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Aims and Scope

The *Journal of Taibah University Medical Sciences* is a bi-monthly peer reviewed publication. Authors are invited to submit articles for publication, reporting original work in clinical and basic medical sciences covering topics from Medicine, Dentistry, Nursing, Pharmacy, and Applied Medical Sciences. Review and Editorial articles are by invitation only. However, those received and found to be of an outstanding nature will be considered for publication. Other regular features within the journal include, Case Reports, Letters to the Editor, and Updates which detail symposia, conferences and workshops located primarily in the Kingdom of Saudi Arabia and the Gulf countries.

The Journal has recently devoted a new Students’ Section where the research projects written by the students shall be entertained. All students from the medical, dental, and allied science faculties are invited to participate in this educational and research activity. The peer review shall be performed by senior academicians with a view to guide and supervise the students in medical research and publications.

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- يبوب المقال على النحو التالي :
 - ١) العنوان ويكون مختصرا ومعبرا. ٢) اسم المؤلف (المؤلفين). ٣) عنوان المؤلف، وإذا كان هناك أكثر من مؤلف تظهر العناوين بنفس ترتيب ظهور الأسماء في البحث. ٤) الملخص: لا يزيد عن ٢٥٠ كلمة وأن يكون مستقلا بذاته ويفني عن قراءة المقال. ٥) الكلمات المفتاحية: وموقعها تحت الملخص. ٦) العنوان البريدي الكامل للمؤلف وتاريخ تقديم البحث.
- الملخص باللغة العربية: يجب أن لا يزيد عن ٢٥٠ كلمة يلخص من خلالها العمل بطريقة بسيطة واضحة ومختصرة ويكون مطابقاً تماماً للملخص باللغة الإنجليزية. أما تقارير الحالات المرضية فيجب أن لا يزيد الملخص فيها عن ١٠٠ كلمة. كما يجب أن يحتوي الملخص العربي على الكلمات المفتاحية باللغة العربية.

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- أي تعديلات أو إضافات أو أي تغييرات غير ناتجة عن أخطاء الطابع يتكفل بها المؤلف.

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ردمد ٣٦١٢ - ١٦٥٨

(يعتبر المؤلفون مسئولون مسؤولية كاملة عن جميع الآراء والمعلومات الواردة في البحوث المنشورة بالمجلة)



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 - Highest degree of each author (only the acronym of the completed degree should be provided)
 - Short Running Title (40 letters)
2. Structured abstract with a maximum of 300 words excluding keywords. No need for structured abstract for Narrative Review and Case Reports.
3. Abstract should have following headings:
 - o Objectives
 - o Methods
 - o Results
 - o Conclusions
 - o Maximum of six Keywords (alphabetical order, separated with semicolons, use capital letters only when it’s necessary)
4. Manuscript should have the following headings:
 - o Introduction
 - o Methods
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 - o Discussion
 - o Conclusions
5. Appropriate reporting guidelines checklist included (see “Reporting guidelines” section above)

6. In-text citations uses superscript Vancouver style with no brackets.
7. Acknowledgment (if any)
8. Funding sources
9. Conflict of interest
10. Ethical approval and statement of Declaration of Helsinki (if applicable)
11. References List (Vancouver style)
12. Legends of Tables and Figures
13. Tables and Figures
14. Maximum word count allowed: Original articles and Systematic Reviews: 5000 words; Brief Reports and Case Reports: 1500 words.
15. The journal is a member of Committee of Publications Ethics (COPE) and accepts a maximum threshold of 20% similarity index. Any submission with similarity index of 20% or more will be considered as plagiarism and the article will not be processed by the journal.
16. All authors should meet all the four criteria stated in the “Authorship” section above.
17. Authors’ contributions: Stated using the relevant CRediT roles. For example:
KIK conceived and designed the study, conducted research, provided research materials, and collected and organized data. HMY analyzed and interpreted data. SYG wrote initial and final draft of article and provided logistic support. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

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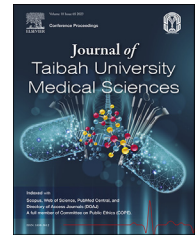




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Abstracts

FREE PAPER SESSION

M-O-101

Assessing Bone Mineral Density in Sickle Cell Disease Patients & Correlation to Frequency of Admission – Prospective - Uni Center Study

Rehab Yusuf AL-ANSARI¹, Amal Shilash², Alexander Woodman³, Sama Khalid Tawfiq², Majed Aldawish², Asem Banjar², Tawasoul Fadoul², Mushref Ali Algarni², Ali Al Orf², Sameerah Motabgani⁴, Ahmad Habib²

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Abstract Background: Sick cell disease (SCD) is an inherited autosomal recessive disorder with decreased bone mineral density (BMD) as a common clinical manifestation of SCD. Saudi Arabia has one of the highest rates of SCD in the world, up to 2.6% of the population.

Objectives: The primary endpoint is the assessment of bone mineral density in patients with SCD. The secondary endpoint is to examine its correlation with hospitalization rates in patients with SCD.

Methods: Observational prospective study in the tertiary care hospital among n = 119 SCD patients. Patients were divided into two groups: Group A - severe SCD patients requiring hospital care - 3 per year; Group B with a smooth course of SCD that did not require frequent hospitalization. Analysis was based on the frequency of hospitalizations with pain crises and assessment of bone mineral density.

Results: Of n = 119 patients, 73.1% had low bone density. The prevalence of low bone density in the spine (62.6%) was higher than in the femur (48.5%). There was no significant difference between the two groups in the prevalence of low bone mineral density (36% vs 38%, p = 0.081). Patients with more frequent hospital visits had significantly higher Mg concentrations (2.30 vs 0.84, p = 0.001), higher GGT (59.44 vs 39.49, p = 0.030), and significantly lower vitamin D (34.82 vs 49.48, p = 0.004). There was no significant correlation between the frequency of the admission or severity of the disease with low bone mineral density value.

Conclusions: The prevalence rate of low BMD was significantly high among SCD patients in our cohort. However, there was no correlation between the BMD findings and the frequency of admissions. Further research is needed to answer the proposed debate about the accuracy of DEXA scanning as diagnostic imaging in patients with SCD with low blood supply to the affected bone."

<https://doi.org/10.1016/j.jtumed.2023.10.008>

M-O-102

Epidemiologic study of hereditary bleeding disorders in Dubai, UAE :A single center experience

Yasmine Tarek Elsherif¹, Maroun Elkoury¹, Faraz Khan¹, Shabeeha K. Rana¹, Mehran Karimi¹

¹ American Hospital Dubai

Abstract Background: Knowing the prevalence of common and rare hereditary bleeding disorders (HBDs) is important in areas where national registry is lacking.

Aim: Reported bleeding disorders in the Gulf region are scarce, so our study aims to evaluate the epidemiologic features of patients with HBDs in Dubai, United Arab Emirates (UAE).

Methods: A cross-sectional retrospective study, conducted on 118 patients with HBDs during the period of 2019 to 2023 in the hematology clinic, American Hospital Dubai (AHD), UAE. These patients were categorized in an Microsoft Word doc. 2013 version 15 and Microsoft EXCEL. 2013 version 15. The information was arranged in tables for easy review where description of the patient clinical manifestation, type of factor and level ,treatment approach and complication were noted of the HBDs including von Willebrand disease (vWD), Hemophilia A and B , factor I (FI) deficiency, factor II (FII) deficiency, factor V (FV) deficiency , factor VII (FVII) deficiency, factor X (FX) deficiency , factor XIII (FXIII) deficiency, Vitamin K–dependent clotting factor deficiency (VKCFD),Platelets dysfunction ,Bleeding of Unknown cause (BUC) and combined deficiencies. The HBDs were diagnosed on the basis of a coagulation profile, coagulation factor assays results obtained by IL ACL TOP 700 model in Mayo Clinic.

Results: It was noted vWD type I was the most recorded out of all the HBDs (n=42, 35.6%) followed by FVII deficiency (n=21, 18%) and factor VIII (FVIII) deficiency (n=20, 17%). Most of the patients received on-demand treatment. RBDs have been reported in 35.5% (n= 42) including combined factor deficiency which was present in 6 patients (5%). Male to female ratio was 69/49 (1.4:1). Fifty-two patients were pediatric (<18 years). The mean age of all participants was 23.9 years, and for pediatric population was 8.49. In this study, consanguine marriage was 16.9% (n=20).

