IJBC
Iranian Journal of Blood & Cancer
Volume 3 Supplement 1 August 2015

انجمن خون و سرطان کودکان ایران
Iranian Pediatric Hematology Oncology Society

The International Congress

Challenges in Pediatric Hematology & Oncology

SEP 16 - 18, 2015
TEHRAN, IRAN

TOPICS

1-Leukemia
2- Lymphoma
3- Histiocytosis
4- Anemia
5- Hemoglobinopathies
6- Bleeding and Thrombotic Disorders
7- Neonatal Hematology
8- Stem Cell Transplantation

Abstract Submission Deadline:
May 20, 2015

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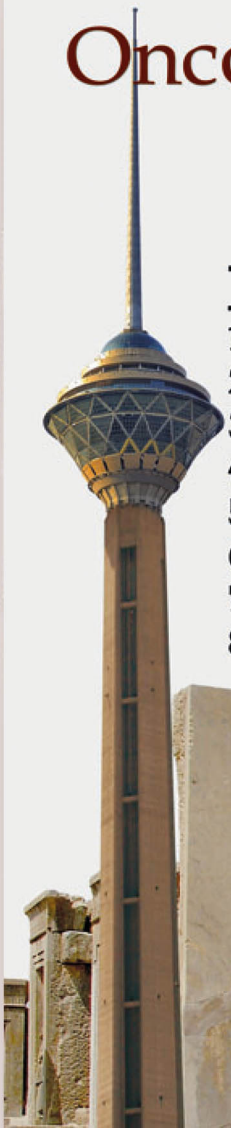

ABOUT IRAN

Iran comprises a land area of over 1. 6 million square Km (the 17th country in the world by land area) it is located in south west of Asia and is one of the middle-east countries. Iran lies down the northern temperate zone, between latitudes 25 degree north and 39 degree 47' north & between longitude 44 degree 02' east and 63 degree 20' east.

Iran is bounded by Turkmenistan, Caspian Sea, Azerbaijan, and Armenia on the North, Afghanistan and Pakistan on the East, Oman Sea and Persian Gulf on the South, and Iraq and Turkey on the West.

Iran, country of Cyrus the great, Dariush, Xerxes, and the poets Ferdousi, Hafez, and Omar Khayyam, is one of the oldest civilizations in Asia and in the entire world.

The International Congress

Challenges in Pediatric Hematology & Oncology

SEP 16 - 18, 2015
TEHRAN, IRAN

Dear colleagues

On behalf of the executive committee of the, we welcome you and extend our warm and cordial invitation to you for the **international congress of challenges in pediatric hematology and oncology** (CPHO) to be held on Iran, Tehran. This congress will be held with the participation of many renowned worldwide hemato-oncologists to exchange information on the latest findings and researches performed in the world to bring scientific that are active in this field closer.

It will be a great pleasure to welcome you to our city. We will organize a challenging and exciting scientific congress together with the international board and scientific committee. The congress will be focus on leukemia, lymphoma, histicytosis, anemia, hemoglobinopathies, bleeding and thrombotic disorders, neonatal hematology and stem cell transplantation

The focus of these symposia will be on areas of controversy, recent progress in molecular understanding and of course recent progress in therapy. Other legislative measures concerning the role of pediatric oncology nurses will change the working situation in pediatric oncology hospitals.

Your participation will undoubtedly add to the scientific value of the congress and will help to answer the numerous questions of the participants in the field of hemato-oncology, especially the questions regarding your lecture appropriately and practically.

We encourage members and non-members to attend in congress, to submit abstracts and enjoy a productive and enjoyable conference.

Looking forward to welcoming you to Tehran in September 2015.

Worldwide cooperation for improving management of childhood blood disorders

With best wishes
Prof. Hassan Abolghasemi
Congress President

Tuesday (15/Sept/2015)	
7.00 pm to 9.00 pm	**Registration and Reception**
Wednesday (16/Sept/2015)	
8.00 am to 9.00 am	Educational Session: **Histiocytosis**
9.00 am to 10.30 am	**How I treat relapsed Acute Leukemia?**
10.30 am to 11.00 am	Coffee break
11.00 to 12.45 pm	Presentations: **Leukemia**
12.45 pm to 2.00 pm	Lunch
2.00 pm to 4.30 pm	Presentation: **Lymphoma**
4.30 pm to 5.00 pm	Coffee break
5.00 pm to 6.00 pm	**Poster round**
7.30 pm to 10.30 pm	**Opening ceremony**
Thursday (17/Sept/2015)	
8.00 am to 9.00 am	Educational Session: **ITP**
9.00 am to 10.30 am	**How I treat Chronic ITP?**
10.30 am to 11.00 am	Coffee break
11.00 to 12.45 pm	Presentations: **Bleeding disorders and thrombosis**
12.45 pm to 2.00 pm	Lunch
2.00 pm to 3.30 pm	Presentation: **Neonatal Hematology**
3.30 pm to 4.30 pm	Presentations: **Anemias**
4.30 pm to 5.00 pm	Coffee break
5.00 pm to 6.00 pm	**Poster round**
Friday (18/Sept/2015)	
8.00 am to 9.00 am	Educational Session: **Hgb Electrophoresis**
9.00 am to 10.30 am	**How I treat Thalassemia?**
10.30 am to 11.00 am	Coffee break
11.00 am to 12.30 pm	Presentations: **Hemoglobinopathies**
12.30 pm to 13.00 pm	**Closed Ceremony**

انجمن خون و سرطان کودکان ایران
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Iranian Journal of Blood and Cancer

The Official Journal of

Iranian Pediatric Hematology and Oncology Society (IPHOS)

Volume 3 Supplement 1 August 2015

About the Journal:

In 2005, Hassan Abolghasemi, professor in Pediatric Hematology and Oncology in collaboration with some of his colleagues, decided to establish a scientific English journal to extend the relationship with other national and international scientists. They obtained the appropriate permissions from the respective authorities and finally, the first issue of "IJBC" was published in autumn 2008, about 4-5 months after receiving the permission.

"Iranian Pediatric Hematology and Oncology Society (IPHOS)", with more than a decade of activity, is proud of being the responsible entity to publish "IJBC". IJBC enjoys the contribution of its credible editorial board members consisting of a diversity of specialists and academics in basic sciences, hematology, immunology, genetics and clinical disciplines of hematology, oncology, radiotherapy, pathology, pediatric surgery and cancer surgery.

Aim & Scope:

IJBC tries to provide a new opportunity for advancing the field of Hematology and Oncology in Iran and make a bridge between Iranian researchers and fellow scientists globally.

This journal is published in print and online and includes high quality manuscripts including basic and clinical investigations of blood disorders and malignant diseases namely: diagnosis, treatment, epidemiology, etiology, biology and molecular aspects as well as clinical genetics of these diseases, as they affect children, adolescents and adults. "IJBC" also includes the studies on transfusion medicine, hematopoietic stem cell transplantation, immunology, genetics and gene-therapy and accepts original papers, systematic reviews, case reports, brief reports and letters to the editor in all aspects of blood transfusion, blood donors recruitment, screening techniques, modern approaches to transfusion, whole blood and blood components applications.

We also cover subjects related to cancer including:

• Risk factors: life style, gene – environment interactions, molecular triggers, and strategies for reducing the risk.

• Public health issues: epidemiology, cost-effectiveness of procedures, health technology assessment, cancer registry, social determinants of health, health needs, quality of life measurements, public and professional education, and cancer control programs.

• Cancer diagnosis and prognosis: normal and abnormal hematopoiesis, benign and malignant tumors, molecular markers; diagnostic imaging; defining tumor margins; detecting minimal residual disease.

• New approaches to cancer therapy: rational drug design, gene therapy, immunotherapy, combination therapy, drug resistance, targeting therapies to the individuals, and alternative/complimentary medicine.

• Experimental systems and techniques: cell culture and animal models, genomic and proteomic approaches to studying cancer, cell proliferation and differentiation, apoptosis and molecular aspects of cytokines.

• Cancer-associated conditions: pain, cachexia, symptoms associated with treatment (hair loss, anemia, gastrointestinal disorders), psychosocial aspects of cancer.

• Ethical and legal issues surrounding cancer research: trial design, genetic screening, communicating with patients and families, death issues, research policy and advocacy.

We also cover articles regarding the history of contemporary medicine in Iran: Manuscripts narrating the history of modern medicine in Iran, the outstanding scientists' contribution to its progress, and the improvement of our health-care system over the past decades are important to us and are welcome.

Legal disclaimer:

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The manuscript should be prepared according to the journal style, using American English spelling and punctuation. Authors must submit their manuscript electronically to expedite the peer review process via online at: www.ijbc.ir

The Peer Review System:

Manuscripts will be reviewed by the editorial board. They will be considered if they are in harmony with the standards of the journal. If the editors find major faults in their structure, they will be rejected. Editorial board deals with manuscripts blindly. Papers are then sent to the corresponding authors for suggested modifications and the corrected version of the manuscript will be peer-reviewed for a second time by two or three external reviewers.

Type of Articles:

Original Articles: Should contain title page, abstract, keywords, introduction, materials and methods, results, discussion, conclusion, acknowledgment, references, tables, and figures, enumerated from the title page. The length of the text should be limited to 3000 words excluding the references and abstract.

Case Reports and Brief Reports: Should not exceed 1500 words. Both should include abstract, keywords, introduction, case presentation, discussion, conclusion acknowledgment, and references. Case reports might have 1 to 4 accompanying figures and/or tables but brief reports should not have more than one figure or table. Necessary documentations of the case(s) like pathology and laboratory test reports should be included in the submission package.

Clinical Trials: should contain patients' informed consent and the approval of the ethics committee of the corresponding institution.

Review Articles: might be requested by the editor, but IJBC will also accept submitted reviews. Both solicited and unsolicited review articles are subjected to editorial review like the original papers.

Letters to the Editor: IJBC accepts letters to the editor. Letters should be less than 500 words. Letters might discuss articles published in the journal during the previous six months or other important aspects related to the field of hematology. Letters will undergo peer-review processing and will be edited for clarity.

Structure of Articles:

Title Page: the first page of the manuscript should contain the following information: (1) Title of the paper; (2) Authors' names; (3) Authors' affiliations; (4) Acknowledgements for research support; (5) Name, address, email, phone number and fax number of corresponding author; (6) Any financial interests, direct or indirect, that exist or may be perceived to exist for individual contributors in connection with manuscript should be disclosed.

Abstract: Abstract for original papers, should contain background, materials and methods, results, and conclusion sections separately. The objective of the study, findings (including its statistical significance), and the conclusion made on the basis of the findings should be clearly presented. For case reports a one paragraph, non-structured abstract or a structured abstract divided to background, report of the case, and conclusion is needed. Nevertheless, the length of an abstract should be less than 250 words (150 words for case reports).

Keywords: each submitted article should contain three to five keywords, which must be chosen from the

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Body of the Manuscript: the article should be organized and formatted according to the following headings: (1) Background, (2) Materials and methods, (3) Results, (4) Discussion, (5) Conclusion(s). Authors may use "patients and methods" instead of "Materials and methods" in papers dealing with human subjects. The body of the paper must be written as concisely as possible. The body of case reports should not exceed 1500 words. They should include introduction, case presentation, discussion, conclusion, acknowledgment, references, and 1 to 4 figures.

Preparing Tables and Figures: Tables and figures must be listed in the order cited in the text using Arabic numerals. Tables should be typed double-spaced; each on a separate page. Tables and figures should not be embedded in the text, but should be uploaded as separate files. A short descriptive title should appear above each table with a clear legend and any footnotes or Captions suitably identified below. Provide headings for all columns and indicate units of measurement, where applicable.

Illustrations: For highest quality reproduction of illustrations we recommend submitting each figure as high resolution TIFF (300 dpi minimum) or EPS (800 dpi) files with any fonts embedded. Color illustrations will be considered for publication.

References: The authors are responsible for the accuracy of references, which must be numbered respectively in the order of citation in the text. Reference citations should be marked by superscript Arabic numbers. Personal communications and unpublished data including manuscripts submitted but not yet accepted for publication should not be used as a reference; however, they may be placed in parentheses in the text. Journal titles should be abbreviated according to Index Medicus. Inclusive page numbers should be given for all references. When there are six or less authors, surnames and initials of all authors should be mentioned. In the case of seven or more authors, the names of the first six authors followed by et al. should be given.

Presented below, are references to a journal, a chapter in a book, and a book, respectively, in the style requested by our journal:

Journal Article: Vega KJ, Pina I, Krevsky B. Heart transplantation is associated with an increased risk for pancreatobiliary disease. Ann Intern Med. 1996; 124:980-3.

Article Not in English: Ryder TE, Haukeland EA, Solhaug JH, Bilateral infrapatellar senseruptur his tidligere frisk kvinne. [English translation] Tidsskr Nor Laegeforen. 1996; 116:41-42

Book: Ringscen MK, Bond D. Gerontology and leadership skills for nurses, 2nd ed. Albany, NY: Delmar Publishers,1996.

Chapter in Book: Phillips SJ, Whisnant JP. Hypertension and Stroke. In: Laragh JH, Brenner BM, editors. Hypertension: pathophysiology, diagnosis, and management, 2nd ed. New York: Raven Press, 1995, 465-78.

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Ethical Guidlines: Ethical Items must be inserted in the "Materials and Methods" Part. If you have to perform some tests on the human, please obtain the consent from all human adult and from the parents or legal guardians of children and also state it in the text (include the name of the reviewers that approved the project). About the experimental animals tests and therapies, please indicate that they complies with National Institutes of Health Guidelines on the laboratory animals.

Conflicts of Interest: Authors must acknowledge any sources of funding and conflicting interest, such as funds, fees and stocks. Also announce that a competing interest will not lead to automatic rejection of the paper.

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ARTICLES OF GUEST SPEAKERS

ARTICLES OF SPEAKERS

POSTERS OF STUDENTS

ARTICLES OF GUEST SPEAKERS

IJBC 2015; Supplementary; P 1
Paper ID: 132

The role of Psychiatrist in cancer management

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1. Department of psychiatry, Iran University of Medical Sciences, Tehran.

Abstract

Aim: Emergence of cancer is a crisis. When a child or teenager is diagnosed with cancer, the patients and their families specially parents and siblings will encounter a lot of problems. The diagnosis may lead to shock, denial, anger, irritability, despair, fear, depression and anxiety in most patients and the people who love them. These psychological reactions could affect level of functioning and quality of life. Financial problems, life style changing, care giving, fear of the future and a lot of other psycho- social stressors put a lot of pressure on the patients and family. Furthermore parents don't know how to ask help from others and even worse they don't know if it is a good decision to tell the truth to their child or children or how to tell them the bad news. The aim of this article is to report the finding of our study about care giver burnout and depression and anxiety in parents and to review the psychosocial aspects of childhood cancer and the role of psychiatric intervention for coping with cancer. Method: In this cross-sectional study on 185 parents of children with ALL, care giver burnout and depression and anxiety were assessed by Hospital Anxiety and Depression Scale (HADS) and caregiver burden inventory. Descriptive analysis, chi-square and t- test were used. Results: the rate of caregiver burnout in 2% was mild, in 18% was moderate and in 71% was severe. In 26% of subjects HADS-A>8 and in 23% HADS-D>8. Conclusion: Our findings showed a high prevalence of psychiatric problems in parents of children with cancer. As a result the importance of psychiatric assessment and intervention for children with cancer and their family is concluded.

Keywords: Psychiatric intervention, cancer, burnout, depression

IJBC 2015; Supplementary; P 2
Paper ID: 145

How I treat childhood ALL?

M Schundeln

Abstract

Lymphoblastic leukemia is the predominant form of childhood malignancies. Survival has improved dramatically within the last decades from less than 20% to at least 80%. Although the standard treatment of ALL is effective, the total treatment is lengthy, complex and has considerable late effects.

ALL relapse, which occurs in approximately 20% of patients, was considered to be fatal until recently. However, approximately 60% of these patients can currently be cured with adequate chemotherapy and stem cell transplantation.

One major challenge for the treatment of patients with pediatric ALL is adequate risk stratification in the initial and relapse treatment. Patients with favorable disease must be identified to spare therapy toxicity. However, it is also important to identify patients at risk for relapse and non-response early on. The first part of the talk will provide an overview of the current I-BFM treatment protocol. Here, the focus will be on the strategies used to spare toxicity in various patient groups. In the second part of the talk, the biology and treatment of relapsed ALL will be presented. With a growing number of promising new drugs, prospective trials are urgently required. Strategies to systematically implement and test these new drugs in treatment regimens will be discussed.

IJBC 2015; Supplementary; P 3-4
Paper ID: 146

How I treat childhood AML?

Drik Reinhardt

Abstract

Pediatric AML is still one of the most life-threatening malignancy in children and adolescent. Intensive chemotherapy amended by an optimized risk stratification, improved supportive care and allogeneic stem cell transplantation (SCT) in high risk AML actually enabled an survival of more than 70% in developed health care systems.

A generally accepted but simplified model of leukemogenesis postulates the combination of type II aberrations (translocations such as t(8;21) or inv(16)) which affected differentiation and maturation and type I mutations (expl. FLT3-ITD) which supported proliferation and impaired apoptosis. Recently, the relevance of the clonal hierarchy and mechanism of evolution comes into the focus.

Diagnostics procedures includes morphology, immunophenotyping and molecular- and cytogenetics. Some study groups already introduced targeted next generation sequencing into routine diagnostics. Central review of morphology, immunophenotyping and genetic analysis at diagnosis and during course of disease (response assessment) seems to improve quality and outcome.

Regarding treatment, the most subgroups of pediatric AML required very intensive therapy based on cytarabine and anthracyclines. Risk group stratification into standard risk, intermediate risk and high risk based on genetics and response.

Recently, in acute promyeloblastic leukemia (APL) with t(15;17) the striking efficacy of arsenic trioxide (ATO) and all-trans retinoic acid (ATRA) combined with a favorable toxicity profile have been shown in adults. Therefore, this combination is recommended in

children, too. Only in high risk APL (>10.000 WBC/µl), an initial course of chemotherapy should be added to reduce cell numbers.

In all other AML, intensive double induction followed by consolidation and re- intensification is necessary. In children with high risk AML, SCT in 1st complete remission is recommended. Cumulative dosages of cytarabine and anthracyclines differs between study groups. Whereas the AML-BFM1, COG1 and UK protocols achieved anthracycline dosages of 400mg/m² and more, the NOPHO1 and Japanese protocols are more based on high dose cytarabine (>50g/m²). The development in the recent decade showed a more and more convergence in terms of risk group definition and the indication of SCT in 1st CR. It is common sense that pediatric AML with t(8;21), NPM1mut, CEBPAdouble mutation or inv(16) could be cured by chemotherapy only. On the other hand several MLL-translocations (t(4;11), t(6;11); t(10;11) and other), complex karyotype, 12p-abberration or exclusive monosomy 7 are considered as high risk AML.

Countries and study groups with less resources should critically review the very aggressive treatment protocols and adapt them to the local conditions in order to optimize survival but to minimize treatment related mortality.

Despite the major progress within the last years, further intensification of therapy seems impossible because of the high frequency of treatment related morbidity and mortality. In consequence there is an urgent need to identify and validate innovative, more precise treatment options either with targeted compounds or introduction of new immunotherapies such as antibody based or engineered cell therapies. Precondition is a broad profiling of leukemic blasts or leukemic stem cells analyzing clonal hierarchy, molecular aberrations and signaling.

New strategies and trial designs are mandatory, to enable further progress and finally allow cure of pediatric AML in a reasonable future.

IJBC 2015; Supplementary; P 5
Paper ID: 147

How I treat childhood MDS?

Drik Reinhardt

Abstract

Myelodysplastic syndrome in pediatrics is a rare disorder of hematopoiesis. In children MDS frequently is characterized by a refractory cytopenia of childhood (RCC) of all lineages.

Intermediate stages are the refractory anemia with excess of blasts (RAEB), RAEB with transition to AML (RAEB-T) and MDS-related AML. Whereas patients with RCC showed a normal karyotype in about 2/3, advanced MDS (RAEB, RAEB-T, MDR-AML) were characterized by typical genetic aberrations such as monosomy 7, 7q- or complex karyotypes. Depending on genetic aberrations there is a variable risk to progress to MDRAML.

Only in a minority, familial cases with mutations in GATA2 or RUNX1 are known. The most crucial point is the correct diagnosis because distinction from severe aplastic anemia (SAA) could be difficult. Minimal procedures are bone marrow aspiration and biopsy analyzed by morphology, immunophenotyping and molecular-/cytogenetics. In addition, several inherited bone marrow failure syndromes (Fanconi-Anemia, dyskeratosis congenital amegakaryocytic thrombocytopenia etc.) should excluded.

The necessity of treatment depends on stage and genetics. In patients with genetic aberrations, treatment should be initiated. In RCC without clinical symptoms, lack of neutropenia (ANC > 1000/µl) or no need of regular transfusions, a wait-and-watch strategy is justified.

The only curative approach is allogeneic stem cell transplantation. In RCC and RAEB, just conditioning chemotherapy prior to SCT seems to be required. An exception might be the subgroup of hypocellular MDS without genetic aberrations, in which an immunosuppressive therapy (IST) similar to the approach in SAA could be performed. In advanced MDS, an induction regimen in order to control blast proliferation could be beneficial; however, the aim is not complete remission but reduction of blasts to levels of less than 10%. Recently, the role of demethylation by agents such as azacytidine showed promising results. In general, the 5-years overall survival in MDS achieved more than 70%, however, advanced MDS remains in a range of 60% survival. Unfortunately, in some subgroups such as advanced MDS with structurally complex karyotype the prognosis is poor (14%).

Further research is urgently needed to learn more about the molecular and epigenetic mechanism in early hematopoiesis, possible inherited predisposition and more precise treatment options, at least in advanced MDS.

IJBC 2015; Supplementary; P 6-7
Paper ID: 148

How I treat non-transfusion dependent Thalassemia?

Ali Taher

Abstract

Introduction Transfusion-dependence in thalassemia has been classically utilized as a factor to distinguish the various phenotypes and their severity. Non-transfusion-dependent thalassemias (NTDT) are those phenotypes which do not require lifelong regular transfusions for survival, although they may require occasional or even frequent transfusions in certain clinical settings and for defined periods of time. Of the numerous NTDT genotypes, the three most studied are: β-thalassemia intermedia (β-TI), α-thalassemia intermedia (HbH disease) and hemoglobin E/β-thalassemia, with the latter being by far the most common, accounting for about 50% of moderate to severe cases. Therapy of NTDT patients conventionally includes a combination of transfusions, splenectomy, fetal hemoglobin induction and iron chelation. Iron chelation is particularly important, as will be demonstrated during this presentation, as it has been shown to be associated with significant morbidity. Pathophysiology & Clinical Complications The clinical picture seen in NTDT is due to an interaction of three hallmark factors: ineffective erythropoiesis, chronic anemia, and iron overload; each associated with certain comorbidities. Ineffective erythropoiesis is the primary factor leading to a chronic anemic state, whereas peripheral hemolysis remains secondary. Moreover, ineffective erythropoiesis leads to compensatory extramedullary hematopoiesis (EMH) and as such is associated mainly with skeletal complications, while peripheral hemolysis has been more implicated in hypercoagulable complications such as pulmonary hypertension (PHT), with secondary heart failure (HF), and thromboembolic phenomena. The chronic anemic state eventually leads to hepcidin suppression by erythroid factors, increased iron absorption from the gut and increased release of recycled iron from the reticuloendothelial system (RES); all culminating in nontransfusional iron overload, mainly in the liver and less so in the heart. Further iron accumulation may occur with occasional transfusions required in acute stress settings or more frequent transfusions required in growing children or adults with vascular complications. Iron overload per se accounts for a significant proportion of morbidity in NTDT. In a recent study of 168 non-chelated patients with NTDT, higher Liver Iron Concentration (LIC) values on magnetic resonance imaging were associated with a significantly increased risk of developing thrombosis, pulmonary hypertension, hypothyroidism, hypogonadism, and osteoporosis. Specifically, LIC levels ≥5 mg Fe/g dry weight were associated with a considerable morbidity risk increase. Although LIC assessment remains the gold standard for quantification of total body iron, in resource-poor countries serum ferritin (SF) measurement may be the only method available for the assessment of iron overload. Observational studies confirm a positive correlation between SF level and LIC in NTDT patients. A recent longitudinal follow-up over a 10-year period (ORIENT study) confirmed these findings, demonstrating that the cumulative incidence of multiple morbidities was highest in patients with SF ≥ 800 ng/ml and lowest patients had SF ≤ 300 ng/ml. However, it should be noted, that close to 50 % of patients with a SF level between 300-800 ng/ml may still have an LIC ≥ 5 mg Fe/g dw, highlighting the need for LIC assessment in this subgroup of patients. Without appropriate treatment, the incidence of morbidities increases with advancing age. Management As mentioned above, the four cornerstone of NTDT management are transfusions, splenectomy, HbF inducers and iron chelation. Traditionally, transfusions were reserved for palliation of late and irreversible anemia-related complications; however, more recent evidence supports an earlier role in NTDT management. The OPTIMAL CARE study, demonstrated that NTDT patients placed on (intermittent

or regular) transfusion regimens suffered fewer complications; mainly EMH, PHT and thrombosis. Thus, there is still merit in considering blood transfusions, with indications including acute stress (Hb level < 5 g/dL, surgery, infection or pregnancy), progressive changes from childhood (declining Hb level in parallel with profound splenomegaly, growth failure, failure of secondary sexual development in parallel with bone age, severe bony changes) and other complications (thrombotic or cerebrovascular disease, PHT, extramedullary hematopoietic pseudotumors, leg ulcers). However, in the absence of randomized trials, there are no specific guidelines on the amount and duration of transfusions needed and thus, therapy should be tailored to individual patient needs. Splenectomy is common practice in NTDT patients, and serves to increase total Hb level by 1-2 g/dl; however, cumulative evidence confirms an association with a variety of adverse outcomes. The spleen functions to scavenge procoagulant platelets and RBCs, which together are the key factors underlying the hypercoagulable state observed in NTDT. Splenectomized NTDT patients are at increased risk for venous thromboembolism, PHT, leg ulcers and silent cerebral infarction than their non-splenectomized counterparts. Nonetheless, splenectomy may be indicated in certain clinical settings such as: hypersplenism (resulting in worsening anemia, leucopenia or thrombocytopenia or their clinical manifestations), worsening anemia leading to poor growth (when transfusion therapy is not possible) or splenomegaly (accompanied by left upper quadrant pain, early satiety or concern of splenic rupture). HbF inducers work by increasing γ-globin production, a β-like globin molecule which can bind excess α- chains, thus decreasing the α/β-chain imbalance inherent to thalassemia, and improving effective erythropoiesis. Hydroxyurea (or hydroxycarbamide) has been the most studied HbF inducer in NTDT. Early case reports documented hematological improvements in β-thalassemia patients treated with hydroxyurea, and since then several studies have evaluated this drug in NTDT. However, the data is conflicting and mostly comes from single-arm trials or retrospective cohort studies. Although HbF inducers are a potentially promising aspect in NTDT treatment, large, randomized, controlled trials are needed before these agents or their derivatives are widely used in management. Finally, iron chelation therapy is currently the cornerstone of managing NTDT patients and minimizing disease related complications. Three chelators are available to treat iron overload: Deferoxamine, Deferiprone, and Deferasirox. Subcutaneous Deferoxamine was the first iron chelator studied in NTDT. The current challenges posed by this drug are two-fold: there is a lack of solid evidence from large studies on the benefit of this drug in NTDT patients, and more importantly, its cumbersome subcutaneous administration may cause compliance challenges. Deferiprone, although orally administered, has only been evaluated in few trials with relatively small sample sizes. Deferasirox on the other hand, has been extensively studied in NTDT and is currently the only iron chelator specifically approved for NTDT patients. This once-daily administered oral iron chelator was the agent used in the largest and first randomized clinical trial of chelation therapy in NTDT (THALASSA). Patients ≥10 years of age with LIC ≥5 mg Fe/g dw received 12 months of therapy with Deferasirox and a significant reduction of LIC was observed, as compared to placebo. LIC decreased by a mean of 2.33 ±0.70 and 4.18 ± 0.69 mg Fe/g dw in patients receiving starting doses of 5 mg/kg/day and 10 mg/kg/day, respectively. Doses were doubled after 6 months for patients with LIC >7 mg Fe/g dry weight and

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Thalassemia intermediate

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Abstract

B thalassaemia is an inherited disorder of haemoglobin (Hb) synthesis wherein mutations of the b globin gene lead to various degrees of defective b chain production, an imbalance in a/b globin chain synthesis, ineffective erythropoiesis, and a spectrum of anaemia . Extremely diverse phenotypes exist within the b thalassaemia syndromes. Almost all patients with thalassaemia syndromes are classified as having thalassaemia minor or thalassaemia major (TM). However, a small number of patients did not fit into this categorical distribution. Sturgeon was the first to describe this group in the American literature. He suggested the term thalassaemia intermedia (TI) to describe patients who had clinical manifestations that are too severe to be termed minor and too mild to be termed major. The understanding about the molecular and pathophysiological mechanisms underlying the disease process in patients with Bthalassemia intermedia has substantially increased over the past decade. The hallmark of disease process in patients with TI includes ineffective erythropoiesis, chronic haemolytic anaemia, and iron overload. Distinction between the various phenotypes of b-thalassemia relies primarily on the clinical severity of the disease,which should be assessed both at initial presentation and over a period of close follow-up.They show mild to moderate anemia and a hemoglobin level ranging between 7 and 10 g/dL,which is sustainable without the need for regular transfusion therapy.The patient's wellbeing, particularly with respect to activity, growth, development, and the early appearance of skeletal changes or other morbidities, is the factor to be taken into consideration before the phenotype is clearly established and the treatment modality is selected. Diagnosis remains largely clinical, a genotype/phenotype association has been described. Most TI patients are homozygotes or compound heterozygotes for b-thalassemia, meaning that both b-globin loci are affected and the disease has a recessive genetic pattern.Less commonly, only a single b-globin locus is affected, the other being completely normal; thus, in these instances, TI is dominantly inherited (Weatherall and Clegg 2001). The phenotype of TI may also result from the increased production of aglobin chains by a triplicated or quadruplicated a-genotype associated with b-heterozygosity . Current evidence highlights that transfusion independence in TI does not come without its own side effects.Knowledge of the various clinical morbidities that could emanate from these underlying mechanisms continues to expand, and it is now apparent that TI patients experience a spectrum of morbidities that remain different from those commonly observed in TM. Complications Iron overload and target organ toxicity, Hypercoagulability and Thromboembolic disease, Pulmonary hypertension, Extramedullary hematopoietic pseudotumors, Leg Ulcers, Endocrine disease, Gallstones. It is evident that without treatment, TI patients experience more frequent morbidity and poorer healthrelated quality of life, Management Transfusion therapy ,Iron chelation, , Fetal hemoglobin induction, Anticoagulation ,Splenectomy, Hematopoietic stem cell transplantation(HSCT). Until ongoing efforts optimise haematopoietic stem cell transplantation or gene therapy as a cure for patients with TI, medical therapy will be the corner stone for management. However, despite a number of available treatment options, there are currently no clear guidelines for managing patients with TI. Here we will present some cases of B thalassemia intermedia and discuss about their issues

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How I Treat Childhood NHL? Newly diagnosed and relapsed patients

Catherine Patte

Abstract

First line treatments

B-cell lymphoma is the most frequent lymphoma in children (50-70% of the cases), with a majority of BL. The latest generally arise in abdomen and head and neck region and present as advanced stage disease. Bone marrow and CNS are involved in about 20-25% of the cases. Treatment of BL must be done by intensive pulse polychemotherapy courses, but is of short duration, relapses occurring early, within the first year. The French LMB studies (Patte JCO 1991 & Blood 2001, Gerrard BJH 2008, Patte Blood 2007, Cairo Blood 2007) told us that :a) HDMTX is a very effective drug for prevention of CNS disease b) B-ALL and CNS positive patients benefited from the increase of the dose of HD MTX and the introduction of HD ARA-C (CYVE courses), c) treatment intensity can be adapted to stage and resection, but also to response to chemotherapy (at D7 and after 3 courses), d) early dose intensity is essential. The German BFM studies (Reiter Blood 1999, Woessman Blood 2005) told us that a) treatment intensity can be adapted to stage and resection, but also to LDH level, b) HDMTX is a very effective systemic drug, and exposition to the drug must be all the more long as the disease is more advanced. They also confirmed that HD Ara-C is important in the advanced diseases. Although stratification and weight of treatment are not exactly the same, results are very similar. Other studies also showed that treatment duration can be of short duration and confirmed that cyclophosphamide, HDMTX and Ara-C are the major drugs in BL, in addition to vincristine, doxorubicin, VP16, corticosteroids. CNS prophylaxis must be done by HDMTX +/- HD Ara-C and IT injections of MTX +/-Ara-C, but not by cranial irradiation. With all these current strategies in Western countries, EFS of BL increase to 80-90%, but immediate toxicity of the treatment needs adequate supportive care. CNS disease, especially in case of blasts in CSF, remains of poorer prognosis, with EFS <80%. Large B-cell lymphomas (DLBCL) represent 10 to 20% of B-cell lymphoma in children and occur more frequently in adolescents. Although relapses can occur later, their outcome is similar to that of BL with the LMB and BFM protocols, except for primary mediastinal large B-cell lymphoma (PMBL) which has a worse outcome and need a different therapeuticn strategy including rituximab.

