

Hematology & Oncology Abstracts

Oral Presentation

Evaluating the Causes of Splenectomy in Patients in Mofid Children Hospital During 6 years

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Objective: Spleen produces specific antibodies and filters out encapsulated organisms and therefore, is important in protecting the body against infection. So we have carried out this study to analyze the causes of splenectomy, therapeutic value of splenectomy, adherence to preventive strategies and prevalence of sepsis and mortality.

Methods and Subjects: In a retrospective study, we reviewed the medical record of children who had splenectomy in the period of 2001 until 2006 in Mofid children hospital. In our study, followed up of 38 patients among 53 patients was possible. Paired T-test were used for data analysis.

Findings: There were 53 patients who underwent splenectomy (32 males and 21 females). The mean age was 9.98 (1.5-23) years old. The indications of splenectomy in these patients include: B-major thalassemia in 43.4%, hereditary spherocytosis in 15.1%, chronic idiopathic thrombocytopenic purpura (ITP) in 11.3%, B- intermediate thalassmia and portal hypertention in 5.7%, splenic cyst, lipid storage disease Gaucheras, and autoimmune lymphoproliferative syndrome (ALPS) each one in 3.8%, rupture of spleen, subcapsular hematoma of spleen, hemophagocytosis-acute lymphoblastic leukemia (T cell- form), and autoimmune hemolytic anemia each one in 1.9%. 100%, 41.5%, 36% of patients had been vaccinated against *S. pneumoniae*, *Neisseria meningitidis* and *Hemophylus Influenza* type B, respectively. The indications of splenectomy in these two groups were hypersplenism and increasing transfusion requirement. There was a significant difference between the mean of preop plt and the mean of postop plt in chronic ITP ($P<0.05$) that shows complete response to splenectomy. There was a significant difference between the mean of preop Hb and the mean of postop Hb in hereditary spherocytosis patients ($P<0.05$) that shows complete response to splenectomy in these patients. In patients with complete follow up, we had 4 deaths, that 2 of them were patients with major

thalassemia because of sepsis and congestion heart failure (50%), 1 of them was a patient with hemophagocytic syndrome-ALL (T- cell) because of sepsis and disseminated intravascular coagulation (25%) and 1 of them was a patient with ALPS because of sepsis (25%).

Conclusion: Splenectomy is one of the cornerstones in the treatment of pediatric hematological diseases. The results obtained showed that splenectomy, which is safe and efficient for the disease and also causes to improvement of life quality.

Key Words: Children; Splenectomy; Thalassemia; Immune thrombocytopenia purpura; Hereditary Spherocytosis

Poster Presentation

Pediatrics Lymphohematopoietic Cancers and Its Relation to Preconception, Pre- and Postnatal Active and Passive Cigarette Smoking of Parents

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Objectives: We have a little evidence about smoking and its relation to childhood cancers; some studies have reported associations between maternal or paternal smoking and a variety of childhood cancers, while others have reported no association. The objective of the present study is to assess the potential association between prenatal exposure to passive smoking as well as maternal active smoking and postnatal exposure to environmental tobacco smoke (ETS) and enhanced incidence of childhood lymphohematopoietic cancers.

Methods and Subjects: In a case-control study, 106 lymphohematopoietic cancer patients (pediatrics age group), and 110 healthy children (controls) were compared for elements like: proportion of current smoking and passive and ex-smoking of father, mother and other close family members of two groups before and during pregnancy and after birth of the child. Controls were matched with cases by age and sex. Cases were sampled by convenience method from pediatric hematology clinic and from the list of province cancer registry; the controls were sampled randomly from urban primary health care centers in Rasht city. The data were gathered by direct interview with parents of children and by usage of a structured questionnaire. The collected data were entered into Stata 10 Software and analyzed by negative binomial and Poisson regression models.

Findings: The most common (65.1%) lymphohematopoietic cancer was ALL (Acute lymphoblastic leukemia). Mean age of children in case and control groups was 8.9±3.7 and 7.6±3.6, respectively. The overall odds of Current Smoking was significantly higher in patients than normal children 1.72 (CI95% 1.24, 3.01) (Negative binomial test, P=0.039). In separated analysis for paternal smoking the odds was significantly higher in patients than normal group, 1.8 (CI95% 1.02, 3.15) (Negative Binomial test, P=0.028) and there is no relation between maternal passive smoking during pregnancy and childhood cancers (Poisson regression model). There is more powerful relation between preconceptional paternal smoking and childhood cancer (negative binomial test, P=0.001). Females are more sensitive to passive and preconceptional smoking effect than males (Poisson regression, P=0.018).

