DISCUSSION

Pediatric pelvis masses can arise from a variety of pelvic structures, including the ovaries, lower genitourinary tract, musculature, neurovascular structures, lymph nodes, and adjacent osseous structures. Ultrasound is almost always the initial imaging modality to assess children with suspected or known pelvic mass. CT is sometimes used for initial diagnosis in children presenting with abdominal pain or large palpable masses. In cases of neoplastic masses, CT is traditionally used for staging and follow-up. MR imaging is sometimes used for further evaluation and lesion characterization. [1, 10]

Compared to other imaging modalities available, the advantages of CT include wide availability, good anatomical detail with fine spatial resolution which can be reformatted to provide multi-planar and 3D reconstructions, simultaneous examination of the lungs in malignancy and superior identification of calcification. An important advantage, compared to MRI, is the relatively quick acquisition time allowing examination with light sedation or no sedation at all, thus avoiding the need for a general anesthesia in many cases. Its main concern in pediatrics is the risk of ionizing radiation where a significant gonadal dose is unavoidable in pelvic exams. [128]

Over the past several years, increasing attention has been focused on the potential for radiation exposure from CT for inducing the development of cancers, including secondary malignant neoplasms in children. The Department of Health and Human Services in the US listed ionizing radiation as a known human carcinogen in 2005. Multiple recent reports have showed that excess solid cancer risks appear to be linear in dose even for equivalent doses in the usual range of 0–150 mSv; with no evidence for a threshold and that the smallest dose has the potential to cause a small increase in risk to humans. It is therefore clear that the carcinogenic potential of radiation at doses encountered in CT needs to be taken seriously by the medical imaging community specially when dealing with children who are inherently more radiosensitive than adults, and becausethey have more remaining years of life during which a radiation-induced cancer could develop.[128, 129]

Until recent times, MRI has been considered as an alternative modality to CT investigation in pediatrics in selected situations. However, cumulative experience has demonstrated that MRI can substitute CT for many reasons ,maybe the most important of which is that it avoids the significant health risk associated with a radiation dose from a CT procedure, more importantly in children surviving cancer who need life-long serial assessments in which the doses will become additive. In addition, MRI provides inherently superior anatomic details and soft tissue contrast and more different types of tissues can be interrogated during a single MRI examination. Also greater patient safety is achieved with the gadolinium based contrast agents used in MRI compared to the iodine-based contrast agents used in CT.[129, 130]

The key issues that have served as impediments to a widespread preference for MRI over CT for pediatrics in the medical community include the higher health care costs ,the less availability , the relatively longer time of examination that necessitated anesthesia for most young uncooperative children and lastly the ordering practices of referring physicians.[129]

The actual costs of MRI equipment, service and exams have decreased significantly over the past 20 years and are expected to continue doing so. More available scanners and radiologists experienced in MRI interpretations are found nowadays. The technical advancements including the 3.0 Tesla magnets, new sequences, multichannel coils and parallel imaging allowed for increased resolution and decreased image acquisition time. However, changes in practice, such as eliminating inappropriate referrals for CT, will exert the most dramatic impact on avoiding the hazards of radiation but will be the most challenging to implement. [129]

MR evaluation of pediatric pelvic masses is advantageous, given its superb anatomic details, excellent soft tissue contrast resolution, and ability to specifically characterize certain types of tissues. Contrast enhanced MR imaging assists in mass characterization, pre-surgical planning, and staging when a malignancy is suspected.[1]

The use of MRI has lagged behind the use of CT in pediatric abdominopelvic oncology because of a variety of factors, including issues related to cancer research protocols, concerns about the sensitivity of MR imaging, and a general lack of awareness among clinical providers as to the feasibility and advantages of current MR imaging techniques. However, nowadays these concepts are changed and many ongoing oncologic clinical trials now incorporate MR imaging in their protocols as an alternative to CT.[131]

Our study was conducted on 20 children presenting clinically with known or suspected pelvic mass with the aim to evaluate the role of MRI in the assessment and diagnosis of these masses. The study included 5 males and 15 females and their age ranged between 2 and 17 years old. The examined pelvic masses included 12 neoplastic and 8 non neoplastic masses of different origins.

The cases with neoplastic masses included two with benign tumors (mature ovarian teratoma and presacral ganglioneuroma) and 10 with malignant tumors (prostatic rhabdomyosarcoma, cervical rhabdomyosarcoma, 4 cases with yolk sac tumor, spindle cell sarcoma, presacral chordoma, pelvi-abdominal Burkitt's lymphoma and recurrent neuroblastoma).

Eight non neoplastic masses were found and included presacral dermoid cyst, pelvi-abdominal lymphangioma, complicated ovarian cyst, and ovarian torsion with secondary necrosis and four cases with hemato (metro) colpos.

In our study, we used axial and sagittal T1 SE, axial and sagittal T2 SE images, Coronal STIR, Axial T1SE with fat saturation before and after IV gadolinium contrast injection in the three orthogonal planes in addition to DWI.

Various protocols for pediatric pelvic MRI are recommended by several authors with some differences in the used sequences, parameters and selected planes. [1, 131, 132].

Marilyn Siegel discussed important points that should be taken into consideration in any MRI examination of the pelvis in pediatrics.[2, 16] Any exam should include both T1 and T2W images. T1W sequences show the contrast differences between soft tissue structures and fat; thus, they provide tissue characterization (i.e., fluid, fat, or blood). They are also useful for detection of lymph node enlargement. T2W images provide superior soft tissue contrast and are particularly useful for showing the presence of necrosis and for

evaluating soft tissue tumor interfaces. Fat-saturated images are useful to differentiate hemorrhagic from fat-containing lesions. Two basic methods of fat suppression are widely available: short-tau inversion-recovery (STIR) and radiofrequency pre-saturation of the lipid peak (fat saturation). Signal from fat is nulled on such techniques whereas pathological lesions are still bright. STIR images in the coronal plane should be obtained from the symphysis pubis to the renal hila to allow assessment of pelvic and retroperitoneal lymph nodes and hydronephrosis. If any abnormalities are seen complementary T1, T2 or MRU sequences are performed.