The present question concerns the use of targeted therapy (rituximab) in childhood NHL. The “Inter-B-NHL ritux 2010” study, an international European-COG trial, is currently running to answer this question.

Lymphoblastic lymphoma (LL) are the second commonest LNH in children (25-30% of the cases): either T lineage (3/4 cases) mostly located in thymus region, or B lineage generally of (sub)cutaneous or bone location. They must be treated with a treatment similar to those designed for high risk leukemia, such as Wollner’s or nonB-BFM protocols (Reiter, Blood 2000). Regimens are based on semi-continuous intensive polychemotherapy followed by maintenance therapy for a total duration of 2 years. Numerous drugs are given (including corticosteroids, vincristine, anthracyclines, cyclophosphamide, cytarabine, and methotrexate), with an emphasis on asparaginase. EICNHL (European Intergroup for children Non Hodgkin’s Lymphoma) conducted the international Euro-Lb02 study based on nonB-BFM90 scheme without cranial irradiation which randomized dexa vs pred in induction and 18 vs 24 months treatment duration. Unfortunately the study had to stop because of toxic death rate higher than expected. Preliminary results showed an EFS rate of 81+2% for the 319 eligible patients (75% T-LL) with a median follow-up of 4.8 years.

Identifying prognostic factors is an important challenge in LL. Several biological characteristics have been described to have prognostic relevance in T-LL: MDD and MRD (Coustan-Smith JCO 2009), loss of heterozygosity (LOH) on chromosome 6q (Bonn Blood 2013), absence of NOTCH1/FBXW7 mutations (Callens JCO 2010). They have to be confirmed on prospective series before being used for treatment stratification.

Anaplastic large cell lymphoma is the third entity encountered in children (~10% of NHL in children). After a retrospective analysis of pooled data of several European groups, prognostic factors were identified (mediastinal or visceral involvement and skin lesions), allowing to design the randomised ALCL99 study based on the B-BFM90 scheme. The 2-year EFS and OS rates were 74.1% and 92.5% respectively (Brugieres JCO 2009, Le Deley, JCO 2010). Following this study patients are currently treated according to the ALCL 99 protocol with HDMTX=3g/m² in 3h infusion and without IT.

More recently, prognostic factors associated with an increased risk of failure have been identified in ALCL: histologic lymphohistiocytic and small cell variant patterns (Lamant JCO 2011), the detection of MDD by RT-PCR for NPM-ALK in blood or BM, the persistence of positive MRD after 4 weeks of treatment and low production of anti-ALK titers (Mussolin, Leukemia, 2013).

Several arguments are in favor of a major role of the immune system in ALK-positive ALC. Interestingly, Vinblastine is a potent immune modulator that enhances anticancer immune response by stimulating dendritic cell (DC) function.

Several new drugs have recently been implemented in the treatment of ALCL: Brentuximab Vedotin (BV, an anti-CD30 monoclonal antibody conjugated to the antimicrotubule cytotoxic monomethyl auristatin-E) is now approved by the Food and Drug Administration (FDA) and European Medical Agency (EMA) for the treatment of systemic ALCL after failure of at least one chemotherapy regimen in adults. ALK inhibitors are also promising drugs, especially Crizotinib. The COG is currently investigating the feasibility of combining BV or crizotinib with ALCL99 chemotherapy in children with newly-diagnosed stage II-IV ALCL whereas the EICNHL is planning a randomized trial with risk stratification based on MDD and antibody levels, to evaluate the efficacy of adding crizotinib to ALCL99 in front-line therapy for intermediate and high-risk ALCL and to compare weekly vinblastine to ALCL99 in low- and intermediate-risk disease.

Treatment of relapsed NHL

Due to the high cure rate obtained with an intensive front-line treatment, relapses are rare, but their treatment remains a challenge. It is generally admitted that HD therapy with stem cell rescue in 2nd RC is necessary. As for front-line, second line treatment will differ according to the subtype, as well as the conditioning regimen and the type of graft. Concerning the type of graft, the review of a large series of pts with NHL transplanted after relapse (T Gross, Bio Blood Marrow Transplant, 2010) indicated no EFS difference between auto vs allo in BL, DLBCL and ALCL patients, but a higher EFS in LL after allograft.

The French experience on B-cell lymphoma relapse was recently published (Jourdain, Haematologica 2015). 31/67 pts achieved CR after 2nd line CT and 41 were transplanted. The 5-y survival rate was 30%. In the multivariate analysis, adverse prognostic factors at relapse were: early relapse (within 6 months), multiple sites of recurrence, BL histology, and features at diagnosis (elevated LDH and advanced stage disease). Among the transplanted patients, those who obtained CR2 had a better survival. In this study, allograft did not do better than autograft and the impact of rituximab could not be assessed.

In LL, the outcome of relapse is also poor. In the BFM series (Bukhardt, JCO 2009), the OS was 14%. In the French LMT 89 study, only the patients who relapsed after 30 months could be salvaged.

The major problem is to achieve a 2nd remission in relapsed poor risk patients with B-cell NHL and LL New therapeutic approach are needed and will have to be investigated in the future (BTK inhibitors, bi specific antibodies for exemple) Contrarily to what is observed in BL and LL, relapse in ALCL can be salvaged, sometimes after several relapses. In France, the benefit of Vinblastine, even after HDCT, has been shown (Brugieres, JCO 2009): the OS of a series of 36 refractory/relapsed pts treated with vinblastine was 65%. The question was the optimal duration of this treatment. Analysis of relapses occurring after ALCL99 study, either during the study, or within the prospective EICNHL relapse study, indicated that consolidation with

allograft benefited to early relapses, while weekly vinblastine benefited to later relapses, with survivals around 60-70%. (Woessman JCO 2011, BHJ 2012 abstract).

In conclusion: Currently the majority of the children and adolescents with LNH are cured of their disease, with treatment validated in prospective national/international trials. But these treatments need adequate supportive care which might not be available in all countries.

Further improvements will need international collaboration to further develop biological studies and define the place of targeted therapies.

Relapses are rare, but are difficult to salvage in BL and LL, contrarily to ALCL. In the latest, relapses can be cured even after several relapses and vinblastine has showed its benefit. In all subtypes, time to relapse is prognostic. The place of innovative therapies has to be investigated.

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How I Treat Childhood HD? Newly diagnosed and relapsed patients

Odile Oberlin

Abstract

Hodgkin lymphoma (HL) accounts for ~ 10 % of all childhood cancers. Five-year survival rates with modern therapies are now approaching > 90-95% as a consequence of its significant sensitivity to both chemotherapy and radiation. However, the risks of late effects associated with radiation and chemotherapy, including infertility, second cancers and cardiac deaths, have become more widely recognized, and treatment decisions are increasingly based on minimizing late-effects risk and late mortality1. The current challenge is to determine how much therapy is needed to ensure survival and how to tailor treatment to the individual to prevent these long term toxicities.

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Improving access to cancer management for children in middle east

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Abstract

Increasing incidence of childhood cancer and reduction of mortality and morbidity of malignant disease achieved in the world .Recent findings showed approximately 200 000 children and adolescents are diagnosed with cancer every year worldwide; of those, 80% live in low middle income countries, which account for 90% of the deaths. We have stepwise process in the development of pediatric cancer programs at least four levels.Some major barrier s influence on treatment outcome in middle east that included:
1. Geogrphic inequality in cancer treatment 2.Non Communicable Diseases (NCD) and Chronic Childhood Disorders are not among the priorities of top 8 health organisations 3.Substantial Barries to access to cancer control: Prevention , Early Detection , Diagnosis/Treatment , Palliation 4.Substantial Quality of cancer control : Economical/Political barriers , Shortage of healthcare workers , Limited access to cancer drugs ,Cultural stigma from cancer 5.Lack of data and cancer registeries (National and Hospital) .
However, available information showing variations in incidence may indicate unique interactions between environmental and genetic factors that could provide clues to cause.
Outcome of children with cancer in Middle East is dictated by presentation and under diagnosis, high abandonment rates, high prevalence of malnutrition and infections, suboptimal supportive and palliative care, and limited access to curative therapies.
MAHAK as charity is comprehensive pediatric treatment and research center. MAHAK preserve civilization and ensures that children with cancer are never refused treatment due to poor financial status. MAHAK involved in cancer care should assume the responsibility to lead the way and be convinced that serious global initiatives will represent the breakthrough in cancer treatment and control for the next years. A global cancer initiative should be recognized as part of the mission of the institution, shared by all cancer programs, and integrated into their strategic planning.
Keywords: Pediatric cancer, outcome, Middle East, MAHAK

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Prevention Program Thalassemia in Iran

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Abstract

Introduction: Major thalassemia is an inherited disease most prevalent in the region of the world known as the thalassemia belt where many countries are located including Iran whose thalassemia prevention program started in 1995. Many different researchers have studied about the success of this program with no unanimous findings.

Materials and methods: A cross-sectional analytical study was conducted at the Iranian Blood Transfusion Organization

A questionnaire was forwarded to all blood centers of the IBTO so as to obtain information about the new cases of thalassemia and the causes of these thalassemic births. Provincial thalassemia societies also received the questionnaires so that screening and prenatal diagnosis (PND) errors would be recorded. The results showed that the fall in affected thalassemia births was estimated to be 82.28% in 2009. In addition, all articles about success in the plan both in english and in Farsi languages were evaluated and compared with obtained data.

Results: The findings show that the program has been successful in the significant reduction of the new thalassemia births though not significant in a few provinces like Sistan & Baluchestan. The role of the network of genetic labs has been also indispensable in the reduction of the new births. However, there is ambiguity over the impact of the program on the attitude and awareness of people across the country about the prevention of inherited diseases.

Conclusions: Despite the success of the Iran thalassemia prevention program, it needs to be modified to be more compatible with the relevant social textures of different provinces including Sistan& Baluchestan where more public awareness raising attempts should be made.

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Immune modulation in ITP

Paul Imbach

Abstract

In 1980 a child with refractory immune thrombocytopenia (ITP), bleeding and secondary hypogammaglobulinaemia due to long term immunosuppressive treatment we administered intravenous immunoglobulin IVIG). His platelet counts dramatically increased. In 13 consecutive children with ITP, but without hypogammaglobulinaemia, similar rapid platelet increases after administration of always the same dose of IVIG were observed. A controlled, randomized multicentre study of corticosteroid versus IVIG treatment confirmed the new therapeutic phenomenon of IVIG. This observation and its mechanisms of actions evoked huge clinical and laboratory research on immunomodulatory effects of IVIG and other immunomodulators worldwide. This is IVIG :Intravenous immunoglobulin is extracted from pooled plasma from 10,000 to 60,000 human blood or plasma donations.The final product of antibody concentrate contains several millions of antibody specificities, mainly natural antibodies and anti-idiotypic antibodies. Natural history of ITP: The clinical manifestion, the severity of bleeding and of platelet count and the natural history of ITP are heterogenous. The patients and the parents are fearful of bleeding and tyrannized by low platelet counts. The outcome of individual patients can not be predicted, although the majority of children have spontaneous resolvement or improvement of ITP. In four prospective Intercontinental cooperative ITP registry-studies (ICIS, see www.itpbasel.ch) of nearly 9000 children and adults with newly diagnosed ITP and follow up of 1-3 years new aspects have been found, i.e. 2/3 of them had initially platelet counts below 20x109/l. The rate of severe bleeding, especially the rate of intracranial bleeding, was low. In the follow up: 30% of the children in ICIS registry I had persistent ITP at 6 month after diagnosis, from which 25,3 % recovered between 6 to 12 months. Therefore, the term chronic ITP has been postponed to ITP duration longer than 12 months. Management: Observation only, if no or mild cutaneaous bleedings are present (in about 1/3 of patients) and a patient is not hyperactive. Standards treatment: - IVIG: 0.4 – 0.8 g/kg bodyweight (b.w.) once - Anti-D immunoglobulin: 50 – 75 microg/kg bw once - Corticosteroids: 4 mg/kg b,w. daily during 2-4 days, then tapering during 3 days - Thrombopoietin agonists: limited available today Emergency treatment: In severe life-threatening bleeding: - Start with Corticosteroids 30 mg/kg bw or dexamethason 1-2 mg/kg bw - Followed by IVIG: 0,8-1,0 g/kg b.w. per dose - As third: Platelet transfusion Refractory ITP: - Thrombopoietin agonists: limited available today - Suppression of B-cells: Anti CD 20-monoclonal antibodies - Suppression of T-cells: cyclosporine A: 2-5 mg/kg b.w. per day, tacrolimus - Vincristine iv: 1,5mg (max. 2 mg)/m2 weekly x 4-6 plus 2 mg corticosteroid/kg b.w. daily x3 per week - Consider also: Interferon, mycophenolate mofetil, high dose corticocsteroids or dexamethasone - Splenectomy - Classic options (individual indications): Azathioprime, cyclophosphamide, other cytostatics Clincial research: As a result of the heterogeneity of ITP and various controversies, current practice (guidelines etc.) does not always follow the various recommendations. Prospective studies are needed. Immunomodulation by IVIG Checking the 8‘215 classified items „IVIG and mechanisms of action“ listed in Pubmed (date of check: 06.08.2015) we recognize synergistic changes between the imbalanced components of the innate and adaptive immune system of ITP are described. Today, beside IVIG other immunomodulators (i.e.cyclosporin, anti-CD20 antibodies and other monoclonal antibodies) revealed similar immunomodulatory effects in ITP and in other autoimmune disorders, such as: - Immune complex formation, activation of dendritic cells, their Fc receptors and their molecules Clinical examples : Children with para- or postinfectious conditions, such as with newly diagnosed ITP, Guillain-Barre´ syndrome or

Kawasaki syndrome. - Anti-idiotypic antibodies neutralization and downregulation of B-cells Clinical examle: A patient with aquired FVIII autoantibody and severe bleeding successfully was treated with IVIG in our hospital. Kazachkine and his group in Paris could demonstrate anti-idiotypic antibodies in IVIG against FVIII which may neutralize the specific autoantibody in patient with acquired FVIII disease. The group could also detect other anti-idiotypic antibodies in IVIG (against thyroglobulin, DNA, peripheral nerve, acetylcholine receptor, endothelial cells and others. Later on, Berchtold et al could demonstrate specific antiidiotypic antibodies in IVIG against ITP. - Antiplatelet T-cell reactivity, suppression of T cells, increase of T regulatory cells and cytokine release Clinical example: After hematopoietic stem cell transplantation IVIG protects against graft-versus-host disease (GVHD) by targeting donor allo-reactive T-cells- beside the regulation, maturation and function of dendritic cells. - Blockade of Fc receptors on effector functions monocytes/macrophages, complement binding/clearing, arrest of apoptosis and Fas inhibition by IVIG Clinical examples: In neurologic autoimmune diseases such as chronic inflammatory demyelinating polyradiculoneuropathy CIDP, multifocal motor neuropathy. In dermatologic autoimmune mucocutaneous blistering diseases, IVIG administration showed progression arrest by complete FcRn dependence and direct effects on keratinocytes. In pemphigus after IVIG alone or in combination with monoclonal anti-CD20 antibodies(rituximab). In patients with Stevens-Johnson syndrome, arrest of apoptotic keratinocyte death resulting from anti-Fas antibodies in IVIG has been shown. . As in ITP the heterogeneity of autoimmune disorders are based on variations from patient to patient, the age, the disease stage, the duration of the disease and from other factors. Therefore, not many controlled studies exist and the off-label uses of IVIG remain frequent. Longterm, prospective registries on rare autoimmune diseases are necessary for finding subgroups of a disease - followed by targeted clinical studies on subgroups.

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How I treat childhood abdominal thrombotic?

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Abstract

The thrombosis in the major vessels of abdomen causes a wide spectrum of clinical pictures ranging from a totally asymptomatic patient to a patient with acute abdominal pain and even impending liver failure in patients with underlying chronic liver disease. The resulting portal vein thrombosis (PVT), splenic vein thrombosis (SVT) and mesenteric vein thrombosis (MVT) have a variety of consequences ranging from acute abdomen to chronic hepatomegaly and even totally asymptomatic patient in whom the only finding is pancytopenia. PVT is a relatively common complication in patients with liver cirrhosis, but might also occur in absence of an overt liver disease. Clinical examination, laboratory investigations, and imaging are helpful to provide a quick diagnosis, as prompt treatment might greatly affect a patient's outcome. The goal of treatment is similar in acute and chronic PVT, and consists in correction of causal factors, prevention of thrombosis extension, and achievement of portal vein patency.. MVT is an acute thrombotic disorder of vessels before formation of main portal vein. It may be associated with nausea, vomiting, increased bowel movements and sometimes bloody diarrhea. After diagnosis the treatment must be initiated with anti-coagulation unless the patient has peritoneal irritation findings in which surgery is indicated. In conclusion consequences of thrombophilia like PVT, SVT and MVT are potentially treatable if only diagnosed early and prompt treatments are initiated. Although anticoagulant treatment is mandatory, some limitations and contraindications may prevent their use. This group of patients must be followed with a consulting surgeon who has experience in this field to decide the best timing for a shunt surgery and for possible acute operation indications.

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Blood Transfusion indication & side effects

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Abstract

Introduction: Haemovigilance is a system with standard program to cover the entire transfusion chain, monitor, evaluate and analyse the data to improve patients' safety. We report the implemented haemovigilance system in hospitals and transfusion reactions (TR) in Iran. Methods and Materials: This was a prospective descriptive study. The national reporting system for transfusion incidents was introduced in January 2009. In the period 2009- 2014, 247 hospitals in a mandatory manner reported transfusion incidents among patients to the blood bank in a mandatory manner. All incidents were anonymously recorded in a standardized report form and registered in 10 categories. Results: A total of 3377 transfusion incidents were reported and categorized as: incorrect blood component transfused (4%), Febrile non hemolytic transfusion reaction (FNHTR) (44%), Immune hemolytic transfusion reactions (12%), nonimmune hemolytic transfusion reactions (7%), allergic reactions (32%), transfusion-related acute lung injury (1%). There were no reports in the categories of infections, post-transfusion purpura, transfusion-acquired viral infection, and transfusion-related graft versus host disease. Conclusion: Haemovigilance is a tool for quality improvement and better surveillance on patient safety. The safety and quality of blood transfusions can be improved if we follow the transfusion chain, prevent and treat transfusion reactions and report adverse reactions to change the protocols.

Key Words: Adverse reaction, Blood Safety, Haemovgilance, Risk

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Educational session: Hgb Electrophoresis

Dr Alireza Korourian[1], Shariat[2]

Abstract

Depends on the primary clinical suspicion many different laboratory tests may be ordered , as simple as a complete blood count to more complex as hemoglobin electrophoresis .judicious use of them help to make a correct diagnosis. Some different methods are available for Separation of different hemoglobin constitution in red blood cells of a patient. In 50's and 60's gel electrophoresis by agar or acetate cellulose in alkaline or acidic PH was the routine. In 70's polyacrylamide and isoelectric focusing was introduced but were not routine in lab duo to labor intensive of procedures. In late 80's and early 90's HPLC was adopted by laboratory as a routine workup for separation of hemoglobin. And last and more accurate technique which introduced in late 90's and early 2000's was capillary electrophoresis. We will discuss our experience in use of this method for 6 years and some details of laboratory technique in different methods of hemoglobin separation as effect of PH , time , voltage , buffers and …..in quality of technique

ARTICLES OF SPEAKERS

IJBC 2015; Supplementary; P 21
Paper ID: 13

Lock therapy in children patients with hematologic malignancies

Prof Soheila Zareifar[1], Prof Mohamad Reza Bordbar[2], Prof Mahdi Shahryari[1] ,Prof Mehran Karimi[3]

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2. assistant professor
3. professor

Abstract

Long-term central venous catheters (CVCs) are essential for modern pediatric practice in oncology wards especially in leukemic patients with long term treatment period. These devices terminate in a large central vein, usually the superior vena cava, and are used for administration of chemotherapy drugs, fluids and blood products; for blood collection and for initiation of chemotherapy. The most common, serious complication of CVC use is central line–associated bloodstream infection (CLABSI). Catheter lock therapy (CLT) may be used in addition to systemic therapy with the aim of reducing bacterial infections. A small dose of antimicrobial agent is instilled to fill the catheter lumen (usually ≤1.5 mL) and is allowed to dwell for an extended period of time (usually hours to days).We prepaired a evidence base guideline for lock therapy antibiotic regimen according to routine antibioticin Iran in Amir oncology hospital of Shiraz for pediatric oncologic patients.

Keywords: lock therapy, central venous catheter, leukemia

IJBC 2015; Supplementary; P 22
Paper ID: 17

Prevention of Persistent aplasia after chemotherapy for acute myeloid leukaemia. Is need chemotherapy titration during treatment ?

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1. assistant professor
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3. associate profesor

Abstract

Persistent aplasia is a rare complication with poor prognosis after intensive chemotherapy for acute leukaemia. We present a 16-year-old boy with acute myeloid leukaemia(AML), was treated after compelet MRC12 protocol chemotherapy induced persistent aplasia and he was candidated for allogeneic peripheral blood stem cell transplantation (PBSCT) from a matched related donor . This phenomenon define that the patient need to chemotherapy titration until stem cell transplantation. The patient unfortunately dead by pulmunary Aspergilosis. Although the incidence and pathogenesis of chemotherapy- induced persistent aplasia are still unknown, but in patient poses matched related donor the physician should be titrated chemotherapy base on minimal residual disease (MRD) markers.

Keywords: persistent aplasia, AML

IJBC 2015; Supplementary; P 23-24
Paper ID: 18

Different phenotypes with Identical Genotype in Thalassemia Syndrome

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Abstract

Thalassemia is a hereditary anemia characterized by reduced or absent beta and alpha globin chains synthesis in beta thalassemia and alpha thalassemia respectively. The resulting relative excess of unbound the other globin chains precipitate in erythroid precursors in the bone marrow, leading to their premature death and, hence, to ineffective erythropoiesis. Thalassemia phenotypes are variable, ranging from mild to severe anemia. Some Patients come to medical attention during early life and some individual present later in life or find incidentally in marital time. Alpha and beta thalassemia are very heterogeneous at the molecular level. In most cases, mutations are single nucleotide substitutions, deletions or insertions of single nucleotides or small oligonucleotides leading to frame shift. In alpha thalassemia most are deletional but some are non deletional. The diversity of phenotype are some related to genotype, coinheritance of alpha deletion or triple alpha and the presence or absence of specific polymorphism such as Xmn-1.However much of the phenotypic variability is explained by these enhancing and/or modifying determinants .But the cause of most phenotypic variability is unknown .Here we present some cases of thalassemia syndrome as a sample with identical genotype and different phenotypes. 1. Beta Thalassemia Minor: Genotype: IVSII-I, α α/ α α, Xmn-1:-/- Case 1: 16 –yr-old male, Hb: 9, RBC: 5.2, MCV: 56, MCH: 20, Ferritin: 55, Hb A: 92, Hb A2:5.2, Hb F: 2.8 Case 2: 18 –yr-old male, Hb: 11.5, RBC: 5.2, MCV: 61, MCH: 21, Ferritin: 75, Hb A: 94, Hb A2:4.2, Hb F: 1.8 2. Alpha Thalassemia Minor: Genotype: --Med/α α,β/ β, Xmn-1:-/- Case 1: 12 –yr-old female, Hb: 8, RBC: 4.8, MCV: 54, MCH: 18, Ferritin: 105, Hb A: 96.2, Hb A2:2.8, Hb F: 1 Case 2: 14 –yr-old female, Hb: 11.5, RBC: 5.1, MCV: 58, MCH: 20, Ferritin: 55, Hb A: 97.2, Hb A2:2, Hb F: 0.8 3. Beta Thalassemia Intermediate: Genotype: IVSII-I, α α α / α α , Xmn-1:+/- Case 1: 22 –yr-old female, Hb: 8, RBC: 5.2, MCV: 66, MCH: 22, Ferritin: 55, Hb A: 92, Hb A2:4.1, Hb F: 2.9 Case 2: 18 –yr-old male, Hb: 11.5, RBC: 5.2, MCV: 61, MCH: 21, Ferritin: 75, Hb A: 91.2, Hb A2:5, Hb F: 3.8 4. Beta Thalassemia Intermediate: Genotype: IVSI-6/IVSI-6, α α / α α , Xmn-1:-/- Case 1: 36 –yr-old male, Hb: 6, RBC: 5.2, MCV: 75, MCH: 22, Ferritin: 350, Hb A: 89, Hb A2:4.2, Hb F: 6.8, Splenectomy, Billirubin total: 9, Direct: 0, 5, Occasional Transfusion Case 2: 32 –yr-old Female, Hb: 10, RBC: 4.9, MCV: 65, MCH: 20, Ferritin: 120, Hb A: 90, Hb A2:4.2, Hb F: 5.8, Splenectomy, Billirubin total: 4, Direct: 0, 5, No Transfusion 5. Beta Thalassemia Major / Intermediate: Genotype: IVSII-I/IVSII-I, α α / α α , Xmn-1:-/- Case 1: 12 –yr-old male, Pre-transfusion Hb: 9, RBC: 5.2, MCV: 55, MCH: 20, Ferritin: 2000, Hb A: 0, Hb A2:4.2, Hb F: 95.8, Spleen: 5 cm.Liver:2cm, Regular Transfusion Case 2: 12 –yr-old male, Hb: 11.5, RBC: 5.2, MCV: 65, MCH: 20, Ferritin: 120, Hb A: 0, Hb A2:4, Hb F: 96, Spleen:-, Liver:-, Bilirubin total: 2, Direct: 0, 5, No Transfusion 6. Hb H Disease Genotype: --Med/-α3.7,β/ β, Xmn-1:-/- Case 1: 38 –yr-old female, Hb: 8, RBC: 4.8, MCV: 56, MCH: 20, Ferritin: 1500, Hb H: 8, Hb A: 90, Hb A2:1.8, Hb F: 0.2, Occasional Transfusion Case 2: 25 –yr-old female, Hb: 11, RBC: 4.9, MCV: 52, MCH: 20, Ferritin: 250, Hb H: 5, Hb A: 93, Hb A2:1.5, Hb F: 0.5, No Transfusion 7. Hb H Disease Genotype: Poly A6/Poly A6,β/ β, Xmn-1:-/- Case 1: 2–yr-old male, Hb: 6, RBC: 4.5, MCV: 50, MCH: 18, Ferritin: 200, Hb H: 2.4, Hb Bart: 1.6, Hb A: 93.5, Hb A2:1.5, Hb F: 1, Regular Transfusion Case 2: 38 –yr-old male, Hb: 10.5,

RBC: 4.9, MCV: 52, MCH: 20, Ferritin: 250, Hb H: 4.5, Hb A: 93, Hb A2:2, Hb F: 0.5, No Transfusion There is also phenotypic discrepancy in hemoglobinopathies such as sickle cell disease and Hb E/Thalassemia that needs to be described in another manuscript.

Keywords: Genotype, Phenotype, Alpha Thalassemia, Beta Thalassemia

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Paper ID: 30

Report of 20 years' Experience on the effect of intensive intrathecal chemotherapy on prognosis of childhood lymphoblastic leukemia with central nervous system (CNS) involvement.

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Abstract

Introduction: Primary CNS involvement and CNS relapse are poor prognostic events in acute leukemia. Due to sever skeletal and endocrine complications of craniospinal radiotherapy, only cranial radiotherapy is advisable; but only 15% of the cases of CNS relapse may remain in remission and second CNS relapse or bone marrow (BM) relapse is usual. So prevention from CNS relapse is very important way to decrease both mortality and morbidity in childhood leukemia. Method: In a Prospective study from June1995 to May 2014; thirty children with CNS involvement and 60 children with CNS relapse of Acute Lymphoblastic Leukemia (ALL) were enrolled in the study with written consent form. They randomly were divided in two groups: 30 cases in group A received triple intrathecal (IT) injections every 2 months as in high risk ALL protocols for three years (including A1: 15 cases of primary CNS involvement; and A2: 15 cases of CNS relapse).Sixty cases in group B received further triple IT injections in the fourth and fifth years (for 2 years after discontinuation of maintenance chemotherapy) including B1: 20 cases of primary CNS involvement and 40 cases of CNS relapses. Each case of group A had two age and sex matched patients in group B. they had 2-15 years follow up. Results: in group A1 5/15 CNS relapses, 3/15 BM relapses and 2/15 death were occurred. Boys had more relapses and deaths than girls (P value

Keywords: Childhood Leukemia, CNS involvement, CNS relapse, CNS prophylaxis, Prognosis.

IJBC 2015; Supplementary; P 26
Paper ID: 34

Retinoblastoma; in a retrospective study of 43 cases of Southwest Iran

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Abstract

Abstract Background Retinoblastoma is the most common embryonic tumor of retina of children under 5-year-old. This tumor include 3-4% of all children malignancies during the age range 0-14 years. In this study, we reported the incidence, therapeutic protocols, and treatment outcome of children with RB in South west Iran. Patients and methods We retrospectively investigated the medical reports clinical and pathological features of RB cases at referral Shafa Hospital of Ahvaz city located in Southwest Iran, between 1994 and 2014. Result Our results showed leukocoria, white discoloration in the pupil, as the most common primary symptom in retinoblastoma patients. Forty-two percent of our patients had bilateral neoplasms. Based on histological investigations, stage IV was most common stage in our patients (79.1%). Conclusion In conclusion, our study showed that leukocoria, stage IV and Choroid involvement as most common symptom, stage and organ involvement, respectively and inherited cases is rare in our population. The results of our study can be as a base for further researches in future.

Keywords: Retinoblastoma,bilateral,leukocoria

IJBC 2015; Supplementary; P 27
Paper ID: 41

Thalassemia intermediate challenges in treatments

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Abstract

Thalassemia Intermediate is clinical definitions. descritive term includes β-thalassemia with a broad spectrum of clinical phenotype between the 2 extremis of asymptomatic thalassemia trait to transfusion depended thalassemia major from mild anemia (Hb9-10gram/dl) without splenomegaly to sever anemia(Hb 6grams /dl)just capable of surviving with blood transfusion ?sever thalassemia intermediate or thalassemia major Diagnosis requires careful observation over a period of time .

Hb levels and identification of herozygos β/globins gene modifiers are arbitrary guidelines as to possible clinical phenotype Patients with Hb bacterial immunization are necessary before Splenectomy Splenectomy is not a treatment of choice Splenectomy increased, coagulable state, secondary right heart disease high risk of the over whelming infections β/thalassemia intermediate due to highly variability of clinical picture and changing phenotype attending physician is in a dilemma to treat or not treat HLA matched sibling donor stem cell s transplantation from bone marrow and umbilical cord are the treatment of choice otherwise medical therapy in patients with Hb level .:Hb

Keywords: intermediate,Thalassemia

IJBC 2015; Supplementary; P 28
Paper ID: 50

The Effect and Side Effect of Hydroxyurea Therapy on Patients With b-Thalassemia: A Systematic Review to December 2012

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Abstract

Hydroxyurea (HU) is being used for patients with transfusion-dependent b-thalassemia major (b-TM) as well as non transfusion-dependent b-TM. As controversy exists regarding efficacy and safety of HU, we searched the published literature on efficacy, effectiveness and toxicity of HU in patients with b-TM. The research sources we used were: Medline, SID, PubMed, Scopus, Request, Web of Knowledge, Springer, Ovid, Cochrane searched up to October 2012. Using search terms sensitive to studies of clinical trials combined with searches on terms related to thalassemia and HU. We selected studies on randomized trials, quasi experimental trials (before and after design), case reports (with 1–5 cases), side effect studies in patients with b-TM, studies related to the mechanism of action and toxicity when used in patients with other hemoglobinopathies. We researched studies in English and Persian. Eligible articles were reviewed by two independent reviewers. Patient's characteristics, duration of trial, outcome and side effects were extracted. The main outcomes were synthesized under a random-effects model. Heterogeneity was assessed using the Q statistic, Tau2 and I2. Subgroup analyses were performed and the statistics data (STATA) software used. More than 500 articles were reviewed. No randomized clinical trial was found. Seventeen trials with before and after designs were found, 16 case reports (1–5 cases), 19 articles for mechanism of action and 16 studies for side effects were published from 1969 to October 2012. Hemoglobin levels after treatment showed modest but significant increase in non transfusion-dependent b-TM (p50.0001) and in transfusion-dependent b-TM (p50.0001).

Keywords: Antimetabolite, b-thalassemia (b-thal), genotype, hemoglobin (Hb), Hb F inducer, hydroxyurea (HU), side effect

IJBC 2015; Supplementary; P 29
Paper ID: 67

Assessment of modern radiation treatments for patients with Hodgkin's lymphoma

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Abstract

Introduction: Hodgkin lymphoma (HL) is a curable malignancy that classified in two histological types (WHO 2008): the nodular lymphocyte predominant and the "classic" HL. The HL malignancy appears commonly in cervical, supraclavicular and mediastinal lymph nodes, while sub-diaphragmatic presentations and bone marrow and hepatic involvement are less common. For patients with early-stage HL, Primary ABVD combination chemotherapy, followed by involved-field irradiation (IF-RT) is the standard treatment. Radiation treatments could be optimized by reducing doses, reducing the radiation field size using involved-node radiotherapy (INRT), delivery of radiation with modern radiation techniques, such as intensity-modulated radiotherapy (IMRT) or breathing-adapted irradiation. In this study, the potential benefits of modern radiation treatments for Hodgkin's lymphoma were investigated. Material and Methods: A computerized search using the databases SID, Google Scholars, Pub med and Science Direct, covering the period from 1998 to 2015, was conducted using the following key search terms: "Hodgkin's lymphoma", "Combined modality therapy" and "Radiotherapy". In total, 38 relevant papers were reviewed. Results: In Filippi AR et al. study, a total of 49 and 41 patients were irradiated by 3D Conformal Radiotherapy (3D-CRT) and image-guided intensity modulated radiotherapy (IG-IMRT), respectively. No differences in relapse-free survival (RFS) were observed between the 2 groups. For 3D-CRT and IG-IMRT patients, three-year RFS was 98.7% and 100%, respectively. Grade 2 toxicity events (mainly mucositis) were observed in 32.7% of 3D-CRT patients and in 9.8% of IG-IMRT patients. In clinical investigation of the deep-inspiration breath-hold technique, there were no significant differences in PTV coverage between the two techniques (free-breathing (FB) vs. deep-inspiration breath-hold (DIBH)). The mean doses delivered to the coronary arteries, heart, and lungs were significantly reduced by 15% to 20% using DIBH compared with FB method, and the lung V20 was reduced by almost one third. Conclusion: Many of studies demonstrated that combined modalities with the employment of new RT techniques are the standard therapeutic approaches for patients with Hodgkin's lymphoma. In therapy strategies, late effects, cardiac toxicity and probable second malignancy risk should be considered.