Conclusion: To best of our knowledge, this is the first comprehensive study that reported HBDs from a referral single center, in Dubai, UAE. Our study showed vWD type I is the most common of all HBDs followed by FVII deficiency which is

higher than hemophilia A which is against previous published literature. On the other hand, our data revealed RBD (35.5%) is not rare in this region. This epidemiologic study guides us to design a comprehensive hemophilia treatment center in order to establish a proper treatment guideline according to the frequency of HBDs in our geographic area.

<https://doi.org/10.1016/j.jtumed.2023.10.009>

M-O-103

Subclinical Atherosclerosis in Childhood Acute Lymphoblastic leukemia Survivors: a Tertiary Center Experience

Laila Sherief¹, Elhamy R. Abd El-khalek², Ibrahim A. Libda³, Osama A. Gaber⁴, Naglaa M. Kamal⁵, Basma K. Soliman³, Wesam A. Mokhtar², Gehad M. Kamar⁶, Marwa Zakaria²

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Abstract Background: An increased risk of cardiovascular complications is reported in survivors of childhood acute lymphoblastic leukemia (ALL). Early identification of impaired vascular health may allow for early interventions to improve outcomes.

Aim: The study was conducted to assess the endothelial dysfunction in ALL survivors using a new marker, serum endocan, and measurement of the mean common carotid arteries intima media thickness (cIMT).

Methods: A case-control study was conducted on 100 childhood ALL survivors (aged 6–18years), with 80 healthy age and sex-matched children as a control group. Lipid profile, hepatitis markers, and serum ferritin were measured, in addition to the measurement of serum endocan. and cIMT by B-mode high-resolution ultrasonography for all study participants.

Results: Triglycerides, total cholesterol, post prandial glucose, and serum ferritin were significantly higher in ALL survivors than controls (p

<https://doi.org/10.1016/j.jtumed.2023.10.010>

M-O-104

The effect of different types of anemia on HbA1c levels in non-diabetics

Basil Alzahrani, Medical student¹, Faisal Saleh Alsharm¹, Hassan khaled Salamatullah¹, Abdullah Osama Abukhodair¹, Jamil Mohammed Baljoon¹, Hebah Malaikah², Mohammed Eldigire Ahmed¹, Suhaib Radi³

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Abstract Background: Diabetes mellitus is one of the most common diseases worldwide with significant morbidity and mortality. HbA1c remains one of the most important methods

for diagnosis and monitoring of the disease. Since HbA1c is a reflection of the glucose attached to red blood cells, factors affecting hemoglobin and red blood cells' half-life can influence HbA1c measurements.

Objective: This study aims to evaluate the effect of different types of anemia including iron deficiency anemia, sickle cell anemia, β -thalassemia trait, and megaloblastic anemia on HbA1c levels in a tertiary hospital over the past 6 years (2016–2022).

Method: This is a retrospective chart review study of 324 patients including those with one of the four types of anemia mentioned above and a control group. The control group were healthy adults with normal HbA1c and hemoglobin, who were not known to have diabetes or anemia. Patients with diabetes or prediabetes based on self-reporting or elevated fasting, random blood sugar, or 2 hours post-prandial blood glucose were excluded.

Results: The mean HbA1c levels were significantly higher in sickle cell anemia at 5.83% (95% CI = 5.39–6.28) and in iron deficiency anemia at 5.75% (95% CI = 5.68–5.82) when compared to the control group at 5.32% (95% CI = 5.22–5.41). However, the mean HbA1c levels in megaloblastic anemia were 5.38% (95% CI = 5.26–5.5) and 5.45% (95% CI = 5.21–5.69) in beta thalassemia trait, which were not significantly different when compared to the control group. HbA1c significantly decreased from 5.75 to 5.44% after treatment in the iron-deficient group with a p-value of

<https://doi.org/10.1016/j.jtumed.2023.10.011>

M-O-106

The Perception of Pediatric Sickle Cell Anemia Patients' Caregivers toward Bone Marrow Transplantation at NGHJ Jeddah, Saudi Arabia

Manar Betar, Medical student¹, Dania Emad Faidah¹, Fay Nabeel Alnafisi¹, Reema Enad Alotabi¹, Dania Mongel¹, Najlaa Al imam¹, Anwar Khan¹

¹King Saud bin Abdulaziz University for health and science

Abstract Background: Sickle cell disease (SCD) is a relatively common genetic disorder in Saudi Arabia characterized by the predominance of sickle hemoglobin (HbS). Although multiple supportive care options exist for patients with SCD, hematopoietic stem cell transplantation (HSCT) is the only cure available and has become highly successful, with an almost 91% overall survival rate. However, pursuing this procedure is still restrained as a curative treatment option.

Objective: This study aimed to evaluate the perception of parents' caregivers at the National Guard Hospital pediatric hematology clinic regarding using HSCT as a curative approach for their children with SCD.

Methods: This is a cross-sectional study of the interviewer-administered survey distributed utilizing electronic devices to caregivers of pediatric patients with SCD. Subjects were recruited from Pediatric Hematology & Oncology clinics at National Guard Hospital Affairs in King Abdulaziz Medical City, Jeddah, Saudi Arabia. An estimated sample size of 100 was initially calculated out of 140 pediatric SCD patients; 72 responses were collected from participants. All study participants gave informed consent. All results were analyzed using

SPSS; moreover, statistics were set at a CI of 95% and a $p < 0.05$. In addition, inferential and descriptive statistics were done.

Results: Of all respondents, 42 (67.8%) would accept HSCT if their hematologist recommended it. However, approximately 7 (11.3%) were not interested in the procedure, and the rest, 13 (21%), were uncertain. The most reported reasons for HSCT rejection among all respondents were attributed to side effects 31 (50.8%), lack of knowledge 8 (13.1%), and misconception toward the procedure 22 (36.1%).

Conclusion: The results of this study were consistent with the fact that most caregivers would follow along with HSCT if it seemed to be fit and was recommended by their hematologists. However, to the best of our knowledge, our study being the first of its kind in the region, further research in the kingdom on the perception of HSCT is needed. Nonetheless, further patient education, an increase in caregivers' knowledge, and enlightenment of the medical team on HSCT as a curative option for sickle cell disease are vital.

<https://doi.org/10.1016/j.jtumed.2023.10.012>

M-O-107

Changes in the Fibrinolytic System of Patients Infected with Severe Acute Respiratory Syndrome Coronavirus2

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Introduction: In this study, coagulation and fibrinolysis parameters and their association with disease severity were investigated in coronavirus disease (COVID-19) patients.

Materials and Methods: COVID-19 patients (n = 446) admitted to our institute between 21 February 2021 and 17 March 2022, were recruited. Clinical data and staging were collected from all patients. Blood samples were collected and analyzed for several parameters of fibrinolysis and coagulation, including alpha-2-antiplasmin ($\alpha 2AP$) and plasminogen, thrombin activatable fibrinolysis inhibitor (TAFI), tissue plasminogen activator (tPA), plasminogen activator inhibitor-1 (PAI-1), D-dimer, and fibrinogen levels.

Results: The TAFI, fibrinogen, and tPA levels were significantly higher in participants who died compared to that of patients who recovered ($p < 0.001$). However, PAI-1, tPA, and TAFI were significantly higher in patients admitted to the ICU than those of the healthy controls ($p < 0.001$ for

PAI-1 and tPA; $p = 0.0331$ for TAFI). Our results showed that stage C and D COVID-19 patients had significantly higher levels of PAI-1 ($p = 0.003$). Furthermore, stage D COVID-19 patients had significantly higher tPA and TAFI values ($p = 0.003$).

Conclusions: Hypofibrinolysis was the most prevalent condition among patients with severe COVID-19. In this study, several coagulation markers were elevated, making them suitable prognostic markers for hypofibrinolysis.

<https://doi.org/10.1016/j.jtumed.2023.10.013>

M-O-108

Oral health status and the impact of Oral health promotion in Children& Adolescents with Bleeding Disorders

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Abstract Background: Understanding the disease-specific risks as well as patient-related barriers of children and adolescents with bleeding disorders is necessary for primary oral health promotion.

Objective: Our goal was to assess the oral health status and the impact of oral health promotion among children and adolescents with inherited and acquired bleeding disorders.

