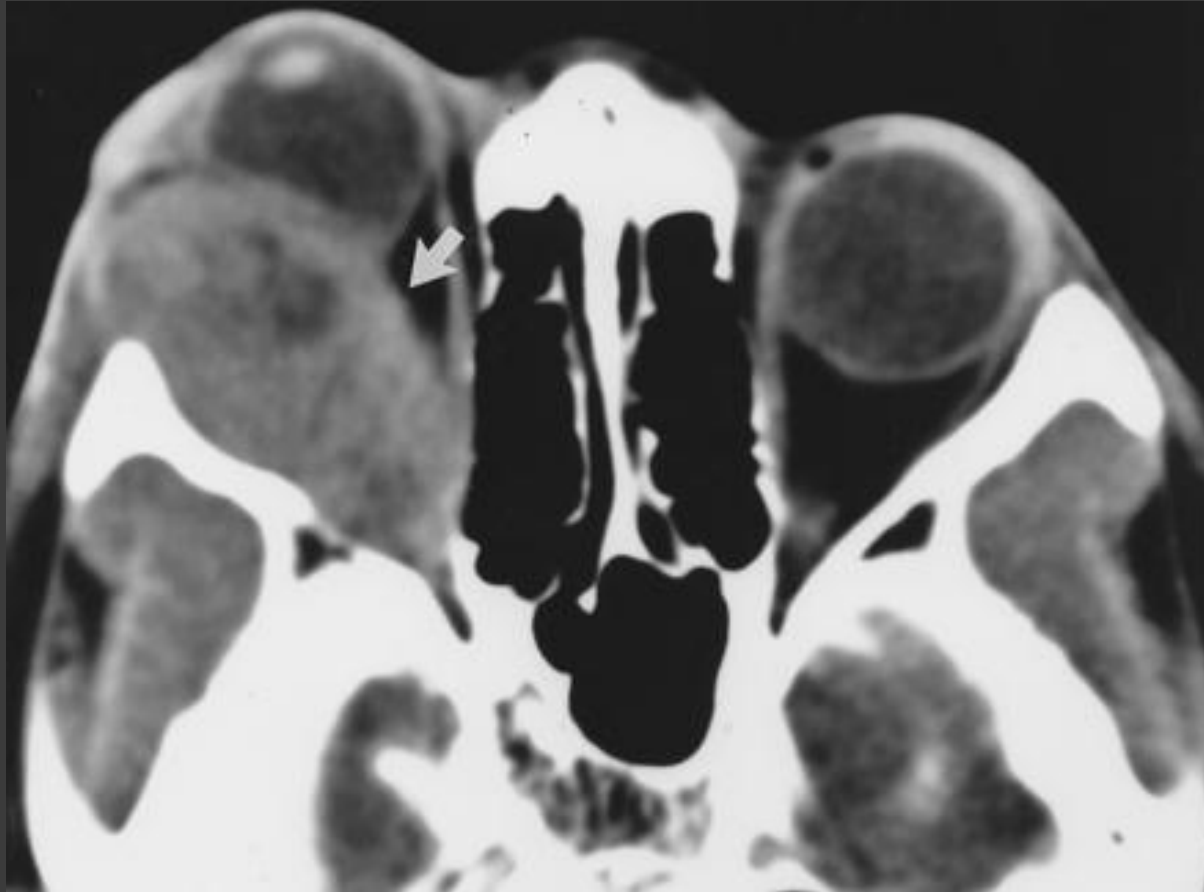
## Findings:

- Hypoechoic paratesticular mass
- Heterogenous in nature



# Companion Patient #2

## Orbital Contrast-enhanced CT Scan



In this case, the patient presented early as the mass caused proptosis and visual changes.

