The Importance of NGOs In Today's Societies

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Governing a caring country regardless of the status of its development, is based on 3 essential factors:

1. Government for making and supervising laws and building the necessary infrastructures for the benefit of society,

2. The Economy and its financial institutions to encourage various economic sectors to boost prosperity and improve living standards,

3. Non-governmental (NGO), non-profit organizations (NPO) which are founded by socially motivated people to engage with the diverse humanitarian and cultural aspects of civil society with a view of curbing existing shortcomings while alleviating various hazards that threaten the public and society at large. To achieve these objectives, first and foremost it is essential to carry out the kind of feasibility studies that can assess and evaluate the various shortcomings amongst the targeted population. This should be carried out by qualified volunteers and co-workers with proper insight into the problems as well as the know, how for managing them. Therefore, there is an unwritten, mutually accepted understanding between government and NGOs in their roles to share such social responsibilities.

NGOs depend permanently on sponsors and sustained financial public supports. In this regard, transparency and accountability is essential for ensuring continuity of support for the work of the NGO from its sponsors. My following comments will describe the formation and development of MAHAK as an example of a public based, successful NGO.

In 1991, in the aftermath of the start of the Iran-Iraq War, the 'Society to Support Children Suffering from Cancer' - "MAHAK" - was founded by a group of charitable Iranians wishing to alleviate pain and suffering from children suffering with cancer, who did not have access to proper medical care. The aim was also to assist their parents so that they could deal with these issues including their terrible psychological burden.

According to MAHAK's mission and vision, every child with cancer has the right to have access to proper diagnosis, treatment and follow-up and prevented from facing death because of poverty .Therefore, MAHAK has committed itself to covering the full expenses of each patient's medical procedures. In line with our motto, MAHAK's doors have thus been always open to all patients regardless of their nationality, race and religion. Based on these lofty goals and ideals, public demand for MAHAK's contribution has undergone a kind of evolution that has, over time, seen its role change form a supporting charity society into a comprehensive 'Pediatric Cancer Treatment and Research Center' (MPCTRC).

In 1991 MAHAK initiated its activities by assigning social worker volunteers to pediatric Hematology/Oncology departments at University Children Hospitals in Tehran to evaluate the medical needs of cancer afflicted children and to reimburse their payments for medicaments and other expenses. Additionally, the wards were also refurbished and made more children friendly. By increasing the

number of patient referrals to Tehran for treatment and follow-up there was a rising need for a hostel to accommodate children and their parents coming from rural areas. Therefore, shortly thereafter, a well equipped hostel was also built and run by the city and local sponsors and donors. At that time University Hematology-Oncology units, as primary referral centers were confronted with budgetary and personnel shortages. Hence, they were unable to provide optimal care for cancer children. Thus it was decided to expand our range of activities and medical services to fill the various existing gaps and fulfill our long standing dream. By coming up with the necessary budget and resources and relying on generous donations of our compatriots, we decided to build a pediatric cancer center adjacent to MAHAKS 's administration offices. The new building was completed in 2005 with 100 ‘single-bed’ rooms, state of the art imaging equipment, 2 linear accelerators ,laboratory facilities,2 surgical theaters, 2 ICUs, emergency room, pain department, aqua treatment as well as a full range of out-patient clinics. In 2007 ‘MPCTRC’ was inaugurated by the late Prof. Vossough and her colleagues who started their comprehensive clinical activity. This led to a surge of referrals which in turn resulted at times with personnel overburden due to the fact that we are the only pediatric cancer center that offers 24/7 services through out the year. Further we have been keen to accept and admit patients with CNS tumors and have assigned a special ward for these patients and have a stand-by neurosurgical staff. In 2011, given the emerging need for BM and Stem Cell Transplantation, as a result of the generous donation provided by one of our benevolent supporters, we built a unit matching all international standards for the mentioned purposes with good therapeutic results in selected candidates. From early on we noticed the importance of the need to share information and include patients’ family regarding their child 's disease and the ongoing procedures. Information and knowledge about a patients’ disease is, however at times, a severe psychological trauma for family members, necessitating consultation and therapeutic support and intervention by dedicated psychologists who have since also become an integrated part of our staff. To carry out our missions, we have expanded our charitable activity throughout the country and have opened new offices in some major cities which are capable of providing full treatment coverage for cancer children in 32 university hospitals. Due to the abundance of patients we have established a hospital based cancer registry for data collection and evaluation. Also, we have an active research center to evaluate our diagnostic and therapeutic results under the guidance of our medical staff. In the past 5 years we have published eight papers in national and international medical journals and our colleagues have actively participated in international oncology meetings and congresses to present our data. In order to match international standards, as an NGO that has being ranked as the 4th NGO among 299 benchmarked by SGS and received the gold award from the International Project Management Association (IPMA), we have organized regular congresses with outstanding oncologist and pain specialists in order to present, share and exchange their experiences with our colleagues Moreover, we are active members of SIOP, SIOP-ASIA , EBMT and have links to St. Jude Children Medical Center, GPOH, I-BFM and Gustave Roussei Medical Center.

It is an obvious factor for us to find ways for coming up with the resources we require in order to shoulder the enormous financial burdens and expenses that we face. These can be secured from the following methods:

- I Medical expenses are partly covered by insurance fees
- II Saving and donation boxes

- III Regular seasonal and annual fund raisings, national and in the USA
- IV Seasonal bazars and festive dinners
- V Donations, collected at our offices in Tehran and other cities
- VI Membership fees
- VII Tax exemptions

In the past 25 years we have had over 24000 patients under our supportive care and since the inauguration of MPCRTC, we have dealt with some 6300 patients who were under treatment or 'follow up'. Our therapeutic results are encouraging and in some diseases comparable with reported international results.

MAHAK is not just a charity and medical center. Indeed, over time, it has become a melting center for young and old volunteers to help organize scheduled events and gatherings. Some are assigned to go to the wards to play and entertain patients and parents. In other words they are keen and have the needed initiative to help whenever they are needed.

MAHAKs' administrative performances and achievements has been awarded with international credits and decorations.

This is the the best result and model for sharing public responsibility, with full accountability and transparency,

The role of non-governmental organizations (NGO) in treating children with cancer

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Charitable associations can play an important role in the treatment and social support of children with cancer. In cancerous patients, especially in children given the sensitivity was considerable public attention to it.

In this regard, non-governmental organizations (NGO) have been actively involved and with the help of donors have been able to aid of patients. Many communities are woring in this way such as Mahak (Tehran), Moshk (Ahvaz), Kasa (Isfahan), Bonyad Omid (Shiraz), Aryan, Sana, koomesh, Mehraneh, Eltiam, Sepas, Daheshpoor, Yas, Arezoo, Navid-e-Sabz and.... but only Mahak institution had established a benchmark empowerment center and had constructed a special hospital for management of children.

We were inspired by the Mahak institution in the north of Iran (Sari) attempt to establish a special center to treating cancerous children. At first stage we had instituted an association (named Maahak) and took the first step with the help of donors. We have prepare an area of 2000 square meters of land donated in this regard. Unlike the Mahak Institute, the financial support for the construction of hospitals did not have much. So we had used the donations from people and students piggy banks. So that since 1392 has collected 40 billion rials cash and non-cash donation with physical progress of 50% for building of 100 boards special hospital. Probably It will tapping in the next two years.

It seems inspired by the valuable experiences we can attempt to establish such centers (four poles and the center of our country). This article intends to share its experience in this regard with the other partners.

Pelvic Rhabdomyosarcoma (RMS) and ileal neo bladder for continent diversion: Abol-Eneine & Ghoneim Technique

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Introduction: Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in children. In this group, Genitourinary RMS needs aggressive radical tumor resection with chemotherapy and sometimes radiation t. Most of the times resection of these tumors leads to radical cystectomy and vaginectomy.

In this report we will introduce the technique of ileal neobladder as an ideal way for continent urinary diversion after radical cystectomy in RMS.

Methods: One year old girl was brought to ER for protruding mass from vagina. Biopsy showed Embrional RMS. US and MRI showed a big tumor that involved bladder and vagina. Cystoscopy and vaginoscopy showed tumor involving the bladder neck up to trigon and vagina up to cervics. Patient was placed under chemotherapy for 6 cycles. Due to unfavorable results, we decided to go for laparotomy. During exploration we found extensive involvement of bladder and vagina, that we could not save. So radical cystectomy and vaginectomy and bilateral end ureterostomy was done. The patient had chemo for nearly one year and after MRI showing no recurrence of tumor ,the patient had ileal neobladder with the Abol- Enein & Ghoneim technique.

Follow up 3 months post-surgery, the patient is doing fine, doing CIC through appendicovesicastomym with no tumor recurrence. Last US showed bilateral mild hydronephrosis.

Disscussion:The principle of treating children with RMS is to save vital abdominal organs. Chemotherapy and radiation are the main primary treatments of these tumors. When there are extensive organ involvement, the only treatment is radical tumor resection with safe margin. In Genitourinary RMS, this means total cystectomy. When this operation leads to total cystectomy there remains the problem of bladder substitution .There are many ways for continent diversion after cystectomy of which ileal neo bladder is safe and feasible operation.

Conclusion:Pelvic RMS is a very aggressive tumor and sometimes the only way of curing, is radical cystectomy. Ileal neobladder is a very feasible and safe operation for bladder substitution and continent diversion after cystectomy in bladder RMS.

Rhabdomyosarcoma in Children- A Case report

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Soft tissue sarcomas (STSs) are a heterogeneous group of neoplasms that arise from primitive mesenchymal cells throughout the body. STSs account for approximately 7% of all cancer cases in children and adolescents younger than 20 years. Traditionally, pediatric STSs are divided into rhabdomyosarcoma and nonrhabdomyosarcoma soft tissue sarcomas (NRSTSs). Rhabdomyosarcoma in children accounts for between 5% to 8% of all childhood cancers. It is the most common soft tissue sarcoma in childhood. About 10–20% of patients present metastases at diagnosis. The treatment of metastatic rhabdomyosarcoma comprises a multidisciplinary approach with a combination of chemotherapy and adequate local treatment, surgery and/or radiotherapy. However, these patients have not benefited from the therapeutic progress achieved in localised rhabdomyosarcoma, as the 5-year overall survival (5Y-OS) still remains between 10% to 35%. Our patient was a 10 years old girl who presented with right thigh mass which excised compeletly. Pathologic diagnosis was rhabdomyosarcoma and adjuvant therapy performed but unfortunately she expired after a year with pulmonary and cerebral metastases.

Management of Unresectable Large Ewing Sarcoma: Role of Surgical Exploration versus Imaging

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Background: Ewing sarcoma is a rare tumor which is also known as the most common chest wall malignancy in children. Surgical management of these masses is widely changed during last decades with the development of neoadjuvant chemotherapy and chest wall reconstruction techniques which let us resecting tumors that were not resectable in previous decades.

Case Presentation: The patient was 12 YO girl Referred to our center due to chest wall mass which was accidentally palpated by parents she was evaluated by pediatricians and after confirming the diagnosis with tru-cut biopsy neoadjuvant chemotherapy was started for her. After chemotherapy CT scan showed a 10x10x6 Cm mass which seem to be unresectable due to involvement of wide area of chest wall, Diaphragm and abdominal wall. Patient undergone wide excision of mass with resection of 12th, 11th and 10th Ribs and lateral part of Posterior abdominal wall. Diaphragm was also partially resected. Then the remnant of diaphragm was fixed to the 9th rib. Chest wall and abdominal wall closed primarily and an onlay dual mesh was fixed over it. Patient tolerated the procedure well and discharged after seven days.

Results: With evolving neoadjuvant chemotherapy and chest wall reconstruction techniques resection of previously unresectable tumors is now possible while radiologic indicators of respectability did not changed. The other mechanism which made this wide resection possible might be the high plasticity of chest and abdominal wall in children that allowed us to do this operation.

Conclusion: New guidelines should be provided for evaluation of chest wall masses in children especially after several report in which wide resection of chest wall showed to be possible or routine surgical exploration should be proposed for evaluation chest wall masses regardless of tumor size.

Genitourinary RMS in Children: A Surgical Challenges

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Rhabdomyosarcoma(RMS) is a cancer made up of cells that normally develop into skeletal muscle.RMS is a cancer of embryonal cells.It can start nearly anywhere in the body. There are 2 main types of RMS, along with some less common types.Embryonal RMS (ERMS) is the most common type. It is usually affects children in their first 5 years of life. Two subtypes of ERMS , botryoid and spindle cell RMS, tend to have a better prognosis that the more common conventional form of ERMS. Alveolar RMS typically affects all age groups equally,but it is most often seen in older children and teens.Anaplastic RMS is an uncommon type that occurs in adults but is very rare in children. Tumors in the bladder or prostate can lead to blood in the urine, while a tumor in the vagina can cause vaginal bleeding. Any of these might grow big enough to make it hard or painful to urinate or have bowel movements. All children and adults with RMS will be treated with surgery to remove the tumor if it is possible to do so without causing major damage or disfigurement.In some cases ,Cht and/or RT may be used first to try to shrink the tumor. The goal is to remove the tumor completely, but this is often not possible. In most children with RMS,it is not possible to remove all of the tumor by surgery.Surgery to treat RMS of the eye or genital areas is usually a biopsy . Cht, and sometimes RT, may be given by surgery to shrink large tumors. All patients with RMS should get Cht. Without it, it is very likely that the cancer will come back at distant sites in the body because small amount of cancer are always present in the other parts of the body when the cancer is first diagnosed. Surgery is the first step in treating RMS,unless the cases with distant metastasis.Complete removal of the main tumor ,along with some surrounding normal tissue is the goal whenever possible. In some cases debulking of the tumor may help other treatments (Cht and RT) to work better.Nearby lymph nodes biopsy is necessary always.If the tumor is large or is in a spot where removing it completely would severely affect the childs appearance or cause other problems, then surgery may be delayed until after a few courses of Cht and possibly RT to try to shrink it,or surgery may not be done at all.The biopsy is generally the first surgery done for RMS.The type of biopsy used is based on imaging test results,location and size of the tumor, the patient" age and health. The common site of treatment failure in patients with localized RMS has been local recurrence.Surgical removal of the entire tumor should considered initially,but only if major functional/cosmetic impairment will not result.So,complete removal of primary tumor with a surrounding margin of normal tissue and sampling possibly involved lymph nodes in draining nodal basin is recommended. Genitourinary region is exception to the rule of normal margins exist. There is little evidence that debulking surgery improves outcome,compaired with biopsy alone. The exact role of delayed primary excision remains undefined in RMS and is most appropriate if it is anticipated that a complete resection is possible and that the modest reduction in radition dose will subsequently decrease the risk for late effects. Only 15% of patients present with Group I, completely resected disease, so RT is used in the majority of cases.RT is an effective method for achieving local control of the tumor for patients with microscopic or gross residual disease after biopsy, initial surgical resection ,or Cht. Group I patients don"t need RT.In more than 50% of group 2 patients local recurrence was due to noncompliance with guidelines or omission of RT.The common type of relapse for patients with group 3 disease is local

failure .Patients with involved lymph nodes also have a higher risk of local and distant failure than do patients whose lymph nodes are uninvolved. As with surgical management of patients with RMS recommendations for RT depend on the site of primary tumor, the postsurgical amount of residual disease (none vs. microscopic vs. macroscopic) , and the presence of involved lymph nodes. Techniques to deliver RT specifically to the tumor while sparing normal tissue (e,g; conformal RT, intensity modulated RT [IMRT] , proton-beam therapy, or brachytherapy are appropriate.Conventional RT remains the standard for treating patients who have RMS with gross residual tumor.Brachytherapy , using either intracavitary or interstitial implants , is another method of local control and has been used in selected situations for children with RMS,especially those with primary tumors at vaginal or vulvar sites and selected bladder/ prostate sites.Patients with initial group III disease, who subsequently have microscopic residual disease after Cht with or without delayed surge are likely to achieve local control with RT.Patients with RMS arising from the perineum or anus usually have advanced disease.These patients are preferentially managed with ChT and RT without aggressive surgery, which may result in loss of sphincter control.Primary sites for RMS of genitourinary system in children are paratesticular area,bladder,prostate,kidney,vulva,vagina, and uterus.Bladder preservation is a major goal of therapy for patients with tumors arising in the bladder and prostate. In rare cases ,the tumor is defined to the dome of the bladder and can be completely resected. Otherwise, to preserve a functional bladder in patients with gross residual tumor,Cht and RT have been used to reduce tumor bulk,followed , when necessary ,by a more limited surgical procedure such as partial cystectomy. Patients with a primary tumor of the bladder/ prostate who present with a pelvic mass resulting to a distended bladder duo to bladder outlet obstruction at diagnosis receive RT to a volume defined by imaging studies after initial Cht to relieve outlet obstruction. This approach to therapy remains generally accepted, with the belief that more effective Cht and RT will continue to increase the frequency of bladder salvage. The initial surgical procedure in most patients consist of a biopsy,which often can be done using ultrasound or cystoscopy, or direct vision transanal route. In selected cases in one series bladder conserving surgery plus brachytherapy for boys with prostate or bladder-neck RMS led to excellent survival, bladder preservation, and short-term functional results .For patients with biopsy - proven ,residual malignant tumor after Cht and RT, appropriate surgical management may include partial cystectomy,prostatectomy, or exentration(usually approached anteriorly with preservation of the rectum).

An alternative strategy, used in European SIOP protocols, has been to avoid major radical surgery when possible and omit external-beam RT if complete disappearance of tumor can be achieved by Cht and conservative surgical procedures. The goal is to preserve a functional bladder and prostate without incurring the late effects of RT or having to perform a total cystectomy/ prostatectomy.

In patients with RMS of bladder/prostate who have received Cht and RT,the presense of well-differentiated rhabdomyoblasts or evidence of maturation in residual tumor is not associated with a high risk of recurrence,additional course of Cht should be given before cystectomy is considered. Surgery should considered only if malignant tumor cells don”t disappear over time after initial Cht and RT.

For patients with primary tumors of vulva/vagina/uterus,the initial surgical therapy is usually a biopsy.Initial radical surgery is not indicated for these tumors. Conservative surgical intervention for

vaginal RMS ,with primary Cht and adjunctive RT(often brachytherapy) for residual disease results in excellent survival rates.

Exenteration is usually not required for primary tumors of cervix, but if needed,it may be done,with rectal preservation possible in most cases.

Girls with GU primary tumors should have their ovaries shielded or possibly moved, in an effort to preserve fertility when they are receiving RT to the lower abdomen and pelvis.

