

# Tailgut Cysts; shedding light on an increasingly identified entity plus selected case review

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# Objectives

- Describe the epidemiology, embryology, anatomy, pathology, clinical presentation and complications
- Illustrate differential diagnoses
- Review imaging features and diagnostic approach
- Outline different treatment modalities
- Review selected cases

# Questions

- The mean age of presentation of tailgut cysts is:
  1. <10 years old
  2. 10-30 years old
  3. 30-60 years old
  4. >60 years old
- The most common complication of tailgut cysts is:
  1. Infection
  2. Malignant degeneration
  3. Fistula formation
  4. Rupture
- The most common presentation of tailgut cysts is:
  1. Rectal pain
  2. Painless rectal bleeding
  3. Constipation
  4. Dysuria
  5. Asymptomatic
- The imaging modality of choice for the diagnosis of tailgut cysts is:
  1. Ultrasound
  2. Computed Tomography
  3. Magnetic Resonance Imaging
  4. PET scan

# Introduction

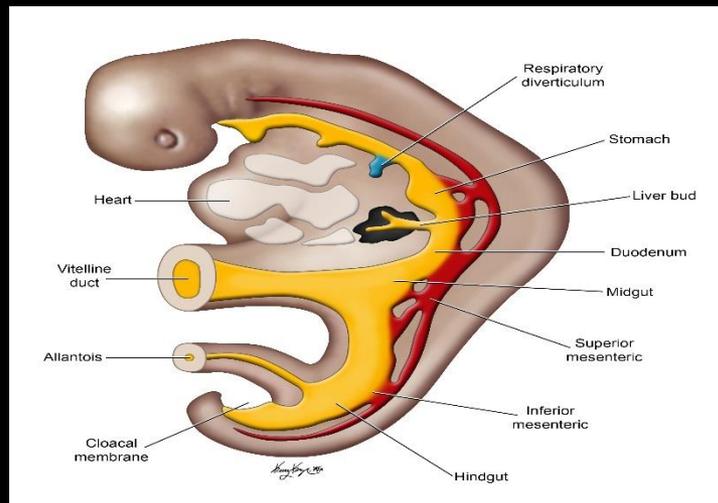
- Tailgut cysts, also called retrorectal cystic hamartomas, are congenital/developmental lesions that arise from vestiges of the embryological hindgut.
- They almost always occur in the retrorectal (presacral) space.
- Retrorectal space is a potential space filled with various elements that can develop different benign and malignant conditions that usually present as masses.
- These masses can be congenital, inflammatory, embryological remnants, neurogenic, or osseous in nature.
- These lesions are rare and can present a challenge both in diagnosis and management.

# Epidemiology

- Tailgut cysts are rare.
- Incidence of retrorectal lesions in general is estimated at 1 in 40,000 based on Mayo Clinic data.
- Congenital lesions account for about 2/3<sup>rd</sup> of retrorectal lesions(excluding inflammatory lesions).
- There is a strong female predilection with a 3:1 female to male ratio.
- They occur at a mean age of 35 years and most commonly occur between ages 30-60.

# Embryology

- In the course of normal development, the embryo forms a true tail which is maximal at 35 days of gestation.
- The embryonic hindgut extends into the tail, constituting the tailgut, which normally undergoes regression by the 8th week of gestation.
- The persistence of the tailgut may remain and lead to the development of tailgut cyst.



# Embryology

- Development:

Embryo starts to fold inward

4th week of gestation

Cloacal membrane becomes ventral to the caudal part of the hindgut and encloses it

