Clinical algorithms for the surgical management of retro-rectal tumours

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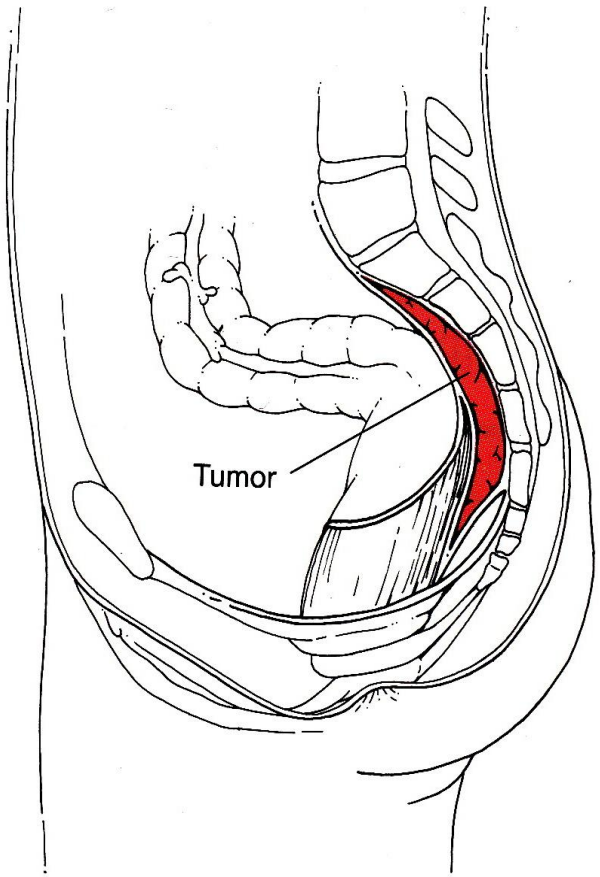
The retrorectal space

- Potential space
- Anterior - mesorectum
- Posterior - sacrum
- Inferior - rectosacral fascia
- Lateral - ligaments, ureters & iliac vessels



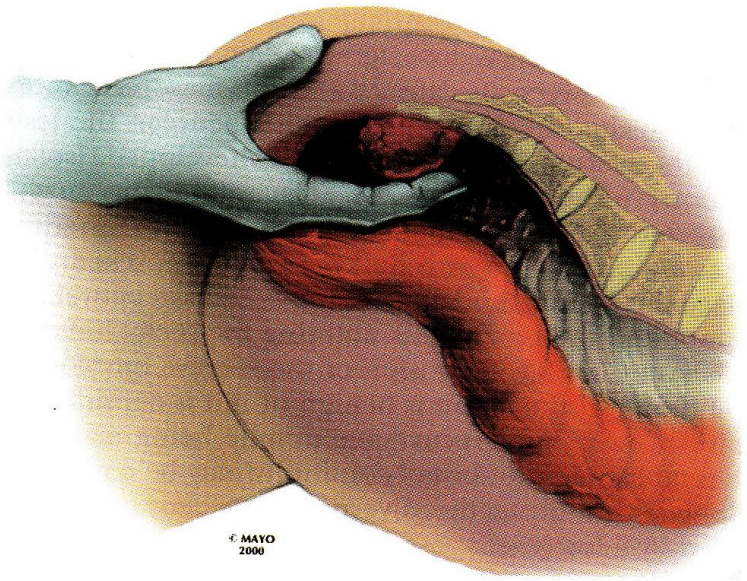
Which team ?

History

- Often non-specific or absent
- Pain - vague, long duration
- Constipation
- Urinary or fecal incontinence
- Perianal discharge
- Obstructed labor

Examination

- Extrarectal mass on digital rectal exam
- Assess fixation
- Determine level
-
- Presence of postanal dimple or previous scars



Classification

I. Congenital
   A. Developmental cysts
      1. Epidermoid
      2. Dermoid
      3. Mucus secreting
      4. Teratomas
   B. Teratocarcinoma
   C. Chordoma
   D. Anterior sacral meningocele

II. Nerve
   A. Ganglioneuroma
   B. Ependymoma
   C. Neurilemmoma
   D. Neurofibroma
   E. Neurofibrosarcoma

III. Cartilage, bone, and muscle
   A. Benign
      1. Osteoma
      2. Osseous cyst (simple or aneurysmal)
      3. Chondroma
      4. Leiomyoma
   B. Malignant
      1. Osteogenic sarcoma
      2. Ewing's tumor
      3. Chondrosarcoma
      4. Giant cell tumor
      5. Leiomyosarcoma

IV. Adipose, fibrous, and endothelial
   A. Lipoma and liposarcoma
   B. Fibroma and fibrosarcoma
   C. Endothelioma and hemangioendothelial sarcoma
   D. Myelolipoma

V. Hematologic and lymphatic
   A. Lymphangioma and lymphangiosarcoma
   B. Plasmacytoma
   C. Hemangioma and hemangiosarcoma
   D. Pericytoma
   E. Lymphoma

VI. Traumatic and inflammatory
   A. Hematoma
   B. Abscess (perineal/pelvic/perirectal/enteric with fistula)
   C. Granuloma

VII. Miscellaneous
   A. Desmoid
   B. Endometrioma
   C. Mesenchymoma
   D. Metastatic carcinoma
   E. Recurrent pelvic carcinoma

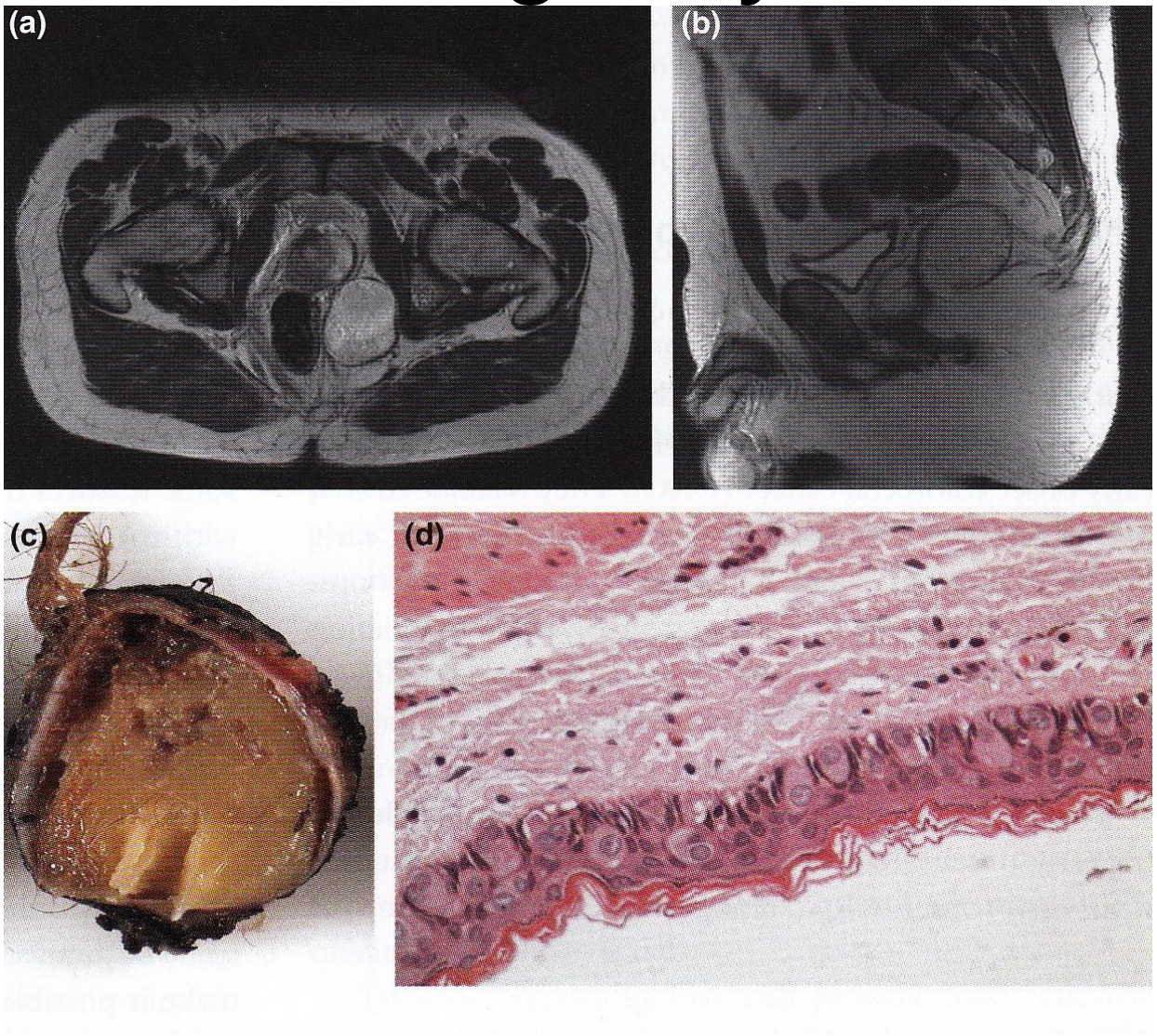
Classification

- Any tissue type within the retrorectal space may give rise to benign or malignant lesions

Tumour specific points

- Developmental cysts e.g. tail gut cysts
- Neurogenic tumours e.g. schwannoma
- Congenital neural abnormality e.g. meningocele
- Sacrococcygeal chordomas

Tailgut cysts

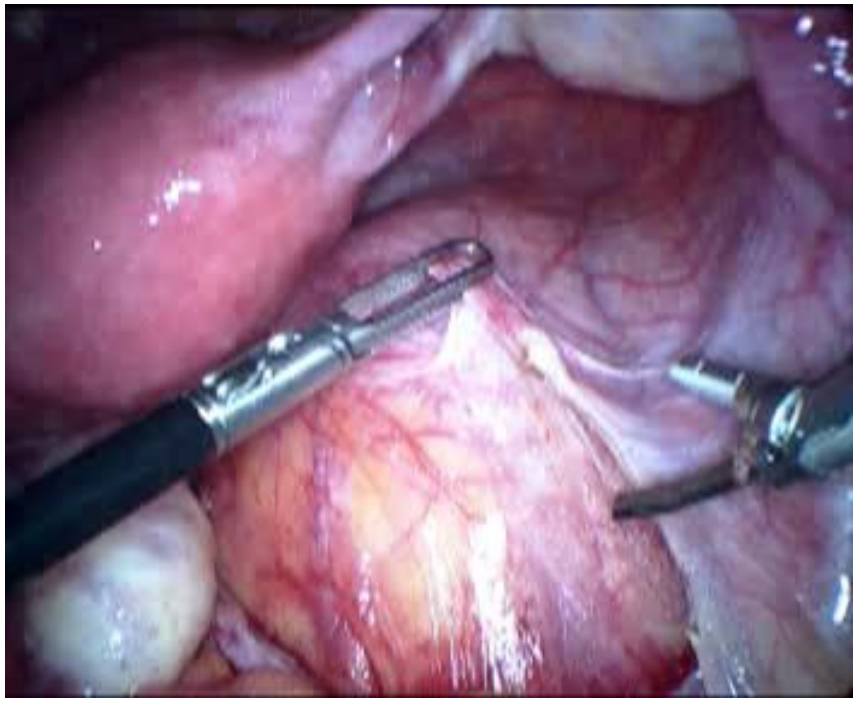


Tailgut cysts

- Remnants of embryonic primitive gut
- Multiloculated, low lying lesions
- Rarely undergo malignant change

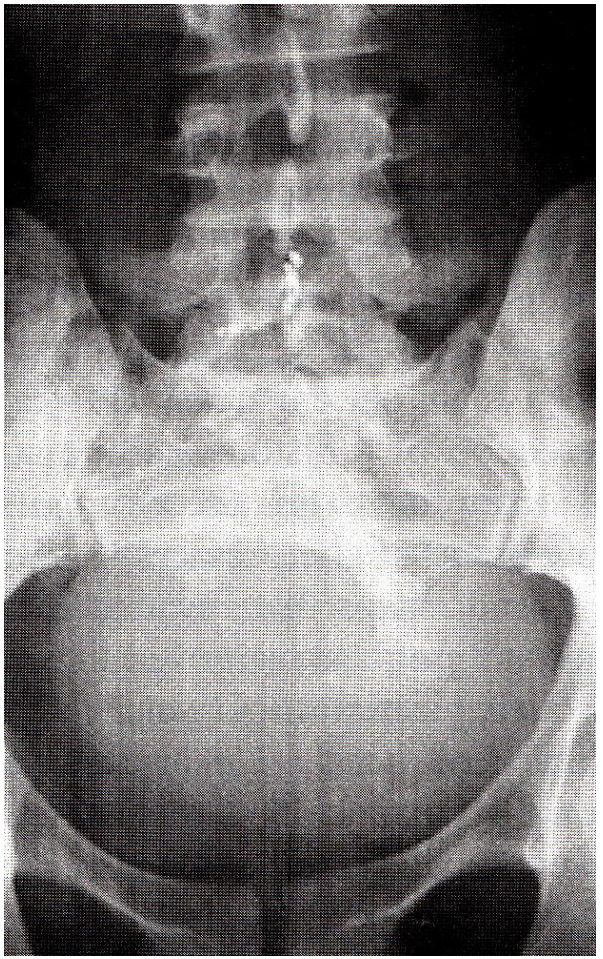
Neurogenic tumours

- Second most common
- 12% of retrorectal tumours
- Arise from peripheral nerves
- 2/3 are benign
- Tend to be large (>7 cm)



