INTRODUCTION

Pelvic masses in children encompass a widely heterogenous group of lesions that may arise from the lower genitourinary tract, ovaries, pelvic muscles, pelvic nerves, vascular structures, lymph nodes, and adjacent pelvic bones. Diagnostic approach to such masses would be a challenging one; however the location of the mass in the pelvis along with the gender and age of the patient can help in narrowing the differential diagnosis.[1-3]

The location of the mass in the pelvis can be characterized as anterior, middle, or posterior based on the sagittal images. The anterior compartment is the space in front of a line drawn parallel to the posterior wall of the bladder. The posterior compartment is the portion behind a line drawn tangential to the posterior margin of the rectum. The middle compartment is positioned between these two lines.[2]

Anterior pelvic masses originate from the bladder and prostate and are nearly always rhabdomyosarcomas (RMS). Masses found in the posterior compartment (presacral masses) arise from variety of tissue origins located in this potential space and the commonest of them are teratomas, developmental cysts and pelvic neuroblastoma. Middle compartment masses are mostly of gynecological origin in female children, the commonest being uterine or vaginal rhabdomyosarcoma and Mullerian duct anomalies (MDA). Masses that may arise from any compartment include extension of bone tumors, vascular masses, neurogenic tumors and nodal masses.[1, 2, 4-6]

Purley cystic pelvic masses in children have a more limited differential diagnosis and could be differentiated according to the site of origin into those arising from lower urinary tract (bladder diverticulum,large prostatic utricle,urachal cyst),the ovaries(funcional cysts,mature teratoma,cystadenoma),the uterus(hydro/hematocmetrocolpos), and the retroperitoneum(lymphangioma,anterior sacral meningiocele and sacrococcygeal teratoma). Also the differential diagnosis may include developmental cysts as dermoid, epidermoid and rectal duplication cysts. [7, 8]

Pelvic lesions in pediatrics can present with different clinical signs and symptoms including lower abdominal and pelvic pain, distension or palpable mass felt by the parents. Also urinary symptoms as urinary tract infections, dysuria, haematuria or bowel problems as constipation or bleeding per rectum may be the presenting complaint. Masses of uterine origin maybe discovered during investigation for primary amenorrhea or abnormal vaginal bleeding.[6, 9, 10]

Clinical examination of children coming with complaint related to the pelvic region may reveal a palpable pelvic mass by abdominal or per rectal examination suggesting the location, size, and proximal extent of the lesion. Also signs of precocious puberty maybe evident suggesting hormone-secreting ovarian tumors.[6, 11, 12]

Diagnostic imaging modalities in assessment of pediatric pelvic masses:

The role of imaging is to confirm the presence of an underlying pelvic mass, determine the site of origin, delineate the extent for preoperative planning and in case of neoplastic lesions, provide information for staging, treatment planning, and serve as a baseline for monitoring response to therapy.[2]

Plain Radiography

Role of plain radiography of the abdomen and pelvis of pediatrics is very limited nowadays however it may show signs suggestive of a pelvic mass like displacement of the air-filled bowel loops or pelvic calcifications. Also bony destruction of the sacrum maybe seen in cases of chordomas and sarcomas. A characteristic X-ray sign ‘‘scimitar’’ sign is described as a rounded concave border of the sacrum without obvious bone destruction and is commonly seen with some types of presacral masses as anterior sacral meningocele and is one of the diagnostic triad for Currarino syndrome (triad of combined anorectal anomaly, sacral bony defect and presacral mass.[10, 13, 14]

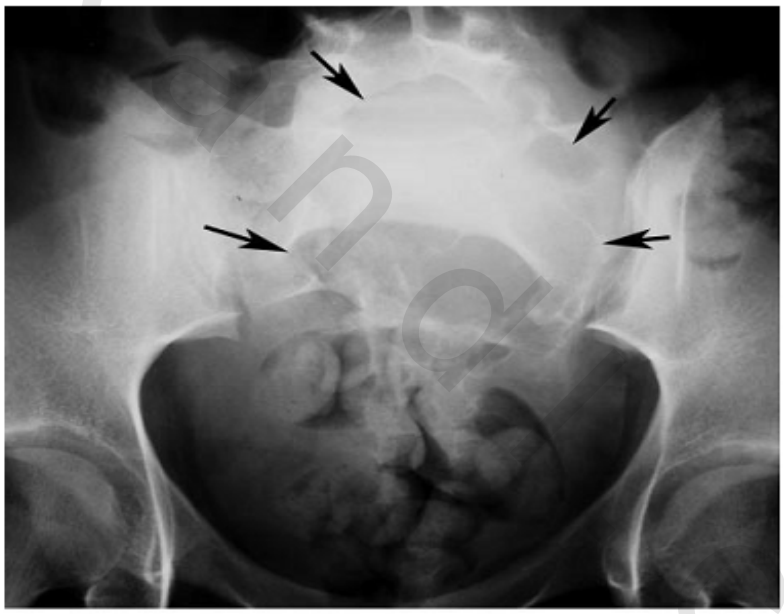


Figure (1): Sacral aneurysmal bone cyst in a 15-year-old boy with pain in the back and lower extremity.Anteroposterior pelvic radiograph demonstrates distortion of the sacrum (arrows).[5]

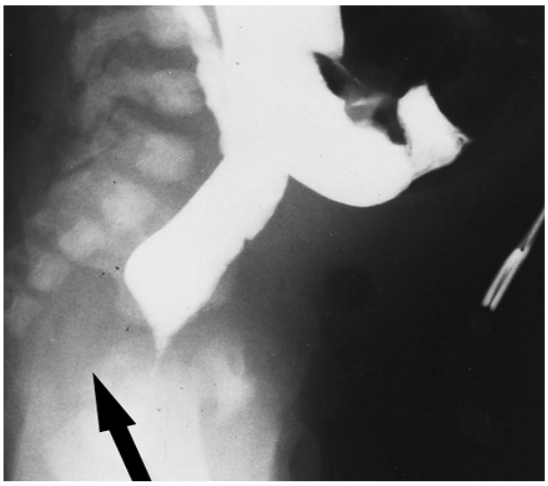


Figure (2): 7 year old girl with presacral deromid.lateral image from barium enema study shows the dermoid at the presacral level with anterior displacement of the rectum (arrow).[5]

Ultrasound(US)

Ultrasound is almost always the initial imaging modality to assess children with suspected or known pelvic mass because of its good resolution, its dynamic nature, lack of ionizing radiation and sedation risks and comparatively lower cost. It gives information about relative size and origin of the mass, differentiates between solid and cystic masses, and shows the fluid content, mass calcifications and the wall details of the mass.[10, 15, 16]

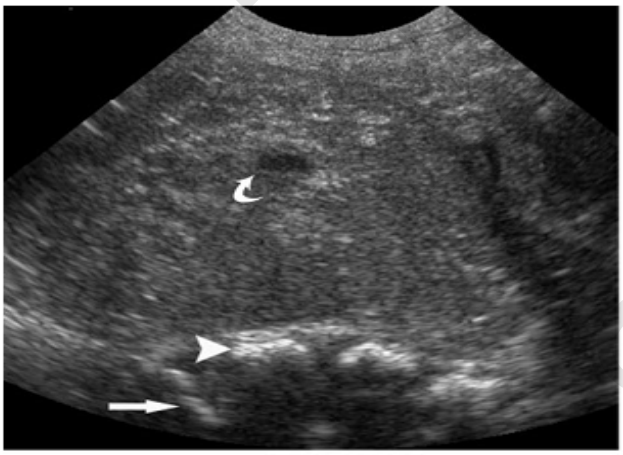


Figure (3): Neuroblastoma detected incidentally at US in a 9-week-old girl with urinary tract infection. Axial US image of the upper pelvis demonstrates a large solid mass that contains a small cystic area (curved arrow); multiple smaller areas of high echogenicity representing calcifications; the right iliac bone (straight arrow); and the sacral vertebrae (arrowhead).[5]

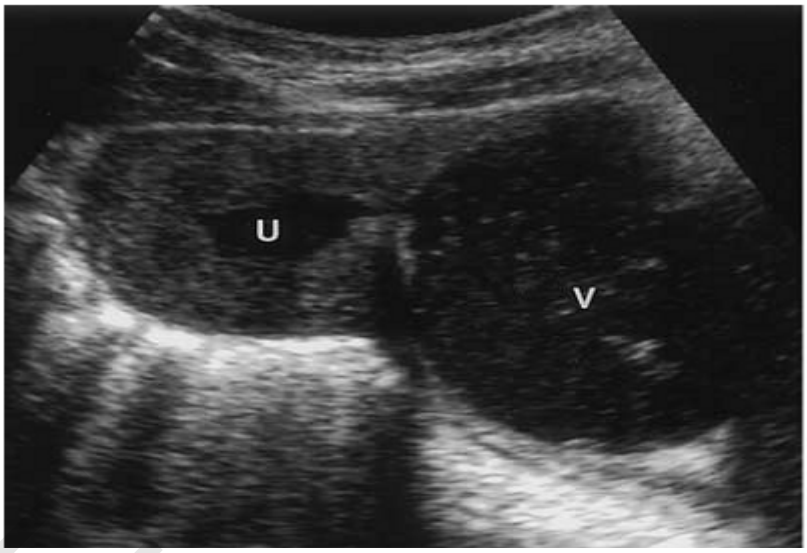


Figure (4): Hematometrocolpos due to an imperforate hymen in a 14-year-old girl with cyclic pelvic pain and primary amenorrhea. Longitudinal US scan shows a thin-walled, distended vagina (V) and the uterine cavity (U) with its thick myometrium.[17]

Multidetector Computed tomography (MDCT)

Both CT and MRI scans are now used in a complementary manner rather than exclusively in the evaluation of pediatric pelvic masses. A CT scan can be used to determine the origin of the lesion,whether it is solid or cystic, evaluate cortical bone destruction and assess involvement of adjacent viscera. CT is still commonly used for staging of pediatric pelvic neoplastic lesions both at first presentation and at follow up. Its main advantages are demonstration of tumoral calcifications,cortical bone changes and revealing pulmonary metastatis.[3, 13, 18, 19]

However, with CT, the use of intravenous iodine-based contrast material (with its potential risks) is required to increase the soft tissue contrast and assess the relationships between the tumor and adjacent vessels. Moreover, CT is associated with substantial radiation exposure to children. X-radiation and gamma-radiation (both ionizing radiation) are now considered known human carcinogens according to the most recent 11th Report on Carcinogens by the National Toxicology Program of the Department of Health and Human Resources. Children are especially susceptible to the carcinogenic effects of ionizing radiation that may lead to the development of second neoplasms in later life because they are inherently more radiosensitive than adults and because they have more remaining years of life during which a radiation-induced cancer could develop.[20-24]

Nevertheless, in the cases who still undergo CT examinations, pediatric radiologists should carefully apply the practice of ALARA (‘as low as reasonably achievable’) to reduce radiation exposure. The concept of ALARA addresses the role for the pediatric radiologist in reducing the amount of radiation which a child is exposed to while maintaining the efficiency and the reliability of the diagnostic modality. Justification is one of the general principles of ALARA in which the exam must be medically indicated. The

examination should be individually tailored with the area of coverage, slice thickness, pitch and milliampere level adjusted to answer the clinical questions. [21, 25, 26]

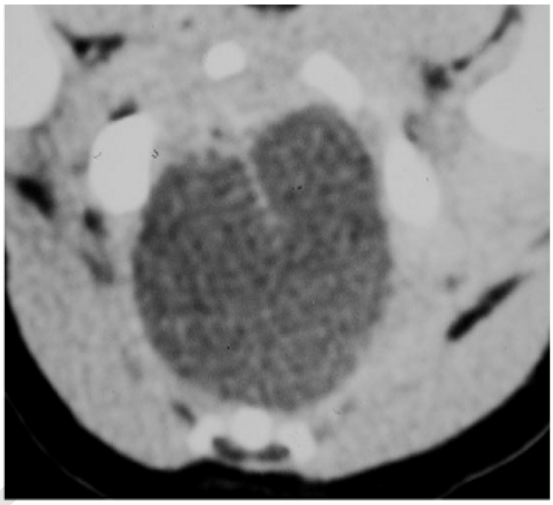


Figure (5): Mature (benign) sacrococcygeal teratoma in an 11-day-old girl. Axial unenhanced CT scan through the upper portion of the lesion shows attenuation similar to that of water, a finding indicative of a predominant cystic component.[5]

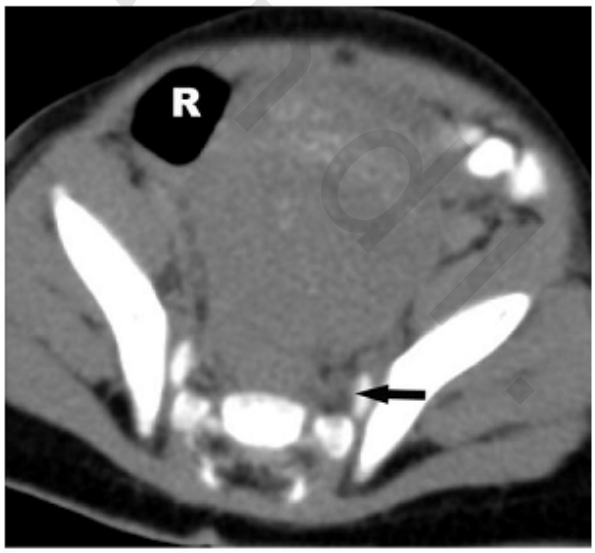


Figure (6): Neuroblastoma detected incidentally at US in a 9-week-old girl with UTI. Axial CT scan of the pelvis, obtained with IV contrast,demonstrates a well-defined presacral mass that contains scattered foci of calcifications. The mass has encroached on the neural foramen (arrow) in the left side and has displaced the rectosigmoid (R).[5]

Magnetic resonance imaging (MRI)

MRI is quickly becoming the modality of choice for assessment of pediatric abdominal and pelvic masses. Currently there is a shift away from CT to MR imaging both because of concerns about risk of radiation and because of additional information provided by MRI in many situations due to its better soft tissue resolution, dynamic imaging capability, and newer techniques such as diffusion-weighted imaging(DWI).[27, 28]

The superior soft tissue resolution aids in determining the planes of resection by suggesting the best surgical approach (e.g. abdominal,trans-sacral or combined approach) and the extent of resection in malignant infiltrating masses (e.g. the need for and level of sacrectomy). MRI is also more sensitive than CT in spinal imaging, showing associated cord anomalies such as a meningocele, nerve root, and foraminal encroachment by tumor, or thecal sac compression. MR imaging is also much superior to CT in assessment of extension of neoplastic lesions with accurate depiction of the pelvic side wall extensions, infiltrations into adjacent viscera and bone marrow changes.[3, 11, 19, 20, 28, 29]

Concerning staging of malignant masses, the tumor spread to the upper abdomen can be reliably evaluated by MRI during evaluation of the primary tumor.[19]

Classification of the common pediatric pelvic masses according to the anatomical location: [1, 2, 5, 7, 12, 30]

Anterior compartment:

Urinary bladder and prostatic masses:

Neoplastic

Malignant

- Rhabdomyosarcoma (the commonest)
- Others: TCC, Rhabdoid tumor

Benign

Papilloma, Hemangioma, Neurofibroma

Non-Neoplastic

- Pseudotumoral cystitis (inflammatory pseudotumor of the urinary bladder).
- Urinary bladder diverticulum, Urachal cyst/diverticulum, Large prostatic utricle, Large uretrocele

Middle compartment:

A) Uterus and vagina:

Neoplastic:

Malignant:

- Rhabdomyosarcoma (commonest)
- Others: Endodermal sinus tumor, Clear cell carcinoma(rare)

Benign

Leiomyoma

Non-Neoplastic:

Hydro/hematometrocolpos

B) Ovarian

Neoplastic:

1- Germ cell tumors

Teratoma (mature/immature/monodermal), Dysgerminoma, Endodermal sinus tumor (yolk sac tumor), embryonal carcinoma

2- Sex cord-stromal tumors

Granulosa cell tumor, Sertoli-Leydig cell tumor, Thecoma-fibroma tumors

3- Epithelial tumors

Serous/Mucinous cystadenoma, Serous/Mucinous cystadenocarcinoma

4- Ovarian lymphoma

5- Other primary tumors: e.g. Small cell carcinoma of the ovary

6- Ovarian secondaries

Non-Neoplastic

- Functional ovarian cyst
- Ovarian torsion
- Tubo-ovarian abscess

Posterior compartment (presacral masses):

Neoplastic:

Germ cell tumors (commonest are teratoma and yolk sac tumor), Chordoma, Neurogenic tumors (Neuroblastoma, neurofibroma, Ganglioneuroma, Schwannoma)

Non-Neoplastic:

Anterior sacral meningocele, Developmental cysts (dermoid, epidermoid, rectal duplication cyst), Cystic lymphangioma

Masses that may occur in any compartment:

A) Vascular masses:

- Vascular malformations (lymphangioma, capillary and venous malformations, arteriovenous fistula)
- Vascular neoplasms (hemangioma)

B) Neurogenic tumors

Neurofibroma, pheochromocytoma

C) Extension of tumors of the bony pelvis

Ewing sarcoma, Osteosarcoma, Giant cell tumor

D) Inflammatory masses

E) Nodal masses:

Lymphoma, nodal deposits

Radiological and clinical features of the most common pediatric pelvic masses:

Anterior compartment

Urinary bladder and prostatic masses

I. Neoplastic

Rhabdomyosarcoma(RMS) is the most common primary malignant neoplasm of the lower urinary tract in children. It is the most common soft tissue sarcoma of childhood representing about 5% to 10% of all malignant solid tumors ranking fourth in frequency after central nervous system neoplasms, neuroblastoma, and Wilms tumor. It can arise almost anywhere in the body, but the most frequent site of origin is the head and neck region, followed by the genitourinary tract, extremity, trunk, and retro peritoneum. [1, 2]

Genitourinary RMS includes tumors originating in the urinary bladder, prostate, testes and para-testicular sites, vagina, and uterus. The tumor has a bimodal age of distribution. The first peak occurs between 2 and 6 years of age and the second occurs between 14 and 18 years of age. Urinary bladder RMS shows evident male predominance of about 2.5:1 ratio.[2, 5, 31]

Two histological types account for the majority of pediatric RMS: the embryonal type, which includes botryoides and spindle cell variants; and the alveolar or adolescent type. Pleomorphic, anaplastic, and undifferentiated sarcomas are rarer types. The embryonal type accounts for 80% of all RMS and has the best prognosis.RMS is one of the small round blue cell tumors that typically are found in children, a group that also includes neuroblastoma, Ewing sarcoma, primitive neuroectodermal tumor, lymphoma, and desmoplastic small round cell tumor. These tumors share similar histological and radiological features but different cytogenetic and immune histochemical characteristics.[5, 31-33]

Children with RMS of the urinary bladder or prostate come to clinical attention because of signs of urinary tract infection, haematuria, dysuria, urinary frequency, or palpable suprapubic mass. Primary tumors of the prostate can almost always be palpated on rectal examination and can produce large pelvic masses resulting in urinary frequency or constipation if significant compression of the bladder or bowel occurs. Late cases may present with a large pelvi-abdominal mass associated with hydronephrosis and renal deterioration.[1, 2, 6, 31]

