اصول انکولوژی در کودکان

دکتر مرداویج آل بویه

Principles of Cancer Therapy in Children and Adolescents

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Malignancies are rather rare ailments in population between the ages of 0-15 years. However they are gaining mounting importance in developed and developing countries and rank as second cause of death after accidents and poisoning. Their incidence is reciprocal to the socio-economic status of the population .Although recent reports from Europe indicate an upward trend in incidence of malignancies in the respective age group, mainly acute lymphoblastic leukemia (ALL). The annual cancer incidence is reported as 15/100000 in the age group of 0-15 years. In Iran the population has doubled to over70.000.000 in the past three decades of which approximately 34 percent are younger than 15 years old. By considering the national improvements in health care services and socio-economic standards the estimated the annual cancer incidence is 3500.This implies to expect higher efforts by official health care services to provide proper preventive measurements, diagnostic facilities and medical care and services for this group of patients. Today's diagnostic and treatment modalities have given rise to long time survival in up to 80 percent of cancer patients. These enormous achievements in the past three decades have been based on deeper understanding of tumor cell biology and growth mechanisms, drug actions and resistance mechanisms. Childhood and adolescence malignancies are in contrast to adult malignancies mostly of mesodemal origin. Carcinogenic evolvement occurs during the embryonic period in a group of tumors such as neuroblastoma, Wilm's tumor, retinoblastoma, rahbdomyosarcoma, germ cell tumors and hepatoblastoma. Clinical manifestation occurs in 50 percent of these tumors between the ages of 0-4 years. ALL is diagnosed between 1-6 years of age and other tumors such as malignant lymphoma and bone tumors in older age groups. Biologic characteristic of hematologic and other malignancies are rapid local growth and distant dissemination. Therefore they are considered as systemic diseases and are treated as such. Based on these biologic behaviors all diagnostic and therapeutic managements will be from the first visit, discussed and designed by close cooperation between pediatric oncologist, experienced pediatric surgeon and radiotherapist; multidisciplinary approach. This modality is based on three essential pillars; poly-chemotherapy, surgery and radiotherapy (neo-adjuvant therapy).Goals are reduction of malignant cell burden , reduction of tumor volume ,thus rendering its operability and destruction of metastatic cells. New management of malignant tumors comprises diagnostic biopsy followed by preoperative chemotherapy to reduce the tumor mass and thereafter in- Toto tumor extirpation. At this stage histological study of tumor tissue will give information about preceded therapeutic response as well as histological malignant grading; response kinetic. Intra-or postoperative imaging procedures will help to identify tumor remnants.PET Ct-Scan will provide additional information about metabolic activities of the tumor. Based on these investigational results further chemo-and radiotherapy will be elaborated and enacted; postoperative adoptive chemo-/radiotherapy. Chemo-and radiotherapy may be given concurrently (synchronic) or alternately (sequential).In case of primary tumor resection the following chemo-and/or radiotherapy will be applied as adjuvant treatment modality. Another treatment modality is radio-sensitization by utilization of drugs like bleomycin, cis-platin or doxorubicin, which is indicated in CNS tumors or soft tissue sarcomas. These drugs will increase tumor sensitivity to the applied irradiation. The goal of poly-chemotherapy is inhibition of tumor cell proliferation and differentiation at various stages of cell cycle (Go, G1, S, G2, M) and act as complementary to each other; cell cycle independent, phase specific and non-specific activity. Among the malignant cells there is a group which is resistant to the chemotherapy, either by intrinsic or extrinsic pathways. Therefore maintenance chemotherapy is mandatory in order to destroy the dormant tumor cells. Currently available cytostatica affect not only the malignant cells but also normal cells. Goal of the neo-adjuvant treatment approach is therefore 1) to minimize acute and late side effects of chemo-/radiotherapy and 2) to preserve organ functions and limb salvage. Recent insights into immune properties and molecular pathways of malignant cells has paved the way for targeted cancer treatment e.g., monoclonal antibodies, tyrosinkinase inhibitors . Considering the complexity of diagnostic steps and elaborate treatment protocols and treatment related acute and chronic complications, the necessity to handle all pediatric malignancies at pediatric oncology centers with experienced medical and nursing staff and well equipped facilities is self-evident. Other provisions are easy accessibility and availability of anti-cancer drugs, antibiotics, antifungal drugs, blood products, growth factors etc. Today most malignancies in children and adolescents are curable with long time survivors. Therefore continuous follow-up of this cohort beyond the set age limit by pediatric oncologists is necessary to ensure timely diagnosis and treatment of different treatment related late effects including second malignancies. In conclusion cure and long term survival of cancer victims will be achieved only by close cooperation of pediatric oncology centers and application of standardized diagnostic and therapeutic approaches based on national consensus.

متاستاز در سرطان کودکان

دکتر مریم قوامی عادل

Tumor metastases in children cancers

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Cancer, a common disease in adults, is rare in children and adolescents. But it is second cause of death after trauma in children older than 1 year. There are Less than 1% of all malignancies in pediatric patients. But, timely diagnosis of childhood cancer is extremely important. Many pediatric malignancies are curable, and in some of them, earlier diagnosis can be associated with a better prognosis. There is an increase in their occurrence either, with an approximately 1% increase per year overall and for some tumors, such as brain tumors, a 2% increased incidence per year.

Tumors are called "malignant" because they have the ability to invade normal tissues (replacing healthy cells with cancer cells) and to metastasize (spread) to other parts of the body. Death from cancer often comes not from the primary site (where the cancer first began) but from the metastases. The development of tumor metastases is the main cause of treatment failure and a significant contributing factor to morbidity and mortality resulting from cancer.

Metastases takes place in many ways: through the lymphatic system, through the bloodstream, by spreading through body spaces such as the bronchi or abdominal cavity, or through implantation. The most common way for cancer to spread is through the lymphatic system. This process is called "embolization". Lymph nodes are dynamic structures that can widely fluctuate in size. Factors of importance are size and location of the nodes.

Cancer can also metastasize through the bloodstream. Cancer cells, like healthy cells, must have a blood supply in order to live, so all cancer cells have access to the bloodstream. Malignant cells can break off from the tumor and travel through the bloodstream until they find a suitable place to start forming a new tumor. (Tumors almost always metastasize through the veins rather than through the arteries.) Sarcomas spread through the bloodstream, as do certain types of carcinomas, like carcinoma of the kidneys, testicular carcinoma, and Wilms' tumor, a type of kidney cancer seen in young children. Cancers may spread by more than one route.

Cancers can also spread by local invasion -- that is, by intruding on the healthy tissue that surrounds the tumor. Some cancers that spread this way do not venture very far from the original site. However, it is possible that a second cancer of the same kind may start to grow at a later time at a completely different site -- the new growth having nothing to do with the first.

A very rare type of metastasis is caused by implantation or inoculation. This can happen accidentally when a biopsy is done or when cancer surgery is performed. However each type of cancer has its own pattern for metastases. Whether or not cancer cells metastasize to other parts of the body depends on many factors, including: type of the cancer, stage of the cancer and original location of the cancer.

By early cancer and its metastases sites detection with proper management we can have much better outcomes.

گزارش ۷ مورد نوروبلاستوما

دکتر سعید طرلان– فاطمه خلیلی – پریسا علیپور

The report of 7cases with neuroblastoma

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Neuroblastoma is the most common extracranial solid malignant of early infants. Its incidence is of 7-12 per 1,000,000in general population. The most common site of the tumor is Adrenal & Retropretoneal. Most patients consult and present at late stages.
The local disease has minor effusion of such tumor.
Stage IV in which we have 10% of whole patients is of interesting pretences of such tumor. This stage opened an immunotherapy era of research.
Abdominal mass is the most common presentation of neuroblastoma and can accompany with weight loss, fatigue ,fever, anemia and ostealgia either metastatic symptoms of neuroblastoma (such as anemia, Horner's syndrome ,skin nodicles ,ostealgia) or paraneoplastic syndrome can be presented.
Initially screening to identify the tumor in early stage .Was expressed by Japanese, but its role in decreasing mortality was not remarkable so now a day there is no insisting on it.
Biochemical markers , imaging and pathology are helpful to diagnosis the tumor. Proper combination of surgery, chemotherapy, radiotherapy and probably immunotherapy have great role in treating such patients .Such combination is based depend on the stage and situation of the patient.
In our center 7 patients were managed and followed up during 7 years .Among them two ones were at stage 4-S.
1) The first patient was presented with intense hepatomegally, abdominal mass and respiratory distress, who was supported with conservator methods .Then the remains in left adrenal were excised when the patient was at 1-years old now he is very well.
2) The 2 second patient was 3months-old, who was presented with multiple skin nodules and adrenal mass which was confirmed neuroblastoma after biopsing the nodules of skin .He was followed up and cured .Now he is very well.
3)The third one was a 2-years –old female presented with protosis &Racoon eye who had retro orbital metastasis.She had a surgery and was undergoing chemotherapy .She had no efficient follow up on her parents behalf and thus she died.
4) The forth one was a male who was admitted at stage IV with bone marrow metastasis and adrenal tumor. He was undergoing chemotherapy.
The initial source of that tumor was adrenal .After chemotherapy [he was undergoing] surgery on the cited source ant its [remnant] was exeised .After chemotherapy he was referred for BMT.
5) A 3-years-old male with bone marrow and pelvic mass metastasis was another patient who had high marker of VMA and cluster cells of bone marrow
He was undergoing chemotherapy and died during that process.
6)The sixth one was a 13-months-old male with huge dumbbell abdomhnal mass which extended to vertebra and spinal cord .After investigating and laparatomy ,we biopsied it .Then the patient was undergoing chemotherapy . Second operation included excision of the remnant tumor .Now he is well and at good situation.
7) The seventh was a 4-years –old child who was admitted at poor condition with huge retropretoenal pelvic mass that caused hydronephrosis .Biopsy was done .He died during chemotherapy.
Among 7patient who were diagnosed &managed in my center, 2patients were at good condition and very well.
The other patients were at stage III &IV. Among these 5,3patients died, 1patient referred for BMT and the other one had no problem and is very well now.
More patiems referred at high stage in our center with abdominal mass or metastasis. However excition of the tumor at stage III or IV was practically impossible ,we decided the surgery after chemotherapy because of more easier and better excision of the tumor . First of all the role of surgery was to diagnosis and after all was to total excision of that tumor.

گزارش یک مورد نوروبلاستوما مدیاستن

دکتر عباس هادی پور

A case report of Mediastinal Neuroblastoma

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Introduction: The term Neuroblastoma is commonly referred to spectrum of neuroblastic tumors (including neuroblatomas, gangliono neuroblastomas, Ganglioneuromas) that arise from primitive sympathetic ganglion cells. It accounts for 15 percent of all pediatric cancer fatalities. Site, stages, age, N-myc oncogen, q-p chromosome deletion, and histological features are common prognostic factors. Mediastinal neuroblastoma is relatively good prognostic factor.

Aims: Introducing a case of Mediastinal ganglion neuroblautomas with unusual clinical presentation. After successful treatment, (surgery and chemotherapy) patient's condition is favorable after 2 years.

Method: A 6 months old female under treatment of bronchitis and prolonged diarrhea was admitted for further evaluation. After CXR and CT-scan with impression of Neuroblastoma, VIP checked & the posterolateral thoracotomy and resection was done and then chemotherapy after surgery.

Results: Mediastinal tumors detected incidentally on CXR in the evaluation of respiratory problem in children. VIP secretion tumor in our patient was a paraneoplastic syndrome that is rarely associated with Neuroblastoma. VIP causes abdominal distension and intractable secretory diarrhea with hypokalemia. Urinary VMA, HVA were increased.

Pt becomes symptom free and normal lab data after treatment.

Conclusion: Our patient was misdiagnosed for two months and treated for bronchitis and prolonged diarrhea. Pneumonia and respiratory symptoms and paracardiac consolidation with not good response to treatments need further & more evaluations such as CT-scan and lat chest X-ray. Such as other reports, VIP producing tumors are lower aggressive than undifferentiated Neuroblastomas. Finally, surgery should be helpful to Mediastonal Neuroblastoma that is usually benign and good prognosis. Our patient is good and symptom free after 2 years follow up.

Evaluation and Outcome of Neuroblastoma in Childrens from 1996 _2007 Alzahra University Hospital

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Introduction:The goal of this study is evaluation of sex, age, clinical presentations, primary sites, sites of metastasis, chemotherapy courses, survival of patients and detect how canwe Improve survival with neuroblastoma in Alzahra hospital isfahan University from 1996 -2007
Results: 29 patiens were evaluated 18 patients were male (62.1 %), 11 patients were female (37.9%) M/F ratio is 1.63. Mean age of male is 29.2 months and female is 20.3 months.
Primary site: retroperitoneal (72.4%), cervical (6.9%), posterior mediastinum (13.8%), pelvis (6.9%). The patients under 1 year were (27.6%), between 1-4 years were (55.2%), and between 4-10 years were (13.8%) and over 10 years were (3.4%). Clinical presentations: abdominal mass(31%), movement disturbances (17.2%), abdominal pain (13.8%), abdominal distention(3.4%), fever (17.2%), cervical mass(10.3%), weight loss(3.4%), pulmonary sign and symptoms (3.4 %). Bone marrow biopsy (44.8%). Liver metastasis (13.8%), Lymph node metastasis. (13.8%), brain metastasis (10.3%), para aortic node metastasis (3.4%).
Operable (51.7%), non operable (48.3%) refused to or incomplete chemotherapy courses (62%), complete course chemotherapy (37.9%) radiotherapy (6.9%), bone marrow transplant (3.45%)

Prognostic markers: Age: most of the patients (52%) were under 5 years
Tumor histology most of the histology showed poor differentiated DNA ploidy we do not have any records, Nmyc gene amplification only in 4 patients detected, other markers such as cytogenetic, Neurotrophin receptor (TrKA) were not detected. Serum markers like ferritin increased in two cases, NSE in four cases, LDH increased in one and Gd2,Cd44 had no detection. Most cases were in high risk groups.
5 years survival revealed (12%) unfortunately most of the parents did not agree for chemotherapy so we do not have regularly follow up of these patients.
Conclusion: We need to teach more parents, detect Prognostic markers specially tumor histology, DNA ploidy ,N-myc amplification Serum markers and risk groups.