The gradient-recall-echo technique results in high signal in flowing blood and is used to evaluate the patency of a blood vessel and to differentiate between vessels and lymph nodes. Arterial and venous phases can be acquired and reformatted in a 3D display which can help to delineate vascular anatomy essential for surgical planning. Contrast enhanced T1W images can help in lesion characterization by better depicting the architecture of tumors and improving contrast between tumor and normal tissues. Images should be taken in at least two or three orthogonal planes: coronal, sagittal, and axial. Slice thickness varies with patient size and the area of interest. A slice thickness of 4 to 8 mm obtained at 5- to 10-mm intervals is usually adequate for a general survey of the pelvis and larger lesions. Thinner slices obtained at 4 mm are particularly useful for evaluating small lesions and for the initial staging of pelvic malignancies.[2, 16]

The receiver coils are very important in optimizing the S/N ratio because they minimize the noise coming from non imaged parts of the body. The smallest possible coil that covers the body part of interest should be used in pediatric MRI. Surface coils designed for specific uses in adults can be adapted to the smaller bodies of children. The head coil can be used to image the abdomen and pelvis of infants. Phased-array coils are usually needed to examine larger children or body parts. They are better than body coils as they allow a larger FOV and a high S/N ratio.[2, 133]

Motion reduction is an important challenge in imaging patients with MRI considering the relatively long examination time and the sensitivity of images to motion artifacts compared to CT. Kassa Darge et al [134] recommended a number of strategies that can help to successfully image the breathing child and reduce motion degradation. The first strategy is to use fast sequences such that good images are obtainable without respiratory motion artifacts such as turbo spin echo, GRE, RARE and trueFISP. Another strategy is to decrease the signal intensity of moving structures which can be done by fat suppression of the abdominal wall fat and placement of a spatial saturation band on the anterior abdominal wall. The third strategy is to use sequences or additions to pulse sequences that reduce image motion. Respiratory triggering synchronizes the signal acquisition to a constant phase of respiration (with the use of abdominal bellows or navigator triggered techniques).

The physical appearance of the MRI scanner and the noises produced during imaging may also be frightening to some children, making cooperation even less likely. To this end, the use of ear plugs, noise-cancelling headphones, and distraction devices may be helpful

The use of sedation is common place in young children undergoing abdominopelvic MRI for the assessment of suspected or known malignancy, because of several factors. Most young children are unable to remain sufficiently motionless for MRI, because imaging times are commonly in the 30 to 60 minute range (This is the time needed for a

complete abdomen and pelvis exam. However localized MRI to the pelvic region, as carried in our study, lasts about 20-30 minutes). In addition, young children often have difficultly cooperating with breath-holding instructions.[131]

In our study we used the same steps for child sedation as described by Sudha Anupindi[133]. For any sedation procedure, children should be NPO for at least 4 hours prior to the examination. A thorough assessment including patient's history, baseline vital signs, weight, airway examination and a cardiopulmonary examination in order to recognize potential factors that may place the patient at increased risk for complications. Oral chloral hydrate is the most common sedative in patients younger than 1 year of age. The dose is 75 mg/kg, not to exceed 2000 mg. In children from 1 to 6 years of age, pentobarbital sodium is the drug of choice. It is given intravenously with an initial dose of 2 mg/kg. The dose can be repeated if necessary up to a maximum dose of 6 mg/kg or 200 mg (whichever is smaller).

DWI uses the motion of water molecules to provide image contrast and to characterize tissues. It has been evaluated for many years as an oncologic imaging biomarker for distinguishing benign from malignant lesions and for evaluating response to therapy, including in the setting of pediatric abdominopelvic malignancy. Its specificity in this role, however, is still controversial. The increased cellularity of many malignant tumors compared with normal tissues impedes the motion of water and presents as restricted diffusion (signal hyperintensity on DWI). The degree of restricted diffusion can be quantified by performing DWI with multiple b values and calculating a map of apparent diffusion coefficient (ADC) values. The number of b values required (e.g., 2, 3, 4 or more) as well as their absolute values (0 to greater than 1000 m/s) have yet to be established for accurate, reproducible ADC calculation in pediatric oncologic imaging. [131]

In our cases, all neoplastic lesions showed different degrees of restricted diffusion while none of the non neoplastic lesions showed any restriction. Milder restriction was noted in the two benign tumors and the lowest ADC values were noted in the case of prostatic RMS.

Ethan A. Smith[131] reviewed the studies assessing the usefulness of DWI in monitoring response to treatment in pediatric neoplasms by analyzing ADC values before and after completion of chemotherapy. The ADC values of tumors changed following chemotherapy, with an increase in the median ADC values (meaning less impeded diffusion and less hyperintense signal on DWI images) seen in most lesions which was thought to represent decreased cellularity of the tumor, thus representing treatment response, as found in a study by MacDonald et al.[135]

Imaging features of selected pelvic masses:

Neoplastic masses:

I Benign

Ovarian mature teratoma

We presented a 12 year old girl with heterogeneous predominantly solid adnexal mass lesion with areas of cystic break down, small spots fat and calcifications. This was associated with marked ascites and two small peritoneal nodules similar to the adnexal lesion. The fatty foci were subtle that they were appreciated only after fat suppression. The solid appearance together with the ascites and peritoneal nodules suggested malignant immature teratoma. However, the pathological analysis of the excised mass and peritoneal nodules showed only mature tissues with predominance of neural elements.

Germ cell tumors include teratoma (mature, immature, and monodermal), dysgerminoma and endodermal sinus tumor (yolk sac tumors) among others. Mature cystic teratoma is the most common (90%) type and also the only benign one. It contains derivatives of at least 2 of the 3 germ cell layers (ectoderm, mesoderm, and endoderm). In the majority of cases it appears as a cystic lesion with variable amounts of fat, fluid, calcifications, hair and other tissues. They are known for variable appearance on different imaging modalities because of their variable contents, however many imaging features are extensively described in the literature and became very specific, sometimes diagnostic for this lesion. The most important of these is the presence of intralesional fat. This can fill the whole lesion, appear in only one part or even be very subtle like a small focus in the cyst wall. Many authors stressed the importance of MRI fat suppression techniques, as chemical fat saturation and Gradient in-phase/out-of phase techniques, to document the presence of fat and thus elucidate the diagnosis. [1, 10, 16, 136]