Keywords: Hodgkin's lymphoma, Combined modality therapy, Deep-inspiration breath-hold, Radiotherapy

IJBC 2015; Supplementary; P 30
Paper ID: 69

Weekly Recombinant Human Erythropoietin in Treatment post-chemotherapy Anemia in Children with a Solid tumor

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Abstract

Background: Generally, Recombinant human erythropoietin (rHuEPO) is used subcutaneously, 3 times a week, for increase hemoglobin levels and decrease transfusion requirements. This study aims to investigate how blood transfusion influences Hemoglobin levels in patients receiving once weekly rHuEPO for 12 weeks.

Materials and Methods: This was a case-control study of 28 patients less than 15 years with anemia and a solid tumor between April 2014 and July 2014. Median age of the patients was 7.32±0.67 years (range, 3-14.5 years). The patients were randomly assigned in two groups of rHuEPO receiving group and control group. 14 Patients in rHuEPO group received 450 IU/kg/dose rHuEPO subcutaneously, once weekly, for 12 weeks. The number of patients received transfusion during the treatment period was compared in the preceding 12 weeks. Also, adverse events (AE) were recorded at the 4th, 8th, and 12th weeks.

Results: Mean hemoglobin levels, before and after study, in rHuEPO group were 7.99±1.32 g/dl and 10.30±0.67 g/dl, respectively ($p<0.05$).

Conclusions: Results revealed that the rHuEPO (450 IU/kg/day, once a week) was effective in increasing hemoglobin levels as well as decreasing blood transfusion requirements in children with anemia following intensive chemotherapy.

Keywords: Anemia, Recombinant Human Erythropoietin, Chemotherapy, Solid tumor

IJBC 2015; Supplementary; P 31
Paper ID: 73

The Correlation between Heart, Liver and Pancreas Hemosiderosis Measured by MRI T2* in Thalassemic Patients from IRAN

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Abstract

Background: Major thalassemic patients need lifelong transfusions. The consequence of these repeated transfusions is iron accumulation in different organs. The main aim of the present study was to investigate the correlation between heart, liver and pancreas hemosiderosis among thalassemic patients in Iran. Patients and Methods: This cross sectional study was conducted at Zafar Adult Thalassemia Center, a referral thalassemia center in Tehran, Iran, from May to November 2014 on 164 major thalassemia patients. All patients were on regular blood transfusion at 2-4 weeks intervals to keep their hemoglobin at a level of 7-9 gr/dl before each transfusion. Demographic data were gathered from patients' history. MRI T2*of liver, heart and pancreas was performed for all patients. **Results:** There was a moderate correlation between pancreatic T2* and cardiac T2* relaxation times ($r = 0.42$, $p < 0.001$), and a weak to moderate correlation between T2* of pancreas and liver ($r = 0.41$, $p< 0.001$). **Conclusion:** Poor correlation between liver and heart, as well as a weak to moderate correlation between pancreas and liver T2* relaxation times indicate that relying on liver MRI T2* to predict the exact condition of pancreas or heart iron overload might not be a reliable approach. Our findings suggest the advantage of using the pancreas and heart MRI T2* as a non invasive method for estimation of iron overload instead of relying on liver MRI T2*.

Keywords: Thalassemia, hemosiderosis, MRI T2*, Iran.

IJBC 2015; Supplementary; P 32
Paper ID: 74

Testicular relapse in childhood acute lymphoblastic leukemia treated with UK-ALL-X protocol

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Abstract

Background: relapse of ALL as medullary or extra medullary in childhood is about 15-20% .but testicular relapse in boys with ALL is rare and about 6% of relapses

Purpose: this descriptive study was designed to assess the incidence of testicular relapse in children treated with UK-ALL-X protocol

Materials and methods: data was obtained from 211 boys with ALL were analyzed . after complete remission of induction and receiving intensification with UK-ALL-X protocol careful examination and follow up on maintenance and after that for relapse of medullary or extra medullary were performed

Results: testicular relapse was documented in one child in total 42 boys who had relapse during maintenance or after complete treatment ,that indicate low incidence of testicular relapse in children treated with UK-ALL-X protocol

Conclusion: testicular relapse which depends on the therapy administered is very low in UK-ALL-X protocol which may be so as administration of intensification with etoposide which is not in other international protocols

Keywords: acute lymphoblastic leukemia. UK-ALL-X protocol testicular relapse ,etoposide

IJBC 2015; Supplementary; P 33-34
Paper ID: 76

A Comparision of Cost-effectiveness Analysis Between Biogeneric Recombinant Activated Factor Vll (Aryoseven™) and Activated Prothrombin Complex Concentrates (FEIBA™) in the treatment of hemophilia A patients with inhibitors in Iran

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7. Pediatric Congenital Hematologic Disorders Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran

Abstract

Background: The current clinical trial aimed to evaluate the costeffectiveness of AryoSeven® -activated factor VII (rFVIIa)- and FEIBA -activated prothrombin complex concentrates (aPCC)- using the Decision Analytic Model from the perspective of the Iranian healthcare system.

Methods: An open label, multi-center, cross-over clinical trial and a Decision Analytic Model were designed. Both of the strategies were used in a random order for two separate joint bleedings in each patient. Patients were categorized into 3 groups based on their prior tendency to each one (group A to AryoSeven® and F for FEIBA) or none of the products (group O). Information regarding the efficiency of medicines and cost of treatment was transferred to the designed model. To determine the premium therapeutic strategy, the Incremental cost-effectiveness ratio (ICER) was also calculated.

Results: Clinical and cost information on 20 patients with 40 bleeding episodes were extracted. Protocol F led to more treatment success in group F than other groups (P= 0.03), however, there was no significant difference in this success between groups on protocol A (P= 0. 53). Also, there was a significant statistical difference between the mean of effectiveness scores in the groups using protocol F (P = 0.01). The effectiveness of protocol F and A was 89% and 72% respectively. The incremental cost-effectiveness ratio analysis (ICER) was US$ 5,146 for managing of one episode of bleeding to get one more unit of effectiveness with using FEIBA vs. AryoSeven in the first line of treatment.

Conclusions: Although the results showed that AryoSeven was more cost-effective compared to FEIBA, but these two strategies are undominated. In other words, both medicines can be applied in the first line

of the treatment if the cost of FEIBA was reduced.

Keywords: Cost-Effectiveness, Hemophilia A, Inhibitor, AryoSeven, FEIBA

IJBC 2015; Supplementary; P 35
Paper ID: 77

A prospective crossover triple blinded trial on safety and efficacy of Iranian rFVIII (Safacto®) versus plasma derived (pilot study)

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Abstract

Background: According to the growth of hemophilic population and infrastructure requirements for a comprehensive approach ,development of recombinant factor has become a milestone. The objective of this study was to assess the safety, efficacy and non inferiority of Safacto (Recombinant factor VIII) in comparison to plasma-derived factor in the treatment of hemophilia A.
Study Design: 10 severe hemophilic A subjects were entered to the study, after initiation of each of 4 consecutive hemarthrosis episodes in each patient was treated by an infusion (40-50 IU/kg) of either plasma derived or recombinant factor VIII in a Triple blinded prospective crossover permuted block randomizing method Clinical efficacy scale score and in vivo recovery of factor VIII was assessed in each of the treated bleeding episode any adverse events were also recorded
Results and Conclusion: Median ± SD level of factor VIII in plasma versus Recombinant was 111.5±39,115±39 respectively, without statistically significant difference. Response scaling method which assessed pain and range of motion, revealed equalized scores along with in vivo recovery ,hence treatment success rate was comparable in both groups. one nonrecurring, mild skin rash reaction occurred coincidence with administration of plasma derived factor . according to the achieved results Safacto (r-FVIII) found to be safe and effective and noninferior in the treatment of hemophilia A related bleeding .
Keywords: hemophilia A ,plasma derived factor, recombinant factor,Safacto

IJBC 2015; Supplementary; P 36
Paper ID: 84

The effect of hepcidin polymorphism C-582 A>G on ferritin level and iron overload in thalassemic patients, Shahr-e-kord, Iran

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Abstract

The effect of hepcidin polymorphism C-582 A>G on ferritin level and iron overload in thalassemic patients, Shahr-e-kord, Iran Kiavash fekri 1 , hossein teimoori 2 ,hossein tavallaei 3 , Zahra sajadpoor4
1-pediatrics oncologist-hematologist ,shahr-e-kord university of medical science
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Background and Aim: β- thalassemia is the kind of hemoglubinopathy resulting from inappropriate production of β glubin chain. Patients need repeated transfusion and this lead to elevated ferritin level and also iron overload. Hepcidin, encoded by HAMP gene, is a recently discovered 25 amino acid appears to play a crucial role in iron homeostasis in humans. Its function is influenced by different factors including polymorphism in promoter region ,the most prevalent C -582A>G .The aim of this study is to evaluate blood ferritin level and iron overload in major β-thalassemic patients with and without hepcidin promoter C -582 A>G gene polymorphism.

Methods: This is a cross-sectional study on 91 β-thalassemic patients in shahrekord and Lordegan hospitals. After extraction of blood DNA of patients, PCR was done to evaluate polymorphism.Then fragments were run by RFLP method. Blood ferritin level was measured.Heart and liver MRI T2* was done to evaluate iron overload.

Results: Blood ferritin level of patients with C -582 A>G polymorphism was lower (220 mg/dl) than AA patients, although statistically non-significant (p-value=0.58) . The cardiac iron over load in AA polymorphism is less than AG according to MRI T2* results vice versa the liver iron over load in AA is greater than AG irrespective of ferritin level.

Conclusion: C -582 A>G polymorphism of hepcidin promoter has a lowering effect on ferritin level although statistically non-significant. Hepcidin polymorphism has an independent effect on cardiac and liver iron overload regardless of ferritin level, so it can be used for prognostic evaluation of thalassemic patients.

Keywords: β -thalasemia major , Hepcidin, polymorphism, ferritin

IJBC 2015; Supplementary; P 37
Paper ID: 89

Clofarabine in Combination With L-Asparginase in Treatment of a patient with Refractory T-cell Acute Lymphoblastic Leukemia

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2. hematology fellowship

Abstract

Clofarabine in Combination With L-Asparginase in Treatment of a patient with Refractory T-cell Acute Lymphoblastic Leukemia Goudarzipour K1, Madani F2 1. Assistant professor of Hematology-oncology, Pediatric Congenital Hematologic Disorder Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran 2. Fellowship of Hematology-oncology Pediatric, Pediatric Congenital Hematologic Disorder Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran (madani_farhad@ymail.com) Clofarabine was the latest new drug to be approved for relapsed or refractory acute lymphoblastic leukemia (ALL). in this case we used clofarabine for 5 Days 40mg/m² in combination with L-Asparginase 10000 mg/m² 17 days after start of chemotherapy because the marrow was M3 type in a newly diagnosed 8 years old boy with T-Cell acute lymphoblastic leukemia refractory to response to 4 drugs regimen of induction phase. Minimal residual disease (MRD) was less than 0.02 % after adding clofarabine in compare with near 20% in day 15 of induction. Despite refractory childhood Leukemia main a significant therapeutic challenge, clofarabine is a safe and active single or in combination therapeutic agent in T-lymphoid refractory pediatric acute lymphoblastic leukemia specially in induction phase. Clofarabine administration has been associated with rapid decrease in blasts of the patient. Keywords: Clofarabine, lymphoblastic leukemia, L-Asparginase

Keywords: Clofarabine, lymphoblastic leukemia, L-Asparginase

IJBC 2015; Supplementary; P 38
Paper ID: 93

Diagnostic, therapeutic and evolutive aspects of infantile acquired aplastic anemia : about a retrospective study of 22 cases

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Abstract

Introduction: The acquired aplastic anemia (AAA) is a rare but serious and severe disease. Evolution could be fatal by medullary isuffisance 's complications. We found it useful to carry out a retrospective study over a period of 7 years and to analyze the diagnostic, therapeutic and evolutive characteristics of myelosuppression in southern Tunisia especially in young patients.

Patients and methods: it has enrolled the acquired aplastic anemia young patients (

Results: 22 cases of infantile acquired aplastic anemia were collected, they were 14 men and 8 women with a median age of 14 years (range 10-18 years). The circumstances of discovery were hemorrhagic syndrome, anemic syndrome and fever respectively in 7%, % and11% of cases. Our patient's distribution according to Camitta) score showed 28% AAA moderate, 36% severe AAA and 36% very severe AAA. The etiological investigation has revealed negative in 17 patients (77%) and labeled idiopathic. It showed a viral infections as in postseronegative hepatitis in 23% of patients. In addition to symptomatic treatment, specific treatment was given to all patients. It involved the allogenic bone marrow (allograft), ciclo associated with SAL, ciclo only and androgen. The therapeutic results according to the used therapeutics are detailed in the following table:

therapy Patients number Death rates(%) 5years Overall survival(%) Terapeutic response(%)
Allogeneic bone marrow 8 1(12) 87 RC=63 RP=25
SAL Ciclo 4 2(50) 50 RC=50 RP=25
Ciclo 9 7(78) 16 RC=25 RP=8
androgens 1 0 100 RC=0 RP=22
Total 22 10(45) 60 RC=36 RP=23

Conclusion: The infantile acquired aplastic anemia is a rare and serious disease. It is a therapeutic emergency requiring a carefull etiological investigation and early management with effective therapeutic.

Keywords: Acquired aplastic anemia, young patients, etiology, treatment

IJBC 2015; Supplementary; P 39
Paper ID: 95

Guidelines for Diagnosis of Factor XIII Deficiency in Iran

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Abstract

Factor XIII deficiency (FXIIID) is an extremely rare hemorrhagic disorder with an approximate 12-fold higher incidence in Iran in comparison to general population of the world. A standard algorithm was suggested for precise diagnosis and classification of FXIIID but due to lack of investment for proper equipment and procedures in Iran, almost all parts of this algorithm cannot be applied for Iranian patients. Thus, this study proposes a guideline for proper molecular and laboratory diagnosis of FXIIID in Iran based on available equipment and procedures. In fact, this study suggests a simple and reliable algorithm for early diagnosis of FXIIID, and can therefore with onetime diagnosis of FXIIID, reduce the rate of morbidity and mortality in patients with FXIIID in Iran.

Keywords: Factor XIII deficiency, Laboratory Diagnosis, Iran

IJBC 2015; Supplementary; P 40
Paper ID: 109

The whys of patient centered care

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1. IBTO

Abstract

The term medification in the health system is equivalent to jurification in the judicial systems. There are many different reasons for this phenomenon including the tendency of physicians to follow older practices or the pressure imposed by the health decision makers for lower health costs. Generally the societies show resistance against change and physicians are no exception. Most frequently the physicians pursue the routine old practice citing the evidence that they are more effective with no necessity for the change in the treatment protocols. It is impossible to serve Patient Centeredness unless physicians find a new perception of what leadership in medical performance is and to shift from the old hakim concept of practitioner that is physician-oriented to a new concept of practitioner in the modern era. Moreover, the advocacy and insurance systems should try to lower the complexity of the bureaucratic strategies and consider the role of health consumers important in decision making for their own treatment.

Keywords: Medification, Patient Centeredness, Patient Centered Care

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Paper ID: 120

Evaluation of Immunotherapy in Pediatric with Severe Aplastic Anemia(SAA)

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Abstract

Prognosis of patients with Severe Aplastic anemia(SAA) has been worse than acute leukemia , and most of these patients did not have long survival before 1970. By bone marrow transplantation (BMT) practicability and then peripheral blood stem cell transplantation(PBSCT),if appropriate donor would be present , BMT is the treatment of choice for SAA.

By proper and occasionally excellent results of ALG and cyclosporine , now perhaps SAA treatment by immunosuppressive drugs are more practicable than BMT in 1-12 years old patients(even by presence of proper donors).

Since April 1996 , we treated SAA patients with cyclosporine , ALG and high dose methyl prednisone in Razi children Hospital (Kermanshah University of Medical Sciences) and Bahrami Children Hospital(TUMS). SAA is defined as Camitta Criteria:

1: Hb

1- ALG: 40 mg/kg/day for 4 days/IV or Rabbit ATG 3 mg/kg/day for 4 days/IV

2- Cyclosporine: 10 mg/kg/day/PO

3: High dose methyl predisone :20 mg/kg/day/IV stared (for3 days ;days 1 to 3) , and then 10 mg/kg/day/IV (for 4 days; days 4 to 7), 5 mg/kg/day/IV (for 5days;days 8 to12), 2 mg/kg/day/IV (for 8days;days 13 to 20) ,2mg/kg/day/IV (for 10days;days 21 to 30),and then continuing PO.

Quality assessment of response to treatment was based up on platelet count, hemoglobin and AGC increment (Plt>100,000/□L, Hg>10gr% and AGC>1,000/□L).

Twenty one patients were treated by above protocol (14 male and 7 female). Complication were included;7 patients with hypertension , 6 patients with diabetes , two patient convulsions and three patient cushingoid signs . In ten patients creatinine raised more than five times than the baseline level but all of them it regress to normal level after decrease of cyclosporine dose. In spite of the complications , responses were excellent.

Four patients expired, one due to brain hemorrhage and three due to septic process.

Two patients did not response to treatment. Among fifteen patients who response to treatment, 11 patients respond after one course of treatment, three patients after second course and one patient after third courses of treatment.

Hemoglobin was increased up to 10 gr/dL after 6 months, platelet counts have increased up to 100,000/□L after 5 months and AGC counts have increased up to 1,000/□L after 3 months.

Althogh statistically twenty one patients are few , but treatment response assessments in all patients (other than four patients who were expired) show that mentioned treatment was a successful method for treatment of patients with severe aplastic anemia.

Keywords: Immunotherapy, Severe Aplastic Anemia, ATG

IJBC 2015; Supplementary; P 42-43
Paper ID: 124

Central Nervous System involvement in pediatric's Hemophagocytic lymphohistiocytosis

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Abstract

Introduction Hemophagocytic lymphohistiocytosis (HLH) is a clinical syndrome caused by the uncontrolled activation of cytotoxic T cells and antigen-presenting cells. The typical histopathological findings include the widespread accumulation of lymphocytes and mature macrophages,sometimes with hemophagocytosis, especially in spleen, lymph nodes, bone marrow, liver, and cerebrospinal Fluid (CSF). HLH may have a relapsing and remitting course, or it may rapidly progress to multiorgan failure and death .The initial signs and symptoms of HLH can mimic other more common conditions including bacterial sepsis, viral infections, autoimmune disease and encephalitis.The early clinical signs associated with HLH include fever, hepatomegaly, splenomegaly, lymphadenopathy, rashes, and neurological abnormalities.Central nervous system (CNS) involvement has been reported in 10 – 73% of all HLH patients, either at presentation or during the course of the disease. Clinical symptoms and CSF findings: Central nervous system (CNS) symptoms such as reduced level of consciousness, meningismus, cranial nerve palsies, and seizures can be found in a third of patients with familial hemmophagocytic lymphohistiocytosis(FHL) at diagnosis. There was a strong association between CNS involvement and familial forms of HLH.About half of the children have a moderately increased cell count and/or protein content and low levels of glucose in the cerebrospinal fluid (CSF).In an Asian cohort with predominantly acquired EBV-associated HLH, only 13% had clinical apparent involvement and 16% had abnormal CSF. CSF microscopy can render additional information, if activated histiocytes or hemophagocytosis is found. CNS involvement is variable in its presentation and widely reported to be a poor prognostic factor. Henter and Nennesmo reported the neurologic findings in a series of 23 HLH patients including irritability, seizures, cranial nerve palsies, ataxia, nystagmus, delayed psychomotor development, meningeal signs and evidence of increased intracranial pressure. Haddad et al. reported a series of 34 patients in which they observed a variety of neurologic symptoms including hypotonia or hypertonia, meningismus, seizures, coma with opisthotonos, and abnormal cardiac or respiratory rate. Most published Reports fail to address the frequency of CNS disease among HLH patients. Janka reported CSF abnormalities in 17 of a series of 33 patients and neurological symptoms in less than 10% of the patients.In a multicenter retrospective study, Arico et al reported CSF abnormalities at the time of diagnosis for 55 of 94 patients (58%).Also Patients being treated for HLH appear to be at risk for neurotoxicity, particularly posterior reversible encephalopathy syndrome (PRES), a poorly understood clinical syndrome including headaches, vomiting, confusion, seizures, cortical blindness and other visual abnormalities, and motor signs. It is estimated that 10–28% of patients who receive cyclosporine(on HLH-2004) experience some neurotoxic side effect .Elevated blood pressure, worsening renal and liver function, increased cyclosporine levels, and also CNS involvement of HLH may be triggers for the neurotoxic side effects of treatment. Children with CNS HLH who either never have systemic symptoms or in whom systemic HLH appears later are often diagnosed with other inflammatory CNS diseases.Neuroradiological findings are not sufficiently specific to confirm the diagnosis of HLH and functional immunological tests are recommended in any child with undefined inflammatory CNS disease to exclude a genetic HLH defect.

Imaging The reported CT findings are diffuse parenchymal atrophy, low attenuated lesions in the white matter and calcifications. Reduction of the volume leads to dilatation of the ventricular system and/or subdural fluid collections. The calcifications appear as gyriform linear areas that are more prominent in the regions of gray-white matter junctions.Some low attenuated parenchymal lesions show nodular or ring enhance- ment after contrast enhancement .The reported MR findings include diffuse leptomeningeal and perivascular enhancement, which corresponds to meningeal and perivascular infiltrations of histiocytes and lymphocytes, patchy areas of an increased T2 signal intensity in the white matter of the both cerebral hemispheres, and a diffuse parenchymal volume loss of the cerebrum and cerebellum.In some cases, nodular or ring enhancement of the parenchymal lesion appears due to the compromised blood-brain barrier that is associated with active demyelination.MRI findings are usually polymorphic, multilobal, and bilateral lesions, typically in periventricular location and less frequently with thalamic or basal ganglia involvement.Abnormalities on brain imaging appeared to roughly parallel the severity of clinical manifestations. Histopathology Leukocytes infiltrating the CNS probably secrete cytokines and other neurotoxic factors, such as tumor necrosis factor-a (TNF-a), which may be responsible for the myelinic alteration observed in neurologically asymptomatic patients. Infiltrating leukocytes could also activate in parallel the numerous resident brain macrophages (the microglial cells) and astrocytes, which in turn, can secrete eurotoxic glutamate and free radicals. The histopathologic findings of HLH in pediatric patients with involvement of the CNS could be classified on the basis of the stages of the disease as determined microscopically, and the stages are characterized by increasing severity: stage I primarily shows only leptomeningeal infiltrates of lymphocytes and histiocytes/macrophages. Stage II shows additional parenchymal involvement with perivascular infiltrations and stage III shows signs of cerebral tissue necrosis and demyelination in addition to the massive tissue infiltration that particularly affects the white matter . Treatments There is significantly increased risk of mortality among patients with CNS manifestations.A recent study of 193 patients with HLH, which identified a trend towards a poorer prognosis for patients with clinical neurological manifestations and abnormal CSF findings. Mashuku et al. also reported poorer outcomes in patients with CNS symptoms and/or deficient NK cell activity. In most protocols, treatment of central nervous system involvement relies both on systemic immunosuppression and intrathecal therapy. Reactivations of HLH in the CNS are common, not rarely without systemic activity. Long term neurological outcome of patients is influenced by the disease and the treatment including HSCT.It appears hematopoeitic stem cell transplantation (HSCT) in patients with HLH can be the only available treatment procedure that is capable of preventing HLH CNS disease progression and that can result in cure when performed early enough after remission induction. Remarkable effect of HSCT on the prevention of subsequent neurological manifestations and the good cognitive and neurological evolution of the transplanted children who are long-term survivors. Interestingly, lasting remissions have also been observed in patients with stable mixed donor-cell chimerism, indicating that even a limited level of immune reconstitution by donor cells is sufficient to control the dysfunctional immune system in patients.HSCT from alternative donors, ie, matched unrelated and haplo-identical related donors can also be considered. However, this approach raises the question of the accuracy of HLH diagnosis in patients without a family history or consanguinity.

Conclusion: CNS involvement in HLH is a frequent finding not only in familial HLH but also in secondary HLH. CNS involvement is highly variable in its presentation and associated imaging findings. CNS involvement is associated with poor outcomes. In view of the low regenerative potential of nervous tissue, superior outcomes may require that carefully monitor neurological manifestations in patients with HLH. Patients who have CNS manifestations of HLH will require an appropriate regimen of intensified chemotherapy and HSCT. It is also important to consider HLH in a child with unexplained neurologic manifestations, especially one with fever, pancytopenia, or hepatosplenomegaly.

Keywords: Central Nervous System(CNS), Hemophagocytic lymphohistiocytosis (FHL), Hematopoeitic stem cell transplantaion(HSCT)

IJBC 2015; Supplementary; P 44
Paper ID: 127

Hematopoietic stem cell transplantation with a reduced-intensity conditioning regimen in pediatric patients with primary hemophagocytic lymphohistiocytosis

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2. -

Abstract

Objective: Over the past years,Hematopoietic stem cell transplantation (HSCT) has become as well-established treatment of choice for Primary hemophagocyticlymphohistiocytosis (HLH).
The toxicity of myeloablative conditioning regimen (MAC) for this patients with per-HSCT comorbidity has led to researchers to attempt reduce intensive conditioning regimen (RIC) with minimize toxicity and graft rejection. This prospective study investigates the result of HSCT using the same RIC for pediatric patients with primary HLH who were referred to our center.
Patients and Methods: 21 patients (11 male, 10 female) with Familial HLH (n=8), Griscelli syndrome (n=7) and Chediak–Higashi syndrome (n=6) who had received HSCT between 2007 and 2015 were enrolled in the study. All patients had experienced at least one HLH crises before transplantation and were treated with chemotherapy protocols and one of them was in HLH crisis.The median age at transplantation was 14.1 months (range: 4–72). All of the patients received Fludarabine in combination with Melphalan and Anti-thymocyte globulin (ATG) as a RIC regimen before HSCT. Cyclosporine with methylprednisolone was used as Graft-versus-host disease (GVHD) prophylaxis regimen.
Results: Engraftment occurred in all patients.The median times to neutrophil and platelet engraftments were 11 days (range: 8-33), and 21 days (range: 10–67), respectively.
Patients were transplanted from HLA-identical sibling (n=9), matched other related (n=7), HLAmatched unrelated donor (n=2)and unrelated mismatched donor (n=3).Peripheral blood (n=13), bone marrow (n=6) and cord blood (n=2) were used as the stem cell source.With the median follow up 31months (range: 3-86) 15 of patients are alive and 13 of them have full chimerism and the other 2 have mixed chimerism. Regardless of having full or mixed chimerism, all alive patients are disease free with no manifestation of primary HLH symptoms.7 patients developed acute GvHD grade II-IV and 4 patients developed chronic GvHD. The causes of death were GvHD and infection.
Conclusion: The results of our study show RIC regimen well tolerated in HLH patients with regard to severe and serious comorbidities in this group of patients. The future large multicenter study is recommended to determine the optimal conditioning regimen for improvement of HSCT results in PID patients.
Keywords: primary, transplantation, lymphohistiocytosis

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Paper ID: 139

Changing traditional written case records in Oncology wards to programmed digital files is not a beneficial measure but a indispensable strategy

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2. -

Abstract

In regard to presence of usual defects in completing traditional medical written case records , incoordination as a state of being scattered data and occasional discrepancy at recordings of clinical or laboratory data of patients , difficulties in maintenance of paper case records in relations to space needed for many years of storage and high vulnerability of these to repeated physical contacts, light and peripheral humidity, inability for continuous availability and occasional loss secondary to difficulties in storage for prolonged years and finally inability to have a quick and on demand conclusion and statistical analysis of data, it seems that establishment of programmed digital files as patient's medical dossiers will be an appropriate response for solving this problem. These files in a programmed and appropriate format can resolve the difficulties and discrepancies in entering the data. They occupied incomparably smaller space which make recording and transportation of a great deal of data in a small portable memory flash. So these data are resistant to environmental physical harmful agents and coincidentally by using an upgradable antiviral programme and giving repeated backups restoration of information every time Is possible. Such a programme if will be designed as web based, every where throughout the world the physician can be an online access to the patient's recent clinical state and the last paraclinical data and make necessary changes in patient's management protocol for responsible nurses.

Keywords: Changing traditional,Oncology,programmed digital

IJBC 2015; Supplementary; P 46
Paper ID: 154

HSCT for relapsed/refractory HD

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Abstract

Autologous stem cell transplantation (ASCT) is the optimal treatment strategy for Hodgkin lymphoma (HL) patients unresponsive to first course of therapy or relapsing after primary treatment . We analyzed our registry data for patients with Hodgkine lymphoma in Taleghani bone marrow center affiliated to Shahid Beheshti university of medical sciences All the patients with HL who underwent a first autologous or a first or second allogeneic HSCT between 1386/6(2007) and 1392/4(2013) were included. Autologous and allogeneic HSCT were performed in 96 and 6 patients, respectively. Three years Overall survival (3yOS) and event free survival was 83% and 67% respectively. We performed allogenic reduced intensity transplantation in 6 refractory or post autologous HSCT relapse . Out of them 3 is in complete remission or stable disease. Grade 2 acute GVHD occurred in one patient who relapsed later and chronic GVHD was also occurred in one patient who is in CR .16 patient passed away ,11 out of them were refractory to autologous HSCT. Disease status at transplantation,disease stage at last relapse , the number of previous chemotherapy courses and age were prognostic factors. OS was favorable even in patients who underwent autologous HSCT in disease status other than complete remission.A first allogeneic HSCT without a previous autologous HSCT was performed only in one patient who relapsed early even after transplantation. In conclusion, autologous HSCT is effective and even curative in patients with relapsed and refractory HL It is preferred to performed as soon as possible in patients who responded to salvage chemotherapy. Allogeneic HSCT is feasible with little complications and might be beneficial in patients who relapsed after autologous HSCT,.

IJBC 2015; Supplementary; P 47
Paper ID: 157

Hemophagocytic Lymphohistiocytosis Complicating Erythroleukemia in a Child with Monosomy 7: A Case Report and Review of the Literature

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Abstract

Hemophagocytic lymphohistiocytosis (HLH), a critical and severe disorder, is characterized by severe hyperinflammation resulting from proliferation of reactive lymphohistiocytes on the basis of various inherited or acquired immune deficiencies. The entity is further classified into two subgroups: Familial (primary) HLH and acquired (secondary) HLH. Hemophagocytic lymphohistiocytosis (HLH) is an extended entity enclosing a variety of macrophage-related disorders characterized by fever, pancytopenia, hepatosplenomegaly, and finally hemophagocytosis in bone marrow, liver, or lymph nodes. Hemophagocytic syndromes associated with malignancy occur in two forms. First, hemophagocytic syndromes (MAHS) can present initially, masking various hematolymphoid malignancies, and second, they may complicate the initial course. Interestingly, different types of malignancies are categorized under each group; specifically, T-cell leukemias and lymphomas are often masked by a hyperinflammatory MAHS state, whereas B-cell leukemias and germ cell tumors are often complicated by MAHS. The patient presented here developed obvious erythrophagocytosis following chemotherapy for AML and also multiple doses of G-CSF he received during prolonged period of pancytopenia. In some cases of MAHS, bone marrow aspirate reveals blasts or dysplastic neutrophils phagocytosing other cells and in these cases specific chromosomal abnormalities are observed on cytologic studies. Malignant neoplasm-associated HLH, a subtype of secondary HLH, is mainly accompanied by malignant lymphoma and, less frequently, by other hematological malignancies and carcinomas. HLH is known as a rare and adverse complication of childhood malignancies including acute myeloid leukemia (AML). There are only four reports available in the literature on the association of AML (M6) and HLH, all of which are described in adult patients.

A variety of chromosomal abnormalities are observed in these associations; however, monosomy 7 has not been previously reported in these cases. Herein, the first case of childhood HLH complicating erythroleukemia in a 5-yearold boy with monosomy 7 is reported. The patient presented here developed obvious erythrophagocytosis following chemotherapy for AML and also multiple doses of G-CSF he received during prolonged period of pancytopenia. In some cases of MAHS, bone marrow aspirate reveals blasts or dysplastic neutrophils phagocytosing other cells and in these cases specific chromosomal abnormalities are observed on cytologic studies. In our patient macrophages were phagocytosing other blood cells mainly erythroid precursors.