تجربه چندساله‌ء نوروبلاستوما در مرکز طبی کودکان

دکتر جواد احمدی

TEN YEARS STUDYT OF CHILDREN WITH NEUROBLASTOMA

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BACKGROUND: The neural origin of neuroblastoma was first proposed by Virchow in 1864. (1).Marchand2 (1891) is credited with recognizing the similarity of neuroblastoma cells to the cells of the sympathetic nervous system. The first recorded successful resection was undertaken by Willard BartlettBof St. Louis, Miss Ouri, in 1914(2). Neuroblastoma is the most common solid extra- cranial malignancy of childhood and the most common malignant tumor in infants (i.e., patients younger than 1 year) (3) The treatment of neuroblastoma requires a multidisciplinary approach. The experience at St. Jude Hospital with combination chemotherapy was published in 1965, (4) but overall results remained poor. The incidence of neuroblastoma varies a little between differential groups.19 in predominantly white populations; the age standardized rate is 7 to 12/1,000,000 and accounts for 6% to 10% of all childhood cancer.
MATERIAL AND METHOD: This study was a retrospective study .We studied all children who were undergone surgery due to neuroblastoma in children medical center in ten years from January 1999 to January 2009.cases that didn't have definite pathologic diagnosis or rosette cell were excluded of the study. Cases that had chemotherapy in the other centers before coming to our center were excluded of the study. Follow up of the patients was from13 moths to 10 years.
RESULS: We had 38 patients in this study, 20(53%) cases were male and 18 (47%) cases were female. Their age was from 5d to 11 years. 15(39%) cases were under 1 year and only one patient (3%) was above 10 years. Sites of tumor were located in adrenal in 21(53%) cases, other abdominal etroperitoneal sites 8(21%) cases , mediastinum in 5(13%) cases , pelvic cavity 3(8%) , and neck 2(5%) .In our study 26(68%) cases had metastatic disease or locally advanced, 9(24%) cases had localized disease, and 3(8%) had stage 4s. (in this study we classified stage I,II as localized disease) The first clinical sign or symptom that patients presented were abdominal mass in 18(47%), progressive paraplegia in 2(5%), sphincter disorder 1(2.6%), multiple skin nodules and abdominal distention in 3(8%), hypertension 1(2.6%), dancing eye or obsomyoclonus in 2(5%), metastasis complications in 11(29%). Complete resection was done in 19 patients in first operation and was done in 5 patients after chemotherapy and tumor size reduction. In 14 patient we could not resects the whole size of tumor neither in first operation nor after chemotherapy because of proximity the tumor to vital organs or due to the severity of the disease .In 7(18.4%) cases we had to do nephrectoy for complete resection of tumor. One of 3 patients with stage4s of disease was died due to respiratory distress before doing any procedure or medication. Two other patients were sent for hepatic artery embolization but because of technical problems they didn't do that. After follow up the hepatomegaly was regressed.
Discussion: Management of neuroblastoma needs a multidisciplinary team include pediatric oncologist, pediatric radiotherapist, and pediatric surgeon. In cases that surgical complete resection is not possible we must not take the patient at risk of massive bleeding or permanent essential nerve injury or death. Complete resection was done in 19 patients in first operation and was done in 5 patients after chemotherapy and tumor size reduction. More over we can give tissue biopsy at first operation in locally advanced disease and in very ill patients, therefore after the other modalities of therapy we can do another surgical procedure for resection of tumor. One of 3 patients with stage4s of disease was died due to respiratory distress before doing any procedure or medication. Two other patients were sent for hepatic artery embolization but because of technical problems they didn't do that. After follow up the hepatomegaly was regressed. Therefore In patients with stage 4s we can wait and watch if the patient has not respiratory or hemorrhagic or obstructive or any complication that needs surgical manipulation.

مقایسهء نتایج و عوارض پورتوکات در عروق ژوگولر داخلی و تحت ترقوه

دکتر بهار اشجعی

THE COMPARISON OF ADVANTEGES AND DISADVANTAGES OF T.I.V.D BASED ON THE SITE OF INSERTION

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Introduction

During the past decades due to successful improvement in management and treatment of very ill patients or patients who are suffering from cancer and use of total and partial parentral nutrition in many patients who cannot have oral nutrition for long time and many paitents like these, the patients need permanent or prolonged IV line or recurrent blood sampling for different causes.

In this report we inserted T.I.V.A.D catheter for these patients and studied the advantages and disadvantage of the device in pediatric age group. Also we compared this disadvantages based on catheter site insertion.

Material and Method

In this study we evaluated the patients who most of them have been suffering with malignancy, and for chemotherapy and blood sampling, port catheter has been implanted. Also This study has been done for patients who were und*er 14 years old and has been treated in pediatric medical center* of Tehran university. Out of these 131 patients 78 were male and 53 were female and average age was 2 to 14 years. Also in 18 patients for prolonged TPN (Total Parentral Nutrition) and 7 patient for corton-therapy and antibiotic therapy after kasai procedure in Biliary Atresia And 3 patient for metabolic disorder and need for intermittent IV therapy and 4 patient for malabsorption due to epidermolysis bullosa and intermittent P.P.N (Partial Parentral Nutrition) the port catheter has been implanted.

Operative Technique

The Patient takes a shower and goes to the bath in the night before surgery. One dose intravenous antibiotic 30' before starting operation for skin suprofits is injected. (often cephazolin 25mg/kg).

This technique is done under general anesthesia and supine position. A role is taken under the shoulder of the patient. Then prep and drape is done from the ear to xyphoid in both side of the patient. Two incisions are done. The site of exploring the vein and inserting the catheter is due to anatomy of the neck and chest and previous operation if has been done (for example external or internal jugular vein and subclavian vein in left or right).The best place for inserting the port is in the lateral of sternum in the same side of vein and under the clavicle (2-4 cm due to the size of the chest wall).

At first we insert the catheter into external or internal jugular or subclavian vein. Then we make a subcutaneous tunnel from vein to the place of insertion the port. And the end of catheter is attached to the port that is placed in the subcutaneous pouch in the lateral of the sternum. After the testing of the outflow and inflow of the device, port and catheter are heparinized .Antibiotics are continued for 48h after surgery (Iv or Oral).

Result

The insertion site was external jugular vein in 41 case and internal jugular vein in 49 cases and subclavian in 43 cases.

We had withdrawal occlusion (out flow obstruction) in 5 patients. And infection was occurred in 3 patients. In 4 patients the port was exposed and in 4 patients the catheter was exposed in its pathway. Complete obstruction of the apparatus was occurred in 10 patients. The cases in which exposure of catheter or port has been occurred we could solve this problem with local

anesthesia and covering the catheter with skin flaps successfully. In cases in which withdrawal occlusion was present we do not do any intervention because the inflow was sufficient.in cases who suffered with infection we extract the port and catheter. In cases due to complete obstruction we exchanged the place of apparatus and in these patients the entrance of catheter was from external and internal jugular vein in 7 cases the insertion site were external jugular vein and in one case the insertion site were internal jugular vein . in patients that insertion site was subclavian vein we did not have complete obstruction except one . in one patient who's catheter insertion was subclavian vein we experienced a serious complication. After explore for complete obstruction we saw disconnection between port and catheter and due to short distance between insertion site and iterance point to the vein that patient was at risk of catheter emboli. Most patients and their parents (117cases) were satisfy from port catheter

Discussion

From the past decades due to successful management of very ill patients or patients who are suffering from cancer and use of total and partial parentral nutrition in many patients who cannot have oral nutrition for long time and many patient like these who need a permanent or prolonged IV line for different causes, insertion of T.V.A.D catheter was suggested by medical doctors and specialists.

By using of this device the peripheral vein of these patients is saved and patients do not suffer from numerous needle sites that are needed for injection of drugs or recurrent sampling of blood.

This device is inserted in a central vein and therefore it works for long time and also because of the wide surface of the port, every time, one needle is inserted into the port from the skin over it very easily. This needle can be used for sampling or injection of drugs at the same time. On the other hand, because of inserting the catheter into a central vein, injection the hypertonic fluids or drugs that are used for chemotherapy cannot damage the blood vessels.

Therefore the patients can be satisfied from this device and we can help them to feel better and improve their life satisfaction. Therefore we suggest using the port catheter in any patient who needs prolong IV injection or recurrent IV-therapy or blood sampling.

Also we suggest these regards in implantation and maintenance of port catheter.

1- Prophylactic Antibiotics.

2- Prep and drape for sterilization.

3- Internal jugular vein is better than external jugular vein for insertion of the catheter.

4- In cases in which exposure of catheter is occurred extraction of catheter is not compulsory and this problem can be solved with local anesthesia and covering the catheter or port with skin flap with sterilization.

5- In withdrawal occlusion cases any extra interventional procedure is not necessary and that catheter can be used for injection.

6-subclavian vein is suitable for insertion of the catheter but in order to have safety guards to prevention of catheter emboli we recommend a subcutaneous tunnel at least about 10 cm.

Demographic and some clinical characteristics of neuroblastoma in Aliasqar Children's Hospital

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Background- Neuroblastoma is the second most common solid tumor in childhood and makes up 8% of the total number of children's cancers. The neuroblastoma can occur in nerve tissue alongside the spinal cord in the neck, chest, abdomen or pelvis, but it most often occurs in one of the adrenal glands. The treatment of neuroblastoma depends on the age of the child, the size and position of the tumor, the tumor biology (including the MYCN status) and whether the neuroblastoma has spread. Treatment modalities are surgery, chemotherapy, high-dose chemotherapy with stem cell rescue, monoclonal antibody and radiotherapy.

Material & Method- It is a retrospective study on all patients with neuroblastoma that were treated in our center between 2000 - 2009. Besides demographic data, stage of tumor, histology of tumor, radiotherapy, chemotherapy and their relation with survival were evaluated. For statistic SPSS was applied.

Results- in these study 30 patients with neuroblastoma were studied. There were 13 (43.3%) girls and 17 (56.7%) boys. Mean age of patients was 29.93±29.17 months (1month-9 years old). Most of patients were stage III or IV (40% in each stage).Chemotherapy used in 26 patients (86.7%) and radiotherapy used in 4 patients (13.3%). The mean survival time was 4.19 years. The five year survival for stage I, II, III and VI were 98%, 80%, 70% and 37% accordingly. There was no significant relation between survival and sex or age of patients. There was neither any significant relation between survival and stage of tumor. This may arouse from this issue that most of the patients (about 80%) were in stage III and IV.

Conclusion- Although neuroblastoma is one of the most common tumors in pediatric age group as you see in this study most of the patients refer in stage III or VI. We recommend national programs for comprehensive screening protocols and education in order not to miss the patients in early stages.

ثبت کانسر در ایران Cancer registry in Iran

دکتر محمدعلی محققی ریاست مرکز تحقیقات سرطان دانشگاه علوم پزشکی تهران (سخنران مدعو) و دکتر علیرضا موسوی جراحی

تاریخ ثبت سرطان در ایران برمیگردد به ده ۶۰ میلادی وقتی که برای اولین بار مرحوم دکتر حبیبی اطلاعات مربوط به بیماران سرطانی را که در مراکز پاتولوژی مختلف در سطح کشور وجود داشت جمع آوری و اولین آمار فراوانی را در مورد سرطان در جمعیت ایران منتشر نمود. بر طبق یافته های آقای دکتر حبیبی فراوان ترین سرطان در آن زمان عبارت بودند از سرطان قسمت فوقانی دستگاه گوارش ولنفوم ها بین هر دو جنس و سرطان پستان و دهانه رحم بین خانمها. بعد از گزارش آقای دکتر حبیبی، فعالیت های مربوط به آمار سرطان در سطح جمعیتی با راه اندازی برنامه ثبت سرطان شرغ دریای خزر که به منظور شناخت عوامل موثر در فراوانی زیاد سرطان مری در منطقه گنبد با همکاری بین سازمان بینالملی سرطان (IARC) و دانشکده بهداشت دانشگاه تهران در سال ۱۹۷۱ شرو ع ابتدا جمعیت منطقه گنبد و بعدا این برنامه به جمعیت دیگر مناطق حاشیه دریای خزر گسترش پیدا کرد و تمامی استانهای حاشیه خزر از جمله گیلان را پوشش داد اطلاعات منتشر شده در مورد سرطان از این برنامه ثبت نشان از میزان بروز بالای سرطان مری در مناصق ترکمن نشین (در بعضی مناطق میزان بروز بالای ۱۰۰ مورد در ۱۰۰۰۰۰ نفر) و مقایسه مناطق حاشیه دریای خزر نشان دهنده این واقعیت بود که در یک فاصله ۴۰۰ کیلومتری میزان بروز سرطان مری از ۱۰۰ در ۱۰۰۰۰۰ به میزان کمتر از زیر ۱۰ در ۱۰۰۰۰۰ در جمعیت استان گیلان تغییر میکرد. برنامه ثبت سرطان کنبد تا اواخر ده ۷۰ میلادی ادامه داشت و اطلاعات ده ساله وقوع سرطان در مناطق شمالی کشور را منتشر نموده است. همزمان با برنامه ثبت سرطن گنبد در استان فارس نیز برنامه ثبت سرطان راه اندازی شد و واطلاعات فراوانی سرطان را بدو ن ارجاع به یک جمعیت خاص بطور سالانه منتشر مینمود. این برنامه هنوز به صورت محدود اطلاعات و گزارشاتی منتشر میکند!؟

در سال ۱۳۷۵ به دنبال درخواست معاون پژوهشی وزارت بهداشت و با همکاری انتستیتو سرطان برنامه کشوری آموزش برنامه ثبت سرطان که بیشتر جنبه تشویق و آموزش مسئولین وزارت بهداشت جهت راه اندازی برنامه ثبت سرطان در سطح استانهای کشور را داشت شروع و گارگاهایی آموزشی برای مدیران سلامت استانها با هدف تشویق و آموزش برگزار گردید که نتیجه این فعالیت ها در سال ۱۳۷۶ با همکاری سازمان بین المللی سرطان تبدیل به یک طرح تحقیقاتی تحت عنوان "برنامه ثبت جمعتی سرطان در سطح شهر تهران" شد و این برنامه تاکنون ادامه داشته و اطلاعات مربوط به بیماران سرطانی را در سطح جمعیت تهران جمع آوری و گزارش مینماید. برنامه ثبت جمعیتی سرطان شهر تهران اولین گزارش خود را در سال ۱۳۸۶ بصورت کتابچه و مقاله علمی منتشر نموده. آمار منتشر شده که از آمارهای معتبر مربوط به سرطان میباشد میزان بروز سرطان در سطح شهر تهران را ۱۲۰ در یکصد هزار در زنان و ۱۵۰ در یک صد هزار در مردان منتشر نموده. سرطان معده به عنوان اولین سرطان در بین مردان و پستان با فراوانی ۳۱ در یکصد هزار اولین سرطان بین خانمها گزارش شده است. برنامه ثبت سرطان جمعیتی تهران در حال حاضر ادامه داشته و و چندین طرح تحقیقاتی را پشتیبانی میکند.

در اواخر ده ۷۰ اداره سرطان وزارت بهداشت برنامه ثبت سرطان بر مبنای گزارش پاتولوژی را با همکاری انجمن پاتولوزیست های ایران راه اندازی کرد که در این روش اطلاعات بصور غیر فعال توسط مراکز پاتولوژی به اداره سرطان گزارش و سپس اداره سرطان به تهیه گزارش جمعی و استانی به صورت سالانه اقدام مینماید.

آخرین فعالیت در رابطه با برنامه ثبت سرطان مربوط به فعالیت های مرکز تحقیقات گوارش و کبد دانشگاه علوم پزشکی تهران میباشد که در سالهای بعد از ۱۳۸۰ شروع و در چندین استان مخصوصا استان اردبیل و گلستان برنامه ثبت سرطان جمعیتی را راه اندازی و این برنامه تاکنون گزارشات دوره ای منتشر نموده است. یافته های این برنامه بصورت حکایت از میزان بروز بالای سرطان مری در جمعیت ترکمن در شرق دریای خزرو میزان بروز بالای سرطان معده در استان اردبیل میباشد.