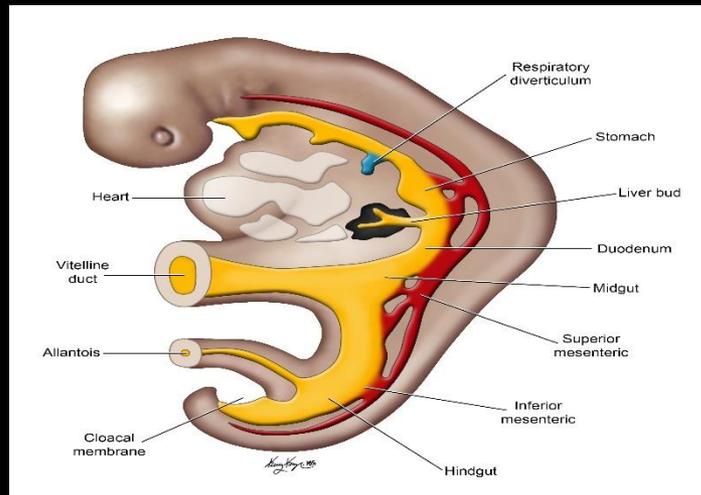
Becomes the Tailgut

8th week of gestation

Normally involutes

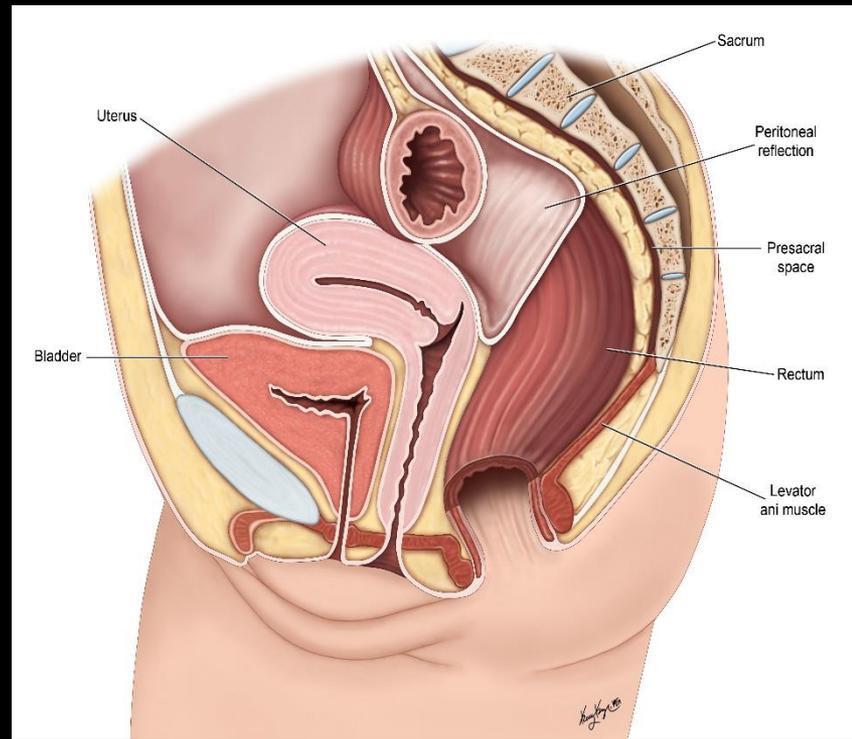
Persists

Tailgut Cyst



# Anatomy

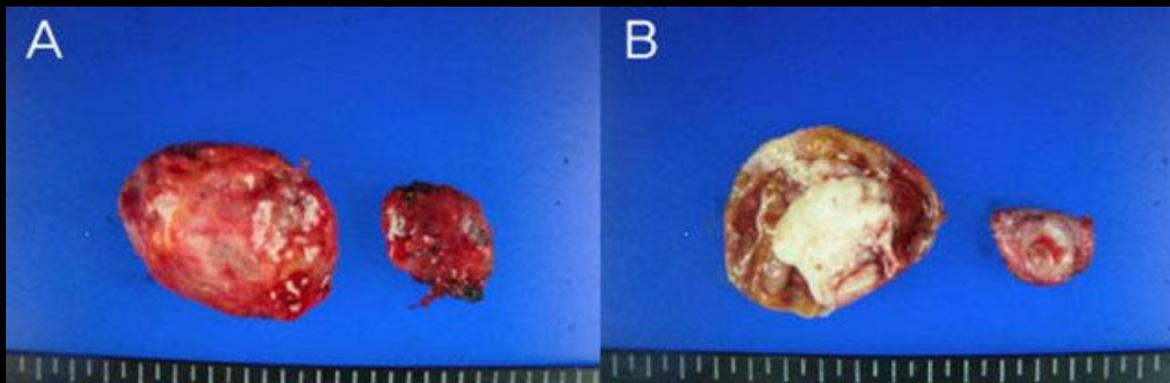
- The retrorectal (presacral) space is a potential space bounded:
  - Anteriorly by the rectum
  - Posteriorly by the sacrum
  - Superiorly by peritoneal reflection
  - Inferiorly by the levator ani and coccygeus muscles
  - The iliac vessels and ureters define the lateral borders.



# Pathology

## ❖ Gross

- Well circumscribed
- Multiloculated
- Thin wall with glistening lining
- Filled with mucoid material
- Size ranges from 1-22 cm; average is about 4 cm