Bladder lesions are initially characterized by US and voiding cystourethrogram (VCUG). RMS, either arising in or invading the bladder base, usually appears as a polypoidal intraluminal mass resembling a cluster of grapes. The appearance on MR imaging, CT, and US is highly variable, as one can see hemorrhagic, cystic, and solid components.RMS of the prostate or bladder base is sometimes seen as bulky pelvic mass the precise origin of which can be difficult to determine, because primary prostatic lesions may invade the bladder base; conversely, the prostate may be invaded by a tumor arising in the bladder. [2, 3, 31]

Recent evidence supports the usefulness of MRI for evaluating pelvic RMS being superior to CT in better delineating tumor extensions within the bladder lumen and into surrounding tissues. The tumor has a nonspecific low to intermediate signal intensity on T1-weighted images and intermediate to high signal intensity on T2-weighted images. If the lesion is hemorrhagic or hyper vascular, the intensity on T1-weighted images may be close to that of fat. T2W images provide excellent contrast between the fluid-filled bladder lumen, hypointense bladder wall, prostate, and surrounding soft tissues. The tumor enhances after the IV contrast administration. Post-contrast dynamic images, acquired before excreted contrast reaches the bladder, are very useful in assessment of heterogeneous tumor enhancement. MRI sagittal images may distinguish prostatic rhabdomyosarcoma that extends into the space of Retzius from tumor invading the bladder. MRI is better than CT in detecting residual disease which leads to appropriate earlier management.[1, 2, 9, 34]

RMS staging is based on primary tumor extension, involvement of lymph nodes, and metastatic disease. Soft tissue extension is readily evaluated in MRI by checking for interruption of the normal low signal within the walls of organs such as the rectum and bladder and by invasion of the peri-vesical or peri-urethral tissues or the ischiorectal fossa

.Both soft tissue extensions and nodal spread are demonstrated by fat-saturated T2 or gadolinium enhanced fat-saturated T1 images.MR imaging suffers from the same limitations as do other cross-sectional imaging modalities in lymph node evaluation so interpretation is limited to the size of the node when assessing for potential underlying involvement. DWI may also assist in the detection of metastatic lymph nodes.[1-3]

Metastatic disease is most commonly found in the lungs, lymph nodes, and cortical bone and is less frequently seen in the bone marrow, liver, and brain. Between 10% and 20% of all patients with RMS have metastases at the time of diagnosis. CT is particularly useful to assess for lung metastasis; however, one must always be aware of the associated radiation exposure.[2, 3]

Other bladder soft tissue masses are relatively uncommon in children. Benign lesions include papilloma, hemangioma, fibroma, paraganglioma, neurofibroma and leiomyoma. Neurofibroma of the bladder is rarely found in isolation, but may occur in patients with NF type 1. Malignant lesions include rhaboid tumor and TCC. Secondary involvement of the bladder has been reported in patients with lymphoma and leukemia. Uncommonly, mass-like inflammatory changes of the bladder wall, known as pseudotumoral cystitis or bladder pseudotumor, may mimic a polypoidal bladder neoplasm.[1-3, 35-37]

Most of these neoplasms are nonspecific in appearance and cannot be distinguished from one another with imaging. The MR features of benign and malignant bladder tumors are similar and include focal or diffuse wall thickening, a sessile or polypoidal mass arising within the bladder wall projecting into the bladder lumen. If the tumor shows intense contrast enhancement, hemangioma and pheochromocytoma should be considered. Tumor distribution along the neurovascular bundles is a finding of neurofibromatosis. One helpful general sign of malignancy is invasion of the adjacent soft tissues. Definitve diagnosis is usually only obtained by patholgoical analysis of cystoscopic biopsy.[2, 3]

Treatment of RMS includes surgical excision and neoadjuvant chemotherapy with the goal of preservation of the bladder function. Careful staging and follow-up imaging are important for prognosis and treatment. The enhanced visualization and delineation of pelvic RMS by MRI lead to its use as the main and sometimes only abdomino-pelvic radiological study for staging and following this disease.[1, 9, 38]

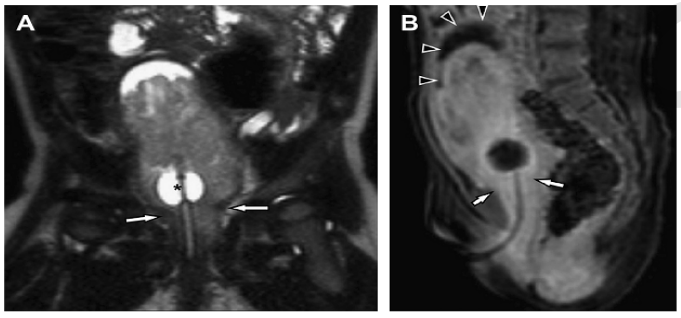


Figure (7): A 2-year-old girl with bladder RMS, embryonal subtype. (A) Coronal T2WI reveals a large, heterogeneous soft-tissue mass at the bladder base, extending into the bladder neck and proximal urethra (arrows). Foley catheter is present (asterisk). (B) Sagittal post contrast fat-saturated T1WI demonstrates diffuse enhancement (arrows). The bladder wall is seen superiorly (arrowheads). [1]

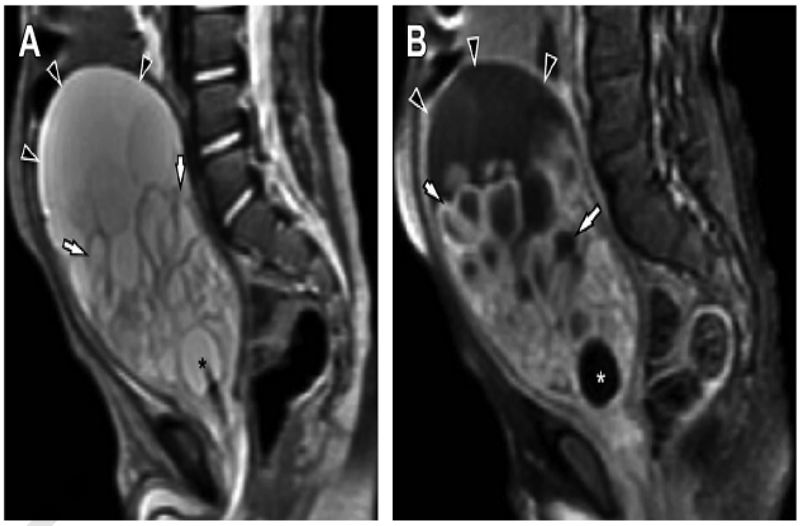


Figure (8): A 27-month-old boy with botryoides RMS of the bladder. (A) Sagittal fat-saturated T2WI shows multiple hyperintense cystic intraluminal masses (arrows) extending from the bladder base. The bladder wall is displaced superiorly (arrowheads). A Foley catheter is present (asterisk). (B) Sagittal post contrast fat-saturated T1WI. Peripheral enhancement of the mass is clearly depicted.(1)

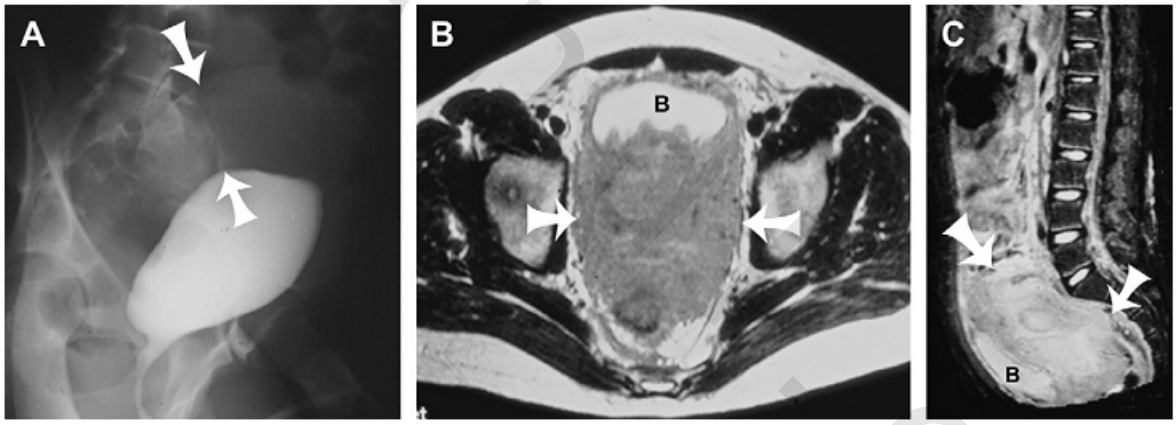


Figure (9): Burkitt's lymphoma of the bladder in a 15-year-old male with frequency. (A) Oblique image from a fluoroscopic VCUG reveals a thickened bladder wall (arrows). (B) Axial contrast-enhanced T1W MR of the pelvis demonstrates a large mass (arrows) along the posterior bladder (B) wall. (C)Sagittal T2W MR confirms a mass (arrows) along the posterior bladder wall. (3)

II. Non-Neoplastic

Non neoplastic pelvic masses related to the urinary bladder and prostate are almost always cystic and include urinary bladder diverticulum, urachal cyst/diverticulum, large uretrocele and prostatic utricle. Ureteroceles represent cystic dilatation of the intravesical segment of the ureter. They may be associated with either a single or a duplex ureter. Uretrocele may be as small as 1 cm, or it may fill the entire bladder and prolapse through

the urethra. Bladder diverticulum in children is a congenital abnormality (termed Hutch diverticulum) and commonly occurs just above the ureterovesical junction. It is associated with ipsilateral reflux causing repeated urinary tract infections. Prostatic utricle is a normal anatomical variant being a small diverticulum from the prostatic urethra between the openings of the ejaculatory ducts. Sometimes it becomes enlarged forming a cystic mass especially in cases with other anomalies as male hypospadias, cryptorchidism and Prune Belly syndrome.[7, 39]

Middle compartment

A) Masses of the uterus and vagina

Ultrasound is the primary screening modality for the evaluation of known or suspected pediatric gynecologic masses. MRI is used progressively more in this field, particularly for the evaluation of complex pelvic masses and congenital anomalies because it is much superior to CT in providing unparalleled spatial and soft tissue resolution with anatomic results helpful for surgical planning.[10, 16]

***I*-Neoplastic:**

Uterine tumors are uncommon in pediatric patients, but when encountered in this age group they are likely to be malignant and the commonest is RMS that may affect uterine body, cervix or vagina. Vaginal RMS is usually found in infants and children younger than 2 years of age while uterine RMS is rare, with peak incidence in the second decade of life and is quite extensive at time of diagnosis. [1, 16, 31]

The clinical presentation of genital RMS in females is presence of a vaginal polyp or cervical polyp, maybe with or without bleeding. Vaginal bleeding may be in the form of 'spotting' in the premenarchal children or extra-menstrual spotting or bleeding for 'several' weeks in the postmenarchal females. At US, a vaginal RMS appears as a large, solid, and heterogeneous or hypoechoic mass posterior to the bladder.[3, 17, 31, 40]

On MR imaging these tumors demonstrate intermediate to high signal intensity on T2-weighted sequences and low to intermediate signal intensity on T1-weighted sequences that is higher than the muscle but lower than the fat. Contrast enhancement is variable, depending on degree of tumor necrosis. Degree and change in tumor enhancement after chemotherapy are also useful in assessing response to treatment. Tumor heterogeneity secondary to necrosis and calcification is common .Vaginal tumors may invade the bladder base or obstruct the ureters, leading to hydronephrosis. The presence of fat planes between the neoplasm and adjacent structures excludes gross invasion, but the absence of fat planes may be normal or caused by adherence or invasion. When the loss of fat planes is associated with an eccentric soft tissue mass, a confident diagnosis of extra-vaginal extension can be made..[1, 3, 16]

CT and MR are used to assess for local invasion and distant metastases including evaluation for retroperitoneal lymphadenopathy, liver, lung, and bone metastases. Lymphadenopathy has a spectrum of appearances ranging from small discrete nodules to large conglomerate masses.[1, 3]

Other less common malignancies encountered in the lower genital tract in children include endodermal sinus tumors and clear cell adenocarcinomas which are usually very large and fill the vagina at time of presentation. They are indistinguishable from RMS on imaging.[1, 3]

Other uterine sarcomas, such as leiomyosarcoma, mixed mesodermal tumors, and endometrial carcinomas have been reported in children but are extremely rare at that age. The most commonly encountered benign tumors in the uterus overall are leiomyomas; however, these are also uncommon in the pediatric population. Most leiomyomas present as well-circumscribed masses that are typically hypointense on T2-weighted sequences relative to myometrium, and demonstrate relatively little enhancement. Occasionally these tumors may demonstrate more pronounced enhancement and increased signal on T2-weighted sequences. Areas of degeneration manifest as lack of enhancement on post contrast sequences.[1, 3, 41]

Conservative surgical management is the recommended approach in non-metastatic RMS of the female genital tract, including the cervix in children and adolescents. Treatment of RMS includes a combination of chemotherapy for 4 to 5 months followed by surgical excision and/or radiation therapy [3, 40]

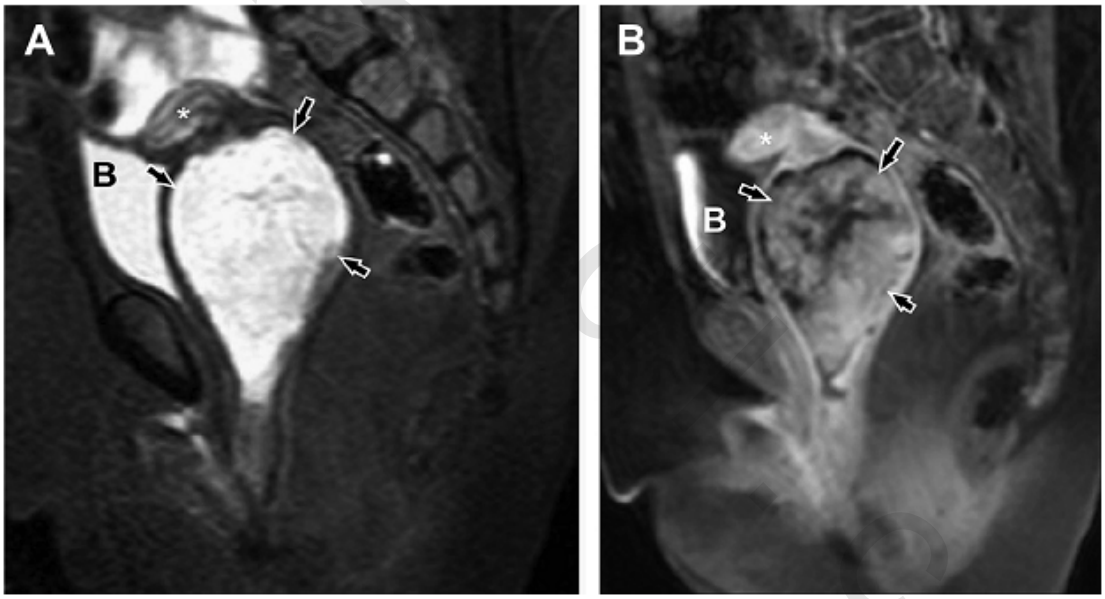


Figure (10): A 2-year-old girl with vaginal botryoides RMS. (A) Sagittal STIR image shows a hyper intense, slightly heterogeneous intra vaginal mass (arrows). The cervix and uterus (asterisk) are superiorly displaced. Mass effect is noted on the fluid-filled bladder (B). (B) Sagittal post contrast fat-saturated T1-weighted image showing intense heterogeneous enhancement of the mass (arrows).The cervix and uterus (asterisk) are separate from the mass.[1]

II Non Neoplastic

Pelvic masses in pediatric female patients may be caused by hydro/hematometrocolpos due to obstructive outflow tract disorders of the vagina. This can manifest at two different periods in life, at birth (hydrocolpos due to accumulated vaginal secretions) and at puberty after menarche (hematometrocolpos due to accumulated menstrual blood).Causes may include: Imperforate hymen, vaginal anomalies with or without Mullerian duct anomalies, urogenital sinus anomaly and cloacal malformation.[10, 42, 43]

Most cases of hydrocolpos in female neonates are due to Cloacal malformations which include a spectrum of complex congenital anomalies with different degrees of persisting fusion of the distal urinary, genital and intestinal tracts. It is commonly associated with genital fusion anomalies and obstruction.US usually reveals distended vagina forming a fluid filled pelvic cystic lesion displacing the urinary bladder anteriorly with characteristic inferior beak. Commonly fluid-debris level is seen due to mixture of vaginal secretions with urine or meconium. Contrast studies through the perineal orifice and colostomy opening has long been considered the reference standard imaging modality. However, in recent years, MRI has been used increasingly to better show the underlying anatomic derangements and associated anomalies. When combined with endoscopy, MRI can make other preoperative conventional imaging unnecessary.[10, 44-47]

Vaginal obstruction is commonly presented after onset of menarche by intermittent cyclic pelvic pain, palpable pelvic mass and primary amenorrhea. Normal menstruation is present in some cases with incomplete obstruction. Many cases are simply caused by imperforate hymen. Others may be due to congenital vaginal hypoplasia, duplication, or septum. Approximately 45% of vaginal septa occur in the upper vagina, 40% in the middle vagina, and 15% in the lower vagina. Some cases are associated with Mullerian duct anomalies as in type I (cervical/vaginal agenesis or hypoplasia) or type III and IV (where uterus didelphys or bicornuate uterus is associated with obstructing vaginal septa).[10, 16, 17, 43]

US and MRI are extremely useful in assessment of hydro/hematometrocolpos.US shows distended vagina with or without distended uterus with fluid collection containing low-level echoes.MR findings of vaginal obstruction are a tubular, fluid-filled, midline mass, representing the dilated vagina and uterus. The vagina typically is larger than the uterus. The distended vagina has a thin and almost imperceptible wall, whereas the uterus has a thicker muscular wall. The appearance of the internal contents is variable, depending on the nature of the fluid. Serous fluid has a low signal intensity on T1-weighted images and high signal intensity on T2-weighted images .The signal intensity increases on T1-weighted images if the contents are hemorrhagic. Additional findings on MRI include hydro- or hematosalpinx, ureteric dilatation and hydronephrosis.[16, 42, 48]

MRI with multiplanar image acquisition provides detailed information regarding uterine anatomy, the cervices, the continuity with the vaginal channels ,duplications, obstruction and the nature of the fluid contents (high signal on T1,T2 and T1-W fat saturated images are indicative of blood). The accuracy of MRI for diagnosing uterine and vaginal malformations is well established and even 100% accuracy has been reported. Information obtained from MRI are crucial for surgical planning .Differentiating simple cases of imperforate hymen from more complex cases with vaginal septum and Mullerian anomalies is of paramount importance due to different surgical techniques. The report on cases with vaginal septum should include its position (high, middle, low) and its thickness. The length

of atretic vaginal segments should be accurately reported, better from the sagittal plane. Identification of the cervix on MR images, especially in the sagittal planes, is crucial for differentiating a high septum from congenital absence of the cervix.[16, 42, 43, 48-51]