بطور خلاصه: برنامه های ثبت سرطان امروز در کشور جزء برنامه های جا افتاده و استیبل فعالیت های مربوط به کنترول سرطان بوده و سهم این برنامه ها در برنامه های کنترولی سرطان در سطح کشور به خوبی مشهود است.

REPORT OF ISFAHAHN CLEFT CARE TEAM ACTIVITES (2005-2009)

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Abstract:

Cleft lip and palate are the most common congenital anomaly in head and neck in the world. About five years ago in 2005 the first team for care of the cleft patients begins to work in Isfahan provenance in the center of Iran. In this paper, we will discuss about the:

1- Why we chose Team management in cleft patient in Isfahan.
2- The specialists that work with each others for treating the patients.
3- The criteria of multidisciplinary cleft clinic.
4- Team protocol for timely team management.
5- Programs for patient's education (Isfahan cleft care team web site - workshops for family- booklets and posters).
6- Programs for patients continue educations.
7- The effect of the early intervention of the team in the management of cleft patients in the first three years.

In a retrospective cross-sectional study on 550 patients between '2005 to 2009', in Isfahan Cleft care team , we will discuss about the team experiences and pitfalls and errors , and the effects of team approach in the first three year in timely interventions in 250 patients ,variables such as (mean age of beginning speech therapy, secondary surgery, alveolar surgery and orthodontic intervention) are evaluated in two groups (early and late intervention) .

Among 250 patients who were evaluated, 56% were male and 44% female. Among 134 patients, 75 patients were refer to speech therapy before 4 year (early intervention) and 59 patients used it after 4 (delayed intervention). 47 patients had secondary surgery for improving hyper nasality. The mean age of early intervention (before 6y.o) was 3.88y.o and for delay intervention after 6 was 15.7y.o (P value < 0.05) 17 patients had alveolar bone graph, the mean age in the early intervention was before 9 and in other group was 16.69 (P value=0.006). 34 patients had used orthodontic treatment before 9, the mean age e in the first group (was 7.66y.o) and in the second group after 9 (was 17.05y.o).

This study showed that early intervention and timely team management will promote the health level of the patients. And we hope by using our experience in the near future in each province in Iran cleft team will be established and connected to each other

Training about nursing care in children with Leukemia Qods Children's Hospital Qazvin University of Medical Sciences

Salimi F. Tavassoli SH. Tavassoli Z. Sadeghipour Roudsari S.

Objective: leukemia is the most common malignancy in childhood. accounting for about 41% of all malignancies that occur in children<15 yr of age were diagnosed with leukemia in USA an annual incidence is 4-5 cases per 100000 children. The Leukemia may be defined as a group of malignant disease in which genetic abnormalities in a hematopoietic cell give rise to an unregulated clonal proliferation of cells. The progeny of these cells have a growth advantage over normal cellular elements, because of their increased rate of proliferation and a decreased rate of spontaneous apoptosis. The result is a disruption of normal marrow function and ultimately marrow failure. The clinical features, laboratory finding and responses to therapy depend on the type of leukemia.

Methods: This study is a review of nursing articles and references.

Results: Patients with cancer may experience a variety of secondary problems, such as infection, reduced WBC counts, bleeding, skin problems, nutritional problems, pain, fatigue, and psychological stress. An important role of nurses on oncology teams is to assess patients for these problems and complications.

Conclusion: Assess patient for evidence of infection
Check vital signs every 4 hours (especially temperature)
Assess Intravenous sites, wounds, skin folds, bony prominences, perineum, and oral cavity.
Placing patient in private room if absolute WBC count<1,000mm3and avoid fresh fruits, raw meat, fish, and vegetables.
Remove fresh flowers and potted plants.
Instruct all personnel in careful hand hygiene before and after entering room
Avoid exposing the area to sunlight or cold weather.
Instruct patient to report oral burning, pain, areas of redness, open lesions on the lips, pain associated with swallowing, or decreased tolerance to temperature extremes of food.
Explain that hair growth usually begins again once therapy is completed.
Encourage patient to wear hat, scarf, or turban.
Discuss potential hair loss and regrowth with patient and family.
Administer prescribed antiemetics, sedatives, and corticosteroids before chemotherapy and afterward as needed.
Ensure adequate fluid hydration before, during, and after drug administration, assess intake and output.
Suggest foods that are preferred and well tolerated by the patient, preferably high-calorie and high-protein foods. Respect ethnic and cultural food preferences.
Assess for bleeding(Petechiae or ecchymosis, hemoglobin or hematocrit)
Support the idea that cancer is a chronic illness that has acute exacerbations rather than one that is synonymous with death and suffering.
Assess the nursing care needs and problems of the patient and the family.
Assess the learning needs, desires, and capabilities of the patient with cancer.
Assess the social support networks available to the patient

Care of Neonates with Esophagial Atresia in NICU

Tavassoli S, Salimi F, mashhadi R, Sadeghipour S.
Qods Children's Hospital Qazvin University of Medical Sciences

Objective: Esophageal atresia is a relatively common congenital malformation occurring in about one in 2500 to 3000 live births. The pathogenesis is multi -factorial and involves multiple genes and complex gene environment interactions.

Methods: this study done by review nursing articles and references.

Results: During 2008, we had four neonates suffering with EA in Qazvin Qods children hospital**.**

Conclusion:
-Instruct nurses about diagnosis of EA in labor room and avoiding feeding.
-Keep baby warm to prevent hypothermia.
-Continues suction of NG Tube and care of airway, oxygen therapy giving correct position to baby before and after surgery and avoid of replace NG Tube and care of endo-tracheal intubation.
-Instruct all personal in careful hand hygiene for prevent of nosocomial infection and report any changes in respiratory and secretions.
-Control vital sign, Io, care of chest tube.
-Attention for any symptoms of pneumothorax, atelectasis and sepsis.
-Encourage family to contact baby's surgeon for dysphasia and respiratory problem.
-Awareness family about GER, recurrent T-E fistula and esophageal stricture.
-Provide pamphlet and booklet about EA and giving to family.

بررسی عوامل مؤثر در تشخیص زودرس آپاندیسیت حاد در کودکان

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مقدمه :نظربه اینکه آپاندیسیت حادشایعترین علت اوژانس بستری کودکان دربیمارستان هامی باشدبمنظورتعیین عوارض آپاندیسیت حاد وارتباط آن بافاصله زمانی شروع علائم تاتشخیص نهائی وعمل جراحی ،میزان عوارض ومدت زمان بستری بیماران ،این بررسی هاانجام شدوبنظرمی رسدکه بررسی علائم درددرکودکان بستری شده ورابطهٔآن باتشخیص های افتراقی مثل UtiوGEو.....مهم می باشدوروش تشخیص آپاندیست حادباتوجه به اقدامات پاراکلینیکی درمراکزمختلف متفاوت است وبررسی علائم درکودکان اندکی بابزرگسالان متفاوت بوده وواکنش به درددرسنین مختلف متنوع می باشدودرتشخیص این بیماری هم نقش مهمی رابعهده دارند.لذادربررسی بیماران نحوهٔبرخوردبادردمهم بوده وگاهی باعث تغییردرنتیجهٔتشخیص شده و از اهمیت خاصی برخوردارمی باشدکه توجه به آنهاتشخیص زودرس رادرپی داشته وزمان بستری طولانی وازایجادعوارض خطرناک جلوگیری نماید.این مسئله انگیزه ای شدتامطالعهٔاپیدمیولوژیکی وتعیین میزان خطای تشخیص ورابطهٔعلائم بالینی جهت تشخیص موردتوجه قرارگرفته واین مطالعه انجام بگیرد.

روش مطالعه :این مطالعه به روش توصیفی مقطعی بمدت ۵سال برروی ۸۲بیماردرسنین زیرسال بستری شده دربیمارستان قدس انجام شده وباتوجه به اینکه امکانات تشخیص درمراکزمختلف ،متفاوت بوده لذاباتوجه به علائم بالینی ووجوددرددرکودکان موردبررسی قرارگرفته است واین کارباطراحی پرسشنامه ای که شامل دوقسمت اطلاعات دموگرافیک (سن ،جنس ،زمان بستری ،زمان عمل و...)وعلائم بالینی وپاراکینیکی مثل سونوگرافی ،عکس ،نتایج آزمایشگاهی و....موردبررسی قرارگرفته وبادسته بندی اطلاعات علائم ،نشانه ها،کیفیت وشدت ومحل دردو...موردپایش قرارگرفته ونتایج زیربدست آمده است .

یافته ها:ازکل بیماران موردبررسی ۴۱/۸٪آپاندیسیت حادبدون عارضه ،۴۹/۴٪آپاندیسیت حادعارضه دارشده وتنها۷/۸٪مواردنرمال گزارش شده است .در۹۸٪مواردحداثردر۱۲ ساعت اول پس ازپذیرش وبستری تحت عمل جراحی قرارگرفته اند.عکس شکم در۷۶/۶٪نرمال و۲۰/۸٪غیرطبیعی ووجودگازدرشکم و۲/۶٪وجودتوده ای رادرشکم نشان می دهد. تب زیر۳۹درجه در۹۴/۹٪مواردیده شده وتنهادر۱/۵٪تب بالای ۳۹درجه دیده شده است .شروع درددر۲۵٪مواردرآپاندیسیت حادبدون عارضه درحداکثر۱۲ساعت قبل ازمراجعه ودرآبسه های شکمی بیش از۳۶ساعت ازدردشان گذشته است .درآبسه های شکمی دردبسیارمبهم گزارش شده است .کیفیت دردبصورت حاددر۷۴/۳٪مواردوراجعه در۲۳٪ودردهمراه باگریه در۱/۴٪وتنهادر۱/۴٪دردنداشته اند.در۱۳٪مواردآپاندیسیت باGEتشخیص افتراقی داشته است وUTIدر۷/۸٪وآدنیت مزانتریک در۶/۵٪مواردبوده است .علامت تهوع واستفراغ توأمدر۵۱/۹٪کودکان و۲۴/۷٪فقط استفراغ داشته اندو۱۱/۷٪بی اشتهائی به تنهائی و۶/۷٪هیچگونه علامت تهوع واستفراغ وبی اشتهائی نداشته اند.علت پرفوریشن آپاندیس در۴,۲۲٪مواردFECALو۷۰/۲٪التهاب آپاندیس و۹٪ناشناخته بوده است .

بحث ونتیجه گیری :باتوجه به اینکه جامعه موردبررسی مااطفال زیر۱۰سال هستندوهمکاری لازم باتیم درمانی وتشخیصی نمی توانندداشته باشندوعدم آگاهی والدین نسبت به درکودکان خوددربیماری هابایددقت بیشتری صورت گیرد.تأثیروجوددردشکم درتشخیص زودرس قابل توجه است چون والدین آگاهی لازم ازاین بیماری رانداشته ودردکودکان خودراموقتاًسرکوب کرده ودردکودک مخفی می شود.وجودعلائمی مثل بی اشتهائی توأم بااستفراغ احتمال تشخیص راافزایش می دهد.درنوپایان شکم تحریک پذیربوده وباکوچکترین تحریکی پایشان رابه داخل شکم جمع می کنند.GEدراطفال چون باآپاندیسیت بیشترین تشخیص افتراقی رادارذلذادراین بیماران بایددقت بیشتری شود. گرفتن عکس شکم درابتدای پذیرش که درآن وجوگازدرشکم رانشان می دهدودرتشخیص آپاندیسیت عارضه دارکمک کننده بوده وباید انجام شود.توجه به درجه حرارت کودک درکودکان مهم بوده وباتوجه به آن تشخیص راسرعت می بخشد.علت ایجادآپاندیسیت درکودکان التهاب آپاندیس بوده که یک واکنش تأخیری می باشدواهمیت زمان رانشان می دهد.کنترل رفتارکودکان نشان دادکه محکم نگه داشتن محل دردنشانهٔخوبی است که بایدبه آن توجه کرد.

پیشنهادمی شودکه :به والدین کودکان آگاهی داده شودکه هرگونه دردوتب رادرکودکانشان سرکوب نکرده وبه آن اهمیت بدهندوآنرا بررسی کنند.همکاران نیزبه علائم درددرکودکان محکم نگه داشتن محل درد،بی اشتهائی توأم بااستفراغ وتحریک پذیری شکم کودک بیشترتوجه نموده وتنهابه علائم پاراکلینیکی اکتفانکنند.چون زمان درکودکان برای جلوگیری ازایجادعوارض ومدت بستری وکاهش هزینه های بسیاربرای خانواده ها ومراکزدرمانی بسیارمؤثربوده،به آنهادقت نمایند.درمواردی که تب وجودنداشته ۵۰٪مواردآپاندیس طبیعی گزارش شده که بایدبه علامت تب توجه کرد.

روش نوین ژن درمانی در سرطان

دکتر مهرناز ناروئی نژاد

Gene therapy for cancer

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In 1970s some scientists proposed that in Some diseases caused by a known monogenic defect, theoretically, diseases could be treated and potentially cured by the insertion and expression of a normal copy of the mutant or deleted gene in host cells.
Then, the first approved gene therapy case in the United States took place in 1990. It was a treatment for a genetic defect of an Immune System deficiency. The effects were only temporary, but successful.

Recently, researchers are studying several ways to treat cancer using gene therapy. Some approaches target healthy cells to enhance their ability to fight cancer. Other approaches target cancer cells, to destroy them or prevent their growth.

The term gene therapy encompasses a wide range of treatment types that all use genetic material to modify cells (either in vitro or in vivo) to help effect a cure. Replacement of mutated genes with healthy genes, improving the patient's immune response to cancer, insertion of genes into cancer cells to make them more sensitive to chemotherapy and radiation therapy, introducing of “suicide genes” into a patient's cancer cells, inactivate the genes involved in the disease process are some approaches which are used for cancer gene therapy.
In general, a gene cannot be directly inserted into a person's cell. It must be delivered to the cell using a carrier, or “vector”. Viruses and liposomes are known as gene transfer agents or vectors. Oncolytic gene therapy vectors are generally viruses that have been genetically engineered to target and destroy cancer cells. Viruses have a unique ability to recognize certain cells and insert genetic material into them. In another approach, non-viral transfection using of naked DNA is used. Clinical trials carried out of intramuscular injection of a naked DNA plasmid have occurred with some success. The use of synthetic oligonucleotides in gene therapy is to inactivate the genes involved in the disease process. There are several methods by which this is achieved. One strategy uses “antisense” (called SiRNA) specific to the target gene to disrupt the transcription of the faulty gene.
As with any new type of therapy, there are serious safety concerns. Each days, newer and safer gene therapy delivery agents have been created and many of cancer patients globally have participated in gene therapy trials with remarkably few treatment side effects. Mammalian models of oncolytic gene therapy have worked remarkably well. In murine models, both colon and bladder cancer have shown survival benefits and reduced
metastasis using oncolytic viral agents.
Overcoming the problems, there are many benefits to oncolytic therapy.
Several very exciting cancer vaccine treatments are in late stage trials, and the field of cancer gene therapy will no doubt be part of the future of cancer therapeutics.

مزایای پورت گذاری در زیر فاسیای پکتورال در کودکان سرطانی

دکتر محسن روزرخ، دکتر احمد خالق نژاد طبری

THE ADVENTAGES OF SUBPECTORAL PORT IMPLANTATION IN CHILDREN WITH MALIGNANT DISEASE

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Backgrounds: The aims of this review are to analyze all the complications encountered with long term use of ports and discuss the possible ways of prevention in children with malignant diseases. With expanding use of ports, however new and frequent complications are being encountered.