Conclusion: The cigarette smoking specially in fathers have a significant effect on children cancers; and also in spite of regular attitude the preconceptional effect is more powerful than passive smoking of mothers, so we need new preventive programs to limit the cigarette smoking of young people for prevention of childhood hematopoietic cancers.

Key Words: Smoking; Lymphohematopoietic cancer; Preconception

Poster Presentation

Combined deficiency of factor V and factor VIII (F5 F8 D) in Gorgan

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Background: Congenital F5 F8 D is estimated to be extremely rare (1:1000,000) in the general population. More than 50 pedigrees with an autosomal recessive bleeding disorder characterized by a combined deficiency of both Factor V and Factor VIII have been reported. This disease affecting males and females in equal numbers. This disorder was reported to be particularly prevalent among middle Eastern Jewish and non-Jewish Iranians, where the incidence was estimated 1 / 100000. In addition, reports show that F5 F8 D is usually associated with fewer symptoms than hemophilia A because the concomitant presence of two coagulation defects does not enhance the hemorrhagic tendency that was observed in each defect separately. Therefore, I encouraged to work on these reports for my patients.

Case Presentation: All the information have been derived from patients files. I have 105 patients of hemophilia and rare bleeding disorders in my center. 4 patients have F5 F8 D. I have 4 patients with F5F8D from 105 patients in our center. (2 males and 2 females). In one female factor V = 25 % and factor VIII = 15 %. In another female Factor V = 12 % and factor VIII = 10 %. In one male factor V = 9.2 % and Factor VIII = 15 %. In another male Factor V = 8 % and Factor VIII = 9.5 %. None of them had bleeding episodes in 3 years ago.

Conclusions: Symptoms of this disorder was reported to be easy bruising. Epistaxes and gum bleeding are not uncommon in affected individuals. Severe symptoms including hemarthrosis is rare. F5 F8 D bleeding episodes are usually treated on demand and do not require prophylaxis. This finding is our results.

Key Words: Factor V; Factor VIII, Rare bleeding disorders

Oral Presentation

To Screen or not to Screen Glucose-6-Phosphate- Dehydrogenase Enzyme

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Objective: Massive newborn screening is important issue to prevent neonatal death, mental retardation and other irreversible clinical manifestations. The purpose of this study was determination cost effective measurement of glucose-6-phosphate dehydrogenase (G-6-PD) enzyme in newborns and its deficiency prevalence rate.

Methods and Subjects: All full term newborns who were born, neonates with icterus and children with acute hemolysis who were admitted, were evaluated for deficiency prevalence and the cost of G-6-PD test. The qualitative color reduction test was performed on healthy newborns and quantitative test was done for newborns with icterus and children with acute hemolysis.

Findings: Three (2%) of 146 screened newborns were G-6-PD deficient and the cost of three screening tests were \$18 and \$856 for remainder of newborns; 31 (11.4%) of 272 newborns readmitted with icterus were G-6-PD deficient and the cost of 31 tests were \$186 and \$1446 for remainder of newborns. The only significant difference between G-6-PD deficient newborns and normal G-6-PD was bilirubin level. Eleven (0.2%) of 5054 hospitalized children were G-6-PD deficient and cost of two night staying for eleven patients were \$1540

and \$706020 for the remainder. The prevalence was estimated to be around 4.5%.

Conclusion: Glucose -6- phosphate dehydrogenase screening in newborns is not cost effective and for prevention of hemolysis during the next years all newborns with icterus who are admitted should be evaluated for G-6-PD enzyme.

Key Words: Screening; Newborns; G-6-PD

Oral Presentation

Immune Thrombocytopenic Purpura (ITP) is Benign Disease in Children

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Objective: Immune thrombocytopenic purpura (ITP) is an autoimmune disease that is characterized by skin-mucous bleeding and decrease in the platelet (less than 150,000). There is a history of viral disease or vaccination before the disease. The aim of this study was evaluation the clinical features, response to treatment and treatment outcomes of children (0-14 years old) with ITP.