IJBC 2015; Supplementary; P 48
Paper ID:

Curability of Hematopoietic Stem Cell Transplant (HSCT) for malignant and non-malignant patients in Amirkola Bone Marrow Transplant center

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Abstract

Background: Hematopoietic stem cell transplant is the main and definitive treatment for thalassemia major and some malignancies. For this purpose we tried to treat some of our thalassemia major patients who had suitable completely match donor with HSCT in thalassemia patients and also in different malignancies with allogenic and autologous HSCT in Amirkola B.M.T ward.

Methods and materials: 31 thalassemia major had underwent to HSCT from august 2010 to November 2014 in Amirkola B.M.T ward.14 Patients were female and 17 male, age distribution was between 3-26 years. The patients divided in two groups on the basis of Lucarreli classification (on the basis of portal fibrosis, hepatosplenomegaly and serum ferritin level). Group I were patients in class I and II and group 2 were patients in class III. Age distribution in group I was between 3 to 17 year and age distribution in group II was between 18 to 26 year.For Malignant Patients,6 cases with acute lymphoblastic leukemia (ALL) treated with allogenic HSCT. 5 out of 4 patients with acute myeloblastic leukemia (AML) had allogenic HSCT and 1 patient had autologous HSCT. 6 patients with Hodgkin disease had autologous HSCT. 1 patient with Non- Hodgkin lymphoma was treated with autologous HSCT and 5 patients had autologous HSCT.
1 patient with myelodysplastic syndrome was treated with allogenic HSCT. From cases of multiple myeloma 2 patients had allogenic HSCT and 23 cases had autologous HSCT. we collected all information about engraftment and B.M.T failure and rejection and survival and event free survival, also mortality and some side effect for instance GVHD. All information collected and analyzed.

Findings: Event free survival in major thalasemia group I was 75% and in group II was 40%. 2 out 16 patients in class I died and 5 patients in class III died. Severe GVHD happened in 6 cases, but only one happened in group I patient.
For Malignant Patients 1 lukemia case relapse happened 1 for ALL and 2 for AML and 13 in Multiple Myeloma and 5 of them died 1-3 years post HSCT.

Conclusion: HSCT is a choice way to relieve thalassemia major patient from their problems but in classI and II specially in young patients has the best result. Also in relapsed malignant patients is effective.

Key words: HSCT, major thalassemia, malignancies, curability

POSTERS

IJBC 2015; Supplementary; P 51
Paper ID: 2

Lifesaving dormancy induction in refractory Acute Myeloblastic Leukemia with monosomy 7

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1. fellowship

Abstract

Refractory AML with monosomy 7 patients have a very poor prognosis. Therefore, rationally designed new therapies, including metronomic chemotherapy regimen with histidin deacetylase inhibitors (Valporic acid, ATRA) are being investigated as potential treatments for this patient population. This is a case report of a patient with primary refractory AML who was saved with oral metronomic and regular adjuvant therapy.

Keywords: Metronomic Chemotherapy, Histone deacetylase Inhibitor, Acute Myeloblastic Leukemia, Valporic Acid, ATRA

IJBC 2015; Supplementary; P 52
Paper ID: 3

Refractory isolated thrombocytopenia with triosomy 8

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1. resident
2. assistant professor

Abstract

Refractory isolated thrombocytopenia (RIT) is an uncommon variant of myelodysplastic syndrome (MDS) that initially presents as chronic pure thrombocytopenia. Because of the lack of distinguishable dysplasia, RTC has often been misdiagnosed as idiopathic thrombocytopenic purpura.We describe a patient with RIT and trisomy 8 for whom a bone marrow mononuclear cell (BMNC) . This RIT patient exhibited an special pattern of MDS.We suggest that RIT be classified as a subtype of MDS on the basis of its specific molecular property.

Keywords: isolated refractory thrombocytopenia,triosomy 8,myelodysplastic syndrome

IJBC 2015; Supplementary; P 53
Paper ID: 5

Ketamine: One of the best analgesic and sedative in Bone marrow procedure, s in oncologic kids

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1. associate professor

Abstract

Bone marrow aspiration and biopsy in outpatient setting is trouble and painfull procedure for pediatric cancerous patients.Performing this procedure with minimum pain and mental sequel is a ideal target for pediatric oncologists. The sedation regimen for bone marrow(BM) procedures is still varied in outpatient setting. We studied Midazolam and Ketamine for premedicants in pediatric anesthesia for BM procedures and find out this way has good effect

Keywords: ketamin,analgesia,bone marrow aspiration biopsy

IJBC 2015; Supplementary; P 54
Paper ID: 6

Implantable port devices, complications and outcome in pediatric cancer patients, a retrospective study in Hamedan University of Medical Sciences

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Abstract

Introduction: Peripheral blood vessels, due to availability are used for many years in cancer patients, however in patients with potentially harmful drugs to skin (vesicant drugs) or difficult accessibility to vessels, the use of implantable port (totally implantable venous access port-TIVAP) devices with central vascular access are important. Materials and methods: In this retrospective study, 85 pediatric cancer patients younger than 14, with TIVAP implantation, were followed for their complications and outcome. Results: Mean days of implanted port usage were 531 ± 358 days in all patients. This period was 287 ± 294 days in complicated patients. Complications included as infection (tunnel infection and catheter related blood-stream infection), malfunction and thrombosis, skin erosion, tube avulsion and tube adhesion to the adjacent vessels were seen in 30.6% of patients. Discussion and conclusion: According to the published data and our experience, the most common complications in TIVAP are infections and catheter malfunction. It is important to notice that in order to prolong its efficacious life, effective sterilization methods, prevention of clot formation and prevention of trauma, are the most useful measures.

Keywords: Totally implanted venous access port, Malignancy, Pediatric, Risk factors

IJBC 2015; Supplementary; P 55
Paper ID: 7

Exchange transfusion in undiffrentiated leukemia with hyperleukocytosis

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1. fellowship
2. assistant professor

Abstract

Hyperleukocytosis refers to a laboratory abnormality that has been variably defined as a total leukemia blood cell count greater than 50 x 109/L (50,000/microL) or 100 x 109/L (100,000/microL). In contrast, leukostasis is a medical emergency most commonly seen in patients with acute myeloid leukemia (AML) or chronic myeloid leukemia (CML) in blast crisis. It is characterized by an extremely elevated blast cell count and symptoms of decreased tissue perfusion.Apheresis is a one of the treatment choice,but in early childhood it isn,t possible. We presented a 2.5 months with undiffrentiated leukemia with 750000 leukocytes which treated by manual exchange transfusion.

Keywords: Hyperleukocytosis, undiffrentiated leukemia, exchange transfusion

IJBC 2015; Supplementary; P 56
Paper ID: 8

Primary Bone lymphoma is a rare tumor in children

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1. fellowship
2. assistant professor

Abstract

Primary lymphoma of bone (PLB) is a rare type of cancer starting in bone, it accounts for about 7% of all primary bone tumours especialy in children. This is distinct from lymphoma which started in the lymph notes and then spread to the bones (bone metastases).We presented a 16 years old girl with femur and humerus primary site and bone marrow metastasis.

Keywords: bone lymphoma , children

IJBC 2015; Supplementary; P 57
Paper ID: 9

Drug induced CNS events in childhood acute lymphoblastic leukemia

Dr Fazl Saleh[1] , Prof Maral Mokhtari[1]

1. fellowship

Abstract

Posterior reversible encephalopathy syndrome (PRES), also known as reversible posterior leukoencephalopathy syndrome (RPLS) and cerebral sinus thrombosis is a two similar clinical situations, that are characterized by headache, confusion, seizures and visual loss. Thease may occur due to a number of drugs such as Asparginase and steroids in acute lymphoblastic leukemia treatment regimen.This two conditions have 2 different therapeutic approach.We presented 2 case of PRES and 1 case of cerebral sinus thrombosis afteracute lymphoblasti leukemia (ALL) treatment.

Keywords: Posterior reversible encephalopathy syndrome , reversible posterior leukoencephalopathy syndrome , Asparginase

IJBC 2015; Supplementary; P 58
Paper ID: 11

What is the role of stem cell transplantation in the treatment of the Burkit leukemia?

Prof Mohamad Reza Bordbar[1] , Prof Mahdi Shahriari[2] , Prof Soheila Zareifar[3] , Dr Fazl Saleh[4]

1) assistant professor
2) associate professor
3) associate profesor
4) fellowship

Abstract

Burkitt lymphoma (Burkitt's tumor, Burkitt's lymphoma, or malignant lymphoma, Burkitt's type) is a cancer of the lymphatic system, particularly B lymphocytes found in the germinal center. Burkitt and Burkitt-like lymphomas have a rapid and aggressive clinical course with frequent bone marrow and central nervous system (CNS) involvement. These are considered to be medical emergencies and require immediate diagnostic and therapeutic intervention. Chemotherapy is the mainstay of treatment for this disease.In this article we discuss about Hematopoetic stem cell transplant(HSCT) for this patients.

Keywords: HSCT, Burkitt's lymphoma ,Burkitt's leukemia

IJBC 2015; Supplementary; P 59
Paper ID: 12

Metronomic chemotherapy in hematologic malignancies

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Abstract

Background: Metronomic chemotherapy, which is continuously administered systemically at close to non-toxic doses, targets the endothelial cells (ECs) that are proliferating during tumor angiogenesis. This leads to harmful effects of an even greatly increased number contiguous tumor cells.
Purpose: The Panel was convened for the purpose of creating package to develop an evidence-based package for the metronomic chemotherapy for pediatric hematologic malignancies.
Methods: We followed previously validated articles for creating evidence-based chemotherapy guideline (package). Working groups focused on articles included metronomic chemotherapy in children and adults. Each working group developed key clinical questions, conducted systematic reviews of the published literature, and compiled evidence summaries. The Grades of Recommendation Assessment, Development, and Evaluation approach was used to generate summaries, and evidence was classified as high, moderate, low, or very low based on methodological considerations.
Results: We reviewed evidence-based systematically literatures and prepared a chemotherapy protocols guideline (package) for hematologic malignancies in children for end stage paients protocols for increased their survival and life expectancy.
Conclusion: This package represents an evidence-based approach to metronomic chemotherapy in children with leukemia and lymphoma. Although some protocols are similar to adult-based protocols, there are key distinctions in multiple areas.
Keywords: Metronomic chemotherapy, children, leukemia ,lymphoma, hematologic malignancies

IJBC 2015; Supplementary; P 60
Paper ID: 14

Basidiobolomycosis mimic lymphoma

Prof Omid Reza Zekatav[1] , Dr Gholamreza Fathpour[2] , Dr Maral Mokhtari[3]

1. associate professor
2. fellowship
3. assistant profesor

Abstract

Basidiobolomycosis is a rare disease caused by the fungus Basidiobolus ranarum, an environmental saprophyte found worldwide. Patients with B ranarum infection may present with subcutaneous, gastrointestinal, or systemic lesions. Gastrointestinal basidiobolomycosis poses diagnostic difficulties, as its clinical presentation is nonspecific, with no identifiable risk factors.We presented a 5 years old boy with B symptoms and abdominal like lymphoma mass that finally was diagnosed Basidiobolomycosis.

Keywords: Basidiobolomycosis, lymphoma, abdominal lymphoma

IJBC 2015; Supplementary; P 61
Paper ID: 15

Isolated Mococutaneus relapse with T-cell acute lymphoblastic leukemia.

Dr Gholamreza Fathpour[1]

1. fellowship

Abstract

Despite high cure rates, approximately 20% of patients with acute lymphoblastic leukemia (ALL) have disease relapse. Isolated recurrence in oral cavity is extremely unusual. We report a case of an isolated relapse occurred in a child with T-lineage ALL. Clinical picture included diffuse maculopapular cutaneus lesions and hard palate mass with gingival hypertrophy without any clinical or hematological alterations. Diagnosis was confirmed by biopsy and immunohistochemical staining. Bone marrow aspiration was normal. He was treated by systemic chemotherapy and disapear lesions deramatically. This case report highlights the relevance of dental care during and after chemotherapy, not only to treat lesions in the oral cavity resulting from the disease itself or from treatment side effects, but also to detect unusual sites of ALL relapse.

Keywords: T-lineage ALL, oral cavity leukemia

IJBC 2015; Supplementary; P 62
Paper ID: 16

Amniotic Membrane in Vincristin extravasation Wound Management

Dr Gholamreza Fathpour[1]

1. fellowship

Abstract

Extravasation is inappropriate or accidental infiltration of chemotherapy into the subcutaneous tissue surrounding the administration site. Hese injuries range from less significant erythematous reactions to skin sloughing and necrosis. Chronic non healing wounds are estimated to affect as many as 1-2% of individuals during their lifetime, and account for billions of dollars of expense annually on both a national and global basis. We report a novel dehydrated amniotic membrane allograft for the treatment of chemotherapy drugs extravastion wounds. We describe the results of EpiFix treatment in one 3 years old girl with acute myelogenic leukemia .Then she referred for a definitive plastic surgery procedure. Healing was observed in a variety of wounds with one to three applications of the dehydrated amniotic membrane material. The material was well tolerated by patients. Healed wounds did not recur in 3 months follow-up. Further investigation of the use of dehydrated amniotic membrane in broader application to various types of dermal wounds should be considered.

Keywords: chemotherapy drugs extravastion, amniotic membrane allograft

IJBC 2015; Supplementary; P 63
Paper ID: 19

Effect of Chewing Gum on Oral Mucositis in Children Undergoing Chemotherapy

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Abstract

Effect of Chewing Gum on Oral Mucositis in Children Undergoing Chemotherapy Abstract Background and objectives: Oral mucositisas an adverse effect of chemotherapy refers to inflammation and ulceration that occurs in the mouth. The transient decrease in saliva production exacerbates oral inflammation. Our goal was to study whether stimulation of salivary flow can protect the oral mucosa against chemotherapy. Methods: This randomized clinical trial was done in Amir Kabir hospital, Arak, Iran. Control group was composed of 65 patients who received mucotoxic drugs. Test group was made up of 65 patients received similar drugs in addition to sugar free gums simultaneously. These patients consumed 6 pieces of gums per day for 15 days. Daily evaluation of the severity of mucositis was done by standardized follow up form during 15 days. Results: Severe oral mucositis occurred in 30 (46%) of 65 patients in the test group and in 26 of 65 (40%) patients in the control group. Difference was not significant.The reduction was chiefly due to a decrease in cases of grade 1-2 oral mucositis (15% vs. 35%). Conclusion: Stimulation in saliva flow can decrease inflammatory injuries of the oral mucosa during chemotherapy. However, it is not effective to decrease severe mucositis. The type of chemotherapy regimen is the main determinant of severe oral mucositis. Keywords: mucositis , gum chewing ,chemotherapy, children

Keywords: mucositis , gum chewing ,chemotherapy, children

IJBC 2015; Supplementary; P 64
Paper ID: 20

Association between serum ferritin level and diastolic cardiac function in patients with major β-thalassemia

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Abstract

Association between serum ferritin level and diastolic cardiac function in patients with major β-thalassemia Abstract Background and objective: Patients with major thalassemia usually suffer from iron over load that is the main cause of heart failure. Prevention of myocardial siderosis is a key step to reduce rate of mortality in thalassemic patients. Many methods have been created for measuring iron level. In addition to usual methods, echocardiography has been gained prominence. Our objective was to study association between echocardiography parameters and serum ferritin level in patients with major thalassemia. Methods: Sixty-six patients with major thalassemia were studied in Amir Kabir hospital, Arak, Iran. Serum ferritin levels were measured during 3 months in patients with no symptoms of infection. It was measured by enzyme-linked immunosorbent assay (ELISA). Ejection Fraction (EF), Fractional Shortening (FS) and Early/Late ratio (E/A) were studied by echocardiography. Results: 52% were female and 48% were male. Range of age was from 3 to 46 years old. Mean of seum ferritin level was1912 ± 1748 ng/dl and its range was from 303 to 8333 ng/dl. There were significant correlations between serum ferritin level and EF(r=0.3 and P=0.05) and also between serum ferritin level and FS. Conclusion: Due to significant association between serum ferritin level and echo parameters, it is beneficial that all patients with major thalassemia undergo echocardiography to gain clearer understating about cardiac function. Key words: Thalassemia, Ferritin, Echo, Heart

Keywords: Thalassemia, Ferritin, Echo, Heart failure

IJBC 2015; Supplementary; P 65
Paper ID: 21

What is strategy forAntracyclin forbided leukemic patients in acute leukemia?

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1. associate professor
2. assistant professor
3. associate profesor
4. fellowship

Abstract

Anthracycline based treatment for acute leukemia can be associated with significant morbidity and mortality among children patients or those with significant co-morbidities. Furthermore, for patients with previous anthracycline exposure or preexisting cardiac disease anthracyclines or after relapse and recived full dose of anthracycline pose an increased risk of cardiotoxicity.

For such patients intensive treatment options are limited.

We reviewed possible strategies for non- anthracycline regimen.

For exaple the FLAG (fludarabine, cytarabine and filgrastim) regimen is a non-anthracycline based chemotherapy useful for relapsed/refractory acute leukemia and as initial therapy or Clofarabine for acute myeloid leukaemia in patients for whom anthracycline-containing combination chemotherapy is inappropriate – first line.

Keywords: FLAG, anthracycline, Clofarabine, fludarabine

IJBC 2015; Supplementary; P 66
Paper ID: 24

Evaluating the Results of Psychotherapy on Anxiety among Mothers of Children with Leukemia

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Abstract

Background: Children with leukemia and their families face a long period of medical treatment and uncertainty about the future. These families may suffer from short and long-term emotional problems. The aim of the present study was to assess the effect of supportive psychotherapy on the anxiety of mothers whose children suffer from leukemia.

Materials and methods: The current research were performed on mothers who had a child suffering from leukemia hospitalized in Children Mofid Hospital, Tehran, Iran, who were chosen randomly. The research method was a pseudo experimental approach with pretest/posttest plan. The pretest Kettles' anxiety questionnaire was given to all the mothers and after seven sessions of supportive psychotherapy the posttest was performed and the grades were compared.

Results: Ten mothers finished all seven therapeutic sessions. There was a statistically significant difference between the pretest and posttest mean scores confirming the mothers' reduced anxiety level ($p < 0.001$).

Conclusion: Finding effective and newer approaches for improving the well being of parents with a sick child is an important challenge of today's medicine research. Based on our findings it is possible to reduce the anxiety among mothers of children with leukemia using supportive psychiatric therapies.

Keywords:
leukemia, mothers, anxiety, psychology, child.

IJBC 2015; Supplementary; P 67-68
Paper ID: 25

Pediatric end of life care barriers: the relationship between nurses' perception of barriers magnitude and their demographic factors

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Abstract

Background: The idea that a child may die is simply unimaginable to most People, yet children die daily. Providing comprehensive and compassionate end-of-life (EOL) care for children within a family-centered and developmentally appropriate context is necessary. Health care professionals face numerous obstacles and challenges while providing care to this unique population of clients and their families, which differ from those cited for adults. Although interdisciplinary care is essential for EOL care quality, nurses play the key role of child-family advocate. Their perceptions of existed barriers which are affected by their personal and professional experiences related to death and dying influence quality of care for children.
Aim: To assess the relationship between demographic characteristics of pediatric nurses and their perceptions of barriers magnitude.
Method: In this cross-sectional, descriptive study a sample of 173 nurses working in pediatric units (pediatric oncology units, pediatric intensive care units, pediatric general units and one pediatric emergency unit) in 2 hospitals (Shahidbahonar and Afzalipour) supervised by Kerman University of Medical Sciences was surveyed. All nurses working in the aforementioned units were surveyed. A translated modified version of National Survey of critical care Nurses' Regarding End-of-Life Care questionnaire was used to assess the relationship between nurses' perception of barriers magnitude and their demographic characteristics.
Results: The item "families not accepting poor child prognosis" belonged to patient-family related category achieved the highest perceived barriers magnitude score (5.04). The next 2 barriers with highest scores were respectively: "no available support person for family such as a religious leader" (4.97) and "poor design of units that do not allow for privacy of dying child or grieving family" (4.96). Both these two items were belonged to organizational related barriers.
The item "continuing to provide advanced treatments to dying children because of financial benefits to hospital" belonged to the organizational related category achieved the lowest PBM score (2.19). Participants' family and closed friends' death experience positively correlated with perceived organizational-related barriers magnitude (P= 0.02). Also participants' nursing experience positively correlated with perceived health-care professional related barriers magnitude (P= 0.04)
Conclusion: This study provides nurses with some valuable insights about effect of nurses' experiences on their perceptions of EOL care barriers. Improving nurses' self-awareness of their perception is required. This may support nurses' awareness of the barriers and learn how to face them. Therefore developing EOL and palliative care education based on Islamic beliefs and Iranian culture may enhance nurses' knowledge and skill, which enable them to face the challenges of EOL care. Also establishing outpatient palliative department and palliative care units in the community may have a positive effect on children's EOL care. Active involvement of chaplain and a psychologist in the treatment team in order to provide spiritual and

psychological support for the family may strengthen EOL care.
Keywords: Relationship, Nurses' perception, End of life care barriers, Terminally ill children

IJBC 2015; Supplementary; P 69
Paper ID: 27

What,s treated Isolated cutaneus T-cell lymphoma?

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Abstract

Cutaneous T-Cell Lymphoma (CTCL) is a subset of non-Hodgkin's T-cell lymphomas which presents with malignant lymphocytic infiltration of the skin. Mycosis Fungoides (MF), the commonest variant of CTCL, is characterized clinically by an indolent clinical course with subsequent evolution of patches, plaques . Isolated CTCL often dosen,t refer to pediatric oncology clinic . The evaluation and treatment of individuals with mycosis fungoides is usually conducted on an outpatient basis. Symptomatic treatments (emollients, antipruritics) are used in combination with specific topical and systemic treatments.I n this article present a 10 years old boy with MF and disscus about different treatment type of this pathology.

Keywords: cutaneus T-cell lymphoma, mycosis fonguides

IJBC 2015; Supplementary; P 70
Paper ID: 28

Sever Hemophilia A in a girl infant with mosaic Turner syndrome and PHPV.

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Abstract

A six months old girl was referred from an ophthalmologist due to post operation bleeding. She was scheduled for operation because of Persistent Hyperplastic Primary Vitreous (PHPV). Workups were done and prolonged PTT with normal platelet count, normal Bleeding Time and PT were detected. There was negative family history of bleeding tendency in both maternal and paternal family, so at the first step Factor XI assay was requested which was normal. Then VW Factor and Factor VIII were assayed. Factor VIII
Keywords: Hemophilia A, Sever Factor VIII deficiency, Turner Syndrome, PHPV

IJBC 2015; Supplementary; P 71
Paper ID: 29

Neonatal screening for anemia in Iranian healthy full term newborns: Is there any indication?

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Abstract

Background: Newborn screening is a systematic application of tests for early detection, diagnosis, and management of certain genetic or metabolic disorders that can lead to mortality, morbidity, if untreated. As mentioned by WHO each year over 330,000 babies are born worldwide with a severe form of hemoglobinopathy. Although Sickle cell anemia is not common in most parts of our country, but high incidence of Alpha thalassemia, G6PD deficiency and spherocytosis are three reasons that newborn screening should be routine in all delivery rooms in order to decrease morbidity. Materials and Methods: 1000 full-term neonates were screened over near 3 years period for detection of hemoglobinopathy or hereditary spherocytosis. This study was conducted in Zeinabiieh and Hafez Hospitals affiliated to Shiraz University of Medical Sciences, Shiraz, Iran. The screening was performed on cord blood samples collected on EDTA and method was approved by ethical Committee. After sample collection, complete blood cell count, osmotic fragility test and hemoglobin electrophoresis were done for each sample. Results: Total prevalence of causes of anemia in this neonatal screening program (including hemoglobinopathies and spherocytosis) was 12.2%; the most prevalent one was Alpha thalassemia (6.4%) followed by hereditary spherocytosis (4.8%) and sickle cell anemia (1.2%). The total analysis for detection of Alpha thalassemia with screening methods by mean corpuscular volume (MCV) ≤ 97fl and mean corpuscular hemoglobin (MCH) ≤ 29 and also hemoglobin level ≤ 16g/dl plus MCHC>35 were the appropriate cut off points for our population. Conclusion: Our study does not approved previous cut off points for the mean corpuscular volume (MCV35 for screening index. We suggested the new cut off points for neonatal screening program for Iranian population that are MCV≤97fl and/or MCH ≤29 for Alpha thalassemia and hemoglobin level ≤16 plus mean corpuscular hemoglobin concentration (MCHC) >35 for hereditary spherocytosis. A successful disease prevention strategy could lead to significant savings in an environment of spiraling healthcare costs and scarcity of blood products. Regarding the results and based on prevalence of hemoglobinopathies, we recommended the neonatal screening program for all neonates in Iran with addition of Sickle prep and Hb electrophoresis in southern Provinces of Iran and a format for neonatal discharge summary was suggested which Hb, and rbc indexes (MCV,MCH,MCHC) should be included in it .

Keywords: Hemoglobinopathy, Neonatal screening program, Alpha thalassemia, hereditary spherocytosis, Sickle cell anemia.

IJBC 2015; Supplementary; P 72
Paper ID: 31

Teaching hematology Oncology cases in bedside versus teaching in the classroom for both residents and medical students.

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Abstract

Introduction: Bedside teaching is defined as teaching in the presence of a patient, it is a vital component of medical education. The aim of this study was to evaluate the effectiveness of lectures with two methods of case based teaching (at the bedside and in the classroom) in the teaching hospitals (for both undergraduates and residents of pediatrics).

Methods: from January 2011 to September 2013; in a pediatric hematology ward and clinic affiliated to Shiraz University of Medical Sciences with undergraduate and pediatric residents, this study was conducted for evaluation of efficacy of teaching methods: lecture, case based teaching, bedside teaching in these two levels. Thirty undergraduates and twenty pediatric residents were asked to study a topic of their curriculum from their text (Approach to anemia in children and hemoglobinopathies) then pretest was taken from learners in the two levels; then either lecture with power point or case presentation or bedside discussion were conducted. One week later posttest was taken. Evaluation of these three methods was done by a questionnaire from learners. Thirty undergraduates and twenty pediatric residents were asked to study a topic of their curriculum from their text then pretest was taken from learners in the two levels; then either lecture with power point or case presentation or bed side discussion were conducted. One week later posttest was taken, and then evaluation of these three methods was done by a questionnaire from learners.

Results: twenty eight (93.33%) of under-graduates and twenty (100%) of pediatric residents had evaluated case based teaching superior to bedside teaching and these two methods superior to lecture method. Comparison of pre-tests and post-tests showed better problem solving by learners after both case based teaching and bedside teaching methods: undergraduates and pediatric residents gained 80% better scores after bedside teaching; 83% better scores after case based teaching and 65% better scores after lecture. Main disadvantages of bedside teaching from learners' point of view was becoming ashamed or disappointed when they had wrong answers or comments in the presence of the patients.

Conclusion: trainees believed that in the case based teaching they are more relaxed and have more self-esteem than at the bedside discussion of the patients. Clinician teacher must involve patients and learners in the process of bedside teaching, by preparing a comfortable situation and by using available technology, for professionalism and role modeling.

Keywords: Anemia, Hemoglobinopathy, Bedside teaching, Medical Students, Pediatric residents.

IJBC 2015; Supplementary; P 73
Paper ID: 32

Renal complications of sickle Syndrome.

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Abstract

Introduction: Sickle cell disease (SCD) is caused by inheritance of the sickle β globin gene, either in the homozygous form (SS), heterozygous (SA) or Sickle β-thalassemia (Sβ). Nephrologic complications are not well defined in subtypes of sickle cell syndrome especially in sickle thalassemia. Patients and Method: The research study takes a cross sectional analysis including all of the patients showing sickle cell syndromes in Hematology-Oncology Clinics affiliated to Shiraz University of Medical Sciences from 2012 to 2013. Ninety seven patients were enrolled in the study after taking informed consent. A random urine sample was taken from each of the patients for microalbuminuria, specific gravity, calcium, creatinine, urinalysis (U/A) and urine culture (U/C). Additionally, a blood sample was also sent for complete blood count (CBC), creatinine, BUN and Cystatin C. Glomerular Filtration rate (GFR) was measured by four methods of Schwart's, Modified Schwart's, Cystatine C and Filler's formula. Results: All 97 patients with mean age of 8.79 ±3.44 years enrolled in the research study and were categorized into three groups: 30 had SS with mean age of 9.6±3.84 years; 31 patients were SA with mean age of 8.27±2.28 year; and finally 36 had Sβ with mean age of 8.55±3.86 years. Enuresis was present in 22.9% patients who were older than 5 years, in the SS group 8/26; in the SA group 7/29; and in the Sβ group 5/32. Microalbuminuria was found in 14 patients. Sixteen patients had hypercalciuria. GFR was in normal range when calculated with creatinine clearance, but glomerular hyperfiltration was present in 56.7% of patients by Schwart's method, 3.1% of patients by modified Schwart's method, 7.2% by Filler's method and 5.2% by cystatin C formula. Mean serum creatinine was in low normal range (0.504±0.139 mg/dl) but mean serum cystatin C (0.925±0.126 mg/dl) was higher than the normal range especially in Sβ patients (p=0.023). Conclussion: Renal complications are not infrequent in sickle cell syndrome, so pediatric nephrology consultation for early detection and management of these complications is critical for prevention of end stage renal failure in adulthood.

Keywords: Sickle cell Anemia, Sickle Thalassemia, Sickle trait, Renal Dysfunction, Glumerular Filtration Rate.

IJBC 2015; Supplementary; P 74
Paper ID: 35

Effect of Isoprenaline on miR-23a Expression in Human Bone Marrow-Derived Mesenchymal Stem Cells

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Abstract

Introduction: MicroRNAs (miRNAs) are small noncoding RNAs that contribute to the control of gene expression. They can affect several cell regulatory pathways, such as cell growth, differentiation, mobility and apoptosis. miRNAs can regulate hematopoietic stem/progenitor cells (HSPC) by modulation of intrinsic cell components or extrinsic factors of the microenvironment were HSPC host. The chemokine Stromal Derived Factor-1 (SDF-1) is an essential factor for both homing and mobilization of Hematopoietic Stem Cells (HSCs) in the bone marrow. A transient increase in SDF-1 levels and following decrease was demonstrated during HSC mobilization induced by Granulocyte-Colony Stimulating factor (G-CSF). Recently, it was demonstrated that adrenergic signals of sympathetic nervous system (SNS) could mobilize HSCs and also miR-23a could potentially inhibit SDF-1 expression. Here, to evaluate the exact contribution of adrenergic signals on miR-23a expression, we treated human mesenchymal stem cells (hMSCs) with a non-selective -adrenergic agonist, Isoprenaline. Materials and Methods: MSCs were isolated and cultured from human bone marrow. The cells were treated with 100 µM isoprenaline and then total RNA was extracted at 12 and 48 hours after treatment, and also from untreated hMSCs as a control. Then, cDNA was generated and miR-23a expression level was quantified using High-Specificity miRNA QPCR Kit. Results: The expression level of miR-23a was decreased significantly at 12 hours post treatment (Ratio=0.2 fold; P

Keywords: mobilization, SDF-1, miR-23a, Isoprenaline

IJBC 2015; Supplementary; P 75
Paper ID: 36

β-Adrenergic Agonist of Isoproterenol Induces VTRNA2-1 Expression in Human Bone Marrow Mesenchymal Stem Cells

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Abstract

Background and Objective: Mobilization of bone marrow–derived hematopoietic stem cells (HSCs) and their progenitors into the peripheral circulation with Granulocyte-Colony Stimulating factor (G-CSF) alone, or in combination with cyclophosphamide, represents the basis for modern bone marrow transplantation procedures. However, the mechanism is not fully understood. β-adrenergic signals from sympathetic nervous system (SNS) recently emerged as a novel regulator of HSC egress from the BM; but, little is known about the exact role of β-adrenergic signals in induction of this process and factors influenced by these signals. The chemokine Stromal Derived Factor- 1 (SDF-1) which is expressed by human bone marrow-derived mesenchymal stem cells (hMSCs), has a key role in mobilization of HSCs. In addition, it was demonstrated that a noncoding RNA (ncRNA) called vault RNA 2-1 (VTRNA2-1) can also regulate the expression of this chemokine in hMSCs. In this study, to investigate the role of VTRNA2-1 in mobilization process, the expression of VTRNA2-1 was evaluated in hMSCs treated by Isoproterenol (a β-adrenergic agonist). Materials and Methods: MSCs were isolated and cultured from human bone marrow. After doing flowcytometric analysis, the cells were treated with 100 μM Isoproterenol. Total RNA was extracted at 12 and 48 hours post treatment, and also from untreated hMSCs as a control. Then VTRNA2-1 expression level was quantified by quantitative Reverse Transcriptase PCR. Results: The expression level of VTRNA2-1 was increased significantly at 12 (Ratio=3.5 fold) and 48 (Ratio=75.2 fold) hours post treatment in comparison with control (P

Keywords: Bone Marrow Transplantation, Mobilization, VTRNA2-1, Isoproterenol

IJBC 2015; Supplementary; P 76
Paper ID: 38

Prognostic significance of 11q23 in Acute Lymphocytic Leukemia children after initial response to chemotherapy on the seventh day

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Abstract

Acute lymphoblastic leukemia (ALL) is the most common form of childhood cancer and lead to cancer-related death in children. The majority of infant with ALL harbor recurring structural chromosomal rearrangements that are important initiating events in leukemogenesis but are insufficient to explain the biology and heterogeneity of disease. Mixed-lineage leukemia-rearranged (MLL-rearranged) at 11q23 occurs in at least two-thirds of infants with ALL. The most common MLL rearrangements are t(4;11) (q21;q23)/MLL-AFF1 (AF4) that found in approximately 50% of cases. In this study, we have investigated 11q23 rearrangement and its relationship with other prognostic factors such as age, sex, organomegally, immunophenotype and therapeutic response. The results show that persons with the 11q23 translocation had blast more than 5% (P = 0.002). There was a significant correlation between the 11q23 translocation with an initial response to chemotherapy. Kay words: Acute lymphoblastic leukemia, 11q23 translocation, Molecular cytogenetic.