Methods: Seventy children and adolescents with inherited and acquired bleeding disorders were recruited from Ain Shams University Pediatric hematology clinic. This study was conducted in two phases; phase I included a detailed dental history, a complete clinical intraoral examination, completion of the Arabic version of child oral health related quality of life (OHRQOL) and child oral health impact profile questionnaires, and an oral health education using a specially designed booklet. Phase II was conducted after 6 months in which the effectiveness of the oral hygiene promotion was evaluated clinically and through the (OHRQOL) questionnaires.

Results: Our cohort included 33 (47.1%) patients with ITP, 27 (38.6%) hemophilia A patients, and 10 (14.3%) with inherited thrombasthenia. Sixty-two (95.4%) patients showed high caries risk assessment. Most of the patients did not brush their teeth (47.7%) with the majority reporting significant carbohydrate intake (95.4%). Forty patients (57.1%) had dental caries, with 90.0% (63) showing fair oral hygiene status with variable degrees of gingivitis (mild in 38.6%, severe in 11.4%). When evaluating the baseline (OHRQOL), we found that the self-image score was significantly inferior among patients with inherited bleeding disorders, while the psychological domain

for family was greatly affected among patients with ITP. Disease duration among patients with inherited bleeding disorders showed a significantly positive correlation with the oral symptomatology domain for child, psychological domain for the child & family as well as the social domain for child. After 6 months, there was a significant reduction in the oral debris index, the modified gingival index, the percentages of cases with oral bleeding, and a significant increase in the percentages of patients who brush once a day. The follow up (OHRQOL) questionnaires showed a significantly improved mean total score (35.67 ± 6.93) compared to the initial mean total score (43.36 ± 10.13) ($p=0.000$).

Conclusion: After 6 months of the oral health education, the Oral health related QOL scores had significantly improved, caries experience decreased, and the gingival health and oral hygiene status were acceptable among patients with bleeding disorders. A team effort is needed to improve the oral health status of these patients.

<https://doi.org/10.1016/j.jtumed.2023.10.014>

M-O-112

Evaluation of safety and efficacy of Emicizumab prophylaxis in Egyptian pediatric patients with Hemophilia A: Single center experience

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"Evaluation of safety and efficacy of Emicizumab prophylaxis in Egyptian pediatric patients with Hemophilia A: Single center experience

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Pediatrics department, Faculty of Medicine, Zagazig University-Zagazig-Egypt.

Abstract Background: Hemophilia A (HA) is an X-linked hereditary bleeding disorder caused by deficiency of coagulation factor (F) VIII activity. Emicizumab is a bispecific monoclonal antibody that mimics the cofactor function of activated FVIII and prevents bleeds in patients with hemophilia A regardless of inhibitor status. Emicizumab prophylaxis is expected to reduce the risk of severe bleeds in those patients with their subsequent complications. However, data about its safety and efficacy in Egyptian people with hemophilia A is limited.

Objective: We aimed to evaluate safety and efficacy of Emicizumab prophylaxis in Egyptian pediatric patients with Hemophilia A.

Methods: This prospective cohort study included 88 patients diagnosed with hemophilia A who started on biweekly subcutaneous Emicizumab 3 mg/kg, following a 4-week induction. The study was conducted at pediatric hematology outpatient clinic of Zagazig University Hospitals, Egypt. For evaluation of efficacy of Emicizumab, breakthrough bleeding episodes as well as annualized bleeding rate were reported for all

patients before and after Emicizumab prophylaxis. For evaluation of the safety of Emicizumab, all adverse events during prophylaxis were reported.

Results: 92.2% of the studied patients had severe hemophilia A. 17% of patients were positive for FVIII inhibitors. Joint bleeds were present in 94 % of the patients. 58% of patients had one target joint, 36.4% had more than one target joint while 5.6% had no target joints. The median annualized bleeding rate was 48 before treatment with Emicizumab versus zero after the use of Emicizumab (p

<https://doi.org/10.1016/j.jtumed.2023.10.015>

M-O-113

Multiplex-PCR is a useful and economical method to detect large exon deletions in severe hemophilia A in developing countries: Experience of Tunisia

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Abstract Background : Philadelphia-negative (Ph neg) chronic myeloproliferative neoplasms (MPNs) are characterized by proliferation of one or more myeloid cell lineages and include polycythemia vera (PV), essential thrombocythemia (ET) and primary myelofibrosis (PMF). Somatic mutations in the JAK2, CALR, and MPL genes have been described as responsible for more than 90% of MPNs patients. In the past, the presence of these mutations was thought to be mutually exclusive. More recently, studies reported the coexistence of two driver mutations.

Objective: The purpose of our study was to determine the frequency of coexisting of mutations: JAK2V617F and CALR mutations, in MPNs Tunisian patients followed in Aziza Othmana Hospital.

Patients and method: Our study is prospective from December 2021 to June 2023.

MPNs Ph-neg patients followed in Aziza Othmana Hospital were included. Mutations in JAK2 V617F and exon 9 of CALR, were simultaneously tested. Total DNA was extracted using the salting out technique. The Allele-Specific PCR was done to detect JAK2V617F. PCR and DNA fragment analysis, were used to detect CALR exon 9 mutations.

Results : 219 patients were included and 11 have simultaneous JAK2V617F and CALR mutations with a frequency of 5.02%: divided into 46.1% in MFP (n=6), 30% in ET (n=4) and 7.6% in PV (n=1).

Conclusion: The coexistence of JAK2V617F and CALR mutations is not a frequent event in MPNs patients in the literature. Based on our descriptive data, this frequency seems to be higher than the few reported data. It may be explained by the genetic specificities of our cohort. Nevertheless, correlation studies with JAK2V617F and CALR mutations and clinical features are necessary, as the evolution to define the impact on outcome for those MPNs patients carrying both mutations.

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M-O-114

Patient Care Strategies for alloantibodies in Type 3 Von Willebrand Disease: A Southern Tunisia Perspective

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Introduction: A minority of patients with type 3 von Willebrand disease (VWD3) develops alloantibodies to exogenous von Willebrand factor (VWF) and /or factor VIII (FVIII). These antibodies pose a significant challenge in the management of patients with VWD3.

Aim: Our study aims to assess the therapeutic approaches and clinical characteristics of patients with VWD3 who had developed inhibitors.

Patients and methods: Study Design: This retrospective study focused on patients diagnosed with VWD3 who developed alloantibodies between January 2018 and August 2023. The study was conducted jointly at the Clinical Hematology Department of Hedi Chaker University Hospital and the Hematology laboratory of Habib Bourguiba University Hospital both located in Sfax, in southern Tunisia. Neutralizing inhibitors' screening and titration were performed using the Bethesda method and non-neutralizing inhibitors were detected through substitution therapy recovery tests. Hemorrhagic events were managed through a multidisciplinary team.

Results : Alloimmunization was observed in 12% (6 out of 49) of VWD3 patients. Specifically, one case developed anti-VWF, two had non-neutralizing inhibitors and three exhibited both anti-VWF and anti-FVIII.

The first patient experienced elbow hemarthrosis resistant to substitution. Anti-VWF antibodies were detected with a titre of 5.8 Bethesda units (BU). Treatment involved using a by-passing agent (Novoseven®) combined with immunosuppressor (Azathioprine -Imurel®) which allowed an inhibitor level decrease to 0 BU and a successful reintroduction of Immunate®.

The 2nd and 3rd patients had non-neutralizing anti-VWF antibodies detected. The 2nd patient's inhibitors were discovered during an episode of gingivorrhagia that did not respond to substitution. The 3rd patient's inhibitors were identified during a preoperative evaluation. Both cases were successfully treated using Novoseven®.

The final three patients had combined anti-VWF and anti-FVIII antibodies. One case revealed these inhibitors during an allergic reaction to the replacement therapy while two cases were presented with severe bleeding syndromes resistant to substitution. The latter two patients were successfully treated with by-passing agents (Novoseven®). The remaining patient received corticosteroid treatment to eradicate the antibodies. Unfortunately, the corticosteroid led to severe gastrointestinal bleeding which was resolved with by-passing agents (Novoseven®) switched to recombinant FVIII.

Conclusion: Alloimmunization in VWD has been increasingly documented over the past decade in our department. There is a paucity of literature providing guidance on its management.

By-passing agents like Novoseven®, recombinant FVIII and immunosuppressive treatments proved to be effective in treating our patients.