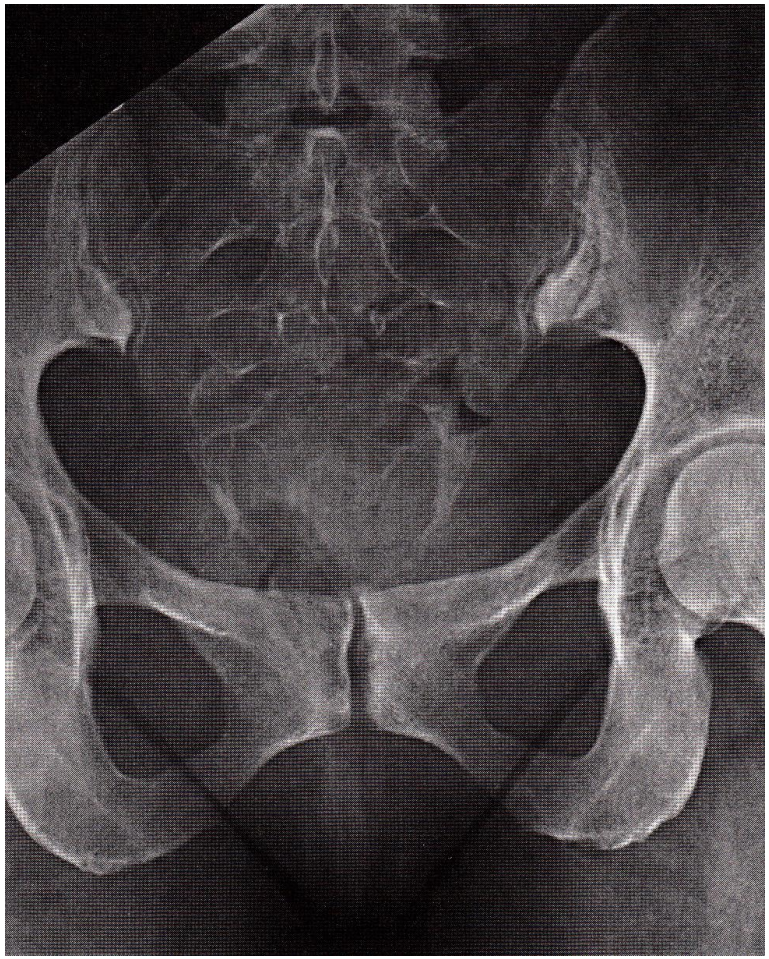
Anterior sacral meningocele

- Beware !!
- Scimitar sacrum - rounded concave border, no bony destruction
- Avoid aspiration - risk of meningitis



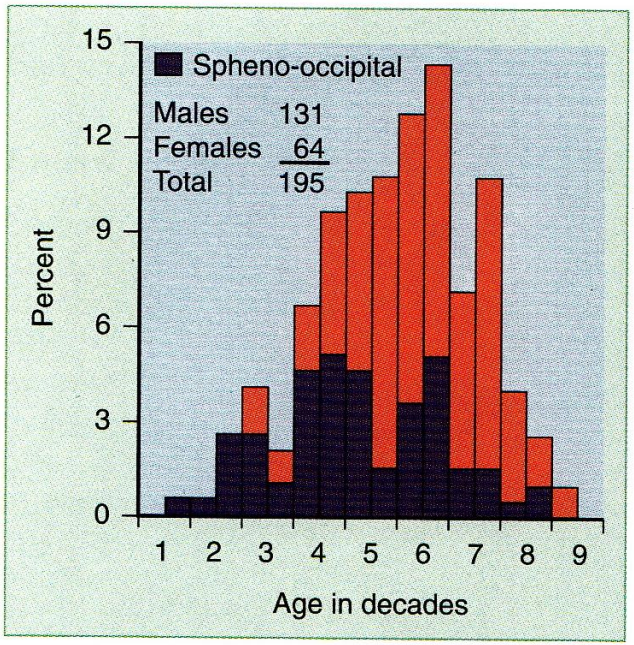
Chordoma

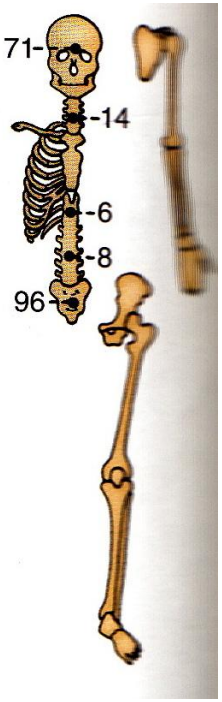
- Long standing vague pain
- “Fang” sign due to sacral bone destruction



Chordoma

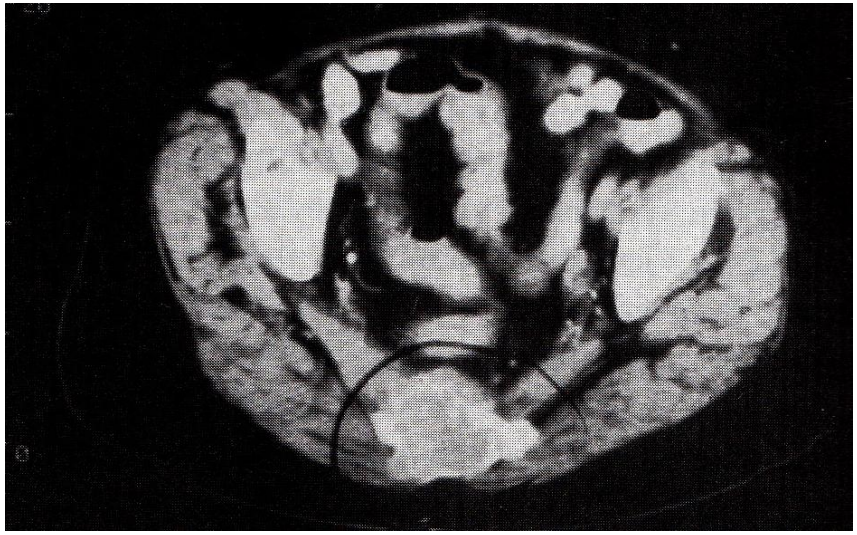
- Most common malignancy
- Arise from primitive notochordal tissue
- Prediliction for the two extermities





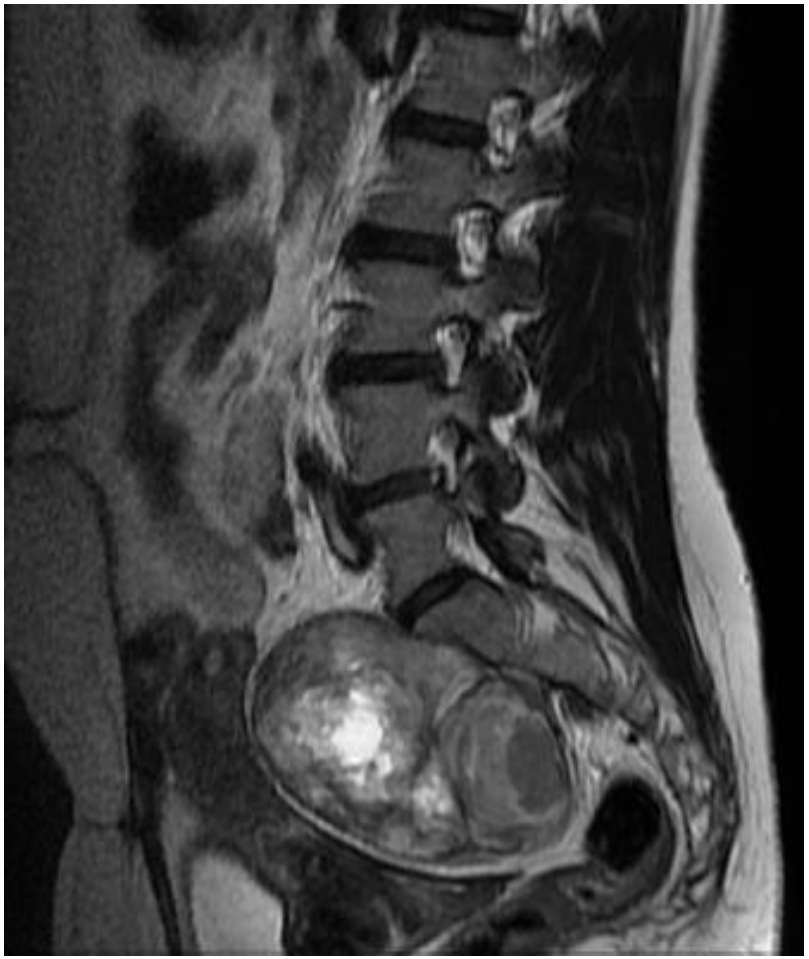
Investigations - CT

- CT scan to distinguish cystic, solid or mixed
- Involvement of adjacent structures
- Bony destruction



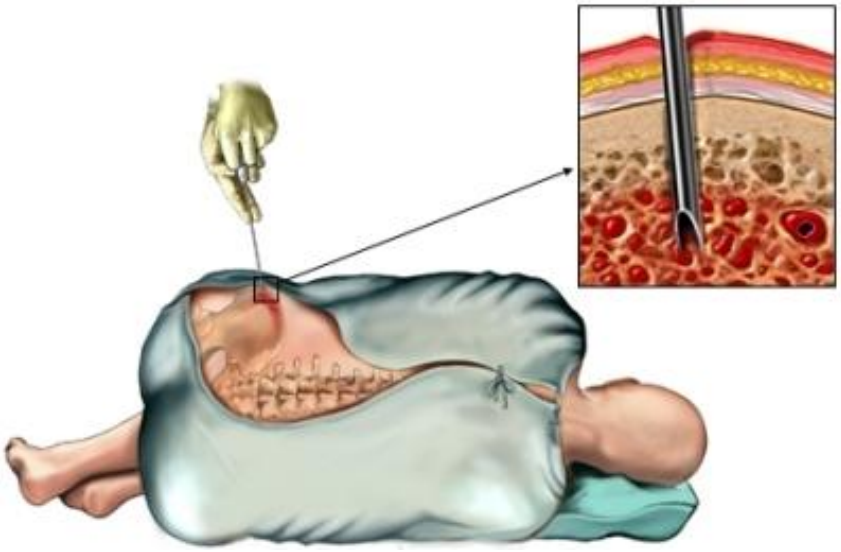
Investigations - MRI

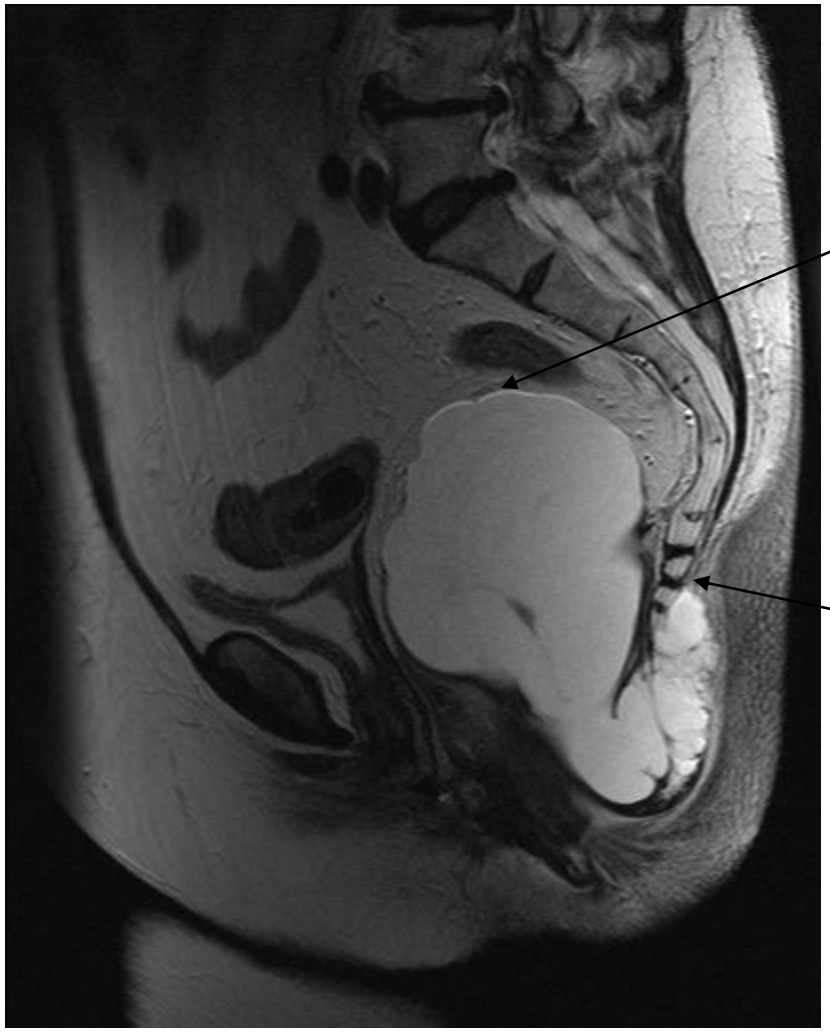
- Improved soft tissue resolution helps plan extent of resection
- Evaluation of marrow involvement
- Identifies nerve root and foraminal involvement



Role of preoperative biopsy

- Will the biopsy change treatment ?
- Not needed if mass resectable
- Do not biopsy if lesion cystic (beware meningocele)
- Consider for osteogenic sarcomas