Keywords: Acute lymphoblastic leukemia, 11q23 translocation, Molecular cytogenetic

IJBC 2015; Supplementary; P 77
Paper ID: 39

Normal HbF, Delta beta-Thalassaemia in south of Iran

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Abstract

A couple of deletional delta beta-thalassaemia with unexplained normal HbF was found during premarriage screening programme in Shiraz. The man a 26year-old man, showed a good clinical picture ,planed marriage with relative(first cousin)21 year old woman. Lab data of man: Hb:11.6,RBC:6680000,MCV:58.2,MCH:17.4,HbA2:2.0,HbF:0.3 Lab data of woman: Hb:10.9, RBC:5500000,MCV:59.9,MCH:17.8,HbA2:2.0HbF:0.3 First for couple was done alfa globulin GAP PCR,reverse dot blot and MLPA for ruled out commom alfa thalassemia gene and also alfa thalassemia gene triplication confirmed normal genotype in both them. Then for couple was done beta gene analysis by MLPA and confirmed delta beta thalassemia carrier in both them. According above results for ruled out major beta thalassemia in fetus , essentially prenatal diagnosis for beta gene analysis by MLPA by chrionic villi sampling in 11-12 weeks of pregnancy is recommended.

Keywords: Delta beta-Thalassaemia ,Normal HbF,south of Iran

IJBC 2015; Supplementary; P 78
Paper ID: 40

Effect of high dose of intravenous Deferoxamine on regional myocardial function in patients with beta thalassemia major by using Strain Imaging Echocardiography.

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Abstract

Introduction: Iron over loading can cause multi organ failure including heart in beta thalassemia major patients. Congestive heart failure is the major cause of death in these patients and chelating therapy can reduce the risk. Strain and Speckled echocardiography can use as a modality to see changes in left ventricular segmental and global myocardial function. The aim of this study was evaluation of the impact of high dose Deferoxamine therapy on lowering serum ferritin and the effect of this type of treatment on myocardial function by Strain and Speckled echocardiography in a short time period follow up.
Method: This study included 21 patients with beta thalassemia major who received 50-80mg/ kg intravenous Deferoxamine for 5days were included in the study from Feb 2013 till Feb 2015.Two-dimensional, M-mode, Doppler and Speckled imaging echocardiography was done before high dose of chelating therapy and 3 months after that.
Results: Ejection fraction was improved after therapy (P
Conclusion: High dose of intravenous Deferoxamine can reduce serum ferritin in patients with beta thalassemia major and improve myocardial systolic function. Strain imaging of epicardium and endocardial surface of left ventricle showed improvement of systolic strain of various segments of left ventricle except apical septal LV wall in 3 months follow up and we concluded that high dose Deferoxamine is effective in lowering serum ferritin and results in improvement of left ventricular function in short time after such therapy and strain imaging echocardiography is a sensitive method to detect the changes in systolic myocardial left ventricular function in a short time period of follow up.
Keywords: Beta thalassemia major, Strain imaging, Deferoxamine, Myocardial function

IJBC 2015; Supplementary; P 79
Paper ID: 42

The Effects of Isoprenaline on SDF-1 Expression in Human Bone Marrow Mesenchymal Stem Cells

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Abstract

Introduction: A pathway critical to the homing and retention of Hematopoietic Stem Cells (HSCs) within the bone marrow (BM) is the CXCR4/CXCL12 chemotactic axis. The chemokine stromal cell–derived factor-1 (SDF-1) is a potent chemo attractant for primitive bone marrow CD34+CD38– cells that include candidate HSCs and express the CXCL12 receptor, CXCR4. SDF-1 is highly expressed in human bone marrow endothelium, reticular cells, endosteal osteoblasts and mesenchymal stem cells (MSCs). It is clear that neurotransmitters can serve as important inducers of mobilization and treatment with norepinephrine results in HSC mobilization. In this study, the effect of Isoprenaline as a beta-adrenergic agonist on SDF-1 expression of human bone marrow MSC was evaluated for the first time. Materials and Methods: MSCs were isolated from human BM samples and cultured in DMEM plus 15% FBS until passage 3. Then the cells were treated in the presence of 100 µM Isoprenaline for 12 and 48 hours. RNA was extracted using Trizol and cDNA synthesized. After setting up RT-PCR for SDF-1, the quantization was performed using qRT-PCR method by \

Keywords: Mesenchymal stem cells, SDF-1, Isoprenaline

IJBC 2015; Supplementary; P 80
Paper ID: 43

The Effects of Beta-Adrenergic Agonist on MMP-9 Expression of Human Bone Marrow Derived Mesenchymal Stem Cells in vitro

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Abstract

Introduction: A complex interplay of cytokines, chemokines, proteolytic enzymes and adhesion molecules maintain hematopoietic stem cells (HSC) anchorage to the niche infrastructure. Proteins of the matrix metalloproteinase (MMP) family are involved in the breakdown of extracellular matrix in normal physiological processes, such as embryonic development, reproduction, and tissue remodeling, as well as in disease processes, such as metastasis. Activation of proteolytic enzymes such as matrix metalloproteinase-9 (MMP-9) results in degradation of anchorage proteins, enabling migration of HSCs from the osteoblastic to the vascular niche. Recently, it was determined adrenergic signals serve as direct chemo attractants to HSPCs and treatment with norepinephrine could mobilize HSCs. The present study, for the first time, was carried out to determine the effect of Isoproterenol as a beta-adrenergic agonist on MMP-9 expression in human bone marrow mesenchymal stem cells (hMSC). Materials and Methods: MSCs from human BM was isolated and cultured in DMEM plus 15% FBS until passage 3. Then the cells were treated with 100 µM isoproterenol as a beta-adrenergic agonist for 48 hour. Total RNA was extracted using Trizol and cDNA synthesized. After setting up RT-PCR for MMP-9, the quantization was performed using qRT-PCR method by \

Keywords: mesenchymal stem cells, MMP-9, Isoproterenol

IJBC 2015; Supplementary; P 81
Paper ID: 44

The relationship between characteristics of parents of children with cancer and their posttraumatic stress symptoms

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Abstract

Abstract Support of parents of children with cancer requires health care personnel to be knowledgeable about the relationship between characteristics of parents of children with cancer and their posttraumatic stress symptoms. This study thus was conducted to fulfill this aim in South-East of Iran. Using the Impact of event Scale- Revised (IES-R) for parents of children with cancer, 200 parents of these children in two hospitals supervised by Kerman University of Medical Sciences was assessed. The Impact of Event Scale-Revised (IES-R) is a 22-item questionnaire that assesses psychological symptoms that occurred within the past week associated with a specific traumatic event. Based on our previous research and because all families experienced it, the child□s diagnosis with cancer was selected as the traumatic event. Each item presents a symptom, and participants use a weighted four-point scale to rate the frequency with which they experienced that symptom during the past week. Three subscale scores, intrusive thoughts, avoidance, and arousal, are obtained. Descriptive statistics (frequency, percentage, mean, and standard deviation) were used to describe the study sample characteristics. The total mean of IES-R was higher among parent of cancerous child with age ranged between 0-1 years compared to the others (mean=2.52, p=0.002, SD=0.78). In this study the mean of category of “hyperarousal” (mean=2.32) was higher in parents of son with cancer (p=0.03, SD=0.76) compared to other. The means of category of „hyperarousal” (mean=2.85, p=0.01, SD=1.09), „intrusion” (mean=2.55, p=0.02, SD=1.00) and “avoidance” (mean=2.23, p=0.02, SD=0.64) were higher among parent of cancerous child with age ranged between 0-1 years compared to the others. The mean of avoidance was higher in parents who they were emotionally supported by his/her wife (mean=1.83, p=0.03, SD=0.81). The mean of category of “avoidance” was lower in parents who they children with cancer had aware about her/his illness (mean=1.3, p=0.01, SD=0.51). There were association between socio-demographic data and posttrumatic stress symptoms. More study is needed to elucidate the Iranian parents□ experience of having children with cancer.

Keywords: Parents of cgildren with cancer, Pediatric oncology, posttrumatic stress syptoms

IJBC 2015; Supplementary; P 82
Paper ID: 45

Psychosocial risks among Iranian parents of children during cancer treatment

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Abstract

Abstract Support of parents of children with cancer requires health care personnel to be knowledgeable about the relationship between characteristics of parents of children with cancer and their psychosocial risks. This study thus was conducted to fulfill this aim in South-East of Iran. Using the Psychosocial Assessment Tool- Revised (PAT-R) for parents of children with cancer, 200 parents of these children in two hospitals supervised by Kerman University of Medical Sciences was assessed. The PAT-R contains subscales such as family problems (consist of 12 questions; Item Response Anchors are 0= no, 1= Yes), parent stress reactions (consist of 3 questions; 0=Not at All, 1= Sometimes, 2= Often, 3= Very Much), family beliefs (consist of 10 questions; 0=Not at all, 1= true Just a little bit true, 2= Pretty much true, 3= Very true), child Problems (consist of 17questions; 0=Never a Concern Sometimes, 1=a Concern Currently, 2= Receiving Help) and sibling Problems (consist of 17 questions; Item Response Anchors are 0= no, 1= Yes). Descriptive statistics (frequency, percentage, mean, and standard deviation) were used to describe the study sample characteristics. In PAT-R, the total mean score was 0.56. A difference was found between parents in category of \

Keywords: Parents of children with cancer, pediatric oncology, psychosocial risks

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Paper ID: 46

Association between iron deficiency anemia and first febrile seizure in children

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Abstract

Association between iron deficiency anemia and first febrile seizure in children
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Objective
Considering the high prevalence of febrile seizure in children and controversial results in present day literature regarding the relationship between febrile seizures and anemia, this study was conducted to evaluate the association between first pediatric febrile seizure and anemia in children between 6 to 60 months old.
Material and Methods: In this case-control study, conducted in 2009, 80 children with febrile seizure(cases) and 80 febrile children without seizure control groups were male, and twenty nine (50%) of the patients in case and control groups were female. Patients were divided in to five subgroups in terms (controls) were evaluated in Sanandaj,Besat hospital; all patients were matched for age, sex, and use of supplemental iron.fifty one (50%) of patients in both case and of age in months. Levels of serum Iron, Total Iron Binding Capacity(TIBC),Ferritin,Hemoglobin, Hematocrit, Platelets count and red blood cell indices(MCV, MCH, MCHC) were determined in all children by Automatic Analizer. Using Iron in last three days, family history of febrile seizure, Temperature degree, and etiology of Fever were analyzed statistically by SPSS, T-TEST, Chi-square and Fisher exact tests were used to analyze data.
Results: Of the case group, 70% (28 patients) and of controls, 30% (12 patients) of children had anemia (p= 0.003). Febrile seizures were found to occur mostly between the ages of 9 to 37 months. There was a considerable relation between febrile seizure occurrence and positive family history of febrile seizure (0.001). Hematologic indices such as, MCV, MCHC and TIBC in both groups (case and control) were different from eachother. But, Hemoglubin, MCH, serum Iron, Ferritin, Platelets count in both groups were not different. Gastroentritis (31.3%) and Upper Respiratory Tract Infection (28.8%) were the most common cause of febrile seizure.
Conclusion: The risk of febrile seizure occurrence in anemic children seems to be higher than that in children who do not suffer from the condition.
Keywords: Febrile seizure, Anemia, Iron deficiency

IJBC 2015; Supplementary; P 84
Paper ID: 47

Development in the management of congenital factor XIII deficiency with factor XIII concentrate

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Abstract

Introduction and objective: Clinical factor XIII (FXIII) deficiency is a rare, inherited, autosomal recessive coagulation disorder. The aim of this case report is to introduce a case FXIII deficiency with late diagnosis, management with fresh frozen plasma, then development in the management with FXIII concentrate for prophylaxis, successful pregnancy, labour and delivery. Material and method:The patient is a 21 year-old girl. Shesuffered from frequent muscular pain since childhood.Diagnostic tests including PT, PTT, INR, and BT were normal in several times. So her disease remained unnoticed until 14 years old due to injury to her left knee and hemarthrosis and hematoma. She hospitalized and after coagulating test containing ureaclot lysis test, her disease diagnosed as factor XIII deficiency. After diagnosis, she received FFP in outpatientdue to muscular pain and hematuriamonthly. FFPhas been used substituted by FXIII concentrate (fibrogammin® P CSL-behring)from 2 years ago.She received regular prophylaxis with 10 IU/kg FXIII monthly. She got pregnant at November 2011.She received regular prophylaxis with FXIII concentrate every 2 weeks until the end of 6th month of pregnancy and then every 4 weeks until the end ofpregnancy.Result: One mild bleeding episode occurred during pregnancy.She operated with caesarian section successfully.The birth weight of neonatal was 2400 g and was healthy.Urea clot lysis test in infantwas normal in 2 month after birth. Conclusion: FXIII deficiency is associated with severe life threatening bleeding, intracranial hemorrhage, impaired wound healing and recurrent pregnancy losses. The algorithm provided for FXIII diagnosis and classification will enable prompt identification and early intervention for controlling potential life threatening complications. In most part of the word, cryoprecipitate and plasma transfusion are the only treatment available. Management developments have revealed the effectiveness and safety of FXIII concentrate for prophylaxis and treatment.

Keywords: Factor XIII, prophylaxis

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Paper ID: 48

Links between coagulation and polyphosphate

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Abstract

Links between coagulation and polyphosphate Hassan Mansouritorghabeh Allergy Research Center, Ghaem Hospital, Mashhad University of Medical Sciences, Mashhad, Iran. Email: Mansouritorghabeh@mums.ac.ir, Tel:+98(513)8012830, Fax:+98(513)8458769 Introduction Polyphosphate is a linear strand of phosphate polymer that connected each other using high energy bonds. It plays as energy reservoir in micro-organisms, as chaperon, and a defense against toxicity of heavy metals. It can be find in dens granules of platelets and release after platelets activation. The patients with abnormal dense granules, there is prone to haemorrhagic events. On the other hands, recent surveys have demonstrated effects of polyphosphate as pro-hemostasis, pro-thrombotic and pro-inhibitory effects of molecule. Here you will find recent advances in roles of polyphosphate in coagulation. Methods To gathering recent contributions on polyphosphate and coagulation, a review of literate was done in PubMed medical search engine using the key words of:"polyphosphate"+"coagulation', "polyphosphate"+"hemostasis", "polyphosphate"+"haemostasis" and "polyphosphate"+"platelets" without any language and time limitations. Results After literature review, overall 22 papers selected and full texts of them retrieved. It has been showed that it influences coagulation via several mechanisms inter alia: First, polyphosphate activates contact pathway, Kalikerin-Kinin pathway, which results to release of bradykinin. It is a vaso-active peptide that after attachment to endothelial cells can promote release of nitric oxide (NO), prostacyclin and a vasodilator entitled endothelia –derived hyperpolarizing factor. Second, it can activate factor V and in collaboration with factor Xa increases thrombin production. Also it can robust fibrin clot and its stability and structure firmness. Third, polyphosphate inhibits tissue factor pathway inhibitor (TFPI). Conclusion Polyphosphate enhance thrombin generation, inhibits anticoagulant activity of heparins, thrombin inhibitors and inhibitors of factor X and shortens formation of blood clot in plasma of hemophilia A & B and finally inhibits antagonist of vitamin K. So, it can be use as potential topical hemostatic mediator in future. Also inhibiting of polyphosphate may be regarded as a novel strategy for preventing thrombosis and inflammation process in future.

Keywords: Hemostasis, polyphosphate, thrombosis, inflamation

IJBC 2015; Supplementary; P 86
Paper ID: 49

Effect of sodium bicarbonate and sodium chloride on renal and hematologic factors in patients with glucose-6-phosphate dehydrogenase deficiency

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Abstract

Introduction: Serum with sodium bicarbonate is one of the medicines that is used for compensation bicarbonatelost and increasing blood pH in metabolic acidosis caused by severe anemia in patient with glucose-6-phosphate dehydrogenase deficiency (G6PD). Some previous studies demonstrate that bicarbonate has some side effects. In this proposal, the aim was comparison the effect of two serum therapies (serum with bicarbonate and without bicarbonate) on some renal and hematologic factors and their side effects in patients with hemolysis caused by glucose-6-phosphate dehydrogenase enzyme deficiency referred to Gorgan Taleghani Hospital. Materials andMethod: In this clinical trial study, 79 patients with Favism were evaluated. The patients randomlyput into two treatment groups, including serum with sodium bicarbonate and serum without bicarbonate (containing sodium chloride), during treatment, patients received blood based on hemoglobin. In both groups, paraclinical and clinical factors including duration of hospitalization, times of Blood transfusion, received blood volume, duration of cleaning UA of hemoglobin, hemoglobin, urine pH and granular casts in UA were evaluated and analyzed. **Results:** The mean age of total 79 patients was 51.22 ± 37.86 months and there were 58 (73.40%) males and 21 (26.60%) females in this investigation. of all studied patients, just one of them has hemoglobin more than 9 mg/ml and didn't receive blood. Among evaluated factors, only duration of hospitalization and urine pH statistically showed a significant difference between two treatment groups, and other factors were statistically almost identical. Conclusion: the result showed that Sodium chloride is more effective than sodium bicarbonate in reducing the duration of hospitalization and with regard to a little clinical difference in received blood volume and duration of cleaning UA of hemoglobin; it suggests that sodium chloride can have a greater impact on improvement of hemolysis caused by G6PD deficiency. The absence of side effects in two therapy groups can be due to controlled injection method and the concentration of soluble drugs as well as the limitation of studied population.

Keywords: G6PD, hemolytic anemia, hemolysis, Favism

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Paper ID: 51

Report on Patients with Non Transfusion-Dependent b-Thalassemia

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Abstract

Hydroxyurea (HU) has been used to treat patients with non transfusion-dependent b-thalassemia major (b-TM) at the Thalassemia Research Center, Sari, Mazandaran Province, Islamic Republic of Iran since 1996. This study was performed to summarize and to share our experience. Medical records of all patients with b-thalassemia (b-thal) attending our center were reviewed in January 2013. Definition of b-TM was based on complete blood count (CBC), hemoglobin (Hb) electrophoresis, and for some patients, by the amplification refractory mutation system-restriction fragment length polymorphism (ARMS-RFLP) method. Patients who had not been transfused before, or had only occasionally had blood transfusions, were selected. Age at first blood transfusion, initial HU therapy and time of study was extracted from the records. The lowest Hb level before using HU and the last Hb value when on the HU regimen as well as the difference, were reported. Number of saved packed red cells was calculated according to duration of HU use and the usual needs of the patients. Hydroxyurea was discontinued before a planned pregnancy and during gestation and lactation periods. Hydroxyurea was discontinued for male patients willing to reproduce. A p value of50.05 was considered statistically significant. It was consistent with 1856 patients/ year, and 3542 units of blood were saved. We found HU to be effective and safe in treating patients with non transfusion-dependent b-TM. We strongly recommend HU therapy.

Keywords: b-Globin gene mutation, compliance, hemoglobin (Hb), hydroxyurea (HU), Iran, non transfusion-dependent b-thalassemia major (b-TM), treatment

IJBC 2015; Supplementary; P 88
Paper ID: 54

CXCR7 Is Expressed in B Acute Lymphoblastic Leukemia (ALL) Cells and Mediates Their Transendothelial Migration

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Abstract

The function of chemokine stromal cell-derived factor (SDF)-1 and its receptor CXCR4 in the pathobiology of hematological malignancies is known, but the role of another, newly identified receptor for SDF-1, RDC1/CXCR7, has not yet been defined. Thus we investigated whether CXCR7 plays a role in hematological malignancy. Using RT-PCR and flow cytometry analysis, we screened the expression of CXCR7 in 14 hematological cell lines and found that it is strongly expressed in 4 out of 5 B cell lines tested (Raji, NC-37, NALM6, Ramos and REH) and weakly in two T cell lines(Jurkat and CEM) and 2 of 7 myeloid cell lines tested. Moreover, CXCR7 was also strongly expressed in all 9 patients diagnosed with B precursor ALL as detected by flow cytometry on peripheral blood cells and Immunohistochemistry on bone marrow biopsy. In contrast, its expression was very weakly in normal lymphocytes, and not at all in T precursor ALL samples, suggesting that CXCR7 is selectively expressed in B-cell ALL. Next we examined whether CXCR7 plays a role in the chemotaxis of these cells and found that CXCR7 antagonist did not inhibit SDF-1-induced chemotaxis, indicating that CXCR7 does not play a significant role in the chemotactic activity of B-cell ALL. However, we found that CXCR7 antagonist inhibited the transendothelial migration (TEM) of B cell lines and primary B precursor ALL cells in a dose-dependent manner. Moreover, transfection of the NALM6 with CXCR7 resulted in downregulation of CXCR4 and the TEM of such transfected cells was more inhibited by CXCR7 antagonist than of wild-type cells. When we compared the roles of CXCR4 and CXCR7 in the TEM of these cells, we found that the CXCR7 antagonist totally inhibited the TEM of B cells at the low dose of 1?M, but the CXCR4 antagonist AMD3100 did not. Moreover, CXCR7 antagonist significantly prevented binding of NC-37 cell line to HUVEC (as shown by adhesion assay) which indicates that CXCR7 is likely required for the interaction of leukemic cells with endothelial cells and their extravasation. In conclusion, we demonstrated that CXCR7 is expressed on B lymphoblasts, and the SDF-1/CXCR7 axis plays a crucial role in the trafficking of these cells, indicating that CXCR7 could be a therapeutic target in patients diagnosed with B precursor ALL.

Keywords: Acute lymphoblactic Leukemia

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Paper ID: 57

Lung involvement in thalassemia

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Abstract

Lung involvement in thalassemia
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Objectives: The aim of this study was to determine the spirometric pattern and its correlation to iron overload in beta thalassemia.
Methods: We studied 40 thalassemic patients with ages ranging from 8 to 29 years. Pulmonary function studies were performed 2 days prior to blood transfusion and were considered pathologic when they fell below 80% of the predicted value.
Result: Twenty two patients (55%) had normal lung function. Respiratory abnormalities were detected in 45% patients. A predominantly restrictive pattern was seen. No significant difference was observed between gender, serum ferittin and hemoglobin PFT'S. There was a significant linear correlation between age and serum ferritin level (P
Conclusion: Restrictive disease are the prominent abnormalities of pulmonary function in thalassemic patients. The deposition and prolonged accumulation of iron didn't play a major role in the pathogenesis of the pulmonary disease, so subsequent studies are essential for obtaining greater conception into pathogenesis of lung dysfunction.
Key words: Thalassemia, spirometry, restrictive disease, ferritin

IJBC 2015; Supplementary; P 90
Paper ID: 58

Cord blood-hematopoietic stem cells proliferation in 3D fibrin scaffold

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Abstract

Background: The use of umbilical cord blood as a source of hematopoietic stem cells for the treatment of over 80 disorders has been established. Given the general availability, the ease of procurement, and progenitor content, cord blood is an attractive replacement for bone marrow. Expansion and proliferation of undifferentiated cord blood- hematopoietic stem cells in vitro is limited and insufficient. Most of the in vitro cell culture systems currently used for expansion and proliferation of hematopoietic stem cells are two dimensional (2D) culture systems. It is important that three dimensional (3D) scaffolds, providing appropriate environment for proliferation and expansion of hematopoietic stem cells. The aim of present study is to use the fibrin 3D biomaterial as a suitable environment for in vitro expansion and proliferation of cord blood- hematopoietic stem cells.
Material and methods: In this study, hematopoietic stem cells were isolated from freshly umbilical cord blood and were recovered by hydroxyl ethyl starch (HES) and phosphate buffer saline (PBS). Also, fibrin glue biomaterial was prepared. Isolated cells were cultured separately on the 3D fibrin glue scaffold and 2D culture environment as control and 2 weeks after expansion, cell proliferation ability was evaluated using MTT assay.
Results: Our findings demonstrated that the cell proliferation of cord blood- hematopoietic stem cells is enhanced in a 3D fibrin scaffold compared to control. This difference was statically significant (p Conclusion: According to the results of the study, it can be said that natural scaffolds such as fibrin can be used as an appropriate environment for hematopoietic stem cells proliferation.
Keywords: cord blood- hematopoietic stem cells, 3D fibrin scaffold, proliferation, MTT assay

IJBC 2015; Supplementary; P 91
Paper ID: 68

The Dramatic effect of vitamin D in improving of ventricular dysfunction in Major thalassemia

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Abstract

Background: Low vitamin D in linked to decreased cardiac function and refractory congestive heart failure. This study aims to investigate effectiveness of high dose vitamin D in improving left ventricular ejection fraction in thalassemia patients with heart failure and vitamin D deficiency. **Materials and Methods:** This was a case-control study of 26 chronically transfused thalassemia patients and ventricular dysfunction with frank vitamin D deficiency between February 2015 and April 2015. Median age of the patients was 20.52± 5.39 years (range, 12.00-30.00 years). A serum vitamin D25-OH levels less than 20 ng/dl was considered frank vitamin D deficiency in our study. An ejection fraction less than 55% indicated poor pump function. The participating patients received 100,000 IU of vitamin D2 weekly for 8 weeks. LV ejection fraction and serum 25 OHD levels data was compared before and after completing treatment. Also, adverse effects were recorded during the study. **Results:** 24 patients were eligible in our study. Two patients had hypoparathyroidism who were treated with calcitriol and vitamin D supplements. Mean serum vitamin D25-OH levels, before and after study, were 11.20±4.80 ng/dl and48.58±13.65 ng/dl, respectively (p=0.03). Mean LV ejection fraction, before and after study, were 22.42±7.56% and 59.83±6.13%, respectively (p=0.03). Mean serum total calcium levels, before and after study, were 8.82±1.02 mg/dl and 10.42±1.56 mg/dl, respectively (p=0.6). Mean serum ferritin levels were 3760±1599 ng/dl (range, 1200-7100 ng/dl). Mean cardiac T2* of the patients were 12.63±6.25 ms. PTH levels of the participating patients were in normal range values (reference value, 15-60 pg/dl). Two patients had constipating and one patient had abdominal pain during the study. All AE were transient. **Conclusion:** Results showed that vitamin D (100,000 IU, weekly for 8 weeks) was effective and safe in improving LV ejection fraction and cardiac dysfunction in major thalassemia patients with vitamin D deficiency. Because of this, major thalassemia patients especially those with ventricular dysfunction should have their vitamin D levels assessed, and replacement should be started if these levels are low.

Keywords: Vitamin D, Thalassemia, Ventricular dysfunction

IJBC 2015; Supplementary; P 92
Paper ID: 75

The effectiveness of Gcsf in remission of chemotherapy induced febrile neutropenia , fever , and hospitalization period in ALL pediatric patients before starting intesification

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Abstract

Abstract Objective : chemotherapy –induced neutropenia is life threating and common side effects in treating of ALL. It predispose to serious infections ,fever ,prolonged hospitalization and delay in continuation chemotherapy and at last relapse of leukemia. The aim of present study was to assess of effectiveness of G_csf in prophylactic starting before intensification of ALL chemotherapy to prevent of severe neutropenia **Materials and methods :** It was prospective randomized clinical study carried out on 68 patients with ALL who received a standard intensification with prednisolone ,cytosar ,doxorobiocine ,etoposide ,and VCR in our children hospital of Tabriz .A random group of 32 patients received Gcsf 10 mic/kg/day for 3 days before starting intesification and and 36 of patients didn,t . Results :patients randomized to Gcsf had longer and more severe neutropenia (median700 v 1280 mm3) and hospitalization (median 5.1 v 3.2) and delay in starting maintenance Conclusion :Adding Granulocyte colony stimulating factor (Gcsf) thrapy before starting intensification not only induce severe neutropenia but also duration neutropenia and cause of delay continuation maintenance. Keywords: acute lymphoblastic leukemia .Gcsf .neutropenia

Keywords: acute lymphoblastic leukemia .Gcsf .neutropenia

IJBC 2015; Supplementary; P 93
Paper ID: 79

Potential benefits of silymarin in β-thalassemia major patients

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Abstract

Silymarin is mixture of flavonolignans extracted from milk thistle (Silybum marianum) plant that has long been used for its antioxidant and hepatoprotective properties. Generally it considered as a liver tonic and has been used to treat acute and chronic liver disease. It also is used for other conditions such as gallbladder disorders, certain types of cancer, and is considered to be protective against diabetes and cardiovascular diseases. A common form of silymarin that is used in most clinical trials is silybin extracts produced by Legalon (Madaus Rottapharm, Cologne, Germany) which contains 52 mg silybin, 22 mg isosilybin, 23 mg silycristin and 28 mg silydianin. Recently researchers have working on silymarin as an iron chelating in thalassemia patients. This substance in addition to iron chelating effect has potential benefits such as antioxidant, Immunoregulatory, antineoplastic, hepato and cardioprotective properties in thalassemia patients. In this paper we discussed about different properties of silymarin and compared silymarin versus other flavonoids and conventional iron chelating agents in thalassemia patients.

Keywords: Antioxidant, Thalassemia, Silymarin

IJBC 2015; Supplementary; P 94
Paper ID: 80

Antiprolifrative effect of valporic acid in leukemic cells in mouse xenograft model

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Abstract

Introduction: The short chain fatty acid valproic acid (VPA, 2-propylpetanoic acid) is approved for the treatment of epilepsia, bipolar disorders and migraine and clinically used for schizophrenia. Currently, VPA is examined in numerous clinical trials for different leukaemias and solid tumour entities. A new generation of HDAC inhibitors is becoming an increasing focus of attention for their ability to induce differentiation and apoptosis in tumor cells and to block the cell cycle. Abnormal expression of cyclin-dependent kinase inhibitors, especially p21, is considered a possible mechanism of the arrested maturation and differentiation seen in leukemia cells. In this study, we evaluated the anticancer effects of VPA on Kasumi-1 cells in acute myeloid leukemia in mouse xenograft tumor model. Methods: flowcytometry were carried out to appraise the effect of VPA on cell viability and apoptosis respectively. Gene expression of p21 and cyclin D1 was measured using Reverse transcriptase PCR and for detection of cyclin D1 and E2F protein expression western blotting were carried out. All the experiments were performed in triplicate and the data are shown as mean ± SD. Statistical significances of difference throughout this study were calculated using a Student's t-test and by one-way variance analysis. P values ‹0.05 were considered significant. Result: In this study we had demonstrated that valproic acid induces G0/G1 arrest of Kasumi-1 cells in acute myeloid leukemia. Our research confirmed that valproic acid inhibits the growth of Kasumi-1 cells in a mouse xenograft tumor model. this occurs via upregulation of histone acetylation in the p21 promoter region, enhancement of p21 expression, suppression of phosphorylation of retinoblastoma protein, blocking of transcription activated by E2F, and induction of G0/G1 arrest. Conclusion: VPA, alone or in combination with other drugs, could be a potent anticancer agent for targeting gastric cancer

Keywords: Valproic acid (VPA), Leukemia, p21, apoptosis, histone deacetylases

IJBC 2015; Supplementary; P 95
Paper ID: 81

Synergistic effect of Valporic acid and ATRA in leukemia cell lines trough restores RARβ2 expression

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Abstract

Introduction: Valproic acid (VPA, 2-propylpetanoic acid) is approved for the treatment of epilepsia, bipolar disorders and migraine and clinically used for schizophrenia. Currently, VPA is examined in numerous clinical trials for different leukaemias and solid tumor entities. A new generation of HDAC inhibitors is becoming an increasing focus of attention for their ability to induce differentiation and apoptosis in tumor cells and to block the cell cycle. All-trans retinoic acid (ATRA) is a potent regulator of cellular growth and differentiation, including normal epithelial cell differentiation, it can function as a chemopreventive agent and is an effective inhibitor of chemical and viral carcinogenesis. Resistance to the growth-inhibitory action of retinoic acid (RA), the bioactive derivative of vitamin A, is common in human tumors. One form of RA resistance has been associated with silencing and hypermethylation of the retinoic acid receptor β2 gene (RARβ2), an RA-regulated tumor suppressor gene. In this study, we evaluated the combination effects of VPA and ATRA on in acute myeloid leukemia cell lines invitro. Methods: MTT assay and flowcytometry were carried out to appraise the effect of VPA, ATRA and combination of them on cell viability and apoptosis respectively. Gene expression of RARβ2 was measured using Real Time PCR. All the experiments were performed in triplicate and the data are shown as mean ± SD. Statistical significances of difference throughout this study were calculated using a Student's t-test and by one-way variance analysis. P values ‹0.05 were considered significant. Result: In this study we had demonstrated that VPA and ATRA separately induces apoptosis and inhibit the growth of leucemic cells lines. In combination we see synergistic effect between VPA and ATRA. This synergism seems to be occurs via upregulation of RARβ2 expression. Conclusion: VPA and ATRA, alone or in combination, could be a potent anticancer agent for targeting gastric cancer.