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M-O-115

Von Willebrand Disease laboratory diagnosis in a low middle income country

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Abstract Background: The development of inhibitory alloantibodies against von Willebrand factor (VWF) is a rare complication encountered during the treatment of type 3 Von Willebrand disease (WBD).

The pathogenic mechanism and risk factors are not well defined. The presence of an inhibitor could lead to poor response to treatment and patients may subsequently develop severe anaphylactic reactions on further exposure to the VWF. However, in the absence of a standardized technique for detecting these antibodies, screening is not systematic.

Owing to the rarity of cases reported in the literature and the absence of recommendations for the management, handling patients with inhibitor remains challenging.

Objective: The aim is to report two cases of type 3 WBD with inhibitor in a developing country and to underline treatment alternatives.

Case1: A 36-year-old woman diagnosed with WBD type 3 at age of 4 years old. She had recurrent epistaxis, treated with cryoprecipitate that showed poor clinical response. VWF inhibitor was suspected and confirmed. She was treated with plasma derived factor VIII (FVIII). During puberty, she experienced heavy menorrhagia treated with hormonal therapy.

Case2: A 15-year-old female born of consanguineous marriage, with WBD since the age of 4 years old. She presented with menorrhagia and was treated with (FVIII/VWF) concentrate. She continued to experience heavy menstrual bleeding and developed an anaphylactic reaction twice after exposure to the VWF and Testing revealed the presence of inhibitor.

Currently, the two patients are successfully treated with hormonal therapy although the screening shows that inhibitor persists.

However, Issues revolved around managing other bleeding manifestations are still challenging (antifibrinolytics, by passing agents and new generation of treatment)

Conclusion: The cases imply that the inhibitor development in WBD could be handled with hormonal therapy especially in a low resource country. Nevertheless, more data are needed regarding the expected outcomes of new treatments that have emerged to address these immune responses including Emicizumab.

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M-O-117

Epidemiological characteristics of congenital bleeding disorders in south Tunisia

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Background and aim: Congenital bleeding disorders are a heterogeneous group of disorders with varying degrees of clinical severity. They include various diseases with variable frequency. In Tunisia, there is a lack of data on bleeding disorders due to the absence of a national registry dedicated to these conditions. Consequently, it remains challenging to determine the precise number of diagnosed patients in the south of Tunisia. This study represents the inaugural presentation of epidemiological data regarding bleeding disorders in the south of Tunisia with the aim of creating a regional register of hemostasis diseases.

Patients and methods: Our study is retrospective. It includes patients with hereditary bleeding disorders diagnosed in laboratory of hematology in Habib Bourguiba hospital and/or treated in clinical hematology or pediatric department of Hedi Chaker Hospital or other regional departments in the south of Tunisia during the period from January 2000 to December 2022. We have described the epidemiological data of our patients.

Results: During the study period, 433 patients with bleeding disorders were included. The distribution of our patients according to the type of bleeding disorder shows that VWD accounts for 21%, hemophilia accounts for 18% and inherited platelet accounts for 8% of cases. Rare bleeding disorders represent 53%. Among them, factor VII and factor XI deficiency were the most common subtypes. The sex ratio for our population was 2. Median age at diagnosis was 5 years (extreme 1 day-85 years). The diagnosis before the age of 1 year was made in 24% of cases. Bleeding was the most frequent diagnostic circumstance (51%), followed by fortuitous discovery of the disease (28%) and then family investigation in 21% of cases.

Conclusions: VWD, hemophilia A, factor VII deficiency and factor XI deficiency are the most frequent diseases in our study. These results are similar to data in the global annual survey published by the World federation of hemophilia in 2021. However, the number of patients with hemophilia seems to be lower, this can be explained by lack of diagnosis of moderate and mild forms. The number of women is equal to men in our study and age at diagnosis seems to be high compared to literature. As a first step, we have compiled epidemiological data on our patients. Our aim is to create a complete register that includes all the clinical, therapeutic and developmental data on our patients.

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M-O-118

Long term outcome of allogeneic stem cell transplantation for patients with Fanconi Anemia

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Introduction: Allogeneic stem cell transplantation (ASCT) remains the only curative treatment for patients with Fanconi anemia (FA). Although it does not prevent the occurrence of secondary malignancies.

Objective: The aim of this study is to identify factors affecting outcomes post ASCT in patients with FA.

Methods: We included patients with FA who underwent ASCT from HLA-sibling donor between January 2003 and December 2021. The diagnosis of FA was confirmed by peripheral blood simple tested for cross-linker hypersensitivity by MMC. Conditioning regimen based on Fludarabine and Cyclophosphamide +/- rabbit antithymocyte globin (ATG). GVHD prophylaxis consisted on cyclosporine and short course of methotrexate.

Results: Twenty-four patients were included. The median age was 12 years (5 - 39) with sex ratio of 0.7. At ASCT, 7 patients had clonal evolution (myelodysplastic syndrome with cytogenetic abnormalities). The median time from FA diagnosis to transplant was 9 months (2 -147). Five patients received ATG with fludarabine based conditioning. Engraftment was observed in all patients. Neutrophile and platelet reconstitution were observed after a median of 12 days (10 -18) and 14 days (9 - 21), respectively. Eighteen patients developed documented infections (13 clinical and 5 microbiological documentation). Three patients developed acute GVHD - grade2. Chronic GVHD was seen in 7 patients (mild :n=5 ; moderate :n=1 ; severe :n=1). Two patients developed a secondary graft failure after 3 and 5 years of ASCT, respectively.

Clonal evolution to acute myeloid leukemia was observed in 2 patients after 32 and 69 months, respectively. A secondary malignancy (esophageal cancer) occurred in 1 patient 18 years after ASCT. After median follow-up of 120 months (3-242), 20 patients were alive and 4 patients died (graft failure (n=2); sepsis (n=1) and nephrotic syndrome (n=1)). Ten-year overall survival (OS) and event free survival (EFS) were 81% and 76%, respectively. In univariate analysis, the only factor of 10-year EFS was age.

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M-O-123

BCL2 overexpression in diffuse large B-cell Lymphoma

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Abstract Background: BCL-2 family proteins are regulators of apoptosis, their overexpression in de novo DLBCL patients is associated with a poor prognosis. This pathway has already been used as a therapeutic target in the fight against cancer -Anti-BCL2 therapies (Venetoclax) are now a standard treatment for acute myeloid leukemia. In this study, we aim to identify the clinical, epidemiological, and prognostic features of our BCL2+ DLBCL patients.

Methods: This is a retrospective, descriptive, and analytical study, from January 2018 to December 2021, and includes newly diagnosed patients with BCL2+ DLBCL during this period.

Results: among a total of 184 DLBCL patients, 36 patients (19.56%) were BCL2+, consisting of 21 women and 15 men with a F/M sex ratio of 1.4. The median age at diagnosis was 56.77 years [19-81 years], and the average diagnostic delay was 5.9 months. Half of the patients had B signs at diagnosis, and 26 (72.2%) had a tumoral syndrome.

Performans status was -1 in 30 patients (83.3%). Histological examination showed a proliferation index greater than 80% in 16 patients (44.44%), CD23 positivity in 4 patients (12%), and BCL6 positivity in 13 patients (36.2%). CT-Scan showed extensive involvement (stage III/IV) in 23 patients (63.9%). A bulky mass was described in 16 patients (44.4%). The IPI score was greater than three in 12 patients (39.9%). Four patients developed venous thrombosis.

Complete blood count revealed anemia in 30.55% of patients, lymphopenia in 16.6% of patients, thrombocytopenia in 8.33% of the patients, and neutropenia was observed in only one patient.

Thirty-one patients were placed on the RCHOP protocol. Primary mediastinal DLBCL cases were given a dose-adjusted R-EPOCH regimen. Unfortunately, three patients died before initiation of therapy. Twenty-one patients (63.6%) achieved remission after first-line therapy. Seven patients were declared refractory, and we lost three patients during follow-up.

We found a significant correlation between BCL2 overexpression and BCL6 positivity ($p=0.026$) and the refractory character ($p=0.045$). The positivity of BCL2 was not significantly associated with death ($p=0.218$). The 4 year-survival of our BCL2+ patients is 64% ($p=0.463$).

Conclusion: This poor prognosis and higher risk of non-response must be taken into account in the guidelines. The BCL2 marker will enable better stratification of DLBCL lymphoma patients and offer them more effective first-line treatment options. In theory, the future is bright for these patients, yet access to drugs may well be our toughest challenge in the years ahead.