Methods and Subjects: In a descriptive (cross-sectional) study, some variable such as age, sex, clinical features, treatment outcome, and the rate of chronic ITP of 66 patients were evaluated from 2002 to May 2008 in the Shahid Sadooghi Hospital of Yazd. The data were analyzed using SPSS statistic software. Chi square test was used for data analysis of the effect of age and sex on this disease.

Findings: Among 66 patients, 30 female and 36 male subjects were from 35 days to 12 years old of age that were divided two age groups: under 7 years old and above 7 years old. 55 patients (83.3%) had purpura and 27 patients (40.9%) had active mucosal bleeding most commonly epistaxis. 31 patients (47%) had a history of common cold, 17 patients (25.8%) a history of vaccination and 2 patients (3%) a history of chickenpox. 66 patients received IVIG with or without corticosteroid, 52 patients had complete response to initial treatment. 50 patients had acute ITP (75.75%) and 42 patients (80.76%) were under 7 years old and 8 patients (57.14%) were above years old (27 were males and 23 were females). 16 had chronic ITP (24.24%) and 10 patients (19.23%) were under 7 years old and 6 patients (42.85%) were above 7 years old (9 were males and 7 were females). Chronic group were treated with IVIG & corticosteroid (7 patients), Anti D (4), Azathioprine and Cyclosporine (2) and Anti D, Azathioprine & cyclosporine (3). Finally Splenectomy was performed in 9 patients. Age had a relationship with chronicity, but sex did not.

Conclusion: ITP in children is a benign disease and responds well to treatment. Vaccination and viral illness play an important role in the etiology of ITP in children.

Key Words: Immune thrombocytopenic purpura; thrombocytopenia; Children; ITP

Poster Presentation

Investigation of Risk Factors of Childhood Leukemia in Fars Province

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Objective: Cancer is the most important cause of death due to disease in industrial countries and leukemia is the most common type of childhood cancer accounting for 30% of all cancer diagnosed in children younger than 15 years. Leukemias divided in to several groups and the most important kinds of them are ALL and AML. Childhood leukemia may originate from a combination of genetic susceptibility factors and environmental exposures. Increasing of childhood incidence in recent years can be the result of increased risk factors. Some known causes of childhood leukemia include incorrect diet, exposure to electromagnetic fields and ionizing radiations, parent's smoking habit and some occupations of them.

Methods and Subjects: To investigate risk factors of childhood leukemia in Fars province we have performed a descriptive study with face to face interviewing with 150 parents of children affected by leukemia in oncology department of Shahid Fagehi hospital and Motahhary clinic. The data was analyzed with SPSS 15.3.

Findings: Most of them were male (62.7%), urban, (61.3%) and were born in winter. 75.3% of them had experience of exposure to diagnostic X-ray at least one time. 49.7% of them (themselves or their parents) have been exposed to chemical materials such as agricultural poisons, lubricant or petrol. 34% of them were exposed to animals. About half of them were affected by viral disorders. 41.3% of the patient's parents had smoking habits and 29.3% of them have been in war between Iran and Iraq and 8% of them were affected by dangerous gases in the war. Family history of cancer was found in 37.3% of the patients. 17.2% and 32.2% of them were exposed to high voltage electricity and different dangerous smokes, respectively. Most of them weight under 3.5 kg and got breast feeding. The mother of 20.7% of them had taken drug during pregnancy.

Conclusion: Most patients are males which is in accordance to the previous findings; being male is a risk factor for leukemia and most of them had experience of exposure to diagnostic X-ray during the postnatal period that shows the importance of it as a risk factor for childhood leukemia. A case-control study on

healthy and affected children to leukemia can get more information about etiology of this cancer.

Key Words: Leukemia; Childhood cancer; Risk Factor; Exposure

Oral Presentation

Should we treat chronic ITP?

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About 20% of patients in children group with immune thrombocytopenic purpura lasted to more than 6-12 months with or without treatment. The platelet count is usually lower than normal (<50000/ μ L). The policy of treatment in acute or chronic ITP sometimes is obscure. The severity of a patient's bleeding tendency can best be evaluated by clinical observation. Given the unreliability of the platelet count, it does not make sense to base treatment on it, unless we plan to treat the platelet less than 1%. But the side effect of treatment and psychiatric stress is more harmful. In a review persisting for treatment without apparent bleeding symptoms and sings with IVIG or Corticosteroids, is unfair. But on the other hand "Watching & Waiting" in most cases, fallow with spontaneous cure. Splenectomy is delayed and should be done on special cases.