Keywords: Valproic acid (VPA), Leukemia, RARβ2, apoptosis, ATRA

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lovastatin induce differentiation and apoptosis in acute myeloid leukemia cells through down regulation of bcl2

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Abstract

Introduction: Lovastatin is a specific, nonreversible competitive inhibitor of HMG-CoA reductase that use as a treatment for hypercholesterolemia. Recent studies shows Lovastatin, induced a retinoic acid-like differentiation response followed by extensive apoptosis in neuroblastoma cell lines at relatively low concentrations of this agent. In this study, we examined the ability of lovastatin to induce apoptosis in acute myeloid leukemia cell lines invitro. Methods: flowcytometry and MTT assay were carried out to appraise the effect of lovastatin on cell viability and apoptosis respectively. Gene expression of bcl2, CD11b and CD18 was measured using Real Time PCR. All the experiments were performed in triplicate and the data are shown as mean ± SD. Statistical significances of difference throughout this study were calculated using a Student's t-test and by one-way variance analysis. P values ‹0.05 were considered significant. **Result:** In this study we had demonstrated that lovastatin induces G1/S arrest in leukemic acute myeloid leukemia cells. Our research confirmed that lovastatin inhibits the growth of acute myeloid leukemia cells, and that this occurs via downregulation of bcl2 and increase expression of CD11b and CD18. Conclusion: lovastatin exposure of acute myeloid leukemia cells induced differentiation response and apoptosis that may contribute to the therapeutic potential of this agent in the treatment of this disease. Lovastatin could be a potent anticancer agent for targeting acute myeloid leukemias.

Keywords: Valproic acid (VPA), Leukemia, p21, apoptosis, histone deacetylases

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Paper ID: 83

Metformin induces apoptosis in leukemia cells through AMPK-dependent or independent mechanism

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Abstract

Introduction: Metformin is an oral drug traditionally used for the treatment of many diseases including type 2 diabetes1 and polycystic ovary syndrome2 and belongs to biguanides. recent studies have demonstrated that metformin could induce apoptosis and inhibit proliferation of many cancer cell types in vitro and in vivo. Metformin is well-tolerated at very high doses, and its safety has also been demonstrated in clinical trials in diabetic children. Acute myeloid leukemia (AML) accounts for 15-20% of childhood leukemias. Although remission is achieved following treatment with front-line chemotherapy, nearly half of the patients are faced with disease relapse associated with chemoresistance. Therefore, therapies that could sustain the remission phase in pediatric AML are urgently needed. Methods: cell viability and apoptosis were estimated trough MTT assay and flowcytometry respectively. The activation status of AMP-activated protein kinase (AMPK) was detected by immunoblotting for phosphorylated (T172) AMPKα subunit. Gene expression of mTOR and surviving was measured using Real Time PCR . All the experiments were performed in triplicate and the data are shown as mean ± SD. Statistical significances of difference throughout this study were calculated using a Student's t-test and by one-way variance analysis. P values ‹0.05 were considered significant. Result: In this study we had demonstrated that Metformin induces G1 cell cycle arrest and apoptosis in AML cells. Metformin is known to induce AMPK activation in a variety of cancer cells. This AMPK activation has also been shown to be essential for Metformin-induced cell death in certain cell types. Metformin caused AMPK activation in MV4;11 and NB4 cells, but failed to induce AMPK phosphorylation in THP-1 cells, indicating that AMPK activation by Metformin is cell-type specific. Metformin down regulate expression of mTOR and survivin gen. Conclusion: Metformin activate AMPK. AMPK inhibit mTOR gen expression and therefore inhibit survivin gen expression. The antiprolifrative effect of Metformin can be induced by inhibition of survivin, Although Differential activation of AMPK by Metformin in different cell lines suggests a possible AMPK-dependent or independent mechanism of metformin-induced cell death.

Keywords: Valproic acid (VPA), Leukemia , p21, apoptosis, histone deacetylases

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Paper ID: 86

The role of microRNAs in classification of acute leukemias of ambiguous lineage

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Abstract

Abstract Introduction: Acute leukemias of ambiguous lineage (ALAL) form a group of leukemias presenting the features of both acute lymphoid leukemia (ALL) and acute myeloid leukemia (AML). Based on Immunologic Classification of Leukemia (EGIL), these leukemias are recognized as biphenotype acute leukemia (BAL). According to WHO classification and based on immunophenotypic characteristics, ALAL is further categorized to acute undetermined leukemia (AUL), mixed phenotype acute leukemia (MPAL) and other ambiguous lineage leukemias.. In this review paper, the role of miRNAs expression in classification of leukemias for better diagnosis and selection of the correct treatment strategy has been discussed. Materials and methods: Relevant English-language literature were searched and retrieved from PubMed (1990–2014) using the terms: microRNAs, classification, acute leukemias of ambiguous lineage. Result: Immunophenotype and cytogenetic aspects are important in recognition of these leukemias; however, there is limited data about these leukemias. This has resulted in mistreatment of patients. Thanks to the recent advances and introducing the role of microRNAs in regulation of gene expression, a new avenue has been opened in the field of classification of ambiguous lineage leukemias. Conclusion: MiRNA-based classification can help define myeloid or lymphoid origin of ambiguous lineage leukemias. Taking miRNAs profiles into account, a better recognition of the disease is achieved, which results in accurate diagnosis and effective treatment of patients.

Keywords: MicroRNAs, classification, acute leukemias of ambiguous lineage

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Paper ID: 87

Prevalence and Related Factors of iron deficiency Anemia in Iranian Children

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Abstract

Introduction: Iron deficiency Anemia is the most common cause of anemia in children and one of the health threatening in all of the world. Anemia affects child development and limits their social activity, education and future life. Anemia is the time that blood hemoglobin level will decrease. It is necessary to prevent and treat it. The present study aimed at determining Prevalence and Related Factors of iron deficiency Anemia in Iranian Children. Method: This review study was carried out with investigation of scientific and research sites such as (Google scholar, SID, ….) without time limit. Results: According to the studies was reviewed Prevalence of iron deficiency Anemia in Iranian Children was different between %1/8, %53/6. Most factors influencing on iron deficiency Anemia in Iranian Children was such as: Insufficient use of meat, cereals, milk, vegetables, eggs and nuts, Inadequate or irregular use of iron from the age of 4-6 months, boy gender, early age of children, Insufficient or don't use of iron during pregnancy by mother, low age and education of mother, using of fish, don't receive supplementation iron, Economic status and life location of family. Conclusion: Prevalence of anemia in child is worrying; this shows the importance of exact recognition and identification of endangered persons with widespread, steady and high-sensitive screening programs. with regard to relatively high prevalence of iron deficiency and its health and socioeconomic complications, it is recommended that screening programs for iron deficiency will be done during the rapidgrowing age of children so that they would be treated if necessary.

Keywords: Related Factors, iron deficiency Anemia, Children

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Acute T Cell Lymphoblastic Leukemia in a Patient with Isovaleric Acidemia

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Abstract

Acute T Cell Lymphoblastic Leukemia in a Patient with Isovaleric Acidemia
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Isovaleric acidemia (IVA) an autosomal recessive disorder, is due to mitochondrial enzyme isovaleryl-coenzyme A dehydrogenase deficiency and is one of the branched-chain aminoacidopathies However, the physiopathology of the bone marrow suppression in this disease is still unclear.
we report the case of an IVA patient who presented with thrombocytopenia and leukocytosis. A 13-year-old male patient known case of IVA from 7 year old of life with mild psychomotor retardation, had recent admission in our Hospital for vomiting associated with dehydration, acidosis, ketonuria after episode of upper respiratory infection. at admission he had thrombocytopenia and leukocytosis that occurred in a sinuous path with increased uric acid that was diagnosed as T-Cell lymphoblastic leukemia with Bone marrow aspiration flowcytometry.
Hematologic problems can be seen in patients with inborn errors of branched-chain amino acid metabolism.Various cytopenias have been reported in IVA. Although findings suggest that the hematologic abnormalities of isovaleric acidemia probably are referable to an inhibition of normal cell maturation rather than a depletion of marrow precursors or increased destruction of mature cells. We believe that the lymphoblast cells observed in our patient's bone marrow were due to maturation arrest in the normal lymphoblastic series caused by IVA.
Although both forms of IVA often occur in the first year of life, IVA may be observed later in childhood as in our patient.
Keywords: Isovaleric Acidemia, leukemia, thrombocytopenia

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Paper ID: 91

Comparing the Heart and Liver Iron Deposition Status in Major β Thalassemia Patients Treated with Two Iron Chelation Drugs of "Deferoxamine and Deferasiroxon" Using MRI T2* Technique

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Abstract

Abstract Background and Objectives: Blood transfusion causes excessive iron store and many complications in major β thalassemia patients. "Deferasirox" is a new chelation drug which is not considered properly, therefore, in this study we have compared the heart and liver Iron deposition status measured by MRI T2* technique in patients that are being treated by two iron chelation drugs of "deferoxamine and deferasirox". Materials and Methods: In this study, 52 patients with major β thalassemia were evaluated. In all patients the liver and heart MRI T2* was done and the efficacy of deferoxamine and deferasirox on the heart and liver iron deposition status measured by MRI T2* technique was compared. The collected data were analyzed by SPSS software and statistical methods: Mann–Whitney U test, Fisher test, and Spearman's rank correlation coefficient. Results: Twenty three female and 29 male were evaluated. Deferasirox and deferasirox group were similar according to the mean of age and ferritin level (p

Keywords: beta-Thalassemia, Deferoxamine, Iron overload, Magnetic Resonance Imaging

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Paper ID: 94

RECURRENT BRONCHIOLITIS REVEALING A VITAMIN B12 DEFICIENCY IN AN INFANT

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Abstract

Vitamin B12 deficiency is rare in children. It affects mainly people over the age of 60. We report the case of a deficit in vitamin B12 in a 7 month old boy discovered through recurrent bronchiolitis and failure to thrive. Blood cell count revealed pancytopenia and a macrocytic anemia with a megaloblastic bone marrow. Viral serologies were negative. Immunological tests showed a decrease in humoral immunity (low levels of IgA).Vitamin dosages done for both mother and child revealed similarly isolated low serum vitamin B12 levels. Maternal pernicious anemia was reported and confirmed by the presence of intrinsic factor blocking antibodies (IFAB) and by an atrophic gastritis, although the blood cell count showed no notable abnormalities. Intramuscular cobalamin injections were quickly given to this exclusively breastfed boy and has allowed his recovery. It's therefore concluded that despite the rarity of the disease in children, complications are severe. It's preferable to recommend an adequate vitamin B12 supplementation for breastfeeding mothers as well as their children in order to help avoid producing irreversible neurological sequelae.

Keywords: Vitamin B12 deficiency, infant, etiology

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Paper ID: 97

Guidelines for the Diagnosis of Paroxysmal Nocturnal Hemoglobinuria by Flow Cytometry Methods

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Abstract

Background: Paroxysmal nocturnal hemoglobinuria (PNH) is a rare hematopoietic stem cell disorder characterized by a somatic mutation in the PIGA gene, leading to a deficiency of proteins linked to the cell membrane via glycophosphatidylinositol (GPI) anchors. While flow cytometry is the method of choice for identifying cells deficient in GPI-linked proteins and is, therefore, necessary for the diagnosis of PNH, to date there has not been an attempt to standardize the methodology used to identify these cells. Methods: In this document, we present a consensus effort that describes flow cytometric procedures for detecting PNH cells. Results: We discuss clinical indications and offer recommendations on data interpretation and reporting but mostly focus on analytical procedures important for analysis. We distinguish between routine analysis (defined as identifying an abnormal population of 1% or more) and high-sensitivity analysis (in which as few as 0.01% PNH cells are detected). Antibody panels and gating strategies necessary for both procedures are presented in detail. We discuss methods for assessing PNH populations in both white blood cells and red blood cells and the relative advantages of measuring each. We present steps needed to validate the more elaborate high-sensitivity techniques, including the need for careful titration of reagents and determination of background rates in normal populations, and discuss technical pitfalls that might affect interpretation. Conclusions: This document should both enable laboratories interested in beginning PNH testing to establish a valid procedure and allow experienced laboratories to improve their techniques.
Keywords: flow cytometry; paroxysmal nocturnal hemoglobinuria; practice guidelines

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Paper ID: 100

Could platelet rich plasma have affected tip cells production in human vascular endothelial cell cultures?

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Abstract

Background: Angiogenesis has key roles in normal growth, wound healing and organ regeneration. It has been suggested that platelet rich plasma (PRP) is a valuable autologous source for therapeutic angiogenesis in various soft and hard-tissue. CD34 has been introduced as a novel marker to detect endothelial cells with a tip cell phenotype in vitro. The purpose of this study was to evaluate PRP potential to induce human umbilical vein endothelial cells (HUVEC) to express CD34 as a tip cell marker.
Methods: HUVEC were cultured and were treated with both PRP and platelet poor plasma (PPP) and incubated for 48 hours. The effects of PRP on the CD34 expression were compared to platelet poor plasma (PPP) as a negative control using flow cytometry. Cell cycle study also was carried out to evaluate the effect of PRP on the proportions of cells in the various phases. NF-kappaB (NF-kB) inhibition was conducted using Small interfering RNA (siRNA) to investigate if there is a role for this signaling pathway.
Results: After incubation with PRP, but not with PPP, an increased expression of CD34 and enhancing both S and G2M phases of cell cycle were detected in HUVECs. NF-κB inhibition, significantly reversed these effects. Our findings support the hypothesis that PRP contribute to angiogenesis function of endothelial cells and may lead to the increase of novel therapeutic interventions for numerous angiogenesis-related diseases.
Keywords: platelet rich plasma (PRP);Tip cells; human umbilical vein endothelial cells; NF-kappaB (NF-kB)

IJBC 2015; Supplementary; P 105
Paper ID: 107

Generic and Copy Drugs in Hematology

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Abstract

Introduction: During recent decades, raising the awareness of persons with thalassaemia and their relatives has triggered an immense demand for standard care and high quality medicines. However, the governmental subsidiary has failed to catch up with the demand especially in developing countries and the decision makers have used generic or copy drugs to compensate for the shortage of budget. The strategy boomed a few questions about the quality of generic or copy drugs. Material and Method: The investigation aims to evaluate the outcome of switching into generic or copy drugs in treating patients with thalassaemia. To reach authentic results, we review clinical trials conducted to evaluate the efficacy of generic and copy drugs together with a few chemical analyses on the drugs. Both clinical data and physicochemical material has been utilized to indiscriminately judge about quality of the drugs. Results: The results of the study shows that a few generic and copy drugs which are used for thalassaemia treatment are contaminated with either expected or unexpected impurities; hence, they may cause postponed adverse drug reactions in the patients. Conclusion: Although affordability is a vital factor for providing patients with medicines in developing countries, decision makers should take more responsibility for the quality of medicines especially used for treatment of patients who are dependent on long time treatment. More restrict regulations should be imposed to assure consumers about the drugs' quality.

Keywords: Quality of Drug. Generic Drug. Copy Drug

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Paper ID: 110

Eltrombopag in pediatric chronic immune thrombocytopenic purpura

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Abstract

Abstract Immune thrombocytopenia (ITP) of childhood is characterized by isolated thrombocytopenia. ITP in children often resolves spontaneously within three months. Approximately 20 percent of affected children will go on to have chronic ITP, defined as ongoing thrombocytopenia more than 12 months from presentation. ITP in the absence of other causes or disorders that may be associated with the thrombocytopenia is known as primary ITP, and is the main focus of this topic review. Pediatric patients with chronic ITP are at ongoing risk of significant bleeding.The goal of treatment in chronic ITP for children is to maintain a safe platelet count that stops or prevents bleeding. The most commonly available and used therapies-corticosteroids and intravenous immunoglobulin (IVIG),Cyclosporine ,azathiopurin,and so on are associated with side effects that are often difficult to tolerate in a pediatric setting.

Keywords: eltrombopag, thrombocytopenia

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Paper ID: 111

A Concise Review of Factor XIII Deficiency in Iran

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Abstract

Factor XIII deficiency (FXIIID) is a rare bleeding disorder (RBD) with an estimated prevalence of 1 in 2 million population worldwide. In Iran, a Middle Eastern country with a high rate of consanguineous marriages, there are approximately 473 patients afflicted with FXIIID. An approximately 12-fold higher prevalence of FXIIID is estimated in Iran in comparison with overall worldwide frequency. In this study, we have undertaken a comprehensive review on different aspects of FXIIID in the Iranian population. The distribution of this disease in different regions of Iran reveals that Sistan and Baluchestan Province has not only the highest number of patients with FXIIID in Iran but the highest global incidence of this condition. Among Iranian patients, umbilical cord bleeding (UCB), hematoma and prolonged wound bleeding are the most frequent clinical manifestations. There are several diseases causing mutation in Iranian patients with FXIIID, with Trp187Arg being the most common mutation in FXIIID in our country. Traditionally, the management of FXIIID in Iran was only based on administration of fresh frozen plasma or cryoprecipitate until 2009, when FXIII concentrate became available for management of the patients. Various studies have evaluated the efficacy and safety of prophylactic regimens in different situations with valuable findings. Although the focus of this study was on Iran, it offers considerable insight into FXIIID, which can be applied more extensively to improve the management and quality of life in the affected patients.

Keywords: Factor XIII Deficiency, Iran, Molecular Spectrum, Clinical Manifestations

IJBC 2015; Supplementary; P 108
Paper ID: 112

The effect of Silybum marianum in treatment and prevention of hepatotoxicity in patients undergoing induction therapy for acute lymphoblastic leukemia

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Abstract

Background and objective: Using Hepatotoxic drugs in induction phase in patients with acute Lymphoblastic Leukemia increases liver enzymes, therefore, maintenance therapy with other hepatotoxic medications seem to have some difficulties. Recently, effects of treatment with Silymarin in monitoring serum levels of liver enzymes in ALL patients were considered. This study aimed to investigate the effects of Silymarin in treatment and prevention of liver toxicity in patients undergoing induction phase. Methods: In this clinical trial, patients diagnosed with Acute Lymphoblastic Leukemia who underwent induction therapy were enrolled, randomly treated with Silymarin and placebo for 28 days. Laboratory tests were done in patients on days 0, 28 and 56 and results were recorded and analyzed by SPSS.19. P

Keywords: Acute lymphoblastic leukemia, hepatotoxicity, Silymarin.

IJBC 2015; Supplementary; P 109
Paper ID: 113

Epidemiology of Childhood Cancer in Northwest of Iran

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Abstract

This is a case series study that has been done on all 83 children below 14 years old suffering from cancer during 2010-2013 who were registered in Ardabil pediatric cancer registry (APCR). The required data were collected by a questionnaire and analyzed by statistical methods in SPSS.19 software. 51 (61.4%) of cases were male and the rest of them were female. The mean age of patients was 5.8□4.2. 60 (72.3%) of cases were from urban area. Results showed that leukemia with 45 (54.2%), CNS with 12% and Neuroblastoma with 8.4% were the most prevalent malignancies in Ardabil province. Based on under 14 years old population estimated from Ardabil province, the cumulative incidence rate was 95.4 patients per one million. Results showed Leukemia was the most common childhood cancer in Ardabil province of Iran. CNS tumors and Neuroblastoma were the second and third ones, respectively. The incidence rate of these cancers was relatively high in Ardabil province. So, childhood cancers should be mentioned as an important issue in health policy making in Ardabil province of Iran.

Keywords: Childhood, Cancer, Incidence Rate, Iran

IJBC 2015; Supplementary; P 110
Paper ID: 114

The effect of intravenous Deferoxamine during blood transfusion time in thalassemia major patients

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Abstract

CONTEXT AND OBJCTIVE: Subcutaneous Deferoxamine intolerance by patients and reject its use Cause to iron overload and its complications. So, uses other methods with easier tolerate and reduce Deferoxamine consumption is necessary. One of these methods is concomitant use of intravenous Deferoxamine and blood transfusion which was investigated in this study. DESIGN AND SETTING: Semi-experimental study at the BU-Ali hospital in Ardabil University of medical science in North West of Iran. METHODS: in a Semi-experimental study, thirty four patients with β-thalassemia major who treated with monthly blood transfusion were randomly selected and followed for nine month. The mean of serum ferritin levels, subcutaneous Deferoxamine and Defripron uses rate in first three mount was considered as the baseline. Then at six months, the patients received intravenous Deferoxamine contaminant with their routine monthly blood transfusion and their chelators. After six mount the mean of ferritin, subcutaneous Deferoxamine and Defripron were checked again in the first and second three mounts compare with baseline. Data analyzed using statistical methods in SPSS.18 software. RESULTS: There were 15 men and 19 women of mean age 20.1±5.7years (range 7-28). The subcutaneous Deferoxamine and Defripron uses rate and ferritin level has been decreased in six mount compare to baseline. CONCLUSSION: Simultaneous intravenous Deferoxamine with routine monthly blood transfusion in beta thalassemia major due to decrease of ferritin level and subcutaneous Deferoxamine and Defripron need without any increasing in complications

Keywords: deferoxamine, Beta-Thalassemia, Ferritin, Blood Transfusion

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Paper ID: 117

Hematopoietic stem cell transplantation results in Taleghani BMT center :8 years exoerience

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Abstract

Transplantation of hematopoietic stem cells (HSCT) had rapid expansion during the last decade. Transplant has been used as a therapeutic option for lots of malignant and non malignant serious diseases.. Scince August 2007 to May 2015 607 HSCT (23% allogeneic, 77% autologous)were performed in Taleghani BMT center..It shows the general increase in HSCT from the beginning.For all transplants peripheral blood stem cells even in very young children(13 KG)was the source of stem cells.Pediatric patients were less than 10% of all transplants.Second and RIC transplants were performed in 9% of allogenic transplantations. Transplants were performed in a various type of diseases: hodgkine lymphoma (154 patients) multiple myeloma(150) ,AML(62) ALL (39),NHL(70) ,germ cell tumor (10) and Thalassemia,Poems,Ewing sarcoma family ,metabolic disorders,neuroblastoma,plasmacell leukemia,aplastic anemia,MDS,Wilms tumor as remaining.. The main indications for allogeneic HSCT were acute leukemias (91%), bone marrow failure syndromes , hemoglobinopathies and metabolic disorders were less in our center(9%) The main indications for autologous transplants were Hodgkine lymphoma and then plasma cell disorders RIC continued to show a progressive increase over the years for lymphomas and older patients. The vast majority (93%) of allo-HSCT sources were from sibling donors and alternative family donors were the remaining.In follow up 3 years DFS for hodgkine disease patients ,mulytiple myeloma,AMLand ALL were 70%,73% 52% ,63% and 28% respectively which is equals to western centers except in ALL.

Keywords: Transplantation,hematopoietic stem cells

IJBC 2015; Supplementary; P 112
Paper ID: 119

A case report of hypereosinophilic syndrome with FIP1L1-PDGFRA positive fusion gene and response to treatment

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Abstract

Definition

Hypereosinophilic syndrome includes a heterogeneous group of disorders defined by:

• A persistent eosinophilia of .1,500/mm3 for longer than 6 months

• Absence of evidence of known causes of eosinophilia despite a comprehensive work-up for such causes

• Signs and symptoms of organ involvement, directly attributable to eosinophilia including hepatomegaly, splenomegaly, heart disease, diffuse or focal central nervous system (CNS) abnormalities, pulmonary fibrosis, fever, weight loss, or anemia (i.e.evidence of end organ damage with histologic demonstration of tissue infiltration by eosinophils or objective evidence of clinical pathology in any organ system associated with eosinophilia and not clearly attributable to another cause).A special variant of hypereosinophilic syndrome with a fusion gene FIP1L1-PDGFRA occurring as a result of interstitial deletion on chromosome 4q12 has recently been described.They respond well to imatinib mesylate treatment, which targets the fusion tyrosine kinase. We repoted a case of hypereosinophilic syndrome :6 years girl with prolonged fever,generalized maculupapular pururitus rash,weigth loss,cervical lymphadenopathy,,abdominal pain,ascitis,hepatosple nomegaly,pleural and pericardial effusion,cardiac involovement,leukocytosis (75%eosinophil).In survey of bone marrow aspiration in genetic laboratory was detected fusion gene FIP1L1-PDGFRA.She was treated with imatinib and prednisolon.her signs and symptoms and hypereosinophilia resolved after 3-4 mounts.

Keywords:

hypereosinophilia,hypereosinophilic syndrome

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Paper ID: 122

Pediatric Hematopoietic Stem Cell Transplantation in Asia-Pacific Region

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Abstract

The Asia Pacific Region has the largest and most diverse population on earth. Although various studies on combined adult and pediatric populations and studies dedicated to pediatric Hematopoietic Stem Cell Transplantation (HSCT) have been conducted in western countries, similar reports on pediatric patients in Asian countries are infrequent.

The Asia Pacific Blood and Marrow Transplantation Group (APBMT) is an international organization which is involved in HSCT, sharing their information and cooperating with basic and clinical research in Asia-Pacific countries. Recent reports on HSCT from Asian countries suggest the progress of HSCT in this region. However, no comprehensive picture of the current state of Pediatric HSCT in Asia has been obtained. In 2012 the number of patients treated annually has reached 15000 for the first time in this region . However, there is still a considerable gap between countries. HSCT activity is associated with economic strength. Also, there are marked differences in donor and stem cell selections among Asian countries. The most significant increases in the past 15 years were observed in Iran and China, which have middle incomes. Even in countries of the high-income group (Japan, Australia and Korea) the number of HSCTs performed has been consistently increasing and is not likely to reach a plateau soon. This suggests that the demand for HSCTs has not been fulfilled in any of these countries. APBMT group seek to help countries in the region without pediatric HSCT centers with establishing centers of their own. Also, we have programs for training of other pediatric HSCT teams, throughout the region, so that new centers could be launched.

Keywords: Stem Cell Transplantation

IJBC 2015; Supplementary; P 114-115
Paper ID: 123

Aleukemic leukemia cutis in Children ; A rare presenatation 2 case presentation

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2.

Abstract

Skin involvement can be the first symptom of hematologic malignancies in children. Cutaneous incvolvement in children with acute lymphoblastic leukemia (ALL) is a rare presentation compared to acute myelogenous leukemia (AML). In the pediatric population, the frequency of leukemia cutis is approximately 10% in pediatric acute myelogenous leukemia and about 1% in pediatric acute lymphoblastic leukemia . Although leukemia cutis tends to present with other features of leukemia, it can occasionally precede the development of blast cells in the marrow and blood. The condition is then known as aleukemic leukemia cutis.The clinical appearance of leukemia cutis is variable. The most common manifestation is described as erythematous or violaceous plaques, papules or nodules involving the face, trunk and extremities. The most frequent location of skin lesions in children with ALL or Lymphoblastic leukemia(LBL) is on the head. Leukemia cutis needs to be differentiated from cutaneous manifestations of leukemia secondary to bone marrow dysfunction (such as petechiae), drug reactions and infections secondary to an immunocompromised state. Leukemia cutis must also be distinguished from the cutaneous lymphomas (i.e., mycosis fungoides and Sézary syndrome). Further studies are needed to evaluate the prognosis of children with Cutaneous involvement at diagnosis. Here in we present 2 cases of aleukemic leukemia cutis

Case 1:

A 12 month old girl with cutaneous involvement was reffered to Mofid children's hospital in SEP-2014 .She admitted duo to progressesive multiple erythematous nodules in areas of head ,frontal and preorbital site since 2-3 months ago. Also in physical examination she had pallor. Laboratory tests revealed; WBC 133000/mm3, Hb: 7.6 gr/dl, Platelet: 61000/mm3, Blast: 91%. Neut: 1% , lymph: 6% Mono: 1% , Eosin:%1. ESR: 25, Biochemstry:Normal, IU/ml. Virology: Nl. Bone Marrow Aspiration(BMA): Full blast, infavour of Acut Lymphoblastic leukemia(ALL).CD Flowcytometry;CD 19: 85%, CD 79: 79%, CD 34: 81%, HLADR: 40%, ,CD10:1%. (CD 10 negative acute B lymphoblastic leukemia - Pro B cell ALL). Molecular analysis for translocation: Normal, CSF: Normal.Uric acid: 10 mg/dl , BMA in day 8: Hypocellular and about 20% blast was seen. CXR: Normal. Abdominal sonography : Normal.Cardiac Echo : Nl. Karyotyp analysis: 46xx. EF: 60%. Brain CT: Normal. Bone survey: Normal. Scalp Skin biopsy report ; Infiltration of tumoral cells ,infavour of lymphoid cells. Treatment plan with ALL- high risk protocol started. After 2 weeks the skin lesions were improved significantly. 25 days later duo to chemotherapy she progressed to pancytopenia , fever, pallor , restlessness, GI bleeding and disseminated intra vascular coagulation and at last she died 30 days after admission duo of sepsis. The result of blood culture was klebsiella.

Case2:

Patient: A 6 years old girl reffered to Mofid children' hospital in Apr-2013 with history of diffuse large ulcers in lower limbs and buttocks since 2-3 months ago without pain , pruritis or pus. In Physical examination

she had good general condition with 5 large ulcers in lower limbs and buttocks. Otherwise , Normal. Biopsy of skin lesion by dermatologist was Suggestive of Langerhans Cell Histiocytosis(LCH). All other evaluations were Normal(including CBC WBC:7000 /mm3, PMN:44%,lymph:49% , Hb:13.9gr/dl,platelet:494000/mm3, ESR:18, ESR, CXR, Abdominal sonoraphy,Biochemistry, Serum Ig level ,virology, bacterial and leishmaniasis tests) . Revision of Biopsy sample & ICH: was not in favour of LCH.So we did Recommendation for deep skin lesion Re Biopsy . But Patient parents Refused. 3 months Later in July -2013 she visied in Ahvaz hospital duo to fever , weakness , lethargy , high WBC: 200000/mm3 and full Blast.CXR showed Mediastinal Mass, BMA and CD flow cytometry :in favour of T-Cell acut lymphoblastic Leukemia . Exchange transfusion ,2 times was done . Then chemotherapy started with T- cell ALL protocol. She had response to chemotherapy but at last she had CNS involvement and she died.

Conclusion: It is is important to think and becarefull about skin involvement ,that a small growing cutaneouslesion may be the presenting form of ALL.

Cutaneous involvement can be an early manifestation of ALL . Cutaneous leukemic infiltrates can be observed in children with standard risk as well as in high-risk ALL. Further studies are needed to evaluate the prognosis of children with such involvement at diagnosis.

Keywords: Pro B Cell leukemia ,T cell Leukemia,Aleukemic leukemia cutis

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Paper ID: 125

Vertebra Plana in Pediatric's Langerhans Cell Histiocytosis(LCH) : Is it necessary to do chemotherapy?

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Abstract

Introduction: Langerhans Cell Histiocytosis (LCH) involvement of the vertebrae is known to occur in 10% to 15% of pediatric spine tumor cases.The number of cases involving the vertebra is likely less than 1 million per year. Spinal lesions commonly affect the thoracic vertebrae, specifically the vertebral body .LCH may eventually cause a complete or incomplete collapse of the vertebral body resulting in the classic finding of "vertebra plana" on imaging. In the absence of this classic finding, a differential diagnosis should be considered, including osteomyelitis, Ewing sarcoma, leukemia, lymphoma, metastatic neuroblastoma, hemangioma, Gaucher disease, osteogenesis imperfecta, and aneurysmal bone cyst.Clinical presentation of LCH of the spine varies from extreme pain, acute or gradual in onset, asymptomatic in which case the lesion is detected as an incidental finding on imaging, Painful scoliosis and in rare instances, neurologic compromise due to cord compression or displacement .Work up of these patients often involves an initial radiograph of the spine to search for signs of vertebra plana; MRI of the spine to rule out a soft-tissue mass, which would suggest a more aggressive cause of vertebral collapse; skeletal survey radiographs or technetium bone scan to search for multiple lesions; and, eventually, biopsy of the lesion to yield a diagnosis.Treatment are including bracing ,radiation therapy, chemotherapy, surgery , either alone or in combination. Here we present 3 pediatric cases of LCH with vertebra plana and different criterias for starting of chemotherapy ;Case 1,with recurrent disease, case 2 with vertebral involvement and soft tissue mass and case 3 just vertebera plana of L1 . Case 1: A 12 years old girl was reffered to our hematology-oncologuy clinic in Mofid Children's hospital ,in year of 2012 . She had history of low back pain without other findings since one year ago. At that time she visited by an orthopedic's surgen.Evaluations including lumbosacral plain radiography and MRI were done. MRI of lumbosacral revealed; Bone marrow edema in L2 body , mild scoliosis with convexity to the left side . Needle biopsy report of lesion was LCH.Recommendation by her orthopedic physician was using of brace and no more treatment.She was good till one year's later .In this time, again she had complained pain in thoracolumbar site. New MRI showed T6 vertebral body compression fracture with significant height loss and posterior cortex bulging compressing in thecal sac,Guided needle core bone biopsy(measuring 0.7 cm in length)of T6 vertebral lesion report was ;LCH .Immunohistochemistry(IHC) study for S100 protein and CD1a were positive. Evaluaions for staging and treatment plan were as below; Bone survey showed concavity in superior and plate of L2.Whole body bone scan; abnormal increased radiotracer uptake in mid thoracic space at about T6 ,Compression fracture and height loss. CXR; normal. Chest CT ;normal.Bone marrow aspiration(BMA); normal . Abdominal sonography; normal. Specefic gravity(SG) of urine; 1025 . CBC , biochemistry, liver , kidney, thyroid functions tests and serum immunoglubolin levels ; normal. LDH; 825 Iu/ml, ESR;30, PT,PTT ;normal. So based of diagnosis of LCH and recurrency of disease after 1 year we decided to do chemotherapy with standard protocol of LCH . She recieved Vinblastion ,prednisone based of response for 12 weeks as an induction phase , then maintenance chemotherapy includings Vinblastin , prednisone

every 3 weeks and 6MP daily for 1 year .Also based of recommendation of neurosurgen , she used of brace.At the time of off therapy ,Bone mineral densitometry (BMD)duo receving of corticosteroids for a long time was done. Repor of BMD showed ; Femur Z score -0.5 ; diagnosis; normal and lumbar Z score ; -0.44 diagnosis;normal ,otherwise evalutions data were normal.Now she is off therapy more than 1 year and follow up is going. Case 2: An 8 years old boy duo to back pain since 1 month ago was reffered to our hematology- oncology clinic in year of 2012.Evaluations results were as below;CBC, biochemistry, liver ,kidney ,thyroid functions tests and serum immunoglubolins level; normal. LDH ; 700IU/ml, ESR;18,PT, PTT; normal.BMA; normal.Urine specific gravity(SG): 1020. CXR; normal, Abdominal sonography: mild splenomegaly(100mm). Bone survey:Collapse of T8 vertebral body with paravertebral soft tissue mass .Whole body bone scan: decreased radiotracer uptake in in T8 vertebra. CT; pathologic fracture of T8 vertenbral body With paravertebral soft tissue without extension to spinal canal. MRI of thoracolumbar spine; vertebra plana in T8.Core needle biopsy and histopathology report of thoracic vertebral mass (measuring 1cm length and 0.1 cm in diameter)showed ; composed of sheets and single histiocytes cells with vesicular nuclei some with nuclear grooves , many eosionophillic cells . Diagnosis; LCH.IHC study ; S100 and CD1 a positive . Recommendation of neurosurgen was using of brace.Chemotherapy started with vinblastin , prednisone . In weeks 6 of Induction phase evaluation for response to treatment was done , then 6 more weeks chemotherapy with prednisone and vinblastine continued .After 12 weeks, maintenance therapy with vinblastin , prednisone , every 3 weeks and 6MP-daily for 1 year were done. BMD at end of treatment showed: L1-L4: z- score; -2.4 ,whole body z score ;–1.1 , which treatment with vitamin D and calcium was done. Now He is good and he is on follow up. Case3: A 4.5 years old boy duo to back pain visited . MRI showed vertebra plana in L1 vertrebral body. Results of Needle core biopsy of L1 vertebral body was LCH and in IHC , S100 protein and CD1a were positive.Based of all evaluation dates and just vertebra plana in L1 and no soft tissue mass we did not do chemotherapy .He is using of brace . He is on follow up . Conclusion: We recommend that any child with suspected solitary LCH of the vertebra undergo a full diagnostic investigation. A biopsy is highly recommended for a diagnosis at the time of presentation and should be attempted in any suspicious lesion .A CT-guided biopsy or an open biopsy would be appropriate.In LCH patients with vertebra plana without soft tissue mass , just observation and using of barce may be the only recommendation ,but careful follow up about events including recurrency of disease such as our case 1 is recommended. Also in patients with vertebra plana and soft tissue mass chemotherapy should be considered.Regular Follow up in pediatric patients with vertebra plana, minimally 3 years shoule be considered.