Early identification of vaginal obstruction by prompt accurate imaging that guide surgery helps to avoid prolonged symptomatic periods in young adolescents and the complications that may subsequently arise including hematometra, hematosalpinx, endometriosis (due to retrograde menstruation), pelvic adhesions and outflow obstruction to the urinary tract if severe obstruction is present. Also infections may be superadded on the accumulated blood forming pyocolpos, pyometros and pyosalpinx.[10, 42, 48, 52]

A well described congenital syndrome associated with hematometrocolpos is Herlyn-Werner-Wunderlich syndrome which includes uterovaginal duplication with unilateral vaginal obstruction, commonly to the right, and ipsilateral renal agenesis. It should be highly suspected in any neonate with a cystic mass detected behind the urinary bladder in association with the absence of one kidney.[10, 48]

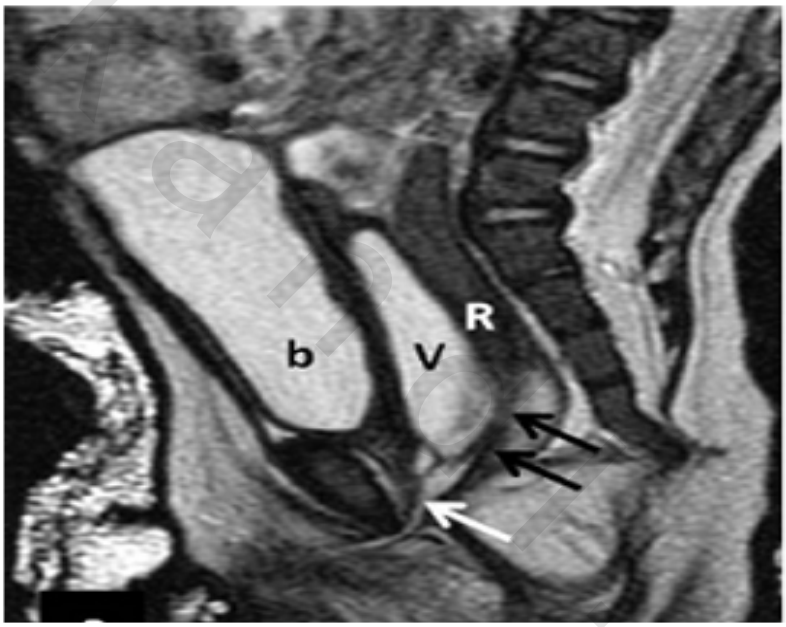


Figure (11): Mid sagittal MRI image in a 12 months old patient with persistent cloaca showing the urinary bladder (b), distended vagina (v), and rectum (R). The three systems converge to meet in the common cloacal channel (white arrow) above the pubococcygeal line (high type cloaca); black arrows point to the long rectal fistula before joining the common cloacal channel.[47]

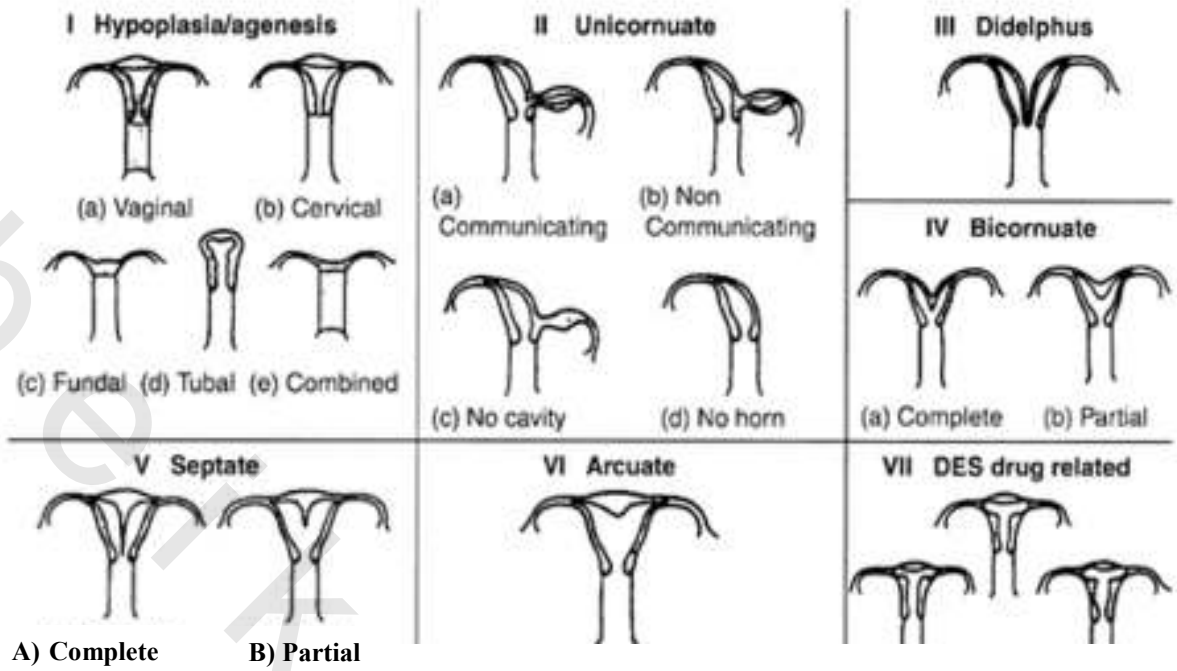


Figure 12: Classification of MDAs on the basis of the American Society for Reproductive Medicine system. DES= diethylstilbestrol.[43]

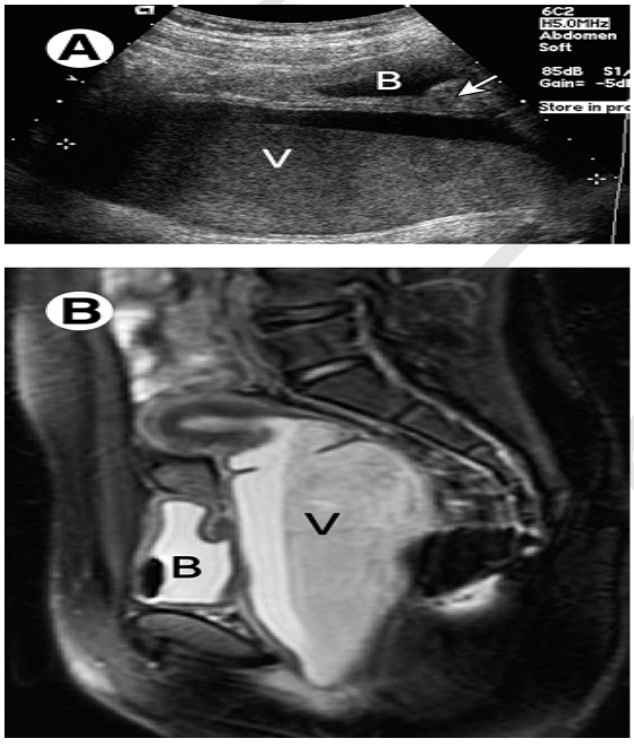


Figure (13): Imperforate hymen in a 13-year-old girl. (A) US shows a distended vagina (V) with a fluid level, which extends well below the level of the bladder (B) neck (arrow). (B) Corresponding T2-weighted MR image demonstrates similar findings to the US.[10]

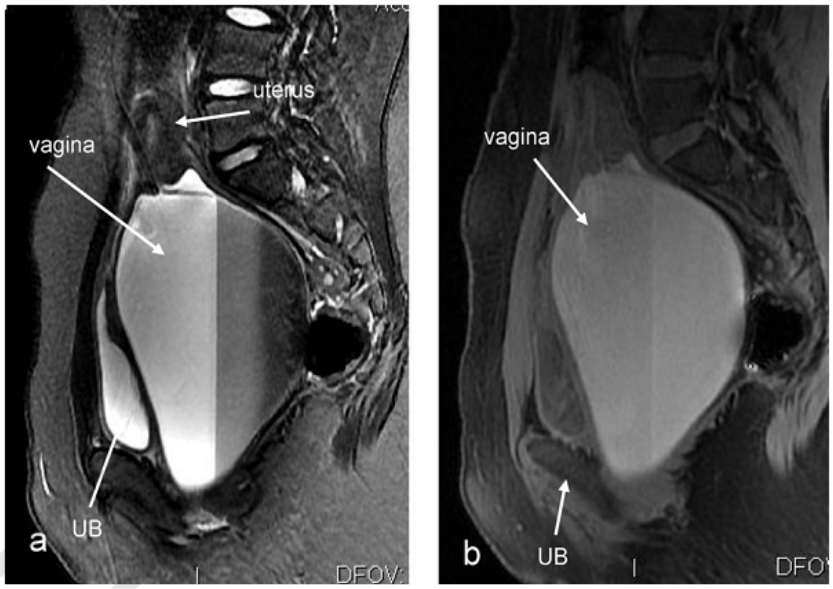


Figure (14): MRI of the pelvis of an 11-year-old girl: (a) sagittal T2, (b) sagittal T1 with fat saturation show a distended vagina containing a fluid/fluid hemorrhagic collection and the presence of hemorrhage at various stages. The uterus is displaced superiorly and is not distended. Imaging findings are suggestive of hematocolpos due to imperforate hymen. (53)

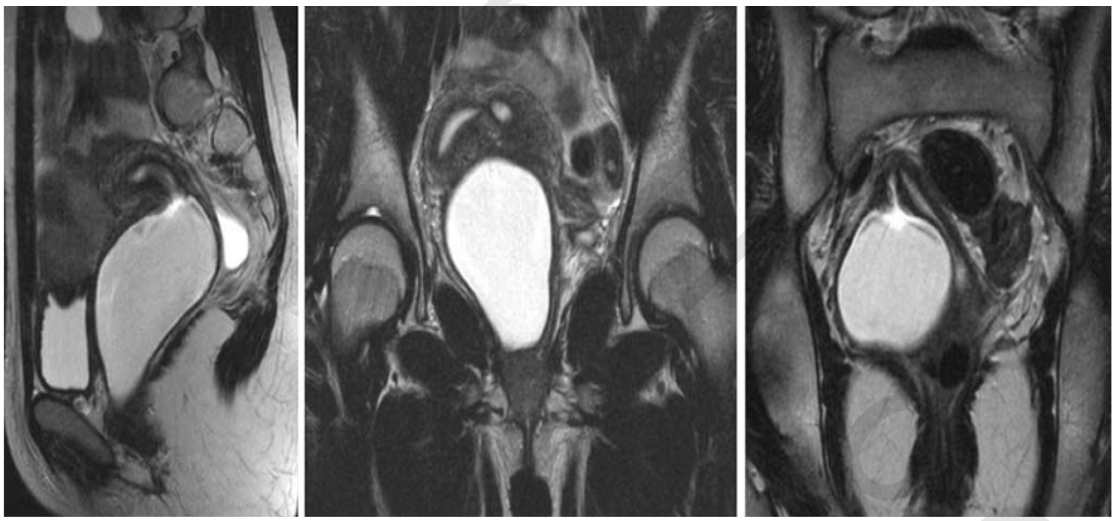


Figure (15): Sagittal and two coronal T2W MR Images show bicornuate bicollis uterus, right hemi-hematocolpos with inhomogeneous high-signal contents. A 13-year-old girl with Herlyn-Werner-Wunderlich syndrome (HWW) (48)

B) Ovarian masses

Ovarian masses in the pediatric age group consist of functional cysts in approximately 60% of cases and neoplasms in 40% of cases. Two-thirds of ovarian neoplasms are benign, one-third are malignant. The commonest ovarian tumor in pediatrics overall is teratomas.[17, 54]

Ovarian masses in pediatrics come to surgical attention in a variety of ways. Patients may present with acute abdominal pain and signs of peritonitis that can be difficult to distinguish from acute appendicitis. Patients may be referred with a large pelvic or abdominal mass and concerns of malignancy. Some cases present with signs of endocrine disturbance as precocious puberty, musculinization or vaginal bleeding. Other patients with large masses may experience ureteral compression and hydronephrosis, bowel obstruction, or respiratory insufficiency. Rarely, Meig's syndrome, with hydrothorax and nonmalignant ascites, may occur with fibromas, thecomas, or granulosa cell tumors of the ovary. [54-56]

Imaging of suspected adnexal mass may start with plain abdominal films that may show pelvic calcifications suggestive of a benign ovarian teratoma or show large pelvic opacity displacing bowel loops.US is the diagnostic study of choice for initial evaluation of potential ovarian pathology in all age groups and allows easy differentiation of solid and cystic components with the detection of septa, excrescences and calcifications.[57]

CT often provides poor density resolution of normal reproductive organs, but can be useful when assessment of the full extent of solid lesion is needed. Direct extension of tumors to adjacent pelvic organs or metastases to the liver and lungs can be demonstrated. This modality may also be useful for the detection of abscesses and lymphadenopathy. MRI offers superb soft tissue contrast resolution and is specially valuable for distinguishing ovarian from uterine lesions.[57]

I. Neoplastic:

Ovarian tumors account for 1% to 3% of malignant neoplasms in girls less than 17 years of age. Two thirds of ovarian tumors in children are benign and one third is malignant. They are classified based on their cell of origin into germ-cell, epithelial, and sex-cord stromal tumors. Germ-cell tumors are the most common and comprise 60% to 90% of pediatric ovarian neoplasms, followed by sex-cord stromal (10%–13%) and epithelial tumors (5%–11%).Ovarian neoplasms often contain cystic and solid components; in general, the solid component is a predictor of malignancy. The most common benign tumors are teratomas, followed by cystadenomas.[1, 3, 16]

In general, ovarian neoplasms often have nonspecific imaging features, making them difficult to differentiate from one another. Familiarity with age of onset, clinical symptoms, laboratory data, and imaging features allow for a more narrow differential diagnosis.[3]

Most ovarian neoplasms present as asymptomatic pelvic or abdominal masses. Isosexual precocity may be a presenting sign in patients with granulosa-theca cell tumors, whereas virilization may be observed in patients with Sertoli-Leydig cell tumors.[12, 16]

Although distinguishing benign versus malignant ovarian neoplasms is often difficult, US features suggesting malignancy include papillary projections, fluid in the Douglas pouch, tumors larger than 10 cm in largest diameter, lesions with irregular walls, and thickened irregular septa. The same features are also present on CT which allows better mass visualization than US and has been for years the initial study of choice for staging, as it allows for visualization of spread into the chest, abdomen, and pelvis. MRI is increasingly used instead due to the lack of ionizing radiation and due its superiority to CT in evaluating the pelvic side wall and adjacent organs for invasion.[3, 16, 58]

Malignant ovarian lesions appear as large solid masses (>50% soft tissue elements) with thick and irregular walls. They have a predominantly low to intermediate signal intensity on T1W images and intermediate or high signal intensity on T2W images. Other findings include foci of necrosis or hemorrhage, calcifications, thick septa and papillary projections. The malignant germ cell and stromal neoplasms spread by contiguous extension, lymphatic spread to regional or distant lymph nodes, or hematogenous dissemination to the lungs or liver. Ovarian carcinomas tend to spread by seeding the peritoneal or omental surfaces. Peritoneal tumor implants appear as nodules on the lateral peritoneal surfaces or in the ligaments and mesenteries of the abdomen .Omental implants appear as discrete nodules or conglomerate soft tissue masses ‘‘omental cake’’ beneath the anterior abdominal wall. Both have medium signal intensity on MRI and may enhance after IV contrast administration.[3, 16, 59, 60]

1-Ovarian germ cell tumors

Germ cell tumors include teratoma, dysgerminoma, endodermal sinus tumors, embryonal carcinomas, and choriocarcinoma. The most common type of pediatric ovarian germ cell tumor is a mature teratoma, which is benign. All of the other types are malignant and have the potential to metastasize to the liver, lungs, and peritoneum.[1]

Ovarian teratomas are congenital lesions containing tissue derivatives from all three germ cell layers (endoderm, ectoderm, and mesoderm). They account for two thirds of all ovarian tumors in children. They are bilateral in 25% of cases. Teratomas are of three main types: mature, immature(containing embryonic neural elements), and malignant .The mature cystic teratoma is the most common type of ovarian teratoma and the most common type of ovarian germ cell neoplasm .[16, 61, 62]

The tumors are unilocular in 88% of cases and are filled with sebaceous material. Squamous epithelium lines the wall of the cyst, and compressed, often hyalinized ovarian stroma covers the external surface .Hair follicles, skin glands, muscle, and other tissues lie within the wall. There is usually a raised protuberance projecting into the cyst cavity “known as the Rokitansky nodule” from which arise most of the contained hair and teeth. [16, 61]

The US findings of teratomas are variable due to its variable contents. They range from purely cystic to completely solid with echogenic calcifications and/or fat, mural nodules, floating debris, fluid levels, or any combination of these. Mural nodules “Rokitansky nodules” are seen as echogenic shadowing nodules projecting into the cyst lumen. Acoustic shadowing on US occurs secondary to calcified material or a matted mixture of sebum and hair and this shadowing can be extensive enough to prevent visualization of much of the underlying lesion which is termed the “tip of the iceberg sign.”[3, 61]

On CT, ovarian teratomas reveal a variable mixture of internal fat, fluid, and calcification, which can be characterized by Hounsfield units. Mural nodules, fat-fluid levels, and floating debris may also be seen. Teratomas rarely rupture into the peritoneal cavity, bladder, small bowel, rectum, sigmoid colon, vagina, or through the abdominal wall. Acute peritonitis and chronic granulomatous peritonitis due to a chronically leaking teratoma are exceedingly rare. Multiple small peritoneal implants and variable ascites may simulate carcinomatosis or tuberculous peritonitis.[3]

Like US and CT, MR imaging reveals a heterogeneous lesion of variable signal intensity. The presence of macroscopic fat in an ovarian tumor is diagnostic of a mature teratoma, and can be exquisitely demonstrated with the use of in-phase/opposed-phase imaging and fat-suppressed MR sequences. The fatty component of the tumor will demonstrate hyperintense signal on T1W and T2W images and will subsequently lose signal with fat suppression. Lesions that contain hemorrhage or proteinaceous material will demonstrate hyperintense signal on T1-weighted sequences both with and without fat suppression. Serous fluid is of low intensity on T1-weighted and bright on T2-weighted sequences. Calcified structures, such as bone and teeth, as well as hair demonstrate low T1 and T2 signal intensity. Contrast administration shows variable enhancement of the solid portions of the lesion and the cyst walls. Other findings seen on CT and US, such as fat-fluid levels, floating debris and rounded mural nodules "dermoid plugs" may also be seen with MR imaging.[1, 3, 16]

Immature teratomas are composed of tissues derived from the three germ layers (as mature type) but differ in that they demonstrate clinically malignant behavior, and are histologically distinguished by the presence of immature or embryonic tissues. Tumor grading is based on the amount of immature tissue present but recently, the amount of yolk sac tumor within immature teratomas has been recognized as both the source of alpha-fetoprotein in affected patients and the major predictor of stage, grade, and rate of recurrence. It is difficult to distinguish between mature and immature teratomas using imaging, but features that suggest immature components include a prominent soft tissue component of the mass with calcifications scattered throughout the lesions as opposed to contained within the Rokitansky protuberance, with small foci of fat and hemorrhage.[61-63]

Only 2% to 10% of ovarian teratomas are malignant. Specifically, central necrosis within a solid mass, thickened irregular septa and papillary projections are suggestive of malignancy. However, there is no imaging feature that is diagnostic of malignancy.[2, 3, 62, 64]