Key Words: Chronic ITP; Treatment; IVIG

Oral Presentation

Correlation between CBC Indexes and Type of the Gene Mutations in Alpha Thalassemia Carrier

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Objective: Thalassemia is one of the most common single gene diseases with a worldwide distribution. It's a heterogeneous disease. Beside the large deletions, it has more than fifty point mutations. Between four to ten percent of the Iranian are carrier for thalassemia affected genes. In this study we found a relationship between the value of MCV/MCH and the type and severity of mutations.

Methods and Subjects: The most important difference between a thalassemia carrier and

normal persons is decreased value of MCV and MCH. Subjects of this study were adult alpha thalassemia heterozygote between 20- 40 years old (208 chromosomes of 33 persons from Khoozestan and 71 persons from Khorasan provin). DNA was isolated from peripheral blood leukocytes using standard procedures. DNA samples were amplified by Gap-PCR.

Findings: In our study we found just one α^0 mutation (MED: 2.9%). All the α^0 mutation had virtually identical range of MCV and MCH. In contrast α^+ mutations were associated with significant differences in mean MCV and MCH. The various Mean MCV/MCH of α^0 carriers were statistically significantly lower than those of α^+ heterozygote.

Conclusion: The result indicated that degree of reduction in MCV and MCH is directly related to the severity of the mutations. Deviations, in cases, were associated with lower number of the carriers.

Key Words: Alpha Thalassemia; Gene Mutations; CBC

Poster Presentation

Evaluation of the Association between Paternal Occupational Exposure and Children Cancer

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Objective: The incidence of childhood cancer has been increasing nearly one percent per year for the past two decades. Leukemia and lymphoma are the most common types of childhood cancers. This study was performed to assess environmental factors (hydrocarbon, agricultural toxin, insecticide) of paternal occupational exposures and children cancers.

Methods and Subjects: This is a case-control, cross sectional study on 78 children with leukemia and non-hodgkin lymphoma and 78 control group between 2002-2005. Data was gathered on questionnaire and analyzed by chi-square test.

Findings: There was 44.9% girls and 55.1% boys in case group which most of them (29.6%) were in 3-5 years age category. Their father's were farmers (57.7%), painters exposed to hydrocarbon (16.7%) and workers (6.4%). Comparison of case and control showed significant difference between fathers occupation in two groups.

Conclusion: This study identified that the rate of malignancy in children's whose fathers occupation are related to hydrocarbon exposure, painting and farming are higher than others. Though fathers should have more attention and protection against the risk factors.

Key Words: Leukemia; Lymphoma; Paternal occupational exposure

Oral Presentation

Rituximab for Child with Chronic Relapsing Autoimmune Hemolytic Anemia

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Objective: Warm-type idiopathic autoimmune hemolytic anemia (AIHA) is a relatively common hematologic disorder resulting from autoantibody production against red blood cells. Steroids represent the first-line therapeutic option, and immunosuppressive agents as well as splenectomy are used for refractory cases. Recently, the anti-CD20 monoclonal antibody rituximab has been shown to control autoimmune hemolysis in patients with refractory chronic disease.

Cases Presentation: We reported the results from a retrospective analysis of 5 children receiving rituximab for steroid-refractory AIHA of the warm type at a mean age of 9 yr (range 3-14 yr). All patients were given methyl-prednisolone as first-line treatment and some of them also received azathioprine and intravenous immunoglobulin. All patients were considered refractory to steroids and/or immunosuppressive drugs and all were given weekly rituximab (375 mg/m²) for four weeks. Two patients required packed red cell transfusions before starting rituximab and all became transfusion-free. At a mean follow-up of 443 d (range 60-1059 d) since the treatment of AIHA with rituximab, all patients are alive, and all of them in complete remission (CR) and two patients had combs' test positive.

Conclusion: Our study show that anti-CD20 rituximab is an effective and safe alternative treatment option for idiopathic AIHA, in particular, for steroid-refractory disease.

Key Words: Autoimmune hemolytic anemia; Steroid-refractory; Rituximab

Poster Presentation

Assessment of the Relation between Sensory Neural Hearing Loss and Desferal Therapy in Thalassemic Patients

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Objective: Thalassemic patients need regular transfusion & iron chelator such as desferal in order

to decrease iron overload. Desferal itself has several side effects like auditory side effects which include sensory neural hearing loss in high frequencies. Despite a lot of researches in this area the etiology is still being debated. This study assesses the above mentioned auditory side effects in these patients.