For previously untreated RMS The surgical options include :

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1- For tumors that are only at the top of the bladder: surgery (wide local excision) is done.
2- For tumors of the prostate or bladder(other than the top):

- Cht and RT are given first to shrink the tumor.If cancer cells remain after Cht and RT , the tumor is removed by surgery. Surgery may include removal the prostate, part of bladder, or pelvic exenteration without removal of the rectum.This may include removal of the lower colon and the bladder. In girls,the cervix,vagina , ovaries, and nearby lymphe nodes may be removed).
- Cht is given first to shrink the tumor. Surgery to remove the tumor, but not the bladder or prostate, is done. Internal radiation therapy may be given after surgery.

3- RMS of the vulva, vagina,uterus or ovary :

*For tumors of the vulva and vagina: treatment may include Cht followed by surgery to remove the tumor. Internal or external RT may be given after surgery.

*For tumors of uterus: Treatment may include Cht with or without RT. Sometimes surgery may be needed to remove any remaining tumor cells.

*For tumors of cervix : Treatment may include Cht followed by surgery to remove any remaining tumor.

*For tumors of ovary: Treatment may include combination Cht followed by surgery to to remove any remaining tumor.

Proton therapy in St.Jude Children" Research Hospital

Primary Pulmonary Fibrosarcoma with Bone Metastasis: a Successful Treatment with Post-Operation Adjuvant Chemotherapy

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Background: Pulmonary fibrosarcoma has been an extremely rare tumor in children. Wide surgical resection of infantile fibrosarcoma would be the treatment of choice.

Case Presentation:Post-operative chemotherapy has shown the benefit in the cases of residual disease after initial surgery and metastatic disease in the literature. We have presented the case of a 70-days old male child with primary infantile fibrosarcoma of the left lung and distant metastasis of skull.

Conclusions: The aim of this publication was to highlight the role of adjuvant chemotherapy to improve outcome of infantile fibrosarcoma with residual tumor and / or metastatic disease.

ALVEOLAR RHABDOMYOSARCOMA ARISING IN A GIANT CONGENITAL MELANOCYTIC NEVUS INA FEMALE INFANT-CASE REPORT

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Giant congenital melanocytic nevus is an uncommon nevus in newborn which defined as a predisposing factor for malignancies such as melanoma. Other malignancies such as rhabdomyosarcoma rarely have seen in a giant CMN.we have some report about rhabdomyosarcoma arising from CMN Rhabdomyosarcoma present with nodules ,pedunculated lesion, ulcers and skin tags in these patients. Except an adult case, other cases are children.Our case is second patient with alveolar type which have poorest prognosis. According to these reports serial examination, parental education and serious investigation for any changes in integrity of CMN is recommended. Size nevi greater than 20 cm, can be a precancerous state for some malignancies such as malignant melanoma.Also rare conditions like rhabdomyosarcoma in the context of giant CMN can occur (Holcomb and Murphy ,2010. Rhabdomyosarcoma is the common pediatric soft tissue sarcoma. This sarcoma has four histologic subtypes. Alveolar tumors arise most common in the trunk and have poorest prognosis (Kleigman, 2011). We will present a case with alveolar rhabdomyosarcoma arisen from giant CMN in a seven-month-old girl that suggest carefully follow up for these patients and review this article.

Abdominal wall Dsmoid tumor

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Dsmoid tumor is an ucommon benign tumor with invasive unexpected behaviour in pediatric population that may involve chest wall or abdominal wall .

Although wide local excsion of tumor is a general accepted approach , recurrance rate in is sometimes high despite compelete resection and appropriate post operative follow up.

In some cases of abdominal wall desmoid tumor , morbodity and mortality is seen despite negative margins of the tumor specimen (pathologic approved) and this reccurant behaviour is accomppany with a lot of problems for both surgeon and patient.

METHODS:

A case report : A 2 years old boy with abdominal wall desmoid tumor (2*3 cm) undergome local excsion with 2 cm margin and primary closure was done . After 6 month recurrant tumor in same place occurred and the child undergone R.L.Q quadranectomy and a little part of L.L.Q resection and repaired with dual mesh and he discharged with Tamoxifen and solindac. He was free of dissease for 2 years and the medical therapy was discontinued. After a 4 month again the symptms of recurrant tumor appeared in periumblical and L.L.Q region . compelete excsion and mesh replacemant was done again and the child was discharged with medical and adjuvant radiotherapy. He was free of tumor for 2 years (in follow up period) and no date is available for 3 years following radiotherapy (Missed follow up)

Again the child is addmmitted with massive tumoral invasion of intra abdominal organ and femoral vessels and abdominal wall and upper G.I.B (hematemesis). The child is reffered to Tehran for cadavric abdominal wall substition that was denied and after few days he expired with sepsis and decompensated massive G.I.B.

RESULTS:

Desmoid tumors are benign tumors with probable invasive and fatal behaoiur . although appropriate surgical resections is recommend , invasive post operative reccurance and anvasion is a really challenging problem with sophisticated management.

CONCLUSIONS:

Desmoid tumor may have somehow invasive bahavoiur despite benign definition of it and post operative management of resections may be challenging.

Congenital Rhabdomyosarcoma of Shoulder

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Abstract: A 16-day-old female was referred with congenital swelling on her right shoulder. On examination, there was a hard, round, ecchymotic, nontender, slightly movable, warm and shiny 10x15 cm mass on the right axillary pits which was extended to the right side of neck and chest wall. The mass separated the shoulder from the chest wall causing paralysis of right hand. Chest X-ray, ultrasound and MRI with contrast demonstrated a soft tissue mass suspected to be a hemangioma. The mass rapidly increased in size despite aggressive steroid therapy with rupture and bleeding. On the 45th post natal day the baby was taken to operating room to control the bleeding and if possible total excision of the mass. The mass was separated easily from the surrounding tissue and was excised along with right upper extremity. At the end of surgery the baby had cardiac arrest, and apparently died of Disseminated Intravascular Coagulation (DIC). The final pathology report was Rhabdomyosarcoma (RMS).

8 years survey for patients with Wilms Tumor

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Here we present the data of last 8 years patients who were treated for Wilm's tumor. There were 49 patients, 31 were male and 19 female. 5 patients were less than one year and 37 patients' were1-5 years and 4 between 5-10 and three were older than years. Nineteen had left side and 25 were right sided and 5 had bilateral Wilm's tumor. 38 had favorable histology and 11 had unfavorable histology. 21 patients were stage one, 10 stage 2, 7 stage 3, 6 stage 4 and 5 stage 5.

Till 18 months ago all patients had histologic diagnosis before chemotherapy either by total nephrectomy, open biopsy or needle biopsy. Within last two years clinical and radiologic findings were used to start chemotherapy and after 6-8 weeks they underwent surgery which made the surgery much easier. In one patient history and physical exam and radiologic findings were in favor of Wilm's tumor and received chemotherapy and was operated on after 8 weeks but the last histology showed to malignant cell.

So we recommend the SIOP approach for patients with Wilm's tumor and prefer to start chemotherapy based on clinical and radiologic findings without biopsy and then nephrectomy after 6-8 weeks.

A case report: Right Wilm's tumor with IVC thrombosis.

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A four year old boy presented with history of abdominal pain from 3 months ago. Routine hematology evaluation was normal.

Abdominal ultrasonography revealed a solid mass 70×76mm in the central part of right kidney with extension to the IVC. Its length was reported 70mm. Doppler ultrasonography also confirmed IVC thrombosis .Thoracic CT scan showed mild bilateral pleural effusion.

Malignant undifferentiated small cell tumor compatible with wilm's tumor was reported in needle biopsy.

After several course of chemotherapy tumor size reduced to 25×15mm but IVC thrombosis has not been changed significantly.

Patient was candidate for operation. Abdominal transverse incision, right radical nephrectomy, thrombectomy and IVC repair was performed.

BILATERAL WILMS TUMOR. REAL CASES, REAL CHALLENGES

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BACKGROUND:Bilateral Wilms tumor accounted for almost 5% of Wilms tumor cases in pediatric population. Management of a child with bilateral Wilms tumor (BWT) is very challenging. Preservation of the maximum amount of renal parenchyma is needed to prevent renal failure that is reported in almost 75% of Synchronous Wilms tumors.

METHODS:this case series article discusses 4 cases of bilateral Wilms tumors, therapeutic approaches, their outcome and lessons learned in this situations. among our four cases of bilateral Wilms tumor the main challenges were need to biopsy, core needle VS open biopsy, primary resection or neo-adjuvant chemotherapy, unilateral nephrectomy on the most involved side and partial nephrectomy on the other side or nephron sparing surgery on both sides, bilateral nephrectomy and renal transplantation. We aimed to discuss these issues based on recent literatures and our experiences.

RESULTS:According to our findings and experiences, the reported risk of renal failure in synchronous bilateral Wilms tumor supposed to be considerable (75% versus 1% in unilateral Wilm's) and the experience of bilateral nephrectomy and renal transplantation is disappointing in pediatrics so we strongly recommend bilateral nephron sparing surgery from the first step and even "on bench nephron sparing" surgery and auto-renal transplantation. We also suggest bilateral core needle biopsy and neo-adjuvant chemotherapy that help us to configure the favorable or unfavorable histology and the response to chemotherapy. Some authors reserve needle biopsy for unresponsive tumors to chemo therapy and some are not recommended biopsy at all. In case of favorable histology, the importance of renal tissue sparing may overcome the free margins as local control may be obtained by chemotherapy and close fallow up and even redo nephron sparing surgery if needed.

CONCLUSIONS:Neo-adjuvant chemotherapy and bilateral nephron-sparing surgery should be considered for all patients who have bilateral Wilm's tumor considering the risk of recurrence in the renal tissue remnants and high risk of end stage renal disease

EXTRARENAL WILMS' TUMOR: CHALLENGES IN DIAGNOSIS AND TREATMENT

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BACKGROUND:Wilms' tumor is one of the most common childhood solid malignancies, which classically arises from primitive metanephric cells, but exceptionally it may arise in places other than kidneys. Extrarenal Wilms' tumor is a rare but challenging entity, considering its diagnosis, histopathology, staging, treatment, and prognosis.

METHODS:In this presentation, we discuss about all these topics in detail after a systematic review of extrarenal Wilms' tumor cases to date in order to provide a clear perspective for confronting this rare disease. We designed a systematic review in Google scholar and PubMed databases using the keywords of Extrarenal Wilms and Extrarenal nephroblastoma in pediatrics (under 18 year old).

RESULTS:Among 193 abstracts there were just 87 pathologically proven primary pure Extrarenal Wilms reports which were reviewed in details. Diagnosis of extrarenal Wilms' tumor is almost always postsurgical, which may jeopardize treatment planning and consulting with parents in the first step. The histopathology of Wilms' tumor is very confusing. While most authors believe that it arises from primitive ectopic nephrogenic rests, teratoid Wilms' tumor leads to the debate whether this tumor is neoplastic or embryonic. Staging of extrarenal Wilms' tumor is also a challenge when we consider the National Wilms' Tumor Study (NWTS) recommendations; all these tumors should be considered as stage II or higher as they are beyond the renal capsule. This will mandate chemotherapy for all patients while most of the reported cases had a favorable histology, and long-term tumor-free survival has been reported even with exclusive surgery in some case reports. Although treatment strategies for extrarenal Wilms' tumor are the same as those for renal Wilms' tumor, different locations and neighboring organs may invoke special considerations and scenarios while planning for surgery and adjuvant therapies. Consulting with the parents is also a problem, considering the rarity of the disease and limited publications.

ONCLUSIONS: ERWT is considered a rare childhood malignancy with atypical presentations. The pathogenesis of ERWT becomes clearer by the popular theory, which suggests the heterotopic metanephric blastema as the precursor of ERWT while the diagnosis, staging, and treatment remain challenging. NWTS protocols are applied for ERWTs due to the rarity of the disease and lack of systematic data. We observed favorable histology in most cases, which made the prognosis good and comparable to that of classic Wilms' tumor with the same stage and histology.

Renal Cell Carcinoma in Children- A Case report

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Renal cell carcinoma is a rare histologic type of tumor in children, with certain features that differ from those in adults. In general, RCC in children tends to present later than Wilms' tumor and is usually associated with signs and/or symptoms of disease. The incidence of renal adenocarcinoma increases with age. It is most frequent in older than 5 years, reaching the incidence of Wilms' tumor in the second decade of life. Whereas the peak of incidence in Wilms' tumor occurs around 3 years of age, renal adenocarcinoma presents between 9 and 15 years of age. We reported a case of RCC in an eight years old boy who underwent left radical nephrectomy after biopsy proved the diagnosis.

Outcomes in Children with Wilms' Tumor: Our Center experience for 2 decade

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Alireza Mirshemirani, Naser Sadeghian, Fatollah Roshanzamir, Mehrdad Izadi, Mehdi Sarafi,

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Background: Wilms' tumor (nephroblastoma) is the most common renal malignancy of childhood. The aim of the study was to evaluate the characteristics of Wilms' tumor and the results of combined modality treatment obtained in our center .

Methods: In this study we collected data in 2 decade. Fifty-five patients diagnosed as having Wilms' tumor were studied in the period between February 1992 and March 2002 and 66 patients in the period between 2006 and 2015. Demographic features, mode of presentation, associated anomalies, the stage of tumor, histopathologic results, and the survival rates were evaluated.

Results: In first decade ,of these 55 patients, 31 were males and 24 were females (M/F = 1.2) in other group 60% female and 40% male. The mean age at the time of diagnosis was 45.2 months for first group and 36 months in other. In first group ,The distribution of 54 operated patients according to the surgical stage was: stage I 32.7%, stage II 16.36%, stage III 38.1%, stage IV 9%, and stage V 1.8% (one patient (1.8%) has not been operated).in second group in 56 patients stage I, 14.3%, stage II 40.8%, stage III 24.5%, stage 10.2%, and stage 10.2%.

Favorable histology was diagnosed in 54.5% and unfavorable histology in 43.6% of the patients in first group but in second group 95.4% were favorable type.

The patients were treated according to National Wilms' Tumor Study protocols. The relapse-free and overall 4 years survival rates were 71% and 86%, respectively in first group.

In second group pulmonary metastasis were seen in 34%, liver metastasis in 2 % , recurrence in 5% .

Conclusion: As a developing country, with similar relapse free and overall survival rates to National Wilms' Tumor Study, our institution showed an improvement in the treatment of patients with Wilms' tumor in recent 20 years, but with more adaptation to the National Wilms' Tumor Study treatment protocols better optimum results seem to be achievable.

Primary renal lymphoma mimicking bilateral Wilm's Tumor

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OBJECTIVES: Primary Renal lymphomas (PRL) are defined as lymphomas arising in the renal parenchyma and not invasion from an adjacent lymphomatous mass .since the kidneys do not contain lymphatic tissue the mechanism of development of PRLs is unclear. Lymphomatous involvement of the kidney is often seen as a part of disseminated disease. The prognosis is usually poor with median survival less than a year .Most of the few cases reported showed rapid systemic progression and a poor prognosis. The disease usually affects adults with an average age of 60 years and slight male preponderance; however it has also been reported in childhood. The most common histological subtype encountered is diffuse large B cell lymphoma (DLBCL)

Case: A 2-year-old male underwent surgery for the presumed diagnosis of bilateral wilms tumor with liver metastases.Bilateral large renal mass which, on biopsy, was diagnosed as a lymphoma.The neoplasm was assessed as primary renal non-Hodgkin high grade lymphoma, diffuse large B-cell type with liver matastasis. Then the patient underwent poly chemotherapy according and fortunately, he cured.

CONCLUSIONS: PRL represents a rare entity which must nevertheless be considered in cases of unusual renal masses or otherwise unexplained renal symptoms. If diagnosed early, cure is possible, and multimodal treatment should be considered.

The Survival of children with Wilm's tumor in, Esfahan (a 7 years study)

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BACKGROUND: The purpose of this study is to evaluate the overall survival and effective factors in 44 infants with Wilms tumor who hospitalized in Emam Hossein and Alzahra hospitals during 7 years from 2008 to 2015.

METHODS: In a retrospective study, we gathered the data from patient documents including: gender, blood group, gestational age, birth weight, birth order (first birth or later in family), breast feeding, kinds of treatment, relapse, metastasis, stage of disease, live or dead and some maternal factors.

RESULTS: The survival analyze revealed that the factors like blood group (p=0.004), birth order (p=0.001), stage of disease (p=0.017) and relapse (p=0.038) can affect the treatment results, death and overall survival.

CONCLUSION: Our findings showed that, in these children with Wilms tumor, the most important cause of death is tumor relapse. So a follow up programme and exact patient observation as well as, a rapid diagnose of tomur relapse can promote the prognosis and increase the overall survival

Left Huge Mediastineal Mass due to Recurrence of Willm's Tumor

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A 17 month boy came with chief complain of palpable abdominal mass after evaluation by hematologist, they reported right renal mass candied for surgery.

During operation right radical nephrectomy without spillage of tumor completely resected.

Pathologic report had shown willm's tumor without evidence of anaplastic compenent in tumor.

Two days after operation chemotherapy was started with diagnosis stag II of willm's tumor and chemotherapy was continued by hematologist in seven courses during six months.

In follow-up, the patient hadn't problem but some effect of chemotherapy such as neutropenia, diarrhea, etc.

One year after nephrectomy, the patient admitted with complain of respiratory distress and fever.

After evaluation, it was recognized huge left mediastineal mass with possibility of recurrence of willm's tumor.

Thoracotomy was performed for him and tumor completely resected pathologic report had shown.

Willm's tumor and positive cytologic finding from fluid in left hemithorax. After surgery chemotherapy had been started for him again for seven courses.

In last four years follow up the patient hadn't any problem.

CONGENITAL MESOBLASTIC NEPHROMA: CASE REPORT AND REVIEW OF THE LITERATURE

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Congenital mesoblastic nephroma (CMN) is the most common renal tumor in neonates, and more than 80% of CMN tumors presenting in the neonatal period. Hypertension has been reported with many renal tumors, with or without hyper-reninemia. There are few reports of CMN and hypertension. Here we describe the clinical presentation in a 37-week gestation infant with hypertension and CMN and review CMN.

Facial Hemangiomas Management: case report and review of article

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Infantile hemangiomas are the most common benign vascular tumors in infancy and childhood. As hemangioma could regress spontaneously, it generally does not require treatment unless proliferation interferes with normal function or gives rise to risk of serious disfigurement and complications unlikely to resolve without treatment. Here we describe a 4 months boy with rapid growing hemangiomatose lesion on his lip and bleeding who was managed successfully with propanolol and corticosteroid. Meanwhile review the literature for the current management of this vascular malformation.

Wilm`s Tumor with extension to IVC and right auricle: Surgical treatment with laparotomy and sternotomy

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Introduction: Wilm`s tumor (WT) with extension to IVC and right auricle as tumor thrombus is one of the challenging problems in treating this tumors. In this scenario the team work of pediatric surgeon and pediatric cardiac surgeon is crucial. In this report we present a child with huge WT with tumor thrombus up to right auricle.