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M-O-126

Refractory diffuse large B-cell Lymphoma

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Abstract Background: Up to 20 % of patients with diffuse large B-cell lymphoma won't respond to first-line therapy. These patients require alternative therapies such as second-line chemioimmunotherapy followed by consolidative autologous hematopoietic stem cell transplantation.

Methods: This is a retrospective, descriptive, and analytical study of newly diagnosed DLBCL patients followed up in an internal medicine and onco-hematology department between January 2018 and December 2021. Refractory DLBCL was defined as progressive disease or stable disease as the best response at any point during chemotherapy (>4 cycles of first-line or 2 cycles of later-line therapy).

Results: We analyzed 160 DLBCL patients treated in our hospital between 2018 and 2021. Among them, 19 patients (11.8%) were declared refractory. The median age at diagnosis of our refractory cases is 52.57 years old [16-72] and the F/M sex ratio is 1.37. The average diagnostic delay was 8.4 months. Of our refractory patients, 57.89% had B-signs at diagnosis, and 84.21% presented with clinically evident tumor syndrome. The performans status -1 in 14 patients. On the CT scan, 68.42% of our refractory cases had a bulky mass, and 14 patients (73.68%) had an III-IV stage disease according to the ANN-ARBOR staging system. Eleven patients had a high IPI (-3).

Eighteen patients are refractory to R-CHOP regimen as first-line treatment, while 1 patient with a mediastinal bulky, who received DA-R-EPOCH protocol, was deemed refractory as well. After the first line, 15 patients were put on the RDHAOX regimen, 2 patients on RGEMOX, and one patient received the R-COPADEM protocol. Six of these patients passed away, seven were lost to follow-up, three achieved complete remission and three were put on a third-line treatment.

Refractory DLBCL was associated with the presence of a bulky mass ($p=0.002$), adrenal involvement ($p=0.006$), pulmonary involvement ($p=0.043$), lymphopenia ($p=0.018$), BCL2 positivity ($p=0.05$), BCL6 positivity ($p=0.049$), and CD5 positivity ($p=0.048$).

The 4-year survival of refractory patients was 59.4% versus 77.7% of nonrefractory cases ($p=0.19$).

Conclusion: Refractory cases require more aggressive therapy. Current data suggest that patients can be consolidated after second-line with hematopoietic stem cell transplantation. Chimeric antigen receptor (CAR) T-cell therapy has dramatically changed the treatment of refractory and relapsed DLBCL, and it is indicated for patients ineligible for transplantation. Tafasitamab, in addition to lenalidomide, antibody-drug conjugates, and bispecific antibodies are the new weapons in this therapeutic strategy. But, unfortunately, are still associated with significant barriers to access.

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M-O-133

Can Social Education Interventions Programs Decrease Incidence of Sickle Cell Anemia? A Success Story from Khaibar, Saudi Arabia.

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Introduction: Saudi Arabia is known to have a high prevalence of sickle cell disease (SCD). One of the areas with high prevalence is Khaibar, which is located north to Almadinah Almunawwarah with more than 180 registered patients with sickle cell disease (SCD). We assumed that our social interventional educational program (SIEP) will help decreasing the prevalence of SCA in Khaibar and will increase the society awareness about SCD and improve the outcome of the Premarital Screening Program (PMSP). **Methods:** A prospective observational study, conducted through Madinah's Hereditary Blood Disorder Charity Society (MHBDCS) in the period from January 2017 to December 2019. Different SIEPs were applied during several visits to Khaibar Governorate. **Results:** A total of 6 visits to Khaibar were carried out in the period of the study including school visits, meetings with general population, meetings with government representatives, medical seminars for local medical teams, meetings with Jomaa prayer leaders, conducting specialized hematology clinics and distribution of written pamphlets. A total of 5620 of Khaibar residents benefited from the programs, the total number of new marriages during the period of the study was 1380, and there were 2400 newborns, of which 22 were diagnosed with SCD but none was born from a new couple marriages in the period of the study. **Conclusion:** SIEPs have great benefits decreasing the incidence of SCD in Khaibar, PMSP can yield better results if it is associated with ongoing SIEPs and not ignoring the possible role of local influencers including religious leaders. **Keywords:** Consanguinity, Education, Khaibar, Premarital Screening, Saudi Arabia, Sickle Cell Disease"

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M-O-134

Heavy Menstrual Bleeding Among Glanzmann Thrombasthenia Ladies, Single-Center Experience

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Introduction : Glanzmann Thrombasthenia (GT) is a rare inherited bleeding disorder due to impaired platelet function. GT in Madinah is the most common bleeding disorder, with a rate of one in 10,000 inhabitants. Menorrhagia is a common manifestation among GT patients. Many experts believe in ceasing the menstrual cycle in these patients. Our patients' culture did not accept this principle. That makes it necessary to develop a protocol that will suit the patients' culture without endangering their lives.

Methods : Data extrapolated from the ongoing AlMadinah GT registry. 136 GT patients were identified. 445 women were found to be menstruating. In our center, we are following the AlMadinah protocol for menstrual cycle control among GT patients. The protocol calls for hormonal therapy to continue daily. The hormone will be stopped at the proposed time, and Antifibrinolytic agents will be administered. One dose of rFVIIa is administered at the first bleed. Hormonal therapy will restart between days three and five. 2017 modification to the protocol includes an observational period for the first three cycles; Either the patient transitions to the 2014 protocol or continues observation, switching between protocols is allowed.

Results : The menstrual cycle has been reported in 45 ladies, 10.5 – 52 years old, with a median of 20 years. 4 (9%) patients could not be interviewed. The age of menarche was 10.5-13 years, with a median of 12 years. The median Length of the menstrual cycle was 7 (5-20) days. Heavy menstrual bleeding (HMB) has been recognized in 35(86%) patients. 22 (53%) girls have enrolled in intermittent hormonal therapy protocol (2014protocol), of which 6 (27%) girls discontinued the protocol due to heavy cycles and continued on continuous hormonal therapy, and 9(41%) girls continued on protocol with acceptable results. 7 (32%) switched to an observational protocol with satisfactory results. 2 (29%) returned to the intermittent hormonal therapy protocol. 16 (39%) patients enrolled in the observational protocol (2017 protocol); of these, 7 (44%) were transferred from the intermittent hormone therapy protocol, and two patients switched to the intermittent hormone therapy protocol. 10 (24%) ladies did not follow any protocol because they were older patients; they had episodes of HMB managed by tranexamic acid and progesterone.

Conclusion : HMB is common among GT ladies and must be managed wisely. The AL Madinah protocol for menstrual cycle control in GT patients is a promising protocol for GT girls from cultures where the menstrual cycle cannot be ceased.

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M-O-136

Red blood cell automated exchange (RBCX) transfusion in sickle cell children: A single center experience from Madinah, KSA.

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Introduction: Sickle cell diseases are a group of the most common inherited hematological disorders, which is considered a major health problem in Saudi Arabia.

Objective: A prospective single-center study describes the challenges and outcomes of red blood cell automated exchange (RBCX) implementation in Hereditary Blood Disorders Center, Madinah, Saudi Arabia.

Methods: RBCX was started in December 2019, and since that date, 425 sessions of RBCX have been done in the center. Patients were sickle cell children indicated for chronic (elective) RBCX or acute (emergency) RBCX, through peripheral or central lines.

Results: From 15th Dec 2019 till 25 th September 2023, 28 sickle cell children were enrolled in the RBCX program; twenty-one patients (75%) were Saudi and seven patients were non Saudi (3 Nigerian, 2 Yemeni, 1 Chad and 1 Genian). Total 425 sessions had been performed with fluctuating frequency per months. Most of sessions were elective 410/425 (96.47%) and had been performed in hematology day care center during working hours, while 15/425 (3.53%) were emergency in PICU or pediatric wards. According to the cause of RBCX; 18/28 (64.3%) patients as a secondary prevention of stroke, 4/28 (14.3%) patients with recurrent severe painful crises, 3/28 patients (10.7%) as a primary stroke prevention, 2/28 patients (7.14%) recurrent acute chest syndrome and 1 patient (3.5%) for priapism. Most of our sessions were performed through peripheral lines (venous

or arterial), and emergent cases were admitted in PICU performed through central venous line. Target hemoglobin S % was set between 15-20% in our elective cases, this target was achieved in 370/ 410 sessions (90.2%). In emergency RBCX target was achieved in 12/15 sessions (80%). 421/425 (99%) sessions passed smoothly without adverse events, hypotension took place in 3 patients (0.7%) and one patient had extravasation after peripheral arterial line (0.23%).