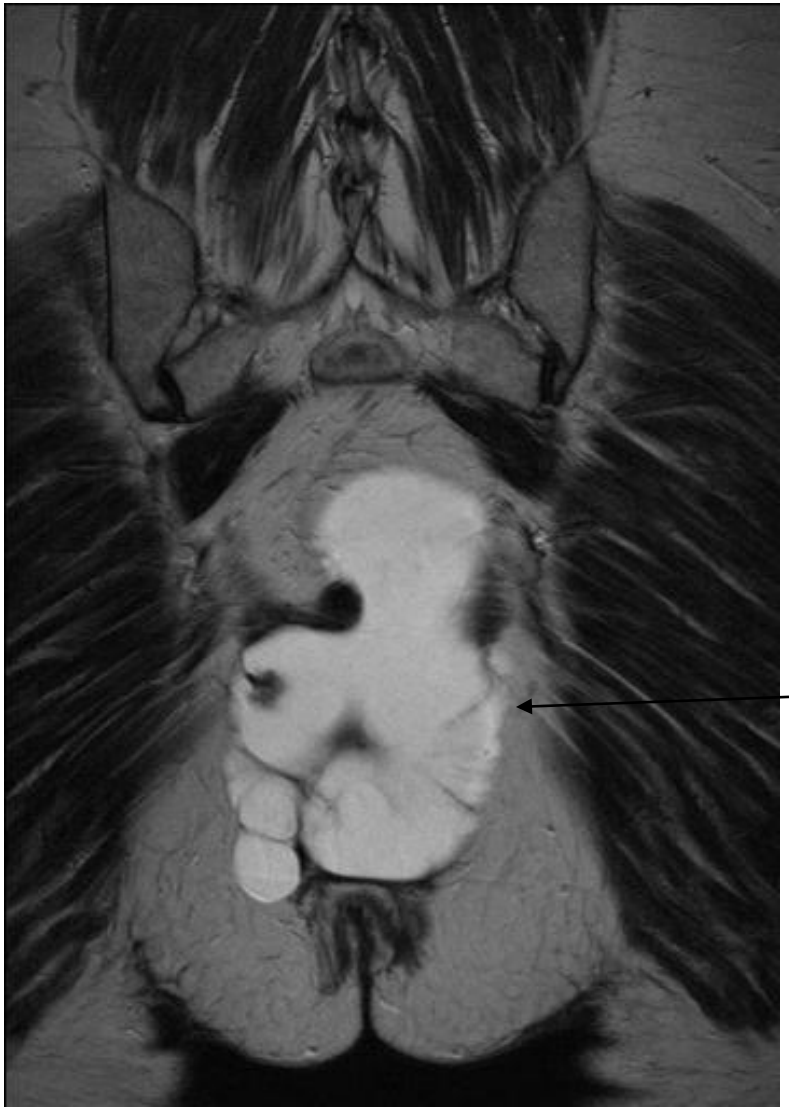




Well encapsulated fluid intensity mass

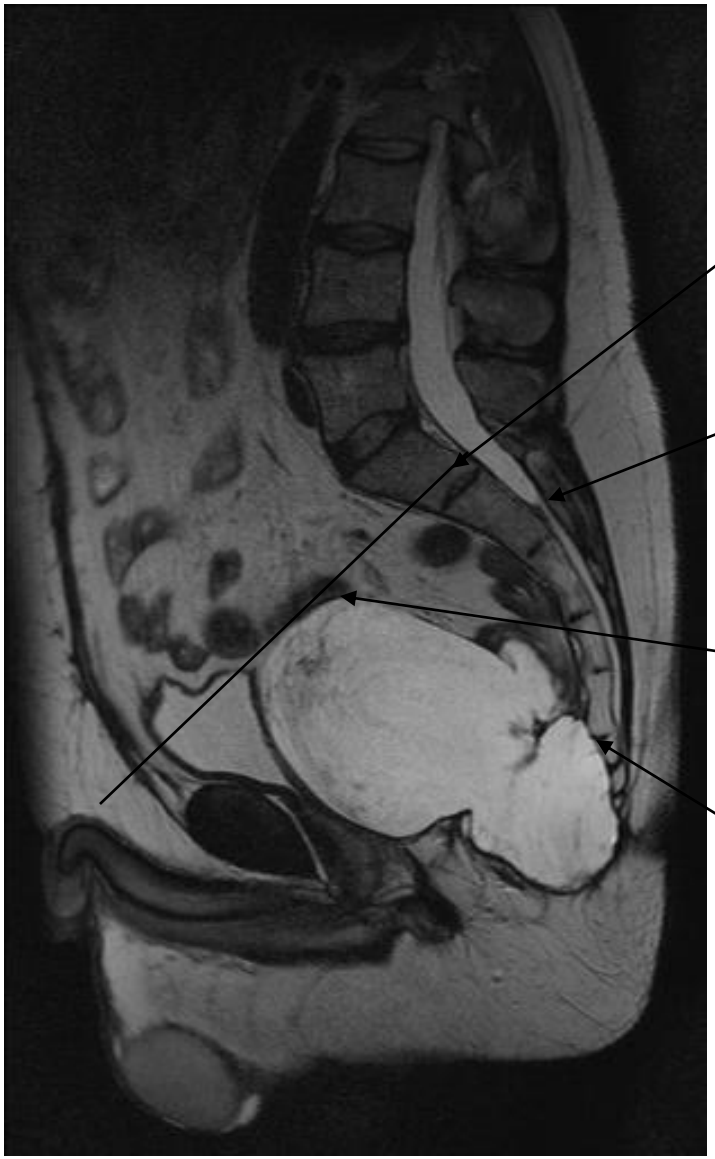
Surrounds the coccyx but no overt bony destruction or invasion

Sagittal image showing the typical appearance of a tail gut cyst (cystic hamartoma).

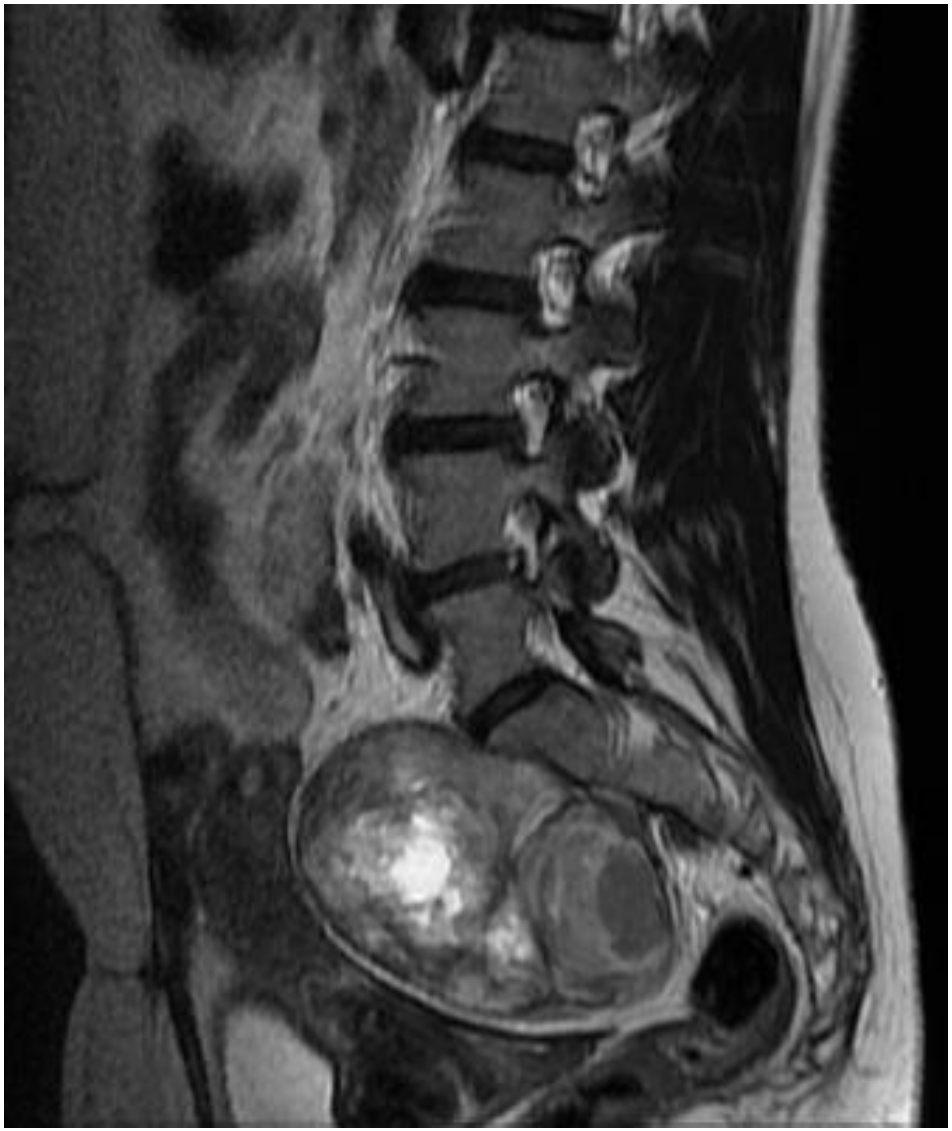


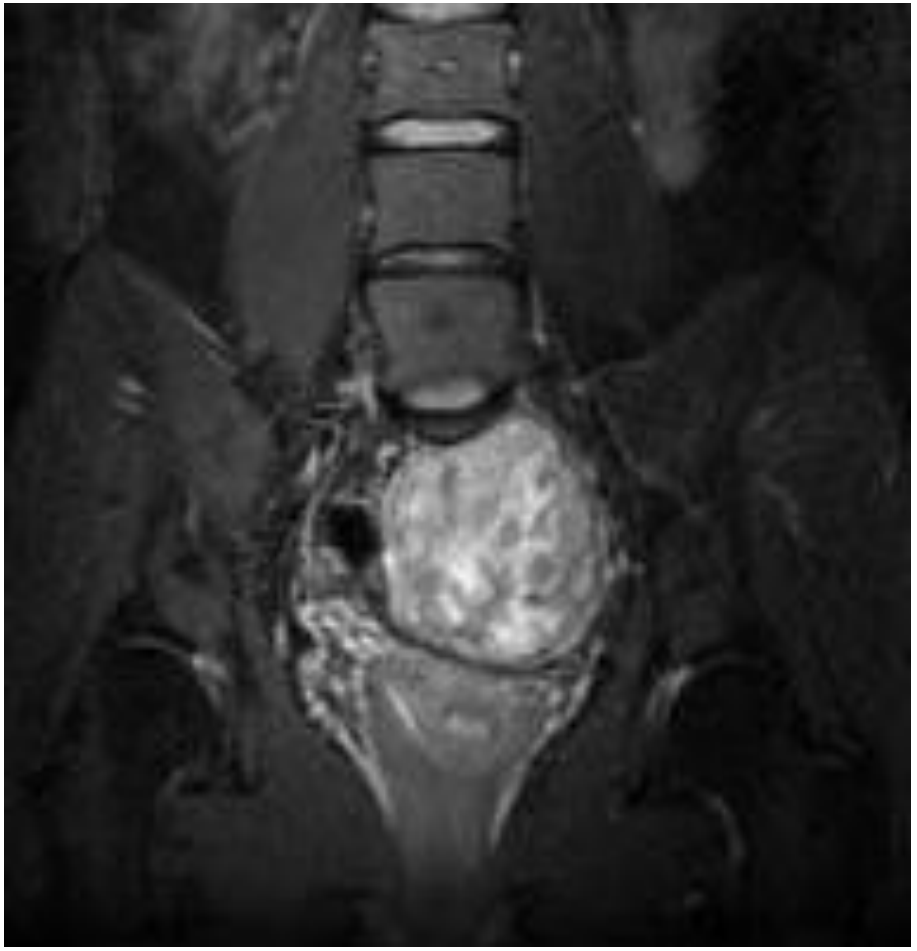
Sharply demarcated, fluid containing mass with internal septations

Coronal image showing the typical appearance of a tail gut cyst (cystic hamartoma)



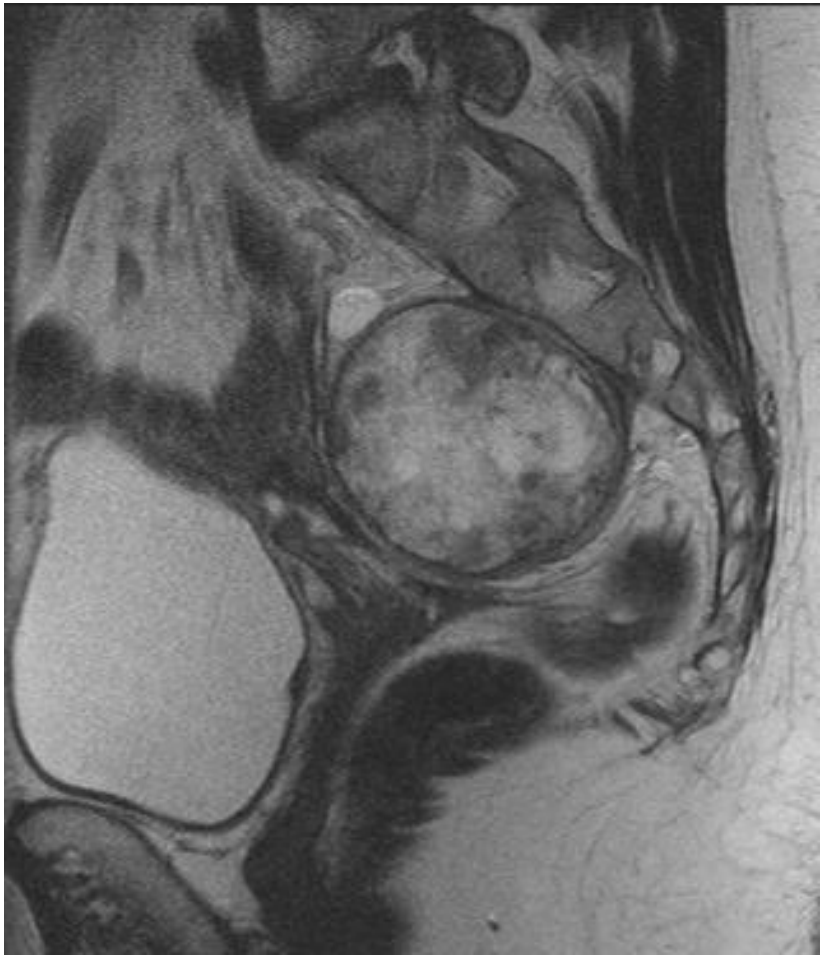
Sagittal image of a recurrent mucinous retrorectal cyst referred after previous debulking procedure and abdomino-perineal excision.



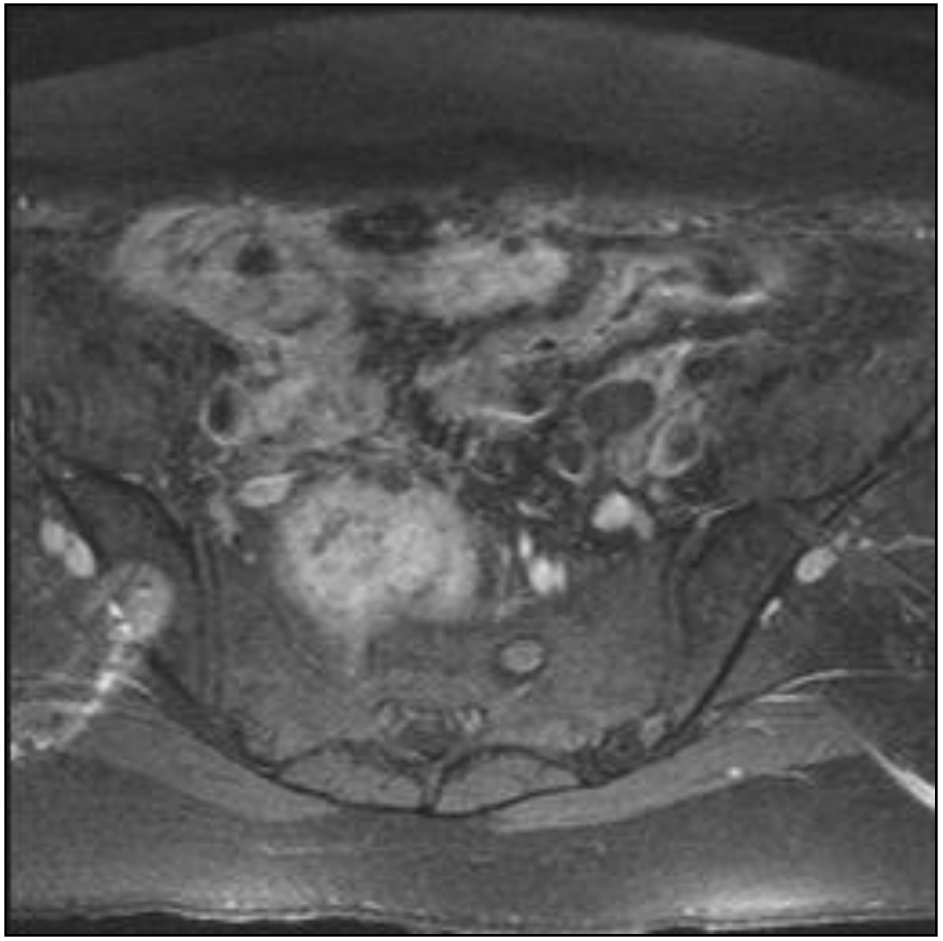


Sagittal and coronal images showing the typical well demarcated heterogenous appearance of a Schwannoma.