Other categories of germ cell tumors include dysgerminoma, yolk sac tumor "endodermal sinus tumor", choriocarcinoma, and embryonal carcinoma. These tumors are indistinguishable by imaging, but MR imaging findings direct the operative approach and provide preoperative staging. Final diagnosis and staging is performed by pathology. Hormonal markers maybe used for tumor type differentiation .For example, endodermal sinus tumors may have elevated levels of a-fetoprotein (AFP), choriocarcinoma show elevated HCG whereas embryonal carcinomas may have elevated levels of both. Other markers include lactate dehydrogenase, CA 19-9, and CA-125.[1, 6, 55, 65]

Dysgerminoma is the most common malignant ovarian neoplasm and usually seen as a solid mass with fine stippled calcifications and intralesional fibrovascular septa which demonstrate low signal intensity on T2-weighted sequences and avid enhancement on post-contrast sequences. They are tumors of adolescence and often large and bilateral in 10±15% of patients.[1, 3, 6]

Endodermal sinus tumors (yolk sac tumors) often have an elevated AFP which can be a useful marker for evaluation of tumor recurrence. Imaging shows a large nonspecific pelvic mass with cystic and solid components. Extension into peritoneum and adjacent structures, including the spinal canal, is not uncommon.[3]

As a general rule, ovarian germ cell tumors in children have an excellent prognosis even when malignant. Apparently benign tumors (non-secreting, no ascites, no lymph nodes or distant tumors) can probably be treated safely with a more conservative surgical procedure (ovary-sparing tumor excision rather than oophorectomy or salpingo-oophorectomy).Chemotherapy is commonly given post operatively in malignant cases with high survival rates.[66]

2-Ovarian stromal tumors

These tumors are the second most common in pediatric age group after germ cell tumors and include Granulosa cell, Sertoli-Leydig cell tumors, thecoma-fibroma, and undifferentiated sex cord stromal tumors.[12, 54, 55, 65]

Granulosa cell tumors secrete estrogen so they typically present with Isosexual precocity, uterine enlargement and bleeding. These neoplasms are highly variable in appearance on cross-sectional imaging ranging from cystic to solid, often being heterogenous because of internal hemorrhage and infarcts. Surgical resection is the mainstay of therapy, usually followed by radiation or chemotherapy.[1, 3, 67]

Sertoli-Leydig cell tumors may present with virilization or oligomenorrhea due to androgen production. These tumors are commonly solid lobulated with hypointense signal on MRI on T1-weighted and T2 weighted sequences due to fibrosis. Rare cases show multicystic appearance with papillary projections.[1, 3, 67, 68]

3- Ovarian epithelial tumors

These are the least common of pediatric ovarian tumors and divided into serous and mucinous subtypes. Benign cystadenoma is the most common epithelial tumor in childhood and adolescence. They are typically large at presentation and can present with ovarian torsion. [55, 65]

Serous cystadenomas usually have a significant unilocular or multilocular cystic component and thin septations. Mucinous cystadenomas are typically multilocular, septated masses, with the multiple cysts containing fluid demonstrating varying signal intensities. Papillary projections and solid, enhancing components may be present and are more predictive of malignancy.[1, 69]

4-Other ovarian tumors

Other rare ovarian neoplasms in children range from aggressive malignancies, such as small cell carcinoma, which is usually associated with hypercalcemia, to benign entities, such as hemangioma. Metastatic disease can also involve the ovaries, either from hematogenous or contiguous spread. Within this group of malignancies are adenocarcinoma of the colon, Burkitt's lymphoma, alveolar rhabdomyosarcoma, Wilms tumor, neuroblastoma, and retinoblastoma. The MR imaging appearance of ovarian metastases can be variable, but may be similar to that of the primary tumor .Imaging may show unilaterally or bilaterally enlarged, often hyperemic, variably enhancing ovaries. History of treated neoplasm is important for suggesting this diagnosis.[1, 3, 65, 70]

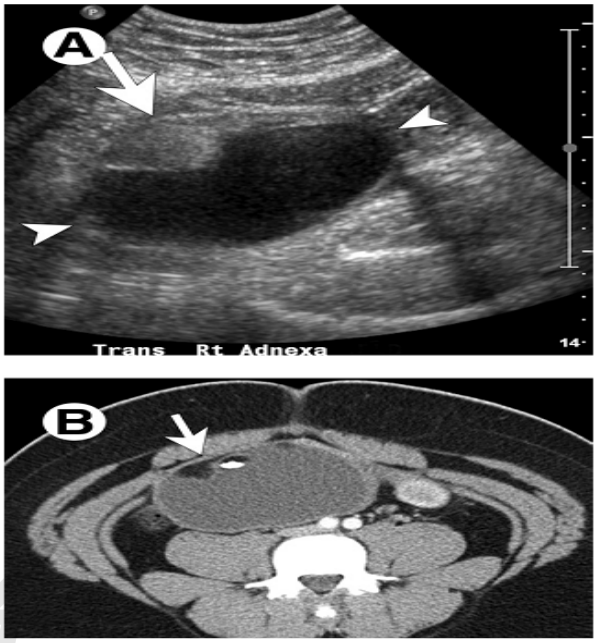


Figure (16): Mature ovarian teratoma. (A) Characteristic US appearance of a mature ovarian teratoma (arrowheads), which appears as a cystic adnexal lesion with an echogenic nodule (arrow),frequently referred to as a dermoid plug or Rokitansky nodule. (B) Corresponding axial CT image displays similar findings to the US image. The arrow points toward the dermoid plug or Rokitansky Nodule.(10)

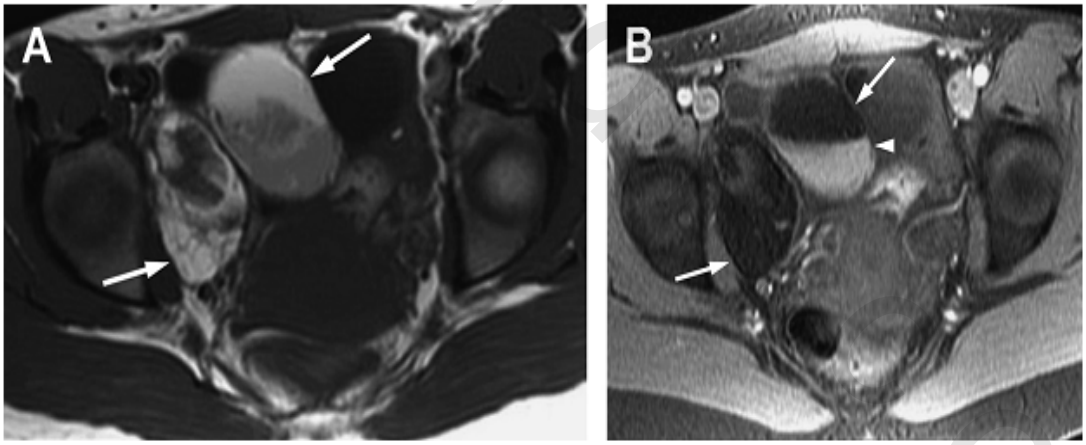


Figure (17): A case with bilateral mature teratomas. Axial T1-weighted image without fat-suppression (A) demonstrates 2 complex pelvic masses. Arrows denote the fatty components of the masses, which are hyperintense and show loss of signal on the fat suppressed image(B).Arrowhead denotes a fluid-fluid level within the mass.(1)

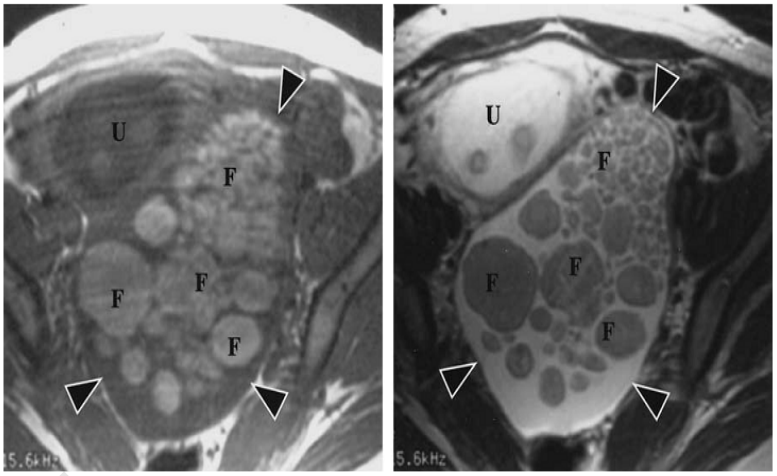


Figure (18): Dermoid cyst with intracystic nondependent spheres of lipid material. Left:T1-weighted MR image shows a large cyst (arrowheads) containing multiple round masses (F). Right:T2-weighted MR image shows the round masses (F) floating in high signal intensity fluid within the cyst (arrowheads).[61]

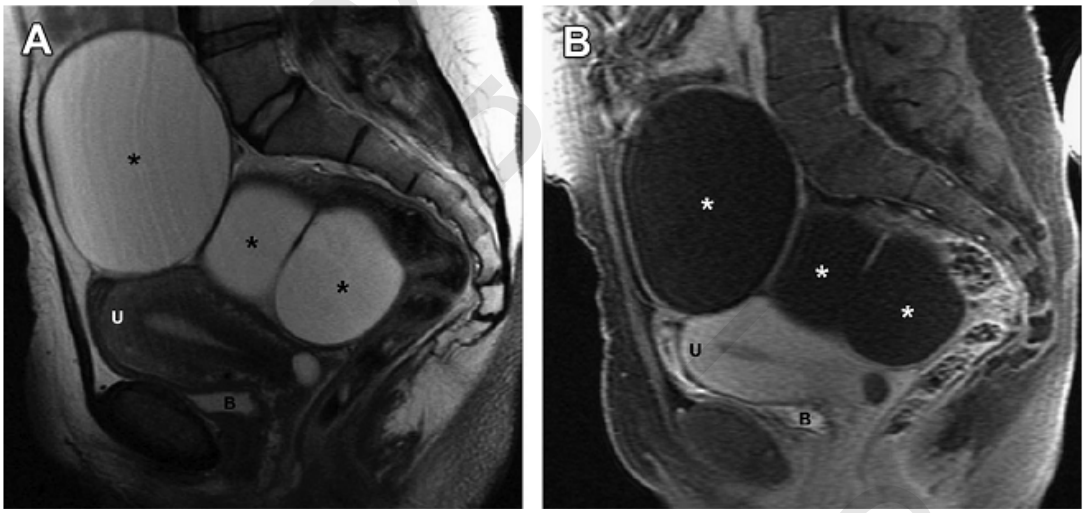


Figure 19: A 17-year-old with a mucinous cystadenoma. Sagittal T2-weighted image (A) demonstrates a multilocular, septated cystic mass (asterisks) just above the uterus. Sagittal T1-weighted fat-suppressed post-contrast image (B) shows peripheral enhancement of the cysts and septations. B, bladder; U, uterus.[1]

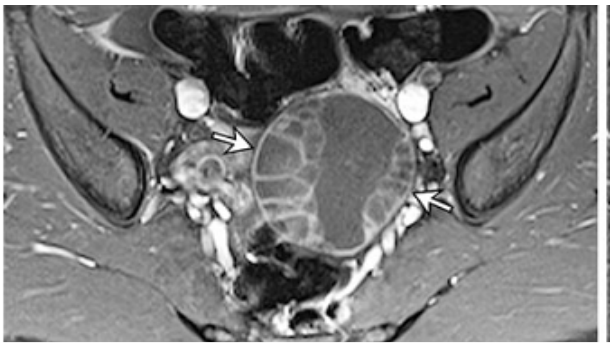


Figure (20): Sex cord stromal tumor. Axial T1-weighted post contrast showing multicystic ovarian mass with marked septal enhancement giving a spongy appearance.[69]

II Non-Neoplastic:

Functional ovarian cysts

This is the most common adnexal abnormality in the pediatric population and the most frequent cause of an abdominal mass in the fetus and in the newborn. An anechoic focus in an ovary is considered a follicle if it is smaller than 3 cm. A mature dominant follicle may fail to involute appropriately and may enlarge into a functional cyst or corpus luteum. Rupture or hemorrhage often brings these patients to medical attention. Ovarian cysts are present in the majority of newborn girls owing to maternal hormonal stimulation, in about 3% of girls less than 8 years of age and are quite common during early adolescence.[1, 55, 65]

Ovarian cysts may be asymptomatic, incidental findings on routine imaging studies, or associated with life threatening hemorrhage, peritonitis, or respiratory compromise or lead to ovarian torsion and possible ovarian loss.[55]

Based on the degree of complexity associated with blood products, clot formation, lysis, and retraction, appearances of complicated ovarian cysts are variable on all cross-sectional imaging modalities. Simple cysts are seen on US as anechoic lesions with thin imperceptible wall and posterior enhancement. Hemorrhagic cysts demonstrate variable appearances from complex masses with septations, fluid-debris levels, or low level echogenicity with increased sound through transmission. They are avascular on color Doppler. A change in appearance over time may provide a clue to diagnosis.[10, 17, 65]

MR imaging can confirm the presence of a cyst when the US imaging characteristics are atypical or when an underlying mass is suspected. A simple ovarian cyst should demonstrate homogeneous T1 hypointense signal and T2 hyperintense signal with uniform peripheral or rim enhancement after the administration of IV gadolinium. The MR appearance of hemorrhagic cysts can vary depending on the age of the clot. Acute blood has a high signal on T1 but low signal intensity on T2 while subacute blood is typically bright on both T1- and T2-weighted images. Fat-suppressed images can help to differentiate between hemorrhagic cysts and fat-containing tumors, such as teratomas. Some cysts may also demonstrate layering of fluid and blood, which is referred to as a ‘‘hematocrit effect”.[1, 16]

Simple ovarian cysts discovered incidentally are commonly only followed by US. In asymptomatic neonates**,** surgical intervention is reserved for complex masses that fail to undergo involution by 4±6 months, and for those cases in which recurrence occurs following aspiration or acute abdominal findings develop. In childhood, small (<1 cm) simple cysts found incidentally should be considered as normal findings requiring no intervention. Larger simple cysts should be monitored conservatively as regression is likely. Further imaging and surgical diagnosis is indicated for acute symptoms to exclude adnexal torsion, for solid or complex masses and large masses that fail to undergo regression to exclude neoplastic lesions. In adolescents, cysts usually need only interval observation as most functional cysts regress after 2-3 menstrual cycles or otherwise would need further evaluation.(71)

It is important to combine imaging findings with thorough clinical and history data to differentiate functional cysts from other possible cystic masses including previously undiagnosed obstructive uterine anomalies, uterine leiomyomas, peritoneal cysts, peri appendiceal abscesses or less common causes in the sexually active adolescent as ectopic pregnancy and tubo-ovarian abscess.(71)

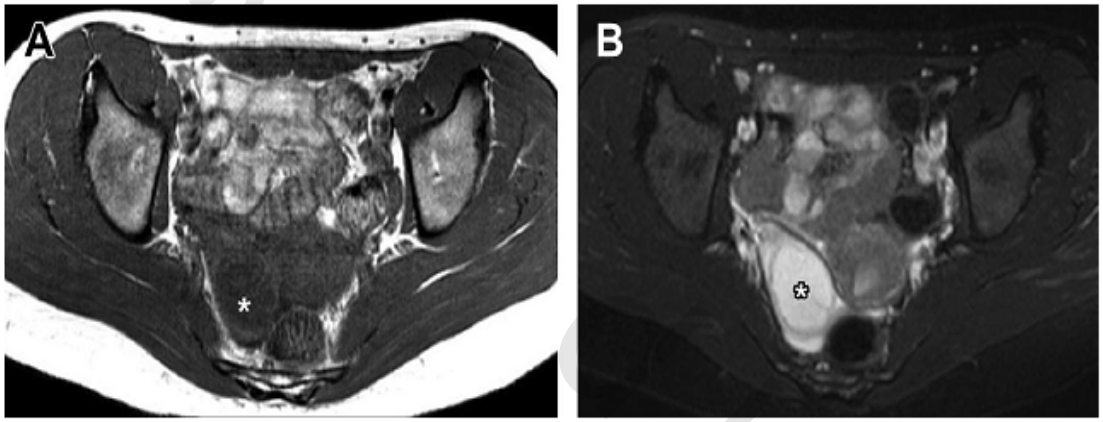


Figure (21): A 16-year-old girl with a simple right ovarian cyst. Axial T1-weighted (A) and axial T2-weighted fat suppressed (B) images of the right ovary demonstrate the characteristic homogeneous T1-weighted hypointense and T2-weighted hyper intense signal of a simple cyst (asterisks). No central enhancement should be present on post contrast imaging.(1)

Ovarian torsion

Ovarian torsion has bimodal age of distribution with the infant peak in response to maternal hormones and the early adolescent peak (9-14 yrs) in response to the maturing reproductive hormonal axis. Ovarian torsion is more common in patients with predisposing lesions such as an ovarian cyst or mass (commonest is mature cystic teratoma).In children, torsion of the normal ovary is also encountered because the fallopian tube is relatively long and the ovary is hence more mobile.(10, 17)

Clinical findings are acute severe lower abdominal pain and vomiting. This severe episode is usually preceded by intermittent pain as the adnexa twists and untwists. Fever is unusual unless necrosis is present. A palpable mass is present in 50% of patients. Ovarian

torsion is more common on the right, and, as a result, many of these patients carry a preoperative diagnosis of acute appendicitis.[10, 17, 54]

On US, adnexal torsion appears as a hypo- or hyperechoic midline mass with good sound transmission reflecting vascular engorgement and stromal edema with multiple enlarged follicles at the periphery .The classic color Doppler finding in ovarian torsion is the absence of arterial flow. Unfortunately, the absence of signal is not diagnostic, since Doppler waveforms are not always obtainable in normal ovaries in young girls and because of double blood supply to the ovary. Indeed, arterial flow (peripheral or even central) can be seen in surgically proven twisted ovaries. A more suggesting sign is decreased or absent venous flow. The sonographic "whirl pool sign" is highly specific where the vascular pedicle itself is seen on grayscale US as an echogenic round or beaked mass with multiple concentric, hypoechoic, target-like stripes associated with circular or coiled vessels. Ovaries without flow in the vascular pedicle at color Doppler sonography are always necrotic or infracted at surgery so this sign is also a helpful marker of ovarian viability.[17, 72-75]

CT and MR imaging are usually not required for diagnosis but can be useful when results of US are indeterminate. CT findings are somewhat nonspecific and include an adnexal mass that may be in the midline, rotated toward the contralateral side, deviation of the uterus to the side of the affected ovary and ascites. Like US, peripheral subcentimetere hypoattenuating structures are seen within the mass representing dilated follicles. Other findings include lack of enhancement, obliteration of fat planes, hematoma, and gas within the torsed mass .[72, 76]