The solid type of mature teratoma found in our case is a very rare occurrence as mentioned by Eric K. Outwater et al [61]. This type of tumor is benign and contains only mature elements, unlike immature teratoma, although both are radiologically almost indistinguishable and occur in a similar age group. The correct diagnosis however can be suggested prior to pathological result as the visualization of intralesional fat differentiates the mass from other types of solid ovarian tumors and the absences of elevated AFP suggest its mature nature. [137]

Few case reports describing findings similar to our case have been published. Marian Wisniewski and Ludwig Deppisc [138] described two cases of solid mature ovarian teratoma, one had also ascites, and both had long term free survival after only surgical removal. Satoshi Kawakami et al[137] described MR features of a 19 yr old woman with a huge solid mature teratoma ,with no ascites or peritoneal implants. Apart from the huge size, the features were very similar to our cases. They described a predominantly solid lesion with small cystic component. Solid component had homogeneous hypointense signal on T1, relatively hyperintense signal on T2 and heterogeneous reticular pattern of contrast enhancement. The cystic component exhibited mixed signal intensities on both T1 and T2 images, which combined with Fat-Sat image, proved mixed fat and high proteinaceous fluid.

The uncommon association of ascites and peritoneal nodules with a totally benign mature teratoma in our case was also described in some previous reports. The reported cases had extensive peritoneal implants, unlike being limited as in our case, while they were of similar predominantly mature neural elements. Two adolescent girls with sizable mixed solid and cystic adnexal mass associated with multiple peritoneal and omental nodules were described by M. El Shafie [139] and Smiti Nanda et al [140] .One of them had also moderate ascites. Both cases had long free survival after only surgical removal of the adnexal mass and all found nodules. Pathological analysis revealed benign mature teratoma and peritoneal implants were all of benign mature tissue predominantly of neural type.

Annette M. Muller et al [141] have reviewed cases of implantation of miliary glial tissue within the peritoneal cavity in patients with ovarian teratomas which has been referred to as gliomatosis peritonei. They reviewed 86 published cases of this rare entity where the ovarian teratomas were of different grades of maturation, but a mature type was very common. Most cases with this association have very good prognosis and survive without adjuvant radio- or chemotherapy. This was the case in our patient who was totally free at about 6 months follow up after her surgery

All reports of similar cases stressed the importance of complete removal and multiple sectioning of solid ovarian tumors, together with any associated peritoneal nodules, to detect any small foci of embryonal immature or malignant tissue which would definitely alter the prognosis as those cases commonly associated with recurrence and poor survival. [137, 140]

Ganglioneuroma

A 2 year old girl had a sizable presacral mass lesion with homogenous low signal in T1 and mildly hyperintense signal in T2. It showed homogenous intense contrast enhancement and mild restricted diffusion. It had smooth borders with clear surrounding fat planes, closely abutting the sacrum yet with no bone or foraminal involvement.

Ganglioneuromas are rare benign tumors of the peripheral nervous system which are derived from the paravertebral sympathetic chain of the posterior mediastinum and retroperitoneum. It is a benign tumor with mature ganglion cells. Imaging features may overlap with those of neuroblastoma and ganglioneuroblastoma, both considered malignant. Patients are commonly asymptomatic as tumors grow slowly and hormonally inactive.[5, 142]

Axel Scherer et al described the imaging appearance of ganglioneuroma on different imaging modalities. The tumors are commonly round or oval with sharply defined margins and relatively large size (ranged in their study from 3 to 11 cm).They were slightly hypodense on unenhanced CT with few discrete punctate calcifications (in contrast to the coarse ones commonly seen in neuroblastoma).Contrast enhancement was slight or inhomogneously moderate.[142]

On MRI,ganlgioneuromas are typically iso or slighlty hypointense compared to the muscles on T1 and intensely hyperintense on T2.Intense contrast enhancement was noted in most cases,more than on CT studies and more in delayed sequences of dynamic studies.Intraspinal extension is rare in ganlgioneuroma but well described,with widening of neural foramina and strict extradural location.This was not present in our patient where no intra formainl extension was noted despite the close abuttment of the mass to the anterior

surface of the sacrum. MRI is recommended for imaging rather than CT for better demonstration of this type of extension. [142, 143]

II Malignant masses:

Cervical Rhabdomyosarcoma

A 4 year old girl presenting by prolapsing mass in the introitus and minimal vaginal bleeding was found to have a large solid mass totally filling and expanding the vagina and uterus showing mainly hypointense signal in T1, hyperintense signal in T2, intense heterogeneous contrast enhancement and restricted diffusion.

The clinical data and imaging findings in our case were classical for cervical rhabdomyosarcoma as described before by Ricki U. Shah et al and Deepa R. Pai. [1, 3]. They mentioned that on MR imaging, these tumors demonstrate intermediate to high signal intensity on T2W sequences and low to intermediate signal intensity on T1W sequences(usually higher than the muscles but lower that the fat). Contrast enhancement is variable depending on the degree of tumor necrosis (with necrotic portions of the tumor not enhancing).

Rhabdomyosarcoma is the most commonly encountered pediatric vaginal and cervical neoplasm. It is usually found in the vagina of infants and children younger than 2 years. Clinically these tumors may present with vaginal bleeding or as a polypoid mass protruding from the vaginal introitus. Uterine rhabdomyosarcoma is rare, with peak incidence in the second decade of life. Histological subtypes include embryonal (the commonest subtype), alveolar and undifferentiated. Sarcoma botryoides is a polypoid subtype of embryonal rhabdomyosarcoma that arises from the submucosa and is seen in a large percentage of cases (which was the type found in our case) .This subtype has the best prognosis, with a 5-year survival rate of 69%. [1, 3]

Prostatic rhabdomyosarcoma

An 11 year old boy diagnosed with prostatic rhabdomyosarcoma had a heterogeneous lesion at the prostate and bladder base with hypointense signal in T1 and hypo to intermediate signal in T2 .It showed moderate heterogenous contrast enhancement and restricted diffusion. Other few small pelvic nodules were noted showing same MR features.