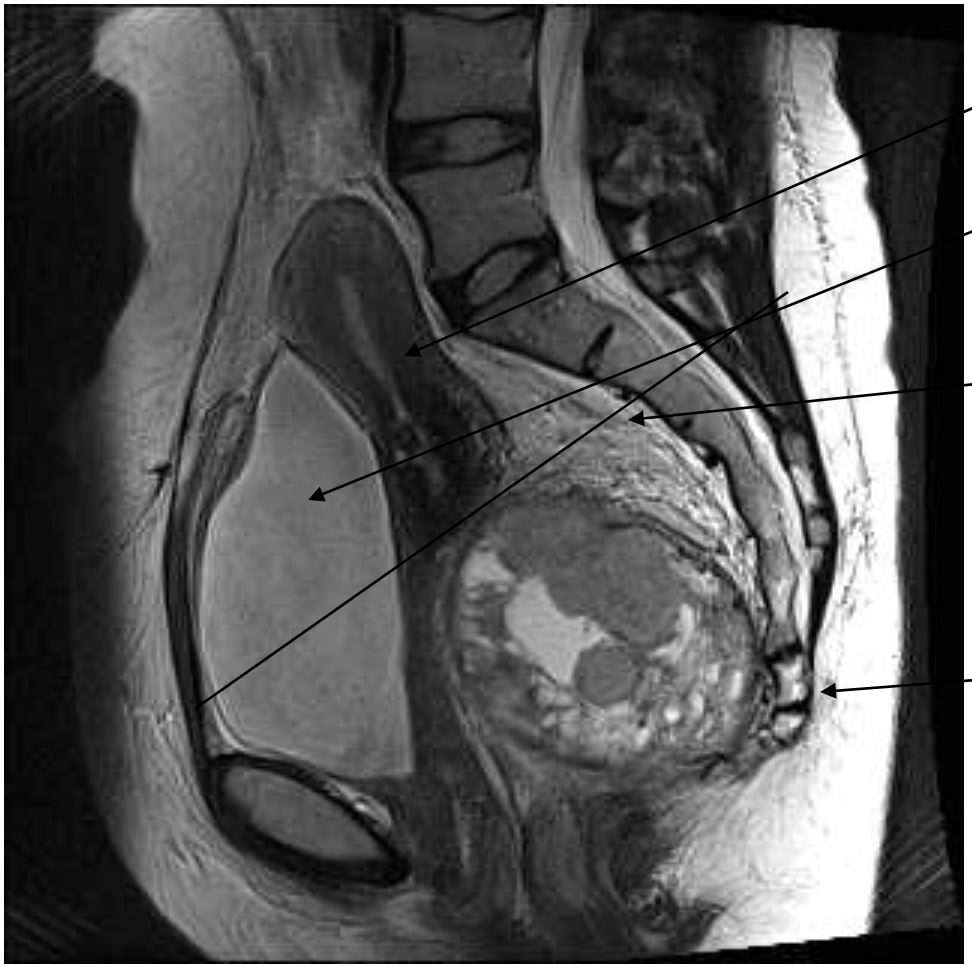
Schwannoma



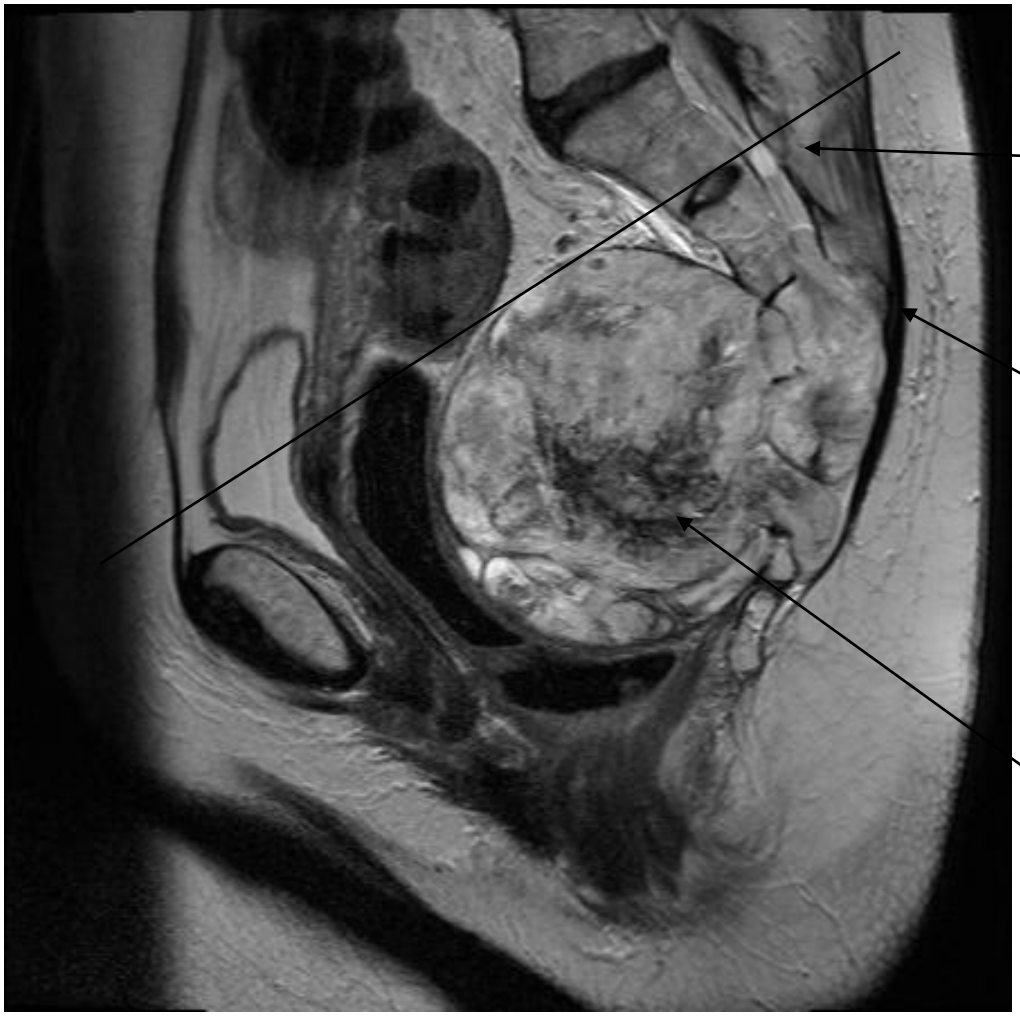
Sagittal image showing heterogenous well-demarcated pre-sacral mass with no gross evidence of bone invasion or destruction.



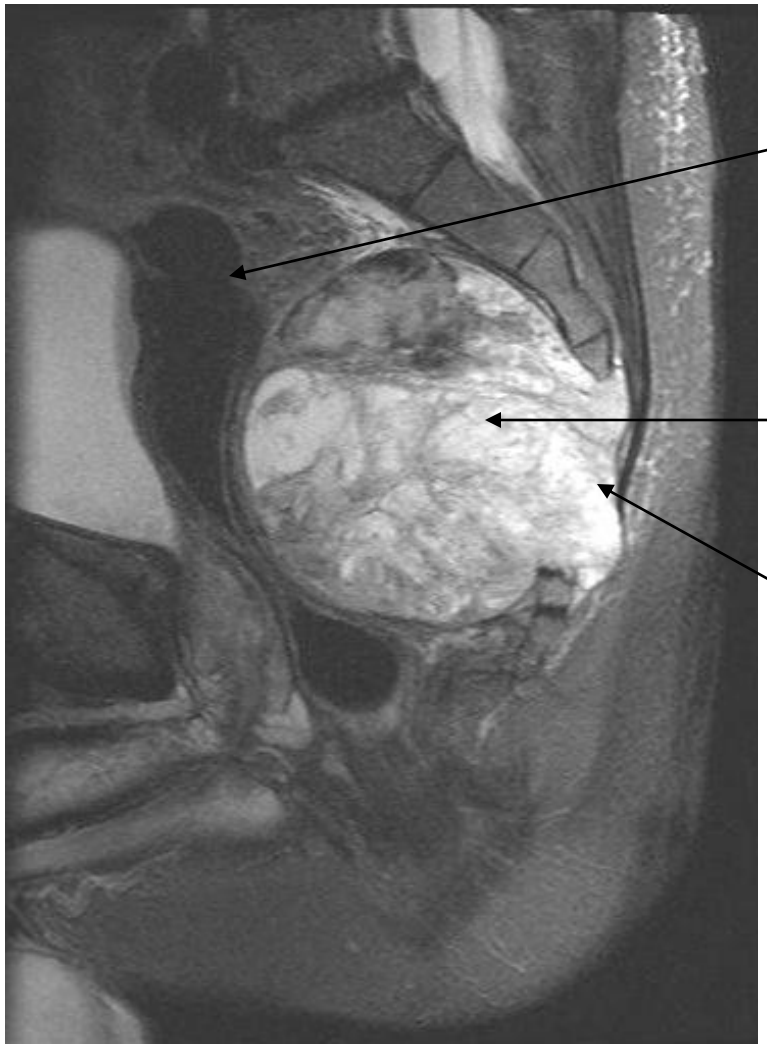
Axial fat saturated post gadolinium sequence showing enhancing right sided pelvic mass arising from the right S2 nerve root



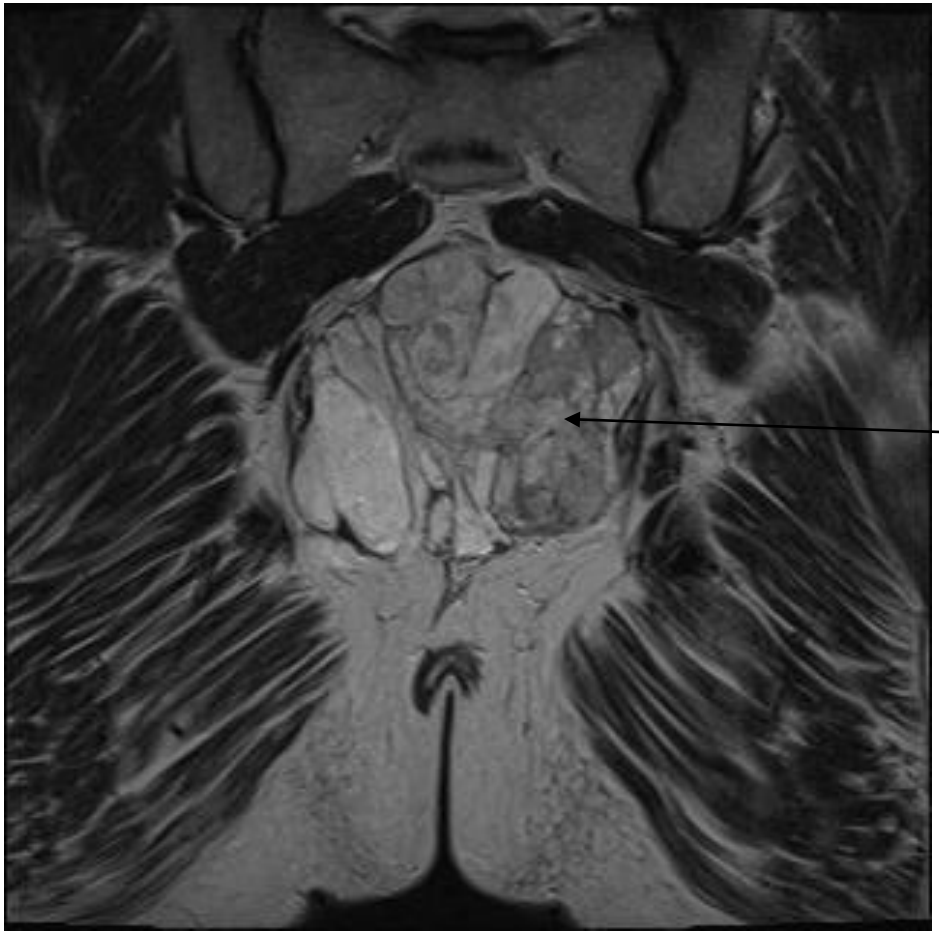
Sagittal T2W image showing a heterogenous retrorectal mass (solitary fibrous tumour) - well encapsulated, compresses the rectum and vagina anteriorly, and abuts but does not invade the coccyx and lower sacrum posteriorly.