MRI is not the primary imaging study for the evaluation of adnexal torsion, but recognition of the MR findings is important, because the lesion can have a subacute course and be unexpectedly seen on imaging examinations. Also, MRI may assist in the diagnosis of acute cases when sonographic findings are atypical or when there is suspicion for an underlying mass acting as a lead point. MR findings of acute torsion include an enlarged edematous adnexa (low T1-weighted and high T2-weighted signal intensity), prominent follicles along the periphery of the ovary, engorged adnexal vessels on the side of the torsion, deviation of the uterus to the side of the twist, and obliteration of the adjacent fat planes. In subacute or chronic torsion, the signal intensity may be low on T1- and T2-weighted images. A low-signal-intensity torsion knot and thick, straight blood vessels that drape around the mass maybe also seen .The ovary usually enhances when torsion is incomplete or intermittent while in complete torsion there's hemorrhagic infarction and absence of enhancement. Foci of high signal intensity corresponding to intralesional hemorrhage may also be noted on T1-weighted images more on the periphery of the mass.[16, 57, 61, 76, 77]

Isolated fallopian tube torsion has also been reported in both premenarchal girls and adults**.** It is a very rare entity however it should be considered in the differential diagnosis for peri menarchal girls who present with acute pelvic pain and who demonstrate a cystic mass in a midline position (either in the cul-de-sac or superior to the uterus) associated with a normal ipsilateral ovary. A specific sign is demonstration of hematosalpinx which is confidently demonstrated on MRI as hyperintense signal of tubal content on fat-sat T1WI combined with lack of enhancement of tubal wall. This condition maybe associated with paratubal cyst or less commonly in normal tubes.[78-80]

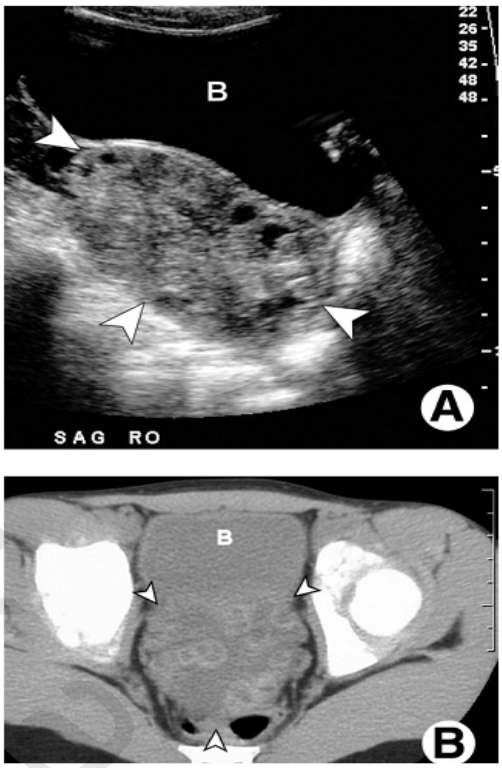


Figure (22): Ovarian torsion. (A) Sagittal US scan shows an enlarged, heterogeneous right ovary (arrowheads) with scattered peripheral cysts. (B) CT demonstrates the torsed right ovary with peripheral cysts.B, bladder.[10]

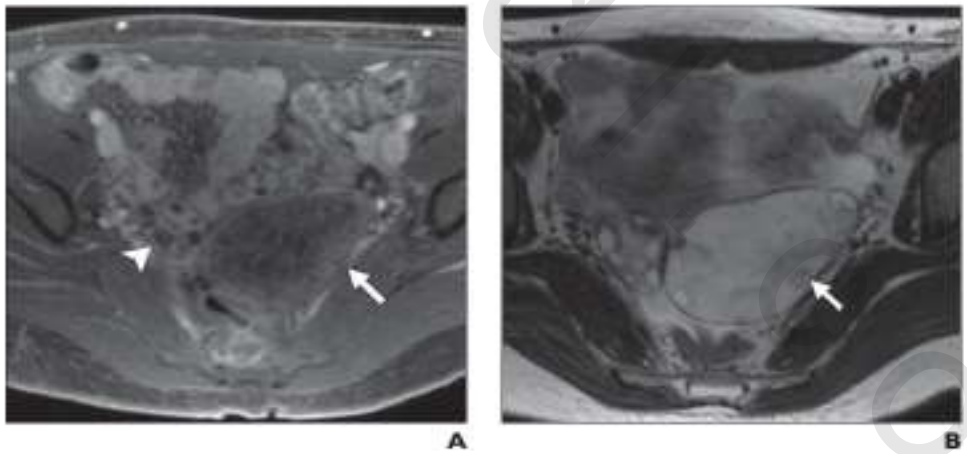


Figure (23): A) Fat-Sat GAD-enhanced T1W MRI shows 7.5-cm left ovary (arrow) with patchy irregular enhancement of afollicular stroma and peripheral follicles. Right ovary (arrowhead) shows normal enhancement. B) Axial fast spin-echo T2W MRI also shows the peripheral follicles and afollicular hyperintense edematous stroma.[77]

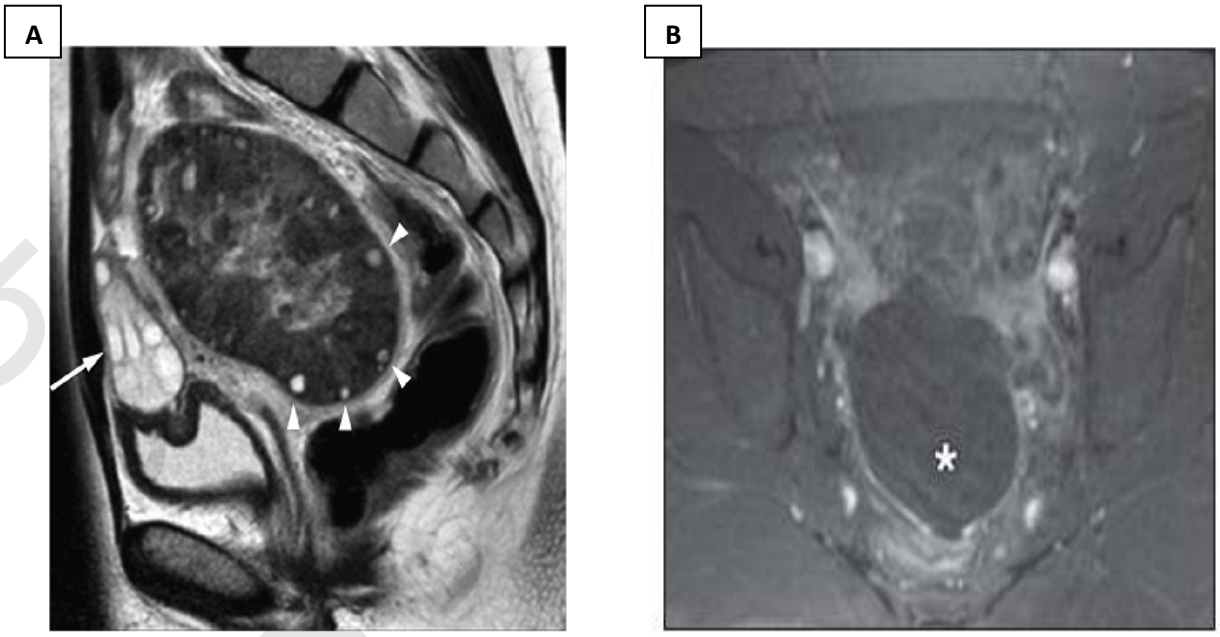


Figure (24): A: An 11-year-old girl with acute abdominal pain and ovarian torsion. Sagittal T2W image demonstrates a hypointense "mass" with peripheral hyperintense follicles (arrowheads), described as a "string of pearls". No underlying mass was detected at surgery. Arrow denotes the patient's other normal ovary.(B)Axial T1W Fat-Sat image after GAD injection from another case with ovarian torsion shows the hypointense featureless large necrotic ovary with no sign of contrast enhancement.[1, 65]

Tubo-ovarian Abscess

Tubo-ovarian abscesses (TOA) are almost always a complication of pelvic inflammatory disease (PID). PID is an infection of the upper genital tract and the majority of cases result from an ascending infection of bacteria from the vagina and cervix. Consequently this condition is very rare in pediatric patients most commonly found in sexually active young adolescents. Otherwise, it may be associated with inflammatory or infectious conditions of the gastrointestinal tract as Crohn's disease or appendicitis. Classic imaging findings of tubo-ovarian abscesses are complex fluid collections with thick walls and rim enhancement in the ovary and tubes often with adjacent inflammatory change and free fluid. Imaging features maybe misleading suggesting other pathology as ovarian neoplasms, torsion, or ectopic pregnancy. CT and MRI are better than US for giving global view of the pelvis to better characterize the adnexal lesion and clarify the inciting pathology.[55, 65, 81]

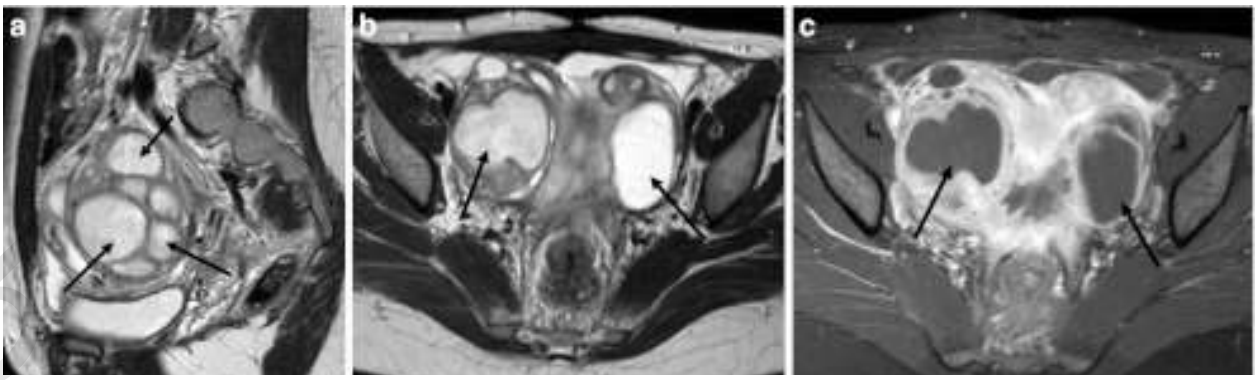


Figure 25: Pelvic inflammatory disease with bilateral TOA on MRI.(a) Sagittal T2 image of the pelvis demonstrates multiple fluid-filled cystic structures within the right adnexa (black arrows) showing thick walls and adjacent fat stranding. (b) Axial T2 image demonstrates bilateral tubo-ovarian abscesses. (c) Axial T1 fat-saturated image after GAD injection demonstrates low signal intensity within the pus-filled cavities and marked enhancement of the inflammatory walls.[82]

Posterior compartment (Presacral masses)

The presacral or retrorectal space is a potential space located between the rectum and the sacrococcygeal part of the spine.The levator ani and coccygeus muscles form the inferior boundary, peritoneal reflections define the superior boundary, and the ureters and iliac vessels define the lateral borders of this space.The presacral space contains a variety of tissues, including fat, mesenchymal tissue,lymph nodes, nerve plexuses, and vessels. Hence, a variety of tumoral lesions may affect this area. [5, 28]

Presacral lesions are generally rare with incidence may be as low as 1 in 40,000 hospital admissions. They may be congenital, developmental, neurogenic, inflammatory, mesenchymal, and osseous (extension of sacral neoplasms). The variety and heterogeneity of these masses has resulted in confusion regarding their classification and management.Recently proposed classification into congenital versus acquired and benign versus malignant, resulting in 4 distinct groups, each consisting of a variety of histologic subtypes but with a similar clinical presentation, diagnosis, treatment, and prognosis, is simple and efficient.The most frequent tumours are congenital and represent 2/3 of all cases, and benign tumours make up 66% of these. Teratoma is the commonest benign tumour and chordoma is the commonest malignant one.[28-30]

Classifications of Pediatric Presacral Masses

- Congenital and developmental masses
  - Germ cell tumors (sacrococcygeal teratoma, germinomatous and nongerminomatous germ cell tumors)
  - Anterior sacral meningocele
  - Developmental cysts (epidermoid cyst, dermoid cyst, enteric cysts [rectal duplication cyst, tailgut cyst])
  - Cystic lymphangioma
  - Lipoma
- Neurogenic masses
  - Neuroblastoma
  - Ganglioneuroblastoma
  - Ganglioneuroma
  - Neurofibroma
  - Schwannoma
- Inflammatory masses
  - Inflammatory bowel disease (ulcerative colitis, Crohn disease)
  - Perirectal abscess
  - Granuloma
- Mesenchymal masses
  - Rhabdomyosarcoma and undifferentiated sarcoma
  - Vascular mass
  - Fibroma
- Lymphomatous masses
  - Lymphoma (usually non-Hodgkin)
  - Posttransplantation lymphomatous disorder
- Extension of sacral bone tumors
  - Giant cell tumor
  - Aneurysmal bone cyst
  - Chordoma
  - Osteoblastoma
  - Ewing sarcoma family
  - Osteogenic sarcoma
- Other presacral masses
  - Hematoma
  - Extension or metastasis to the presacral space from another site

Figure (26): Classification of pediatric Presacral masses.[5]

The clinical presentation is determined by etiology, size and location of the lesion, and the age of the patient at diagnosis. Possible symptoms include constipation, palpable mass, urinary symptoms, and lower back pain. Careful rectal examination is essential and may give some indication of the likely diagnosis. [1, 5, 11, 28, 83]

Both CT and MRI scans are now used in a complementary manner rather than exclusively in the evaluation of presacral lesions. A CT scan can be used to determine whether a lesion is solid or cystic, evaluate cortical bone destruction, show lung deposits in malignant lesions and assess involvement of adjacent viscera. MRI, because of excellent soft tissue resolution, aids in determining the planes of resection, spatial relationship to surrounding structures, and associated cord abnormalities, as well as the extent of bone marrow involvement. Other diagnostic modalities that can help in the diagnosis and management of presacral lesions include flexible endoscopy and endorectal US.[1, 5, 84]

The purpose of diagnostic investigations, particularly cross-sectional imaging, is to accurately evaluate the location and nature of the lesion and determine the extent of involvement of the surrounding structures. This, in turn, helps to determine the surgical approach(anterior versus posterior versus combined) and the intraoperative extent of resection (local excision versus en bloc resection).The surgical approach and extent of resection is determined by the location, nature, and size of the lesion while taking into account whether or not the sacrum, pelvic sidewall, or adjacent viscera are involved. [1, 5, 28, 84]

An appropriate differential diagnosis may be formulated by considering the specific imaging features of a given case: In particular, involvement of the sacrum (either remodeling or destruction) and the presence or absence of a solid, soft-tissue component. Typically, osteochondral and neurogenic tumors remodel or destroy the sacrum, whereas sacral involvement is less common in patients with a mesenchymal tumor. Ewing sarcomas and chordomas are typically associated with a large soft-tissue mass. Teratoma shows areas of fat that are hyperintense on both T1 and T2 and lose signal on fat suppresion.Osseous or chondroid matrix is indicative of osteosarcoma or chondrosarcomas; neurofibromas may have a target appearance at MR imaging and hemangiomas have areas of increased signal intensity on T1-weighted MR images.[84]

A consensus exists that all presacral tumors should be resected, even if the patient is asymptomatic and even though many tumors are benign . It is the only possible curative treatment for malignant tumors in this area, which are known to be chemoradioresistant. Resection is necessary for benign tumors because of a tendency towards malignant degeneration and for prevention of future infection of cystic lesions.Several surgical approaches are used for the resection of presacral tumors: anterior (abdominal), posterior (trans-sacral) and combined . The choice is dictated by the patholgoical grade of malignancy, size and location of the tumor and its relationship to contiguous structures**.** Small low lying lesions below the level of the S3 vertebra can be removed through a posterior approach.Tumors extending above the level of the S3 vertebra can be removed either through an anterior trans-abdominal incision or through a combined anterior and posterior approach depending on the need for concurrent sacrococcygeal resection.[11, 28]

I. Neoplastic:

1-Sacrococcygeal teratoma (SCT):

Sacrococcygeal teratoma is the most common presacral tumor in children and the most common solid tumor in neonates, with a prevalence of one in 35,000–40,000 births. Sacrococcygeal teratomas are three to four times more common in girls, and 60% of tumors are benign. Histologically teratomas are classified as mature or immature. Mature teratoma comprises only mature elements derived from at least 2 of 3 germ cell layers (such as the skin, hair, fat tissue, cartilage, bone, glands, etc.). Immature teratoma contains immature elements such as neuroepithelial tissue and immature mesenchym. [62, 84, 85]

The classification system of SCT is based on Altman's survey of the Surgical Section of the American Academy of Pediatrics proposed in 1973.SCT is classified into four types based on the relative amounts of internal and external tumor: type I are predominantly external (47%), type II are dumbbell shaped with external and intra-pelvic components(34%), type III have a small external component with most of the tumor extending into the pelvis and abdomen (9%), and type IV are entirely presacral (10%).The probability of malignancy increase in less apparent lesions (grade III and IV) and with increase of the age of the child. At birth, the risk is about 1.5 per cent, but masses discovered after the age of 1 year had a risk up to 40% of being malignant. [30, 86-88]

Most SCT are diagnosed on antenatal US scans or are visible mass at the gluteal region at birth, making the diagnosis obvious. Lesions with a large intra-pelvic component may cause urinary or bowel obstruction in the neonate. The diagnosis of purely intra-pelvic teratomas may be delayed. Children commonly have long history of constipation, urinary

retention, an abdominal mass, or symptoms of malignancy, such as failure to thrive. Serum markers (alfa fetoprotein and beta HCG) before the operation are helpful for later comparison.[85, 89]

About 18% of patients with SCT have other congenital anomalies. These may include spinal dysraphism; sacral agenesis; and, occasionally, dislocation of the hips caused by a large tumor. Renal cystic dysplasia, imperforate anus and gastroschisis were also noted in few cases.[90]

Pelvic x rays may only show pressure erosions in longstanding cases as benign tumors do not invade or destroy the sacrum. The angle of the sacrum may also be increased. In cases of Currarino syndrome, a presacral teratoma is associated with scimitar deformity of the sacrum and anorectal anomaly. At US, SCT shows different imaging patterns ranging from a complex mass to a predominantly cystic mass, which may be unilocular or contain multiple septations. Echogenic foci maybe noted if fat or calcifications are present. A small solid nodule may be present within the cyst. Fluid-fluid levels are seen less commonly than they are in ovarian teratomas.[14, 69, 90]

On MR images, SCT can be seen a cystic mass that contains fat, calcification, bone, or teeth. Usually there are no associated osseous anomalies. In general, predominantly fluid-filled teratomas (< 50% solid elements) are benign whereas tumors that contain predominantly solid components are more likely to be malignant .Fluid-filled, cystic SCT have low signal intensity on T1-weighted and high signal intensity on T2-weighted MR images. The signal intensity can be greater than that of muscle on T1W images if the contents contain blood or proteinaceous material. Fat components have high signal intensity on T1- and T2W images and low signal intensity on fat-suppression sequence. Calcifications, bone, or hair appear as foci of low signal intensity on all imaging sequences. Although SCT arise from the coccyx, abnormalities of the spine are uncommon.[5, 84]