Bladder-prostate RMS has a bimodal age of distribution with the first peak occurs between 2 and 6 years, and the second occurs between 14 and 18 years. It commonly arises in the bladder trigone or neck. Cases of primary prostatic RMS may occur in boys. The appearance on MR imaging, CT, and US is highly variable, as one can see hemorrhagic, cystic, and solid components. A common imaging appearance described by Marilyn J. Siegel et al [2]and Deepa R. Pai et al [1] is of a polypoid intra luminal mass resembling a cluster of grapes with nonspecific low to intermediate signal intensity on T1 and intermediate to high signal intensity on T2 images with heterogeneous contrast enhancement. For tumors arising from the bladder, T2-weighted images provide excellent contrast between the fluid-filled bladder lumen, hypointense bladder wall, prostate, and surrounding soft tissues. Post contrast dynamic images, acquired before excreted contrast reaches the bladder, are very useful in assessment of tumor enhancement.

Careful staging and follow up imaging is important for the prognosis and treatment of pelvic RMS. Recent evidence supports the usefulness of MRI as stated by Rick R. Van Rijn et al[144] and Deepa R. Pai[1]. MR images are better than CT in delineation of tumor extension. MRI sagittal images may distinguish prostatic RMS that extends into the space of Retzius from tumors invading the bladder. MR is the imaging modality of choice to evaluate for local invasion as well as regional lymph node enlargement. DWI may also assist in the detection of metastatic lymph nodes. In a study assessing the role of MRI in the diagnosis and follow up of pediatric pelvic RMS by Anthony Finelli [9], MRI detected all lesions shown by CT. On the other hand, MRI detected residual disease in 1 case that was missed by CT which enabled appropriate earlier management. In 2 other patients, MRI was superior to CT for delineating the local extent of disease, especially urethral involvement.

Yolk sac tumor

One case had a right adnexal yolk sac tumor appearing as mixed solid and cystic lesion showing low T1 and intermediate to high T2 signal, with moderate diffuse contrast enhancement. This solid component also exhibits type 2 DCE curve and diffusion restriction. Small extra ovarian extension was noted through a suspected focal capsular tear together with pelvic omental stranding and mild pelvic ascites.

Three other cases had extra ovarian tumors: two cases in the form of multiple pelvic nodules and one with infiltrative perineal mass associated with bilateral multiple inguinal nodal deposits and one lumbar vertebral deposit. All cases showed hyperintense signal in T2, strong contrast enhancement and evident restricted diffusion. The signal on T1 images was hypointense in 2 cases and hyperintense in one case.

Yolk sac tumor (endodermal sinus tumor) is one of malignant germ cell tumors with reported very poor prognosis. Ricki U. Shah et al [3] mentioned that these are rare tumors in the pediatric population and the average age at presentation is between 18 and 19 years. P. Dallenbach [145] also mentioned a median age at presentation of 18 to 25 years. This is different from our findings where the age in our cases ranged from 4 to 13 years. He mentioned that serum AFP is a useful marker for diagnosis and management of YST, which is used to check for complete remission or recurrence. This tumor grows rapidly and has a poor prognosis, one of the worst in the group of germ cell tumors.[64]

MR imaging features of yolk sac tumor were described in many reports [64, 84, 93, 145], however none of these were specific for this kind of tumors. YST are commonly large at presentation with ill defined margins and common infiltration into the related peritoneum, viscera and spinal canal. Large tumors commonly have a cystic component and areas of hemorrhage and necrosis are frequent. Hyperintense foci at T1 images are commonly caused by hemorrhage and signal voids maybe seen due to hypervascularity. The sign of T1 hyperintensity was noted in two of our cases: in the pelvic nodules in one case and in the metastatic inguinal nodes in another case.YST demonstrate heterogeneous contrast enhancement which is typically higher than the myometrium (which was also evident in our ovarian case).

P. Dallenbach[145] concluded that no specific imaging findings have been described to distinguish YST from other ovarian masses preoperatively. In case of ovarian location of YST, where germ cell tumors are the commonest encountered neoplasms in pediatric age

group, Seung Eun Jung[64]pointed to the other important differential which is dysgerminoma. Both are malignant germ cell tumors presenting as large, predominantly solid masses. Dysgerminoma may have peculiar appearance of multilobulated lesion with prominent fibrovascular septa and speckled calcifications.

Spindle cell sarcoma

A 17 year old girl had a sizable heterogenous destructive soft tissue mass involving the left iliac bone with infiltration of the related muscles and forming a large pelvic soft tissue component. It showed heterogenous MR signal mainly hypointense in T1 and hyperintense in T2 with moderate peripheral contrast enhancement with a large central necrotic area.

Only 1% to 6% of soft tissue masses in children are malignant. Of the malignant tumors in the 0- to 5-year age group, fibrosarcoma is most common, followed by rhabdomyosarcoma. In the older age group, 6 to 15 years old, malignant fibrous histiocytoma is most common, followed by synovial sarcoma and rhabdomyosarcoma.[111] Spindle cell sarcoma is a wide group of sarcomas of monomorphic spindle cell type with different clinical and morphological features and include some variants of synovial sarcomas, leiomyosarcoma, fibromyxoid sarcomas and malignant peripheral nerve sheath tumors.[146]

Rebecca Stein-Wexler [111]explained that in approaching a child who has a soft tissue mass, clinical history and physical examination play a critical role in diagnosis, and patient age also can narrow the differential. The typical presenting complaint is a palpable mass; larger masses are more likely to present with pain. Many benign and malignant soft tissue masses have common imaging characteristics. Many lesions are iso- or hypointense to muscle on T1 and hyperintense on T2 images. Although some investigators have found MR imaging capable of determining the benignity of specific lesions, accuracy often is uncertain, necessitating biopsy.

Benign lesions are more likely to appear well-circumscribed, homogeneous on T2W imaging, and with no surrounding edema. Enhancement characteristics may facilitate diagnosis of malignant lesions, which may show less rapid enhancement and more rapid washout. Even when MR fails to differentiate between benignity and malignancy, it does provide essential information, delineating the extent of the lesion, extension beyond facial planes, and involvement of adjacent structures, such as the neurovascular bundle, joints, and bone. It is useful in assessing response to therapy, although differentiation of residual or recurrent tumor from postoperative edema, hemorrhage, or inflammation often is difficult. If high T2W signal is present without mass effect, differential considerations include seroma, hematoma, post-radiation change, packing material, fat necrosis, or hygroma. Mass effect increases concern for tumor recurrence as does contrast enhancement. Contrast is also helpful for defining the cystic and solid nature of the lesion and for locating site of viable tissue for biopsy. GE imaging may help assess the presence of high flow, which assists in differentiation of vascular lesions. [111, 147]

M. van Vliet et al [147] mentioned that initial assessment of the soft tissue tumors should be done with imaging before biopsy as biopsy tracts may produce artifacts, but more importantly because imaging features can be used to choose the optimal biopsy location (e.g., enhancing area of a large tumor) and the optimal biopsy route to prevent

spreading of tumor cells through multiple compartments. They stated that MRI is the imaging modality of choice for such lesions.