Methods: 633 children aged 2 months – 14 years required port insertion during this study period (March 2000 – June 2009) for malignant disease. The most common underling malignancy was ALL (52.9%).All ports were implanted in the operating room under general anesthesia with open technique using internal jugular veins. Ports implanted subcutaneous in 180 cases in the early years of study and 453 ports implanted under pectoral fascia thereafter for better fixation and prevention of common complications. All information of sex, age, underlying malignancy, duration of catheter usage and complication analyzed by SPSS software as an uncontrolled clinical trial.

Results: From 2000-2009, 633 children with malignant disease requiring chemotherapy included this study.271 cases were female and 362 were male. Mean age of patients was 5.4 years (2months-14 years).The time the port stayed in-situ was more than 6 months in 474 cases , 3-6 month in 100 cases and less than 3 months in 109 cases .Port complications are noted in 64 cases including infection in17 cases, catheter tip dislodgment in 11,skin necrosis in 20 cases, and thrombosis in 4 cases, inability to remove catheter tip from jugular vein in 4 cases and others in 8 cases .

Conclusions:

In the first 4 years of this study, we routinely inserted ports sucutaneously, but due to skin necrosis and port chamber exit in 20 cases(usually by single team), procedure changed to subpectoral fascia implantation(by other single team).

We didn`t have skin necrosis in 453 cases with sub-pectoral implanted.

Removal of catheter tip simply done in majority of patients but with increasing time of usage in treated patients a newly complication reported in this study that rarely reported before. Many of our patients especially ALL cases after complete treatment and cessation of chemotherapy referred us for removal of port catheter, that rarely seen in adult patients. This procedure simply is done in operating room with a light sedation with incision on previous scar of port without vein insertion explore. In our series we have difficulties for removal of intravascular portion in 4 cases that need to explore of neck in one and limited thoracotomy in one case and two other cases removed by cardiologist intervention.

Abdominal Inflammatory Pseudotumor: case report and review of the literature

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Introduction: Inflammatory Pseudo-tumor is a rare benign mass lesion of various sites that mimics a true neoplasm. It is an unusual cause of chronic abdominal pain in children.
Aims: Inflammatory Pseudo-tumors can simulate true neoplasm both clinically and morphologically so surgeon should be aware for avoiding radical surgery.
Method (case): A 4-year-old girl admitted to the shahid Rajaei hospital of Qazvin province on January 2005 because of abdominal mass. She had a history of Five months abdominal pain. Abdominal CT Scan with contrast report was soft tissue sarcoma.
In laparotomy there was a hard, nodular mass which involved Jejunum, omentum with extension to bladder and sigmoid colon. The mass resected with a JeJunal segment and involved omentum with primary anastomosis.
Results: Patient discharged on 5^{th} post operative day uneventfully. Four years after operation follow up showed patient is well, free from recurrent abdominal lesion.
Conclusion: Abdominal Pseudo-tumor is not a Common lesion of abdomen. Most of the cases in the literature have been in the lung.
It is important to be aware of such benign lesion which may simulate malignant tumors, so that unnecessary radical treatment can be avoided.

Surgical treatment of hepatic tumors in children

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PURPOSE: Hepatectomy remains a complex operation even in experienced hands. The objective of the present study was to describe our experience in liver resections, in the light of liver transplantation, emphasizing the indications for surgery, surgical techniques, complications, and results.
METHODS: The medical records of 32 children who underwent liver resection for primary or metastatic hepatic tumors were reviewed. Ultrasonography, computed tomographic (CT) scan, and needle biopsy were the initial methods used to diagnose malignant tumors. After neoadjuvant chemotherapy, tumor resectability was Re evaluated by repeat CT scan and (PRE TEXT) in hepatoblastoma. Surgery was performed after complete evaluation for distant metastasis. Hepatic resection dictated by evaluation (Right or Left lobectomy or segmentectomy) after Hilar dissection and vascular isolation. Vascular anomaly it was suspected by Doppler ultrasound, CT angiography were performed. The hepatic artery and portal vein were dissected and ligated near their entrance to the liver parenchyma to avoid jeopardize the normal lobe.
RESULTS: Thirty two children with hepatic tumors underwent surgical treatment, 32 patients underwent liver resections. There were 22 cases of hepatoblastoma,10 children presented with other tumors, 2 inflamatory psedutumor, 2Mesenchymal tumor, 3 hepatocellular carcinomas, 1 GIST tumor, Hemangioendothelioma, an malignant epithelial tumor,. Hepatic resections included 28 right lobectomies, 5 right trisegmentectomies, and 3left lobectomies. Two case of heptoplastoma. That was unresectable, liver transplantation was performed. The overall mortality rate was 14.9%, and all deaths were related to recurrence of malignant disease. The mortality rate of hepatoblastoma patients was less than other malignant tumors (P = .04). CONCLUSION: The resection of hepatic tumors in children requires expertise in pediatric surgical practice, and many lessons learned from liver transplantation can be applied to hepatectomies. The present series showed no mortality directly related to the surgery and a low complication rate.

Hepatoblastoma: Epidemiology and Survival

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Background: Hepatoblastoma is the most common primary liver tumor in children, accounting for just over 1% of pediatric cancers. The etiology is unknown, but it has been associated with Beckwith-Weidmann syndrome, familial adenomatosis polyposis and low birth weight. The prognosis for patients with resectable tumors is fairly good; however, the outcome for those with non-resectable or recurrent disease is poor.
Methods and Materials: This is a cross –sectional non- randomized descriptive study. 30 patients were included according to the criteria. Treatment protocols were 8 pts (VCR+ADR+CIS), 6 pts (5-FU+VCR+END), 8 pts (VCR+ADR+END), 6 pts (5-FU) and the remaining other protocols.
Results: There were 18 males and 12 females (M/F=1.5/1). Median age was 49 mo (5-144 mo).We found 3 cases of precocious puberty and 1 case of tyrosinemia. Staging of patients were stage 1 (11 pts), stage 2 (0 pt), stage 3 (17 pts), stage 4 (2pts). Clinical manifestations were abdominal mass (100%), abdominal distention (63.3%), anorexia (56.7%), weight loss (50%), abdominal pain(53.3%), vomiting (16.7%), pallor (20%), jaundice (33.7%), fever (26.7%), diarrhea (13.3%), constipation (16.7%) and splenomegaly (20%).Median AFP were 53200 (range 2083-166000), median B-HCG 43.85 (range 8-150) and LFT in normal range. Hepatic involvement was right lobe 16 pts, left lobe 7 pts and both lobes 7 pts. Total surgical resection was done in 10 patients. 5-yr survival was 71.9 ± 10 percent with CI= 95%.
Conclusion: Hepatoblastoma in our center is a rare tumor (1%). The primary treatment is surgical resection. Prognosis of patients with respectable tumor and low stage are good.

Our experience with congenital mesoblastic Nephroma

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Congenital Mesoblastic Nephroma is the most common renal tumor in infants with a mean age at diagnosis of 3.5 months. The typical presentation is a newborn with abdominal mass, but in recent years a number of cases have been recognized prenatally with polyhydramnius .

The Neoplasm has three histologic subtypes:

Classic, cellular and mixed.

The cellular variant is the most common subtype

Complete Excision is curative for most patients with CMN.

Local recurrence and metastasis can occur. Particularly with the cellular varient of CMN.

Since seven years ago we had six Infants with Congenital Mesoblastic Nephroma. 2 females, 4 males. The youngest 3 days old and oldest 6 months old. The avarage age between three days to six months.

5 of them the left kidney were affected with tumor and one right kidney.

The pathologic subtype of all was cellular. All of them were treated by radical nephrectomy.

None of them had metastasis prior or post operation and no local recurrence.

None of them needs chemotherapy or Radiotherapy.

All of them are well now.

Wilms' Tumor: A 10 Year Retrospective Study

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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 in Tehran.
Methods: Fifty-five patients diagnosed as having Wilms' tumor were studied in the period between February 1992 and March 2002. Demographic features, mode of presentation, associated
Anomalies, the stage of tumor, histopathologic results, and the survival rates were evaluated.
Results: Of these 55 patients, 31 were males and 24 were females (M/F = 1.2). The mean age at the time of diagnosis was 45.2 months. 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].
Favorable histology was diagnosed in 54.5% and unfavorable histology in 43.6% of the patients.
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.
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 10 years, but with more adaptation to the National Wilms' Tumor Study treatment protocols better optimum results seem to be achievable.

تومور ویلمز و گزارش ۸۸ مورد

دکتر محمد هادی رفیعی

Wilms' tumor in children: 88 cases reported·

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Introduction: Wilms' tumor is an embryonal renal-tumor, the most common primary, malignant renal tumor in childhood, accounting for 5-6% of all types of cancer during this period. Wilms' tumor usually presents as an abdominal mass. Abdominal pain, fever , anemia , hematuria and hypertension are seen in 25-30% of children The average age at presentation is 42-47 months for unilateral wilms' tumor and 30-33 months for bilateral wilms' tumor . Diagnostic testing includes abdominal U.S, CT, MRI and biopsy for definitive diagnosis .CXR and chest CT may be required. 6% of wilms' tumors contain anaplasia. (Unfavorable histology) 10-15% of patients with wilms' tumor experience recurrence. Nephroblastoma is bilateral or multicentric in 5-10% of cases.
Thanks to early diagnosis, improvement in surgical and anesthesiological techniques, chemotherapy and radiotherapy and the close communications between the surgeon, pathologist and oncologist, the rate of survival has increased dramatically.
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 1370-1385 in training centers, Isfahan University. 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 (i.e.45.5%) survived and 48 (i.e.54.5%) died. Of the total number of the patients, 45.5% were male and 54.5% female. Of this number, 36 patients (i.e.41 %) were at stage 1. Eight (i.e.9.1 %) at stage II . 32(i.e.36.3%) at stage III. 12(i.e.13.6%) at stage IV and three (i.e.3.4%) at stage V. The mean age of the patients was 48 months for the females, 32.8 months for the males and 40.4 months for the total. The age range of the patients was 2.5-10 years and the highest disease frequency (i.e.44.1%) was for the age range 2-4 years. The mean age for stages I-IV was 43.7, 21, 24.2 and 42 months respectively. In 38.2% of the cases the tumor had affected the right kidney, in 58.8% the left kidney and in 3% bilaterally. The clinical signs and symptoms have included abdominal mass in 88.2%, fever in 23.5%, weight loss in 20.5%, gross haematuria in 23.5%, and microscopic haematuria in 29.4% abdominal pain in 29.4%, anemia in 47% and hypertension in 15%. In 17.6% of the cases the tumor was seen to cross the midline. In 11.8% of the cases the tumor recurred.
Associated anomalies, hypospadias, undescended testis and horse shoe kidney were seen in 8.8% of the cases. 91.1 % of the patients had favorable and 8.9% unfavorable histology. In 23% the cases, treatment included surgery only, in 50% surgery and chemotherapy combined and in 27% a combination of surgery, chemotherapy and radiotherapy. Four years survival was 95% and six years survival 32%. Taking into account the stage of the disease in stage I six years survival was 95% , in stage II 4.75 years survival was 95%, in stage III 1.5 years survival was 95% and in stage IV one year's survival was 95% .
Discussion: The long -term survival rate for the Wilms' tumor at low stages was 90% and at high stages 70%. The survival rare for our patients, in particular the long -term, was lower than the above statistics, i.e. 32% compared to 70%. The conclusion drawn from this study is that arrangements should be made to firstly enable diagnosis at early stages and secondly establishing close communications between the surgeon, pathologist and oncologist is essential to increase the long -term survival rate.

Nephron-Sparing Procedures In Wilms tumor
Report of two cases

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Wilms tumor or nephroblastoma comprises 87% of all pediatric renal tumors. It is the most common GU malignancy in children[1,2,3] The tumor is classified by the SIOP and NWTSG into five stages according to extent, infiltration of neighboring organs ,local or distant metastases and bilateral involvement. The treatment of WT, including chemotherapy, surgery and radiotherapy (when appropriate) differs substantially between the NWTSG protocol recommending primary surgery and the SIOP protocol favoring preoperative chemotherapy. The protocols by which our patients have been treated over the years were closely related to the NWTSG strategies performing primary surgery. Kidney-sparing surgery by tumor enucleation or resection of at least one kidney after preoperative chemotherapy is recommended approach in bilateral WILMS tumors according to both groups. A recent published study Showed a good outcome with a 5-year survival rate of 73% and a 10-year survival rate of 70%.[7]. The author considered the cumulative effect of surgical reduction of functioning kidney tissue, chemotherapy and/or radiotherapy responsible for renal dysfunction [7]. In contrast to bilateral WT patients, total nephrectomy is recommended in unilateral cases as the surgical procedure of choice. In contrast to bilateral WT patients, total nephrectomy is recommended in unilateral cases as the surgical procedure of choice. Based on excellent survival rate of more than 90% the question must be asked whether some carefully selected forms of unilateral nephroblastoma (easily respectable tumors, confined to one kidney pole, and excellent response to preoperative chemotherapy) could be treated by kidney tissue sparing surgery with the same favorable outcome. In the group of patients with unilateral WT presented here, we followed a renal salvage surgical strategy removing the tumor together with a rim of healthy renal parenchyma [8]. The major argument to prefer kidney-sparing surgery in selected patients comes from the local situs during surgery, when only a rather small and/or easily respectable remaining tumor is situated on one kidney pole. Resection of tumor through distant healthy tissue, thereby sparing at least half of the kidney, is a very strong option in these cases. Local radiotherapy should not be necessary; therefore it is important that para-aortic lymph nodes are free of tumor invasion. The danger of focal glomerulosclerosis caused by hyper filtration after unilateral ureteronephrectomy is discussed in a controversial manner in the literature [9]. Ritchey et al. reported a low risk of renal failure in children and adolescents with unilateral nephrectomy ranging from 0.2% to 0.4% [10]. On the other hand microalbuminuria ,proteinuria and a decreased GFR could be seen in adults as long term sequel of renal agenesis or unilateral nephrectomy[11,12].Thus ,kidney-sparing surgery in unilateral WTs may minimize the risk of late glomerulosclerosis and reduced renal function. A further argument for renal salvage procedures in unilateral WT is the advantage of renal tissue preservation in the case of secondary controlateral nephrectomy ,i.e. due to metachronous WT. Moorman-voestermans reported a patient suffering from unilateral WT who was saved from dialysis by partial nephrectomy because the contolateral kidney had to be removed after a renal trauma [13].
The major argument of critics concerning renal salvage procedures in unilateral WT is that, partial nephrectomy bears the risk of leaving a tumor remnant in situ ; especially when the tumor is anaplastic,the outcome would be markedly worse[14,15] .It has to be pointed out that a characteristic of anaplastic nephroblastomas is their resistance to conventional chemotherapy. Therefore only in selected group of patients ,who responed to preoperative chemotherapy (>50% tumor volume reduction rate).have to be considered to partial nephrectomy.Intra-operative opening of the tumor is similar to tumor rupture with local spillage ,which then needs postoperative radiotherapy of tumor bed[16,17] Therefore, concerning the surgical technique in partial nephrectomy , it is important to be radical,which means that the surgeon always has to resect a rim of normal renal tissue. If the resection margin turns out to be uncertain,intra-operative ultrasound should be carried out to clearly mark the resection line.