Arrow - Right Orbital Mass

### Findings:

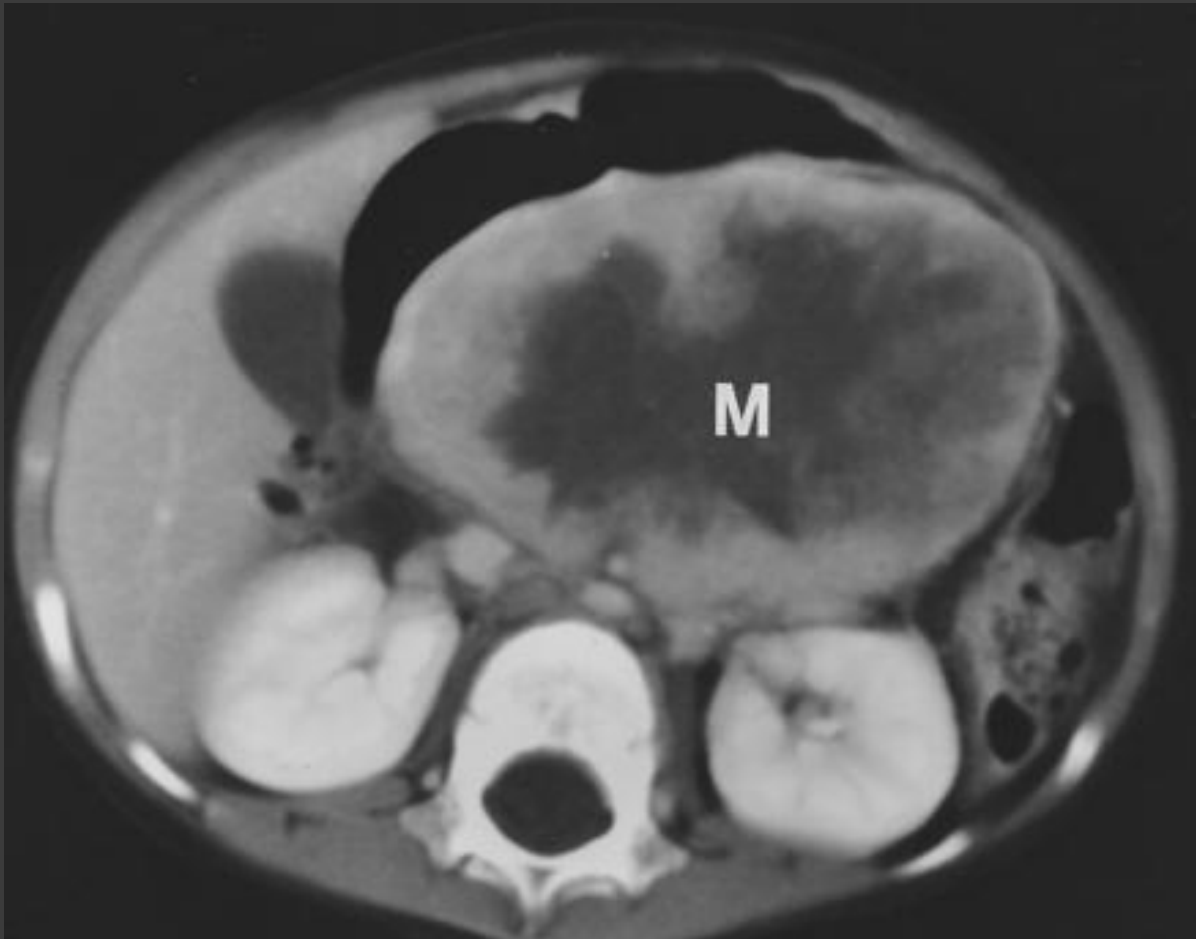
- Heterogenous mass causing proptosis.

Image Source: <http://www.ajronline.org/content/176/6/1563.full>



# Companion Patient #3

## Pancreas - Contrast-enhanced CT Scan



**M – Large Mass**

### Findings:

- Large centrally necrotic mass
- (biopsy showed pancreatic mass)

Image Source: <http://www.ajronline.org/content/176/6/1563.full>



# Prognosis of Rhabdomyosarcoma

Prognosis is different depending on the location and the age of the patient

## LOCATION

- ⦿ In patients with a localized disease – Prognosis good.
  - The 5-year survival rate - 80%
- ⦿ In patients with metastatic disease – Prognosis poor
  - The 5-year survival rate - less than 30%

## AGE

- ⦿ Highest is children 1-4 years of age – 77%
- ⦿ Poor in infants and adolescents – 47%





# Summary

- Rhabdomyosarcoma is a malignant tumor that can occur almost anywhere in the body.
- Signs / Symptoms depend on the location of the tumor.
- Treatment varies depending on the location and the stage of the tumor.
- Prognosis is better for localized disease compared to metastatic one and better for children 1-4 years of age compared to adolescents.



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