Presacral chordoma

A 7 year old boy presented by a sizable presacral soft tissue mass lesion with small intra-osseous, intra-thecal and retro-sacral components. It showed isointense signal in T1, hyperintense signal in T2, moderate contrast enhancement and restricted diffusion.

Chordoma arises from notochordal rests and are limited to the clivus, spine, and sacrum on the basis of their cell of origin .It is the most common primary malignant sacral neoplasm but it accounts for only 2%– 4% of primary bone tumors. Occurrence in children is very rare as it is most commonly are manifested in the fourth through the seventh decades of life. Because they are slowly growing, patients typically have nonspecific symptoms, including low back and sacral pain, retention of urine, and radicular pain. Given this subtle manifestation, patients commonly present with a large, bulky mass. [5, 84].

Kendra S. Hain et al [84] described common features of sacral chordomas, most of which are non specific. These include a destructive lytic lesion that may extend across the sacroiliac joints with calcifications commonly seen in X-rays and CT. Usually, a large presacral soft-tissue component is present, with soft tissues extending into the sacral canal. Chordoma has low to intermediate signal intensity on T1 and high signal intensity on T2 images, with variable, often only moderate, contrast enhancement.

These non specific features similar to our case led to our radiological diagnosis of a peri-sacral malignant mesenchymal neoplastic lesion. Chordoma was suggested based on the anatomical location, almost midline position, destructive nature and combination of bony and soft tissue lesions.

Differential diagnosis for a similar large destructive sacral lesion includes sarcomas, giant cell tumor, ependymoma or a metastatic tumor.[94]

Burkitt's lymphoma

A 15-year-old girl had multiple pelvic and abdominal masses with hypointense signal in both T1 and T2, mild contrast enhancement and restricted diffusion. The largest of these were the two hugely enlarged ovaries showing peripheral T2 hyperintense follicles. This was associated with mild ascites and enlarged lymph nodes. Core biopsy revealed the diagnosis and follow up MRI was done after 4 months to assess response to chemotherapy. It showed remarkable response with resolution of all masses except for single residual mass on the left side of the pelvis. The residual mass has become homogeneously hypointense in both T1 and T2 with no restricted diffusion and only minimal contrast enhancement .These features suggested residual fibrotic adnexal mass. This was confirmed on PET/CT done 1 week later that showed no residual metabolic activity.

The MRI features of the infiltrated ovaries in our case were the same as those described by J CRAWSHAW et al. [114].However their case report was of a 28 years old adult female who was found to have primary ovarian NHL with no other lymphomatous masses except for few regional nodes. Also the same features were found by Ferrozzi et al[148]who reported 5 cases of ovarian NHL in the adults. The MRI findings included in

both studies solid bilateral masses, which were low signal intensity on T1, mildly high signal intensity on T2W images and showed homogeneous mild to moderate contrast enhancement.

Akihuto Mitsomori [149]described ,in a rather old case report, the MR appearance of ovarian lymphoma in a 12 year old girl presenting with dysuria and a palpable large abdominal mass. One ovary was enlarged appearing as solid mass with T2 intermediate signal intensity and hyperintense septal structures that showed marked enhancement on GAD injection .At the periphery of the tumor, several small cysts with thin walls were seen in a linear arrangement which were found on ovarian specimen to be normal ovarian follicles in the preserved ovarian cortex. In the contralateral ovary three unconnected nodules were evident on T2W images which had the same signal intensity as the other enlarged ovary.

Burkitt's lymphoma is the most frequent subtype of NHL in childhood and accounts for approximately 34% of these cases. It is the most rapidly growing tumor in children, with a doubling time of approximately 24 hours, so prompt recognition and initiation of therapy are essential. Children who are 5–9 year old represent more than one third of the cases. Affection of the abdomen and pelvic occur in about 45% of cases .Common presenting symptoms include abdominal pain, palpable mass, nausea and vomiting, intestinal obstruction due to bowel compression or intussusception, and acute appendicitis. Weight loss, fever, and other systemic features present more often with disseminated disease but are less commonly seen than in other types of lymphoma in childhood.[113]

David M. Biko et al mentioned that involvement of the lymph nodes in the mesentery and retroperitoneum by Burkitt's lymphoma commonly presents as abdominal and pelvic masses which may be large and single or multiple .[113].Some masses may have necrotic centers containing fluid or, in rare cases, air. Calcification of nodal masses has also been reported. Ascites is a common finding that occurs in 25% of patients. They stated that lymphomatous involvement of the peritoneum is not frequent in children and is manifested by nodularity along the peritoneal reflections usually associated with widespread bowel or mesenteric inflammation. Imaging findings can be confused with other more aggressive pathologies like DSRCT in children and adolescents which may have a similar clinical presentation (which was the suggested diagnosis in our case at first).

PET/CT has become the preferred functional imaging technique at many institutions both for initial staging and for evaluation response to treatment in children with Burkitt's lymphoma . It provides essential clinical information and can help to differentiate between residual tumor and fibrotic tissue during the course of chemotherapy, thereby providing a more accurate diagnosis than does either CT or MR alone. However, because of concerns about radiation risk to pediatric oncology patients the role of MRI is likely to increase. Many studies are investigation the role of MR with special regard to Whole body techniques and DWI in staging and follow up of pediatric patients with lymphoma.[23, 113, 150]

Neuroblastoma

A 2 year old boy with known history of treated neuroblastoma had a denovo small presacral mass at follow up showing hyper intense signal in T1, intermediate signal in T2 with mild contrast enhancement.