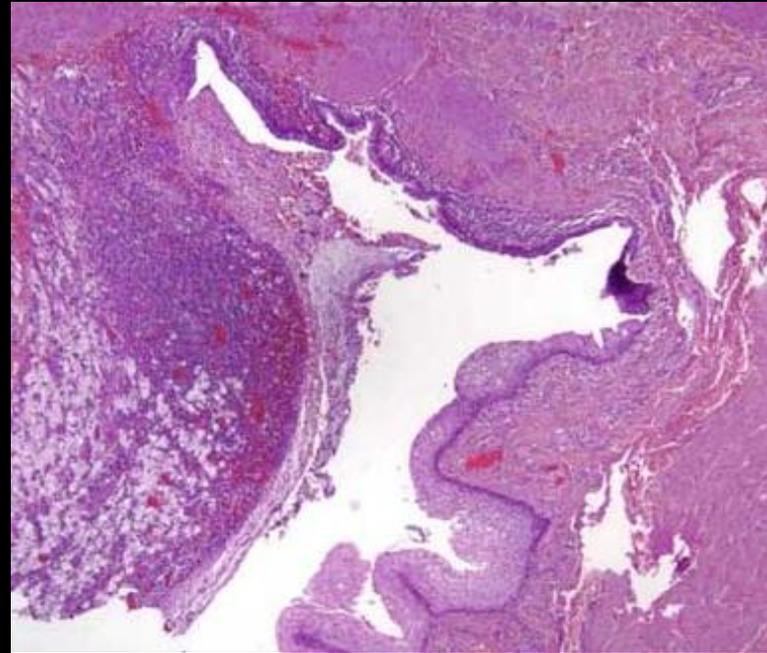


Gross appearance (A) and cut surface (B) of resected tailgut cysts

# Pathology

## ❖ Microscopic

- Can be lined by any type of adult or fetal GI tract epithelium.
  - a) Stratified squamous is most common
  - b) Transitional
  - c) Cuboidal
  - d) Mucinous
- Epithelium is underlined by fibroconnective tissue stroma.
- Scattered, disorganized smooth muscle bundles.