In contrast tumor enucleation is preferred in bilateral WT .Cozzi recommends this procedure as nephron-sparing surgery in unilateral WT,too,whereas Gulielmi et al, showed that tumor enucleation of an unilateral WT is a “nononcologic”procedure,because only in cases with a thick histologically confirmed tumor pseudocapsule is complete tumorectomy predictable[10,9].
As mentioned above an important indicator of the feasibility of partial nephrectomy is the tumor response rate after preparative chemotherapy of at least 50% [20]. In thease cases even patients with very large tumor con be treated by partial nephrectomy.
Moorman-voestermans et al. described a 87% accuracy rate in predicting the possibility of renal salvage procedures in unilateral WT by preoperative imaging techniques[13].However our experience showed that only the intra-operative situs is decisive for the definitive surgical approach.
We had two cases of Partial nephrectomy in our patients.The 1st one was a 7 months of old male with left upper pole tumor ,and the 2nd was a 2years of old male with right lower pole tumor.Both tumor was removed with free margin renal tissue .In both cases the histologic study revealed favorable pathology.The three years follow-up was uneventful. .
Conclusion;
PN in unilateral WT in general cannot be recommended yet, but promises to be a reasonable alternative surgical strategy to tumor nephrectomy in some patients. It preserves renal tissues and potentially can save the child from dialysis in cases of controlateral nephrectomy resulting from renal trauma or metachronous WT. However resection through healthy kidney tissue is mandatory, because tumor spillage due to incomplete resection bears the risk of local relapse, which would make nephrectomy and postoperative local radiotherapy necessary. Important prerequisites for a decision for a partial nephrectomy in unilateral WT are a preoperative tumor volume reduction rate of at least 50% measured by abdominal ultrasound, confinement of the tumor to one renal pole and or easy respectability, preservation of at least 50% of the renal tissue and histologically free para-aortic lymph nodes. While tumor enucleation in unilateral WT cannot be recommended as a safe strategy, in bilateral WT, tumor enucleation or kidney-sparing resection after preoperative chemotherapy remains the surgical procedure of choice,because it prevents the child from having dialysis and renal transplantation.

تجربهء چندسالهء تومور ویلمز مرکز علی اصغر

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The evaluation of the Treatment of Wilm's Tumor in Ali Asghar children Hospital in a Period of 10 years.

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Introduction: Wilm's tumor is the common renal malignancy observed in childhood, With an incidence of 8 in 1 million children younger than 15 years of the age (1,2). the management of the Wilm's changed dramatically in the advent of radiotherapy and later chemotherapy. surgical treatment alone led to a survival of only 20% , but the addition of other treatment modalities has led to an overall survival of greater than 85%. Nephrectomy plays only a part in the overall management of these patients. Early diagnosis and treatment also has effective role on the survival of the patients. The majority of children with wilm's tumor present with an asymptomatic abdominal mass, but hematuria is a feature in 15%, fever in 20% and abdominal pain in 10% . Hypertension occurs in 10%. A minority have feature of Aniridia, hemihypertrophy or genital anomalies. The diagnosis could be performed by ultrasound, IVP, Contrast CT Scan and MRI. Regarding the histopathology in NWTS-S (National Wilm's Tumor Study) Staging system, classified in 5 groups as stage I , II , III , IV and V. The stage V identifies the bilateral wilm's tumor.

The aim of this retrospective study is to evaluate the clinical presentation, treatment and survival of the patients who admitted to Ali Asghar children Hospital during last 10 years.

Material and Method: From January 1999 to January 2009, 72 patients medical record who admitted with wilm's tumor to Ali Asghar Hospital, have been evaluated. 38 (52.8%) were girls and 34 (47.2%) were boys. The youngest was 2 days old and the oldest was 10 years old with 3,4 years of mean age. The site of the tumor was at right in 37 (51.4%) and were at left in 31 (43.1%) patients. In 3 cases (4.2%) were bilateral and one case was unknown in his medical record. The clinical presentation were abdominal mass in 65 (90.28%) patients, abdominal pain in 16 (22.22%), Hematuria in 14 (19.42%), Disuria in 10 (13.89%), loss of weight in 15 (20.83%), Fever in 3 (4.17%), Hypertension in 2 (2.78%), Anemia in 23 (31.94%) cases. In one patient respiratory distress due to lung metastases have been detected. The disease was recognized by mother and father in 34 cases and by physician in 17 patients and accidentally after trauma in 2 cases. In one case has been diagnosed by antenatal ultrasound.

For certain diagnosis, ultrasound, have been done for all cases, CT Scan in 58 patients and MRI in 2 patients have been done, to rule out the lung metastases, chest X Ray have been done for all cases. The least duration between diagnosis and treatment was 1 day in 40 (55.5%) cases and the longest time was 75 days.

The surgical treatment was radical Nephrectomy (33 left and 38 rights) in unilateral group and radical nephrectomy in advanced kidney and partial nephrectomy for the other kidney in bilateral type were performed. In 2 cases with renal vein and IVC thrombosis, evacuation has been done.

The Results: Regarding to the time of operation the least time for follow up was one month and the longest was 10 years. In periodic examination until January 2009, 51 patients were alive. In one case, 4.5 months after operation, recurence of the tumor have been seen. In the other one, after right radical nephrectomy, wilm's tumor has been seen in left kidney. In one case, left femur metastases have been detected after left nephrectomy. One case died 8 months after operation. We could not follow up 19 cases.

The early chemotherapy has no effect on survival of the patients.

Conclusion: Early diagnosis and treatment have important effect on survival of the patients with wilm's tumor. If the patient is in stage I, he or she does not need chemotherapy. The radyotherapy has most effect on survival for advanced cases.

While the time of starting chemotherapy does not affect the survival of the patients.

تجربهء چندسالهء تومور ویلمز در مرکز محک

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Abstract: Wilms tumor is the most common childhood renal tumor accounting for about 6% of pediatric malignant disease. Most patients with Wilms tumor can be cured with treatment and subsequently lead normal life. The multidisciplinary management of Wilms tumor has resulted striking improvement in survival of more than 85% nowadays and has become a paradigm for successful cancer therapy. We describe the results of patients treated according to National Wilms Tumor studies (NWTS) 3-5, with surgical staging, Central pathology review and multimodality treatment. This is a historical cohort study on the all patients who had Wilms tumor. We used the existing files of all patients who had admitted to Ali Asghar Children's hospital with Wilms tumor in the years of 1990-2003. The patients evaluated for age, sex, histologic type of cancer, metastasis, outlook of relapse and outcome after 5 years from diagnosis. We analyzed 175 files of Wilms tumor. They are 49.7% are male and 50.3% female. Mean age (±S.D.) of patients at diagnosis was 3.8±0.4 year. The 5-year survival rate of these patients was approximately 76±4%. History of cancer in first degree of relative was 11.5% and family marriage was 36.4%. Tumor involvement were 45.3% in right kidney, 51.5% in left kidney and both kidney involvement in 3.2%. Congenital anomalies in association with Wilms tumor were urologic problem (1.5%), hemihypertrophy (0.5%), sporadic aniridia (0.5%) and without abnormalities (97.5%). Histologic type of tumor were 32.6% favorable, 65.2% unfavorable and 2.2% intermediate. Stage II and III were the most common (35.4 and 32.4%, respectively). Tumor relapse were occurred in 25.4%.

Key words: Wilms tumor, hemihypertrophy, epidemiology, survival

Cancer genetics

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Cancer genetics has for many years focused on genes are involved in cancer procedure and mutational events that have their primary effect within the cancer cell. Recently that focus has widened, with evidence of the importance of environmental genetics or epigenetic events and of cellular interactions in cancer development. The role of oncogenes, tumor suppressors and common genetic variation in determining the range of individual susceptibility within the population is increasingly recognized, and will be addressed using information from the Human Genome Project.

Clinically relevant genetics knowledge is essential for appropriate assessment and management of inherited cancer risk, and for effective management and communication with patients. The knowledge regarding basic cancer genetics concepts early in the process of introduction of predictive genetic testing for breast/ovarian and hereditary non-polyposis colorectal cancer (HNPCC) syndromes and other common cancers are necessary for physicians.

Our research directions in cancer/testis genes, the genes which are specifically expressed in testis and different cancers such as TSGA10 will highlight determinants of cancer that lie outside the cancer cell, for example, immune responses, suggest new targets for early detection, and inform the design of strategies for prevention in groups at increased risk and new cancer therapy methods.

Principles of performing biopsy in children's cancer

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Introduction: biopsy is a procedure in which tissue samples are removed (with a needle or during surgery) from the body for examination under a microscope; to determine if cancer or other abnormal cells are present. Biopsy procedures are separated into two categories that consist of targeted lesion biopsies or non lesion organ biopsies. Biopsies of identified lesions may be performed for primary diagnosis or for confirmation of a recurrent neoplasm.
Type of biopsy: there are many type of biopsy, such as: FNA Biopsy, Cone Biopsy, Core Biopsy, Suction assisted Core Biopsy, Endoscopic Biopsy, Punch Biopsy, Surface Biopsy and Surgical Biopsy (or Excisional Biopsy). The surgeon will determine the most appropriate method of biopsy, which would be based on various factors, including:

- the tissue, organ or body part to be sampled
- how suspicious the abnormality appears
- the size, shape and other characteristics of the abnormality
- the location of the abnormality
- the number of abnormalities
- other medical conditions a patient may have
- the preference of the patient, and
- the imaging and biopsy systems available at a given hospital or healthcare location

Biopsies are usually guided by the method that identifies the abnormality best. Palpable lumps can be felt and therefore no additional guidance is needed in most cases but sometimes the lesions discover by an imaging, for example Ultra sonography, MRI or CT, in these cases usually the biopsy will be done under the guide of some modalities that will show the lesion the best.
In this brief lecture; Indications, type of biopsy, preparation of the patient and the site of biopsy, the technique of the procedure and complications will be discussed.

Congenital Rhabdomyosarcoma of right shoulder: a case report and review of literatures for decision making

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Purpose: Rhabdomyosarcoma is the most common soft tissue malignancy of childhood, but may occur extremely rarely in the neonatal period.
We report a congenital rhabdomyosarcoma of right shoulder and summarize recent information on congenital rhabdomyosarcoma.

Case presentation:
A 16 day old female baby was referred with congenital swelling on right shoulder. Baby was the 3rd born child to a healthy mother at 38th weeks of gestation by normal vaginal delivery followed by dystochia. On examination, there was a hard, round, ecchymotic, nontender, slightly movable, warm & shiny mass of size 10x15 cm on right axillary pits that was extended to neck right chest wall. The mass separated between shoulder & chest wall with paralysis of right hand. CXR showed a round soft tissue shadow. Ultrasound showed a huge, heterogenic hypoechogenic mass with 20 Cm in size and enhanced vascular pattern and suspected to hemangioma. MRI with contrast demonstrated a soft tissue mass suspected to multicentric hemengioma. All laboratory profiles were normal. Patient was treated with corticosteroid. The mass rapidly increased in size despite aggressive steroid therapy with rupture and bleeding. On 45th post natal day neonate was taken to operating room to control the bleeding and if possible total excision of the mass. The mass separated easily from surrounding and excised with right upper extremity amputation. At the end of surgery the baby had cardiac arrest, so she died because of DIC. The final pathology report was rhabdomyosarcoma.

Conclusion:
Congenital huge rhabdomyosarcoma (RMS) is very rare. The treatment for neonatal RMS requires a multidisciplinary approach where surgery and chemotherapy both have their own specific role. When the tumor size is huge we should consider biopsy and chemotherapy followed by surgery.

Deltopectoral vein cut-down to reach central veins in children Who undergo chemotherapy

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Introduction: Although routine vein access in children's chemotherapy is Subclavian or Jugular
Veins but we choose Deltopectoral vein and here are the results of our experiences.
Method: A random prospective study was done in patients who need chemthrapy. 18 out of 42 patients that need vein access for chemotherapy during from 1385-1388 underwent Deltopectoral cut-down.in rest of patients other veins were used (jugular subclavian and saphenus veins) and these two groups were compared.
Results: 42 patients undergone catheterization in order to reach central veins By using percutanous (seldinger method), cut down. Vein expose prospective Results were recorded and revealed that bleeding and operation time are less in Deltopectoral cutdown in comparison to other methods.
Moreover there is no need to specific equipments and post operative complications such as pneumothorax, hemothorax and thrombosis were not seen.
Conclusion: Deltopectoral cut down in deltopectoral fold of shoulder can an alternative method insertion of chemothrapy infusion pomps,with less complications.

Mediastinal masses, case series and its approach

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Mediastinal tumors are relatively uncommon in children. Here we present our experience with these lesions, their presentations and treatment. We also discuss the algorhythm for approaching the mediastinal cysts and solid lesions.

We reviewed the charts of the patients with mediastinal lesions who were operated within last two years in Namazi and Shiraz Mother and Child Hospital. The presentation, pathology and surgical approach were reviewed.

Results: There were 15 patients with mediastinal masses. The age-range was from 6 months to 16 years. There were 9 Males and 6 females. Twelve patients presented with respiratory distress. One patient developed lower limb weakness. Two had dysphagea. The histologic study showed 6 lymphoma, 2 PNET, 2 Neuroblastoma, 2 lymphangioma, 1 endodermal sinus tumor, I thymic hyperplasia and one ganglioneuroma. All patients underwent total to subtotal resection in first or second operation except for the lymphoma patients who were only biopsied.

The systematic approach will be discussed.

ندول تیروئید در کودکان: تجربهء مرکز مفید

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Thyroid nodules in childhood: a single institute Experience

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Background/purpose: Thyroid nodules are rare in children, and multiple diagnostic modalities are used to evaluate the thyroid mass. The aim of this study was to determine the result of management of thyroid nodule in children, and the special attention to the role of fine needle aspiration biopsy (FNAB) in diagnosis.
Materials & methods: 32 children who underwent surgery for thyroid nodules in Mofid children's hospital within 10 years (1995 to 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 and were analyzed statistically for associations with the presence of thyroid cancer.
Results: 25 patients (78.1%) girls, and 7(21.9%) were boys. Mean age was 10.9 years (range of 8 to 14 years). 24(75%) patient had benign and 8 (25%) had malignant tumors. 18(56.25%) nodules were located in right lobe. Statistical analysis revealed sensitivity, specificity, accuracy, and positive and negative predictive values respectively 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.
Conclusion: Clinical judgment as determined by serial physical findings continuous to be the most important factor in the management of the thyroid nodules in children. FNAB is the most accurate method of investigation and its accuracy is improved by USG guidance.

آیا کیمو رادیوتراپی در کودکان با سرطان کولون مفید است یا مضر؟

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Is chemo-radiotherapy has benefit or harmful in children with Adenocarcinoma of colon: Case series

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During past 10 years we have 4 patients with adenocarcinoma of colon (ACC). Of these, 3 patients received chemo-radiotherapy (CR) and 1 patient without CR. One of the 3 patients who used CR developed secondary tumor. All patients who received CR expired, but that patient who has not received CR is alive (5 years after operation) without recurrence. Therefore, is CR appropriate for this very malignant pediatric carcinoma or harmful (besides cost of CR)?
Because of rarity of this tumor and few our cases we cannot promptly suggestive best management. But these cases can add information about ACC.