Marilyn J. Siegel mentioned that MR imaging is a useful tool to monitor tumor regression and detect recurrence after therapy of neuroblastoma. Demonstration of a residual mass with low signal intensity on T1- and T2-weighted images suggests the diagnosis of fibrosis, although high signal on the T2-weighted sequence may be seen with either residual tumor or fibrosis with an inflammatory component.[91]

Nour-Eldin A. Nour-Eldin et al [19] mentioned that there are still some limitations for MRI in the inability of differentiating viable tumor and nonviable residual lesion, and in the detection of calcified lesions. DWI can certainly improve diagnostic accuracy of MRI in this aspect. Also complementary use of MRI and other metabolic imaging methods such as MIBG scintigraphy or PET scans probably increases diagnostic accuracy and, subsequently, improves clinical outcome of children with NBL. Whole body MRI (WBMRI) is a new imaging method which appears quite useful in detecting distant metastasis, assessing initial treatment responses and identifying tumor recurrence of neuroblastoma. It has very promising results in staging and monitoring NBL, although its clinical impact is still under investigation

Non -Neoplastic masses:

Hemato(metro)colpos

Four adolescent girls (age 13-15 years) with pelvic pain, associated with primary amenorrhea in three of them, were found on MRI to have vaginal and/or uterine distension due to vaginal obstruction.

Imperforate hymen was the cause of the vaginal obstruction in two of the cases who both presented by primary amenorrhea and recurrent pelvic pain. One case showed mild distention of both the uterine cavity and vagina and the other showed marked vaginal distension with normal-sized uterus lying on its top.

The age, clinical presentation and imaging features were similar to those described in the literature. Marilyn J. Siegel mentioned that in adolescent girls, vaginal obstruction is most often the result of a simple imperforate membrane or septum. Affected patients present with cyclic lower abdominal pain or a mass and a history of absent menses. Other presentations may include urine retention and low back pain. 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 and high signal intensity on T2W images. The signal intensity increases on T1-weighted images if the contents are hemorrhagic. [16]

Cem Dane et al mentioned that imperforate hymen occurs in approximately 1 in 2000 females, although true incidence is difficult to obtain. It is rarely diagnosed in the neonatal

period, and most cases occur in the teenagers with symptoms of an expanding pelvic mass. The diagnosis is straightforward clinically as examination always reveals a bulging bluish membrane on retracting the labia. Imperforate hymen is usually not associated with any other Mullerian abnormalities thus extensive investigation for urogenital anomalies is often unnecessary in these cases.[151]

Our third case has a different anatomical abnormality which was atresia of the cervix and upper vagina .The uterus was well developed while its cavity and both fallopian tubes were distended by accumulated hemorrhagic contents. The distal vagina was normal in appearance. This matched with class IB of Mullerian duct anomalies.[43]

This case also had overdistended urinary bladder and bilateral megaureteres that were suggested to be sequel to associated neurogenic bladder or chronic reflux disease. Cem Dane et al and Sabah Servaes et al mentioned that the differential diagnosis of primary amenorrhea , pelvic pain and fluid filled pelvic mass with low level echoes on US in prepubertal girls include transverse vaginal septum, vaginal agenesis and cervical atresia.

MRI is very helpful to differentiate imperforate hymen from a low transverse vaginal septum and a high vaginal septum from cervical agenesis. MRI will delineate these abnormalities and is considered the gold standard for diagnosis.[10, 151]

Our forth case with vaginal obstruction complained of recurrent pelvic pain with otherwise regular normal menses. MRI revealed didelphys uterus with two separate uterine bodies, two cervices and two vaginae separated by longitudinal septum. One hemivagina (left) was obstructed by a low transverse septum and distended by hemorrhagic contents. Absent left kidney was noted (ipsilateral to the obstructed vagina).

This condition of obstructive uterovaginal duplication anomaly (sometimes called the blind hemivagina malformation) is the consequence of a failure in the fusion process of the 2 Mullerian ducts at around 9 weeks of gestation. The incidence of this malformation is estimated within the range of 0.1% to 3.8%. It is associated with congenital urinary tract anomalies in 50% of the cases. The most recognized genetic theory explaining this combination is an abnormal development of the Wolffian ducts, also named mesonephrenic ducts. The association of this Mullerian duct anomaly with ipsilateral renal agenesis, as in our case, is well described in the literature and referred to as Herner-Werner-Wunderlich syndrome and more recently by the acronym OHVIRA (obstructed hemivagina with ipsilateral renal anomaly). [10, 48, 50, 152]

L. Ballesio described a case having almost the exact same clinical and imaging findings as our case [50].Anita Mandava et al described a 14 year old girl who had unusual presentation by acute urine retention with fever and vomiting. In addition to the left vaginal distention she also had distended cervix, uterus and fallopian tube with several endometrial peritoneal deposits over the left fallopian tube and left ovary and in the recto-uterine pouch.[152]. Eleven cases were described by Cinzia Orazi et al [48] and a larger study of similar 32 cases with the same syndrome were described by Carmen Capito et al [52]. The latter study included both pre-pubertal and pubertal cases where the former group was diagnosed surgically after prenatal or postnatal US revealed ipsilateral renal agenesis or multicystic dysplastic kidney together with a cystic pelvic mass.

This syndrome usually presents after menarche with progressive pelvic pain, sometimes with regular menses, and a palpable pelvic mass due to hemi-hematocolpos. Hematocolpos is usually suspected months after menarche and the diagnosis is generally made only if suspicion of the existence of this syndrome is raised .Clinical presentation may be further delayed, with vaginal discharge of smelly longstanding partially clotted blood, if a narrow communication between the two vaginas exists. A right-sided prevalence of the obstructed system has been described. The vaginal introitus is not involved because of its origin from the urogenital sinus.[50, 52]

US is usually the first modality used in these cases and can suggest the correct diagnosis. MRI provides more detailed information regarding uterine morphology, the cervices, continuity with each vaginal channel (obstructed and non-obstructed), and the bloody nature of the content. Early and accurate diagnosis of this syndrome is important so that adequate and prompt surgical therapy (excision of the vaginal septum) can provide relief of pain and prevent further complications. It is advisable look for an obstructed Mullerian system whenever a multicystic dysplastic kidney or the absence of a kidney is discovered in a fetus, or girl postnatally.[48]