Low power view of squamous and glandular epithelium

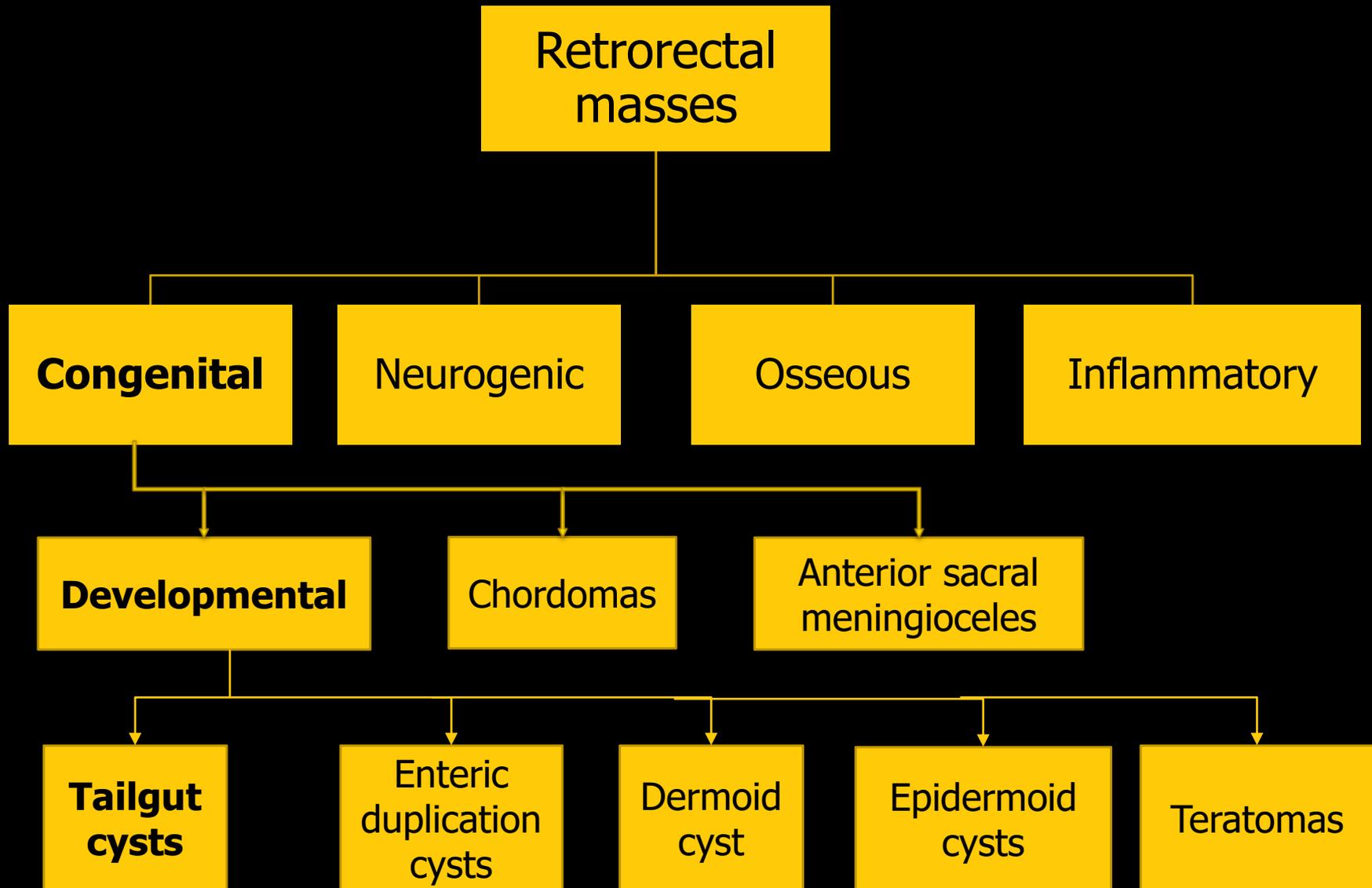
# Clinical Presentation

- According to the largest reported case series of 53 cases by Hjermsstad and Helwig, half of the patients presented with symptoms, likely from local mass effect.
- These symptoms include included rectal pain, constipation, painless rectal bleeding, dysuria and urinary frequency.
- Physical examination can demonstrate a funnel-shaped dimple in the post-anal midline.
- Digital rectal examination may reveal a smooth, firm mass in the presacral space, bulging into the rectal lumen.

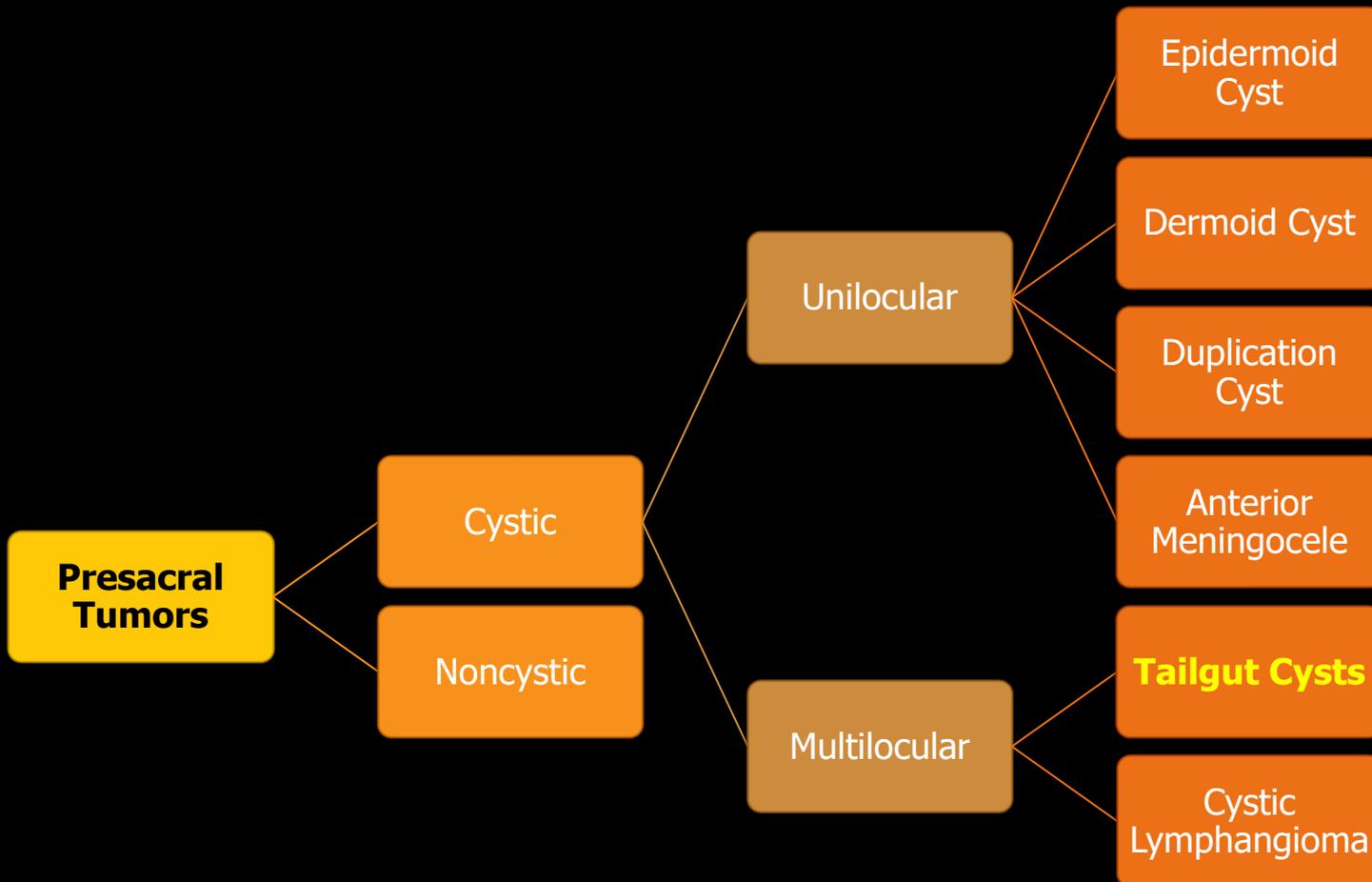
# Complications

- Infection; the most common complication, occurring in 30-50% of patients → may result in abscess formation or perianal fistula with discharge of pus.
- Bleeding; Not very common.
- Malignant degeneration into adenocarcinomas, carcinoids, and sarcomas → The largest series in the literature, by Hjermsstad and Helwig, reported a 2 % incidence. **However**, incidence was reported at 13% by a recent Mayo Clinic report.