Keywords: Langerhans Cell Histiocytosis(LCH), Vertebra Plana ,Chemotherapy

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Paper ID: 126

The mechanism of Busulfan treatment in chronic myeloid leukemia imatinib resistant patients by its effects on Growth Factor Receptor-bound protein 2

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Abstract

Background: Chronic myeloid leukemia (CML) therapy has based on chemotherapy(with busulfan) since 1980 until tyrosine kinase inhibitors(TKIs) emergence. TKIs,such as imatinib , has significantly improved outcomes for CML patients, but their main limitation is resistant patients, due to different reasons, that oblige physicians to use of conventional chemotherapy with busulfan. although, busulfan has used for many decades but its mechanism in CML treatment was not understood.
so, the aim of this study is identification of the mechanism of busulfan effect in CML patients.
Methods: In this review article we surveyed available literature in PubMed database during 2000 to 2015 to find original articles, bibliographic reviews and books.
Results:BCR-ABL fusion, generated with reciprocal translocation between 9 and 22 chromosomes, is a tyrosine kinase that is main cause of CML formation.BCR-ABL increase growth factor independent cell growth with activation Ras-MAPK pathway thorough BCR-ABL-GBR2-SOS protein complex.
also, it has been found that MiR 200 can target Gbr2 and also it is upregulated by busulfan.
Conclusion: given that MiR 200 unregulated by busulfan therapy and it can targeted GBR2, so it was concluded that busulfan can down regulated Gbr2 thorough up regulating MiR 200.also evaluation of secondary Gbr2 regulator,MiR 376, in CML patients has been suggested in future studies.

Keywords:
CML,Busulfan,imatinib resistance,Gbr2

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Paper ID: 128

Effect of shift –base of vanadium on necrosis and apoptosis of k562 erythroleukemia cell line

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Abstract

BACKGROUND: Cancer is the most popular agent of mortality in human being, after cardiovascular system disease and accident, so there is great interest on research of cancer in the diagnostic, treatment and prophylactic points of view. In the present study, the apoptotic and necrotic effects of shift –base of vanadium on k562 erythroleukemia cell line were studied. In the first step the effective dose range was determined by MTT test, and then 150,250 and 350µg/ml of compounds were used to determined necrotic as well as apoptotic death of cells, using commercial available Annexin V/PI kit and flow cytometry. The results of MTT test showed that the effective dose range of these compound is 25-400µg/ml, the highest cell death was happened in 350µg/ml concentration after 48 hours incubation(83-89%),In comparison to doxorubicin(as standard), these compounds killed cells for necrosis rather than apoptosis pathway. RESULTS : the results of apoptosis and flowcytometry examination showed that in mentioned densities the best effect is after 12 hour care with vanadium compounds,and the more apoptosis occure in dose of 350 µg/ml is 37.96%,also the results from MTT indicate that 150,250,350 µg/ml of vanadium compounds on cells of k562 cell line cause a worthy and meaningful decrease in the number of existing cells,in away that after 48 hours of decrease,cellular percent in comparison to cells which not affected by shift –base of vanadium compounds was meaningful at this level p

Keywords: shift –base of vanadium , k562, Apoptosis , MTT,Flow cytometry.

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Paper ID: 133

Marriage and child bearing in patients with Beta-thalassemia major in South-East of Iran

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Abstract

Background Expecting a family is an important component and a great goal for better quality of life for most of adults with thalassemia major. The aim of the present study was to examine the marital status of adults with thalassemia major. Materials and Methods This cross-sectional study examined the marital status of patients with transfusion-dependent thalassemia aged over 15 years. Patients' demographic characteristics including age, gender, marital status, duration of marriage, divorce, having or not having children and spouse's health status were recorded. Information about the disease including cardiac and endocrine complications, ferritin level, splenectomy and viral hepatitis were also recorded. Results Of 228 patients with transfusion-dependent thalassemia major aged over 15 years treated at this medical center, 32 patients were married comprised 14% of thalassemia major patients over 15 years. The mean age of married patients was 25.18 ± 4.74 years. Among the married patients, 8 (25%) were females and 24 (75%) patients were males. The mean age of marriage was 22.76 ± 4.16 years. The minimum and maximum marriage age was 15 and 33 years, respectively. The mean duration of marriage was 2.47 ± 2.82 years. Only 8 (25%) patients (one female and 7 males) had children. Conclusion Therapeutic advances have led to significantly increased survival and improved quality of life and fertility of patients with thalassemia major. According to the results, 14% of patients over 15 years were married which was slightly higher as compared with other similar studies.

Keywords: Thalassemia Major, Iron Chelation, Marital Status

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Paper ID: 185

Twice daily dosing of deferasirox significantly improves clinical efficacy in transfusion dependent thalassemias who were inadequate responders to standard once daily dose

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Abstract

Introduction: Clinical efficacy of deferasirox (DFX); a once daily oral iron chelator in patients with transfusional iron overload depends on factors such as iron burden, rate of transfusion and appropriate dose. At present, the recommended dose approved as a label indication is 30 mg/kg/day, though doses up to 40 were studied in patients with cardiac siderosis with acceptable safety profiles. Recent several studies have shown in a non-clinical trial setting that a proportion of patients could not achieve satisfactory iron balance at 30 mg/kg/day or even at higher dose. These patients were labeled as inadequate responder (IR). Difference in bioavailability with a lower area under the curve (AUC) of each IR individual was the only main mechanism identified earlier (Neufeld E, et al. Blood 2009). Objectives: We hypothesize that adjusting DFX dose from once daily into two dividing dose per day might improve drug exposure and chelating efficacy in such patients. Methods: A retrospective cohort of clinical response to deferasirox in Iranian thalassemia patients was performed after an informed consent. • A standard guideline for the use of deferasirox at our hospital including SF and MRI monitoring, adverse events (AEs) evaluation and standard laboratory evaluation was set by both authors and this was applied to all patients participated into this study. Patients with IR was primarily defined as (1) having a rising serum ferritin (SF) trend or (2) having a reduction of SF less than 30% of baseline levels (BL) at least 3 consecutive months, with more than two SF measurements higher than 1500 ng/mL; and (3) receiving once daily DFX at an average dosage > 35 mg/kg/day for at least 6 months. • DFX administration schedule was switched to twice daily with the same total dose per day. • CBC, renal function, urine analysis were performed every 3 weeks to monitor possible adverse effects. SF and liver function test were checked every six weeks. • Tolerability and compliance to DFX were evaluated by direct history taking and drug account prescribed during study period. Results: Total 8 patients (3 males, 37.5%) were eligible with a mean age of 18.6 yrs (range; 10-28 yrs). There was a statistically significant decrease in serum ferritin levels with twice daily use of deferasirox compared to once daily use of the same dose (2319.00 vs. 1284.19 mg/dl, P=0.002). The initial and follow-up ALT and serum creatinine levels did not differ significantly (p>0.05). None of the patient required a dose reduction or cessation of the drug related to a toxicity. there was no statically significant difference for liver MRIT2 before and after twice daily treatment (P=0.056). But difference between Heart MRIT2 before and after twice-daily treatment was very significant (P=0.001). Conclusion: dividing DFX to twice daily dose might provide a better bioavailability in selected patients with sustainable therapeutic levels of DFX throughout 24 hr-exposure resulting in a better clinical efficacy. Further pharmacokinetic and pharmacogenetic study in IR patients is warranted and this can provide additional insights on the next level of tailoring iron chelation therapy in patients with transfusional iron overload.

POSTERS OF STUDENTS

IJBC 2015; Supplementary; P 123
Paper ID: 22

Drug resistance and multidrug resistance in blood cancer ; differences and similarities

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Abstract

Introdution: Although chemotherapy of tumors has been proved to be successful, drug resistance is one of the most essential reasons for the failure of cancer treatment like as blood cancer. The aim of this project is to investigate the differences and similarities of drug resistance in order to find new therapeutic strategies to conquer this complication. Materials and Methods: In this review article we used related articles from various reliable biological and medical journals from Pubmed, NCBI, Science Direct and Elsevier databases. **Results:** Drug resistance is the phenomenon is which patients lose their proper response to one or more medication(s). The resistance may occur in response to a single cytotoxic drug. In this regards, the reason for drug resistance may be alteration in the targets of the chemotherapeutic drugs, so that they may no longer bind to or affect their target molecules. Ligands for chemotherapeutic agents may be portions of DNA, microtubules and RNA polymerases. Multidrug resistance is failure to response to several drugs that are different in their chemical structures and molecular mechanisms.This phenomenon is one of the most important reasons for relapse and failure to therapy. Several mechanisms are involved in multidrug resistance including resistance to extracellular matrix, ABC transporters, drug inactivation, alteration in drug targets, DNArepair mechanisms, deficiency in sphingolipid and ceramide metabolisms, deficiency in apoptotic pathways and cell compartmentalization. Conclusion:This article helps a better understanding of drug resistance in treatment of patients with blood cancer . Determination of differences and similarities between these two phenotypes may help selecting proper chemotherapeutic protocols for patients and improve new cancer therapeutic

Keywords: Chemotherapy,drug resistance,multidrug resistance,blood cancer

IJBC 2015; Supplementary; P 124
Paper ID: 23

The role of ABC transporters in drug resistance of brain tumors

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Abstract

Background and purpose: Although chemotherapy of brain tumors in addition to surgery and radiation therapy Was successful for cancer management, multidrug resistance(MDR) is considered as one of the most important obstacles in treatment of brain tumors .MDR is a complex phenomenon through which cancer cells become resistant to variety of anti-cancer drugs which are functionally and structurally unrelated and discrete. This article aimed foru nderstanding the mechanisms of generating MDR and investigating the importance of ATP Binding Cassette(ABC) transporters in this phenomenon. Materials and Methods: In this review article we used related articles from various reliable biological and medical journals from Pubmed, NCBI , Science Direct and Elsevier databases. Results:The major mechanisms through which MDR isproduced are the loss of drug carrier proteins on the surface of the cell, change or mutation in drug specific targets such as tubulin and topoisomeraseII, decrease in absorption of water soluble drugs, DNA damage repair, decrease in apoptosis and increase in energy dependent efflux of hydrophobic drugs and cancer stem cells. One of the most important mechanisms of MDR which has recently attracted attention is the overexpression of ABC transporters specially in blood-brain barrier that efflux chemical substances from the cells.P-glycoprotein (P-gp) from the ABC family, and multidrug resistance proteins (MRPs) are very important in MDR of brain tumors. To overcome MDR, alternative treatments can be added to the main treatment protocol. Administration of flavonoids and retinoids, for instance, can be a key success for management of glioblastoma. Conclusion: This article helps a better understanding of MDR in patients with brain tumor in order to develop improved therapies in this regards

Keywords: multidrug resistance(MDR), brain tumor, blood-brain barrier, glioblastoma

IJBC 2015; Supplementary; P 125
Paper ID: 33

The effect of cinnamon on Cancer treatment

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Abstract

Introdution: Plants with their biological properties are to safeguard the health and treatment of diseases. In the past, most people had no knowledge about the constituent elements of the plants, but they used them to treat diseases. Nowadays, the natural products from plants provide better patterns for designing potential therapeutic agents than synthetic drugs. The incidence rates of different cancers have increased dramatically worldwide. This disease is characterized by the abnormal growth of undifferentiated cells and many efforts have been recently done to find anticancer reagents. Medical herbals play a vital role in the prevention and treatment of cancer and are usually available and economical to use. Understanding the anticancer properties of plants have always been critical. Cinnamon is one of these plants. Materials **and Methods:** In this review article we used related articles from various reliable biological and medical journals from Pubmed, NCBI, Science Direct and Elsevier databases to understand the health benefits of cinnamon in cancer. Results: The genus cinnamon is from the family of Lauraceae. Members of this family are evergreen trees which mostly grow in south-eastern Asia, Australia and South America. Two popular types of cinnamon exist with the scientific names of Cinnamomum Zeylanicum and Cinnamon Aramaticum. The chemical compounds of cinnamon are essential oils, carbohydrates, tannins (proanthocyanidins), crude fiber and minerals. Proanthocyanidins are greatest the phytochemicals and polyphenols compounds that are present in the bark. In vivo and in vitro studies have shown that cinnamon compounds may target molecules such as NFκB, AP1, Caspases, pro apoptotic genes (Bim, Bax, Bak), angiogenesis factors (VEGF), master regulators in cancer progression (HIF 1α, COX2) and MAP kinase signaling pathways to cause cell cycle arrest and cell growth inhibition, induce apoptosis and the reduce of aggreresive behavior of tumor and tumor angiogenesis therefore inhibiting cancer progress. Conclusion: Evidence suggest that cinnamon, due to its polyphenol compounds and antioxidant properties, may be used for cancer treatment.
Keywords: Cinnamon, polyphenol, blood cancerneeds to be described in another manuscript.

IJBC 2015; Supplementary; P 126
Paper ID: 37

relation of npm1 mutation with AML prognosis in children and adult

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Abstract

AML(acute myeloid leukemia) is the second-most common form of leukemia in children, after acute lymphoblastic leukemia (ALL). NPM1 is the most frequently mutated genein AML and the NPM1 mutant has an important role in leukemogenic.

Nucleoplasmin (NPM1) is a nucleolar phosphoprotein which shuttles between the nucleus and cytoplasm during the cell cycle. It was originally identified as a nucleolar phosphoprotein expressed at high levels in the granular region of the nucleolus. NPM1 plays an essential role in cell growth and proliferation by regulating cell cycle progression and centrosome duplication.

NPM1 mutations associate with a higher white blood cell count at presentation and higher blasts percentage at diagnosis; Some authors have found a significantly higher platelet count at the time of diagnosis in patients with NPM1 mutations.

NPM1 mutation has been associated with higher rate of complete remission compared to NK (normal karyotype) AML with wild type NPM in both adults and children.

Some data suggests that NPMc+ (cytoplasmic form) AML has an increased sensitivity to chemotherapeutic agents secondary to its interaction with (NF-κB).

Base on the results of several researches , patients having only an NPM1 mutation had a significantly better overall and disease-free survival and a lower cumulative incidence of relapse. On the other hand overall survival and disease free survival of NPM1-mutated AML were significantly higher than patients with wild type NPM1.

NPM1-mutated AML displayed increased expression of numerous HOX genes such as HOX B2 and B6. These results suggest that HOXB6 and B2 overexpression are biologically important markers of NPM1-mutated leukemia.

Keywords: AML, children, NPM1, leukemia

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Paper ID: 52

Gene expression analysis of Foxo3a gene in pediatric acute lymphoblastic leukemia

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Abstract

Introduction: Acute Lymphoblastic Leukemia (ALL) is a malignant proliferation of Lymphoid Cells blocked at an early stage of differentiation. The disease is divided into pre (precursor) B-cell and pre T-cell ALL. It is the most common pediatric malignancy with peak prevalence age between 2-5 years. ALL could be caused by repeated alterations in Proto-Oncogenes and tumor suppressor genes. The Forkhead box transcription factor 3, FoxO3a, is an essential tumor suppressor that regulates the mechanisms of tumorogenesis and leukemogenesis. It has been reported that therapy-resistant ALL cells inactivate FoxO3a to scape apoptosis. This study was aimed to investigate the expression of FoxO3a in the children with ALL.

Methods & materials: Real Time PCR was used to detect FoxO3a mRNA expression in blood samples from 50 ALL patients and 50 healthy children as control group. Normalization of the results was done in comparison with TATA- box binding protein gene.

Results & discussion: From literature review, the role of the FOXO3a gene in B-cell and T-cell development is obvious; we expected that FoxO3a expression in ALL patients would be lower than that in healthy matched children. The result of our experiment could provide a powerful prognostic molecular marker in early diagnosis of ALL in children.

Keywords: acute lymphoblastic leukemia, pre B-cell, pre T-cell, FOXO3a, Real-Time PCR

IJBC 2015; Supplementary; P 128
Paper ID: 53

Gene expression analysis of SPI1 gene in pediatric acute lymphoblastic leukemia

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Abstract

Introduction: Leukemia is a malignant disease of the bone marrow and blood, characterized by the uncontrolled growth of progenitors of white blood cells. While acute lymphoblastic leukemia (ALL) would happen during life span, its incidence peak is between 2 and 5 years of age. ALL named since, it is an acute form of leukemia, or cancer of the white blood cells, characterized by the overproduction and accumulation of cancerous, immature white blood cells—known as lymphoblast. Mutation in proto-oncogenes are common known cause of human leukemias. One of the members of this group of genes is SPI1, the transcription factor (TF) related to Ets family proteins expressed in myeloid and B lymphoid cells. SPI1 required for generating lymphoid progenitor cells from hematopoietic stem cells. The aim of this study was to estimate the expression of SPI1 gene in pediatric ALL.

Methods & materials: We used real-time RT-PCR to compare PU.1/SPI1 expression between 50 ALL patient samples and 50 normal blood specimens. Cases and controls were matched by age and sex.

Results & discussion: It is predicted that the expression of SPI1 transcription factor gene show increase in ALL patients compare to healthy controls, as this TF involved in the development of T and B lymphocyte. SPI1 would be considered as an molecular marker in early diagnosis of ALL in children.

Keywords: acute lymphoblastic leukemia, transcription factor, SPI1, Real-Time PCR.

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Paper ID: 56

How Nicotine can cause leukemia

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Abstract

Background: Cigarette smoking is an environmental risk factor for many disease, that accounts for a tenth of adult mortality rate around world .Nicotine, as a most important compound in cigarette , is a ligand for n-Nicotinic acetylcholine receptors(nAChR) that plays a principle role in many human cancer formation such as lung,breast,bladder and colon. the aim of this study was determination of effects and leukomogenic roles of nicotine usage. Methods:In this review article we surveyed available literature in PubMed database during 2000 to 2015 to find original articles, bibliographic reviews and books. Results: Our studies showed that the cigarette smoking is a significant risk factor for the development of Acute myeloid leukemia in adults. also it has been indicated that α7-nAChR expressed on Hematopoietic stem cells(HSCs). homopentamers of α7-nAChR are the important channels for Ca2+-dependent mechanisms that can activated some signaling pathways such as PKA, PKC, PI3K/Akt, and MAPK. furthermore, any disruption in this pathways can be a risk factor for leukemia formation.moreover, among them PI3K/Akt seems to be has a most important and well-documented role in AML generation. Conclusion: these information reveal that nicotine usage can disrupt some signaling pathways in HSCs thorough α7-nAChR and subsequently cause leukemia. so, it can be assumed specifically blockage of this receptor can reduce the risk of nicotine related cancers. but these findings need more experimental study for confirmation and used as therapeutic tools.

Keywords: Leukemia,HSCs, Nicotine

IJBC 2015; Supplementary; P 130
Paper ID: 66

Role of microRNAs in Outcome of Pediatric Acute Leukemia Pateints

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Abstract

Introduction: Treatment outcomes of pediatric cancers have improved greatly, resulting in overall survival rates greater than 70%. Acute lymphoblastic leukemia (ALL) ,the most common malignancies in children, now can achieve complete remission with current treatment regimens but leukemia relapse, the main cause of treatment failure, still occurs in a significant proportion of patients. Although acute myelogenous leukemia (AML) accounts for 16 % of leukemia in children, it is responsible for over half of all pediatric leukemia deaths. MicroRNAs (miRNAs) are a group of small 19-22 nucleotides-long non-coding RNAs which post-transcriptionally control the expression level of almost 60% of protein-coding genes. Deregulation of miRNAs is often correlated with tumorigenesis.

Material and methods: We searched different databases by PubMed by using microRNAs and acute leukemia as key words. We reviewed related articles published in English after the year 2010 irrespective of type of the articles.

Results: In pediatric AML, level of microRNAs, miR-125b, miR-335 and miR-375 significantly associated with poor survival. Reduced level of miR-29a was also found to be associated with unfavorable outcome. In childhood ALL, microRNAs miR-27a, miR-150, miR-191, miR-223, miR-335, miR-342, miR-486, miR-487 and miR-708 were markedly associated with better outcome. However expression level of miR-7, miR-128b, miR-216, and let-7i associated with early relapse and poor treatment response.

Conclusion: The initial screening of microRNAs differentially expressed from normal in Acute leukemia's suggesting the potential roles of them in leukemogenesis. microRNAs expression signatures may be useful for predicting prognosis and relapse in childhood acute leukemia and directing personalized treatment.

Keywords: MicroRNA, acute lymphoblastic leukemia, acute myeloid leukemia

IJBC 2015; Supplementary; P 131
Paper ID: 71

Challenge of dramatic increased invasive mycosis in pediatrics with hematological disorders

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Abstract

Introduction: Hematopoietic disorders(HD) in pediatrics are disruptions or malfunctions in the formation of one or more types of blood cells. Aplastic anemia, Thalassemias, Leukemia and Lymphomas are different kinds of HD. Invasive mycoses(IM) are major cause of morbidity and mortality of HD. Definite diagnosis of IF in febrile neutropenic patients is challenging and time-consuming and delay of antifungal treatment leads to higher mortality rates. This work aimed at description of most important types of mycosis in these patients.
Method: The literature published about IF in hematological disorders over the past 25 years was searched. We present list and tables of the most IF in patients with hematopoietic dysfunction that published during time.
Results: Aspergillosis and candidiasis remain the most common causes of IF in patients with HD. Less common IM are cryptococcosis or mucormycosis, Cultures of tissue, blood, or other fluids are used to make a definitive diagnosis of IF; however, histopathologic or microbiologic tests are slow, so treatment may be initiated empirically based on clinical suspicion. Uniformly effective prophylaxis therapeutic strategies for IM are lacking. Researchers are developing new serology and molecular diagnostic techniques (e.g. ELISA and PCR) for identify fungal species quickly and accurately. Corticosteroid therapy and neutropenia increase risks of IM. Because many patients can't mount an effective immune response, mortality rate is over 50%.
Discussion: Recent reports have indicated that the incidence rates of IM (8%-16%) have increased among HD particularly recipients of allogeneic hematopoietic cell transplants after engraftment. High-dose corticosteroid therapy was an important prognostic variable of outcomes after IM in patients. Although neutropenia has been considered to be the primary risk for IM, multiple studies indicated that IM may occur in non-neutropenic hosts. Developing new diagnostic technique for identify fungal species quickly and accurately is necessary to establish good treatment and reduce levels morbidity and mortality.
Keywords: Invasive Mycosis, Fungal infection, Hematopoietic disorders, Anemia, Leukemia, Lymphomas

IJBC 2015; Supplementary; P 132
Paper ID: 78

Cytogenetic analysis and assessment of miRNA let-7a and its target NRAS in a Childs with myelodysplastic syndrome (MDS)

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Abstract

Abstract Introduction Myelodysplastic syndrome comprise a heterogeneous group of clonal hematopoietic stem cell malignancies that characterized by a maturation defect in myelopoetic progenitor cells ,peripheral cytopenia(s) and clonal instability with enhanced risk of transformation to acute myeloid leukemia (AML). They are commonly diseases of the elderly and exhibit a dramatic increase in incidence with age. However, in children, MDS is often seen in association with genetic disorders and inherited bone marrow failure syndromes. Here we report 3, nonsyndromic MDS children in a family with the history of the disease in tree generation. Material and method Clinical assessment wase done by hematologist for MDS confirmation in these patients. Genetic counseling was done and the pedigree of the family drawn. Karyotype analysis on the basis of G-banding technique at high resolution was performed for all of the members of the patient's family and the expression of the miRNA let-7a and its target gene NRAS was assessment by real time PCR technique . Result Karyotype analysis reveals an abnormality in one of this patient and the assessment of the expression of the miRNA let-7a and its target shows a significances decrease compared with normal cases. Discussion The incidence of the MDS increase in elderly and it is not common in young people under 50 years old. The cytogenetic study of the patients shows abnormality just in one of them. One of the patient was 9 years old boy and his karyotype revealed abnormality. Such cytogenetic abnormality was not seen in other members of the family, so it may be a candidate for novel mutation. About 50 percent of the patient with MDS have normal karyotype so the diagnosis, prognosis and the treatment of such patient is difficult; for the better assessment and classify the MDS new molecular marker should be used. One of these markers were miRNA that recently widely uses for prognosis of the cancer and they have the potential for the detection of the disease in early stages.

Keywords: myelodysplastic syndrome, Cytogenetics, miRNA Let-7a, NRAS

IJBC 2015; Supplementary; P 133
Paper ID: 85

Inherited antithrombin deficiency causing thrombophilia : a case report

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Abstract

Introduction Antithrombin (AT) is a potent inactivator of thrombin and factor Xa and the major inhibitor of blood coagulation. Deficiency of AT represents a risk factor for thromboembolic diseases. It can be acquired or inherited which is uncommun with an incidence relatively rare in general population(1:10.000). Two types of deficiency are known ; quantitative (type I) or qualitative (type II). Case report We present the case of a 2-years old boy, without any particular medical history, who suffered an acute ischemic stroke. The etiological investigation showed a very low level of antithrombin which was confirmed with a subsequent assay on a fresh specimen (9% and 4% of the normal value). To evaluate the antithrombin level, we used a functional AT assay which is amidolytic (chromogenic) assay. The Quick time, the partial thromboplastin time and the fibrinogen were normal. Concentrations of protein C, S and Leiden V factor were normal. Discussion Congenital antithrombin deficiency is inherited as an autosomal dominant trait. Homozygous congenital antithrombin deficiency is rare. It is accompanied by a high risk of thrombosis, typically noted in the neonatal period or early infancy. This condition is rarely compatible with life. The familial study of our patient showed that he is homozygous for the AT deficiency. His parents which are cousins are both heterozygous (53% for the father and 48% for the mother without any thrombosis) and her sister's AT level was normal (94%).

Keywords: antithrombin deficiency,homozygous,thrombophilia

IJBC 2015; Supplementary; P 134
Paper ID: 92

Hematological and demographical features of childhood Acute Lymphoblastic Leukemia referred to Ahwaz Shafa hospital during 2006 to 2011

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Abstract

Background: Acute lymphoblastic leukemia (ALL) is a hematological malignancy that accounts for approximately 2% of all lymphomas. Although ALL occurs both in childhood and adolescent, it is the most frequent neoplasm in children fewer than fifteen years old. Due to relatively high incidence rate of ALL in Khuzestan, evaluations of hematological features of these patients are useful diagnostic and prognostic tool. Materials and Methods: This retrospective study was conducted in Ahvaz Shafa hospital, during 2006 to 2011. The data from all of fewer than fifteen years old ALL patients was collected and analyzed with SPSS 16 software. Results: From 175 ALL patients 63.4% and %36.6 was male and female, respectively. The highest incidence rate of ALL was found in age group 1-month to 5 years (50.3%) and after then 5-10 years (28.6%) and the lowest one was 10-15 years (21.1%). We found leukocytosis, thrombocytopenia and anemia in 52.6%, 94.9% and 87.4% respectively of our population,. Conclusion: Our findings showed that the poor prognosis factors include male sex, anemia, leukocytosis and thrombocytopenia had a high prevalence in the study population. It can be cause of the high percent of fewer than five years old patients. In concerning tour findings, conducting a simple routine and regular CBC test can be very helpfulness in diagnosis, management and prognosis scoring in ALL patients.

Keywords: lymphoblastic leukemia, childhood, hematological features, Shafa hospital

IJBC 2015; Supplementary; P 135
Paper ID: 99

2 years' experience on Automated Continuous Flow Centrifugation Therapeutic Plasma Exchange in Mofid Children Hospital

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Abstract

Objective: Therapeutic plasma exchange (TPE) is going to be used progressively in pediatric diseases around the world by removal of Antibodies, paraproteins, toxins and other large pathologic molecules from patient`s plasma. Advanced Automated Continuous Flow Centrifugation (ACFC) machines have been developed to provide this treatment modality with less extracorporeal volume and shorter time , which made it more suitable for children. In this article we review TPE and its different methods, and indications and also report a 2 years' hospital based experience with ACFC in our hospital since 2013-2015.
Method and Material: We provided a COBE Spectra automated machine in our hospital In 2013, and after a training course for our nurses and physicians started to respond to any consultation which requested to use it. We accepted to perform TPE according to the last guideline of The American Society for Apheresis (ASFA) regarding to its category of indication, method ,frequency and number of sessions, and replacement fluids . Consequently their own physicians monitored their clinical and laboratory responses and now according to their judgment and records after 2 years we report the results of application of this method in our hospital which is the greatest single hospital based data in this field in children on our knowledge from Iran.
Results: We applied this TPE in 18 children aged from 11 months To 13 Years (mean 7.3 Y) and female-to-male ratio of 7/11. Frequency of disease groups were: neurologic (55%), nephrologic (33%), hematologic (11%). The frequency of Indication categories according to ASFA guidelines were 44.4%, 27.7%, 22.2% & 5.5% in category I to IV respectively.Mean number of TPE sessions were 8.3 (3 to 22). 44% of courses were primed with packed RBC due to low hematocrit or low body weight. Replacement fluids were Fresh Frozen Plasma in 39% and Albumin 5% in 61% .all of the patients were improved and the rate of complete and partial clinical improvement were 25%(n=2) & 75%(n=6) in category I ,80%(n=4) & 20%(n=1) in II , 25%(n=1) & 75%(n=3) in III and 100%(n=1) in IV according to clinical and paraclinical assessments by their own specialists.
Conclusion: Although it was a preliminary data on small sample size of children , ACFC -TPE led to excellent improvement especially in unexpected category II,III and IV diseases in children, who have not responded to other modalities.so it should be considered as an effective and promising alternative treatment in children.
Keywords: plasmaphresis, albumine, FFP, packed RBC.

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Paper ID: 104

Proton Therapy for Pediatric Conditions with Geant4

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Abstract

Proton beam therapy plays an important role in the management of a variety of pediatric conditions including medulloblastoma, craniopharyngioma, ependymoma, low-grade glioma, germ cell tumors, retinoblastoma, pediatric lymphoma, Ewing's sarcoma, and rhabdomyosarcoma. There is data to suggest that proton beam therapy reduces long-term morbidity from irradiation compared with photon therapy, including reducing the risk of development of secondary cancers in children. There are special simulation and treatment planning considerations when treating children compared with adults including the potential need for anesthesia in the youngest of this childhood patient population. Pediatric patients should be treated at specialized centers by a subspecialized, multidisciplinary team and be placed on clinical trials whenever feasible. Proton beam therapy shall continue to play an important role in the safe and effective radiotherapeutic management of a variety of pediatric conditions.