Method: 3.5 year old girl was admitted with a very large abdominal mass. Ultrasound and CT scan showed a right sided tumor of 117 * 84 mm in right kidney, destroying completely the right kidney architecture, with tumor thrombus in IVC extending to right auricle. Suspicious diagnosis of WT and the extension of the tumor thrombus was made , so we decided to do needle biopsy that confirmed the diagnosis of WT.

Patient underwent chemotherapy and subsequent CT showed smaller tumor, with tumor thrombus in IVC and right auricle. We decided to go for laparotomy and mid sternotomy for complete removal of the tumor and hopefully all the tumor and tumor thrombus were completely removed. The tumor thrombus had not invaded the IVC wall.

Patient was discharged in good condition and went for further chemotherapy. In follow up 2 years after surgery patient is doing well without any tumor recurrence.

Discussion: Treatment of WT is mainly radical surgery and chemotherapy, based on tumor histology and staging. In cases of tumor thrombus in IVC , surgery can be done with removal of tumor thrombus, but when the involvement extends up to right auricle , the patient needs more extensive surgery that includes the help of pediatric cardiac surgeon , with laparotomy and midsternotomy.

Conclusion: Wilm`s tumor with extension to IVC and right auricle is challenging scenario in patients with WT. in these cases the recommendation is to do chemotherapy followed by radical tumor resection through laparotomy and then removal of tumor thrombus with mid sternotomy .

Evaluation of Wilm's tumor treatment in Qods Children's Hospital

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Purpose: To determine the prognosis of child with Wilm's tumor after radical surgery and chemotherapy

Patients and methods: 15 patients with Wilm's tumor between 1 to 4 years that operated in Qods Children's Hospital between 2000 and 20016 collected and evaluated for survival. After surgery all patients received chemotherapeutic agents

Results: Of 15 patients 14 patients had stage 2 or 3 and1 patients had stage 5. Follow up of patients cleared 13 patients are survived, and 2 patients died (1 from tumor)

Conclusion: Survival of patients with Wilm's tumor without metastasis is very good if radical surgery and chemotherapy treatments are done appropriately and adequately

One century of nephroblastoma treatment evolution

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In the beginning of the 20th-century nephroblastoma-treatment was surgery, its lethality was roughly 25%, therefore many treatment approaches were investigated. Patients could receive irradiation and drugs; alone or as combined treatment, pre- and postoperatively or even without surgery. By introducing local surgical state-of-the-art guidelines, survival rates increased and reached 20%. Introduction of postoperative irradiation treatment gave another boost to survival reaching 50% in the early 50's. This was when North American colleagues suspected preoperative treatment to facilitate metastasis-development and pushed the upfront-surgery-approach. Nowadays more than 88% of patients survive with a stratification according to local tumor extent, subtype, metastasis and molecular risk factors. In continental Europe physicians started to evaluate the role of preoperative treatment after pure radiologic diagnosis of a renal malignancy at an age of 6 months to 16 years. Initial preoperative irradiation was later on substituted by chemotherapy with Dactinomycin and Vincristin (AV). SIOP investigators thus achieved significantly decreased risk for tumor-rupture and additional down-staging by decreasing lymphnode-involvement. They successfully treated patients having lung metastasis without pulmonary radiotherapy, if they achieved a complete metastatic response. Pathologist investigated histologic subtypes, showing that regression and surviving components in the primary tumor, reflects response to AV and is important for stratification. Recently the increasing importance of persistent blastema for prognosis emerged from these investigations.

80 to 85% of all patients in the SIOP are treated without anthracycline or irradiation, maintaining an overall survival above 88% for the whole group. The biggest group of patients, having a stage-I-intermediate-risk nephroblastoma, need only 2x4 Weeks of AV. Similarly, North-American groups were able to define low risk patient groups. Molecular risk factors such as LOH 1p, 16q and gain of 1q are gradually gaining importance in refining stratification. However, both groups are still struggling to improve treatment for patients with advanced stage diffuse-anaplastic nephroblastoma.

Epidemiological features and survival of Wilms' tumor in children in Isfahan city

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Introduction: Wilms' tumor is the most common primary, malignant, renal tumor in childhood, accounting for 5-6% of all types of cancer during this period. Today, the rate of survival has increased dramatically, partly due to large multicenter studies conducted by the National Wilms' Tumor Study Group that have been resulted new management methods with less morbidity.

Purpose: This study was done because there is not enough information about epidemiological feature and survival of these patients in Iran.

Materials and Methods: This is a retrospective cohort study conducted on all the patients diagnosed as suffering from wilms' tumor and who had been hospitalized and treated during 15 years in training centers, Isfahan. When the initial data was collected from the hospital medical records, follow up on the complications, recurrence and mortality was conducted.

Results: From the 88 cases studied, 40 survived and 45.5% of the patients were male. The most clinical sign was abdominal mass (88.2%) and 91.1% of the patients had favorable and 8.9% unfavorable histology .In 23% of the cases, treatment included surgery only, in 50% surgery and chemotherapy combined and in 27% a combination of surgery, chemotherapy and radiotherapy. Over ally 4 years survival was 95% and 6 years survival 32%.

Discussion: Because survival rate particular the long –term, for the wilms' tumor in our patients was lower than the other centers (32% vs.70-90%). The conclusion drawn from this study is that arrangements should be made to firstly for more researches and modifications in current methods and secondly establishing close communications between the surgeon, pathologist and oncologist is essential to increase the long –term survival rate.

Clinical Evaluation and Prognosis of Wilms' Tumor in Tabriz Children Hospital in Recent Six Years

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BACKGROUND:

Wilms tumor is one of the most common solid malignant tumors in childhood and comprises 6% of all pediatric tumors approximately. In this survey, we evaluated clinical presentation and prognosis of this malignancy in our setting.

METHODS:

In this retrospective study, 52 patients that referred to children hospital evaluated for their clinical manifestation, operation finding, pathological finding, and outcomes. Accompanying disease, risk factors and pre-natal diagnosis also were reviewed. Extent of disease and occurrence of metastasis during treatment were evaluated. Response to therapy and outcome of disease were compared.

RESULTS:

There were 25(48%) male and 27(52%) female patients. Ages of these children were 32.2±23.2 months (3 days-84 months). Three (6%) patients had pre-natal diagnosis and three of them had underlying disease or syndrome that compromised them to wilm' tumor. Presenting signs were abdominal pain in 34(65%) and hematuria in 20(39%) acute abdomen in 5(9%) paleness in6(11%) and urinary retention in 2(4%) patients. Fever were presenting sign in older patients in two cases. Two patients had pulmonary metastasis at admission. Nine(17%) patients went under chemo-radiotherapy after completion of therapy because relapse of tumor.

CONCLUSIONS:

Our results are comparable with other studies but we had more hematuria and less hypertension than others did.

Hypertension after Bilateral Nephron Sparing Surgery for Bilateral Wilm's (case report)

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BACKGROUND: Nephron sparing surgery for unilateral Wilms tumor has been debated recently and is being used to preserve kidney tissue and function. However, nephron sparing surgery is feasible only for selected cases with higher local relapse rate have been observed.there is a significant reduction of nephrons with development of renal hypertension and progressive renal failure. We analyzed outcomes after bilateral partial nephrectomy and unilateral partial plus contralateral total nephrectomy in our patients with bilateral Wilms tumor.

METHODS: We analyzed on our 4 patients (8 kidneys)with bilateral Wilms tumor and 8 unilateral complete resection. Kidney size was measured using volumetric analysis ct scan imaging. Patients were matched with children who had undergone imaging of the abdomen for other malignancies.

RESULTS: Mean kidney volumes after unilateral partial plus total contralateral nephrectomy (60.9 cm3) were significantly greater than the reference kidneys, whereas controls were equal to the bilateral partial nephrectomy group (40.7 cm3). Total kidney volume was significantly larger after bilateral partial nephrectomy (98.1 cm3) vs unilateral partial plus total contralateral nephrectomy (60.9 cm3). Two patients 4 kidney had renal hypertension after unilateral partial plus total contralateral nephrectomy but only 2 after bilateral partial nephrectomyand 1 kidney in total cases. Overall survival and relapse rates were not equal between the groups. There were beter in group with complete rsection.(6/8

CONCLUSIONS: Our findings suggest that patients with bilateral Wilms tumor benefit from bilateral nephron sparing surgery. Hypertension is less common after bilateral partial nephrectomy, However rates of local relapse or disease associated death are separately between the groups.

Maximizing lymph node sampling differ the prognosis during surgical resection of Wilms tumor.(10 years experience)

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BACKGROUND: Sampling lymph nodes (LNs) had decreased recurrence and improved survival for Wilms tumor (WT). We are to show how we can deliver the most numbers of lymph node.en bloc +sampling vs. sampling

.METHODS: We conducted a retrospective chart review from 2005 to 2016 of WT resection cases, examining the type of LN dissection, the specimens submitted to pathology, number of LNs evaluated, and complications associated with the procedure.

RESULTS: We identified 15 children with WT;11had unilateral disease and 4 had bilateral disease. With unilateral disease, more LNs were identified by separate versus en bloc sampling (5.2 ± 0.6 vs. 4.4 ± 1.2 nodes). Both the methods identified fewer LNs compared with en bloc+separate sampling (12.5 ± 2.7 nodes,).

CONCLUSIONS: En bloc+separate sampling yields the most LNs during resection of WT. We recommend using this technique to facilitate the maximum number of LNs evaluated in WT.

Renal function in survivors Wilms tumor with unilateral radical nephrectoctomy with low stages

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BACKGROUND: In the current study, we evaluated the prevalence of hypertension and impaired renal function in long-term survivors of UWT who were treated with nephrotoxic chemotherapy or ionizing radiation METHODS: Eligibility included age ≤5 years at the time of diagnosis of UWT, maintenance of disease remission after unilateral nephrectomy without receipt of abdominal irradiation or nephrotoxic chemotherapy. Renal function was assessed by urinalysis and estimated glomerular filtration rate (eGFR). Patients receiving antihypertensive medication or those with blood pressure readings of >140/90 mm Hg were considered to be hypertensive RESULTS:A total of 8 patients with a median age at diagnosis of 2.2 years (range, 0.2-5.1 years) met eligibility criteria. The median length of follow-up was 6.6 years (range, 1.0-7.8 years). All but 1 patient had stage I/II disease. Fifty-eight patients (90.7%) patients had WT with favorable histology.3 patients had an eGFR <80 mL/minute/1.73m, 2 of whom also had proteinuria No patient had an eGFR <50 mL/minute/1.73m2. Five patients had hypertension, 3 of whom were receiving antihypertensive medications. At the time of last follow-up, no patient had developed end-stage renal disease CONCLUSIONS:Patients with UWT who were treated with out unilateral radical nephrectomy with nephrotoxic chemotherapy or ionizing radiation appear to be at low risk of developing significant long-term renal dysfunction. For this patient population, the routine use of partial nephrectomy does not appear justified but monitoring for a rare patient who develops subtle renal insufficiency.

Renal Function of Patients With Synchronous Bilateral Wilms Tumor Undergoing Surgery

BACKGROUND: Wilms tumor in both kidneys are present in Five percent of children with disease. The treatment challenge is to achieve a high cure rate while maintaining long-term renal function. We retrospectively reviewed our cases with nephron sparing surgery (NSS) in patients with synchronous bilateral Wilms tumor (BWT) operated on between 2005 and 2016. METHODS:Imaging studies, surgical approach, adjuvant therapy, and pathology reports were reviewed. Outcomes evaluated included surgical complications, tumor recurrence, patient survival, and renal function, as assessed by estimated glomerular filtration rate RESULTS: A total of 4 (8kidneys)patients with BWT underwent bilateral NSS; only 1 patients underwent unilateral nephrectomy with contralateral NSS. Postoperative complications included prolonged urine leak (1), infection (2), intussusception (0), and transient renal insufficiency (1). In the long-term, allpatients had local tumor recurrence (managed with repeat NSS in 4and completion nephrectomy in 4) and 1 had an episode of intestinal obstruction requiring surgical intervention. Overall survival was 50.7% (mean follow-up, 4.1 years). Of the 2 patients who died both had anaplastic histology. All of the patients had an estimated glomerular filtration rate more than 45 mL/min/1.73 m2 at the last follow-up.

CONCLUSIONS: In patients with synchronous, BWT, bilateral NSS is safe and preserving maximal renal parenchyma survival howevr rate of recurrence and survival arise tendency to complete resction and waiting for growthing more transplantation criteria.

Clinico-pathologic and Ultrasonographic features of abdominal tumors in Iranian children

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Background: Since the abdominal tumors are one of the common causes of childhood death, studying the clinico-pathologic features of tumors is important for early diagnosis. The aim of this study was to determine these features in Iranian children and to evaluate the accuracy of Ultrasonography in determining the abdominal masses in children.

Method: In this retrospective case series study data about the sex, age, primary chief complaint, physical examination, imaging report, Pathology finding of 156 children with abdominal tumor, who admitted to the children medical center from 2005 to 2011 were recorded and analyzed.

Results: Male to female ratio was 0.69. The most common type of tumor in this study was Willm's (37.5%) and Neuroblastoma (35.7%). The mean age in children with Willm's tumor and Neuroblastoma was 38.95 and 26.65 months old respectively. Ultrasonography has lower accuracy in patients with tenderness, children with Willm's tumor, female patients and children under 5 years old.

Conclusion:The observed differences in tumor type and distribution in comparison with previous studies may be attributed to genetic and geographic variations. In addition, this study shows that the accuracy of Ultrasonography in children with abdominal tumors depends on children's sex, age, pain and type of tumor.

A huge Cystic Mesenchymal Hamartoma of the liver in a 7 month infant

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BACKGROUND: Mesenchymal hamartoma (MH)is a rare benign de¬velopmental tumor of the liver, which is found in the first two years of life in about 8 percent of cases .MH account for 6-8% of liver tumor and 18% to 29% of benign liver tumors in pediatric.The most common benign tumor is hamartoma and MH is the second one. It is slightly predominance in male (3:2 M/F) . Its old names are lymphangiomas, bile duct hamartomas, mesenchymomas, psuedocystic mesenchymal tumors and cystic hamartomas. The right lobe of the liver is more involved. The histogenesis of MH has not been well understood but various numerical and structural chromosomal abnormalities was documented in literatures. Most lesions in ultrasonography and computerized tomography imaging contain well-marginated large mass with cysts of varying sizes or mixed solid and cystic tumor measuring from one centimeters up to 30 cm. It has two main variant, cystic and solid. In this literature we report very large sizes MH in a 7 month boy that in our literature review in this age this size of tumor are very rare and notable.

METHODS: A 7 –month- old boy presented with a very large non tender abdominal mass. On examination, the liver was palpable 7 cm below the right subcostal margin .Other systemic examination was normal. Laboratory studies of liver function were normal. Tumor markers level including: beta -human chorionic gonadotropin (β-HCG) and CA 125 were normal but the Alpha-fetoprotein (AFP) was more than 350 ng/ml. In the ultrasound examination there was a large liver mass with solid and cystic component and internal septation in the right lobe. There was no cyst or abnormality in the kidneys or other organ. A clinical diagnosis of malignancy was made because the huge size of cyst and its mixed component and for surgical planning we requested abdominal computed tomography. The CT scan showed a 350*270*240 millimeter large, complex solid –cystic mass with internal septation in the right lobe of the liver. The tumor was resected and its gross specimen is shown in figure 3 but it isn't the true size of it because the cystic part of mass aspirated by surgeon to shrink it before resection. A pathological diagnosis of a cystic mesenchymal hamartoma was made. Follow up was done by ultrasonography and blood analysis and up to one year after surgery, there are no signs of recurrence.

RESULTS: About one-third of primary pediatrics liver tumors are benign that can be of mesenchy¬mal or epithelial origin. [According to the original description by Edmondson (1956), MH was a cystic mesenchymal lesion, chiefly composed of connective tissue containing much serous fluid].Most of MH are in the right lobe of liver like our case.MH are vary in size. According to our literature review most of the large size of MH were seen in children older than one years old and we only find one case of MH in the 3 month old child that was 17 cm in size.We think that the present case of MH is rare for its very large size in a 7 month old child .Chang, H. J ,et al. in their case series demonstrate that MHs of the older patients were more commonly cystic and the solid form had a higher serum level of AFP but in our case a cystic MH were seen in a child with high level of AFP. The high level of AFP level in our case may be physiologic because its level not reaches the adult level until 8 months of age.

Some clinician believe that MH is subsequently decrease in size need to watchful waiting in symptomatic patients but in our case because the natural course of progressive enlargement and compression effect we decided to resects it.

CONCLUSIONS: Some clinician believe that MH is subsequently decrease in size need to watchful waiting in symptomatic patients but in our case because the natural course of progressive enlargement and compression effect we decided to resects it.

Abdominal Inflammatory Myofibroblastic Tumor: Report on Four Cases and Review of Literature

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Background: The Abdominal Inflammatory Myofibroblastic Tumor (AIMT) is a rare tumor with unknown etiology which usually occurs in children and adolescents. It is composed of

myofibroblastic spindle cells intermixed with inflammatory cells. We present four cases of

AIMT.

Cases Presentation: We herein present four cases of AIMT in different ages (range: 3.5 to 13

years) and in different organs (stomach, periduodenal, mesenteric, and colon). There were two

females and two males. The main symptoms were abdominal pain/mass/obstruction, vomiting,

and weight loss. In all four patients, diagnosis was made by laparatomy and pathologic

examination of excised mass lesion. Three patients underwent complete excision and no

residual disease was present, one patient received chemotherapy due to tumor recurrences.

The patients were followed up in average for four years.

Conclusion: As the imaging and laboratory tests are non-specific, the diagnosis of AIMT is

rarely made before surgery. AIMT should, therefore, be considered when a mass arises in an

unusual location in the pediatric age group. Complete surgical resection should be performed

whenever possible and the child should be kept on long-term follow-up.

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Risk factors and prevention of cancer in infants and children

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Unfortunately cancer occur at any age including during infancy. According to recent statistic, roughly 23 of every 100,000 babies are diagnosed with cancer each year.

Mother smoking during pregnancy and drinking alcohol, exposure to insecticides, and have neen exposed to farm animals and to sick pets more chance to take babies cancer.

In developing countries, approximately 1/600 children develop cancer before they are fifteen years old.

Half of all childhood cancer are diagnosed during of first five years of life, the 30% of cases are mainly Embryonaltumorsincluding:

Neuroblastoma, Wilms tumor, Embryonal Rhabdomyosarcoma, Retinoblastoma and Hepatoblastoma. Various syndromes confer an increased risk of childhood cancers.