Rhabdomyosarcoma

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Rhabdomyosarcoma is a primitive soft tissue tumor that arises from mesenchymal tissues. The most common sites of origin are the head and neck (36%), extremities (19%), genitourinary tract (21%), and trunk (9%), although the tumor can arise virtually anywhere. The clinical presentation of the tumor depends on the site of origin. The diagnosis is confirmed by the findings of incisional or excisional biopsy after evaluation by MRI, CT of the affected area and the chest, and bone marrow biopsy. The tumor grows locally into surrounding structures and metastasizes widely to lung, regional lymph nodes, liver, brain, and bone marrow. The staging system for rhabdomyosarcoma is based on the tumor, nodes, and metastasis (TNM) system as established by the Soft Tissue Sarcoma Committee of the Children's Oncology Group. It is shown in Table 1-1. Surgery is an important component of the staging strategy and involves biopsy of the lesion and evaluation of lymphatics. Primary resection should be undertaken when complete excision can be performed without causing disability. If this is not possible, the lesion is biopsied and intensive chemotherapy is administered. It is important to plan the biopsy so that it does not interfere with subsequent resection. After the tumor has decreased in size, resection of gross residual disease should be performed. Radiation therapy is effective in achieving local control when microscopic or gross residual disease exists after initial treatment. Patients with completely resected tumors of embryonal histologic type do well without radiation therapy, but radiation therapy benefits patients with group I tumors of alveolar or undifferentiated histologic type.

Prognosis:

The prognosis for rhabdomyosarcoma is related to the site of origin, resectability, presence of metastases, number of metastatic sites, and histopathologic features. Primary sites associated with more favorable prognoses include the orbit and nonparameningeal head and neck, paratestis and vagina (nonbladder, nonprostate genitourinary), and the biliary tract. Patients with tumors <5 cm in size have better survival than children with larger tumors, and children with metastatic disease at diagnosis have the poorest prognosis. Tumor histologic type influences prognosis; the embryonal variant is a favorable type, whereas the alveolar type has an unfavorable prognosis.

Treatment, out comes & complications after resection of hepatoblastoma

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Background/Purpose:
Hepatobelastoma comprise only 1% of all cancer in childhood and is the most common malignant liver tumor, which accounts for 79% of hepatic neoplasia in children, less than 15 years of children. The aim of this study was to review the treatment, out comes & complications after resection of hepatoblastoma.

Methods:
From 1997-2009, 19 patients with hepatoblastoma were treated in Alzahra Hospital, Isfahan. I review retrospectively, focusing on the treatment, out comes & complications.

Results:
Between 1997-2009, 19 children were treated for hepatoblastoma (10 females & 9 male), The age range was 4 months to 16 years old (median, 27 months).The right lobe were involved in 13 patients(68.42%),
the left lobe in 3 children (15.8%) & 2 lobes 2 patients (10.52%),right lobe and lung metastasis 1 children(5.26%).Treatment includes biopsy ,chemotherapy ,surgery & chemotherapy after resection. Surgical procedures includes ,right segmentectomy 10+2, right lobectomy 5 ,left lobectomy 2 patients, non anatomic resection 1 & extended hepatectomy 1. Complication occurred intraoperative (3), hemorrhage from the contralateral lobe 1,bleeding from IVC (repaired vena cave (2)).The length of follow up was (6 month,12 years).
Survival rate at:
1 years was 16 (84.16%), 2 years survival 12 (62.86%), 3 years survival 10 (52.6%), 4 years survival 3 (15.78%),5 years survival 0.

Conclusion:
The chemotherapy was found to greatly contribute to improved survival rate of hepatoblastoma patients. Preoperative chemotherapy resulted in an increased resectability of the tumor. As postoperation chemotherapy played an important rate in the increased cure rate of cases with either an incomplete tumor resection or metastasis.

MODERN RADIATION TREATMENTS IN PEDIATRIC ONCOLOGY

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ABSTRACT

Background. Radiation treatments in pediatric oncology have considerably evolved in the last ten years owing to tremendous advancements in imaging modalities and radiation delivery techniques. These improvements should lead to better cure rates and fewer late complications in children treated for cancer.

Patients and Methods. Children affected with various cancers. All imaging procedures and treatments were performed in children in the treatment position with customized immobilization masks. A 3 dimensional conformal therapy (3D-conformal radiotherapy) was devised using a modern treatment planning system and delivered with a linear accelerator.

Results. Two hundred and seventy children with multifarious cancers have been treated at Mahak hospital since January 2008. All imaging modalities (CT scan and MRI) were performed on children in the treatment position. Most children were given a 3D conformal therapy using 6 or 15 MV photons. In a few cases, patients received treatments using an electron beam (5 to 14 MeV). Portal X rays were systematically performed at the start of treatments to ensure the accuracy of radiation fields.

Conclusions. Modern radiation treatments can be delivered to children with great accuracy and fewer side effects.

Keywords: conformal radiotherapy, pediatric cancer, treatment planning system

Transfusion in childhood cancer

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Transfusion therapy has a significant role in successful management of children with cancer and recipients of hematopoietic stem cell transplant. Invasion or replacement of bone marrow by malignant cells and myelosuppression following chemotherapy are the main causes of pancytopenia in children with cancer. Cancer related anemia is multifactorial and often has both acute and chronic components including:

1- Replacement of the bone marrow with malignant cells
2- Myelosuppression due to chemotherapy
3- Anemia of inflammation
4- Blood loss due to thrombocytopenia or cancer surgery
5- Hemolysis

According the study conducted in many large pediatric oncology centers in Europe, it has been shown over 80% of patients were anemic regardless of tumor type. Packed red cell transfusions are indicated for patients whose Hb is less than 7-8gr/dl or whose Hb is greater than 8 gr/dl but are cardiovascularly unstable or in respiratory failure or bleeding.
Residual transfusion risk of major viral infections for HCV, HIV and HBV after screening with serology and NAT tests are 1 in 1.5×10^6, 1 in 1.7×10^6 and 1 in 174000 receptively. In order to reduce the risk of transfusion transmitted infections unnecessary transfusion should be avoided and alternative strategies for assuring blood safety should be in consideration. It is important that the use of rHuEPO for the treatment or Prevention of cancer associated anemia in children is carried out within Controlled, randomized clinical trials. Whether rHuEPO therapy will eventually replace RBC transfusions is difficult to predict and current evidence of its efficacy is limited in children. Patients with hematologic malignancies and Hodgkin′s disease and also the cancer patients undergoing intense chemotherapy or receiving fludarabine should receive irradiated Blood components which is currently available in Tehran Blood transfusion center. Cancer patients undergoing intensive chemotherapy are also at risk for significant CMV infection. Most physicians prescribe blood components with a minimal risk of transmitting CMV. In practice this is done either cellular blood components collected from donors who are seronegetive for antibody to CMV, or those components effectively leukoredced to a level of <5000000 white blood cells (WBCs) per unites or combining both.
Platelet is another main component of Blood which is frequently used to treat bleeding caused by thrombocytopenia in pediatric cancers. Platelet count of Patients is better to be maintained ≥ 100000 /μ l for CNS bleeding or planned CNS surgery, ≥ 50000 /μl for actively bleeding patients or undergoing other major surgeries and as prophylactic transfusion (prevention of bleeding), Platelet administration should be carried out when the platelet count is less than< 10000 / μ l . For children over 1o kg a dose of 1 platelet unit per 10 kg should increase the platelet count by approximately 50000 / μl .Aphaeresis platelet is the product obtained when donor blood is processed by automated aphaeresis equipment, which extracts the desired component and returned the remainder of the blood to the donor. One aphaeresis platelet unit represents at least one"dose"of platelet for a large child or average adult and is comparable to a pool of 4 to 6 units of whole-blood –derived platelets.

Cancer Diagnosis and Treatment at Molecular Level (Specifically Lung Cancer)

Sheikhnejad R

Lung cancer is the leading cause of cancer mortality worldwide. More patients die from lung cancer than prostate, breast, and colorectal cancer combined. Approximately 85% of the patients with lung cancer will have non–small cell lung cancer (NSCLC), which is frequently subdivided into the squamous, adenocarcinoma, large cell. The most common histologies are: adenocarcinoma (~50% of cases), squamous cell (~20%), and large cell (~10%).

While surgery remains the optimal treatment for early stage non–small cell lung cancer (NSCLC), 5-year survival rates for resected NSCLC without additional treatment range from 23% for Stage IIIA disease to 67% for Stage IA disease. Recurrences that account for mortality occur most commonly at distant extrathoracic sites. The goal of effective systemic therapy in patients with resected NSCLC is to eradicate micrometastatic disease, reduce the risk of recurrence, and improve survival.

Lung carcinoma is usually classified by microscopic morphology and immunohistochemistry. The development of microarray methods makes it possible to search for molecular markers of cancer classification and outcome prediction. Molecular classification is not in clinic yet but this will eventually provide a better tool for staging and prognosis. Identification of cancer biomarkers will provide better pharmaceutical targets to treat cancer at molecular level as well. Some novel "targeted" therapies have been introduced into adjuvant protocols. Agents such as Iressa (gefitinib) and Tarceva (erlotinib) that target the epidermal growth factor receptor (EGFR) and the vascular endothelial growth factor (VEGF) are in clinic. Several prognostic biomarkers have also been studied, including k-ras mutation and p53 mutation. Investigators have used gene-expression profiling to develop the lung metagene model in an effort to refine assessment of prognosis and identify patients at the highest risk of recurrence.

KRAS accounts for 90% of *RAS* mutations in lung adenocarcinomas, and approximately 97% of *KRAS* mutations in NSCLC involve codons 12 or 13; mutations are uncommon in squamous cell carcinomas. Gene expression profiling has been used as molecular diagnostic tool for classification and staging of cancers. It can be also effectively used to determine the molecular targets for personalized treatment. We have used a panel of 11 genes expression signature to characterize surgically removed human lung cancers specimens as well as patient's noncancerous lung tissues. The panel includes 6 well studied oncogenes such as bcl-2, c-myc, ki-ras, c-ha-ras, her-2/neu and Tgf-α that represent excellent therapeutic targets. Other genes are, p53 (best known tumor suppressor), MDM2 (known for regulating p53), Mmp1 and Mmp14 (metastatic genes) and MDR1 (a well known drug resistant gene).

گزارش یک مورد نادر آترزی مری به همراه تنگی قسمت دیستال مری

دکتر منصور شیخ – دکتر منصورمولاییان

Congenital Esophageal Stenosis with Esophageal Atresia / Tracheoesophageal Fistula

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Introduction: Congenital Esophageal Stenosis (CES) can be associated with Esophageal Atresia / Tracheoesophageal Fistula (EA/TEF). It is relatively rare.

It is frequently difficult to make a pre and post operative diagnosis of the distal CES associated with EA/TEF. Children with EA/TEF sometimes have post repair esophageal problems such as anastomotics stricture, gastroesophageal reflux and poor esophageal peristalsis. CES associated with EA/TEF can cause symptomatic obstruction after repair of EA/TEF and sometimes make second surgical intervention necessary. However pre or intra operative diagnosis of the distal CES associated with EA/TEF is difficult. Although radiographic finding of CES have been well described in infants and young children as a fixed intrinsic narrowing of the esophagus. Little is known about CES associated with EA/TEF. An abnormal connection between the respiratory and digestive tubes during embryonic development results in the creation of a TEF “and an abnormal incorporation of the respiratory tissue in the esophageal wall results in CES. For this reason ‘CES has been associated with children with EA/TEF.

Case report: A neonate ‘3.100kg weight and was delivered by C/S. The neonate was diagnosed with EA/TEF after delivery. He was 2 days old when operated on. The anastomosis has been done without tension via Rt thoracotomy approach. I passed the NG tube through anastomotic site easily. 3 days after surgery in retoplureal drain salivary secretion was observed. 7 days after operation barium study showed stenosis in distal part of esophagus. Leakage of salivary stopped after 2 weeks. Afterwards was candidate for reoperation to resect stenosis. This procedure was done by Rt thoracotomy and end to end anastomosis. During this period he was under coverage of suitable antibiotic therapy .4 day after second surgery he tolerated milk per oral and discharged the hospital. During recent visits his general condition was favorable and had gained weight.

کنترل درد در کودکان
دکتر سهیل بن رضوی

Pain Control

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All children, including neonates, experience pain. Therefore, the careful recognition and management of pediatric pain represents an important component of the perioperative management of all pediatric surgical patients. A range of pain management options are available that can improve the child's as well as the parents' comfort. The use of a pacifier, which may be dipped in sucrose, has been shown to decrease crying time and neonatal pain scores after minor procedures. Additional analgesic modalities include the use of topical anesthetic ointment (cream containing a eutectic mixture of local anesthetics) and the use of regional anesthesia, such as caudal blocks for hernias and epidural or incisional catheter infusions (On-Q system) for large abdominal or thoracic incisions. For situations in which more pain is expected, IV narcotic agents should be used. Morphine and fentanyl have an acceptable safety margin and can be administered judiciously to neonates and children. A recent randomized trial showed that administration of a morphine infusion in neonates receiving artificial ventilation decreased the incidence of intraventricular hemorrhage by 50%. In neonatal surgical patients who have been given large concentrations of narcotics over a prolonged period, transient physical dependence should not only be expected but anticipated. When narcotics are discontinued, symptoms of narcotic withdrawal may develop, including irritability, restlessness, episodes of hypertension, and tachycardia. Early recognition of these signs is essential, as is timely treatment using a thoughtful weaning schedule, appropriate assessment criteria, and administration of naloxone and other agents. In the postoperative period, patient-controlled analgesia is another excellent method of pain control. Additional means of achieving adequate pain control in children include the use of epidural analgesia and paraspinal blockade, which can be commenced at the time of surgery. By ensuring that the pediatric surgical patient has adequate analgesia, the surgeon ensures that the patient receives the most humane and thorough treatment, and provides important reassurance to all other members of the health care team and to the family that pain control is a very high priority.

Timing in pediatric surgery operations. Which is the difference?

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OBJECTIVE: To access the difference between the duration of common operations in children performed by a pediatric surgical team and a general surgical team in a developing province of Iran. METHODS: This retrospective study was carried out in Zanjan during 2009 in a single teaching hospital having below standard operating, anesthetic and recovery facilities. Operations were carried out by 2 different surgical teams (team A: pediatric surgical and team B: general surgery) with different sets of anesthetists. The duration of the procedures was recorded.
RESULTS: In a 2-month period a total of 10 surgical procedures at random were compared from both groups. No operations were done in children under 1-year of age by team B. The average age was 8 month-old and average weight was 6 kg for team A and for team B the average age was 5 years-old and the average weight was 12 kg respectively. The average operating time was 50 min. for team A and 73 min. for team B. Unfortunately, anesthesia time and recovery time were not recorded separately.
CONCLUSION: Based on the above experience, we recommend that a variety of common pediatric operations are better to be performed by a pediatric surgical team, which not only includes a pediatric surgeon but an exclusive pediatric surgical scrub nurse as well. In this way, the procedures are standardized and routines established in order to achieve better results.