Keywords: Pediatrics, Proton beam therapy, Medulloblastoma, Craniopharyngioma, Ependymoma, Low-grade gliomas, Germ cell tumors

IJBC 2015; Supplementary; P 137
Paper ID: 105

Comparative proton beam therapy and photons with Geant4

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Abstract

comparative treatment plans are shown of proton beam therapy and photons for the treatment of multiple malignancies. Data is presented across multiple tumor sites including craniospinal, brain, prostate, spine, liver, and mediastinum showing the ability of proton beam therapy to reduce dose to normal organs at risk compared with photons in the properly selected patients. In the future, the list of clinical indications for proton beam therapy should continue to expand as there is growing recognition of the ability of proton beam therapy to adequately cover the intended target volume while sparing nearby normal tissues. Such sparing of nearby normal tissues from irradiation should reduce side effects of treatment and reduce the risk of developing secondary malignancies

Keywords: Proton beam therapy, Passive scatter, Pencil beam, Intensity modulated proton therapy, Comparative plan, Craniospinal irradiation

IJBC 2015; Supplementary; P 138
Paper ID: 106

Proton Therapy for Mediastinal Tumors with Geant4

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Abstract

Mediastinal tumors, especially lymphomas, are highly sensitive to the cancer-killing effects of chemotherapy and radiation therapy. These tumors are in close proximity to several normal organs at risk including heart, lungs, breast, parenchyma, esophagus, and spinal cord. It is essential to employ radiation therapy techniques that not only adequately treat the region of tumor but also shield nearby healthy tissues from the harmful effects of radiation. There is a growing body of evidence that suggests, for the properly selected patient, that proton beam therapy can be a valuable tool to deliver a conformal dose of treatment to the tumor while shielding nearby normal tissues from the harmful effects of unnecessary irradiation.

Keywords:
Mediastinum, Lymphoma, Proton beam therapy, Secondary malignancy, Coronary artery disease, 4D CT scan

IJBC 2015; Supplementary; P 139
Paper ID: 116

Umbilical Bleeding - A Presenting Feature for Congenital Afibrinogenemia

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Abstract

Introduction: Congenital afibrinogenemia is a rare blood disease that occurs due to low levels of fibrinogen and defined as plasma levels 120 sec, control 12 sec), Prothrombin time/ INR (>10 sec, control 1 sec), Activated partial thromboplastin time (>180 sec, control 33 sec), thrombin time (>240 sec, control 17 sec) were all abnormally prolonged. Factor X level was 73 mg / dL (normal 70-120 mg / dL) and level of factor XIII was abnormal. Fibrinogen level was

Keywords: congenital, a fibrinogenemia, umbilical bleeding

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Paper ID: 121

evaluate of prevalence of anemia in infant and pre schollers

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Abstract

introduction: Anemia is a disorder that caused by decrease in amount of hemogolobin or number of RBC and led to the capacity moving oxygen in blood. anemia is also a symptoms of background disease. bsed on standards of world Health organization childrens less than 11,5 g/dl Hb are anemic. based on MCV they divided to 3 groups as Microcytic (less than 80 fi), Normocytic (between 80-100 fi),Macrocytic (more than 100 fi).prevalence of anemia in developing countries are more than in developed countries and estimated that ,is in children about 43%. anemia seen in all races and all ages and in both gender of male and female .present study aimed to evaluate the prevalence of anemia and the associated with background disease among infants and pre-schoolers.

Method: in this study clinical data were retrospectively collected and was done during October2014 - March 2015 in shushtar katam- al-anbia hospital.we assed CBC index in 520 patient include: RBC ,HB, HCT, MCV, MCH, MCHC, PLT, and age and sex and clinical sign that diagnosis via medico. then data analyzed by spss.

Result: Our finding showed that there was 323 patient (62%) with anemia. microcyte hypochorome 73.4%(237 patient) with 59.9% male and 40.1% female, normocyte normochorome 11.5%(37 patient) with 54.1% male and 45.9% femal and 0.6% are macrocyte.average of hematocrite is 33.63 . average of patients age is 5.5. most background disease diagnosed is pneumonia with 30% (96 patient) and then Gastrointestinal with 18%(58 patient)

Conclusion: our finding show that microcyte hypochrome anemia is most common anemia in children that has background disease also prevalence in male more than female. since anemia in lactivorous and childhood time has unreturnable effect ,can use as data and result of this study for focus on programs of prevention and treatment strickens anemia.

Keywords: Anemia, background disease, CBC index, world Health organization

IJBC 2015; Supplementary; P 141
Paper ID: 130

The role of GSTM1 and GSTT1 in disease severity in β-Thalassemia

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1. free

Abstract

Thalassemia is a widespread disease with high mortality and morbidity rates and a wide range of symptoms. As a result of repetitive transfusions and ineffective erythropoiesis, iron overload and consequently destructive oxidation occur in various organs and due to high reactive oxidant species (ROS) concentrations, erythrocytes of a thalassemic patient are at a higher risk of apoptosis. Glutathione-S transferase (GST) enzyme family forms a defense mechanism against such destruction. These enzymes are encoded by 16 genes and are divided into five classes: α (GSTA), π (GSTP), μ (GSTM), θ (GSTT) and ζ (GSTZ). The null genotype in GTSM1 and GSTT1, with neither allele active, leads to lack of enzyme activity and increased sensitivity to oxidative stress. SNPs and deletions in the mentioned loci have been correlated with a decreased glutathione activity or total loss of it. These polymorphisms as well as the null genotype can play a role in tissue damage, including heart and liver, which is caused by iron overload in β-thalassemia patients. Finding these polymorphisms and mutations in beta-thalassemia patients can contribute to a more accurate prognosis and probability of different symptoms for each patient.

Keywords: GSTM1, GSTT1, β-Thalassemia

IJBC 2015; Supplementary; P 142
Paper ID: 131

The relationship between COL IAI polymorphisms and bone disorders in β-Thalassemia

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Abstract

Osteopenia and osteoporosis are key players in morbidity among thalassemia major (TM) patients. There is a host of different factors leading to bone density reduction in TM, one of which could be a mutation in the gene for type I collagen α-chain (COL IAI). Type I collagen is the most abundant structural protein in the bone matrix and its polymorphisms have been associated with bone mineral density (BMD). The regulator sequence of the mentioned gene is a known binding site for SP1 transcription factor, where a G>T mutation is reported to have a strong connection with osteoporosis in TM patients. The resulting altered SP1 affinity, the T allele carriers, produces more α1 collagen chain causing an imbalance between the α1:α2 chains, which in turn affects bone density. So finding these polymorphisms and mutations in beta-thalassemia patients can contribute to a more accurate prognosis and probability of different symptoms for each patient.

Keywords: COL IAI, polymorphisms, bone disorders ,β-Thalassemia

IJBC 2015; Supplementary; P 143
Paper ID: 135

Expression of Long Noncoding RNAs In pediatric leukemia

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Abstract

Introduction and Objectives: Long non-coding RNAs (lncRNA) are a type of non-coding RNAs (ncRNAs). They play several biologic roles such as imprinting, epigenetic regulation, apoptosis and cell cycling. In this paper, we highlighted new approaches for ascribing lncRNAs expression with clinical characteristics, recurrent mutation, and outcome in childhood leukemia.

Methods: In order to write this review, we used highly cited articles with keywords such as long noncoding RNAs, childhood Leukemia, gene expression presented in credible databases, Pubmed, Scopus, Google, Medline from2010 to 2015.

Result: The studies show that different expression of some long non-coding RNAs are associated with childhood leukemia.

Conclusion: Some of LncRNAs have functional roles in leukemogenesis. Therefore, they can be used as the biomarkers for prognosis of leukemia.

Keywords: Long noncoding RNA, childhood Leukemia, mutation.

IJBC 2015; Supplementary; P 144
Paper ID: 136

Childhood Anemia and Mental Retardation

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Abstract

Background: Previous studies questioned the link between early childhood anemia and detrimental child development. Objective: We conducted this study to examine the association between early childhood anemia and mild or moderate mental retardation at 10 y of age. Design: The study linked early childhood nutrition data collected by the Special Supplemental Program for Women, Infants, and Children (WIC) and school records. Hemoglobin values were used to determine the relation between anemia in early life and children's assignment in special education classes for mild or moderate mental retardation. Subjects were all participants in the WIC program. A computer program was used to link data from birth, WIC, and school records. Results: Logistic regression demonstrated an increased likelihood of mild or moderate mental retardation associated with anemia, independent of birth weight, maternal education, sex, race-ethnicity, the mother's age, or the child's age at entry into the WIC program. Conclusion: These findings support the proposition that efforts to prevent mild and moderate mental retardation should include providing children with adequate nutrition during early childhood.

Keywords: Iron deficiency, anemia, mental retardation

IJBC 2015; Supplementary; P 145
Paper ID: 137

Iron Deficiency and Contribution With Attention-Deficit/Hyperactivity Disorder

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Abstract

Background: Iron deficiency lead to abnormal dopaminergic neurotransmission and may contribute to attention-deficit/hyperactivity disorder (ADHD). In this study we evaluated iron deficiency in children with ADHD versus iron deficiency in control group. Materials and methods: In this case – control study, we evaluated thirty children with ADHD with the mean age of 9.2 ± 2.2 years and 27 controls (mean ± SD, 9.5 ± 2.8 years). Serum ferritin levels evaluating iron stores have been obtained. Results: The mean serum ferritin levels in the children with ADHD were lower than in the control group (21 ± 11 ng/mL versus 41 ± 20 ng/mL; P < .001). Serum ferritin levels were abnormal (< .001). In addition, low serum ferritin levels were correlated with more severe general ADHD symptoms measured with Conners' Parent Rating Scale (Pearson correlation coefficient; P < .02) Conclusions: These results suggest that low ferritin levels contribute to ADHD and that ADHD children may benefit from iron supplementation.

Keywords: Iron deficiency, ADHD, Children

IJBC 2015; Supplementary; P 146
Paper ID: 138

Effects of Endosulfan on some blood parameters in male rats

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Abstract

Background: Endosulfan is an organochlorine insecticide and herbicide widely used for control of insects and worms. This toxin in humans and animals by oral, inhalation and skin absorption is significant. This study aimed to investigate the effects of Endosulfan on blood parameters in rats.
Methods: In this experimental study, 30 rats were divided into five groups. The control group did not receive any substance, saline control group and experimental groups 1, 2 and 3, respectively Endosulfan did not receive any substance, saline control group and experimental groups 1, 2 and 3, respectively Endosulfan with doses of 5, 10 and 20 mg per kg of body weight once every three days for 21 days by gavage. At the end of the experiment, rats were anesthetized with chloroform, blood was collected from the heart and blood factors were evaluated by standard methods. Data ANOVA and Duncan test were analyzed.
Results: Analysis of the results showed that white blood cells and monocytes, a significant increase (05/0 ≥ p) and red blood cells, lymphocytes, and platelets in the experimental group had a significant reduction (05/0 ≥ p) relative to controls.
Conclusion: Endosulfan in significant long-term effects on blood factors that can cause serious injury and even cause anemia and related diseases and therefore its use should be handled.
Keywords: Endosulfan, blood parameters, rats

IJBC 2015; Supplementary; P 147
Paper ID: 140

Intravenous Immunoglobulin (IVIG) in Rhesus (Rh) and ABO Isoimmunization in Neonatal Hemolytic Jaundice

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Abstract

ABSTRACT Objective: Several studies have shown that prescribing intravenous immunoglobulin (IVIG) can reduce the severity of neonatal hemolytic jaundice due to Rhesus (Rh) and ABO incompatibility as well as the need for exchange transfusion. The present study aimed to evaluate the effect of IVIG in treatment of neonatal hemolytic jaundice caused by Rh and ABO isoimmunization. Materials and Methods: This clinical trial study was conducted on 80 newborns with neonatal hemolytic jaundice caused by Rh and ABO isoimmunization hospitalized in the neonatal ward of Mahdieh Hospital in Tehran, Iran from 2012 to 2013. The infants were randomly assigned to case (phototherapy and IVIG) and control (phototherapy) groups. IVIG was prescribed with the dosage of 500mg/kg during 4 hours and which could be repeated for 3 doses, if required. The severity of jaundice, duration of hospitalization for phototherapy, need for exchange transfusion and IVIG complications were studied and recorded in all patients. The data were analyzed using SPSS software, Chi square and independent T-tests. Results: In patients who received both IVIG and phototherapy, the duration of phototherapy and hospitalization as well as the need for exchange transfusion were significantly lower than the control group. Total serum bilirubin level was reduced to the average of 2.01 ± 0.17 and 0.14 ± 0.23 mg/dl in case and control groups, respectively, which the difference was statistically significant (p=0.001). The total serum bilirubin levels of the case and control groups correspondingly were reduced to 7.6±2.3 and 6±1.3 mg/dl at discharge which was significantly different (p=0.001). The duration of hospitalization, the mean duration of taking phototherapy in case group were significantly shorter than control group, 4.02±1.1 days VS 7.05±1.5 days (P=0.001) and 120.9±41.1 hrs VS 156.4±31.6 hrs (P=0.001) respectively. 15 infants required exchange transfusion; 2(5%) of case group and 13(31%) of control group which the difference is statistically meaningful (P=0.002) No IVIG complications were observed. Conclusions: Prescribing IVIG is an effective and safe treatment that can reduce the severity of jaundice, the need for exchange transfusion and duration of phototherapy in neonatal hemolytic jaundice due to Rh and ABO incompatibility.

Keywords: hemolytic jaundice, intravenous immunoglobulin, Rh and ABO isoimmunization, phototherapy

IJBC 2015; Supplementary; P 148
Paper ID: 141

Cloning and transient expression of cytoprotective factor, HO-1, in mesenchymal stem cells using the adenoviral expression system through Gateway technology

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Abstract

Abstract Background and Objectives Heme oxegenase1 (HO-1) is one of the potent cytoprotective factor. The goal of this study was to clone and transiently overexpression the human HO-1 gene in MSCs using the adenoviral expression system based on gateway technology. Material and Methods In order to induce expression of HO-1, A549 cell lines were exposed to UV for 1 hour. Full length cDNA of HO-1 was isolated and cloned into pENTR TOPO/D vector by TOPO cloning reaction. To construct the expression clone, a LR recombination reaction was carried out between the entry clone and destination vector, pAd/CMV/V5-DEST. The Recombinant virus was produced in appropriate mammalian cell line. The recombinant virus expressing HO-1 was infected to MSCs. Results The results showed that human recombinant HO-1 was successfully cloned and the accuracy of the gene and its frame in the vector were confirmed by DNA sequencing. Expression of HO-1 in MSCs was confirmed by RT-PCR and western blot analysis. The results indicated that the expression of HO-1 is transient. Conclusions HO-1-modified MSCs by using an adenoviral vector may provide a novel strategy to promote the efficiency of cell therapies flowing transplantation.
Keywords: Mesenchymal stem cell, HO-1, Adenoviral vector, Transfection, Gateway technology

IJBC 2015; Supplementary; P 149
Paper ID: 142

PAX3 and PAX7 Polymorphism in Children with Rhabdomyosarcoma: a 10-year histopathological and genetic study

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Abstract

Background: Rhabdomyosarcoma is the most common soft tissue sarcoma among children which has two major subtypes: embryonal rhabdomyosarcoma (ERMS) with more frequency and better prognosis and alveolar rhabdomyosarcoma (ARMS) with aggressive behavior and less survival rate. Distinction between these subtypes is mandatory to choose proper treatment. Histopathologic study is the main method, but nowadays cytogenetic studies like PCR are also used. The aim of this study was to evaluate the frequency of PAX3 and PAX7 mutations in children with rhabdomyosarcoma.
Method: 34 paraffin blocks were gathered during a 10-year period from our pathology department. Tumoral areas dissected and embedded in paraffin blocks for PCR study (Tissue dissection method). Pure tissue and pure RNA extraction, cDNA synthesis and PCR process were performed according to iNTRon biotechnology company kits' protocols. All of these cases were analyzed for PAX3 and PAX7 mutations.
Results: 34 cases of rhabdomyosarcomas were diagnosed in ten years in pathology department, 32 cases of them were ERMS and two of them were ARMS. None of the ERMS samples was t (2; 13) or t (1; 13) positive. Also, two ARMS cases were negative for PAX3 and PAX7 mutations.
Conclusions: This study revealed lack of PAX3 and/or PAX7 mutations in ERMS. However, careful morphological evaluation cannot replace by the PCR-based t(2;13) and t(1;13) assay of childhood sarcomas, but can be used to make certain current histopathological diagnosis.
Keywords: Children, Rhabdomyosarcoma, PAX-3, PAX-7.

IJBC 2015; Supplementary; P 150
Paper ID: 175

Iron Deficiency and Cognitive Test Scores

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Abstract

Background: Iron deficiency anemia in infants can cause developmental problems. Objective. To investigate the relationship between iron deficiency and cognitive test scores among a sample of school-aged children.
Materials and methods: In this cross-sectional study children 6 to 16 years old and contains measures of iron status including transferrin saturation, free erythrocyte protoporphyrin, and serum ferritin were evaluated. Children were considered iron-deficient if any 2 of these values were abnormal for age and gender, and standard hemoglobin values were used to detect anemia. Scores from standardized tests were compared for children with normal iron status, iron deficiency without anemia, and iron deficiency with anemia.
Results: Among the 234 children in the sample, 2% were iron-deficient. The prevalence of iron deficiency was highest among adolescent girls (5.7%). Average math scores were lower for children with iron deficiency with and without anemia, compared with children with normal iron status (86.4 and 87.4 vs 93.7). By logistic regression, children with iron deficiency had greater than twice the risk of scoring below average in math than did children with normal iron status. This elevated risk was present even for iron-deficient children without anemia.
Conclusions: We demonstrated lower standardized math scores among iron-deficient school-aged children including those with iron deficiency without anemia. Screening for iron deficiency without anemia may be warranted for children at risk.
Keywords: iron deficiency, anemia, cognition

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The role of Pax5 in leukemia: diagnosis and prognosis significance

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Abstract

Pax5 transcription factor, also known as B-cell specific activator protein (BSAP), plays a dual role in the hematopoietic system. Pax5 expression is essential in B-cell precursors for normal differentiation and maturation of B-cells. On the other hand, it inhibits the differentiation and progress toward other lineages. The expression of this factor is involved in several aspects of B-cell differentiation, including commitment, immunoglobulin gene rearrangement, BCR signal transduction and B-cell survival, so that the deletion or inactivating mutations of Pax5 cause cell arrest in Pro-B-cell stage. In recent years, point mutations, deletions and various rearrangements in Pax5 gene have been reported in several types of human cancers. However, no clear relationship has been found between these aberrations and disease prognosis. Specific expression of Pax5 in B-cells can raise it as a marker for the diagnosis and differentiation of B-cell leukemias and lymphomas as well as account for remission or relapse. Extensive studies on Pax5 along with other genes and immunomarkers are necessary for decisive results in this regard.

Keywords: Pax5 Leukemogenesis Fusion genes. B-cell development Hematological diseases

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The role of genetic factors in the development of coagulation disorders in patients with acute lymphoid leukemia

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Abstract

Acute lymphoblastic leukemia is the most common childhood leukemia. Thrombotic disorders have been reported in children with ALL from 1% to 36% depending on different protocol. It also covers a wide range of events with no sign that this rate compared with the rate in other populations (1 in every 100/000 people) is a high figure. At least 5% of the risk of venous thromboembolism embolism (VTE) in pediatric ALL, and about 30-70% of cases is asymptomatic signs. On the other hand, more than 50% of symptomatic VTE causes CNS involvement and only 30% with central venous lines (CVLs) are associated. In addition to chemotherapy, including an L-asparginase (L-ASP), genetic factors such as perfect quantitative protein C and S, and antithrombin, presence of polymorphisms of coagulation factor V and II, polymorphism 4G / 5G gene PAI-1 (plasminogen activator inhibitor- 1) , MTHFR C677T and MTHFR TT677 mutations , increased risk of thrombotic disorders in patients with ALL. Since these polymorphisms are among the risk factors for thrombosis, and with respect to the incidence of VTE in patients with ALL, so Knowledge of the prevalence of them enable physicians to predict of thrombotic events before treatment that this policy will be appropriate treatment according to the patient's condition and resulting the prevention of thrombotic disorders, especially in cardiovascular and pulmonary embolism is the central system.

Keywords: acute lymphoid leukemia, genetic factors , coagulation disorders

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petechia and purpura algorithm

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Abstract

children with suspected bleeding disorders might present for evaluation in a variety of ways:concernings signs & symptoms,abnormal screening laboratory results,or a known family history of hemostasis defects that requires consultation.the variety in severity of different bleeding disorders also results in children presenting at diffrent ages.the goal for the consulting hematologist is to approach the child with a broad differential diagnosis in mind,narrow the options by carefully assessing the medical & family history as well as pertinent physical findings,and order the most appropriate laboratory tests to reach a diagnosis &initiate definitive therapy,if indicated.so an algorithmic approach seems to be necessary & much helpfull.

Keywords: petechia,purpura,platlete,PTT,anomaly

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Non-accidental Injury and Bleeding Disorder

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Abstract

Background: Forty two children with non-accidental injury, most of whom had bruising, were evaluated to exclude a bleeding disorder.

Materials and methods: In this cross-sectional study, following investigations were undertaken in each child: full blood count; platelet count, prothrombin time; partial thromboplastin time; fibrinogen; and a bleeding time.

Results: The results of these initial investigations were abnormal in six children (16%). One child had a severe coagulopathy secondary to spontaneously acquired inhibitory activity to coagulation factors which led to spontaneous bruising and noticeable signs of injury after a minor accident. The remaining children had several features supporting a diagnosis of non-accidental injury. In two patients it was associated with bleeding disorders in the form of von Willebrand's disease. The remaining children initially had an abnormal laboratory finding--a prolonged partial thromboplastin time--which resulted in lengthy discussions during subsequent legal proceedings.

Conclusion: Evidence of a bleeding disorder is not uncommon in non-accidental injury and the two conditions are not mutually exclusive.

Keywords: non-accidental, injury, bleeding

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Evaluation of positive blood cultures in children with malignant disease

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Abstract

Introduction With the increasing intensity chemotherapy for childhood malignancy, severe infections in these patients are becoming increasingly more common. The majority of severe infections are septicemic, and they are frequently fatal. However, institutions vary in their use of blood cultures for the evaluation of bacteremia in children with episodes of fever and neutropenia. Hence, it is recommended to identify the frequency of documented infections, organisms associated with these infections and efficacy of a standardized antibiotic regimen in children with malignant disease under defined conditions. Materials and methods The records of all the blood cultures drawn from children with malignancy at the Shafa Hospital, Ahvaz, Iran were examined during 2014-2015. All the patients with hematologic malignancy and malignant solid tumors were included. Patients with fever and neutropenia criteria were investigated in the term of isolated bacteria and susceptibilities to antibiotics. Results During the observation period in 62 children with fever and neutropenia criteria; positive blood culture was seen in 37 with leukemia and lymphoma and 25 with malignant solid tumors. These occurred in 39 male and 23 female patients, of a mean age of 2.3 years (range 0.5–15). The most frequent diagnoses were ALL (24.1% of patients), lymphoma (11.2%) and neuroblastoma (9.6%). Staphylococci (30.6%) (Both coagulase-negative and coagulase-positive strains), Pseudomonas and E.coli (17.7% each) were the organisms most frequently isolated from patients with episodes of fever and neutropenia. High degree of antibiotic resistance is shown for Cefixime (94.7%) and Vancomycin (47.3%) in Gram-Positive Blood Cultures and Vancomycin (97.6%), Cefixime (76.7%) and Ceftriaxone (67.4%) in Gram-Negative Blood Cultures. Conclusion Neutropenia is common during chemotherapy treatment for hematologic cancers. The risk of developing infection associated with neutropenia is directly related to both the severity and duration of neutropenia. Our analysis highlights the need to refine the role of obtaining blood cultures and proper use of antibiotics in pediatric cancer patients with fever and neutropenia.

Keywords: pediatric neoplasm, neutropenia, fever, infection

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Earlypresenttionof MacrophageAvtivationSyndrome in a newly diagnosed systemic Juvenile Rhematoid arthritis- a case report

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Abstract

Macrophage activation syndrome (MAS) is a rare and fatal complication of rheumatic disorders in children and its presentation as the first symptom of rheumatic disorders is rarer. We describe a 9-year -old girl in whom MAS developed as a first complication and symptom of systemic juvenile rheumatoid arthritis (S-JRA). She suffered from fever and rash followed by multiple joints swelling for about one month before admission. Physical examination revealed cervical lymphadenopathy and hepatosplenomegaly. Laboratory findings were: abnormal liver enzymes and increased ferritin levels, coagulopathies resembling disseminated intravascular coagulation, anemia and thrombocytopenia.
Hyperplasia of hemophagocytic macrophages was remarkable in her bone marrow. Dexamethasone therapy resulted in clinical and laboratory improvements.

Keywords: macrophage activation syndrome, juvenile rheumatoid arthritis, rash

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Paper ID: 184

Comparison of Oral versus Intravenous Busulfan in Patients Undergoing Hematopoietic Stem Cell Transplantation

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Abstract

Comparison of Oral versus Intravenous Busulfan in Patients Undergoing Hematopoietic Stem Cell Transplantation Background: Hematopoietic stem cell transplantation (HSCT) is a potentially curative therapy of hematologic and non hematologic malignancies. Before HSCT, patients undergo a conditioning regimen to eradicate the malignant or abnormal bone marrow (BM) and suppress the immune system. Busulfan(BU) is an alkylating agent that has been used since 1950s. Oral BU is commonly used as conditioning regimen. But, BU has a narrow therapeutic window. The high BU exposure is associated with toxicity such as Hepatic Veno Occlusive Disease (HVOD) or sinusoidal obstruction syndrome (HVOD/SOS), regimen-related toxicity (RRT) while, under exposure is associated with graft rejection and relapse. So intravenous BU (IVBU) was introduced for the treatment of patients as an alternative to its oral type. IV BU provides proficient myeloablation and ensuring high rates of engraftment in Pediatrics and adult patients. The aim of comparison of oral versus intravenous Busulfan in in patients undergoing HSCT to determine the efficacy and practicality of HSCT with IV BU. Methods: In this review article we surveyed to gather the available literature in the Pubmed database during 2000 to 2015 to find original articles, bibliographies and review books. Results: Regarding to studies, oral BU were switched to the IVBU because of The therapeutic potential of oral BU has been compromised, due to unpredictable levels of absorption, especially in children. In particular, attaining adequate BU exposure is also problematic because need to administer a large number of BU tablets. Therefore, may be problematic Especially, in patients with difficult swallowing a large of number of tablets and those with anticipatory vomiting. Oral BU need for drug monitoring and dose adjustment, unlike IV BU. IV BU reduce both intra – and inter individual variability of BU pharmacokinetics and unpredictable standard dosages, it can offer constant dosing and pharmacokinetics profiles. Conclusion: So, an IV formulation of BU was developed to get over the problems associated with oral BU, It has convincing evidence of suitable pharmacokinetics behaviors and can be more convenient for patients and caregiver. The result of this study suggests that the use of IV BU can replace its oral formulation as part of conditioning regimens with no deleterious effect on transplantation outcomes. Also the use of IV BU in new conditioning regimens has been suggested in the future. Key words: Busulfan , myeloablation, hematopoietic stem cell transplantation,

Keywords: Busulfan , myeloablation, hematopoietic stem cell transplantation

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Paper ID: 186

Maternal Height with Child Mortality, Anthropometric Failure, and Anemia

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Abstract

Background: Prior research has focused on risk factors such as maternal behaviors, dietary factors, and immediate environmental conditions.

Objective: To examine the association between maternal height and child mortality, anthropometric failure, and anemia.

Material and methods: We retrieved data from the 2012-2013. The study population constitutes a nationally representative cross-sectional sample of children aged 0 to 59 months and born after January 2012 (n = 1321) to mothers aged 15 to 49 years. Information on children was obtained by a face-to-face interview with mothers, with a response rate of 94.5%. Height was measured with an adjustable measuring board calibrated in millimeters. Demographic and socioeconomic variables were considered as covariates. Modified Poisson regression models that account for multistage survey design and sampling weights were estimated. Mortality was the primary end point; underweight, stunting, wasting, and anemia were included as secondary outcomes. Results In adjusted models, a 1-cm increase in maternal height was associated with a decreased risk of child mortality (relative risk [RR], 0.978; 95% confidence interval [CI], 0.970-0.987; P < .001), underweight (RR, 0.971; 95% CI, 0.968-0.974; P < .001), stunting (RR, 0.971; 95% CI, 0.968-0.0973; P < .001), wasting (RR, 0.989; 95% CI, 0.984-0.994; P < .001), and anemia (RR, 0.998; 95% CI, 0.997-0.999; P = .02). Children born to mothers who were less than 145 cm in height were 1.71 times more likely to die (95% CI, 1.37-2.13) (absolute probability, 0.09; 95% CI, 0.07-0.12) compared with mothers who were at least 160 cm in height (absolute probability, 0.05; 95% CI, 0.04-0.07). Similar patterns were observed for anthropometric failure related to underweight and stunting. Paternal height was not associated with child mortality or anemia but was associated with child anthropometric failure.

Conclusion: Maternal height was inversely associated with child mortality and anthropometric failure.

Keywords: anemia, maternal height, child mortality

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Prevalence of Iron Deficiency Based on Lab. Indexes in Children with None-organic Growth Referred to Imam Khomeini Hospital

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Abstract

Introduction: The iron deficiency anemia is one of the most common anemia in children with poor diets. Appetite in infants with abnormal growth is more affected by iron deficiency anemia, aggravating the anemia in such situations. Anemia can cause depletion of iron deposits and, as a result, the level of iron in serum will decrease. MCV, hemoglobin and the level of ferritin will also decrease.
Method: This prospective analytical study was carried out on 195 children (86 males & 94 females) using Gomes method for analysis of weight, height and growth abnormalities. Data were collected using questionnaires, physical examinations by physicians, laboratory tests for MCV, iron, Hb and level of serum ferritin.
Results: 145(80.6%) out of all the 195 cases showed low levels of wasting, while 27 ones(15%) had moderate, and only 4.4% people were diagnosed with severe wasting. According to Hb level, 97 (67%) out of 145 were among patients with mild wasting, 20 (73%) out of 27 among children with moderate wasting, and 3 (75.) out of 4 amongst those with severe wasting showed iron deficiency anemia. Concerning ferritin level in serum, 123 (84.8.) out of 145 individuals with mid wasting, 26 out of 27(96.3.) among those with moderate wasting, and finally 3(75.) out of 4 of those with severe wasting demonstrated a decrease, (P=71). The iron deficiency anemia using MCV index was detected in 40% of mild wasting infants and 48% of moderate wasting children and finally in 75% of those with Severe wasting.
Conclusion: As the correlation between MCV levels and the anemia was statistically significant, this index is a better means for anemia assessment than other indexes such as ferritin and Hb levels.
Keywords:
iron, anemia, children, nonorganic disturbance

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Autoimmune lymphoproliferative syndrome: a case study 14 year old boy

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Abstract

Introduction: Autoimmune lymphoproliferative syndrome (ALPS) is a rare genetic disorder leading to autoimmune cytopenia. Because of differential diagnosis for lymphadenopathy, splenomegaly, and cytopenia, diagnostic criteria for ALPS and other lymphoproliferative disorders, such as hemophagocytic lymphohistiocytosis (HLH) have been developed. In this case report, we describe a 14-year-old boy with ALPS after a presumptive diagnosis of HLH.

Case presentation: A 14-year-old boy was admitted with seven months of left side of abdomen pain and continuous fever. He had palpable cervical and axillary lymphadenopathy. His spleen was palpable 5 cm below the left costal margin, and liver edge was palpable 4 cm below the right costal margin. Hemoglobin was 6.9 g/dl, hematocrit was 27.9%, platelet =57000 /mm3, and ferritin=354 ng/ml. IGM=290 mg/dl (normal= 40-230 mg/dl), IGG=3300 mg/dl (normal = 700-1600 mg/dl), CD3=185mg/dl (normal=90-180 mg/dl), and CD4=50 mg/dl (normal=10-40 mg/dl). Double negative (DN) T cells were positive. He also showed hypofibrinogenemia with Plasma fibrinogen=150 mg/dl (normal=180-540 mg/dl). Serum vitaminB12 was normal. Abdominal computed tomography showed huge splenomegaly. After viral etiologies were ruled out, a diagnosis of HLH was considered because he met 4 of 8 criteria. A repeat ferritin measurement 1 month later was 876ng/ml providing a fifth criterion for HLH. Treatment of HLH was initiated. To catch a more definitive tissue identification and because the patient was experiencing discomfort from her significantly enlarged spleen, a splenectomy was performed. The pathology report revealed an enlarged spleen showing hypersplenism with widened splenic cords and rare to absent hemophagocytic activity. It was also noted that the spleen had atypical T-cell hyperplasia with DNTs, consistent with ALPS. He was treated with prednisone 15 mg once daily and immunomodulator 150 mg three times a day, the dose of prednisone was reduced gradually. Till now, the patient has been treated and observed for 7 months. DISCUSSION ALPS and HLH have many overlapping clinical and laboratory features. Both are lymphoproliferative syndromes, present in childhood and are characterized by persistent lymphadenopathy/splenomegaly with evidence of hyper inflammation and cytopenia. However, distinguishing between ALPS and HLH is vital because the managements are very different. Ferritin level can help differentiate ALPS from HLH, because the ferritin level in ALPS patients is generally lower than 3000 ng/mL, which is reportedly more specific for HLH. Conclusion: We have highlighted this case to emphasize the necessity of considering rare disorders, particularly ALPS, in the differential diagnosis of patients presenting with lymphadenopathy, splenomegaly and cytopenia.

Keywords: ALPS, HLH, lymphadenopathy