Children with WAGR syndrome have the 33% chances of developing Wilms tumor. Children with Beckwith- Wiedemann syndrome have a higher risk of developing Wilms, Hepatoblastoma and Neuroblastoma.

Cancer maybe a genetic disorder but chances of getting cancer are affected by the environment in which baby live, women should stop smoking before they start trying to conceive, because that increase the chances of the child having a low birth weight. Children born lighter than average often then put on weight quickly, but in the form of fat instead of muscle and develop fat around the middle, which raise cancer risk.

In the child's early life, breast feeding has recognized anticancer effects. Baby should not eat any solid food until six month old. Nor be given any sweet drinks especially sugar-laden drinks which promote weight gain.

Exposure to one of these factors is the risk of cancer in children:

□□□Abdominal XRY during pregnancy

□□□Radioactive exposure like Chernobyl nuclear reactor disaster in 1986

□□□Non ionizing radiation

□□□Electric and magnetic fields In England studied that children who live in within 600 meters of a high voltage overhead electric power line, magnetic field make childhood leukemia. Australia and New Zealand have a relatively high incidence of childhood Melanoma which maybe due to UV radiation. Infection: Liver carcinoma (Hepatitis B) and Kaposi sarcoma (HIV and HHBV) these association can only for a tiny proportion of childhood cancer in western countries.

Evaluation the need for intensive care after surgery in children with Wilms' tumor

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BACKGROUND: Childhood cancer may need to have intensive care. This need is felt after the surgery and tumor removal more and more. One of the cancers that may require intensive care after surgery has been Wilms' tumor. The aim of this study was to evaluate the need for intensive care after surgery in children with Wilms' tumor.

METHODS: This cross-sectional study was done through reviewing of documentation for 42 children that undergo Wilms' tumor surgery and were registered their need to take intensive care. Results were analyzed using spss17 statistical software.

RESULTS: The results showed that after surgery 5 of the 42 children were transferred to intensive care unit. 1 case of them was a neonate with fetal origin Wilms' tumor that has been hospitalized in the NICU. 4 Children who have been in the PICU had underline disease in vital organs such as the lung, heart and liver.

CONCLUSIONS: However, require to intensive care in this study is not impressive, reserve ICU bed for surgery in children who have underlying disease in vital organs is essential.

Identified symptoms of Wilms' tumor by parents and the interval between the detection of symptoms to see a physician

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BACKGROUND: Wilms' tumor is a malignancy of the kidney that occurs in childhood. The symptoms caused by the tumor are identified mostly by the parents. Urgent action after identify marks are important in the prognosis. The aim of this study was to determine the type of symptoms that have been identified by parents and the interval of time between the detection of symptoms to see a physician.

METHODS: This cross-sectional study was conducted through interviews with parents and reviewing of documentation for children that undergo Wilms' tumor. Results were analyzed using spss17 statistical software.

RESULTS: The results showed that the most of cases were admitted with complaints of touching the mass by parents and next in rank was hematuria and abdominal pain, 1 hemihypertrophy and 2 cases are detected incidentally by a physician. The parents have to see a physician with significant interval after the identification of symptoms by themselves.

CONCLUSIONS: As the child of an immediate visit to the doctor Profited, It is essential that awareness through public education be given to families

Bilateral Wilm's tumor or acute lymphoblastic leukemia?

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A 14 months old years girl, was admitted in a general hospital with abdominal distention and irritability. In imaging study with sonography,bilateral enlargement of kidney was reported.

At the first admition , a biopsy of the left kidney was taken under general anestesia with surgery. The pathology report of sample biopsy was bilateral wilm's tumor and patient was candidate for chemotherapy.

Because , patient family were not satisfied with this diagnosis and the course of treatment; she was brought to a hospital in another city; the physician was become suspicious after reviewing the CBC;diff which was as follows: white blood count=11600; 72%lymph 18%poly ,Hemoglobin=11.3; platelets=17000. So bone marrow aspiration was done which suggested leukemia. After all she was brought to Secondary hospital for the course of treatment. After another bone marrow aspiration and flowcytometry; Pre-B-cell leukemia became the definite diagnosis.

The flowcytometry revealed tranlocation (2,14); deletion(6)q; translocation t(9,22)(q34,q11).

The case became a high risk ALL; due to :

1)t(2;14)

2)t(9,22)

3)Infantile leukemia

4)Bilateral infiltrative kidney masses and BMA suggestive of acute lymphoblastic leukemia.

The first CSF analysis was normal. The pathology of the biopsy done in Tehran with immunohistochemistry showed

CD10+ in tumor cells, Ki67 + in 85%, CD19 diffusely positive and LCA positive in tumor cells; all these leading and confirming are diagnosis: lymphoma or leukemia infiltrative with B cells.

So the treatment was started. Up until today she has had 17 sessions of chemotherapy and her latest CBC is: white blood cells=6300 (44% lymph), hemoglobin =10.3 and platelet count was 465000.

Discussion:As for this case report; we wanted to show ; although the presentation of ALL in pediatric medicine is rare as an bilateral abdominal mass, but it could happen and it can be mistaken with wilms tumor; which is common in early ages and its most common presentation is abdominal mass. Therefore the most common presentations should not make us forget to look

in the basic lab results; such as a simple CBC. With covering all the aspects of a complete history and physical examination and reviewing all the lab results; we can make a definte and true diagnosis.

In our case; we wanted to report an atypical case presentation of acute lymphoblastic leukemia, which was misdiagnosed with bilateral wilm's tumor.

Unusual thoracic tumor and treatment in educational center of taleghanigorgan1394

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BACKGROUND: Mediastinal tumors are occasionally diagnosed however most of them remain silent during infancy. Plain radiographs of the thorax or ultrasonography may show widening of superior mediastinal shadow or mass in either hemithorax. CT scan provides information about the location of tumor its cystic or solid nature depicting calcifications, necrotic areas and widening of spinal foramina in cases with intra spinal extension.mediastinal Hodgkin is less frequent and occurs more often in adolescents.

METHODS: Patient is 13 years old boy that presented with fevere coughing and weight loss for a month prior to admission .ct scan showed large well defined posterior mediastinal mass with 65mm size and which came in close contact with great vessels as SVC.patient underwent right postrolateral thoracotomy and complete excision of tumor.the patient post operative evolution was favorable .the histopathologic result showed the Hodgkin lymphoma .

RESULTS: Hodgkin lymphoma may be localized primarily in the mediastinum and may causes compressive effects.Biopsies of extra thoracic nodes are sometimes possible. If not the chamberlain operation or thoracoscopy should be used. in localized cases complete excision is possible. CONCLUSIONS: Surgery is not usually the primary treatment of Hodgkin lymphoma except in very localized cases. However it may occasionally be needed in children

A comparison of axilary versos anterior chest wall placement of ports in cancerous pediatric patients in educational center of taleghani gorgan 1394

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BACKGROUND:A few data are available from analyses of complications of central venous access ports for chemotherapy in children. We compared morbidities in patients with surgically implanted central venous ports that were placed in the subcutanouse tissues of the mid axillary line at the anterior border of latissimus muscle versus the anterior chest wall.

METHODS:Between 2013 and 2016 a total 64 patients with leukemia and other cancers were taken to operating room for placement of central venous port for delivery of chemotherapy .port location was determined with patient and surgoen preference .patient demographics were collected. Complications identified as thrombosis ,infections ,port and catheter problems. Basic descriptive statics were generated. patients with axillary ports were then compared to those with chest wall ports by appropriate t- test or x2 tests.

RESULTS: During this period 24 of 63 ports were placed in axillary position. There was no difference in thrombotic and infective complications between two groups but cosmetic results in axillary line was better than anterior chest wall position and breast deformity was lower in axillary position .

CONCLUSIONS: Subcutanouse ports can safely be placed in mid axillary line.

Axillary ports spare the negative cosmetic outcomes and breast deformity of chest wall ports

Gross tumor Resection inAbdominal Neuroblastoma

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Neuroblastoma is the most common abdominal malignancy of childhood, occurring in about 1 in 10000 children.In general these tumors are highy malignant and the majority (about two-thirds) present with locally advanced or metastatic disease.The most commonly used staging system for these tumors is INSS. A consistent surgical approach is used. This entails a systematic dissection of the involved vessels prior to removal of the tumor.Macroscopically complete or near complete tumor clearance are achieved in the majority of the cases.The described technique is safe and reproducible and allows tumor clearance in the majority of affected children. In addition, a number of biological markers – MYCN amplification,!p deletion, 17 q gain- have further defined inherent tumor behaviour.Tumors which show these features are more aggressive and are associated with reduced survival. A number of biochemical markers in the serum also help to define tumors With more aggressive behaviour- elevated levels of LDH,NSE,and ferritin denote a worse outcome. The aim of the technique is to display all the major vessels as they traverse the tumor.*En bloc* excision is clearly not an option when vascular encasement is present. these tumors don"t usually invade beyond the tunica advantitia of major blood vessels. Consequently, a plan of dissection may developed between the tumor and tunica media.This is most easily accomplished using a scalpel. There are three phases to the procedure- vessel display,vessel clearance,and tumor removal. The first phase is to display part of the wall of each of the vessels which traverse the tumor,in continuity. The vessels are subsequently cleared circumferentially and mobilized from the tumor,after which the tumor may be removed. These phases are not absolutely distinct during the course of a long operation. For those with an abdominal primary tumor, the abdomen is opened through a transverse supra-umblical incision. Full laparotomy is performed before evaluating the extent of the disease.On whichever side the tumor arises, the colon is reflected medially to display the tumor.On the left side,, the spleen and the pancreas are also mobilized and all the viscera are placed in an intestinal bag.The use of table mounted retractor is of considerable benefit. Dissection is commenced distally below the lower limit of the tumor .The surgeon and assistant each pick up the tunica adventitia of blood vessel and this is then incised along the middle of the vessel to enter the sub adventitial plane.This establishes the plan of dissection for the reminder of the procedure. In this manner ,the dissection advances to the common iliac artery ,then along the distal aorta ,always incising along the middle of the artery in the 12 o"clock position.The vessels encountered proximally are inferior mesenteric artery,gonadal artery,left renal vein,left renal artery ,SMA ,coeliac artery, and the median arcuate ligament.The dissection continues until the superior limit of the tumor is reached.If the surgeon has reached this stage of the operation whereby part of the aortic wall is visible in continuity from proximal to distal limits of the tumor, then the likelihood is that the tumor can be excised. Placing a sling around the thorasic aorta provides reassurance for the reminder of the operation. In general , clearance of the aorta follows clearance of the visceral arteries.It is not unusual to divide a number of lumbar arteries. The aim is to preserve as many as possible.The typical appearance folloing tumor clearance is that ,the coeliac,its branches, SMA,IMA,and IVC are clearly seen.No remaining tumor is present.

Post operative diarrhea is a probem in 30% of patients.It is due to SMA and Coeliac arteries clearance. It persist in the majority. Prononced ascites,adhesion obstruction aortic injuries which require patch repair, troublesome caval injury ,late loss of kidney , effects on potency and ejaculation are in the list of complications.

Conclusions: The technique reported here allows a planned, systematic and consistent approach to resection, regardless of the position of the tumor. It is easily taught and learned and has enabled excision of the majority of the tumors.

Functional Adranal Masses in Children: Two Case Report

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Neuroblastoma accounts for more than 90% of adrenal masses in children.The reminder 10% include functional and nonfunctional and metastatic masses.The functional masses include adrenal adenoma, adrenocortical carcinoma(ACC) and pheochromocytoma.

Patients who have an incidentally discovered adrenal mass should undergo hormone evaluation, including dexamethasone suppression test,aldostrone levels,and measurement of plasma free metanephrines. Surgical treatment is indicated for all functional adrenal cortical tumors and pheochromocytoma.

ACC accout for 0.1% of all childhood malignancies.There is F/M predominance of about 2:1.The tumors occur equally on the right and left sides and are hormonally functional in 80% to 100% of cases.Half of ACC occurs in children younger than 5yrs of age. Southren Brazilians have a 15 fold greater incidence of ACC compared with other populations.

Virilization is the most frequent presentation (66%)' whereas the reminder of children will usually have Cushing"s symptoms.Virilization is secondary to secretion of the adrenal androgens.Featurs include axillary and pubic hair ,deepening of the voice. Acne , a rapid acceleration of height, hirsutism, enlargement of the penis or clitoromegaly, and development of body odor.Feminization may occur in 2% to 25% of patients and results from an overproduction of estrogens, particularly estradiol. Nonfunctional tumors in children are infrequent. Only about 5% of pediatric adrenocortical tumors produce no clinical evidence of hormone excess. Accordingly, these patients usually are first seen late in disease with abdominal pain or fullness.

Evaluation should be the detection of elevated androgens.These include measurement of plasma T, urinary and plasma DHEA,and DHAE-s.Urinary 17-ketosteroids are also important because usually two thirds of 17-ketosteroids are derived from adrenal androgens.

Imaging evaluation shou ld be plain abdominal x-ray , ultrasound,bdominal CT ,MRI,and finaly adrenal scintigraphy.In scintigraphy bilateral symetric images indicate hyperplasia,unilateral uptake suggests adenoma,and nonvisualisation is suggestive of carcinoma.

Surgical resection offers the only chance to cure.In the case of extensive disease wide en-bloc resection of the tumor,lymph nodes and involved organs is indicated. For less extensive disease MIS have been advocated.

Pediatric series report the incidence of t responses have been reported between 30%andumormetastasis at diagnosis as being between 5% and 64%.Mitotane is an adrenolytic agent that selectively causes adrenal gland necrosis and has been the most widely used Cht agent.It is used for metastatic disease ,for incompletely excised tumors, and for hormonal effects of the tumors.In the pediatric literature ,tumor responses have been reported between 30% and 40%.Additional regimens

That have been shown some promise include the combination of cisplatin,etoposide,and taxol.The role of RT in children has not been well established.

Patients who are untreated for ACC have a mean survival of 2,9 months.These tumors are highly lethal, with nonfunctional tumors demonstrating a worse prognosis.A delay in diagnosis leads to a worse prognosis as well. The prognosis depends on the child's age and the resectibility of the tumor.

In one review of 55 children with ACC the survival rates were more than 67% if the tumors were completely excised, but no survivors were found after partial resection.

Case 1: A 2 yrs of old boy reffered for treatment of LIH. The physical exam revealed the stimulated penis in this boy. Endocrine w/u demonstrate increased androgens. The ultrsound scan detected a 5*7*8 well defined adrenal mass in the left side. The tumor was removed completely without rupture.The histologic study revealed adrenal cortical tumor. Case 2: A 8 months of old girl reffered for treatment of abdominal mass inLUQ.The pubic area was hairy. She had been operated for biobsy by needle in another hospital,and the response was pheochromocytoma. She had not the clinical sign of Cushing disease.The ultrasound scan confirmed the adrenal mass.She was operated and the adrenal mass with adherent splenic flexure segment of the colon was removed without rupture. The histologic study confirmed adrenal cortical tumor.

Large Adrenocortical Carcinoma: Case Report

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Introduction:Adrenocortical carcinoma (ACC) incidence is about 1/1,000,000 people. ACC in childhood is a rare case in which the patient's symptoms are different as adulthood, mostly present with virilization. The childhood ACC also has better prognosis after resection. Even though, after complete resection recurrence is common.

Case Presentation: A 4 years old girl came with chief complaint of gradual hoarseness and hirsutism in face, genitalia, and lower extremities from 4 months ago. She had family history of lung cancer in her uncle, breast cancer in her cousins, and also had a cousin with lung, stomach, bone and brain malignancies. The abdomen was not distended and no organomegaly found. Abdominal ultrasonography and CT scan revealed 9.5*10*10 cm mass in right adrenal gland, left adrenal gland spared. Biochemical parameters are within normal range. Free plasma metanephrin was 19 pg/ml (normal range of <90 pg/ml) and 24h urine VMA level was 2.55 mg (normal range of 1-2.6 mg for 3-6 year old). She underwent surgery by suspect of right adrenal mass. Intraoperatively the mass found so adhesive to right lobe of liver, diaphragm, and also inferior vena cava (IVC) which gently removed and the mass completely excised.

Results: The histopathological examination reported neoplastic proliferation of poleomorphic cells with round, temporary bizarre nuclei and poorly-circumscribed cytoplasm in a vague nodular manner. Vimentin and Melan A were positive while Synapthophysin, Chromogranin and CK were negative through IHC microscopic evaluation.

Conclusion: ACC is a rare, poor prognosis neoplasm (esp. in adults). Although positive family history of malignancies was not very provocative in ACC cases, the report shows great relation. Regarding high rates of recurrence in children serial ultrasonography profoundly suggested to patient.

A case report: Pelvic Neuroblastoma with common iliac artery and vein involvement

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A 3, year old boy presented with lower abdominal and pelvic mass. Radiologic investigation has been shown a 106×77×75 mm retroperitoneal solid mass with aorta and IVC displacement. In core needle biopsy Neuroblastoma was diagnosed. After 8 courses of chemotherapy tumor size was reduced to 75×60×45 mm but left Common iliac artery and right common iliac vein were involved by tumor, and also left common iliac artery had been displaced to anterior. Patient was candidate to operation. After releasing common iliac artery and vein from the tumor, complete resection was done.

Huge neurofibroma of neck and mediastinum

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Here we present a 7 years old girl with a huge cervical mass since 2 month prior to admission, the patient had no respiratory destress but had hoarseness

chest x ray showed a huge mediastinal mass, biopsy was taken which was in favor of neurofibroma and no malignancy was seen. CT scan and MRI were performed. the patient underwent operation and debulking of the neck tumor was performed, the mass was about 10 x10 cm and was very hard causing shifting of the trachea, six month later the neck was re explored and had severe adhesion to carotid, internal jugular vein, trachea, thyroid, esophagus and surrounding muscles.

the mass was excised however the extension to the mediastinum left intact, 3 month later right posterolateral thoracotomy was performed and the huge posterior mediastinal mass was excised.

One year later re exploration of the neck was performed and the huge mass 8x10 cm was dissected from the carotid and subclavian vessels were performed with Ligasture and the mass was almost totally excised. all pathologies were in favor of neurofibroma and the last followup after six month showed no recurrence of the tumor.

Review of Patients with Teratoma in Last 8 Years

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The files of patients who were treated as teratoma since 2008 in Shiraz University of Medical Sciences is reviewed. There were 52 patients, 38 female and 14 male. Most of the patients where operated on less than one year of age.

36 patients had mature cystic teratoma, 8 immature teratoma, 6 germ cell tumor and 2 had lipoma, 7 patients needed laparotomy in addition to sacrococcygeal approach but for 45 patients sacrococcygeal approach was the only procedure which was performed. Of 7 patients who had laparotomy 5 patients had either gem cell tumor or immature teratoma. All patients with gem cell tumor and 4 patients with immature teratoma had increased alfa feto protein and needed chemotherapy and responded well to adjuvant therapy.