Malignant teratomas are predominantly solid heterogenous masses with intermediate signal intensity (equal to muscle) on T1-weighted and T2-weighted images. Hemorrhage and necrosis are common. T1-weighted images demonstrate intense heterogeneous enhancement. Margins are usually poorly defined and infiltrating into adjacent soft tissues. Other signs of malignancy include intra-spinal involvement, soft tissue stranding or mass, lymph node enlargement, and sacral destruction. MRI is superior to CT in detecting invasion of pelvic soft tissue structures and identifying intra-spinal involvement .After treatment, malignant SCT may regress entirely, leave a small fibrotic mass, or convert to a mature teratoma .Metastases are to the lungs and occasionally to the liver or retroperitoneal lymph nodes. CT scanning is the best study to assess the presence or absence of metastatic pulmonary disease. Tumor spread to the upper abdomen can be reliably evaluated by MRI during evaluation of the primary tumor.[1, 2, 5, 91]

There are no diagnostic methods, short of surgical resection and histological evaluation, that allow the absolute exclusion of malignant neoplasm. However, whether a tumor is benign versus malignant may, to some extent, be predicted on the basis of the clinical findings and imaging characteristics. Factors usually associated with benign lesions include early (neonatal) presentation, female patient, predominantly external location of the tumor, predominantly cystic composition, and the presence of large areas of calcifications. Factors associated with malignant lesions include clinical presentation

beyond infancy, male patient, predominantly presacral location, predominantly solid composition (particularly with areas of hemorrhage or necrosis) and lack of calcification.[92]

The management for all benign tumors (mature and immature teratomas) and select completely resectable malignant tumors is surgery alone. Modern-day chemotherapy is extremely effective in infants and children with irresectable and metastatic disease and these children have a very high survival rate. The use of neoadjuvant chemotherapy allows vital organ preservation and there is no role for resection of vital structures at the time of initial presentation.[87]

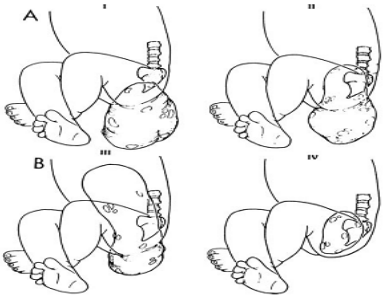


Figure (27): Schematic diagram showing the Altman classification system for sacrococcygeal teratomas. [5]

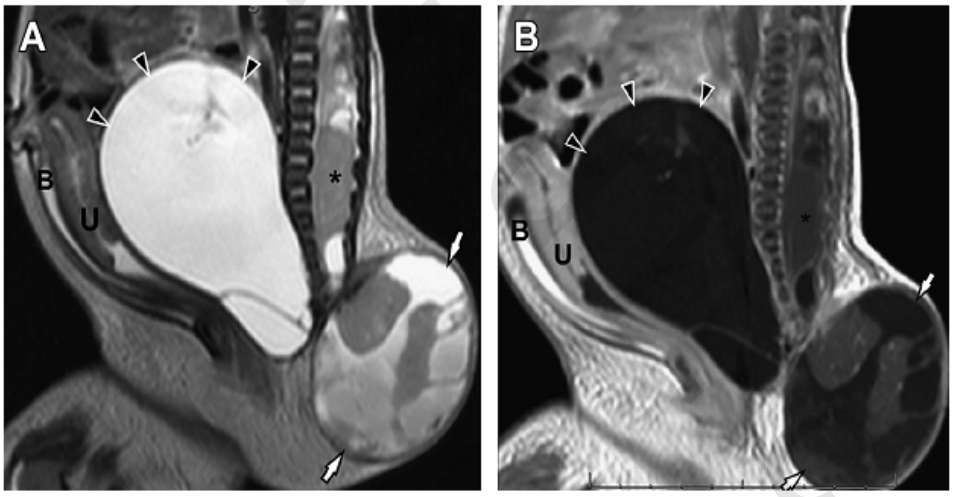


Figure (28): A newborn girl with a large sacrococcygeal teratoma. (A) Sagittal fat-saturated T2-weighted image demonstrates large external mass (arrows), with both cystic and solid components and large predominantly cystic intrapelvic mass extending to the lower abdomen (arrowheads). Mass effect on the uterus (U) and bladder (B) is noted. (B) Sagittal postcontrast fat-saturated T1-weighted image shows only minimal enhancementof solid components. Intraspinal extension of the mass at the level of the conus medullaris (asterisk).[1]

2- Yolk sac tumor (endodermal sinus tumor)

This is another type of germ cell tumor that may occur in the sacrococcygeal region. It commonly has a more aggressive imaging appearance than teratomas and is characterized by more heterogeneous soft tissues and areas of hemorrhage and necrosis.[84, 93]

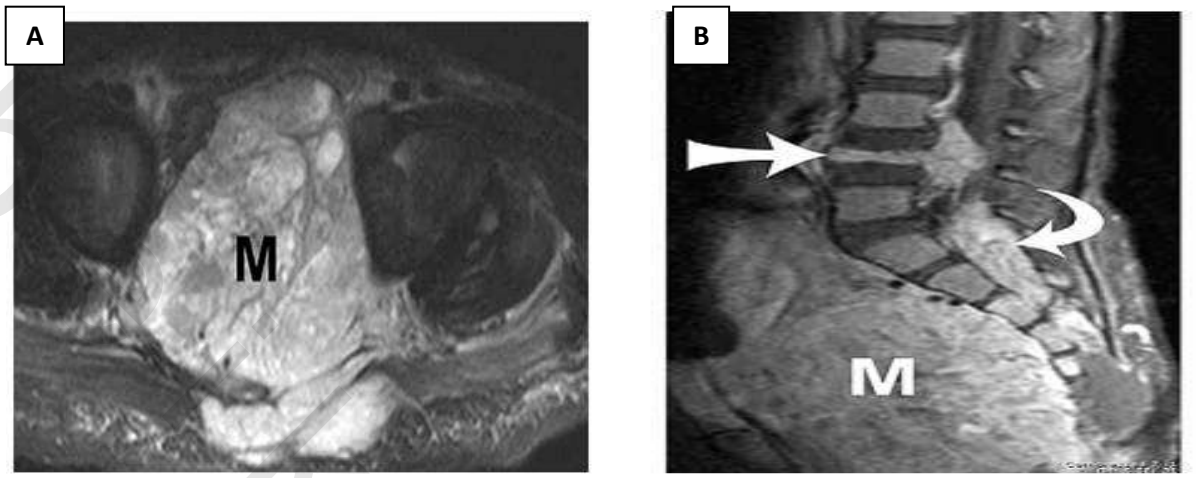


Figure 29: Yolk sac tumor in a 2-year-old female with perineal skin changes (A) Axial and (B) Sagittal contrast-enhanced fat-suppressed T1-weighted MR images reveal a heterogeneous, enhancing pelvic mass (M). Spine involvement is causing vertebra plana (arrow) and epidural extension (curved arrow).[3]

3-Primary sacral bone tumours:

Although primary tumors of the sacrum are uncommon, when encountered they may extend into the presacral space. Benign tumors include for example giant cell tumors and aneurysmal bone cysts and malignant tumors include Ewing sarcoma, osteosarcoma and chordoma. However, metastatic involvement of the spine by tumors with an extra spinal origin is more common. [1, 94]

A) Chordoma

Chordoma is the most common primary malignant sacral neoplasm .It is most commonly manifested in the fourth through the seventh decades of life, but they can occur in children. It appears as destructive lytic midline lesion with multiple amorphous cartilaginous calcifications. It usually has a large destructive presacral soft-tissue component with destruction of the sacrum and coccyx, crossing to the sacroiliac joints and extensions within the sacral foramina. Chordomas demonstrate low to intermediate signal intensity on T1, significantly high signal intensity on T2 images and variable contrast enhancement. Treatment consists of resection and radiation therapy.[2, 5, 84, 94]

B) Ewing sarcoma

Ewing sarcoma of the bone, extraosseous Ewing sarcoma, and primitive neuroectodermal tumor belong to one group of small round cell tumors: the Ewing′s sarcoma family of tumors (ESFTs) with similar histologic and radiologic features but different cytogenetic and immunohistologic characteristics.These tumors typically fill the bone marrow cavity, destroy the cortex, and produce a soft tissue mass that is usually

larger than the bone component. Another pattern of development is eccentric extraosseous extension of the tumor secondary to erosion of the cortex. Radiography and CT show lytic, sclerotic, or mixed features in the tumor. CT depicts the soft tissue component and the bone lesions better than radiography demonstrating permeative bone lysis, osseous expansion, or sclerosis.[5, 95]

MR imaging clearly depicts both intra- and extraosseous components of the tumor, including paraspinal, extradural, and presacral involvement .The extraosseous component is usually the predominant part of the tumor and may include thin fibrous septa. Radiation and chemotherapy are the standard treatments for patients with Ewing sarcoma, although those with neurologic instability or compromise may require surgical decompression and stabilization.[1, 5, 96]

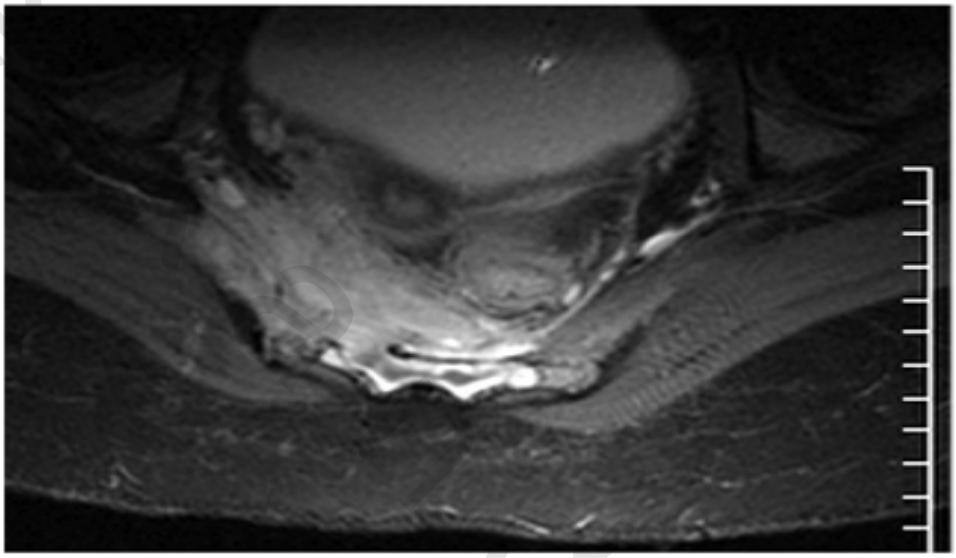


Figure (30): Ewing sarcoma. An ill defined high-signal-intensity presacral mass with bone involvement and invasion of the sacrum.[84]

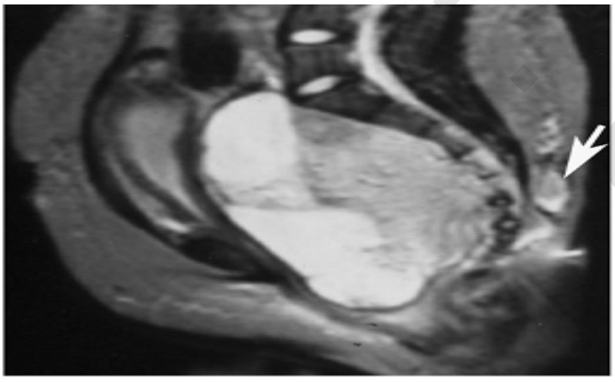


Figure (31): Ewing sarcoma. Sagittal T2-weighted MR demonstrates a heterogeneous presacral mass with extension posterior to the sacrum (arrow).[5]

C)Giant cell tumor

This bone tumor is the second most common neoplasm of the sacrum, after chordoma. Sacral lesions are usually eccentric and may extend across the sacroiliac joint. Giant cell tumors are frequently lytic and destructive and do not contain calcifications or septa.They generally appear heterogeneous on CT and MR. Low-attenuation areas on CT scans represent necrosis. Hemorrhage is present when there is high signal intensity on both T1- and T2-weighted images. Fibrotic tissue appears as an area of low signal intensity on both T1- and T2-weighted images. Both CT and MR images show contrast enhancement of the tumor. [5, 96]

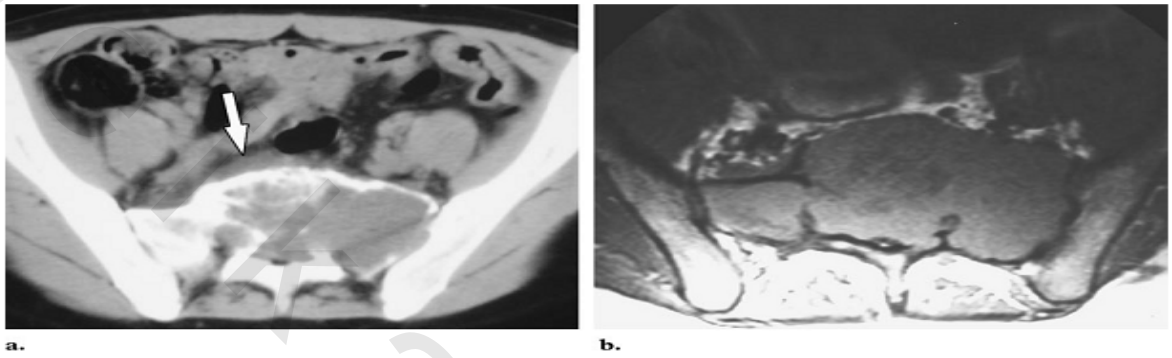


Figure (32): Giant cell tumor in a 15-year-old girl with hip pain.(a)Axial pelvic CT scan obtained with intravenous contrast material shows a large and expansile osteolytic mass (arrow) in the upper sacrum, with involvement of the sacral neural foramina and spinal canal and extension into the presacral space.(b)Axial T1-weighted MR image shows an area of low signal intensity in the mass, a finding that indicates penetration into the bone marrow. The spinal canal is markedly narrowed, and the left sacroiliac joint is irregular.[5]

4-Neurogenic masses

A) Neuroblastoma

Neuroblastic tumors (neuroblastoma, ganglioneuroblastoma, Ganglioneuroma) are the most common extra cranial solid tumors occurring in children**.** They arise from primordial neural crest cells, precursors of the sympathetic nervous system. Although Ganglioneuroma is a localized tumor and tends to be relatively homogeneous, the imaging characteristics of this group of tumors are similar. The histological type cannot be discriminated with imaging but is based on pathological findings only**.**[20, 91]

Neuroblastoma can arise anywhere along the sympathetic chain from the neck through the pelvis. Approximately 70% of neuroblastomas are found in the abdomen (adrenal gland and retroperitoneal area), 20% in the posterior mediastinum, 5% in the neck region, and only 2%–3% in the pelvis. Pelvic lesions are found most commonly in the presacral region, or arising near the distal aorta and its bifurcation where they are referred to as "Organ of Zuckerkandl (O. Z.) Tumors ".These tumors of pelvic origin have a twofold improved survival over adrenal primary tumors even when in an advanced stage at diagnosis and despite of the technical problems in their surgical removal.[1, 5, 97, 98]

The median age at diagnosis of children with neuroblastoma is approximately 19 months and 98% are younger than 10 years of age. The clinical manifestations of neuroblastoma are nonspecific and maybe related to the primary tumor, a paraneoplastic syndrome, or metastases. Biochemical markers are VMA, HMA and dopamine in random urine sample, serum ferritin, LDH and NSE .Several cytogenetic abnormalities have been shown to have a significant impact on the clinical behaviour of the tumor and have been developed into important prognostic markers including MYCN oncogene amplification and aneuploidy.[99, 100]

Metastatic deposits are seen in up to 80% of patient .Metastasis are predominantly to the liver, bone marrow, skin, lymph nodes and cortical bone. Lung deposits are less common. Detection and staging of neuroblastoma can be accomplished with CT, MR imaging, and nuclear medicine. Bone scintigraphy with Technetium-99m and skeletal radiography are used to determine the areas of involvement. MIBG studies are sufficiently sensitive to have become a mainstay for assessment of the entire body, including the skeletal system.[5, 19, 101]

On MR imaging, neuroblastoma is predominantly solid with heterogeneous signal intensity and variable degrees of enhancement. They commonly show low signal intensity on T1-weighted images and high signal intensity on T2-weighted image. Hemorrhagic areas may have high signal intensity on T1-weighted images. Cystic and necrotic areas may be seen as hyper intense on T2-weighted images, while areas of calcification are devoid of signal. Presacral tumors may extend into the spinal canal through the neural foramina and cause erosion and remodeling of bone. MR imaging allows for staging and pre surgical planning, including details regarding tumor size, vessel encasement, and the relationship between the tumor and the sacral plexus and pelvic floor. Iliac vessel encasement and crossing of the sciatic notch by the tumor are important surgical risk factors of respectability.[1, 5, 19]

After therapy, MR imaging can be used to monitor tumor regression and detect recurrence. MRI has become one of the most valuable modalities for imaging and staging of NBL, such that, in many centers, it has replaced the use of CT. MRI is better for visualization of anatomic details of the primary tumor, including relationships with the surrounding viscera and blood vessels .This is helpful particularly in infants whose lack of intra peritoneal fat can make CT inadequate. MRI is also superior to CT for characterizing epidural extension or leptomeningeal disease, for detecting bone marrow invasion (and distinguishing it from cortical bone involvement). Care must be taken in infants and young children to avoid mistaking the diffuse low signal intensity of normal hematopoietic marrow on T1-weighted sequences for metastatic disease. MRI is particularly helpful for evaluating an unexpected MIBG-avid focus in skeleton or soft tissue during follow-up surveillance.[2, 19, 100, 101]

Complementary use of MRI and other metabolic imaging method such as MIBG scintigraphy or PET probably increases diagnostic accuracy and, subsequently, improves clinical outcomes. Whole-body MRI (WBMRI) is an emerging imaging method useful in staging and monitoring NBL. DWI may improve diagnostic accuracy of MRI by differentiating viable tumor and nonviable residual lesion. NBL is known to demonstrate diffusion restriction because of high cellular density.[19, 20, 102, 103]

The staging of neuroblastoma was classically based on the International Neuroblastoma Staging System (INSS) that was developed in 1988 and modified in 1993 and is still used. In 2009, The INRG Task Force developed a new staging system based on tumor imaging rather than extent of surgical resection. As the surgical risk factors are based on radiological images, it was decided to use the term "image-defined risk factors" (IDRFs), which are risk factor that may make total tumor excision risky or difficult at the time at diagnosis. Risk factors that may be found in imaging of pelvic tumors include tumor encasement of aorta, inferior vena cava or iliac vessels, tumor crossing of the sciatic notch or intra-spinal extension (provided that more than one-third of spinal canal in axial plane is invaded, the peri-medullary leptomeningeal spaces are not visible, or the spinal cord signal intensity is abnormal).[19, 20]