Patients on elective RBCX for primary prevention of stroke had no stroke recorded with a success rate 100%; In patients with secondary prevention of stroke one patient experienced stroke 1/18 (5.5%) with success rate in secondary prevention of stroke was 94.5%. Patients became more active, improved in school achievement, decreased numbers of hospital admissions and ER visits.

Conclusion: Our experience has shown the effectiveness of RBCX with minimal side effects or complications. The rate of thrombophlebitis and infection rate were minimal, too

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M-O-137

Case Series with Haemoglobin Hope in Egyptian Family

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Introduction: Hb Hope, is a rare variant of β -globin chain. It results from mutation which is located on codon 136 on exon 3 of beta globin gene (beta 136(H14)Gly>Asp. It has low oxygen affinity and a reduced stability as compared with that of Hb A. Subjects and methods: We report this rare Hb variant (Hb Hope) in an Egyptian family which co-inherited with other B- thalassemia gene mutations. High performance liquid chromatography (HPLC), Capillary electrophoresis, cellulose acetate electrophoresis and PCR multiplex were used to detect the abnormalities. Results: the mother and 3 of her children had Hb hope which co-inherited with different beta thalassemia gene mutations. Hb Hope interferes with HBA1c assay by HPLC, it included with Hb A in alkaline gel electrophoresis. It appears in zone 10 in capillary electrophoresis. Conclusion: Hb hope is a rare hemoglobin variant. Its detections need more than one technique of Hb electrophoresis. Molecular analysis of the globin genes is essential in those with Hb hope variant due to the high incidence of co-inheritance with different globin gene mutations

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M-O-138

Coexistence of JAK2V617F and CALR mutation in Tunisian cohort with Philadelphia-negative chronic myeloproliferative neoplasms

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Introduction: Multiplex Ligation-Dependent Probe Amplification (MLPA) is a useful method in screening large deletions

but it is expensive for developing countries. Others strategies are recommended such as multiplex-PCR.

Aims : the aim of our study was to search large deletions in Tunisian severe hemophilia A using multiplex-PCR as a reference method.

Methods : After written informed consents, peripheral blood samples were obtained from 15 unrelated Tunisian severe hemophilia A followed in HTC Aziza Othmana Hospital without intron 22 and 1 inversions. Genomic DNA extraction was conducted with salting out method. Searching large deletions was performed by multiplex-PCR method.

Result : We describe here our experience using multiplex-PCR for identification of large deletions in 15 Tunisian severe HA without intron 22 and 1 inversions. All 26 exons were analysed for gross exon deletion mutations in eight multiplex-PCR assays. 3 cases showed gross exon deletions (table 1).

	HA 21	HA 89	HA 95
Severity	severe	severe	severe
Presence of inhibitor	+	+	+
Mutation	Large deletion	Large deletion	Large deletion
Localisation	Exon 1-13	Exon 1-22	Exon 19-22

Discussion : Our preliminary result reveals the presence of large exon deletions in 3 severe hemophilia A. All of them have inhibitors. Further samples should be studied

Although intron 22 and 1 inversions are controversial risk factors, large deletions are considered high-risk factors. Our preliminary result confirms that patients with inhibitors have large deletions. Searching large deletions in newly diagnosed patients with severe HA maybe considered to screen for high inhibitor risk.

MLPA protocol is expensive for developing countries. Routinely used single-PCR with 35 reactions for each patient is an alternative but it is a time consuming method. Multiplex-PCR with 8 reactions is a rapid and cheaper method."

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M-O-143

The Multidimensional Impact of Sickle Cell Disease in Children on Parents and Family Functioning: A Cross Sectional Study

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Background and aim: Sickle cell disease (SCD) is a genetic blood disorder that affects millions of people worldwide. In Saudi Arabia, SCD is a significant public health concern, with a high prevalence rate of 2.6% and Alahsa being one of the most endemic areas. The impact of SCD on parents and family functioning has not been studied in Saudi Arabia. This study aims to examine the multidimensional impact of SCD on parents and family functioning in Alahsa, Saudi Arabia.

Methods: A cross-sectional survey involved a total of 164 parents of children diagnosed with SCD. The impact of a child's condition on the functioning of the family was measured by PedsQL 2.0 Family Impact Module, and the authors' own questionnaire was used to collect sociodemographic and medical data. Higher scores on a scale ranging from 1 to 100 indicate better family functioning and parents' quality of life (QoL).

Results: Our study showed that the parents included in the study reported the highest scores for the cognitive functioning domain (total score: 72.2, SD = 20.6) and the family relationships domain (total score: 72 SD = 19.9) and reported the lowest scores for the worry domain (total score: 60.9, SD = 26.4) and the emotional function domain (total score: 68.7, SD = 23.2). Frequent hospital admissions, prior stays in the Intensive Care Unit (ICU), and low hemoglobin levels were associated with increased impact across all domains. On the other hand, consistent follow-up care and the use of Hydroxyurea were associated with improved quality of life and family functioning.

Conclusion: Child SCD demonstrates a considerable impact on the quality of life of parents and family functions. A multidimensional care is essential for both children with SCD and their parents, addressing their various needs and challenges. Therefore, studies on the determinants of high QoL seem to be crucial for the planning of interventions aimed at maximizing families' QoL and functioning."

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M-O-147

Effect of Low Dose of Hydroxyurea in SCD Children with Arab-Indian phenotype- Local Experience

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QCH

Abstract Background: Hydroxyurea has consistently been shown in multiple prospective research studies to improve the clinical complications associated with sickle cell disease (SCD) such as vaso-occlusive crisis and acute chest syndrome. To ensure safe and effective use of hydroxyurea in patients with SCD, close monitoring of patient clinical status and laboratory parameters is necessary. The aims of this study were to document the Effect of Low Dose of Hydroxyurea in SCD Children with Arab-Indian phenotype at Qatif central hospital- Local experience.

Methods: Retrospective descriptive cohort study, as a comparison of clinical improvement before and after starting hydroxyurea within a period of 24 months, starting from June 2019 to June 2021 at Qatif Central Hospital (QCH), Eastern province, Saudi Arabia. The sample included 341 children less than 14 years were included (boys and girls).

Results: Out of 341 patients who is on hydroxyurea, 217 were eligible to our study, 136 (62.6%) were male and 81 (37.3%) were female, 56.2% has co-existing alpha trait, 39.1% has co-existing G6PD deficiency and all are still alive in low dose hydroxyurea 10-15 mg/kg/day, mean 12.5 mg/kg/day.

Over a period of 24 months, hydroxyurea has significantly reduced the number of ER visit per year by 57%, also decreased the number of PVOC by 60% and inpatient hospital stay length per year by 46%. Moreover, 24 months post-HU,

we found that the incidence of ACS reduced by 74% and blood transfusion by 87%.

Hydroxyurea has also a great effect on the laboratory parameters, it increased an average of haemoglobin level by 1 g/dl, MCV by 8 fL HgbF by 4%, 18 months after starting the medication. Hydroxyurea also found to decrease WBC and absolute neutrophil counts (ANC) with drop in ANC by $1.6 \times 10^3/uL$ and decreasing LDH by 82 U/L and retics 1.15% over the same period.

Conclusion: The present investigation substantiated the efficacy of low-dose hydroxyurea in the management of sickle cell disease (SCD) among children with Arab-Indian phenotypes."

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M-O-148

Cancer Treatment Outcomes in Pediatric Patients with Glucose-6-phosphate dehydrogenase (G6PD) Deficiency

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Introduction: As the number of paediatric oncology patients has been dramatically rising in Saudi Arabia, with numerous cases being diagnosed as G6PD deficient without previous notice, the treatment for these cases is a concern. The safety and efficacy of cancer treatment in these circumstances present significant risks, as there is a lack of established protocols or guidelines about chemotherapy and supportive therapy. Due to the potential risk of hemolysis, there may be an increased need for additional blood transfusions.

Our main aim is to determine the cancer treatment (chemotherapy, supportive, and pre-medication) outcome for G6PD-deficient patients in King Abdulaziz Medical City- Jeddah, and review the prevalence of oncology in children with G6PD deficiency.