The followup of these patients where performed via calling them. there where 2 mortalities and 4 patients had reoperation due to recurrent of the tumors, all other patients where continent and had no neurologic deficit. This study showed that most patients who presented in older age had more chance of malignancy and also the patients with intra-pelvic mass also had more chance of malignancy and need more aggressive therapy.

Postoperative Pain Control

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Accompanying the development of the new drugs , Pediatric Postoperative Pain Control (PPOPC) has developed . But it is still remained undertreated because of Difficulty of pain assessment and apprehension regarding cardiorespiratory depression. Pain Control is important to improve both clinical outcome and patient comfort. For more successful postoperative pain control, it control should begin with Preoperative assessment of anxiety, assessment of pain and assessment the efficacy and safety of analgesic techniques and drugs. Factors that affect the PPOPC:Anesthesia and operative fields are unfamiliar and unpleasant environments for children, Separation from mother, hunger, fear of strange places, and perioperative pain can cause stress and result in indistinct behavioral and physiological changes.Steps for PPOPC: 1-Preoperative assessment of anxiety, 2-Assessment of pain and the efficacy and safety of analgesic techniques and drugs in pediatric patients.3-Postoperative pain assessmen 4 Pain management Postoperative Pain Assessment:There are numerous assessment tools, but many of them are, complicated, not well validated and difficult to use in clinical practice. For choosing pain assessment tools, should take consideration into; Age, stage of development, clinical condition, cultural appropriateness, language, familiarity, and training requirements of tools. However, no individual tool can be universally recommended in children of all ages.Physiologic changes as a tool for Pain Assessment:Heart rate, which is the simplest and the most appropriate indicator. Respiratory rate, blood pressure, sweating, decreases in oxygen saturation and vagal tone. Sometimes the Physiological parameters are not reliable and may be influenced by associated clinical condition such as: sepsis, distress, hypoxemia, hypovolemia, and fever. Behavioral changes: is another tools for pain assessment, but these are also influenced by other states of distress, such as; hanger, fear, anxiety. Composite tools for assessment For reducing these effects using both physiologic and behavioral changes, for assessment may be helpful, such as:The COMFORT scale, Children Hospital of Eastern Ontario Pain scale, Face, Leg, Activity, Cry, Consolability (FLACC). No single composite scale is clearly superior to the others. One standard approach of assessment of pain is QUESTT: Q – Question the child U – Use pain rating scales E – Evaluate child's behavior S – Secure parent's involvement T – Take cause of pain into account T – Take earliest action Pain management: There are many different modalities to treat the pediatric pain, but before opting for an appropriate modality of pain relief, one must evaluate; the relative risks or benefits, it's analgesic efficacy, safety, side effects, costs and the course of recovery. The child should be prepared properly for that particular method of pain relief. A good psychological preparation of the child as well as parents, proper premedication and smooth anesthesia course always helps in reducing the anxiety and needs of pain medications in the post-operative period. Realistic aims for post op pain management are:To recognize pain in children, to prevent pain where it is predictable, to minimize moderate and severe pain safely in all children, to bring pain rapidly under control, to continue pain control after discharge from hospital, Prevention of pain whenever possible and using multi-modal analgesia work well for nearly all cases and can be adapted for day cases, major cases, the critically ill child, or the very young patient. Many acute pain services use techniques of concurrent or co-analgesia based on four classes of analgesics, namely; local anesthetics, opioids, non-steroidal anti-inflammatory drugs (NSAIDs), and acetaminophen. In particular, a local/regional analgesic technique should be used in all cases unless: there is a specific reason not to use and the opioid-sparing effects of Local anaesthetics, NSAIDs, and Acetaminophen (paracetamol) are useful. Indeed, for many day-case procedures, opioids may be omitted because combinations of the other three classes provide good pain control, in most cases regional anaesthesia is nearly always conducted in anaesthetized children, but some high risk neonates have lower perioperative morbidity after inguinal surgery when awake spinal anaesthesia is used.The treatment modalities:A:: Child should be made comfortable and less distressed, before surgery and during hospital stay. these measures include; presence of parent with the child, nursing in a comfortable environment, allowing the child to adopt most comfortable position and feeding if permissible B: Systemic drug therapy: Non-narcotic analgesics • Paracetamol (Acetaminophen 15-20 mgkg oraly every 4 hours •Ibuprofen 4-10mgkg/dose every 6-8 hours in >6 m/age • Diclofenac 1-1.5 mgkg / 12 hourly •Ketorolac 0.2 -0.5 mg/kg q6h for 48h iv or im • Ketamine <1mg/kg iv Narcotic analgesics •Morphine 0.1 mg /kg im or 0.05 mg/kg iv infusion •Codeine 1 mg/kg oral •Pethidine 1.5-2 mg/kg •Fentanyl 1-2 mg/kg Intravenous analgesia using opioids; After an intravenous bolus dose of 0.1 mgkg injection morphine, the child gets relief from pain for 1-3 hours. Intravenous analgesia can be given by two different ways as i) Continuous I. V. infusion : Initial dose of 0.05 mgkg IV morphine, followed by an infusion of 0.015-0.025 mgkg /hr, in children < 6 months and 0.025-0.030 mgkg /hr, in older children. Apnea monitors' and pulse oximeters should be used specially, if Opioids are being used in infants < 6 months of age, or In children with acute or chronic respiratory dysfunction etcii) Ppatient controlled analgesia (PCA). Following appropriate pre-operative teaching, children > 6 years of age can learn to use a PCA pump, It is important that the child understand the expectation of PCA is pain control, not elimination of pain. C. Regional techniques:Epidural injection Sacral epidural Intercostal block Nerve block D. Non-pharmacological approaches: Various non-pharmacological approaches eg. psychological interventions like hypnosis, behavioral therapy, Acupuncture, Transcutaneous electrical nerve stimulation (TENS) have been described for post-operative analgesia.

Sacrococcygeal teratoma: 10-year experience in Tabriz Children Hospital

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Background: To evaluate our experience with 35 patients with sacrococcygeal teratoma (SCT) in our hospital over a period of 10 years between 2004 and 2014 and determine the outcome of the management and recommendations for treatment strategies.

Patients and methods: A retrospective study was conducted at our Pediatric Surgical Unit, Tabriz Children Hospital. The medical records were reviewed for age at presentation, clinical manifestations and investigations, time of surgical approach, histopathology, recurrences, bladder and anorectal function, and cosmetic outcome.

Results: Thirty-five patients with SCT were referred to the Pediatric Surgical Unit. The time of referral was as follows: immediately after birth in the case of ten patients; during the first week for 17 patients (one of them died before surgery because of hemodynamic instability and other associated congenital anomalies and was excluded from the study); later in infancy for 22 patients; and at one and half years of age for one child. The lesion was excised in the case of 34 patients. Teratomas were of type I (n=5), type II (n=15), type III (n=12), and type IV (n=2) (Altman's classification). The age of patients at surgery ranged from 2 days to 1.5 years. Histological analysis of results revealed mature teratoma (n=24), immature teratoma (n=7), and malignant teratoma (n=3). The coccyx wasn't removed in three cases during the early period of the study. The follow-up period ranged from 3 months to 10 years. Recurrence occurred in five (12%) cases, wound infection in four (9.7%), and diarrhea in two (4.8%) cases. The a-fetoprotein level was high in 30 cases and was normal in 4 patients; it decreased after excision. Fetal diagnosis was made in 12 cases by means of a prenatal sonographic scan.

Conclusions: Prenatal diagnosis of SCT is very important and it is recommended to save the baby from obstructed labor. Early diagnosis allows early surgical intervention and avoids malignant transformation. The coccyx should be excised to decrease the risk of recurrence. Skin flap modification is feasible for large teratomas with healthy skin.

Congenital Mid-Face Teratoma – A Case Report

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Head and neck teratoma is a rare entity. Its prognosis mostly depends on the risk of neonatal respiratory distress, its extension and potential malignancy. Surgical management must be as complete as possible to avoid recurrences and malignant transformation.

We describe the case of a female neonate with a very large facial mass, deforming the maxilla, etmoid, and frontal bone, without any intracranial extension. This teratoma was discovered at 36 gestational weeks by ultrasonography. Fetus had multiple cardiac anomalies (ASD, VSD, sever PS, and tricuspid atresia) at the same time. C/S labour performed at 40 gestational weeks because of maternal HTN and respiratory distress. At birth a huge mid-facial mass was evident with a grossly deformity of nose and maxilla. Palpation showed a heterogeneous mass with scattered hard nodules. The tumour was removed at the age of 8 days because of respiratory problems. The postoperative course was uneventful. The diagnosis was mature teratoma.

Pediatric Germ Cell Tumors; A 10-year Experience

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BACKGROUND: The aim of this study was to evaluate the outcome of germ cell tumors in patients admitted to our center during a ten year period

METHODS: In a retrospective descriptive study, patients with the pathological diagnosis of germ cell tumor (GCT) were included. All records were evaluated and patients followed by personal visit in clinic or phone call. Data regarding age, sex, tumor site, bio-chemical assay, pathology, treatment and outcomes were gathered. For qualitative variables we computed frequency and percentage and for quantitative variables, mean and standard deviation. Survival analysis was performed using Kaplan-Meier. All statistical analyses were performed by SPSS version16.0.

RESULTS:Forty four patients consisted of 32 girls (72.7%) and 12 boys (27.3%). Their median age was 23 months. The most common pathological tumor types were 18 (40.9%) mature teratomas and 14 (31.8%) yolk sac tumors. Extra gonadal tumors were more prevalent (32 cases) and consisted of 21 (47.7%) sacrcoccygeal, 7 (15.9%) retroperitoneal, 2 (4.4%) mediastinal and 2 (4.4%) cervical tumors. In gonadal tumors 9 patients had ovarian and 3 patients testicular involvement. Staging at the time of diagnosis revealed stage one in 23 (52.3%) cases. All patients were treated surgically and the most common procedure was total resection in 41 (93.2%) patients. Fifteen (34.1%) patients received chemotherapy. In follow-up 31 (77.5%) patients were in complete remission, 9 (22.5%) had died, and 4 cases did not appear to follow-up visits. The median survival was 16 months (IQR 4-49 months). The highest mortality rate was found in patients with yolk sac tumors (8 of 13 cases).

CONCLUSIONS: The patients with extra-gonadal GCT and a high AFP level have the worst prognosis and lower survival rate. Combination of surgery and chemotherapy can lead to a better prognosis.

Testicular tumours in children: a single-institutional experience

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OBJECTIVE: To report our experience of testicular and paratesticular tumours in children in our centre.

Testicular tumours in children are rare, and historically yolk sac tumour has been described as the most common lesion in children.

PATIENTS AND METHODS: We reviewed retrospectively the records of children treated for testicular tumours from 2006 to 2015. The patients' age, clinical presentation, side of tumor, diagnostic procedures, treatment methods, histopathological findings, and outcome were recorded. Patients aged>168 months and those with metastatic lesions were excluded.

RESULTS: In all, 13 patients met our criteria, with a mean age of 47 months (range 2 month to 13 years). Pathological analysis revealed teratoma in one patients, yolk sac tumour in five, , and paratesticular rhabdomyosarcomas in four,malignant high grade lymphoma in one,Leydig cell in one,Sertoli cell adenoma in one. The most common clinical presentation was a painless testicular mass. Depending on the clinical presentation and pathology, scrotal ultrasonography, tumour markers (alpha-fetoprotein and beta-human chorionic gonadotrophin), and staging computed tomography (CT) were obtained in all patients. All patients had a radical orchidectomy. Five patients had elevated tumour markers that normalized after orchidectomy. CT revealed retroperitoneal adenopathy in one patient with rhabdomyosarcoma that underwent retroperitoneal lymph node dissection.11 cases were stage I ,one case stage II and one case stage III.12 cases were right side only one case left side . Chemotherapy was administered to all patients except one case with yolk sac tumor and teratoma.

CONCLUSION: most of the lesions were malignat tumours, with the most common histological subtype being yolk sac tumor. paratesticular lesions occurred at a significant frequency. frozen-section analysis at the time of surgery confirms a benign lesion but we would continue to advocate an initial radical inguinal approach in doubtful cases.which time testis-sparing could be considered if the preoperative evaluation was favourable,

Neuroectodermal tumors (PNETs)

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Primitive neuroectodermal tumors (PNETs) are a group of highly malignant tumors composed of small round cells of neuroectodermal origin that affect soft tissue and bone. Primitive neuroectodermal tumors (PNETs) exhibit great diversity in their clinical manifestations and pathologic similarities with other small, round cell tumors. This has made classifying this family of tumors challenging and controversial. Batsakis et al (1996) divided the primitive neuroectodermal tumor (PNET) family of tumors into the following 3 groups based on the tissue of origin: CNS primitive neuroectodermal tumors (PNETs) - Tumors derived from the central nervous system Neuroblastoma - Tumors derived from the autonomic nervous system Peripheral primitive neuroectodermal tumors (pPNETs) - Tumors derived from tissues outside the central and autonomic nervous system Peripheral primitive neuroectodermal tumors (pPNETs) are also classified as part of the Ewing family of tumors (EFTs); peripheral primitive neuroectodermal tumors (pPNETs) and Ewing family of tumors (EFTs) are often referred to interchangeably in the literature. Generally, Ewing family of tumors (EFTs) and peripheral primitive neuroectodermal tumors (pPNETs) represent different manifestations of the same tumor and have similar genetic alterations. Ewing sarcoma, however, is more common in bone, while peripheral primitive neuroectodermal tumors (pPNETs) are more common in soft tissues. Immunohistochemical and cytogenetic studies suggest that these tumors all have a common origin

The following tumors are classified as peripheral primitive neuroectodermal tumors (pPNETs): Ewing sarcoma (osseus and extraosseous) Malignant peripheral primitive neuroectodermal tumors (pPNETs) or peripheral neuroepithelioma of bone and soft tissues Askin tumor (peripheral neuroepithelioma of the thoracopulmonary region) Other less common tumors (eg, neuroectodermal tumor, ectomesenchymoma, peripheral medulloepithelioma) Based on molecular cytogenetic analysis, both EFTs and peripheral primitive neuroectodermal tumors (pPNETs) are known to share the same reciprocal translocations, most commonly between chromosomes 11 and 22. Further advances in immunohistochemical analyses have helped further distinguish PNETs and Ewing family of tumors (EFTs) from other small, round, poorly differentiated tumors, including rhabdomyosarcoma, neuroblastoma, and lymphoma. Peripheral primitive neuroectodermal tumors (pPNETs) often exhibit aggressive clinical behavior, with worse outcomes than other small, round cell tumours. Clinical feature Most peripheral primitive neuroectodermal tumors (pPNETs) manifest in the thoracopulmonary region (Askin tumor), pelvis, abdomen, and extremities. Surgical management:Obtaining a complete resection of disease with negative margins is paramount in surgically treating primitive neuroectodermal tumors (PNETs) in the head and neck. In some cases, however, the aggressive nature and diffuse spread of these tumors precludes complete surgical excision. Furthermore, complete surgical resection may not be possible when vital structures are involved. Case report:A 13 years old girl was referred to our centre with abdominal wall mass from 9month ago, the tumour gradually has grown up and in serial ultrasound exams was diagnosed as umbilical hernia . In our hospital the patient was operated on.The tumour was 5*3.5*2.5 cm tan-red tissue with nodular surface and the diagnosis was malignant round cell tumour ,compatible with extra skeletal EWING sarcoma(PNET).

Hematopoietic stem cell transplantation for pediatric solid tumors

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While advances in the treatment of pediatric cancers have increased cure rates, patients with metastatic, refractory, or recurrent disease continue to have poor prognoses, and these "high-risk" patients are candidates for more aggressive therapy, including autologous HSCT, in an effort to improve event-free survival (EFS) and overall survival (OS).Prior to the introduction of high-dose chemotherapy (HDC) with autologous stem cell rescue (also called autologous HSCT), marrow tolerance was the limiting factor in the escalation of chemotherapy for the treatment of malignancies. With the ability to safely harvest, store and re-infuse a patient's own hematopoietic stem cells, doses of cytotoxic therapies for cancer could safely proceed beyond marrow tolerance, thereby allowing more intense treatment of certain malignancies. Neuroblastoma is the most common extracranial solid tumor of childhood.Conventional treatment of children with high risk neuroblastoma has generally resulted in a DFS of less than 20%.Efforts to improve outcome, focused on intensifying induction therapy in addition to using high-dose therapy with autologous Stem Cell support for consolidation, resulted in a 3-year DFS of up to 43% . While clearly demonstrated to improve outcomes in patients with metastatic neuroblastoma, autologous hematopoietic stem cell transplantation is also frequently used to treat patients with other high-risk diseases such as Ewing sarcoma, osteosarcoma, rhabdomyosarcoma, Wilms' tumor, retinoblastoma, germ cell tumors, lymphomas and brain tumors. CNS tumors comprise the second largest group of pediatric cancers. Because the prognosis is dismal for those children failing surgery ,radiation therapy,and/or conventional chemotherapy, autologous HSCT allows for escalation of chemotherapy doses above those limited by myeloablation and has been tried in patients with high-risk brain tumors in an attempt to eradicate residual tumor cells and improve cure rates. Patients with solid tumors with less than a 30% chance of long-term survival with conventional chemotherapy, only if a dose–response relationship to chemotherapy is established, should be considered for autologous or allogeneic (if bone marrow is invaded) stem cell transplantation in remission or for minimal residual disease. Table 1. lists the high-risk solid tumors that may respond to intensive therapy followed by autologous stem cell transplantation.

Supratentorial astrocytoma	Recurrence
Medulloblastoma	Recurrence
Ependymoma	Recurrence
Ewing sarcoma/PNET	Recurrence
	Primary metastatic
Rhabdomyosarcoma	Recurrence
	Primary stage IV
Neuroblastoma	Recurrence Stage IV n-myc positive
Wilms tumor	Recurrence
Retinoblastoma	Recurrence Primary extraocular

Epidemiological features of Iranian common pediatric malignancies: literature review of past and current data

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Introduction Population based cancer registries as the most important database of malignancies revealed that the common types of pediatric cancers accounts for leukemia (34%), brain tumors (23%) and lymphomas (12%). Other types of childhood cancers are less common.

Worldwide estimations show that pediatric malignancies has incidence of more than 175,000 cases per year annually. The mortality rate of these patients nearly accounts for 96,000 child per year. The highest variations in incidence of these patients occurs at the time of comparing high income countries with low income ones. The mortality rate of developed countries is 20% of cases, unfortunately in low resource and poorest world countries it estimated as 80% and 90% respectively. These variations through mortality depend on delayed diagnosis, risk factors in population subgroups, quality of treatments and follow-up of patients. The idea of this study is to evaluate the epidemiological features of common pediatric malignancies in Iranian population. In this regard the literature and review of Iranian papers around childhood cancers was the main fundamental of this study.