# Differential Diagnoses



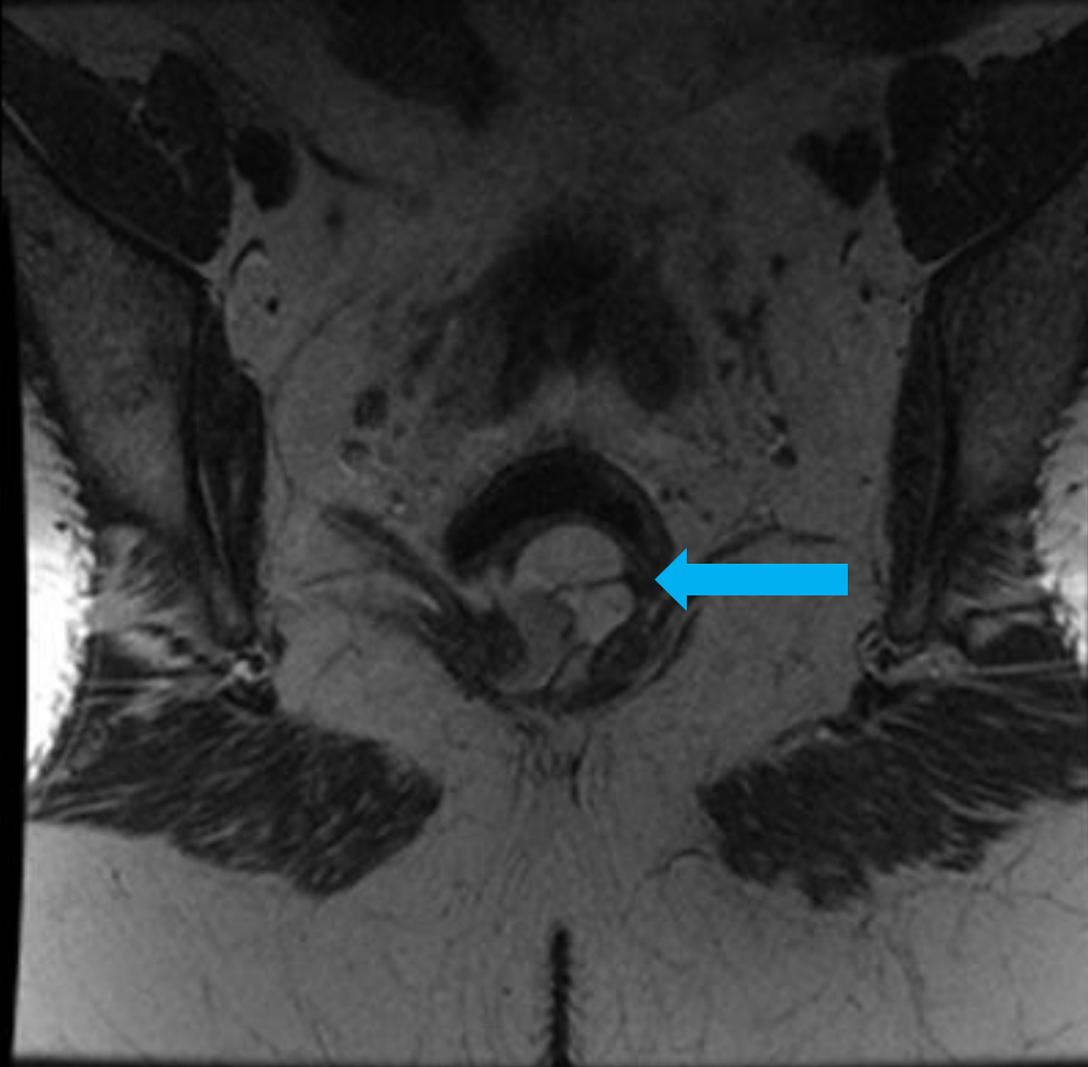
# Imaging Approach



# Imaging Features

- Classic findings:
  - Multilocular, retrorectal.
  - Low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.
- Alternatively:
  - High signal intensity on T1-weighted images due to presence of mucinous materials, high protein content, or hemorrhage in the cyst.