Method : Retrospective cohort study, comparing oncology pediatric patients with G6PD deficiency to patients without the deficiency, at King Abdulaziz Medical City (KAMC)-Jeddah from January 2018 to December 2022 (KAIMRC approval obtained).Our sample size was 116 patients, 58 with G6PDD and 58 with normal enzyme activity. We reviewed the laboratory data change from baseline and the number of blood transfusions needed as part of treatment during the treatment period.

Result: In G6PDD, it was discovered that the mean 3.43 ± 3.3 , and the correlation coefficient between the number of blood transfusions required and the necessity for them was $+0.416$. In patient with normal activity of G6PD enzyme the correlation coefficient was -0.416 , mean 1.14 ± 1.27 .

Conclusion : There was a correlation between G6PDD and blood transfusion in paediatric patients with oncology haematology disease, despite that further investigation is required."

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M-O-150

myWBDR Application Users' Satisfaction

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Background: The World Bleeding Disorders Registry (WBDR) is the only global registry collecting standardized clinical data on people with hemophilia (PWH) and people with von Willebrand disease (VWD). It comes with an app, "myWBDR", that allows patients to register their bleeding events, level of pain and treatment, that can be tracked by the Hemophilia Treatment Center (HTC).

Aim: To evaluate the satisfaction of "myWBDR" users.

Methods: We conducted a prospective study in which all users included in the registry in our center.

Extracted data are: age of the user (PWH or their parents).

They were contacted and surveyed in order to evaluate the app.

The survey contains 10 questions and results were analyzed using SPSS 26.

Results: In our center, 54 PWH are currently included in the WBDR (from the 27th of July 2022 to the 20th of September 2023) with 30 users of "myWBDR" application.

20 PWH were using the app themselves, and for the other third, one of the parents was in charge of the app. The median age of the app users was 30.5 [13-56]. Most of our users (26, 87%) prefer the app over the written patient report. Only 3 users (10%) find the application complicated, one of them is a father (49 years old) and the other two are PWH aged 35 and 45 years old.

The most important benefits of the app according to 18 users are the possibility to track their bleeding history, and its availability at any time in respectively 11 and 7 users.

Most of the users (26, 87%) find the app helpful or very helpful. The 2 users who find the app useless are the same who prefer the written patient report.

Although, more of the half of our users (16, 53%), complain with some difficulties to log in, some bugs, and the non availability of the app when they are not connected to Internet.

Conclusions: In summary, the "myWBDR" application has shown promise as a valuable tool for PWH and their parents in our HTC, offering benefits and constant accessibility.

It is noteworthy that the application appeals especially to young users

It is valuable to start tracking changes in health status of PWH through myPROBE and EQ-5D-5L, furnished by the app, and to organize workshops focused on the supervised learning of the use of those surveys in order to furnish a personalized experience to PWH and to enhance their engagement and healthcare management.

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M-O-151

Allogeneic Hematopoietic Stem Cell Transplantation In Aplastic Anemia: Experience Of The Pediatric Hematology And Oncology Department In Rabat (Morocco)

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Introduction: Aplastic anemia (AA) is a quantitative bone marrow failure due to a total or a partial hematopoietic tissue loss. It might be constitutional or acquired. Most cases of acquired

AA are idiopathic. It is a serious, life-threatening disease. The only curative treatment for severe forms is allogeneic hematopoietic stem cell transplantation (HSCT).

Aim: describe the epidemiological, clinical, biological, therapeutic and evolutionary profile of patients treated with allogeneic HSCT for severe aplastic anemia at our department.

Methods: Retrospective descriptive study of 07 patients with severe aplastic anemia treated with allogeneic HSCT in our transplant unit over a 3-year period (July 2020 to September 2023).

Results: We enrolled 7 patients, 5 boys and 2 girls. The median age at diagnosis was 10 years (02 to 16 years). Parental consanguinity was found in 3 patients. Anemia and bleeding manifestations revealed the disease in all cases. An infectious syndrome was present in 4 of them. AA was idiopathic in 4 patients, post-hepatic in 2 and Fanconi's constitutional aplastic anemia in one case. The median time from diagnosis to transplantation was 12 months (03 to 137 months). Allogeneic HSCT was performed from HLA-identical related donors in all patients. These were siblings in 6 cases and a mother in one case. Donor-recipient ABO incompatibility was present in 2 patients; It was a major mismatch in one case. The source of stem cells was from bone marrow in 6 patients and peripheral blood in 1 case. The conditioning regimen included rabbit anti-thymocyte globulin with cyclophosphamide in 6 patients, and Fludarabine with cyclophosphamide in the patient with Fanconi anemia. Graft-versus-host disease (GVHD) prophylaxis was conducted with cyclosporine and methotrexate in 6 patients, and cyclosporine with Mycophenolate mofetil in the case of Fanconi anemia. The median time to neutrophil recovery was 24 days (13 to 32 days), and 22 days for platelets (18 to 28 days). After a median follow-up of 19 months (9 to 38 months), late graft rejection was observed in only one patient, who also developed chronic cutaneous GVH with late post-transplant EBV and CMV reactivation; however, he is still alive. The other 6 patients are in good health.

Conclusion: Data from our series confirm that allogeneic HSCT should always be the first choice of treatment for AA, when an HLA-identical related donor is available. The time between diagnosis and marrow transplantation significantly influences the degree of acute and late complications.

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M-O-152

Observational, PRO study to Evaluate Quality of Life for Hemophilia patients on Emicizumab, KSA Single center study

Ohoud Kashari

Background: Emicizumab (Hemlibra) is a novel molecule, and its widespread prophylactic use is expected to have dramatic impact on reducing the bleeding frequency in the community of hemophilia A patients, thereby significantly improving their clinical condition and, ultimately, their quality of life.

Objectives: To assess the Quality of Life for Hemophilia patients before and after six months of Emicizumab usage in a real-world setting.

Methods: A prospective PRO study was planned on pediatric patients older than four years with HA with or without inhibitors who received Emicizumab for at least six months at a

Center for Blood Disorders for Pediatric Patients, Alaziziyah Children Hospital, Jeddah, Saudi Arabia. Validated Arabic versions of HAL, Heam-A-Qol, and EQ-5D-5L questionnaires were prepared. All patients were asked to mention their responses before and After Hemlibra Usage.

Results: Among 23 children with HA who started Emicizumab, the prospective PROs study was done on 10 Hemophilia patients who met the eligible criteria. All patients were males. The percent of patients with no problems before and after Hemlibra usage was calculated. As for mobility, the percentage of patients with no mobility problems before Hemlibra use was 10%, which was increased to 80%. Of them, 70% had no problems with self-care before use, which was increased to 100% after use. Patients with no problems in usual leisure and activities were 30%, which increased to 70% after use. One-fifth of patients (20%) had no Pain and Discomfort before use; this percentage increased to 90% after use. The overall positive emotional feelings were 29% before use compared to 78% after use. As for the positive feelings about Hemophilia, 38% of patients had positive feelings before Hemlibra use; this percentage improved to 80%. Positive feelings about Self-perception also increased from 22% before use to 66% after use. Feelings about School/Work improved from 13% before use to 83% after use, and feelings about treatment improved from 27% to 72% after use. The positive feelings of patients about the future before Hemlibra use was 45%, which increased to 92% after use.

Conclusion: Patients' feelings, overall self-perception, daily activities, and feelings towards hemophilia treatment improved after six months of using Hemlibra. Patients started living everyday lives and became less worried about their case worsening and could control their bleeding after Hemlibra usage. However, there is room for enhancement in the rest of the work/school activities.

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M-O-154

Circumcision in haemophilia Moroccan Protocol

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Introduction: Circumcision is based on religious and socio-cultural considerations and a sense of belonging. In emerging countries, the biggest obstacle for hemophiliacs is the consumption of antihemophilic products (AHP), which are expensive. It is therefore important to think about a less costly and safer protocol

The aim of our work is to adapt a protocol for the circumcision of a child with hemophilia, to reduce the cost with few or no haemorrhagic incidents.

Patients and methods: This is a prospective study of 33 cases including all types of hemophilia (A and/or B), all ages and all degrees of hemophilia, with a negative inhibitor status.

Our protocol consists of an injection of Factor VIII or Factor IX: 20 IU/Kg, one hour before the circumcision. After leaving the operation room, the child is placed for observation with

the parents. Injections are only given if bleeding occurs. Follow-up is carried out at home. We have evaluated the total quantity and cost of the anti-haemophilic product used.