Literature and Review Databases of different provinces in Iran have some defects in pediatric cancers' registries. As some referral childhood centers never report their data or even their reports are deficient. So the loneliest and trustable information around this subject is papers that published by scientific groups. The pediatric population of 9 Iranian provinces in 1996 was nearly 7,000,000 individuals that 420 ones diagnosed as cancer patients. In that time the Age Specific Rate of Iranian pediatric cancer was 6 per 100,000 individual.

In 1996, the most common Iranian pediatric malignancies were leukemia (20%), brain tumors (15%), lymphoma (15%), sarcoma (8%), Wilms tumor (7%), retinoblastoma (6%) and soft tissue sarcoma (5%)

respectively. In 2010, the most common childhood Iranian cancers were leukemia, lymphoma and brain tumors.

The male/female ratio in Iranian pediatric cases is nearly 1.1-1.5. This ratio is in concordance with worldwide report. There are variations in the incidence rate of childhood malignancies between Iranian provinces. The mortality rate of pediatric cases less than one year old is higher than other age groups.

Conclusion One of the rare diseases among patients less than 15 years old is cancer that has specific characteristics. Iran as one of the developing regions in knowledge and science have to improve it's global pediatric cancer registry database. The main limitation in the epidemiological features of childhood cancers is that there is not definite reports of pediatric malignancies from childhood referral cancer treatment centers. However beside of establishing national childhood cancer registry database, more researches are needed for suggesting new strategies in improving the quality of diagnosis and treatment of pediatric cancers.

Mediastinal Teratoma in infant(case report)

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Abstract

Background: Mediastinal teratomas compose 20% of childhood mediastinal tumors, 10% of all teratomas and it is an uncommon cause of respiratory distress in neonatal period.

Method: We report a 6month female infant with mediastinal teratoma presenting with severe respiratory distress and recurrent cardiac arrest.chest radiography showed huge mediastinal mass with severe shift of heart and mediastinum to left side.she underwent emergency right thoracotomy because of recurrent cardiac arrest and The 6x 5x8 cm mass adjacent to the right upper lung was totally resected.

Results:infant improved very rapidly after excision of mediastinal mass. Histopathologically immature teratoma was identified. Chemotherapy was administered. At the 6th month of her follow up,she was very good with normal breathing and normal c.x.r. the AFP level was concordance with her age. No recurrence was observed.

Conclusions: Most of mediastinal teratomas grow slowly and do not manifest emergent condition. Chest X-ray, thorax ultrasonography, computed tomography and magnetic resonance imaging are useful in identifying the localization and association of mass with peripheral tissues. Treatment of the mediastinal teratomas in children is total surgical excision.

Case report : An Infant With A Mature Sacrococcygeal Teratoma

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A teratoma is a tumor with tissue or organ components resembling normal derivatives of more than one germ layer. teratoma may be mature ,immature or malignant.Sacrococcygeal . Extragonadal teratomas of childhood are rare tumours and may occur in unusual locations . sacrococcygeal teratoma(SCT) generally a rare condition, with a reported incidence of approximately one in 35,000 – 40,000 live births. (SCT) is a most common tumour in the newborn period.

Aim To demonstrate the clinical presentation and management and one year follow up .

of a mature SCT in an infant and review the literature

Patient and Method

We present A case of a infant with mature gluteal or lateralized SCT

Results

A 3-mounths old infant girl is presented with a cystic mass or (SCT) prominent on the left gluteal side after primary investigation and oncologic and radiologic consultations on a team management strategy she had complete surgical excision through an posterior sagittal approach (including coccygectomy). blunt and sharp dissection of tumor that was pushed rectum and bladder anteriorly. suturing sphincter to pre sacral and skin trimming and primary wound closure with an invert y shape line was performed . she had uneventful postoperative recovery. Pathologic report was mature cystic teratoma with a normal coccyx . with a team management and during, one year follow up, using clinical, biochemical and radiological assessment revealed no evidence of recurrence.

Conclusion : we are recommended : team management for appropriate investigation and surgical approach and periodic biochemical monitoring and long term follow up in these atypical patients

A rare case of rectal Adenocarcinoma In A Nine Year Old Girl

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back ground and aims : Adenocarcinoma of the colon is very rare malignancy in children.

Incidence of this tumor in children is the tenth in a million of live births

In this study we present a nine years old Female child with adenocarcinoma of the rectum that underwent emergency laparatomy .and review the literature.

Case presentation: A Nine -year-old female patient who is admitted for rectal bleeding in gastroenterology ward for undergoing colonoscopy. In colonoscopy a large sessile in ten c.m s of anal Verge recognized and tried to sampling. In next few hours after colonoscopy ,The patient developed anemia and abdominal distention and signs of peritonitis . abdominal X. R revealed free air in abdomen .The patient underwent an emergent exploratory laparotomy.At laparotomy Perforation was seen in the rectum with sessile polyps on the five to ten cms region of anal Verge ,those have been covered 2/3 diameter of posterio lateral of rectum resection of affected proximal of rectum with possible relative safe margins ,proximal end colostomy and Hartmann Poach was performed .

the colon sample was sent for Pathologic assessment . the report was rectal adenocarcinoma.

After completed investigation and consultation with colleagues oncologist 2weeks later on the another elective operation, resection of the The remainder of rectum as a low Anterior Resection by stapler and a Protective ileostomy was performed. the patients followed with chemotherapy, And two months later, ileostomy was closed

In the next patient follow-up, examination,Tumor markers assessments were normal , and currently there is no evidence of recurrence of the tumor.

Conclusion: Colorectal adenocarcinoma is rare in children.Treatment is Complete resection of the tumor followed by chemotherapy.so that our patient. that on near one year follow up , has no evidence of tumor recurrence.

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A Challenging topic about A benign tumor with invasive behaviour

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BACKGROUND:

Desmoid tumors are known to be benign tumors and Wide local excsion with safe margin is generally prefferd as a surgical approach . However , in practice it may have a challenging behaviour with unexpecting results despite standard surgical techniqus and postoperative follow ups .

Local recurrances , intra abdominal invasion , chest wall or abdominal wall closure and consequent dissabilities after multiple opertaions are the most challenging issues of this benign tumor .

METHODS : (Case Report)

A 6- years old girls with prevoius history of abdominal wall desmoid tumor surgery in iraq two years a go , was come with a 5*5 cm round mass in anterior abdominal wall.

A tumor was resected again with recurrent desmoid tumor diagnosis with safe margins (pathologic approved) and intra abdominal invasion (liver right lobe and omentum) removed intra operatively , abdominal wall defect was tension-free closed with prosthetic dual mesh .

Achild sent to P.ICU after surgery .

3 dayes after surgery , she went on surgery with dehiscence of the mesh at the superior and lateral border of it despite the tension free fashion of mesh closure . the aviscerated small intestine reduced to the abdomen and the fascia refixed to the mesh borders.

In this period of addmission the patient had seroma collection 3 times that aspirated under guide of songraphy.The patient was discharged with tamoxifen and solindac .

RESULTS:

Desmoid tumors is a benign tumor but it may have an invasive and recurrent behaviour . Microscopic negative tumor margins and likely use of prosthetic material is accompany with high incidance of collections and infection as a forign body .because of these reasons it is suggested not to use prosthetic materials for abdominal closure and skin closure in this session and incsional hernia is acceptable .

CONCLUSIONS:

Desmoid tumor is a challenging tumor in managemet and trreatment , modified diagnostic , operative and post operativestrategies is being needed.

Abdominal Burkitt's lymphoma in Children

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BACKGROUND: Burkitt's lymphoma is a high grade B-cell neoplasm, which is a kind of small non-cleaved cell lymphoma. It is presumably the fastest growing human malignancy, and the patients are prone to develop tumor-lysis syndrome. Here we present findings on our patients with Burkitt lymphoma.

METHODS: This descriptive retrospective study included 46 children with abdominal Burkitt's lymphoma who were treated during 15 years from June 1998 to Dec 2013 at Mofid Children's Hospital.

RESULTS: Of fourty six patients 32 (70%) were boys and 14 (30%) were girls with ages ranging from 2 to 14 years. Surgical exploration was carried out in all cases, the lesions were located in the small intestine (N=17), large intestine (N=15), ileocecal region (N=12), and stomach (N=2). We performed a complete mass resection in 16 cases, debulking in 10 and lymph node/mass biopsy in 20 cases. Pathologic examination revealed Burkitt's lymphoma for all patients. The majority were stageIIE and stage IIIE (24 and 19 respectively). Post-operative complications were seen in five cases with the most common being persistent ileus. All patients received a sort of systemic chemotherapy. The mean follow up duration was 6 years. Death occurred in 7(15%) of our patients, due to tumor lysis syndrome and acute renal failure.

CONCLUSIONS: The extent ofdisease at presentation is the most important prognostic factor in abdominal Burkitt's lymphoma. Children with BL are at a high risk of tumor lysis syndrome before or during chemotherapy. Surgery still plays an important role in this pathology, and chemotherapy offers an excellent chance for long term disease free survival.

ABDOMINAL MASS SECONDARY TO HUMAN TOXOCARIASI: A case report

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INTRODUCTION: Toxocariasis is a widespread helminth infection that leads to visceral larva migrans in humans. It has an extensive and various clinical spectrums.

Case presentation: A 2.5 year old female referred for abdominal mass. She had history of pharyngitis from 2 weeks ago and abdominal mass identified by physician incidentally. She was quite asymptomatic without any abdominal painor fever, cachexia or loosing appetite. Physical examination indicated an irregular solid mass in RLQ approximate 8x11 cm. The patient was anemic with hemoglobin 10.1gr/dl and 4% eosinophil.She had leukocytosis. Abdominal sonography revealed an echohetrogeny large mass in RLQ and in liver and retroperitoneal.Abdominal CT scan revealed a huge mass. Laparatomy was carried out, and exploration showed massive retroperitoneal mass which involved right liver lob, bladder , ileocecal valve ,small and large intestine . 50cm of small intestine, cecum, appendix, ascending colon ,some mesenteric lymph node measured 2×1.5×1cm were removed,and ileocolostomy was performed. Histopathological examination verified the serological test against toxocara. .

conclusion: Some clinical features of human toxocariasis can mimic tumors .Symptoms , signs and investigations are well-matched with malignancy so surgical exploration remains undesirable.

Kaposiform Hemangioendothelioma Of A Neonate's Left Arm

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Kaposiform hemangioendothelioma is an aggressive endothelial-derived spindle cell neoplasm that occurs nearly exclusively during childhood and teenage years. The lesion grows rapidly and is often associated with Kasabach-Merritt syndrome.

In this case report a 24 days old male neonate who presented with an ill-defined deeply situated violaceous mass on his left arm is described. He had also anemia and life-threatening thrombocytopenia. Despite hospitalization in intensive care unit (ICU) and transfusion of platelets and packed red blood cells as well as medical managements such as oral prednisolone, intravenous (IV) methylprednisolone and interferon alpha, thrombocytopenia persisted, so surgical excision was done.

Meticulous surgery with preservation of vessels and nerves was performed.

Also shaving of the mass on the surface of the left brachial artery was done.

The histopathological findings were distinctive and characteristic of kaposiform hemangioendothelioma. Following surgery, the infant did not have any complications and was discharged from the hospital in good conditions.

Now after seven years He is completely fine and ok.

Abdominal Mass (Burkitt lymphoma)

Javad Seyedi ;Marzieh Sabzechian

Pediatric Surgery Research Center, Shahid Beheshti University of Medical Sciences

Case report: Pediatric patient 11 years old, with chife complain nausa and vomiting had two week ago.

After sonography and CTscan showed hudge mass,FNA quided CT presented nuroblastoma,suspicious lymphoma

After operation showed mass mesenteric and not vertebral origen, therefore incsional biopsy from mesanter and omentom performed

Neonatal Neck Mass (PNET)

Javad Seyedi ; Marzieh Sabzechian

Pediatric Surgery Research Center, Shahid Beheshti University of Medical Sciences

Case report: Nenatal femal delivered with cesarian cection .birth weight was 2900 g. The neonate had cyanosis, breathing disteress ,tachypnea and granting after the birth.

In left neck a mass with size 2#2 was found.

Abdominal & chest examination was normal.pripheral vascular was normal too.

Increaseing size of the tumor was quickly enough that extubation was imposiible so surgery was necessary.

A mass with ill defined borders was found in surgery.

Important Renal cystic diseases in children with emphasis on cystic nephroma, cystic partially diffrentiated nephroblastoma and localized multicystic renal dysplasia

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The important cystic lesions of kidney in children are:

cystic nephroma (CN) cystic partially diffrentiated nephroblastoma (CPDN) and localized multicystic renal dysplasia (MCKD), Cystic nephroblastoma, cystic clear cell sarcoma,

cystic congenital mesoblastic nephroma

diffrantiation between these lesions are challenging and sometimes the correct diagnosis become possible after operation.

We will discuss about CN, CPDN, MCKD and cystic nephroblastoma (Clinical presentation, diagnostic modalities, treatment and prognosis)

METHODS:

3 cases that will present prospectively will discuss.

RESULTS:

The results will explain with each cases

CONCLUSIONS:

The correct diagnosis of kidney cystic lesions of the children are necessary for better treatment

PLEUROPULMONARY BLASTOMA (PPB), A CASE DISCUSSION AND INTRODUCING INTERNATIONAL PPB REGISTRY

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BACKGROUND:

Pleuropulmonary blastoma (PPB) is a rare childhood malignancy which is believed to rise from Pleuropulmonary mesenchyme. PPB have seen in children less than 6 years old mostly. Since 1988 that the first case of PPB was reported, less than 500 cases are recorded up to now. This presentation is a case based discussion of a PPB and an introduction to the international Pleuropulmonary blastoma registry (IPPBR)(www.ppbregistry.org)

METHODS:

A 2 year old boy was presented with tachypnea and dyspnea. Primary evaluation revealed a left white lung with mediastinal right shifting in chest x-ray. Ct scan showed a huge left hemithoracic solid mass and core needle biopsy was compatible with PPB.

RESULTS:

According to the risks of primary surgical intervention, neoadjuvant chemotherapy was initiated based on international protocols for PPB III and the case was submitted and enrolled in PPB international registry. A rapid reduction in size of tumor was noted after the first course of chemotherapy so it was continued for two more courses. Further images showed a well-defined pelural mass in anterior thoracic wall near fourth and fifth ribs. The patient become asymptomatic and prepared for surgical resection. Enblock tumor excision was performed including two involved anterior ribs and thoracic wall reconstruction with anterior serratus muscle flap. All the margins were tumor free. Post-operative course was uneventful and all the specimens and also parental blood and sputum samples were transmitted to international laboratory of PPB registry (US) to confirm the diagnosis and genetic evaluation to define the risk of disease inheritability and transmission.

CONCLUSIONS:

Although current guidelines are suggested primary resection in PPB but neoadjuant chemotherapy may be very helpful in huge tumors even in type III. International collaborations may help to improve treatment of rare diseases and offer complementary high tech paraclinical studies to all physicians and families around the world.

Thyroidectomy in pediatric patients

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The chart of patients who underwent thyroid surgery since 2008 were reviewed. there were 30 patients. 10 patients had total thyrodictomy due to hyperparathyroidesm. 8 patients had multi nodular goiter and 4 more patients had hashimato thyroiditis.

There were 5 patients with papillary carcinoma of thyroid. All of the patients with papillary carcinoma of thyroid were more than 6 years of age and there were 3 female and 2 male.

3 patients underwent lobectomy of thyroid but because of the result of pathology were positive for malignancy reoperation was performed for total thyroidectomy.

3 more patients also needed reoperation due to lymph node enlargement.

fine needle asperation were performed for 3 patients preoperatively and all were negative for malignancy however the end pathology results showed malignant cells.

for one patients frozen section was performed which was negative for malignancy. this study showed that malignancy is common in patients with thyroid nodule and should be treated surgically and usually need total thyrodictomy.

Here we try to discuss the approach for cold nodule and options of therapy for malignant one

Large Liver Embryonal Sarcoma: Anatomical Resection of Huge Mass

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Background: Liver tumors are common in children and are one of the causes of patient referral to pediatric surgery clinics. Resecting liver tumors is evolved during recent decade with introduction of Couinad liver segments classification and also development of new devices. The other factor which is associated with improved ability to resect such tumors is increasing surgeons experience with liver surgery and performing anatomical and extra-anatomical resections.

Case Presentation: The Patient was a 13 YO Girl came with chief complain of abdominal pain, post parendial vomiting and abdominal distension in recent weeks. On admission patient had a palpable RUQ Mass. CT scan was done which showed a 200x156x172 mm mass which occupied the Rt Liver lobe. Preoperative diagnoses proposed for her were Mesenchymal hamartoma and atypical hemangioma. Patient undergone exploratory laparotomy and Rt Anatomical Hepatectomy was done for her. Patient tolerates the procedure well and discharged in good condition.

Results: This case is the largest such liver tumor that is reported in the literature. But the importance of this case is that Anatomical resection of liver masses is independent of tumor size and exploration should be done in such cases. As here despite the huge size anatomic resection was possible.

Conclusion: Resection of liver tumors is became a subspeciality in pediatric surgery. Having adequate knowledge of liver segments made these procedures possible and these procedure should be done in tertiary referral enters. *irsps.org*

Survival and long-term outcomes in children with hepatoblastoma: our center experience

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BACKGROUND/PURPOSE:
This is a retrospectively study to evaluate and compare the clinical features, treatment strategy, pathology, and outcome of all patients with hepatoblastoma treated at Mofid hospital over a 10-year period(2006-2015) The aim of this study was to review the outcome and complications after resection of hepatoblastoma treated over one decade in our institution..

METHODS: 40 patients with hepatoblastoma were idntified in this study. All available clinical, surgical, radiologic, and pathologic data were reviewed and analyzed.
RESULTS: Between 2006 and 2015, 40 children were treated for hepatoblastoma. The age range was 3 to 96 months (median 20 months). The right lobe was involved in 61%, the left lobe in 9.55, and in 28.6%% the main bulk of the tumor was centrally located. Surgical procedures included the following: complete surgical resection then chemotherapy in 65% , surgical biopsy „Neoadjuvant Chemotherapy then complete surgical resection in 20% and percutaneous biopsy ,Neoadjuvant Chemotherapy then complete surgical resection in 15%. Intraoperative bleeding was a common complication but only one patient expired during surgery..Rupture of inferior vena cava and portal vein were seen in 2 patient that repaired. Microscopic residue of tumor was seen in 22.7% of patients. Postoperative complications occurred in 8 patients including biloma in 5 patients ,subphrenic abscess in 2 cases, and adhesion obstruction in one .Pulmonary metastasis was seen in 2 cases that in one case led to pulmonary resction.7 patients underwent redo resection. That this procedure occurred 2 times for one of them.
CONCLUSIONS: Reresection of a positive resection margin does not necessarily have to be performed, because postoperative chemotherapy showed good results. Resection of lung metastases can be curative if there is local control of the primary tumor. Preoperative chemotherapy followed by complete surgical excision according to International Society of Pediatric Oncology guidelines yields excellent results.