# Imaging Findings



Oblique axial T2 weighted image shows T2 hyperintense lesion dissecting through the right levator ani musculature

# Imaging Findings



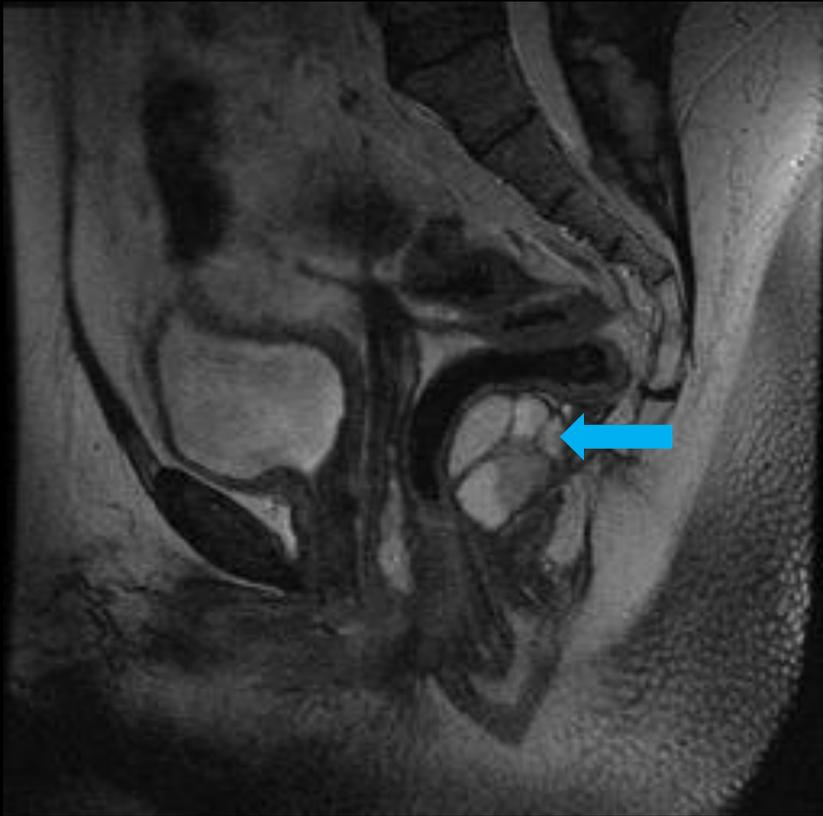
Sagittal T2 weighted image demonstrates a multiloculated, T2 hyperintense lesion posterior to the rectum

# Treatment

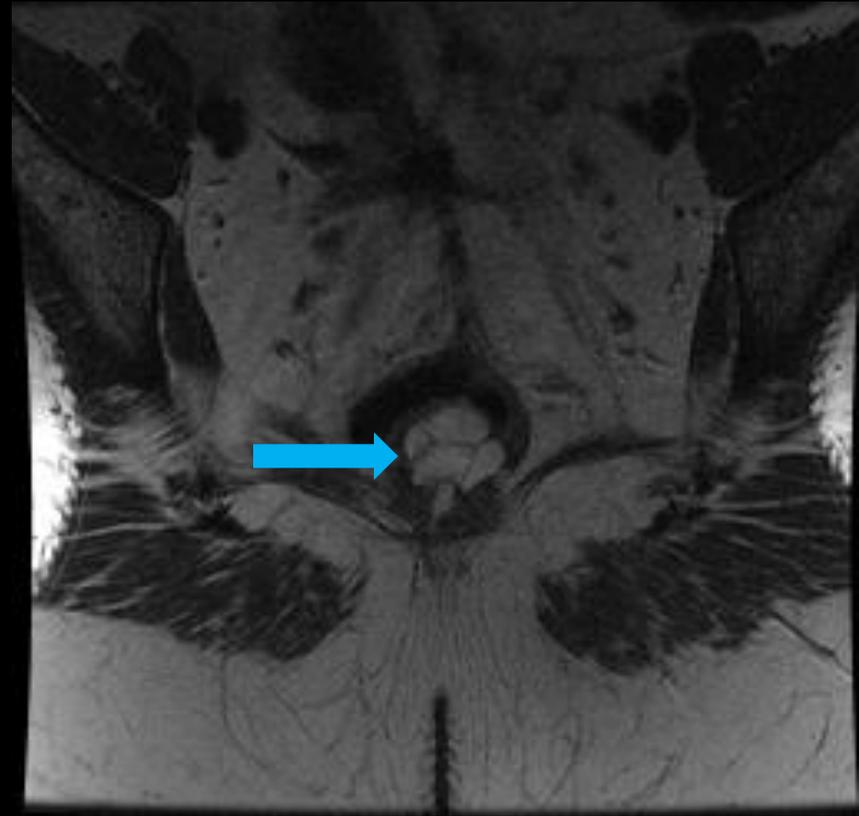
- ❖ Once diagnosed, complete excision is the treatment of choice for both asymptomatic and symptomatic patients to prevent potential complications.
- ❖ Excision may be accomplished by transanal, transrectal, transabdominal or combined approach.
- ❖ The most described approach is posterior parasacral incision with preservation of the coccyx unless en bloc resection is required for malignancy or the cyst is densely adherent to the coccyx.