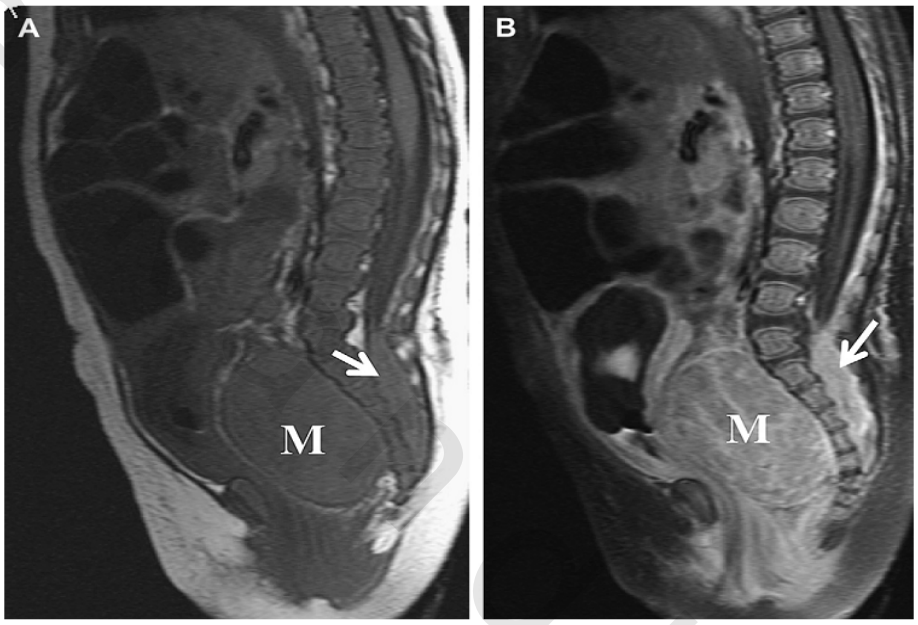


Figure (33): Pelvic neuroblastoma in a 3-month old girl. (A) T1-weighted sagittal image shows a large presacral mass (M) with signal intensity similar to soft tissue. (B) On a fat suppressed T1-weighted image with gadolinium enhancement, the tumor shows heterogeneous enhancement. Panels A and B both show intra spinal tumor extension (arrows).[91]

Descriptions of New INRG Tumor Stages

Tumor Stage	Description
L1	Localized tumor not involving vital structures, as defined by the list of IDRFs, and confined to one body compartment
L2	Local-regional tumor with presence of one or more IDRFs
M	Distant metastatic disease (except stage MS tumor)
MS	Metastatic disease in children younger than 18 months, with metastases confined to skin, liver, and/or bone marrow

Figure (34): The new staging system for neuroblastoma developed by the INRG in 2009.[20]

B) Neurofibroma

Neurofibromas are benign neural tumors that consist of fibroblasts, Schwann cells, and neural elements that expand and diffusely infiltrate a nerve. They may occur singly, or they may occur in multiples as in neurofibromatosis type1, a common autosomal dominant disease that affects one person in 2000 – 4000. Pelvic neurofibromas often follow the distribution of the sciatic and femoral nerves. Large ones may extend into the presacral space through the sciatic notch or femoral canal .Bladder and rectal involvement due to mass effect can be seen, or may present as circumferential or focal infiltration. In NF1 abdominopelvic lesions are usually bilaterally remarkably symmetrical in location, size and extension and of almost identical attenuation and demarcation.[1, 5, 37]

At CT, a neurofibroma may resemble lymphadenopathy and may have attenuation lower than that of soft tissue .On T1-weighted MR images; the lesion appears as an area with homogeneously isointense or mildly hyper intense signal in comparison with the signal intensity of muscle. On T2-weighted images, the lesion has high signal and commonly show a "target like" appearance with a hyper intense rim of myxoid material and a central zone of low signal .This sign is highly specific for benign neurogenic tumors .Relatively homogeneous enhancement is noted on post contrast sequences, although peripheral enhancement may be seen. Malignant peripheral nerve sheath tumor occurs in 2% to 29% of patients with NF1. Imaging findings suggestive of malignant degeneration include rapid growth of existing tumor, loss of typical target sign, development of hemorrhage, heterogeneous enhancement pattern indicating necrosis or asymmetry of size and enhancement of bilateral lesions.[1, 5, 84, 104]

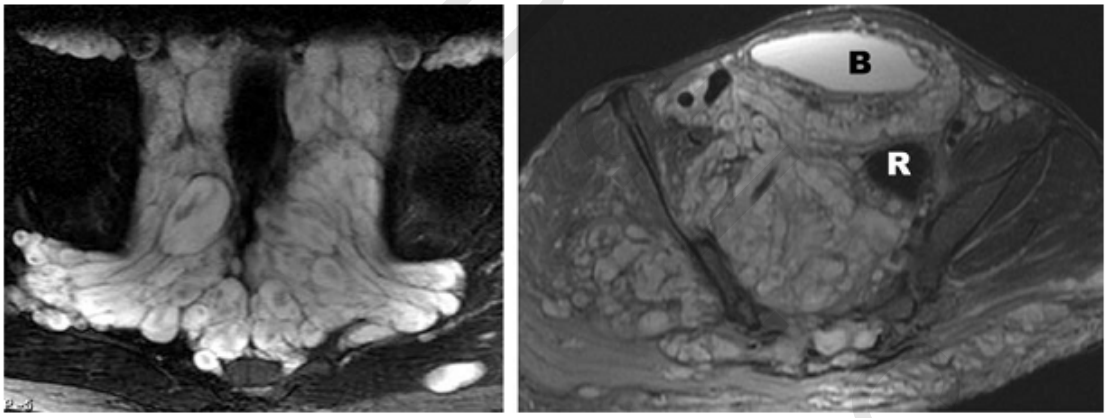


Figure (35): NF1. T2W images with fat suppression in two patients. Left: multiple bilateral target signs indicative of intra-pelvic neurofibromas, which extend into the inguinal regions. A subcutaneous lesion also is visible in the left buttock. Right: multiple lesions also showing the characteristic target sign and abnormal soft tissue has filled and expanded the spinal canal. Evident mass effect on the rectosigmoid (R) and the bladder(B).[5]

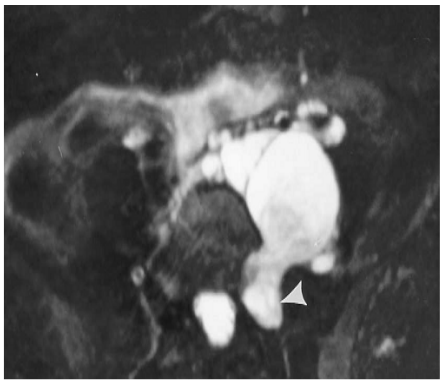


Figure (36): Neurofibromatosis. Axial T2-weighted FSE image shows high signal intensity neurofibromas in the left para spinal area involving the sacral foramen (arrowheads).[2]

II. Non-Neoplastic

1-Developmental cysts

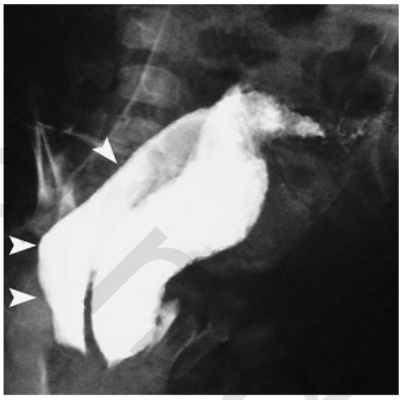
These are the most common congenital condition in the presacral space and include epidermoid, dermoid, enteric and rectal duplication cysts. Dermoid and epidermoid cysts result from the abnormal closure of the ectodermal tube. Epidermoid cysts are unilocular lesions composed only of stratified squamous cells while dermoid cysts also have skin appendages like sweat glands, hair follicles, or sebaceous cysts. Both types may be associated with a posterior anal dimple or sinus when they communicate with the skin. Rectal duplication cysts are diagnosed based on three histological criteria: continuity or contiguity with the rectum, a well defined muscular wall with a myenteric plexus, and a mucosal lining. This lining is usually similar to rectal mucosa, but can sometimes contain ectopic tissue such as gastric mucosa. Rectal duplication cysts may communicate internally with the anorectal lumen or externally with the skin surface.[5, 28, 84]

Patients usually present with symptoms related to mass effect from local compression on the rectum and lower urinary tract. The most important complications of developmental cysts are infection and fistula formation. Dermoid and epidermoid have a high rate of infection (up to 30%) and infected cysts can be easily mistaken for peri-rectal abscess, pilonidal disease, or peri-anal fistula. Bleeding related to ectopic gastric mucosa or mucosal irritation is rare, as is malignant degeneration.[1, 5, 11]

Developmental presacral cysts may cause a widening of the retrorectal space, an effect that is visible on lateral radiographs. Communication between a duplication cyst and the intestinal lumen may be observed during a barium enema study.US shows thin-walled uni- or multilocular cystic masses with internal echoes related to mucoid material or inflammatory debris. US images also may show the “bowel signature” of rectal duplication cysts seen as areas of hypoechogenicity in the musculature of the cyst wall. Developmental cysts show low attenuation at CT, with no associated enhancement, and associated thin calcifications are rare. If they are secondarily infected, they tend to be thick-walled with surrounding inflammatory change, and they may contain air if a fistula is present.[5, 15]

Typically, developmental cysts are thin walled fluid filled rounded lesion hypointense on T1-weighted MR images, (although they may be hyperintense if they contain mucoid material) and hyperintense on T2-weighted images. Duplication cysts are seen closely abutting the rectal wall and maybe seen connected with its lumen. Fat content of dermoid cyst is an important differentiating point form other pelvic cystic lesions, however, it can be sometimes very minimal and is visualized only on fat suppression techniques. Both dermoid and epidermoid cyst show restricted diffusion due to dense keratinoid content. Developing malignancy is exceedingly rare, reported only in adults and presenting as irregular wall thickening with low signal intensity on both T1- and T2-weighted images with heterogeneous contrast enhancement. Associated abnormalities in the spine and sacrum should be borne in mind when assessing presacral masses,that is why MRI is the investigation of choice.[2, 15, 84, 105, 106]

The differential diagnosis of cystic presacral masses in a neonate includes,in addition to developmental cysts, cystic sacrococcygeal teratoma, lymphangioma, anterior sacral meningocele, chordoma, and cystic neuroblastoma.[107, 108]



Figur (37): Rectal duplication cyst in a 6-month-old girl. Oblique radiograph of the pelvis, obtained during a barium enema examination, demonstrates a well-defined retro-rectal tubular communication (arrowheads) with the rectum.[5]

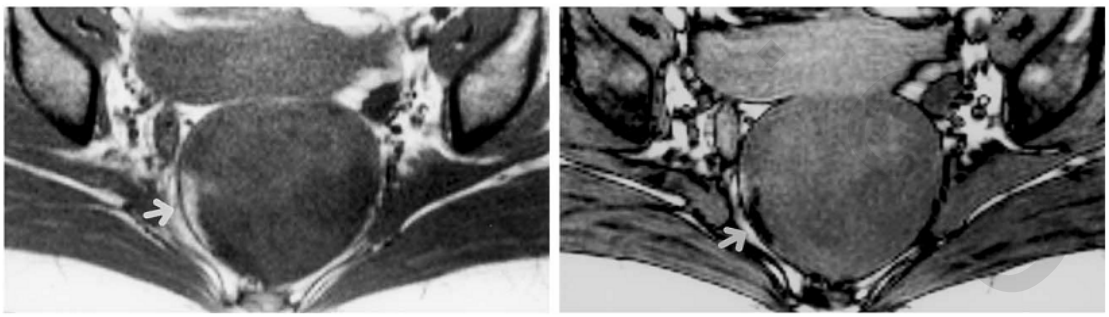


Figure (38): Presacral dermoid cyst.Left:T1-weighted GE in-phase image shows predominantly hypointense presacral mass with small high signal intensity content along the wall (arrow). Right: T1-weighted gradient-echo opposed-phase image shows significant signal reduction of the content along the wall confirming its fat content (arrow). [105]

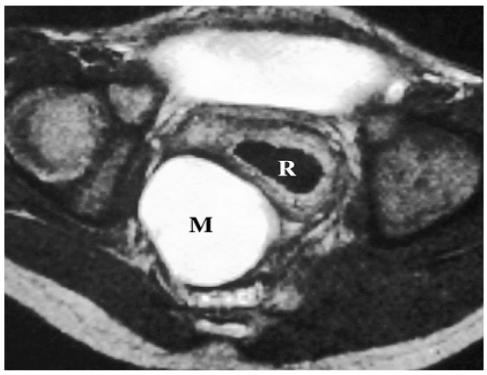


Figure (39): Rectal duplication cyst. Fat-suppressed T2-weighted image shows a high signal intensity mass (M) adjacent to the rectum (R).[91]

2- Anterior sacral meningocele

This is a rare condition (around 1:40.000) resulting from herniation of the dural sac through a congenital defect in the vertebral body. It is most common in the sacral region and lumbosacral junction. It can be located ventrally in the presacral space or ventro-laterally through anenlarged vertebral foramen or coalesced foramina into the pelvic retroperitoneal and infraperitoneal space.When the contents of the herniated sac contain neural elements in addition to meninges and cerebrospinal fluid, the mass is termed a "myelomeningocele"; when fat and cerebrospinal fluid are present, the mass is termed a "lipomeningocele".[2, 5, 13]

Symptoms are usually due to mass effect on pelvic organs and nerve roots, neurologic compromise, meningitis, or rupture of the meningocele.Also maybe back pain, numbness in the lower limbs, and headache. The headache maybe associated with defecation due to a compression-induced increase in CSF pressure .All cases can be felt on digital rectal examinatio as a soft, cystic retrorectal mass that appears to be affixed to the ventral sacral wall.[13, 28]

X ray will often reveal the defect in the sacrum,the pathognomonic "Scimitar sacrum," which is a unilateral sickle-shaped smooth distortion of the sacral bone. Other findings may include widening of isolated sacral foramina, an increase in interpedicular distance ,flattening of the pedicles, and abnormalities of curvature, vertebral scalloping, hypoplasia and aplasia.Pelvic US images in neonates may demonstrate a presacral cystic mass connected with the thecal sac but are not sufficient for surgical planning. Osseous abnormalities are better evaluated with CT, which also can depict the neck of the meningocele.[5, 13]

MR imaging is the modality of choice for the assessment of anterior sacral meningoceles because of its excellent depiction of neural elements. Sacral defects, neck, hernial sac, nerve roots (which appear as areas of intermediate signal intensity in T2-weighted images), and dysraphism are best assessed with MR imaging .The hernial sac show same signal intensity as CSF,homogneously hypointense on T1 and hyperintense on T2 while mixed fluid signals maybe noticed due to fluid movements during imaging.The thickened filmum terminale and any assoicated masses are also well depicted.[1, 13, 84]

An anterior sacral meningocele may be accompanied by other anomalies or syndromes, including uterine, anorectal, renal, and bladder malformations; Marfan syndrome; and type 1 neurofibromatosis.An interesting association with anterior sacral meningocele is the so-called Currarino triad. This association consists of sacral defect, a presacral mass and different forms of anorectal malformation.The syndrome is often familial, with autosomal dominant inheritance.[13, 14, 28, 109]

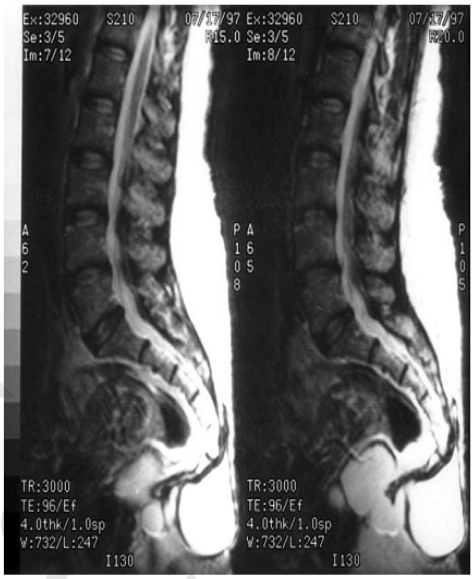


Figure (40): Sagittal T2-weighted MR images of the sacral spine demonstrating a caudal anterior sacral meningocele extendinginto the presacral space. [13]

3- Lymphangioma

Lympangioma(cystic lymphatic malformations/cystic hygroma) are rare benign congenital vascular malformations that result from failure to establish normal lymphatic channel anatomy.Retroperitoneal cystic lymphatic malformations are usually detected incidentally, but they may be complicated by infection, hemorrhage,rupture, or mass effect.US images typically show a sharply marginated uni- or multilocular cystic lesion containing anechoic fluid with enhanced through transmission. Internally increased echogenicity is seen in the presence of infection or hemorrhage.CT scans better demonstrate the anatomic extent of the lesion. On CT scans, lymphangionas are thin-walled, multi-septated cystic mass with homogenous fluid content of attenuation values that may range between those of fat and those of fluid.[5, 110]

MR imaging also can depict the extent of the malformation and is superior to other imaging methods because of its excellent display of soft tissue contrast information. Cyst size determines the MR imaging appearance. Microcystic lesions may appear solid, hypointense on T1, and markedly hyperintense on T2. Enhancement is absent or diffuse, further complicating differentiation from a solid mass. However, the macrocystic form is usually readily identified as clearly defined cysts that are homogenously hypointense on T1 and hyperintense on T2. Different fluid signal intensities and fluid-fluid levels result from hemorrhage or infection. Vascular flow voids do not occur. Septa often enhance, yielding the characteristic ‘‘rings and arcs’’ appearance.[5, 111]

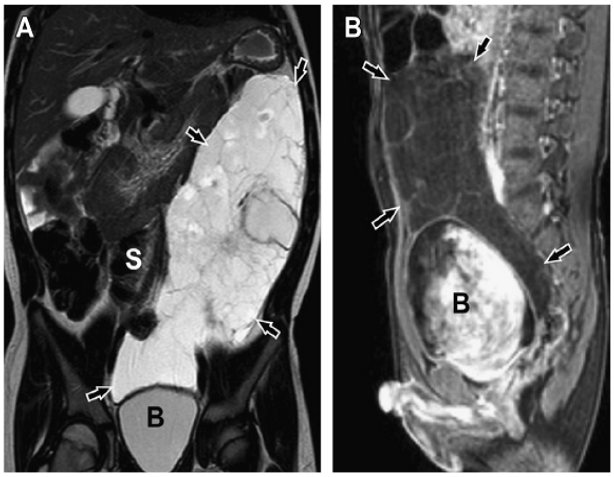


Figure (41): A 3-year-old boy with large abdominopelvic lymphatic malformation. (A) Coronal T2WI demonstrates homogeneous fluid signal intensity with multiple thin septations and significant mass effect on the bowel including the sigmoid colon (S). (B) Sagittal postcontrast fat-saturated T1WI reveals minimal enhancement of the internal septations. Contrast material is seen in the bladder (B).[1]

Other plevic masses:

Lymphoma

Non-Hodgkin lymphoma is seen more frequentlythan is Hodgkin disease in children younger than10 years and generally occurs as an aggressive malignancy in children. Burkitt, Burkitt-like, and B-cell lymphoma are the most common types that occur as a primary abdominal masses. Lymph node involvement may be discrete or may develop as part of a conglomerate mass. If pelvic involvement occurs, it is usually associated with iliac lymph node disease.Symptoms can be systemic or caused by a local mass effect and may include abdominal pain, diarrhea, vomiting, peritonitis, and ascites. Lymphoma often is first assessed with US which commonly demonstrates a solid hypoechoic mass. At CT, lymphomas have soft-tissue attenuation with homogeneous enhancement that is occasionally peripheral. The appearance of post-transplantation lymphoma is indistinguishable from that of lymphoma unrelated to transplantation.[5, 84]