PREPUBERTAL BREAST MASSES

Leili Mohajerzadeh, Ahmad Khaleghnejad Tabari, Mohsen Rouzrokh,

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OBJECTIVES:

Breast masses in children and adolescents are uncommon and most often benign Prepubertal breast masses are nearly always benign. Primary or metastatic tumors of the breast are rare but can present in prepubertal children .The majority of lesions are benign. Cancer metastatic to the breast has been reported in children with primary hepatocellular carcinoma, Hodgkin lymphoma, non-Hodgkin lymphoma, neuroblastoma, and rhabdomyosarcoma, particularly the alveolar variant.
Cases:
We present three cases of prepubertal breast masses. Case one was 1.5 year old male with mammary hamartoma . To the best of our knowledge, this is a rare case of mammary hamartoma occurring in a male child.

Case 2 & 3 are boys with metastatic tumors followed PNET and Hodgkin lymphoma.

Conclusion: Breast development at the onset of thelarche starts with a firm, disklike area of tissue under the areolar complex that can be mistaken for a mass, but if the diagnosis is uncertain, fine-needle or open biopsy may be indicated.

Primary retroperitoneal Yolk sac tumor (PRYST)

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Introduction: Germ Cell Tumors (GCTs) arise from primordial germ cells, which migrate during embryogenesis from the yolk sac through the mesentery to the gonads. Most childhood extragonadal GCTs arise in midline sites. Yolk sac tumors is the only malignant germ cell tumor type occurring in the sacrococcygeal region of infants. Extragonadal YST is rare, particularly in the retroperitoneum.
These tumors consist of friable, pale gray, mucoid tissue in which variable amounts of hemorrhage and necrosis are present. Yolk sac tumors produce alpha-fetoprotein (AFP), while germinomas (seminomas and dysgerminomas), and especially choriocarcinomas, produce beta-human chorionic gonadotropin (beta-HCG), resulting in elevated serum levels of these substances. Elevated alpha-fetoprotein (AFP) ≥ 10,000 ng/mL was associated with worse outcome.

Case Presentation: An 18-months-old male presented with sacrococcygeal buldging and abdominal distension for one month. The patient was admitted to Amirkola Children Hospital. An abdominal computed tomography (CT) scan revealed a solid mass located in the retro peritoneum which extended from anterior of sacrum to left diaphragm. The α-fetoprotein (AFP) serum levels were elevated to10.000 ng /ml. A palliative tumor resection was performed during the first exploratory laparotomy and perineal resection on October 28, 2009. The histopathology report revealed a malignant YST. Chemotherapy was started intravenously by Bleomycin, Etoposide and Platinum. The α-fetoprotein (AFP) serum levels were decreased rapidly after 3 course of chemotherapy, but gradually elevated after 9 course of chemotherapy. Unfortunately CT and MRI were normal at that time. After that the patient underwent to surgery for 5 times. The α-fetoprotein (AFP) did not become to normal level by chemotherapy even by different drugs. Finally widespread metastasis to lung and skull became obvious, despite of intensive chemotherapy and the patient was dead.

Conclusion: The Yolk sac tumor (YST) represents a highly malignant germ cell neoplasm in children and is generally characterized by a high serum α-fetoprotein (AFP) level. The serial measurement serum AFP is useful for monitoring its clinical course and response to treatment. We cannot trust to normal imaging study when AFP is abnormal.

Inflammatory pseudotumor; two case report

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Inflammatory pseudotumor(IPT) or plasma cell granuiomas are synonymous for an inflammatory solid tumor that contains spindie cells,myofibroblasts,plasmacells.Inflammatory pseudotumor can occur in children and young adults,in many different organ systems. Common sites of presentation include lung,orbit,mesentery and spleen. Inte stinal involvement is rare. The tumor behavior resembles cancer,but histologically without malignancy. Our cases were 2 cases with sigmoid tumor and ileocecal tumor, that presented with complete obstruction.Complete resection with primary anastomosis was performed because of obstruction.

Inflammatory myofibroblastic tumor of the small intestine: a case report

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INTRODUCTION: Inflammatory myofibroblastic tumor (IMT) is a rare benign tumor. Usually seen in children and adolescents, this inflammatory tumor can affect all the organs. The definitive diagnosis of IMT is possible through surgery and pathology. The removal of symptoms generally requires the resection of tumor.

PRESENTATION OF CASE: In this case, a five-year-old child experienced the sudden onset of symptoms and the enlargement of abdominal mass 20 days before referral. The patient did not have any symptoms of nausea, vomit, and abdominal pain. The medical ultrasound indicated a mass sized 10*8cm in the abdominal cavity. Observed in the abdominal cavity, the mass was extended from below the pancreas to the area near pelvic cavity. In the laparotomy, a large and sticky solid mass, attached to the ileum with the mesenteric origin, sized 10*8cm was observed and completely resected. The sample was sent for pathological analysis to confirm the diagnosis of inflammatory myofibroblastic tumor.

DISCUSSION: This tumor rarely emerges in the small intestine, and there are few patients with intestinal manifestation. In this case report, the tumor had an origin of the small intestine mesenteric and it had invasion to the ileum.

CONCLUSION: Despite using some radiographic methods such as medical ultrasound and computerized tomography (CT) scan to diagnose the disease, the definitive diagnosis is merely possible thorough complete surgical resection. The final confirmation is also based on the results reported from the sample sent for pathology.

Pediatric Thyroid Cancer

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Background: Thyroid carcinoma in pediatric patients usually manifests as an asymptomatic neck mass, with a reported incidence of cervical lymphadenopathy that ranges from 35-83%, Therefore, they present in advanced stage than adults.The aims of our study was to evaluate disease demographic, clinical presentation, management and outcome of patients under 15 years old with thyroid cancer in Alzahra Hospital from 2000-2016.

Materials and Methods: This study was performed on 19 patients under 15 years old with thyroid cancer undergoing thyroidectomy in Alzahra Hospital from 2000-2016. The age, sex, clinical presentation, pathology of cancer, type of surgery, complication and outcome were retrieved from the medical records.

Results: Among 19 patients, 13(68%) case presents with neck mass, 3(15%) with hoarseness and three patients were found in screening. Overall, 16 patients (84%) had unilateral disease. Ten subjects (52%) undergone total thyroidectomy and in others near total thyroidectomy was done; neck lymph node dissection was performed in all patients (10 patients had unilateral and three of them had bilateral involvement of neck lymph nodes).twelve (63%) patients got I_{131} .

During the 16 years follow up period two patients passed away, one of died case had lung metastasis (in total 3 cases developed lung metastasis).

Conclusion: We conclude that, thyroid cancer in pediatric is more invasive comparing with adults. The standard procedure is total thyroidectomy with lymph node dissection and I_{131} base on type of thyroid cancer.

CLINICAL IMPLICATIONS AND SURGICAL MANAGEMENT OF ACUTE ABDOMENS IN PEDIATRIC PATIENTS WITH MALIGNANCY; A REVIEW OF 138 CASES.

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Background: Pediatric oncology patients developed similar abdominal emergencies as immunocompetent children in addition to acute abdomen from malignancy or secondary treatment. Cytotoxic chemotherapy, radiotherapy and extensive surgical resection all contribute to the risk of gastrointestinal symptoms. Immunocompromised children may lack the inflammatory signs of acute abdomen making prompt diagnosis of pediatric oncologic abdominal emergencies challenging.

Method: Here we review of clinical finding and surgical management of 138 cases with abdominal emergencies secondary to malignancy including gastrointestinal hemorrhage, infection, mechanical obstruction, and perforation as well as management strategies for these condition.

Result: The most common abdominal emergencies secondary to malignancy are inflammatory process, obstruction, and GI hemorrhage respectively. Pain is the most common and sensitive clinical symptom; however, Prompt identification of abdominal emergencies in these patients can be a challenge due to decreased signs and symptoms of inflammation in the immunocompromised host in addition to interference with other problems of primary disease.

Conclusion: A high index of suspicion and a prompt multidisciplinary approach are essential for optimal patient care. Early initiation of aggressive medical management reduces the need for invasive surgical treatment and concomitantly improves mortality rates.

Hepatic resection in small tumors in non liver surgery center in children with liver tumors (10 years experience)

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BACKGROUND: Liver resection is the most frequently used method to treat hepatocarcinoma in pediatric patients. Approximately 70 % of hepatocarcinomas in children are malignant. And primary hepatic malignancy is one of the ten most common malignancies in children.

METHODS:From January 2005 until December 2016 we have encountered 6 case of hepatoblastoma which had lab data , rdiologic workup, biopsy. They were 2 girls and 4 boys with mean age of 3.5 (range 1-4 years)and there tumor diameter were mean of 4.5 cm (rang 3-6) at least involving 2 segment underwent resection with clear margin of 1 cm through technique of in flow and out flow control and finger fracture with fine clamp and major duct and vasculatures ligation.

RESULTS:2 major bleeding which were controlled ,2 post-operative biloma manged with drained inserted previousely.We were able to resect the tumor in children with mean blood loss of 57.7 cc (range 40-130 cc)1 abcess formation .3 post operative bleeding controlled conservatively.we had clear margin in all resection .the mean survival were 65% in 4.5 (range 2-6years).also there were no intraoperative dead .

CONCLUSIONS:The most common primary hepatic malignancy in pediatric patients, accounts for approximately 80 % of primary hepatic malignancies. Hepatoblastoma is commonly seen in children under 3 years of age, is rarely observed in those beyond 5 years of age, and is predominantly present in males (male/female ratio 2.5:1). Surgical removal is important in treating pediatric hepatoblastoma and beacause of difficulty in sound liver resection in non master center creating criteria and choice of patient for obtaining good out come.

Prevention of complications which canot be managed.

Thyroid Nodules in Childhood: a Single Institute Experience

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Objective: Thyroid nodules are rare in children. Multiple diagnostic modalities are used to evaluate the thyroid mass. The aim of this study was to determine results of management of thyroid nodules in children with special attention to the role of fine needle aspiration biopsy (FNAB) in diagnosis.

Methods: Thirty-two children who underwent surgery for thyroid nodules in Mofid Children's Hospital within 10 years (1996to 2005) were retrospectively studied. From clinical records we obtained data about demographic characteristics, clinical manifestations, ultrasonography (USG) findings, and FNAB results, pathological reports, surgical therapy and complications. Data was analyzed statistically for association with thyroid cancer.

Findings: Twenty-five patients (78.1%) were girls, and 7 (21.9%) boys. Mean age was 10.9 (range 8 to 14) years. 24 (75%) patients had benign and 8 (25%) malignant tumors. 18 (56.25%) nodules were located in the right lobe. Statistical analysis revealed sensitivity, specificity, accuracy, and positive and negative predictive values as follows: 80%, 65%, 63%, 25%, and 86% for USG; 35%, 41%, 40%, 18%, and 66% for RNS; 91%, 94%, 90%, 74%, and 96% for FNAB respectively.

Conclusion: Clinical judgment as determined by serial physical findings with USG continues to be the most important factor in the management of thyroid nodules in children. FNAB is the most accurate method of investigation and its accuracy is improved by USG guidance.
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CONGENITAL SACRAL MESENCHYMAL CHONDROSARCOMA IN A NEONATE; CASE REPORT AND REVIEW OF LITERATURE

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METHODS: Mesenchymal chondrosarcomas are rare malignant tumors in children, especially in neonates. The authors present a case of congenital mesenchymal chondrosarcoma in a one-day neonate located in sacrum. According to the authors' literature searches, this case is the first congenital sacral mesenchymal chondrosarcoma. We also reviewed the papers published in English literatures

Sacrococcygeal yolk sac tumor developing after mature teratoma:case report

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Abstract

Purpose: mature(MTs)and immature(ITs) teratomas are histologically considered to be benign and are usually treated by gross complete resection. However, malignant germ cell tumors, such as yolksactumors, may occur at the same location after sacrococcygeal teratoma (SCT) resection.

Case:We evaluated the clinicopathological characteristics of yolk sac tumors (YSTs) developing after sacrococcygeal mature teratoma (SCT) resection. The patients were diagnosed with YST within 12 months after initial surgery. The patient underwent gross complete resection with coccygectomy in neonatal period and had histologically mature teratoma without microscopic YST foci . The surgical margins were undocumented. She observed without chemotherapy. But one year later tumor recurred with liver and lung metastases. Redo resection showed yolk sac tumors.

Conclusions: YST can develop after SCT resection not only in patients with previously reported risk factors. We recommend that patients undergo serum AFP monitoring with Intervals of measurement≤4months to detect subclinical localized YSTs after resection.

فرازهای برجسته تاریخ جراحی جهان

مصطفی جابر انصاری - ۱۳۹٤

هر چه کردم جستجو در کار خویش　　تا ره آوردی مگر آرم به پیش

غیر برک سبز زاری مختصر　　هیچ مقدورم نشد زان تحفه بیش

یک حکایت بشنو از تاریخ گوی　　تا بری زین راز سر پوشیده بوی

گر گران و گر شتابنده بود　　آنکه جوینده ست یا بنده بود

" .تردیدی نیست که گذشته چراغ راه آینده است. جورج سانتایانا استاد دانشگاه هاروارد چنین می‌گوید: " ملتی که تاریخ خود را نشناسد، لاجرم محکوم به تکرار آن است
Those who can not remember the past, are condemned to repeat it

ما ایرانیان از جمله مللی هستیم که در طول حیات خود بارها تاریخ را تکرار کرده‌ایم؛ زیرا با تاریخ تمدن و علم کشور خود و سایر ملل آشنایی کافی نداریم. نگارش تاریخ پزشکی، چیزی ورای آشنایی با زندگی پزشکان مشهور یا توصیف ساده‌ای از پیشرفت روش‌های بالینی یا جراحی است. اکثر تاریخ نگاران و همینطور تاریخ نگاران طب و جراحی، تمرکزشان روی وقایع مهم تاریخی و سیر تحولات بوده است. این سلیقه، اگر چه در جای خود قابل احترام است، اما نتایج کاربردی کمتری به همراه دارد و اغلب .موجب کسالت خواننده غیرمتخصص می‌شود

ویل دورانت (٥ نوامبر ۱۸۸٥ – ۷ نوامبر ۱۹۸۱)، فیلسوف، تاریخ نگار و نویسنده آمریکایی در آثار خود علاوه بر توجه بروی وقایع تاریخی، توجه خاصی به سیر تمدن بشری، و مهمتر از آن به عوامل تمدن ساز در طول تاریخ داشته است. او می‌نویسد: "تمدن رودی است با دو ساحل ". او به این مسئله اشاره دارد که تاریخ نگاران اغلب نهایت توجه و دقت خود را معطوف رودخانهٔ در جریان تاریخ می‌نمایند؛ که معمولاً پرآشوب و پرهیاهوست؛ و اجازهٔ برداشت‌ها و تفاسیر صحیح را نمی‌دهد. در مقابل دیدگاه دیگری وجود دارد، که در آن حواشی تاریخ و تمدن ساحل‌ها، به اندازهٔ متن تاریخ اهمیت دارند. به عقیده او همه مردمانی که در طول تاریخ خانه و مجسمه ساختند، شعر سرائیده اند، و . . . در شکل‌گیری تمدن نقش داشته‌اند. ویل دورانت در کتاب " درس‌هائی از تاریخ که در سالهای آخر زندگی خود تألیف نمود، چنین می‌نویسد: **" تاریخ ملت‌ها را باید با توجه به پدیده‌های علمی جدید نوشت "**

در اکثر کتب تاریخ پزشکی و جراحی، شرحی از زندگی پزشکان بزرگ و آثار آنها ارائه می‌شود. بنابراین کتاب تبدیل می‌شود به مجموعه‌ای بی‌ربط از شرح حال پزشکان. در حالیکه با نگاه نوین به تاریخ پزشکی می‌باید علاوه بر پرداختن به سیر تحولات و محتوی زیست شناسی رشته جراحی، نتایج اجتماعی، اقتصادی و سیاسی هر مرحله را در کنار تولیدکنندگان آن تحلیل نماید. این ممکن نیست مگر تاریخ پزشکی و جراحی را در بستر تغییرات سیاسی، اجتماعی ، اقتصادی، و بهداشتی هر دوران بررسی نمائیم؛ و این .درست همان کاری است که پروفسور روتکو در اثر خود انجام داده است

فرایند با اهمیت حرفه اي شدن جراحي: براي قرنهاي متمادي، عمل‌های جراحي به عنوان روشهاي فاقد اهميت و تنها در حد يك روش ترسناك و به عنوان جزء بسيار كوچكي از . حيطه وسيع پزشكي، تلقی می شدند

از چه زماني جراحي از يك روش صرفاً عملي و فيزيكي به روشي كاملاً مبتني بر علم و تحقيقات دقيق علمي در مطالعات باليني مستقل تبديل شد؟

.تقابل پيچيده‌اي از عوامل اجتماعي، سياسي، اقتصادي و سازماني در فرايند حرفه‌اي شدن جراحان و حرفة جراحی دخيل بوده است

جراحي چگونه مستقل شد؟

آيا اين فرايند از يك كشور اروپايي به كشور ديگر سرايت كرد، يا به عنوان فرايندي همزمان تمام علم پزشكي را در اروپا فرا گرفت؟ آيا جراحان ايالات متحده مراحل مشابهي را طي كردند كه همتايان اروپايي آ نها قبلاً تجربه كرده بودند؟ قبل از عصر نوين تفاوت قابل ملاحظه‌اي بين پزشك و جراح وجود داشت. پزشكان همواره به عنوان افرادي فرهيخته كه تحصيلات دانشگاهي در حوزة طب داشتند در نظر گرفته مي‌شدند. در حالي كه جراحان عنوان هنرمندان يا استادكاراني را داشتند كه بدون تحصيلات عالي بودند. اينان مهارتهاي خود را با شاگردي نزد استادان ماهر فرا مي‌گرفتند. در چه زماني اين تشكلات قديمي كشورها خاتمه يافت و جراحي چهرة اجتماعي و جايگاه ويژة خود را در پزشكي پيدا كرد؟ از همه مهمتر آن كه در چه زماني تخصص در رشتة جراحي جزء جدايي ناپذير مسئوليت جراحان شد؟

The Role of Play in Children's Palliative Care

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BACKGROUND:
It is generally agreed that play is an important part of children's development, education and learning, and that play develops with the mind, and the mind develops with play. Play is the language of children. Play is the way that children learn about their world, become competent at skills, learn to cooperate with others and gain mastery over their emotions. Play is what motivates children to get out of bed in the morning. Think of children and one thinks of play. Mention toys and one thinks of children. However, thoughts of very sick children or children with severe limitations or restrictions on their mobility, their stamina or their cognitive capacity do not as quickly turn to thoughts of toys and play. As responsible adults our overwhelming impulse when faced with a very sick and vulnerable child is to find ways to improve that child's health, take away distressing symptoms and to provide the most appropriate medications and therapies. It does not sit well with us that children suffer and die, but they do. While still not universally accessible , all children with life-threatening and life-limiting conditions have the right to receive palliative care which aims to ameliorate the unpleasant symptoms of the illness or condition and improve the quality of life of the sick child and that child's family .
Palliative care providers walk the journey from diagnoses, through to the end-of-life and into bereavement along with the child and the child's family, providing appropriate and compassionate support whenever and wherever it is needed. However, despite the devastating consequences of a terminal diagnosis, the child continues to have all the unique and ever-evolving needs, desires and the rights of any other child .The overwhelming urge and desire Children 2014, 1 304 to play still exists, indeed, it could be argued that because of the benefits of play, these children have an even greater need to play than other children
RESULTS:Palliative care aims to optimize quality of life in the face of an ultimately terminal condition. The World Health Organization (WHO) defines palliative care for children as "the active total care of the child's body, mind and spirit, (which) also involves giving support to the family. It begins when illness is diagnosed, and continues regardless of whether or not a child receives treatment directed at the disease. Health providers must evaluate and alleviate a child's physical, psychological, and social distress.The Therapeutic Role of Play in Children's Palliative CareAll children receiving palliative care need to play and whilst not all of them will need play therapy, the need for this type of therapeutic intervention will be higher within this more vulnerable population. Conclusions:A child receiving palliative care not only has the right but has an overwhelming need for time to be a child, to engage in childish pursuits such as developmentally appropriate play. The dying child needs activities that have no chance of failure as a way to balance the feelings of defeat and loss of mastery that comes with a diagnosis of a life-limiting illness. More than anything, these children need opportunities to communicate openly and honestly, to be enabled to express their very real fears or their feelings of anger. They need to know that there is someone on their care team that they can talk to about anything that concerns them, including about dying. Children without the necessary verbal skills, the maturity or the full comprehension of what they are experiencing only have the medium of play through which to communicate these anxieties and concerns. Those involved in their care need to have the skill to "listen" to what the child is telling them through their play and to respond appropriately.