Our four cases showed different signal intensities of the retained menstrual blood being hyperintense in T1 and T2 in two cases, hyperintense in T1 and hypointense in T2 in one case while it was hypointense in T1 and hyperintense in T2 in the fourth case. No signal suppression was seen on Fat-Sat images. This is explained by the accumulated blood being of mixed different chorological stages. DWI was carried in 3 of the four cases and showed evident restriction. This however should not be expected in all cases of hemato(metro)colpos as blood contents have variable diffusion properties according to its chronological stage .This is well describe in neuroradiology as hematoma is commonly seen restricted in the hyperacute and late subacute stages only while it is not restricted in the other stages. [153]

Mullerian duct anomalies are a broad and complex spectrum of congenital abnormalities resulting from disruption of Mullerian duct development during embryogenesis .There is considerable controversy regarding their classification with the most widely accepted one being that developed by the American Society of Reproductive Medicine. Various types include uterine hypoplasia and/or agenesis, unicornuate uterus, didelphys, bicornuate, septate, and arcuate uterus, in addition to a peculiar anomaly of uterine morphology that is related to diethylstilbestrol. Due to the different embryologic origins of the upper two-thirds of the vagina, which originates from the Mullerian ducts, and the lower third, which originates from the urogenital sinus, vaginal anomalies, such as hypoplasia/aplasia, duplication, or septa, may or may not coexist with uterine anomalies.[10, 43]

Sabah Servaes et al [10]mentioned that despite the wide acceptance of the current MDA classification system, it has several drawbacks the most noticeable is the inability to classify uteri demonstrating anomalies of 2 or more class and that many of the characteristics that need to be described may not be clearly identifiable in very young patients. Vaginal anomalies, which are usually responsible for many of the symptoms in the pediatric population, are not systematically discussed. In addition, there is a wide variety in the criteria required for accurate diagnosis, particularly when dealing with "duplication" anomalies. Hence they recommended that the more important thing than

ascribing a specific class is to describe the anatomy thoroughly, including the number of cervices, uterine horns, communication of any atrophic horn, and MR signal characteristics, if present, of any uterine or vaginal septum, to guide any surgery or management.

Our four cases of hematometrocolpos elucidated and confirmed the vital role of MRI for assessment of suspected uterine and vaginal abnormalities. As stated previously by Eftychia Kapsalaki et al, MRI can be very enlightening in such cases and not only confirms the diagnosis but accurately localizes the collection and identifies its content and the level of the obstruction as well. It provides important information regarding coexisting abnormalities of the uterus and the urinary tract contributing to the avoidance of unnecessary surgical procedures and supplying a baseline image for the follow up. The accuracy of MRI for diagnosing uterine malformations is well established and even 100% accuracy has been reported. [16, 43, 51, 53]

In cases with suspected gynecological abnormality, Ashish P. Wasnik et al [154] recommended that the MRI protocol should begin with a coronal rapid spin echo T2-weighted sequence(as SSFSE or HASTE) with wide field of view to including most of the kidneys, due to the common coexistent risk of renal anomalies in the setting of suspected genitourinary developmental disorders. Subsequently, a sagittal T2-weighted fast spin-echo sequence is used to delineate the uterine lie then axial and coronal images are taken in the short and long axes of the uterus.T1W images with and without fat-sat are also essential. Contrast injection is only done if indicated like in case of suspicious of associated mass lesion.

Ovarian cyst

A 7 year old girl presenting with right sided pelvic pain for few weeks had a right ovarian cystic lesion showing thin wall(< 3 mm) with homogenous T1 hypo and T2 hyper intense fluid content together with few thin hypointense septa (not exceeding 3 mm). No soft tissue or abnormal enhancement was detected after GAD injection and no diffusion restriction. The diagnosis was complicated functional ovarian cyst with no suspicious features.

These imaging features matched with those described for benign functional ovarian cysts by Deepa R. Pai [1]and Mehmet Ruhi Onur et al[8]

Ovarian cysts may be physiological, functional or neoplastic. Physiological cysts include the normal follicle cyst and corpus luteum which are less than 3 cm in diameter. Functional cysts result from failed involution of the follicle or corpus luteum and occasionally present with intra-cystic hemorrhage. Other ovarian cystic lesions include cystic neoplasms as mature cystic teratoma and serous and mucinous cystadenomas. Functional ovarian cysts are the most common adnexal masses seen in children. They 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. Any cystic lesion identified between infancy and the onset of puberty should be viewed with a higher degree of suspicion, as functional ovarian cysts are only rarely encountered in this age group. [1, 71]

As described by Mehmet Ruhi Onur et al [8], Ovarian cysts appear on US as well-circumscribed, thin-walled anechoic cystic lesions arising from adnexal region. CT has

limited role in the characterization of ovarian cysts. Ovarian cysts appear hypointense on T1 and hyperintense on T2W MRI. Contrast enhanced MRI can reveal nodular or papillary enhanced solid components of cystic neoplasm which are usually large. Complications of ovarian cysts include rupture and torsion. Purely cystic lesions less than 4 cm without solid component may be observed with follow-up US since they have a tendency to regress.

Ovarian torsion

A 5 year old girl had an enlarged right ovary with abnormal persistent hyperintensity in both T1 and T2 weighted images with hypointense rim. No contrast enhancement was noted after GAD injection and it showed evident diffusion restriction. She presented by acute unilateral pelvic pain few weeks prior to the MRI study. Hemorrhagic infarction and total ovarian gangrene were found at surgery with no associated adnexal mass.

Lesley L. Breech[78] mentioned that adnexal torsion accounts for about 2.7% of all cases with acute abdominal pain in children. In contrast to the adult where torsion is likely due to an ovarian mass, the pediatric patient may have normal ovaries in up to 25% of cases of adnexal torsion. In the rest of cases, the responsible ovarian lesion is commonly benign as large simple cyst or cystic teratoma. This diagnosis should be strongly considered in a child or adolescent who presents with intermittent lower quadrant pain, associated nausea and vomiting, and an adnexal mass.