مراقبتهای پرستاری در تومور ویلمز

افسانه افتخاری منش سوپروایزر آموزشی بیمارستان کودکان بهرامی

Nursing Care in Wilm's Tumor

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در زندگی روزهایی سپری شده اند که در آنها بیماری مشکل اصلی کودک و والدین بوده و آنها را به شدت نگران می کند .بخصوص هنگامی که کودک خود را در بستر بیماری می بیند، باید بداند که والدین، پزشک وپرستار در کنار او بوده تا هرچه سریعتر بهبود یابد ومصرف داروها،عمل کردن به دستورات پزشک وپرستار و توصیه های پدر ومادر تنها راه باز یافتن سلامتی دوباره اوست.طبق آمارهای ارائه شده در کشور آمریکا سالیانه ۵۰۰کودک مبتلابه تومور ویلمز با حدود تقریبی یک کودک در ۱۰۰۰۰گزارش شده است. تومور ویلمز در ۸۰٪موارد بین سنین ۵-۳سالگی شیوع دارد.

یافته های کلینیکی ممکن است به صورت توده شکمی ،درد شکم،علایم گوارشی، افزایش فشار خون و به ندرت هماچوری باشد. در ده های اخیر هماهنگی بین جراحان اطفال، انکولوژیست، نفرولوژیست ،رادیو لوژیست و پرستاران به دلیل متعدد بودن عوارض ناشی از بیماری ،متعدد بودن درمانها و ارتقاء سطح سلامت کودکان تا حد بسیار زیادی توصیه شده است.

همچنین تشخیص صحیح همراه با مشخص نمودن مرحله بیماری و پروتکل های در مانی از عوامل بسیار مهم در درمان می باشند.بدلیل آنکه تومور ویلمز بیشترین تومور کلیه در بچه های سن ۶ماه وزیر ۱۲سال می باشد پرستاران باید توجه زیادی به کودکانی که با تشخیص توده شکمی بستری می شوند داشته باشند.

زندگی با بیماری مزمن نه تنها بر کودک بیمار و خانواده او بلکه بر افراد حرفه ای ارائه دهنده مراقبت نظیر پرستاران ،اثرات زیادی بر جا می گذارد.

در یک مطالعه طولانی مدت ۱۳۶۲کودک مبتلا به سرطان مورد بررسی قرار گرفتند که۲۳/۴٪کودکان یک بار تهدید شدید زندگی را بر اثر پروستیجرهای درمانی تجربه نموده و۶۴٪دیگر (مانند کودکان مبتلا به تومور ویلمز) این تهدید را جدی تر تجربه نموده اند .

بنابراین پرستاران باید با برقراری ارتباط مناسب با کودک و خانواده اشان و سایر اعضاء تیم در مانی و با ارائه آموزشهای لازم تاثیر مهمی در سلامتی روانی وجسمی آنها گذاشته و برای مراقبت، ارتقاء سطح سلامتی و بهبود کیفیت زندگی آنها برنامه ریزی و نقش حمایتی داشته باشند.

مراقبت از پورت در کودکان

منیره خاموش چشم، آذر عرب
بخش جراحی ۱ بیمارستان کودکان مفید

CARE OF THE IMPLANTABLE CATHETER-PORT SYSTEM IN CHILDREN

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Shaeed Beheshti University of Medical Sciences

DISCRIPTION: polysite implantable access ports allow repeated and prolonged access to the vascular system (intra venous or intra_arterial) peritoneal cavity or intra vertebral space (intra thecal and peridural space)
POLYSIT venous access ports are used to administer chemotherapy, antibiotics, antiviral drugs and hemophilia patients. They can be used for parenteral nutrition, collection of blood samples and transfusion of blood
POSSIBLE COMPLICATION:
Embolism, pneumothorax, hemothorax, hydrothorax
-Infection -artriovenous fistula
-occlusion of the catheter
-Inflammation/skin necrosis over the implantation site.
INSERTION:
-only qualified doctors can insert and implantable access port.
-before insertion, purge the device of any air and fill with N/S or heparinised solution.
-take care during insertion of the device to avoid any mechanical alteration of the catheter.
-strictly follow the connection technique described under “*INSERTION TECHNIQUES” *to avoid damaging the catheter and to ensure correct connection.
Use and maintenance:
-before use, inspect and perform asepsis of the injection site.
-locate the needle perpendicularly to the skin and advance as far as the base of the port.
-check the patency of the device and correct positioning of the needle by obtaining blood reflux (polysite venous access ports only) and by injection of N/S without excessive pressure or local extravasations.

ژرم سل تومور: تجربهء ۱۰ ساله

دکتر احمد خالق نژاد طبری– دکتر علیرضا میرشمیرانی – دکتر محسن روزرخ – دکتر لیلی مهاجرزاده– دکتـر شـقایق حساس یگانه – دکتر نسیبه خالق نژاد طبری – دکتر علی علی عسگری

Pediatric Germ Cell Tumor: A retrospective study in Mofid hospital

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Introduction: Pediatric germ cell tumors (GCTs) are a heterogeneous group of rare neoplasms . They occur at a rate of 2.4 cases per million children and account for approximately 1% of cancers diagnosed in children younger than 15 years. These neoplasms occur in both gonadal and extragonadal sites, with extragonadal and testicular tumors predominating in children younger than 3 years and gonadal tumors predominating during and after puberty.
The aim of this study was identify of treatment outcome in pediatric patients with germ cell tumor in mofid hospital.
Materials and methods: this retrospective study we studied units of hospitalized germ cell tumor patients in mofid hospital from 1378 until 1388. And we investigated treatment outcome in clinic or by calling them.
Results: 44 patients were investigated. 32(72.7%) patients were female and 12(27.3%) patients were male. Mean age of children was 23 month.(ranged in age from 1 months to 144 month). Pathology report of tumors included : Mature teratoma 18(6.8%) cases,yolk sac tumor 14(31.8%),immature teratoma 6(13.6%) cases , mixed germ cell tumors 3(6.8%) cases, malignant teratoma in 1(2.3%) case ,dysgerminoma 1(2.3%) case,choriocarcinoma 1(2.3%) case . The most common sites of tumor were sacrococcygeal tumor in 20(45.5%) cases, ovary 9(20.5%) cases and retroperitoneum in 5(11.4%) cases. Staging of disease at diagnosis: 23(52.3%) patients in stage I, 3(6.8%) patients in stage II, 2(4.5%) cases in stage III, 11(25%) in stage IV. In 5(11.4%) patients staging was undetermined. Surgical management included: complete surgical resection in 41(93.2%) cases, partially resection in 1(2.3%) case and open biopsy in 2(4.5%) patients. 15(34.1%) patients underwent chemotherapy. Condition of 4(9.1%) patients were undetermined. but treatment overview in another patients included: complete remission in 26(59.1%) cases, locally recurrence in 3(6.8%) patients, distant recurrence in 2(4.5%) and 9(20.5%0 patients expired. Mean survey was 14 month and upper limit of follow up was 102 months.
Conclusion: Advances in surgical treatment, together with the use of multiagent chemotherapy regimens, have resulted in a dramatic improvement in the outcome for children with malignant germ cell tumor. International studies using integrated multi modal treatment strategies are reporting impressive survival rates, ranging from 75% to 90%. The number of relapsed patients is, ranging from 20% to 30%. In our study; survival rate was 70.3% and number of recurrence was 5(11.3%).

جراحی لنفوم بورکیت داخل شکمی در کودکان: تجربه ۱۰ ساله

دکتر جواد غروبی، دکتر علی رضا میرشمیرانی، دکتر بی بی شهین شمسیان، دکتر جعفر کورانلو

Surgery of Intra-abdominal Burkitt's lymphoma in children: A 10 year experience

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Abstract

Aim of study: The role of surgery in the management of Burkitt's lymphoma within the last 20 years has been changed substantially. Along with the assignment of adjusted therapy and specific treatment protocols, surgical procedures have been restricted to defined situation including abdominal emergencies, diagnostic biopsy, and total tumor excision and second- look operations, then operation in intra abdominal Burkitt's lymphoma remains controversial and different opinion are present in the literature.

Materials and Methods: In a descriptive retrospective study, 34 children with intra-abdominal Burkitt's lymphoma have been operated and followed in Mofid children's Hospital from 1995 to 2004. All patients underwent laparotomy and the surgical procedure was classified as total excision, debulking, or incisional biopsy. Collected data from the patient's medical records were analyzed and variables such as age, gender, diagnostic tools, tumor localization, surgical and medical treatment and outcome were evaluated.

Results: The patients consisted of 26 (76%) boys, and 8 (24%) girls with the ages ranged form 2 to 14 years. Of 34 patients 9 (26.47%) underwent emergency surgery because of acute abdomen (intestinal obstruction 6, intussusceptions 4, appendicitis 1), and 25 (73.52%) patients were referred to our clinic because of abdominal mass, pain, anorexia, and weight loss. 11(32.35%) patients had localized tumor which total resection was performed, and in 23 (67.64%) children extensive intra-abdominal tumor were detected. 5 of 23 patients underwent debulking procedure and in 18 cases only biopsy were carried out. Second-look operation was performed in 9 patients of our biopsy group. All patient received chemotherapy after operative recovery. The survival rates at 3 to 10 years were: 100% in total resection group, 78% in biopsy group and 40% in the debulking group. There were 7(20.58%) mortality in our study.

Conclusion: In patients with proven localized disease, total resection should be attempted, if not partial resections seems feasible, surgical intervention should be restricted to the least necessary procedures for life-threading organs, it is desirable to limit the surgery to tissue biopsy only.

10 years evaluation of ovarian teratoma (case report)

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Introduction:

Ovarian teratoma is one of the most tumors in childhood.It composes almost 50% of pediatrics tumors.The most presentations are abdominal pain and appears between 6 and 15 years.Its presentation is seldom in below than 3 years of age.5 to 10% of presentations are bilateral.Ultrasonography showes solid and cystic masses.

Cases report:

We evaluated 21 girls whom referred to DR Sheikh hospital (mashhad)between 1999-2009 with ovarian tumor and went under surgery.Age of all of them was below than 14.

In 13 cases report of pathology exam showed teratoma.Their age at the operation was 1.5-14 years.8 cases had mass in right side and others had it in left side.Ovophorectomy had been done for 7 cases and just only resection of tumor had been done in 6 cases and ovarian tissue preserved in them.Sonography showed solid-cystic ovarian mass in all of them .Plane abdominal X ray showed calsification in 9 patients.Follow up did for 11 patients and 2 cases were not available.Sonography showed normal ovary at the same or other side and tumoral mass had not seen.2 cases married and one of them has a child now.

Conclusion:

We didn’t find any synchronous or metachronous tumors and no differences between patients who had ovophorectomy and preserved normal ovary.

دکتر فریبا جهانگیری، دکتر جواد نصیری، دکتر خدیجهء ارجمندی، دکتر شهلا انصاری، دکتر غلامرضا باهوش

Demographic data of childern with Germ cell tumors (GCTs) in Ali Asghar hospital

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Background & ame: (GCTs) are a heterogeneous group of rare neoplasms and account for approximately 1% of cancers diagnosed in children . These tumors occur in gonadal & extragonadal sits. (GCTs) exhibit a broad spectrum of clinical presentation & histopathologic features & carry varying risks for malignancy depending on the type of lesion . Optimal outcomes are echived with complete surgical resection accurate histologic examination & selective use of chemotherapy.

Method : Between 1999 & 2009 the record file of all patients with diagnosis of (GCTs) admited in hematology – oncology & surgery ward in Ali Asghar hospital were evaluated .The demographic data ,clinical sign at presantation & the last time of follow up recorded .The data are presented as frequency .

Results: there were 18 cases of (GCTs). 11(61%) was femal & 7(39%) was male, 5 patient died, 7 patients are still alive, 6 (33%) pathents was lost to follow up. Presenting complain was as follow : urinary retention in 4 (22%) , Abdominal mass in 2(11%) , Mass in buttock in 3(17%) , Testicular mass in 3(17%) , Abdominal pain in 2(11%). Agitation in 1 (5/6%), Abdominal mass combined with Abdominal pain in 3(17%). In pathology: 10 cases were yolk sac tumor 56%, 4 (22%) was mature teratoma , 2 were mixed : one case yolk sac tumor with teratoma & another case chorio carsinoma with yolk sac (11%) One case was dysgerminoma (5/6%). Location of tumors :ovary in 8 case (44%) , Testis in 3 case (17%) , Retroperitoneum in 2 case (11%) , Sacrococcygeal in 2 case (11%) , Abdominal and sacrococcygeal in 2 case 11% & Thorasic in 1 case (5/6%) .

Conclusion: ovary was the most common site of tumor. Urinary retention & abdominal mass with pain was the most common sign & yolksac was the most common pathology there was no relation between sex & onset of symptoms with survival. Because we have no close fallow up in some patients in our series we cannot determine the survival.

Occurrence of teratomas in infants and children

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Introduction / Purpose:
Teratomas are embryonic neoplasms that originate from Totipotential cells. These lesions can be monodermal or composed of cells from each of the three germ layers (ectoderm, endoderm and mesoderm). They present in different locations of the body from brain to sacrococcygeal area. The aim of this study is to assess the incidence of different types of teratomas in two major pediatric surgery centers affiliated to Tehran University of Medical Sciences and comparing it with literatures.

Methods:
This retrospective study was conducted on all the patients with teratoma who underwent surgical treatment in two major hospitals from 1982 to Feb 2010, considering location and histology of the tumor, age and gender of the patients.

Results:
This study was performed on 116 cases in these two hospitals from 1982 to Feb 2010. 85 of 116 cases had sacrococcygeal teratoma while 11 suffered from teratoma of the ovaries, five teratoma were found in peritoneum, two in oropharynx, six in testis, four in vagina ,one in neck area and two cases of fetus in fetu . From these 71 were neonate and 45 were either infant or more than two years old. Three patients died before operation and three of 113 cases who received surgical treatment died because of the complications afterwards. 83% of teratoma was diagnosed as benign tumors and 17% were malignant.

Conclusion:
Most of the patients in this study were female. A high percentage of the tumors were benign and mature and the most common location was sacrococcygeal area. There were small differences between our findings and the Literature.

Toxocariasis

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Toxocariasis or larva migrants caused by toxocara cani (dog) or toxocara cati (cat) which they lives in dogs or cats but eggs can be eaten by contaminated soil.
Larval stage of parasite can infect various organ of intermediate host like human.
Liver, lung, eye…..can be involved by larva.
Symptoms and signs depend on involved organ, immune state and severity of infection.
Three cases in our center (Imam Khomeiny Hospital of Ahvaz) are very unusual cases.
The first case was an acute abdomen due to idiopathic gangrenous perforated ileum, after resection and anastomosis in 2nd post operation day recurred peritonitis due to gangrene of other part of intestine and 3rd and 4th operation due to gangrene of some other part of small intestine which leads to death of patient.
Pathologic report was toxocara.
The second case was an abdominal mass (retroperitoneal) resection showed toxocariasis.
The third case is an eleven years old boy with fever for one month and weight loss and abdominal mass and eosinophylia.
Ct scan showed a left colon mass and partial obstruction, mass completely resected and colostomy and mucus fistula done.