Methods and Subjects: This was a cross sectional study. 53 beta thalassemia major patients referred to special medical center in Tehran who were under regular blood transfusion regular desferal consumption at least for 5 years were included in this study (35 females & 18 males). Hearing loss was assessed using audiometry & PTA. Patients with a history of auditory problems, ototoxic drug intake or any other hearing impairments and patients who had interruption in desferal consumption or irregular blood transfusion were excluded from the study.

Findings: No relation between desferal dosage or duration of it's consumption and sensory neural hearing loss was found.

Conclusion: These results reveal that although no relation between desferal dosage and it's duration of consumption with SNHL was found, perhaps a more precise examination will (reveal)? more information, further it is recommended to repeat the study on a larger population.

KeyWords: Thalassemia; Sensory neural hearing loss; Desferal

Oral Presentation

Why Bone Marrow Transplantation and for Whom

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Transplantation of allogeneic and autologous hematopoietic stem cells has become an increasingly safe and effective procedure in recent years and is now established as one of the most important curative strategies in patients with hematological malignancies. It also has an important role to play in the management of acquired marrow failure, hemoglobinopathies, congenital immunodeficiency and metabolic disease. Briefly, indications for BMT in children are as follows, high risk ALL in first CR, high-and intermediate risk relapsed ALL in second CR, all in third or more CR, poor risk AML in first CR, AML in second CR, chronic myeloid leukemia, myelodysplasia including myelomonocytic leukemia, relapsed burkitt's NHL, relapsed diffuse large cell NHL, relapsed anaplastic large cell lymphoma, relapsed T cell lymphoblastic NHL, relapsed or refractory Hojkin disease, high risk solid tumors like stage 4 neuroblastoma, high risk Ewing sarcoma, high risk or relapsed medulloblastoma, refractory or relapsed Wilms or Germ cell tumors, Aplastic anemia, Fanconi anemia, Diskeratosi congenita, Congenital amegakaryocytic

thrombocytopenia, Schwachman-diamond syndrome, Diamond blackfan anemia, Kostmann syndrome, Thalassemia, Sickle cell anemia, Primary immunodeficiencies, Osteopetrosis and certain metabolic disease. The goal of this article is increasing knowledge of pediatrician for indications of BMT in pediatric field for their better judgment for referring patients to BMT centers.

KeyWords: BMT; Indication; Pediatric

Oral Presentation

Irradiation of Blood Components for Prevention of Transfusion Associated Graft-Versus-Host Disease

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Graft-versus-host disease (GVHD) results from the engraftment of immunocompetent donor T lymphocytes into a recipient whose immune system is unable to reject them. It is a common sequela of bone marrow transplantation (BMT), but is also recognized as a rare risk associated with blood transfusion. Virtually all cellular blood components

have been implicated in reported cases of TA-GVHD. The syndrome has developed after transfusion of whole blood, red blood cells, platelets, fresh (nonfrozen) plasma and leukocytes harvested from both normal donors and donors with chronic myelocytic leukemia. Because the treatment of TA-GVHD is almost always ineffective, efforts are directed at prevention and minimizing risk by reducing or inactivating transfused donor lymphocytes. Available in blood banks for physically removing T lymphocytes (washing or filtration) do not provide effective prophylaxis against TA-GVHD. Current 3-log leukocyte depletion filters do not remove sufficient lymphocytes to prevent TA-GVHD. Inactivation of transfused lymphocytes by gamma irradiation of blood components remains the most effective method for inhibiting lymphocyte blast transformation and mitotic activity and hence preventing TA-GVHD. The well defined indications for the use of irradiated blood and blood products are listed below and other issues will be discussed in detail. Clearly established indications: Congenital T-cell defects (known or suspected), Immunologic immaturity (fetus or premature infant), Intrauterine transfusion, Exchange transfusion, Acquired T-cell defects, Bone marrow or peripheral blood stem cell transplant recipients (allogeneic or autologous), Hodgkin's disease, Haplotype sharing between donor and recipient, Transfusions from biologic relatives, Transfusion of HLA-matched platelets.

Key Words: Irradiation; Blood components; TA-GVHD