# Case 1

- 57 year old African American female, asymptomatic.



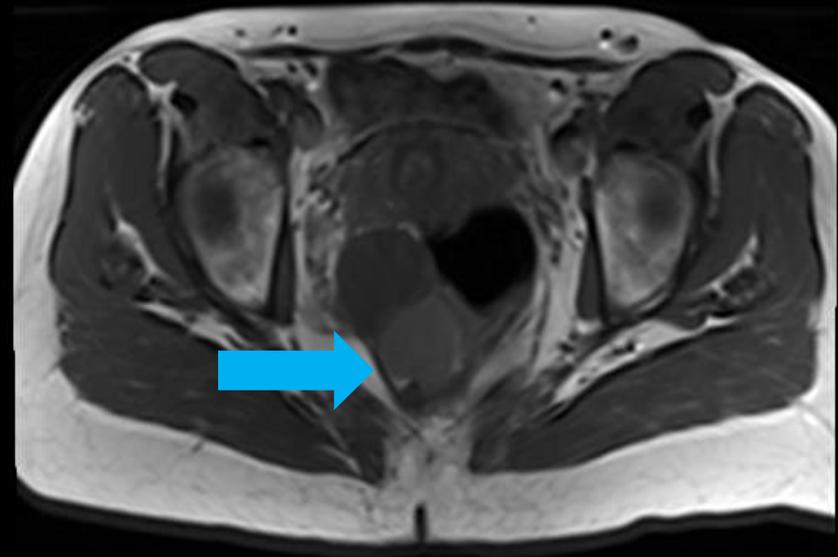
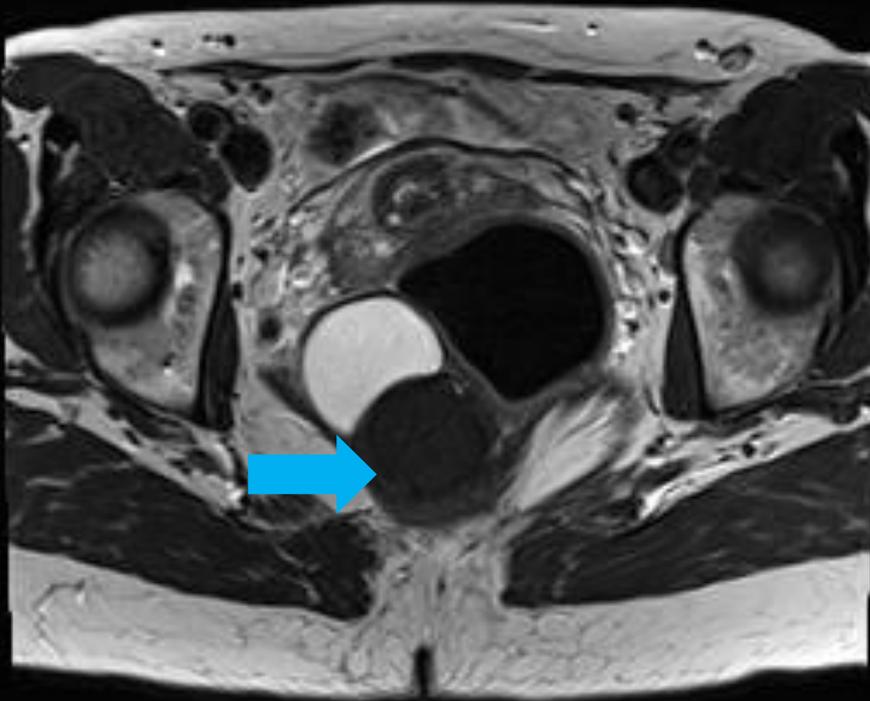
a) Multilocular cystic mass hyperintense on T2 sagittal in presacral space (blue arrow)



b) Part of the cystic lesion dissects through the right levator ani musculature (blue arrow) on T2 axial