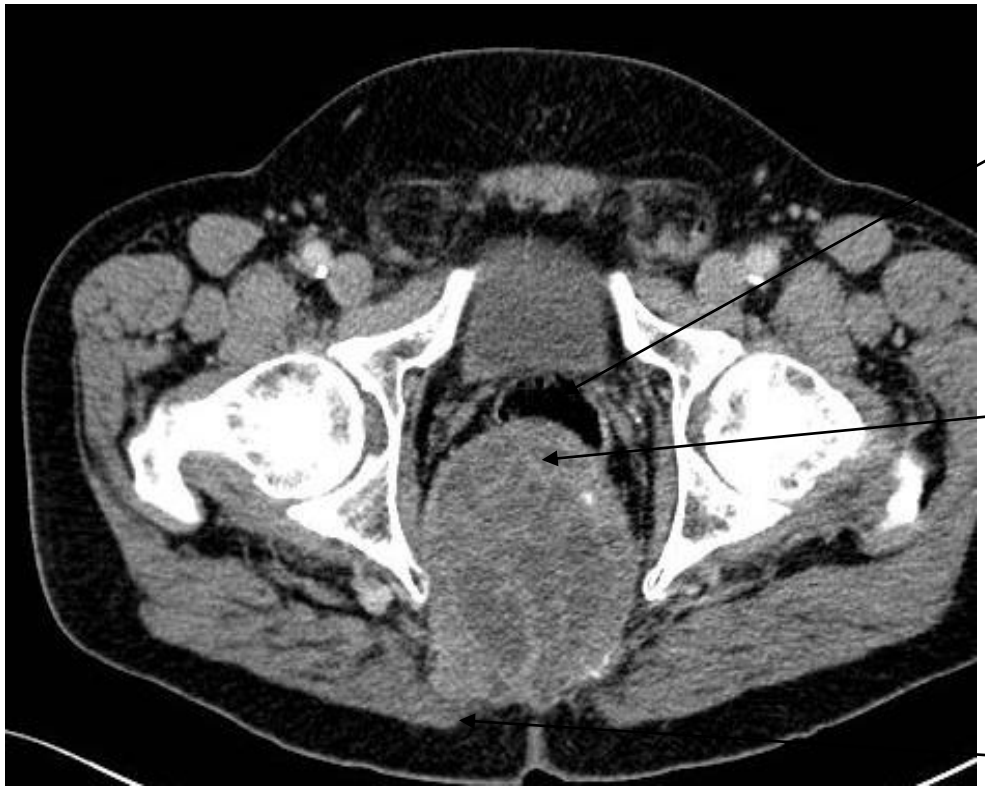
T2W sagittal image showing a retrorectal tumour (chordoma). Although the most cephalad tumour extent lies at the level of S1/2 (line



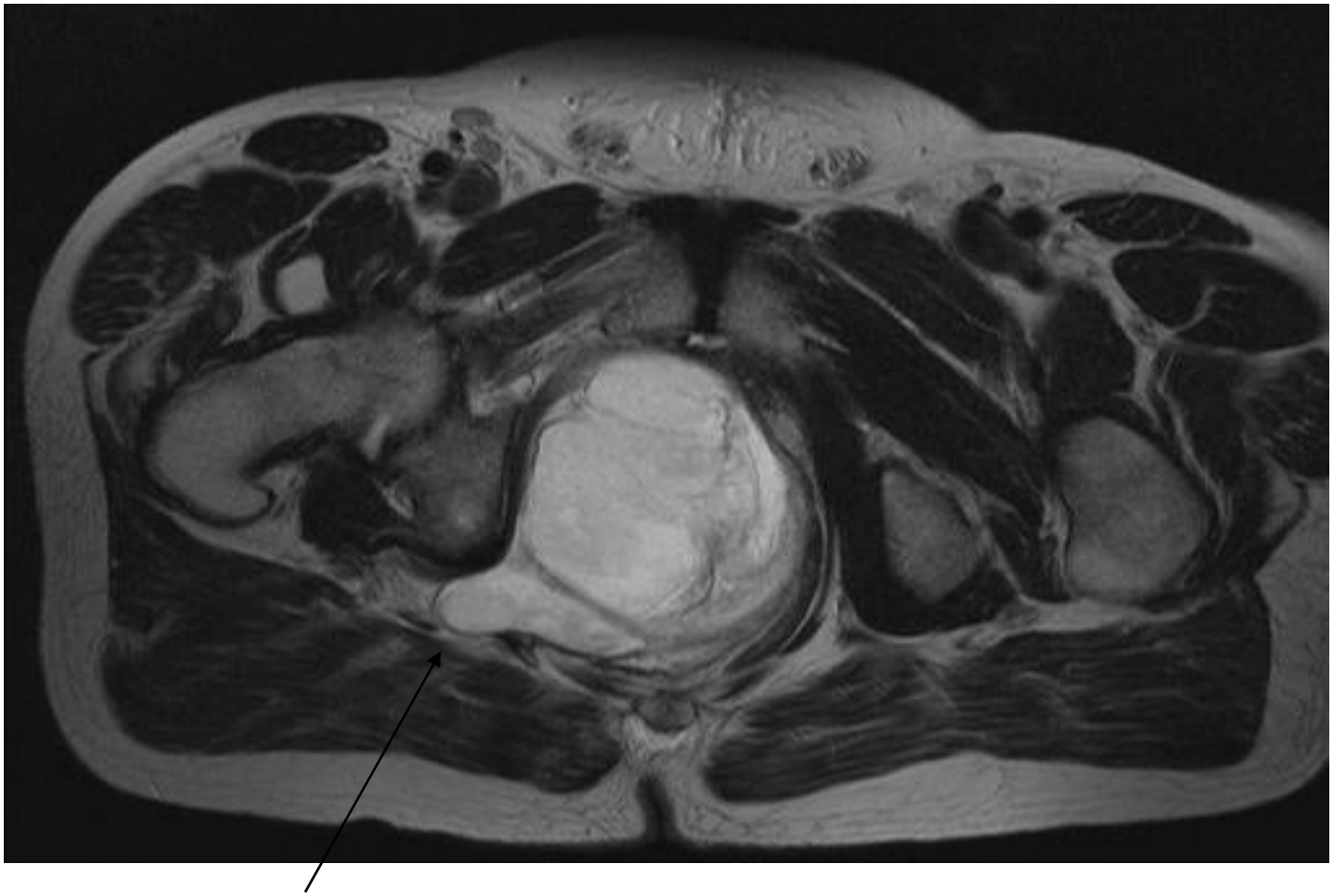
Sagittal image showing the typical heterogenous high signal intensity of a chordoma



Coronal T2W image of a multi-septated heterogenous high signal intensity retrorectal tumour (chordoma).

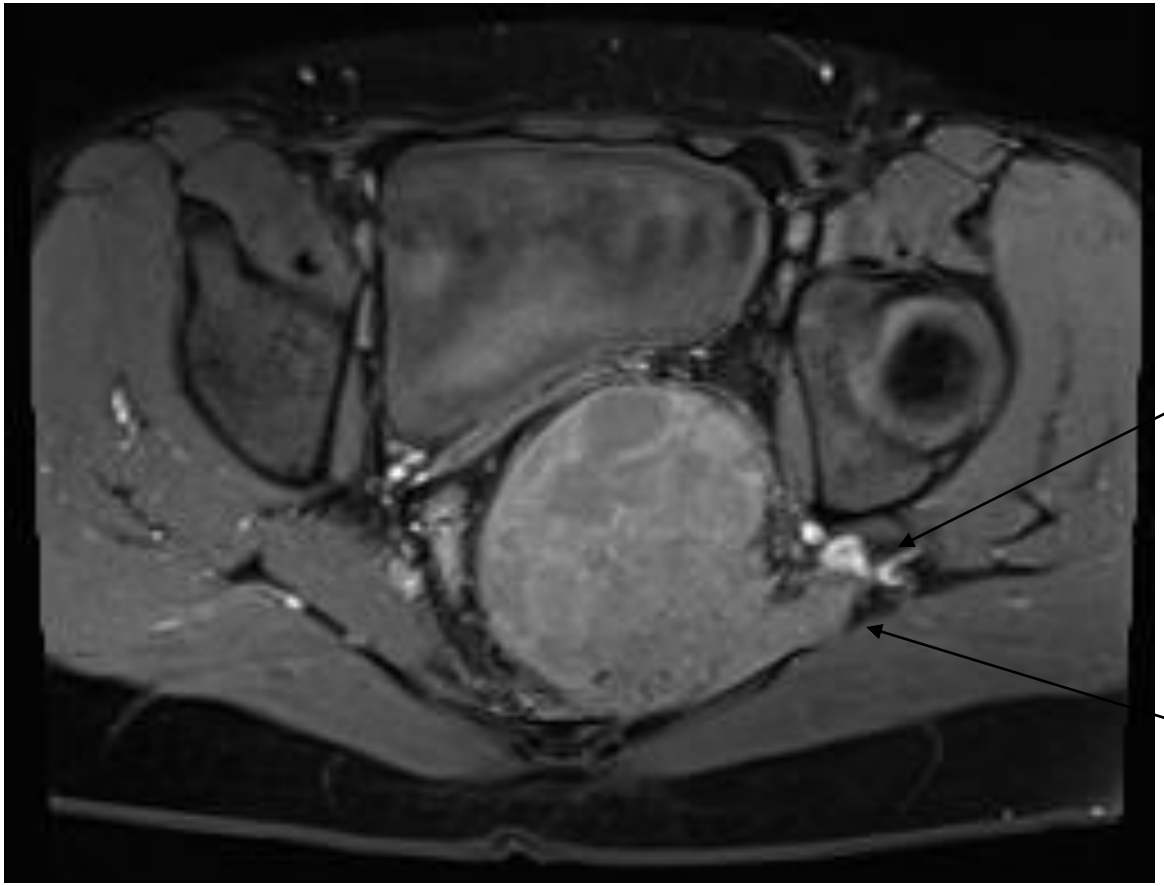


Axial CT image of chordoma showing anterior displacement of the rectum and posterior bulge into the medial aspect right gluteal muscle.



Extension into greater sciatic notch

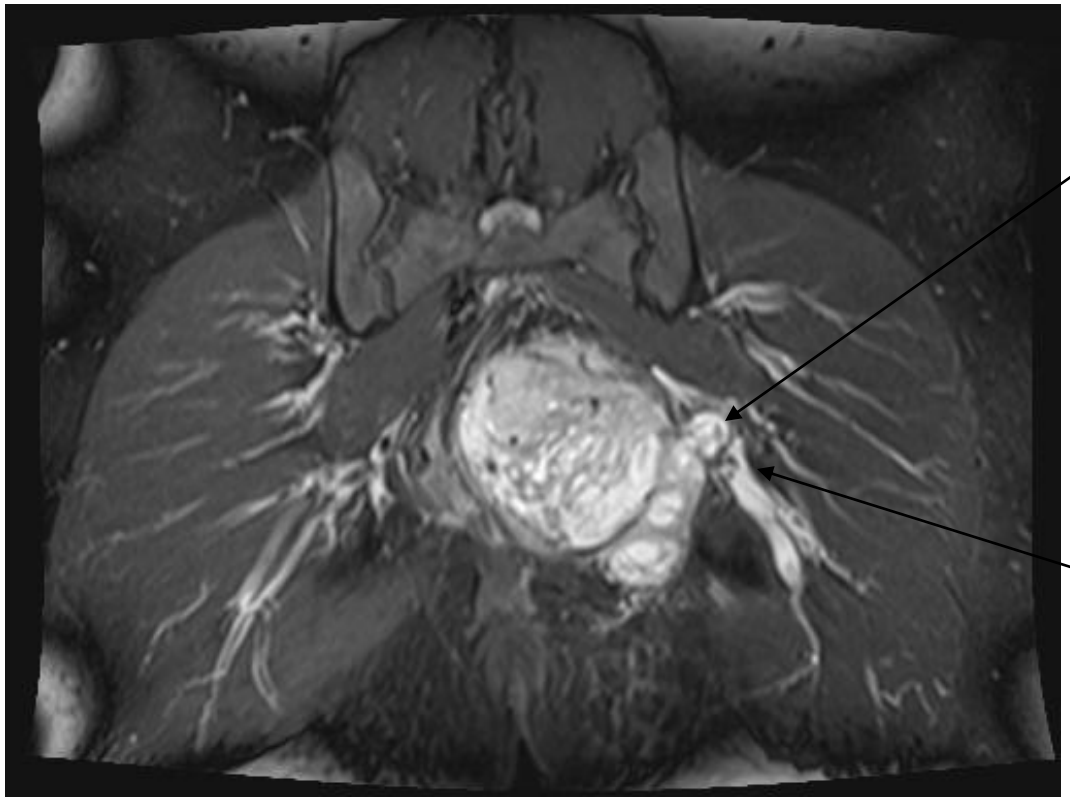
Axial T2W image showing a 2cm extension of a malignant retrorectal tumour (myxoid liposarcoma) into the right greater sciatic notch (arrow).



Important to identify close proximity of internal iliac vascular bundle on pre-operative imaging

Tumour exiting through left greater sciatic notch

Axial post gadolinium fat saturated T1W image demonstrating crucial additional information for the assessment of operability. Tumour extends into the left greater sciatic notch. Appears well encapsulated rather than frankly infiltrative so has potential to “shell out”.