The effects of motivation interview on decrease the stress of children parents need to surgery

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Abstract

Context and causes

Today humans dealt with different unsecure life which is his stress comes up of its. And todays all of these reasons that caused by mashinc system of life that make its double.

In this research the topic of study is about the effects of motivation interview on decrease the stress of children parents need to surgery.

Method

This research is a kind of similar test that is designed by a pretest and the real test which is used by a controlling group that made with a sudden simple test for 30 people.

In this test among 30 parents 15 of them were managed by sudden test and 15 of others tested by controlled groups. The process of motivation interview with parents of sick children's passed by the stress action test.and they filled eshpigel Berger paper.

After doing the motivation interview for groups both of each group were under the level of stress by passing the test and after this process they filled the eshpegel paper test.

In this aspect the average of estimating test related to the analyzing the average score of the collection tests.(for getting to the average used of circumstance software.)

Results

The Consequences of search shows that motivation interview caused to decrease the level of stress for parents needed to surgery.

Conclusion

The result of searching and analyzing of quriyans shows the effect of interviews and some ways to decrease the stress of patient children's need to surgery.

Scientific challenge: Pediatrics trauma may result malignancies

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Introduction:

Many causes can provoke pediatrics malignancies.Some causes are well known and some are in doubt.In this paper we propose probable relationship between trauma and malignancies.

Case Presentation:

Case 1:6month old male has reffered with tunor in histhigh following vaccine injection in the first trimester of age.The pain of region was present in visits have done within 3 month later.Sonography showed a mass in region .whith suspection of abcess aspiration has done in operation room.Biopst findings indicated rhabdomyosarcoma.

Case 2: A 3 years old male has reffered to clinic by pain of arm.Parents presented history of bicycle crash in 4 month ago which resulted arm trauma. He had pain whitin 4 month in region. Biopsy had done and rhabdomyosarcoma has diagnosed.

Case 3: A 5 years old male has falling down on perineum region 6 month ago.he have had pain during this period.A mass has f ound in Perineum. Biopsy had done and rhabdomyosarcoma has diagnosed.

Case 4: A 10 years old female with history of blunt trauma on left breast region 8 month ago.physical examination has found a mass and biopsy findings indicated adenocarcinoma.

Case 5: A 7 years old female who had history of blunt trauma in left renal region following car crash in 5 month ago has reffered. She had pain in region within 5 month ago.At the time of referral all of paraclinical evaluation included sonography and CT was normal and she discharged after 48 hours.The pain has remained for 5 month and repeated CT has shown tumor and biopsy confirmed wilms tumor.

Conclusion: Since history of trauma ,we suggest all children with this history nust evaluate in aspect of tumors.

Solid pseudo-papillary tumor of the pancreas

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Abstract:

Background: Categorized as benign non-metastatic masses, solid pseudopapillary tumors of pancreas (SPTP) often present with non-specific signs and symptoms most commonly as pain, palpable mass, obstructive jaundice, bloating, and dyspepsia. Although rare, the SPTP are slightly more common in female adolescents. Definite diagnosis would be made accordingly to histopathologicfindings including excessive necrosis, cystic changes, solid areas mimicking endocrine tumors, and cystic areas with papillary structure. Surgical resection of encapsulated tumor would be the most definite treatment for SPTP with more that 95% complete response rate; however, neoadjuvant chemotherapy might be occasionally used as well.

Case presentation: Three cases with SPTP have been referred for more investigations. The first patient has been admitted with 1-year history of progressive obstructive jaundice signs, which were managed medically and have been severely aggravated following abdominal trauma. The two other patients have presented with abdominal pain and tenderness followed by nausea, non-biliary vomiting, and leukocytosis in one of them; and progressive jaundice in the other one. Primary diagnosis has been made through abdominal CT scan and they have underwentWhipple surgery for complete mass resection with maintaining lateral elements including the CBD and IVC. Final diagnosis has been confirmed through histopathologic findings as necrosis, pseudopapillae, covered with single to multiple layers of epithelial cells. Besides, the IHC has revealed positive results for α1-antitrypsin, vimentin, calretinin, CD10; and negative results for CEA, CA19-9 and chromogranin. One patient required post-operative chemotherapy and all three patients experienced complete response after 5 year follow up.

Study of Advantages and Disadvantages of Totally Implantable Venous Access Device in childrens with malignant disesase

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INTRODUCTION

Totally implantable venous access devices (TIVAD) or implantable catheter ports are devices which can be implanted subcutaneously. They enable prolonged and repeated access to the vascular system, into the peritoneal cavity or intravertebral space. This device is particularly useful for repeated medical injection, for blood sampling or transfusion of blood and blood derivatives and for total parenteral nutrition (TPN). Nowadays the most usage of TIVAD is for patients who are suffering from cancer and are candidates for chemotherapy.This group of patients are suffering from not only the deteriorative disease but also the complications of chemotherapy and other therapeutic plans. Therfor TIVAD must be a safe device with low rate of complications.Although many patients benefit from the insertion of TIVAD without any secondary effects, any surgical implantation can nevertheless lead to complications.

MATERIAL & METHODS

In this study, we investigated the advantages and disadvantages of TIVAD catheter in pediatric age group. All patients who were operated for insertion of TIVAD from January 2004 to January 2014 were evaluated in our study. A total of 188 cases, 2 to 14 years old, were included in our study. We implanted TIVAD in these patients for chemotherapy in 173 cases (92.02%), for prolonged TPN in 6 cases (3.19%), for corticosteroid and antibiotic therapy after Kasai operation in 2 cases (1.o6%), for intermittent IV therapy in 2 cases (1.06%) and for need to partial parenteral nutrition in 5 case (2.65%). The complications of TIVAD at the time of operation or after of insertion were reported in the patients files and we evauated them from patients files.

RESULTS: Out of 188 cases, 17 cases (9.04%) had some kind of complications and 171 cases (90.95%) had no complication. There was no mortality. Most patients and their parents 175 , (93.05%) were satisfied from TIVAD. It seems that TIVAD can be a useful device for many chronic patients who need an IV access for multiple injections. Pneumothorax was occurred in 1 case (0.53%) , hemothorax in 2 cases (1.06%) , exravasation of drug in 3 cases (1.59 %), infection in 2 cases (1.06%) , occlution of catheter in 5 cases (2.65%),hematoma in 2 cases (1.06%) , vegetation at the tip of the catheter in 1 case (0.53%) , difficult extraction of catheter due to adhesion to adjacent tissues in 1 case (0.53%).

CONCLUSION :TIVAD is a device for reducing the pain and complications of intermittent drugs infusion which are used for chemotherapy in children who are suffering from malignancy or chronic deteriorative diseases therefor it must be inserted by an expert pediatric surgeon for reducing these complications. Morover the most complications were occurred after insertion at the time of needle insertion for repeated usage therefor some expert nurses are needed for reducing these preventative complication

COMPARING OF PORTTOCATHETER INSERTION THROUGH EXTERNAL JAGULAR VEIN VS.INTERNAL IN CHILDHOOD TUMOR CHEMOTHERAPY

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BACKGROUND: Low intraoperative complication rates by surgical approaches with cephalic vein or external jugular vein cut-down techniques that are related to. We are to report a prospective evaluation of 33 consecutive external jugular vein cut-down approaches for totally implantable venous access devices implantation.

METHODS: thirty three consecutive patients (18 M, 15 F, mean age 54.2) suffering from solid tumors (38) or hematologic diseases (20) were consecutively submitted to totally implantable venous access devices insertion through external jugular vein cut-down approach .

RESULTS: All devices were surgically implanted; no instances of intraoperative complications were detected. After a minimum follow-up of three month, only 3 case of wound hematoma and 5 case of device malfunction due to incorrect catheter angulation and inappropriate care during usage were noted

CONCLUSIONS:external jugular vein cut-down approach should be considered as a tool for long-term central vein catheters positioning, both as an alternative or primary approach.

WHICH LOCATION IS THE BEST FOR IMPLANTATION OF PORT SYSTEMS IN CHILDREN WITH MALIGNANCY? A single center experience EXPERIENCE

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ABSTRACT: Objective: The best location for implantation of port systems (LIPS) in children with cancer remains controversy. The proper location of an implanted reservoir might be a way for reduction of complications. The aim of this study was to compare three LIP systems including subcutaneous pocket implantation (SCP), subpectoral fascia pocket implantation (SPFP) and over costal ribs port fixation (OCP) in children with malignancies.

Methods: Between 2003 to 2013 a consecutive sample of patients (mean age: 18 ± 28.8 months, ranging from 1 to 180 months; 450 female, 593 male) with a variety of malignant diseases were enrolled into this cross-sectional, single-center study. All suitable size catheters were inserted through the internal jugular vein in the superior vena cava above the right atrium (level of Louis angle of the sternum)under general anesthesia. Then, patients' complications were assessed among three groups to select the best LIPS.

Findings: The ports were placed in SCP (n= 282), SPFP (n=342) and OCP (n= 436) groups, while 17 patients received a second device after removal of the first one, due to failure of the first implantation. The mean follow up was 412days ranging from 8 to 2102 days. Common complications were recorded in SCP, SPFP and OCP groups as follows: catheter displacement(4.3%; 4.1% ; 0.9%), skin necrosis(7.1%; 1.2% ; 0.4%), port exposure (4.3%; 0.3% ; 0.23%), port related infection(3.9%; 2.3% ; 1.8%), catheter obstruction(6.4%; 6.1% ; 4.6%), pocket hematoma (2.1%; 2.3% ; 0.9%), the reservoir rotation (4.6%; 2.3% ; 0%), difficulty in port injection (0%; 1.2% ; 0.9%), wound Infection(5.7%; 2.9% ; 0.5%), vein thrombosis(2.1%; 1.8% ; 0.4%), pocket Infection(4.3%; 2.9% ; 1.4%),andrespectively. There were significant association between complications such as the reservoir rotation, catheter displacement, skin necrosis, port exposure, wound infection and wound bleeding in the 3 procedures applied.

Conclusions:The findings suggest that the OCP was the best location for implantation of port device for management of children with various malignant diseases as study showed lesser complications.

complications and outcome of Port—Caths inserted in Ali Asghar children Hospital (2011 -2015)

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Abstract ;

Central venous catheters (CVCs), such as the tunneled catheters and the totally implanted ports, play a major role in general medicine and oncology. Aside from the complications (pneumothorax, hemorrhage) associated with their initial insertion, many of these CVCs are associated with the long-term risks of infection and thrombosis. Despite routine flushing with heparin or saline, some of CVCs result in thrombosis of the blood vessel,infection, and obstruction.

Complications related to Port--Cath were studied retrospectively during a period of 48 months in 92 patients, aged 1 month-18 years, with : malignancy . nephrotic syndrom .short bowl syndrom and CF. There were 102 Port--Cath inserted, (in10 patients catheter were extracted and reinserted) 95% of them were inserted.under sonographic guidance. No complications occurred at the time of insertion. No complications occurred in 81 patients . In 11 patients there were 3 events of local infection & in one patient a combination of systemic and local infection ; which cannot treated successfully with antibiotics. 4 obstructions and 2 skin necrosis occurred . one local bleeding had occurred in which catheter kept unused for some days.There was no mortality.

Conclusion ;

Port Cath implantation is associated with some risk of serious complications. Many of immediate perioperative and short-term complications such as accidental arterial puncture, hematoma, , pneumothorax or vessel perforation can be reduced by sonographic guide .This procedure must be done by an expert surgeon and carefull management of the catheter should be maintained to decrease the risk of complications. therefor some expert nurses are needed to reduce these complications.

Types and frequency of ovarian masses in children over a 10-year period

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Background: Ovarian masses represent a range of pathology from benign cyst to highly aggressive malignant tumors. It has been estimated that gynecologic malignancy account for approximately 2% of all types of cancer in children, 60-70% of these lesions arise in the ovary.

Methods: All ovarian masses which were resected or biopsied in Mofid Children's Hospital from 2002 to 2012 were reviewed retrospectively. Patient's age, presenting symptoms, surgical procedures, pathological diagnosis, postoperative treatment, and outcome were obtained from medical records.

Results: Fifty-seven girls (aged 40.2±57months with the range of 1 day to 15 years) underwent different types of ovarian operations (24 salpingo-oophorectomies, 10 oophorectomies, 21 ovarian cystectomies, and 2 ovarian biopsies). 50 children had unilateral ovarian mass (49.1% right and 38.6 left, respectively). The most common presenting symptoms were acute abdominal pain in 46%.Twenty one (37%) of our patients had ovarian torsion. Four (7%) patients had benign tumors, and 8 (14%) had malignant tumors. There were no age differences between those with benign type (8.2±2.6years) and malignant tumors (6.1±5.3years) (P=0.683).

Conclusion: Ovarian tumors are rare in children. Most are benign, in children presenting with acute abdominal pain, ovarian mass particularly neoplastic tumors should be suspected. An important proportion of these patients may require postoperative chemotherapy.

Anesthetic considerations of child with intravascular extension of wilms tumor ,a case study

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BACKGROUND: Extension of abdominal tumors through the IVC presents a unique challenge to both the surgeon and the anesthesiologist . In the pediatric population , the most frequent intraabdominal tumor associated with intraluminal venous extension is wilms tumor .The aim of this study is to consider and manage safety anesthesia in children with Wilms tumor.

METHODS: This study was a case report of intravascular extension of wilms tumor

The 7 years-old, 25 kg boy presented with a right abdominal mass. There was no associate symptoms. Ultrasonography revealed a large echogen mass in right abdominal area with pressure effect on liver and spreading into the pelvic area. Pre operative tests were normal .The patient was candidate for laparotomy with probable wilms tumor diagnosis. Anesthesia was induced and continued as routine.

RESULTS: .Forty five minutes later during closure of renal vein and emptying the IVC , Spo2 decreased suddenly and the airway resistance increased substantialy. After a while the JVP increased and the patient became hypotensive. CPR began but the patient's condition deteriorated and deceased after 45 min of CPR.

CONCLUSIONS : Preoperative echocardiographic studies with emphasis on intravascular and intra-atrial spread of tumor in these patients may decrease morbidity and mortality.

The method of pain control i n children with Wilms' tumor surgery

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BACKGROUND: Wilms' tumor is a malignancy of the kidney that occurs in childhood. One of the stages of this mass treatment is surgery to remove the tumor. Children's pain control after surgery is a challenge for nursing care of them. The aim of this study was to evaluate drug methods of pain control in children that undergo surgery.

METHODS: This cross-sectional study was done through reviewing of documentation for 28 children that undergo Wilms' tumor surgery and methods of pain control were derived from the nursing records. Results were analyzed using spss17 statistical software.

RESULTS: The results showed that in 7 children was used of pethidine injection combined with One of the acetaminophen dosage forms. in 3 children only injectable form of acetaminophen is used. Pain of 19 children was controlled with acetaminophen Supp. and in 3 children with acetaminophen syrup. All these drugs have been used for 24 hours after surgery. There is no housing in 4 children. Any one of non-pharmacologic methods to pain management is not mentioned.

Port-a-Cath-related complications in children with cancer admitted to Tabriz children's hospital

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BACKGROUND: The use of cytotoxic drugs causes to vein damage and its wall inflammation. Applying the port-a-cath to access central venous is one of the methods that can overcome this problem. Despite of the numerous advantages of port-a-cath, this method has some complications. The main purpose of current study is investigation on the port-a-cath-related complications in children with cancer.

METHODS: This cross-sectional study was done through reviewing of documentation for children with port-a-cath who admitted to Tabriz children's hospital and was recorded port-a-cath-related complications. Results were analyzed using spss17 statistical software.

RESULTS: According to the results, most complications from using the port include the closure of the port with clots, infection, dislocation of port-a-cath and infiltration of drugs, disability to touch the port and spin it. The closure of the port with clots and infection were the most common complication of port and other complications were in later stages.

CONCLUSIONS; experiments showed that staff training in the use of port-a-cath and aseptic conditions will have significant effect in reducing complications from using the port-a-cath. Also education of patients and their family about how to deal with the port-a-cath and its protection can be useful.

CONCLUSIONS: Pain after surgery is very common in children. It is essential that nurses have sufficient knowledge about the pharmacologic and non-pharmacologic pain control ways in children.