The imaging findings in our case were somewhat similar to those previously described by Marilyn J. Siegel[16] and Sung E. Rha et al [77]. They stated that MR findings of acute torsion include an enlarged edematous adnexa (low on T1 and high on T2), prominent follicles along the periphery of the ovary, engorged adnexal vessels on the side of the torsion, thickened fallopian tubes, 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. The ovary usually enhances when torsion is incomplete or intermittent but with complete torsion and infarction, there is absence of enhancement. Signs that are specific to hemorrhagic infarction, which indicate ovaries non-salvageability, include foci of high signal intensity corresponding to intra-lesional hemorrhage on T1-weighted images, hemorrhage in the thickened fallopian tube and hemoperitoneum

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. If the torsion is partial and intermittent with spontaneous untwisting, symptoms may subside, only to return within hours, days, or week. If the adnexal torsion is complete and goes undiagnosed and untreated, hemorrhagic infarction may occur in the involved ovary (as in our case) and may lead to peritonitis and death. Local peritonitis may cause intestinal obstruction. Early diagnosis can help prevent irreversible damage to the adnexal structures and may thus allow conservative, ovary sparing treatment in female pediatric and adolescent patients. [16, 77]

Lymphangioma

We presented a 15 year old boy with a huge pelvi-abdominal multilocular cystic lesion containing locules of variable signal intensities predominantly hyperintense in T1 and hypointense in T2 with multiple fluid levels. No enhancing soft tissue could be detected and no diffusion restriction.

Lymphatic malformations, also referred to as cystic lymphangioma or cystic hygroma, are benign congenital vascular malformations that result from failure to establish normal lymphatic channel anatomy. Acquired cases occur because of either trauma or inflammation that obliterates the lymphatic vessels. Retroperitoneal cystic lymphatic malformations are usually detected incidentally, but they may be complicated by infection, hemorrhage, rupture, or mass effect.[5]

The imaging features of our case are similar to those previously described by Deepa R. Pai [1]and Rebecca Stein-Wexler [111]. They described multilocular cystic lesions hypointense on T1 and hyperintense on T2 .Fluid-fluid levels and variable internal signal intensities result from hemorrhage or infection. Vascular flow voids do not occur. Peripheral and septal enhancement is common yielding the characteristic ''rings and arcs'' appearance, however lymphatic malformations do not internally enhance unless there was treatment. This is an important point to differentiate them from venous malformations which fill with contrast material on delayed imaging.

Presacral Dermoid cyst

A 2 year old girl had a large well defined presacral mass with mixed signal intensities in both T1 and T2 and partial signal loss with fat suppression. Such features denoted mixed fat and fluid components. No contrast enhancement was seen and no restricted diffusion. No associated spinal dysraphism was noted. The condition was diagnosed successfully as a simple dermoid cyst.

As mentioned by Akihiro Nishie et al, dermoid cyst is a cyst of developmental origin and histologically characterized with squamous cell lining, keratinoid or mucoid content, occasionally associated with fatty component and calcified structure. When the presence of fat and/or calcification is evident, radiological diagnosis is readily made. In cases with little fat or calcification the lesion presents as a 'nonspecific' cyst, radiologically, because keratinoid or mucoid substance yields water density on CT, and long T1 and T2 characteristics on conventional MR images which result in a diagnostic dilemma. Chemical shift fat suppression and DWI are very helpful sequences for better mass characterization. Fat content is easily appreciated as it is suppressed in Fat-Sat images and the keratinoid substance causes restricted diffusion with low ADC values. [105]

A dermoid cyst is one of the lesions commonly present in Currarino triad, with associated anorectal anomalies and sacral bony defects. Differential diagnosis of fat containing lesions in the presacral space includes also sacrococcygeal teratoma and lipomeningocele. Other cystic lesions in the presacral space that need to be excluded include rectal duplication cyst, anterior sacral meningocele, lymphangioma and neuro-enteric cyst. [5, 105, 109]

Pediatric body MRI has made many advances since its inception. It offers exquisite soft tissue contrast without exposure to ionizing radiation, which is important in the pediatric population. The advancement to 3.0 T magnets, new sequences, multichannel coils and parallel imaging allows for increased resolution and decreased image acquisition times.MRI is quickly becoming the modality of choice for pediatric abdominal masses. It plays an important role in the diagnosis of disease, staging and monitoring response to therapy.

SUMMARY

Our study was conducted on 20 children presenting clinically with known or suspected pelvic mass with the aim to evaluate the role of MRI in the assessment and diagnosis of these masses.

All patients were subjected to full history taking (from parents and referring physician), thorough clinical examination and review of the laboratory investigations and of available previous imaging studies. Anesthesia was used in uncooperative patients (less than 7 years old). The MR study was performed on closed 1.5 T magnet machine using combination of T1 and T2 SE sequences in different orthogonal planes including fat suppressed and IV GAD contrast enhanced studies. DWI was also used in most of the cases.

The study included 5 males and 15 females and their age ranged between 2 and 17 years old. The examined pelvic masses included 12 neoplastic and 8 non neoplastic masses of different origins.

The cases with neoplastic masses included two with benign tumors (mature ovarian teratoma and presacral ganglioneuroma) and 10 with malignant tumors (prostatic rhabdomyosarcoma, cervical rhabdomyosarcoma, 4 cases with yolk sac tumor, spindle cell sarcoma, presacral chordoma, pelvi-abdominal Burkitt's lymphoma and recurrent neuroblastoma). Eight non neoplastic masses were found and included presacral dermoid cyst, pelvi-abdominal lymphangioma, complicated ovarian cyst, ovarian torsion with secondary necrosis and four cases with hemato (metro) colpos.

MR proved very useful in depicting the anatomic details of examined masses providing excellent soft tissue contrast and spatial resolution with the ability to specifically characterize certain types of tissues like fat and hemorrhagic contents. Imaging features were specific to certain diagnoses in cases of non neoplastic masses and in the case of ovarian teratoma. However, the unusual presentation of ovarian teratoma with solid mass together with ascites and peritoneal nodules led to the impression of immature type which was excluded by pathological results. MRI was also excellent at depicting the cause of obstruction and associated anomalies in cases of hemato (metro) colpos

Even in the neoplastic cases where MRI imaging features were not specific enough for certain diagnosis, they were very helpful to confirm presence of underlying neoplastic process, to suggest either its benign or aggressive nature and to accurately demonstrates its extent which was essential for surgical removal or setting a baseline for follow up studies after neoadjuvant treatment.