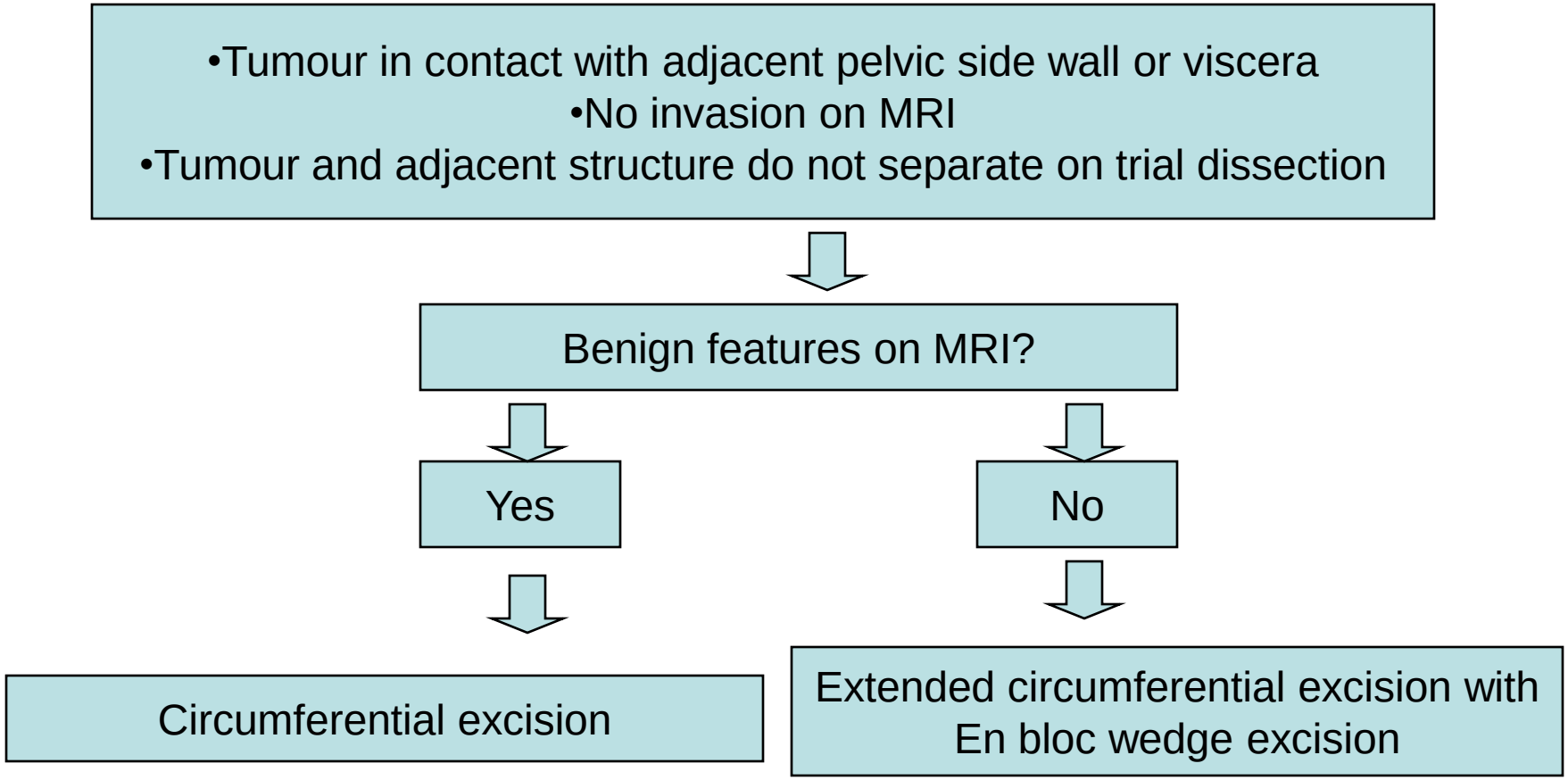


Tumour exiting through left greater sciatic notch

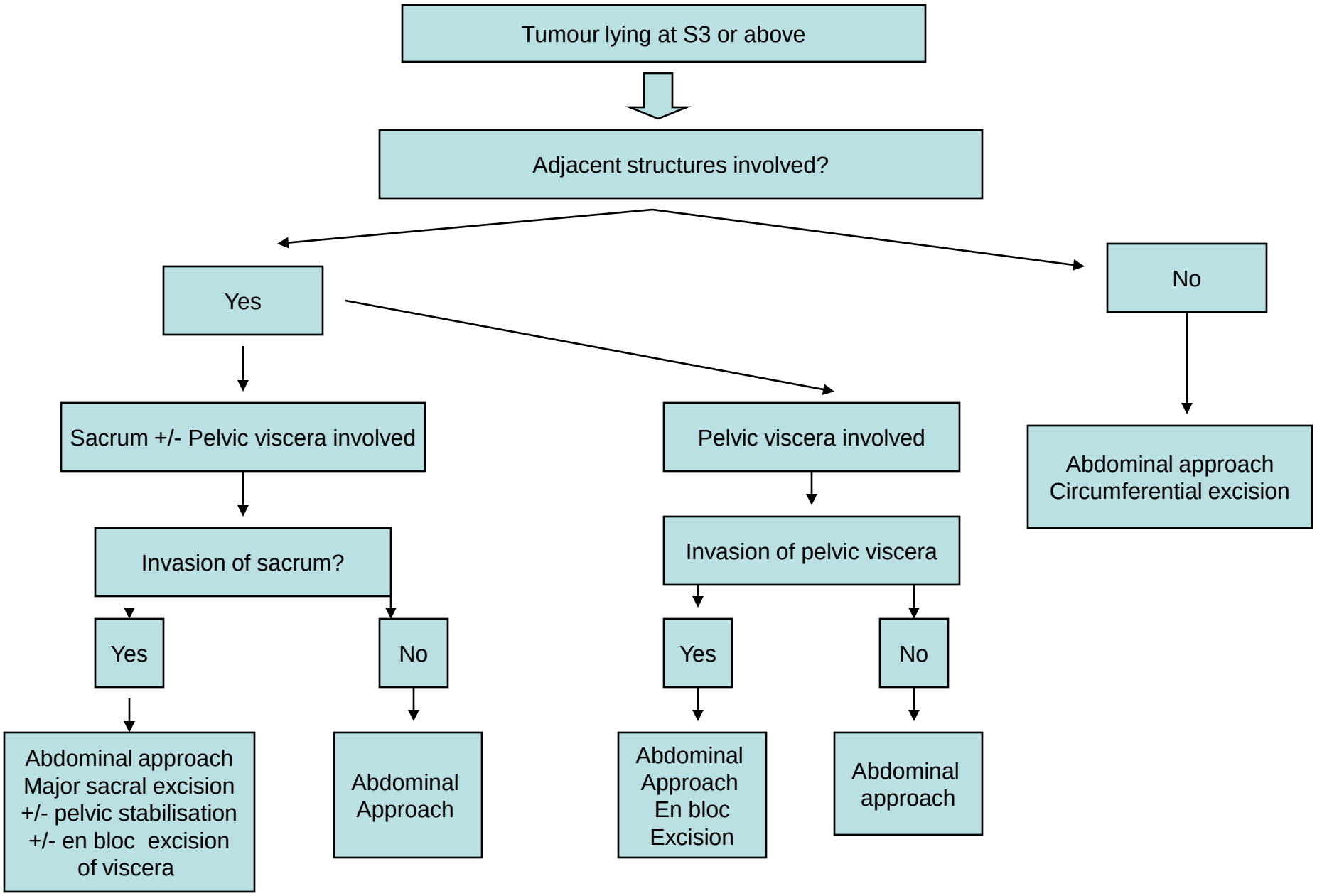
Close proximity of internal iliac vessels to mass

Coronal image showing retrorectal tumour extending into the left greater sciatic notch and demonstrating the potential intra-operative problem of the immediately adjacent vascular structures.

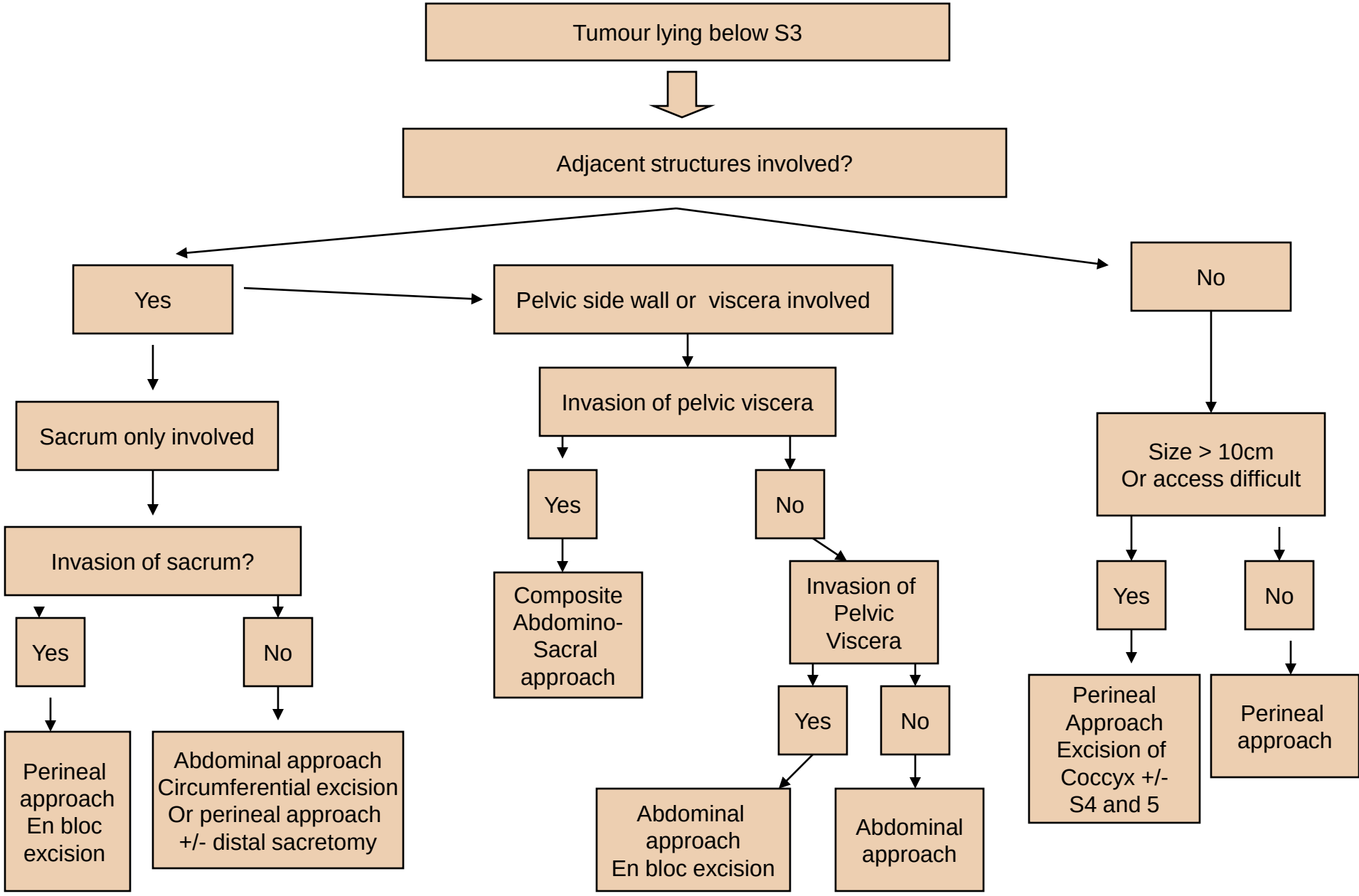
Algorithm for intra-operative decision-making for an adherent tumour with no radiological evidence of sacral invasion



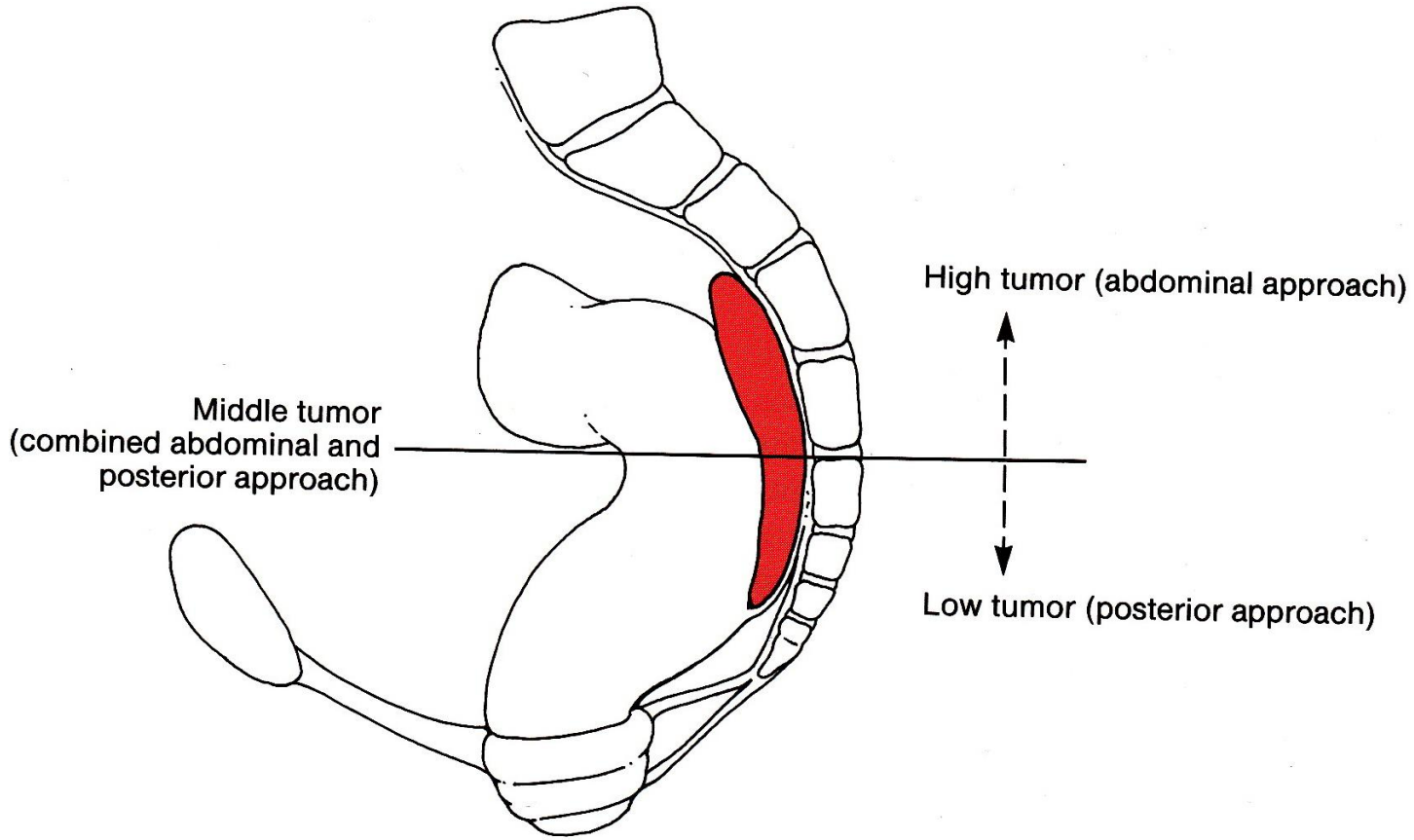
Algorithm for surgical decision making in retrorectal tumours lying at S3 or above



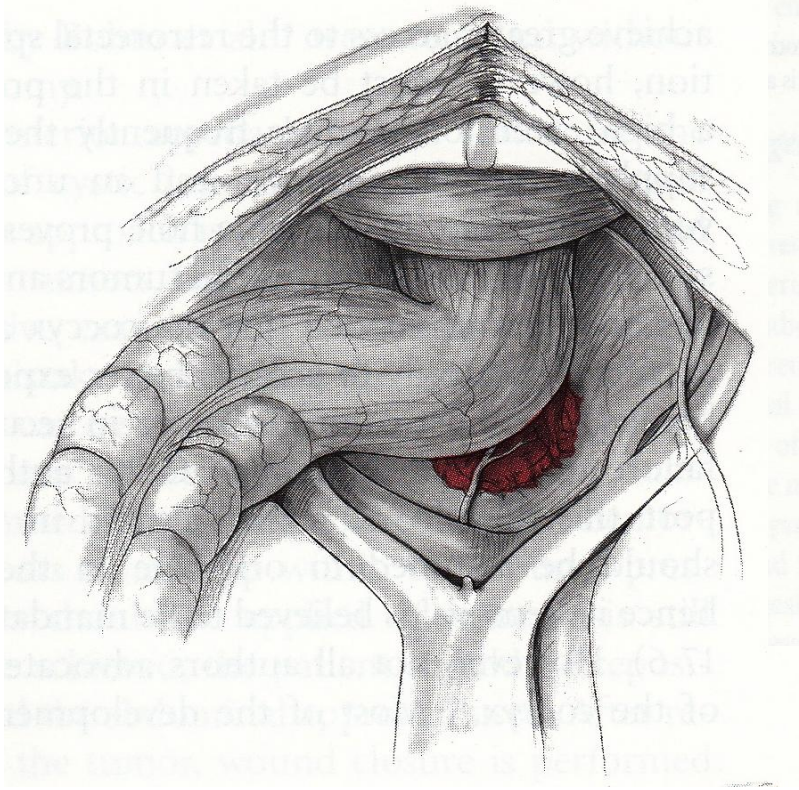
Algorithm for surgical decision making in retrorectal tumours below S3