At MR imaging, the mass appears heterogeneous, with low signal intensity on T1W images and high signal intensity on T2W images. Conventional imaging is not sensitive for bone marrow involvement, whereas MR imaging is extremely sensitive. Bone marrow infiltration appears as low signal intensity on T1W images and high signal intensity on T2W images and enhances with contrast.[5, 112]

NHL may involve the ovaries forming large nonspecific unilateral or bilateral cystic, solid, or mixed masses .On MRI the mass is usually of low signal intensity onT1 and enhanced avidly following intravenous gadolinium. On T2, the masses are of intermediate signal intensity, well defined and with high signal intensity lesions in the periphery, consistent with follicles. They show internal enhancing septations. These masses are very rapidly progressive and may be huge engulfing most of pelvic viscera. The

importance of considering lymphoma in the differential diagnosis of adnexal lesions in pediatrics is that the management of such patients is different from that of patients with other types of ovarian tumors, as urgent chemotherapy is the initial treatment of choice.[112-114]

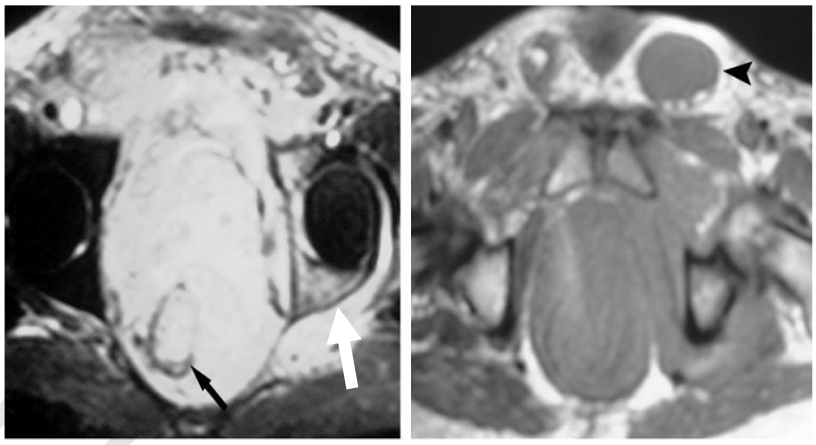


Figure (42): Burkitt's lymphoma .Axial T2 and T1 MR images reveal a large heterogeneous pelvic mass that has surrounded the rectum (black arrow) . Also noted left inguinal lymphadenopathy (arrowhead) and left acetabular involvement(white arrow).[5]

Inflammatory masses

Pelvic inflammatory masses and abscesses in children may be secondary to complicated appendicitis, inflammatory bowel disease, or postoperative complications, or may be related to the extension of a mass from an upper abdominal primary site .Infection is almost always evident clinically by high fever, malaise and elevated leucocutic count.Conventional radiographs may show intralesional air, including air-fluid levels. At US, abscesses may have complex fluid contents; may appear homogeneously echogenic, mimicking a soft-tissue mass; or may have multiple septae. However, CT is more helpful for the detection and localization of an isolated area of infection in the abdomen or pelvis.[5, 115]

MR may be used in some cases if the clinical presentations are misleading, or upon suspicion of underlying mass or osteomyelitis. The central portion of an abscess is iso to hypointense on T1W images and hyperintense on T2W images. Marginal enhancement is common but when irregular may raise concern for the presence of a necrotic tumor. The penumbra sign (an area of relative T1-W hyperintensity positioned between the intermediate or low signal abscess and the adjacent soft tissue edema) is a useful sign that indicates soft tissue infection rather than tumor. The surrounding fatty tissue around abscesses commonly show wide are of inflammatory changes in the form of marked high intensity both on T2WI and T2 SPIR and low intensity on T1.[111, 116, 117]

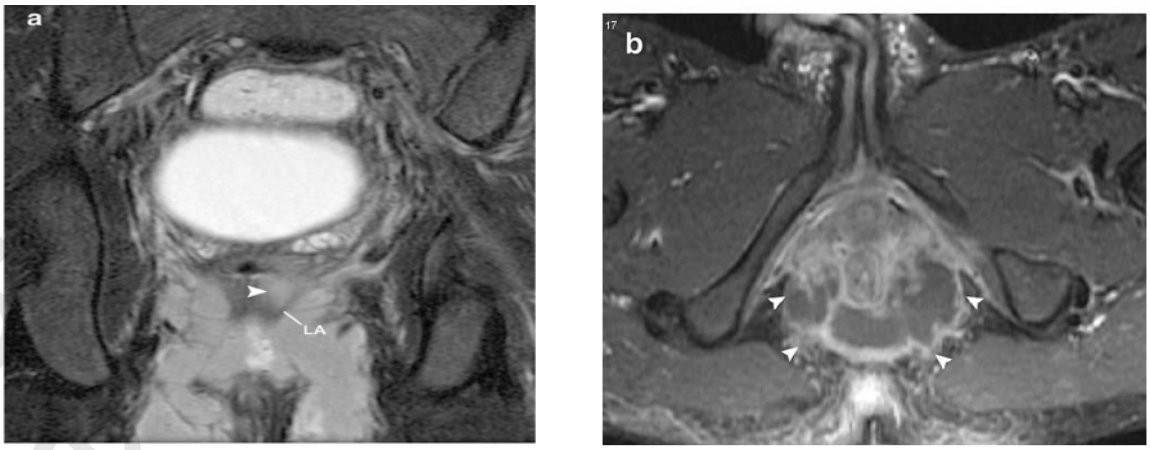


Figure (43): A child with Crohn's disease. Large abscesses throughout the ischio-anal fossa with extension above the levator ani. Coronal T2-weighted (a) and axial post-GAD(b) images show abscesses and inflammation almost completely replacing the normal fat in the bilateral ischio-anal fossa(arrowheads). Levator ani muscle(arrow in a). [118]

Vascular masses

Vascular masses include vascular malformations and vascular neoplasms such as hamangioma. Vascular malformationsthat may occur in the presacral region include lymphatic malformations (described earlier), venous or capillary malformations, and arteriovenous malformations(AVMs), which may occur either alone or in combination.[5]

Venous malformations may primarily arise in the pelvis or extend into the pelvis from the abdomen or lower extremities. Complex combined malformations,such as those seen in Klippel-Trenaunay syndrome, commonly affect the lower limbs, with occasional extension into the perineum or pelvis.Fast spin-echo sequences are used to delineate the anatomy. STIR images provide homogeneous fat suppression to determine the extent of the malformation. T1-weighted images are useful for evaluation of internal hemorrhage and thrombosis. Gradient-echo sequences are especially helpful for evaluation of hemosiderin and calcification or phleboliths within the lesion. Phleboliths are exclusively present in venous malformations. Dynamic postcontrast imaging not only reveals the pattern and comparative timing of arterial and venous enhancement but also provides anatomic information about extent and drainage. Venous malformations fill with contrast material on delayed imaging.[1, 119]

AVMs result from abnormal vessel development which bypasses the capillary bed. They are composed of a focal or diffuse network of interconnecting arteries and veins, with a central nidus of tortuous, dysplastic vessels. Although present at birth, they usually manifest later on. MR imaging identifies high flow vessels as signal voids on spin-echo imaging or as increased signal with gradient-recalled echo. Feeding and draining vessels without a focal soft tissue mass are seen,along with enlarged central vascular channels.Surrounding tissue may demonstrate edema and enhancement, but there is no associated mass. The skin may appear thickened, and bony involvement results in lytic changes with cortical thinning. Treatment is transarterial hyperselective embolization.[111, 120]

The MR imaging appearance of haemangioma varies with lesion phase. During proliferation and plateau, hemagiomas appear isointense to muscle on T1 and hyperintense on T2 where the typical appearance is of a focal, well-marginated, lobulated mass . Margins may be indistinct at T1W. Enhancement is homogeneous and diffuse. Enlarged, high-flow vessels, within and around the mass, demonstrate increased signal on gradient-recalled echo and signal voids on spin-echo sequences. Enlarged veins also are seen. Central low-signal-intensity dots are common, probably representing fibrous septa viewed in cross section, or perhaps thrombosed vascular channels. Areas of increased signal intensity on T1- and T2-weighted images correlate with hemorrhage and areas of fat deposition.With involution,vascularity and enhancement decrease, and with progressive fibrofatty replacement, only a few scattered hyperintense vessels remain.[84, 111, 120, 121]

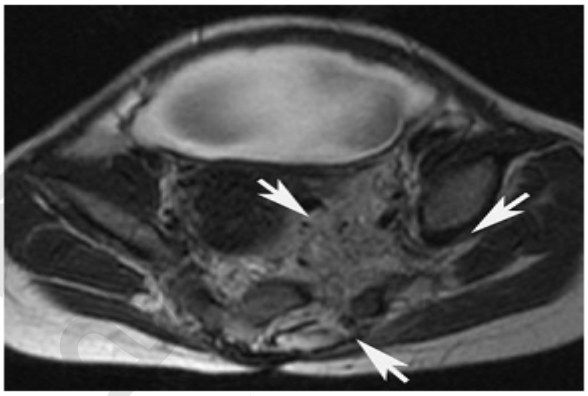


Figure (44): Vascular malformation in an infant girl. Axial T2-weighted MR image demonstrates a mixed signal-intensity mass (arrows) that involves the posterior sacrum as well as the presacral space.[5]

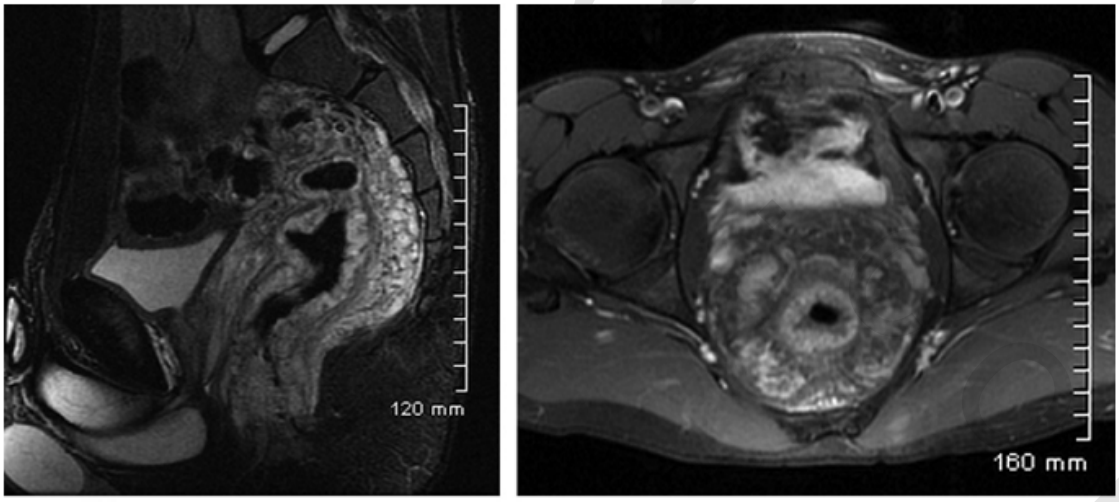


Figure (45): Perirectal hemangioma. (a) Sagittal STIR MR image shows a perirectal mass with heterogeneous high signal intensity. (b) Axial contrast-enhanced T1-weighted MR image shows that the mass demonstrates heterogeneous enhancement. [84]

Desmoplastic small round cell tumous(DSRCT)

This is a relatively rare malignancy of unknown origin belonging to the "small round blue cell " tumor group. It mainly affects adolescent males. The most striking histological feature of DSCRT is the desmoplastic stroma that envelops the tumor cells. This stroma is generally densely collagenous or fibromyxoid and encases well defined nests of primitive undifferentiated cells. The most common presentations are multiple, low-attenuation soft tissue masses in the omentum or mesentery or along abdominopelvic peritoneal surfaces, without a distinct organ of origin. Tumor calcification, liver metastases, abdominal lymphadenopathy, ascites, urinary tract obstruction, and bowel obstruction also have been documented. Some cases show a dominant mass in the retro-vesical or recto-uterine space. FDG-PET/CT imaging can provide useful information on the tumor stage and identify occult lesions that cannot be obtained using CT or MRI .[122, 123]

Abdominopelvic DSRCT often appears as lesions with heterogeneous isointense or hypointense areas on T1, heterogeneous hyperintense on T2, and heterogeneous enhancement on contrast-enhanced images. Some cases may show nodular hypointense lesions on T2W and relatively mild enhancement on contrast enhanced T1W images, which reflect densely desmoplastic tissue. Fluid–fluid levels on T2-weighted MR images and increases in the signal intensity on T1-weighted MR images reflect the presence of hemorrhagic tumor necrosis.[122, 124, 125]

The differential diagnosis for DSRCT includes tumors, often malignant, that produce bulky mesenteric masses, such as rhabdomyosarcoma, lymphoma, neuroblastoma, PNET, mesothelioma, peritoneal leiomyosarcomatosis, and intra abdominal desmoid tumor. A diagnosis of DSRCT usually can be favored by a combination of factors, including adolescent age at presentation; frequent tumor calcification; extensive peritoneal involvement at the time of diagnosis and absence of definite organ of origin.[123]

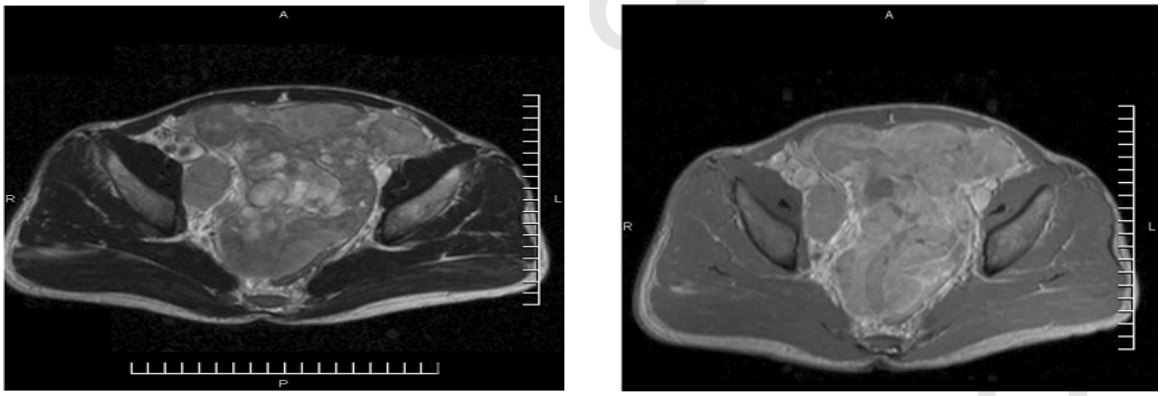


Figure (46): Abdominopelvic desmoplastic small round cell tumor. Left:The dominant tumor shows mixed signal on axial T2-weighted image. Right: The dominant tumor shows mildly heterogeneous enhancement on contrast-enhanced axial T1-weighted image. [122]

Currarino triad

Currarino triad is a form of caudal regression syndrome. It is also known as ASP triad (anorectal malformation, sacrococcygeal osseous defect, and presacral mass).This is a rare syndrome characterized by autosomal dominant genetic inheritance in more than 50% of cases. It consists of an anorectal malformation (anal atresia, anorectal stenosis, anal ectopia, recto-urethral fistula, recto-vaginal fistula), sacral bone abnormality (commonly "sickle-shaped" deformity) and a presacral mass (teratoma, anterior sacral meningocele, dermoid cyst, hamartoma, enteric duplication cyst, or more than one type of these masses). Incomplete forms of this syndrome exist, especially in relatives of patients with ASP syndrome.[5, 14, 126]

These patients present with constipation of variable severity (which may result from a combination of compression by the mass, neurogenic dysfunction and anal stenosis itself), recurrent UTI or meningitis. There are also other anomalies which may be associated with the triad such as duplication of the urogenital tract, tethered cord, and different types of fistulas.[109, 127]

Early recognition of a presacral mass in CS is important to prevent complications such as meningitis, neurological impairment, and malignant degeneration of a presacral teratoma. Among the diagnostic imaging modalities, MRI is superior to other tools in evaluating the presacral mass and detecting possible associated intraspinal abnormalities. It has been suggested that screening pelvic radiograph for all first degree relatives and genetic counseling for family members of CS is mandatory and if any family members have a sacral anomaly, then they should have further imaging, preferably MRI, to identify any other features of this syndrome.[14, 109]

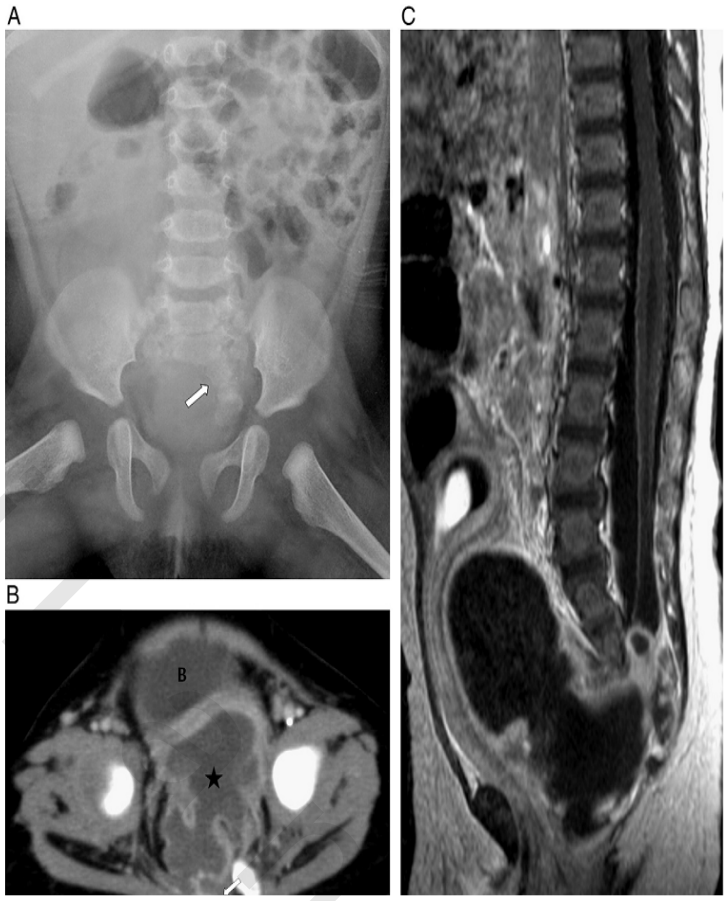


Figure (47): Currariona syndrome.A 6 month gold girl with long history of unexplained fever and gradually developing abdominal distension.(A) Plain radiograph shows a typical appearance of sacrococcygeal bony scimitar-like defect (arrow). (B) Postcontrast CT demonstrates a multilobulated presacral mass (★) with enhancing thick wall which extends into the spinal canal through the sacral bone defect (arrow). Note urinary bladder (B) displaced anteriorly. (C) GAD-enhanced T1W sagittal image shows prominent enhancement at the periphery of the mass and along the surface of the spinal cord which is low lying with syrinx. This suggested infected presacral meningocele associated with arachnoiditis. Bacterial meningitis was shortly confirmed but meanwhile the condition rapidly deteriorated with acute cerebral infarction, septic shock and the child succumbed to brain death.[109]