گزارش یک مورد تومور ویلمز با تهاجم به دهلیز قلبی

دکتر منصور ملاییان، دکتر محمدرضا میرزاآقاییان، دکتر محمدعلی احسانی، دکتر سروش غفوریان

Wilms Tumor with Vascular Extension Successful Treatment of intracaval and atrial extension A case report

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Progress in the treatment of Wilms tumor is one of the most significant achievements in the field of pediatric oncology. Current combination multimodal therapy has produced a 2-year disease-free survival rate approaching 86%.[1]. Renal vein involvement (extra-renal) is found in 11.3% of patients and further spread into IVC and/or atrium occurs in an additional 4%.[1,2].The surgical management of WT with vascular extension is associated with considerable morbidity [2,3].Pre-operative chemotherapy seems to be gaining favor as the first line therapy in the management of WT[2,4-7].there are several reports of chemotherapy being used to manage intravascular tumors, especially in Europe and in some North America centers, following the SIOP protocol[1,2,5,8]. Those centers following the NWTS guidelines still prefer surgery before chemotherapy [9]. The current report describes our experience with a patient who presented with WT and involvement of IVC and right atrium.

Case report

A 4-year-old girl presented with a history of abdominal distension and weight loss of three months duration. On physical examination there was a large mass filling the right upper and lower of the abdomen. The reminder of the examination was unremarkable except for a blood pressure of 130/90 mmHg and a meso-cardiac systolic ejection. Liver function tests was normal .The IV pyelogram showed distortion of right kidney .Abdominal US revealed a homogeneous solid mass arising from the upper portion of right kidney and crossing the midline and extending to the left kidney .A thrombus was seen in the retro-hepatic portion of IVC ,Extending into the right atrium. There was as cites in the abdomen. Thoracic CT was normal. Echocardiography revealed decreased ejection fraction of about 36% and decreased function of the left ventricle. There was a solid mass in the right atrium. Open biopsy was performed by small incision, and preoperative chemotherapy was administered according the SIOP protocol. The tumor was reduced in size extensively, but the tumor thrombus remained unchanged. The tumor was nonpalpable at the time of operation .The patient admitted to the hospital due to cardiac arrhythmia and treated medically at the period of chemotherapy. Laparatomy revealed tumor originated from upper pole and renal and IVC were filled with tumor thrombus up to right atrium. Right total nephrectomy was performed, and tumor thrombus from the IVC at the level of right renal vein up to the right atrium, were removed by use of extracorporeal circulation , cardiac arrest and profound hypothermia as one piece .The thrombus had greatest diameter at atrial end about 5 cm and tapered gradually down to the level of right renal vein. The thrombus was free-floating in type and only had minimal adhesion to the atrial wall. Recovery was uneventful .Chemotherapy was completed. The patient was free of disease during one year postoperatively.

Discussion

Renal vein involvement (extra-renal) is found in 11, 3% of patients and further spread into the IVC and/or atrium occurs in additional 4% [1]. The involvement of the IVC in WT cases is usually an intra-luminal extension of the tumor mass rather than an invasion by the malignant tumor, although this situation can occur in rare instances. This intra-luminal extension may be excised by cavotomy when blood flow is present. Depending on the level of thrombus, cardiac arrest, profound hypothermia and cardiac bypass may be necessary. There are rare instances in which the tumor thrombus invades the vena cava wall, so that thrombectomy is not feasible. In this situation cavectomy is a good surgical strategy. Once again, depending on the level of thrombus, cardiac bypass ,as described above ,might be necessary. Although intravascular and intra-atrial extensions are rare, we believe that every patient with a renal tumor should undergo preoperative Doppler examination of the IVC because the surgical procedure is completely different, when such extension exists. There are signs and symptoms that suggest intravascular extension, such as varicocele, hepatomegaly and collateral circulation. Albuminuria, hematuia ,or presence of hypertension, penile, genital or leg edema, should alert the physician of an underlying caval obstruction. Ritchey et al, in a retrospective review of 77 patients with intravascular extension, observed only 5 with clinical findings suggesting this condition [1]. Imaging studies (tomography, ultrasound and Doppler) can demonstrate the thrombus. Preoperative chemotherapy can result in total thrombus resolution but not in all patients [2-4]. The extent to which the vena cava is compromised will determine the surgical approach. For infrahepatic thrombus, with or without invasion of vessel wall, a transperitoneal approach is indicated. In cases with intra-atrial extension extracorporeal circulation ,cardiac arrest and profound hypothermia are required ,regardless of whether blood flow is present[1-3,10].Some emphasis on vena caval extension with blood flow around the thrombus, because surgical procedure is different[10]. It is possible that in some cases of WT with intravascular extension, the thrombus has no tumor cells after preoperative chemotherapy. However, according to

the basic principles of surgical treatment of WT, complete resection of all macroscopic disease must be attempted. Akyuz et al observed a high number of recurrences in patients who had residual tumor thrombus [11].Thus cavectomy is indicated when there is no blood flow in the vena cava, and extracorporeal circulation will become necessary depending on the level of the thrombus.

It is known that the presence of thrombus does not modify the prognosis, which will depend only on tumor pathology and stage [2]. Thus, the inherent risks of cavectomy, with or without cardiac bypass, are justified, because tumor resection is essence for treatment. In the case of caval involvement renal failure have been reported ofter unilateral nephrectomy[12].The pathophysiology of caval involvement and the likely cause of renal failure in this situation is briefly discussed. The collateral venous drainage of the left kidney is better than right kidney and the left kidney is better protected against venous hypertension in the event of caval involvement by the tumor[12].A sustained increase of venous pressure of the IVC can cause atrophy of the proximal convoluted tubules, glomerulus and distal tubules and result in renal failure[12]. The collateral veins draining the kidney in case of IVC obstruction are via the intercostals veins, vertebral veins, hemorrhoidal veins and superficial epigastric vessels. The (L) renal vein has a better collateral venous drainage and receives the ascending lumbar vein which connects it with hemiazygos and the azygos system of veins. Others, viz., the adrenal, inferior phrenic gonadal, ureteric also contribute to the collateral circulation.The collateral venous drainage of the right kidney is limited to capsular ,adrenal and ureteric veins [12]. Preoperative accurate assessment of caval invasion is a must before surgery.[13,14,7] In NWTS where prenephrectomy chemotherapy is not employed. It will be necessary to accurately stage the disease and remove the entire involved segment [13] .The knowledge of the venous anatomy and patho physiology of renal shutdown is a must before surgery. In SIOP Group where presurgical chemotherapy is used, a longer course of chemotherapy can clear the IVC of the tumor before surgery.[14,7] Caval involvement of WT must be adequately treated with chemotherapy and the integrity of the renal reserve assessed intra-operatively to prevent renal failure in the postoperative period .When the left kidney is affected by the tumor with caval thrombusis it may be prudent not to be disturb the capsule of right kidney. The preoperative assessment included abdominal and chest computed tomography (CT) scan with 3-dimensional reconstruction,and vena cava Doppler in order to define the tumor and the cranial thrombus extension.Patiens presenting more complex cases (atrium involvement) are evaluated with magnetic resonance (MR). The cranial limit of thrombus are classified according to the Neves and Zincke system classified according to cranial extension as level I-renal(thrombus extending less than 5 cm above the ostium of the renal vein); level II, infrahepatic (below the intrahepatic vena cava); level III ,retrohepatic (thrombus involving the intrahepatic portion of IVC, but below the diaphragm); and level IV,atrial (above the diaphragm, could reach the right atrium).[15]Surgical exposure is achieved by chevron bilateral subcostal laparatomy and,When necessary ,is extended to the xiphoid process for a maximum exposure of suprahepatic vena cava(Mercedes incision) ,which ensures greater exposition. After incision, the cavity and organs are examined.Initially, the renal hilum is approached ; the renal artery is tied and sectioned, maintaining the renal vein intact.Then,the IVC is dissected verifying the thrombus extension.[15]. For thrombus level I, no additional maneuver is necessary. The vascular Satinsky clamp is positioned partially closing the IVC, pumping the thrombus toward the kidney and the renal vein is sectioned with a safety edge. The IVC is sutured by running suture with polypropylene. In thrombus level II , the infra renal vena cava as well the infra diaphragmatic vena cava , the hepatic hilum and the opposite renal vein are clamped with vascular clamps[15].

This permitted collateral circulation control,leaving lumbar veins without control only.A self-retaining retractor is necessary to an adequate exposure.

After controlling collateral circulation, a short longitudinal anterolateral cavotomy is performed and thrombus is removed completely.Then the renal vein is sectioned and IVC is sutured in running fashion with polypropylene.

For thrombus level III, the liver is mobilized completely by dividing the falciform,triangular and coronary ligaments. The collateral circulation is controlled by vascular clamps as in thrombus level II. The femoral artery and right internal jugular vein is dissected to perform cardiopulmonary bypass(CPB). The CPB are followed by a deep hypothermic circulatory arrest(DHCA). Then, cavotomy is performed to remove the thrombus. Sternotomy is not necessary in these cases. To ensure for all thrombi removal, a fogarty catheter is used.Afterwards, the renal nein is sectioned and the vena cava is repaired. The CPB is restarted and blood is progressively heated until the patient become normothermic.

The procedure is similar for cases with thrombus level IV (right atrium); however, a sternotomy is necessary to perform conventional CPB. The aorta and SVC is canulated for CPB with DHCA. The cardio-thorasic team performs right atriutomy to remove thrombus. A cavotomy is performed to remove the thrombus simultaneously. After repair of IVC and right atrium, CPB is started again, and the patient is rewarmed until cardiac recovery is achieved [15]

Retroperitoneal Teratoma

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Retroperitoneal Teratoma occur outside the pelvis,often in a suprarenal location. They represent about 5% of all childhood Teratoma & 75% occur in children younger than 5 years.
It can compress the gastrointestinal tract. Abdominal X-Ray may show calcification or bony structures within the tumor.
US & CT & serum markers are the investigations used.malignancy is uncommon.
I will discuss this type of teratoma & presentation a six month female with immature Teratoma,operated in Atieh hospital with para clinical investigation & approach to diagnosis & laparatomy findings.

A very rare tumor of testis presented as an acute scrotum

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Abstract:
Juvenile Granulosa Cell Tumor (JGCT) is a rarely diagnosed subset of benign sex cord-stromal testis tumors. Although it accounts for only 1.2% of all prepubertal testis tumors
Case: A case of 7-month-old male infant referred to the pediatric surgery clinic with an acute right scrotum. In clinical evaluation it was suspicious to a testicular mass. After frozen section and permanent pathology the diagnosis was a very rare tumor of testis, Juvenile Granulosa Cell Tumor.

نقش جراحی در سرطان های کودکان

دکتر محمدتقی ارزانیان (سخنران مدعو)

The role of surgical treatment in pediatric oncology

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Pediatric surgical oncology is one of the oldest form of cancer treatment and also rapidly developing field. Over the past 50 years, great progress has been made in diagnosis, treatment, and survival of children with cancer. In the 1960s, the survival rate for children with malignant solid tumors was below 25%, whereas it has exceeded 75% at present. This dramatic change is attributable to the creation of multidisciplinary therapy, the improvement of surgical techniques, the successful use of chemotherapy, the application of radiation therapy and the progress in understanding of tumor molecular biology.

Surgery plays a key role in the process of diagnosing cancer and finding out how far it has spread. This process is called *staging*. Advances in surgical techniques have allowed surgeons to operate on a growing number of patients and have good outcomes. Today, operations that involve less cutting and damage to nearby organs and tissues (less invasive surgery) often can be done to remove tumors while saving as much normal tissue and function as possible. Surgery offers the greatest chance for cure for many types of cancer, especially those that have not spread to other parts of the body. Most people with cancer will have some type of surgery. Surgery can be done for many reasons. Some types of surgery are very minor, while others are much bigger operations.

The more common types of cancer surgeries are as belows:

1-Preventive (prophylactic) surgery: Preventive surgery is done to remove body tissue that is likely to become cancer (malignant), even though there are no signs of cancer at the time of the surgery. For example, pre-cancerous polyps may be removed from the colon.

2-Diagnostic surgery: This type of surgery is used to get a tissue sample to tell if cancer is present or to tell what type of cancer it is.

3- Staging surgery: staging surgery is done to find out how much cancer there is and how far it has spread

4-**Curative surgery**: Curative surgery is done when a tumor is found in only one area, and it is likely that all of the tumor can be removed. Curative surgery can be the main treatment for the cancer. It may be used alone or along with chemotherapy or radiation therapy, which can be given before or after the operation

5- Debulking (cytoreductive) surgery: Debulking surgery is done to remove some, but not all, of the tumor. It is done when removing all of the tumor would cause too much damage to an organ or near-by tissues

6-Palliative surgery: This type of surgery is used to treat problems caused by advanced cancer. It is not done to cure the cancer. Palliative surgery can also be used to correct a problem that is causing discomfort or disability.

7- Supportive surgery: Supportive surgery is done to help with other types of treatment. For example, a vascular access device such as a port-a-cath can be surgically placed into a large vein. The port can then be used to give treatments or draw blood for testing, instead of putting needles in the arms.

8- Restorative (reconstructive) surgery: This type of surgery is used to improve the way a person looks after major cancer surgery, or to restore the function of an organ or body part after surgery.

Surgery to diagnose and stage cancer:

***Biopsy* , Fine needle aspiration biopsy, Core needle biopsy, Excisional or incisional biopsy, Open surgical exploration (laparotomy, thoracotomy, or mediastinotomy).**

Laparoscopic surgery: The role of laparoscopic surgery in cancer treatment is not yet clear. Doctors are now studying whether it is safe and effective to use laparoscopic surgeries for many cancers of the bladder, colon, prostate, and kidney, among others. It may prove to be as safe and work as well as standard surgery while cutting less and causing less damage to healthy tissues (being less invasive). Some studies have hinted at this being the case. But larger, long-term studies still need to be done.

Thorascopic surgery :A thoracoscope is a narrow, rigid tube with a camera connected at one end that can be put through a small cut (incision) into the chest after the lung is collapsed. This allows the doctor to see inside the chest. Tissue samples of any areas of concern on the lining of the chest wall can be taken out (biopsied), fluid can be drained, and small tumors on the surface of the lung can be removed with small stapling devices. This approach leads to less cutting and has even been used to remove parts (lobes) of the lung that contain cancer. Studies have shown that for early stage lung cancer, results using this approach are much the same as removing part of the lung through a cut (incision) in the side of the chest (this surgery is called a thoracotomy)

Conclusion: The pediatric surgical oncologist must have a good collaboration with all those involved in the care of the patient and participate in multidisciplinary board discussions to determine the optimal surgical care for the patient. In addition, the qualified pediatric surgical oncologist should be familiar with the surgical principles for the peculiarities of tumors and current treatment protocols. Surgical specialists with pediatric expertise (i.e. training and certification, if available) in neurosurgery, urology, orthopedics, ophthalmology, otolaryngology, dentistry, and gynecology a board-certified radiation oncologist trained are needed in this field. The improvement of pediatric tumor surgery in further exploration and the improvement of surgical skills will improve the survival and long-term life quality of the patients.