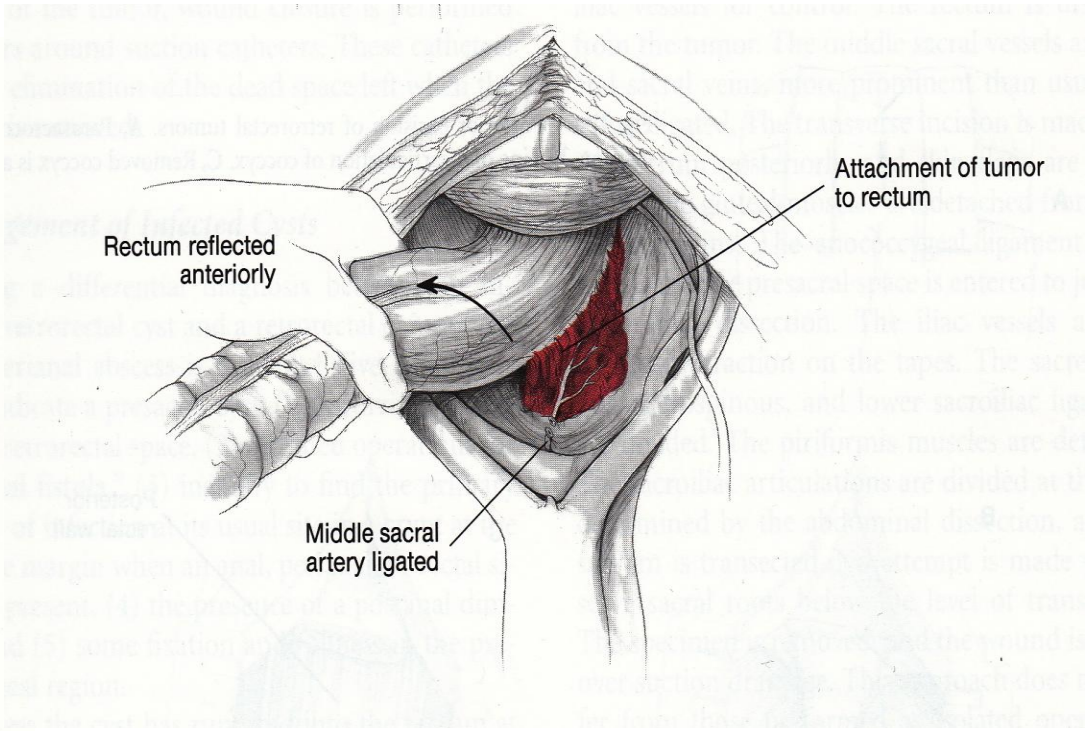


Level of tumour and choice of approach

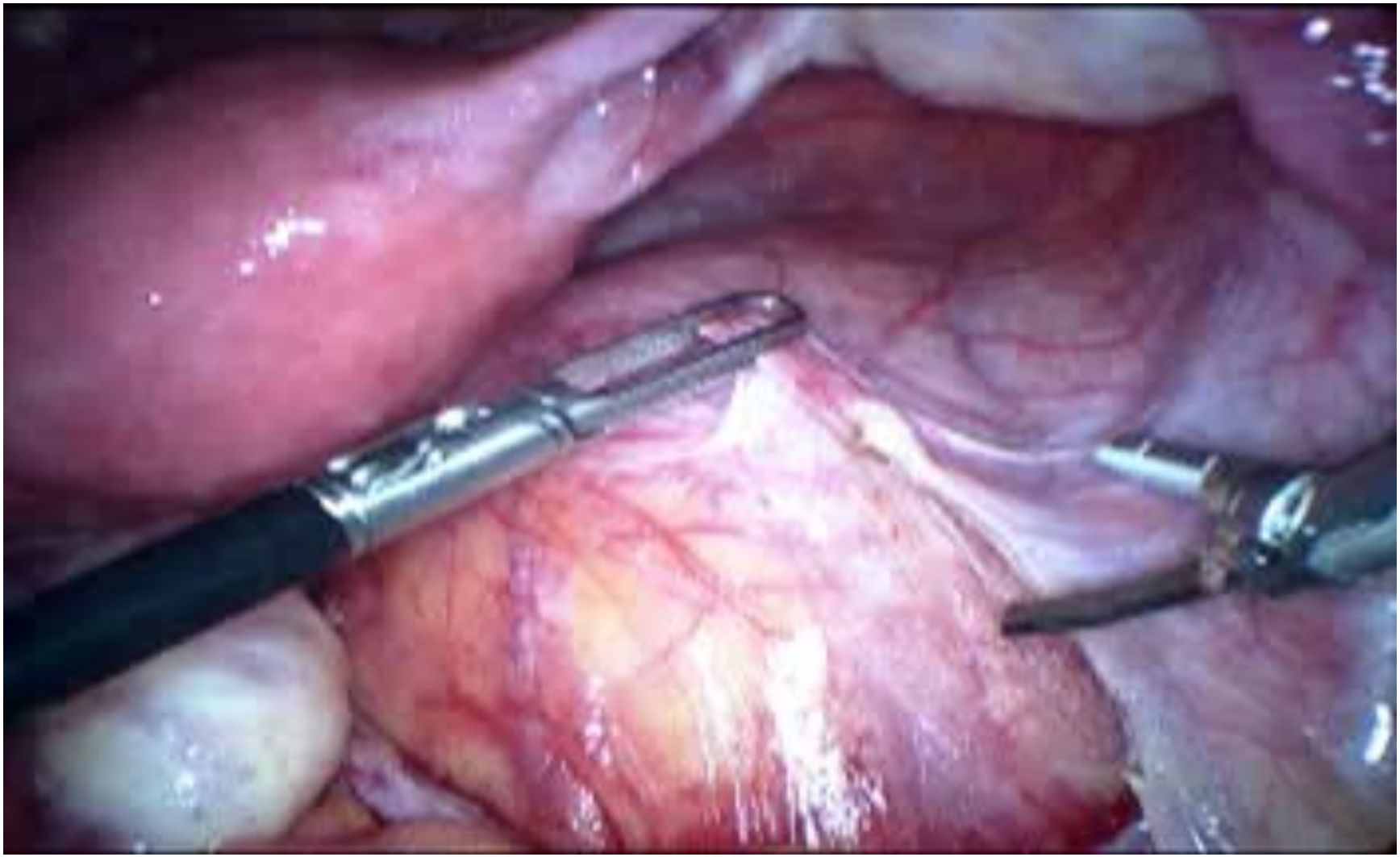


Abdominal approach

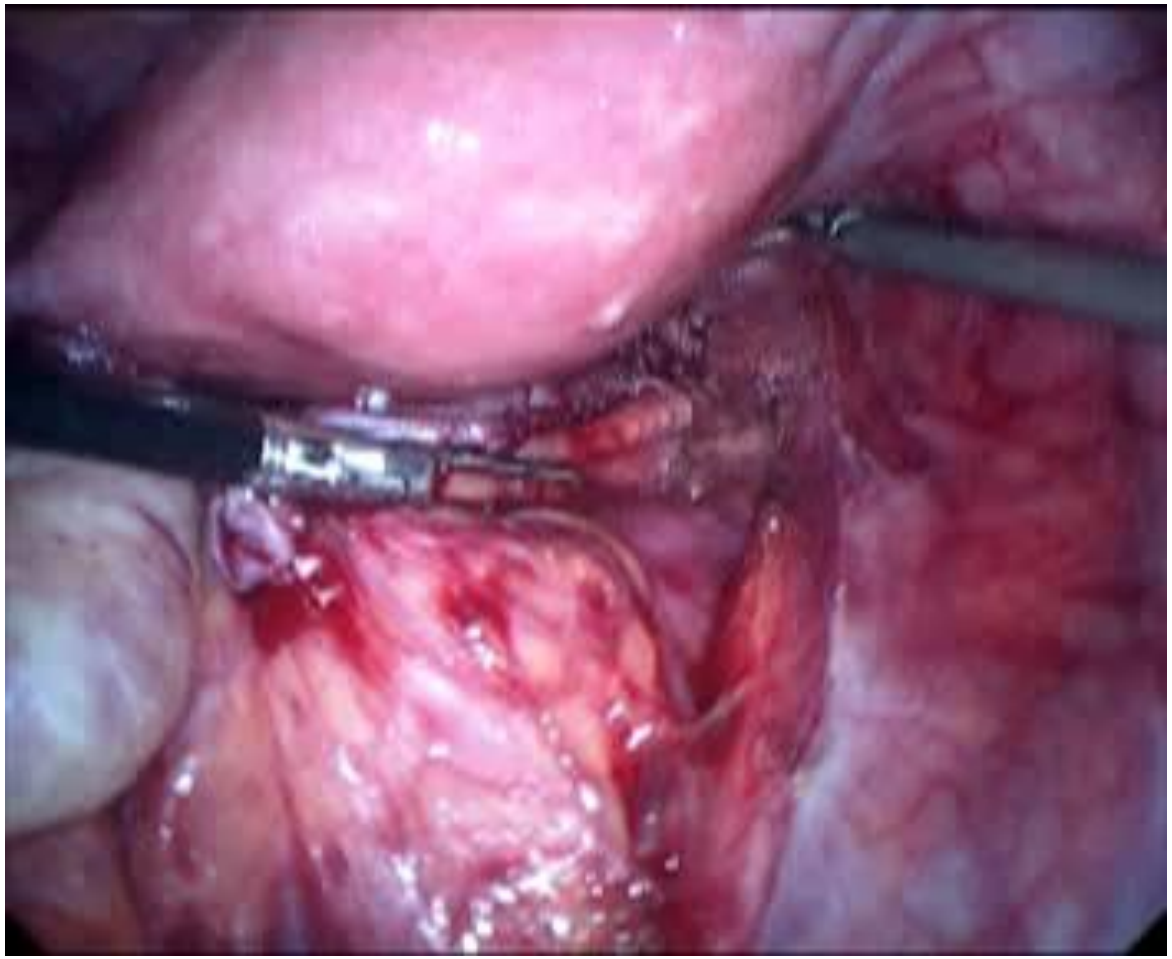




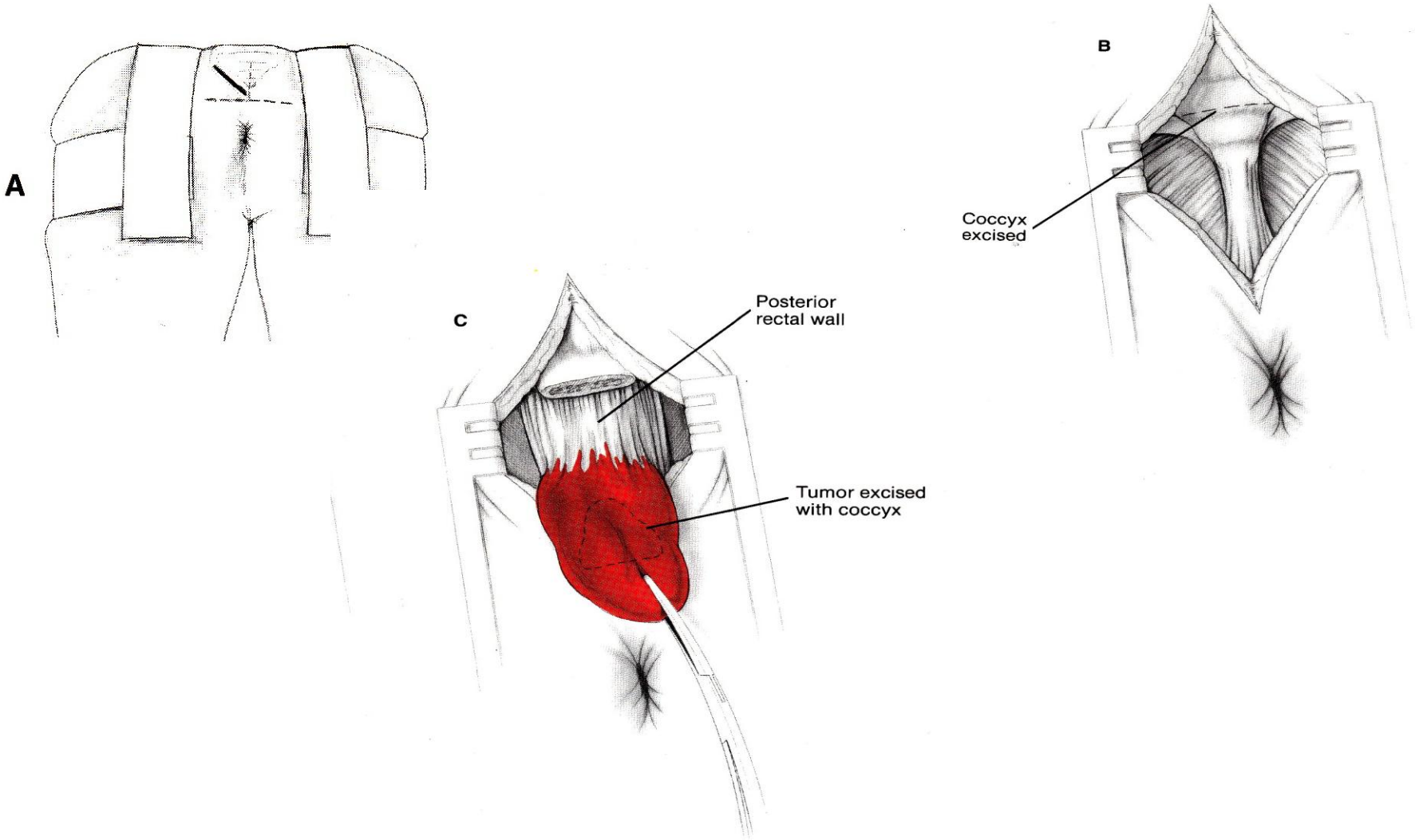
Retrorectal schwanoma

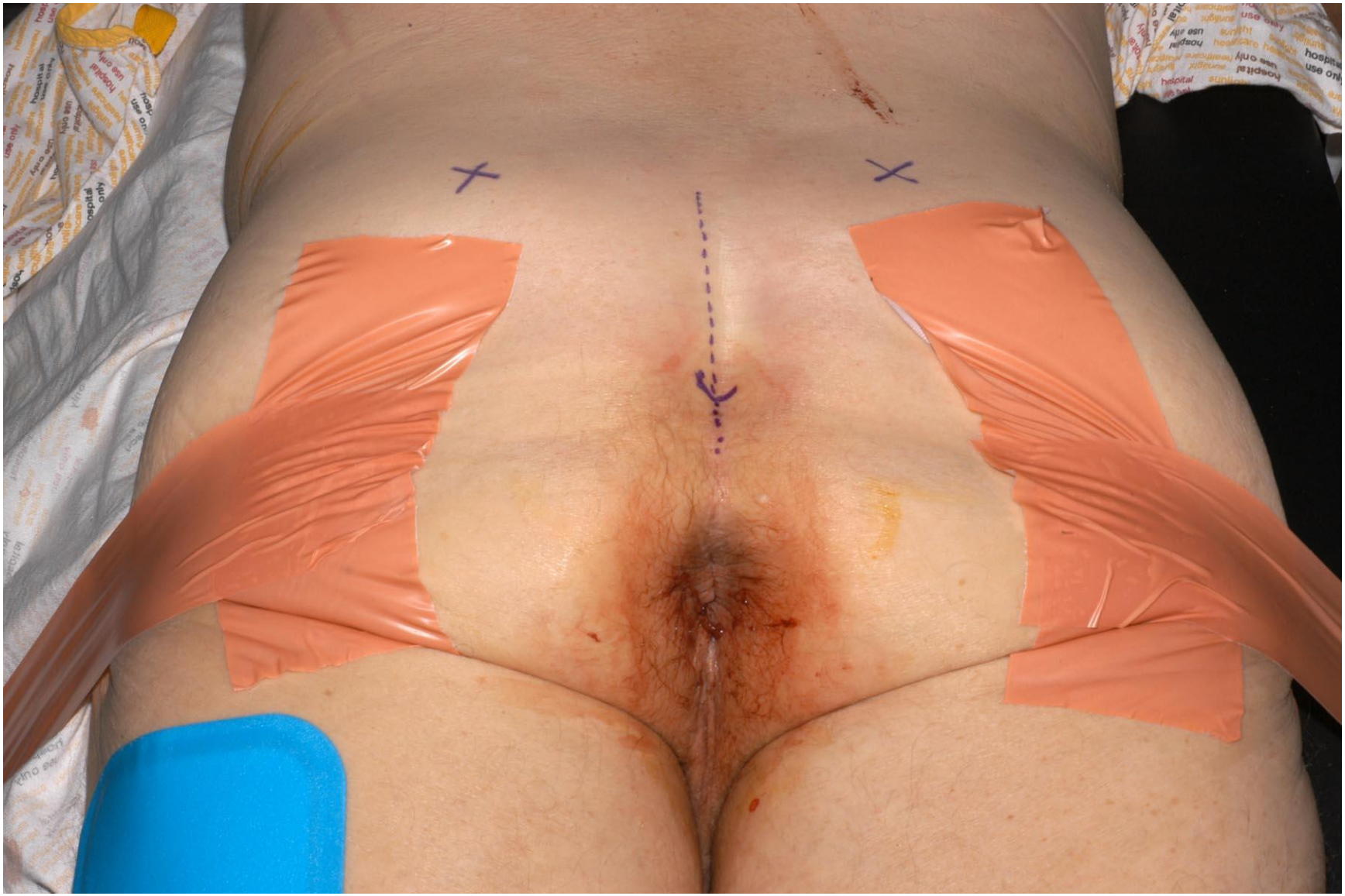


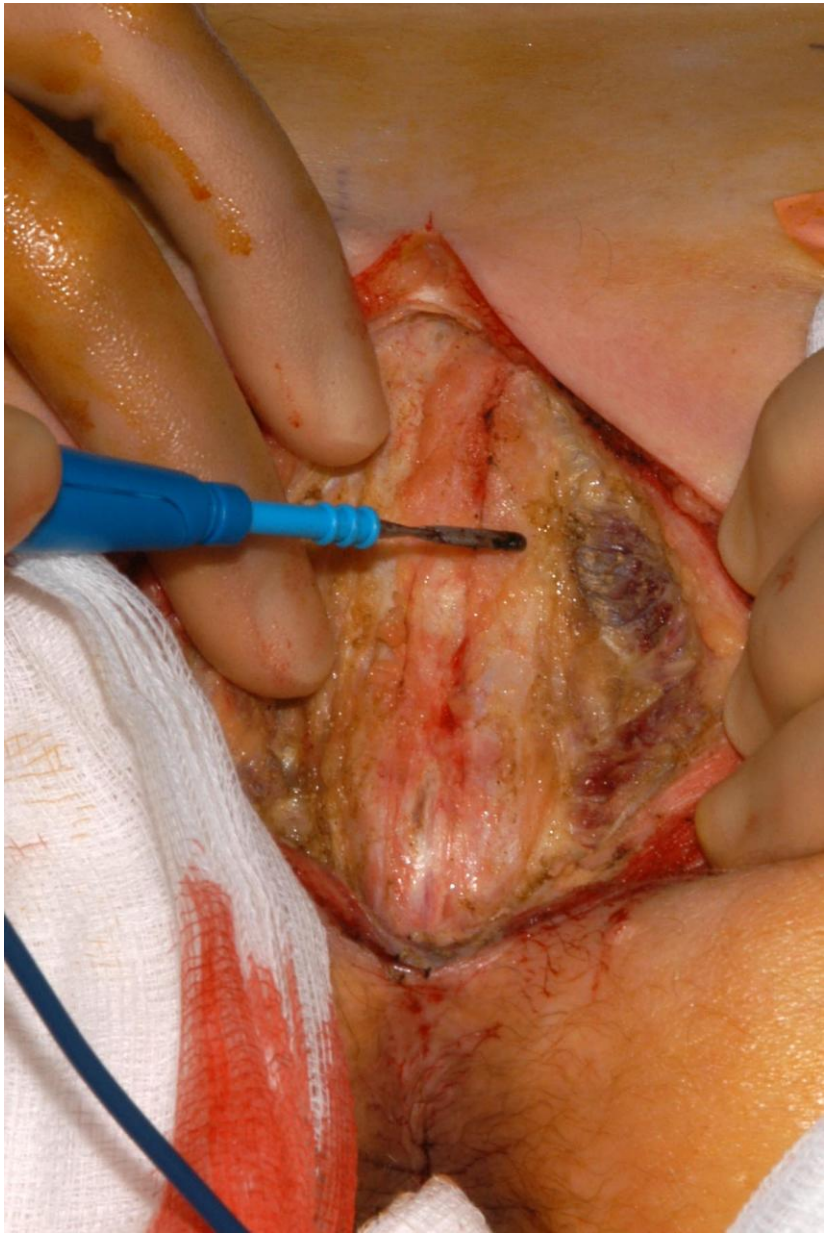
Preservation of adjacent structures

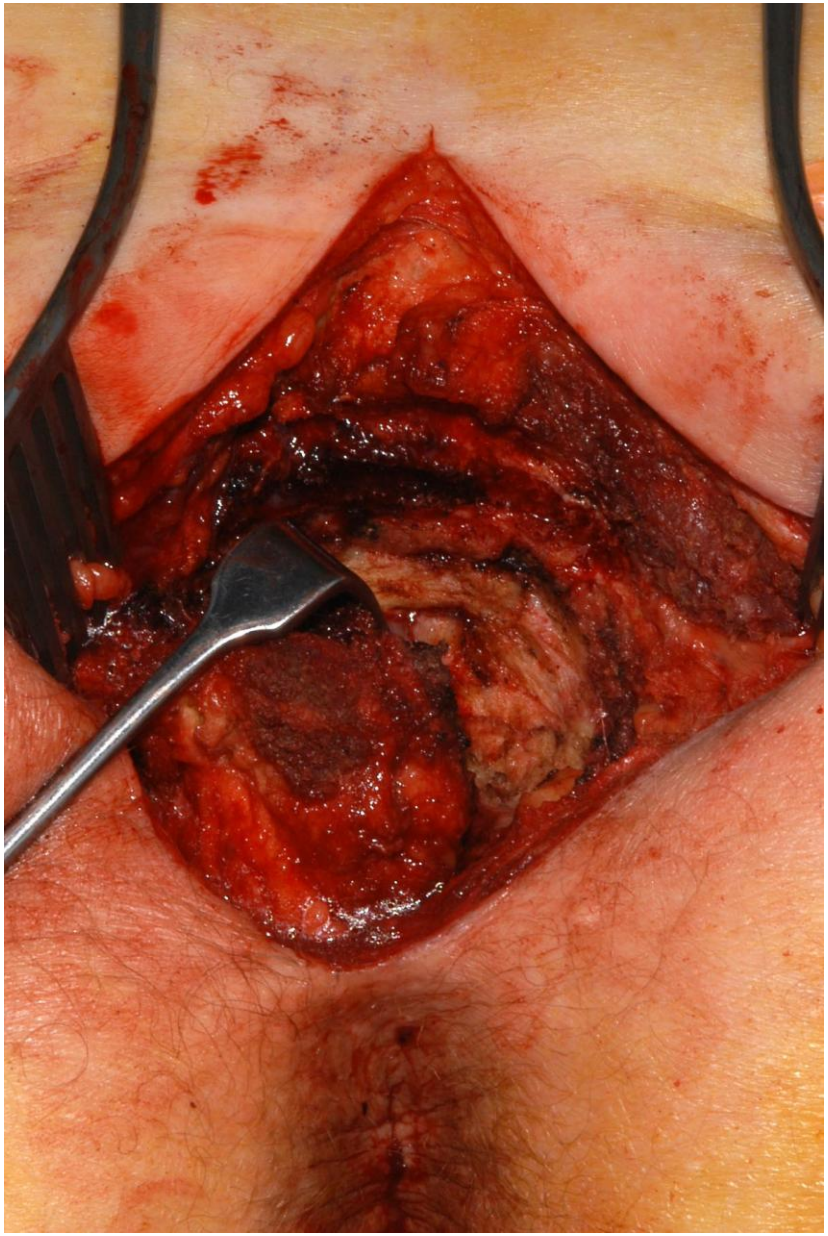


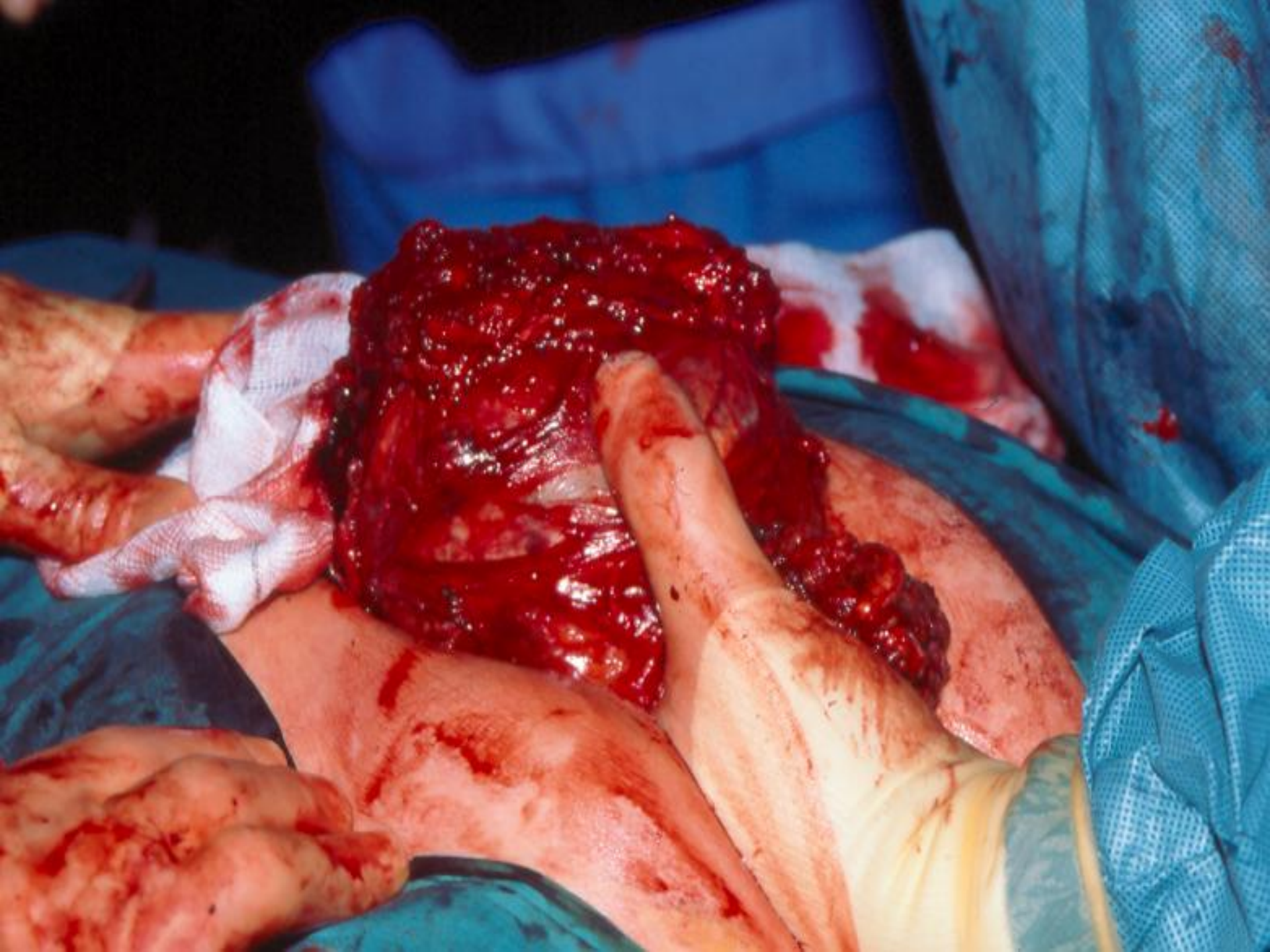
Posterior approach











Congenital cyst	23
Schwannoma	13
Chordoma	9
Ganglioneuroma	3
Liposarcoma	3
Gist	2
Solitary fibrous tumour	2
Angiomyxoma,Mucin secreting,Leomyoma,Leomyosarcoma, Rhabdomyosarcoma	5
Neuroendocrine , Neurofibroma,Dermoid with extramammary paget's,Myeolipoma	5
Others	10
Total	75

Surgical approach

Tumor	Abdominal	Perineal	Abdo-peri	Total
Congenital cyst	7	15	1	23
Schwannoma	13	-	-	13
Chordoma	-	3	6	9
Ganglioneuroma	3	-	-	3
Liposarcoma	1	2	-	3
GIST	1	1	-	2
Solitary fibrous tumour	2	--	-	2

Summary

- Rare tumors but be aware of them
- Avoid biopsy if possible
- Imaging is crucial

- Approach determined by level of tumor

Tumor	No of cases	Sex ratio F/M	Age grou
Congenital cyst	23	18/5	20-88
Schwannoma	13	8/5	27-78
Chordoma	9	3/6	45-77
Ganglioneuroma	3	3/0	21-55
Liposarcoma	3	2/1	44-59
Gist	2	1/1	54-57
Solitary fibrous tumour	2	2/0	38-69
Angiomyxoma,Mucin secreting,Leiomyoma,Leomyosarcoma, Rhabdomyosarcoma	5		
Neuroendocrine , Neurofibroma,Dermoid with extramammary paget's,Myeolipoma	5		
Others	10		
Total	75	F>M	