## Case 2

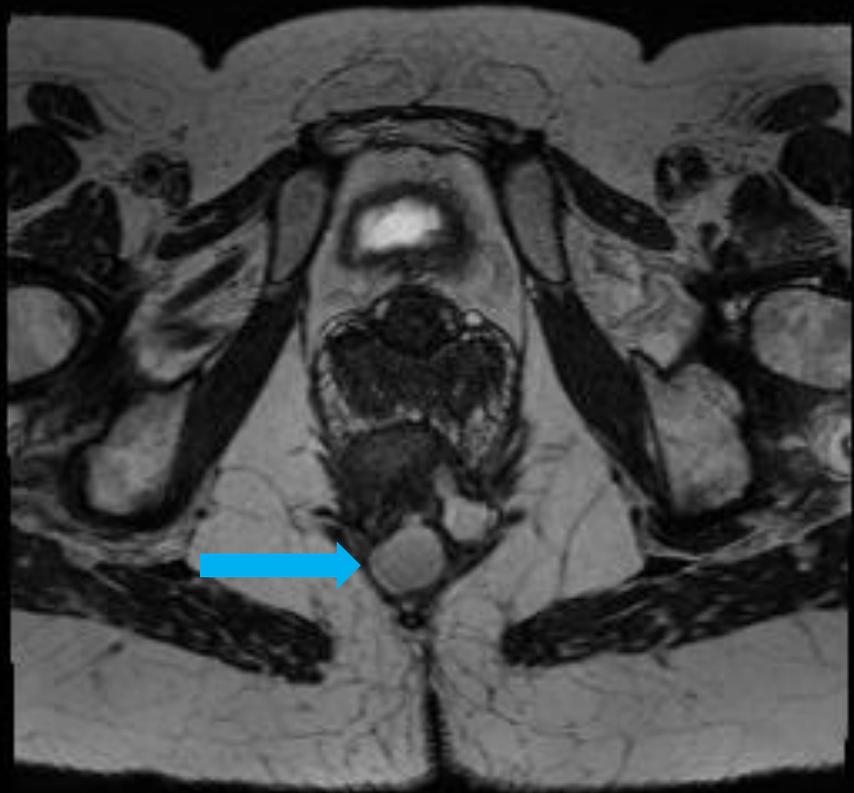
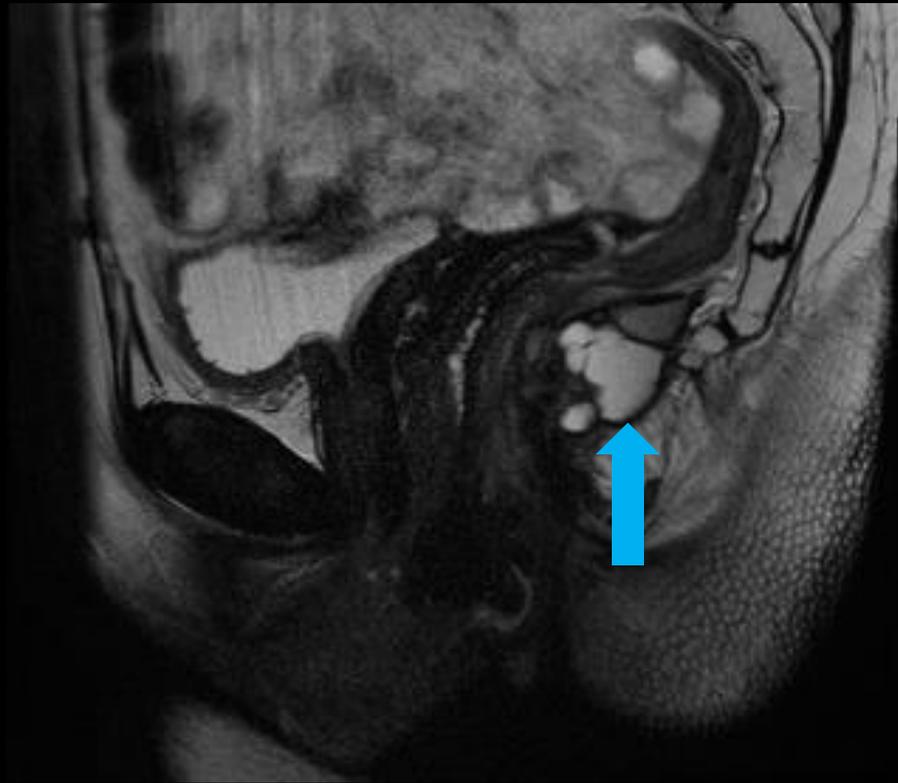
➤ 48 year old white female, asymptomatic



Several loculations demonstrate low signal intensity of T2 and high signal intensity on T1 weighted images due to presence of mucinous materials, high protein content, or hemorrhage in the cyst (blue arrows)

# Case 3

➤ 51 year old white female, asymptomatic



Sagittal and Axial T2 images demonstrate a multiloculated, cystic mass causing slight anterior displacement of the rectum (blue arrows)

# Questions

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# Conclusion & Clinical Implications

- Tailgut cysts are rare but important lesions to recognize.
- Mean age of presentation is 35 years old with a strong female predominance.
- Almost half the cases are asymptomatic, discovered incidentally during imaging for another reason.
- The most common symptoms are back/rectal pain, rectal bleeding and urinary frequency.
- Possible complications include infection, fistula formation, and the possibility of malignant transformation.
- U/S, CT and MRI can be used to detect these cysts with MRI increasingly becoming the modality of choice.
- Typical MRI findings: Multilocular lesion with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images but presence of mucus can alter the picture.
- The treatment of choice for both symptomatic and asymptomatic patients is complete excision to prevent the previously mentioned complications especially malignant transformation.

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