Results: We enrolled 33 hemophiliacs, 28 hemophilia A and 5 hemophilia B. 10 cases had major hemophilia, 21 had moderate hemophilia and 2 had minor hemophilia. Patients ranged in age from 1 month to 17 years. The study revealed minor bleeding events. No bleeding incidents were noted between day 1 and day 5 post-operative. Bleeding most often occurred around day 5 (crust formation) and day 10 (crust fall-off). Children under 6 months and over 6 years of age bleed exceptionally. The haemorrhagic incidents that have occurred are all post-traumatic. The most significant bleeding was from the frenulum of the penis, which was stopped by surgery. Our protocol reduced the cost of circumcision by approximately 86%.

Conclusion: Our protocol consists of an injection of 20 IU/Kg of factor before circumcision. During post-operation the factor is only injected in the event of bleeding. Our protocol is based on close monitoring and therapeutic education of the parents.

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POSTER DISCUSSION

M-P-105

Assessment of peripheral blood CD26+ leukemic stem cells in CML Patients : a preliminary study

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Background : Chronic myeloid leukemia (CML) is a hematopoietic stem cell (SC) neoplasm characterized by an acquired genetic alteration ; the t(9;22)(BCR-ABL1) fusion gene. Bone marrow Ph+ CML CD34+/CD38- leukemic stem cells (LSCs) were found to specifically co-express CD26 (dipeptidyl-peptidase-IV).

Aims : We performed a flow cytometric assay to assess CD26+ CML leukemic stem cells (LSCs) in peripheral blood (PB) for its utility for detection of minimal residual disease (MRD) in CML patients on TKI therapy.

Methods : Standard qRT-PCR technique were performed to evaluate MRD

Results : A total of 45 patients with CP-CML were enrolled in this study. PB CD26+ LSCs were detectable in 30/45 (66.7%) and 15/45 (33.3%) CP-CML patients in first-line and second TKI treatment respectively (median=388 cells/μl [365.99-894.80], 95%IC and median=780 cells/μl [401.28-2750.98], 95%IC ; p=0.11). According to the MRD, PBCD26+ LSCs was significantly higher in CML patient with failure than optimal molecular (median=2560 cells/μl vs 388 cells/μl ; p=0.05), expressed as a BCR-ABL1 ratio.

Conclusion: Our finding highlights that CD26+ LSCs identification can be a useful marker for monitoring patients with CP-CML when a BCR-ABL1 quantitative assay is not available.

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M-P-109

Acquired Hemophilia: A Report on 2 Cases

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Introduction: Hemophilia is a recessive genetic disorder linked to the X chromosome. It involves a partial or complete deficiency of factor VIII (hemophilia A) or factor IX (hemophilia B), leading to a coagulation disorder and recurrent bleeding episodes. Joint and muscle bleeding gradually lead to disabling arthropathies and muscle atrophy, resulting in social, economic, occupational, and even psychological consequences. However, prophylactic treatment has revolutionized the prognosis and significantly improved the quality of life for individuals with hemophilia. The aim of our work is to identify challenges that prevent all eligible children from following this therapeutic model and to propose potential solutions.

Methods and Materials: This study is an exhaustive descriptive analysis of all hemophiliac children under follow-up at the Farhat Hached Hematology Department in Sousse, Tunisia.

Results: A total of 130 hemophiliacs are being followed in our department, of which 27% (36 cases) are children. The average age is 7 years (ranging from 1 to 17 years). Among them, 75% (27 cases) have hemophilia A, while 25% (9 cases) have hemophilia B. Out of the cases, 55% (20) are classified as moderate, 27% (10) as severe, and 16% (6) as mild. About 11% (4) of cases are on intermittent prophylaxis, with children aged - 10 years self-administering the factor. Only 36% (13) of cases are on prophylactic treatment, due to several reasons:

- Continuous unavailability of the factor, preventing the implementation of prophylactic model.
- Insufficient social support that doesn't allow for this option.
- Difficulty in obtaining venous access, causing anxiety for both the child and parents.
- Lack of parental adherence to this therapeutic model due to fear of puncturing their children.

Conclusion: Undoubtedly, prophylactic treatment has significantly improved the quality of life for individuals with hemophilia, enabling normal schooling and appropriate physical activities. Parental trust and adherence, along with the child's comfort, are essential for the success of this therapeutic model. To achieve this goal, we propose:

- Increasing educational workshops for self-treatment.
- Providing ongoing education to parents about the benefits of this therapeutic model.
- Raising awareness among authorities regarding the necessity for a consistent and sufficient supply of the factor

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M-P-110

Bleeding disorders in heavy menstrual bleeding; Debunking myths and revealing facts

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Background: Individuals with bleeding disorders suffer from bleeding symptoms such as mucocutaneous and orifical bleeding after trauma or spontaneously. Gynecologic and obstetrical bleeding are additional symptoms in females with bleeding disorders especially heavy menstrual bleeding [HMB] in reproductive age group and estimated to be 15 to 30% [1]. Most females feel ashamed to discuss the problem of HMB with their families and do not seek medical advice. HMB **markedly affects the quality of life in those females as they are often unrecognized and remain undiagnosed.** [2]

Aim: To determine inherited bleeding disorders as a contributing factor in females with HMB in Egyptian tertiary hospital.

Patients and methods: This study included 100 females in reproductive age [14-45] documented to have HMB by using PBAC score [PBAC -100]. Thrombocytopenia, systemic illness, and local causes e.g: uterine fibroids were excluded. Bleeding assessment tool ISTH-BAT was used to quantify the bleeding score. We assessed haemoglobin level [CBC], activated partial thromboplastin time [aPTT], prothrombin time [PT], and levels of FV, FVII, FVIII, FIX, Ristocetin Cofactor Activity, VWF antigen assay, Platelet aggregation tests [ADP, ristocetin].

Results: The mean of PBAC score was 234 [100 – 950]. The bleeding score mean was 9 [4 – 22]. Mean of haemoglobin level was 9.8 ± 1.2 . Seventy-eight [78%] females out of 100 had bleeding disorders. The bleeding disorders were: 30 cases VWD, 24 cases clotting factor deficiencies [9 FVII deficiency, 8 FVIII deficiency, 5 FV deficiency, 2 FIX deficiency], 24 cases platelet disorders [20 Glanzmann thrombasthenia, 3 Platelet storage pool disease and, 1 Bernard-Soulier Syndrome]. Other mucocutaneous bleeding symptoms were epistaxis, bleeding after dental procedures, and ecchymosis.

Conclusion: Early recognition of bleeding disorders is essential for management and avoidance of complications. Awareness through campaigns to girls in secondary schools and their mothers about HMB by PBAC score to recognize the difference between normal and abnormal bleeding. It is crucial to alert the general practitioners, physicians, and gynecologists about rare bleeding disorders as a cause for HMB. Moreover, such cases should be referred to a hematologist for proper management

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M-P-111

Follow-Up Of Hemophiles And Management Of Complications

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Introduction: Hemophilia is a recessively inherited genetic disorder linked to the X chromosome. Genes may be absent or damaged, leading to absence or deficiency of the coagulation factor. The disease causes difficulties in daily life, especially

for children who do not yet fully understand or accept their difference.

Material and method: This was an exhaustive descriptive study of all children with haemophilia followed at the Farhat Hached haematology department in Sousse, Tunisia.

Results: A total of 130 haemophiliacs of all ages are monitored in the department, 27% (36) of them were children. The average age is 7 years (extremes 1 and 17 years). 75% of cases (27) are hereditary, while 25% (9) are sporadic. A number of problems arise when caring for children with hemophilia, starting with the announcement of the diagnosis to the parents, particularly those with no history of hemophilia. On the other hand, giving parents enough time and simplifying the process will help them gain confidence and self-assurance in their dealings with the disease. What's more, they will become a competent and effective contributor to their child's care.

In our series, 75% (27) were hemophiliacs A and 25% (9) hemophiliacs B. In the majority of cases, that is 60% (17), the diagnosis was made in infancy, and in 71% (20) during a life-threatening haemorrhagic syndrome or during the evolution of the haemophilia. Hence the need to increase public awareness of the need for systematic screening of boys with a family history of haemophilia, with a view to early diagnosis and prevention of haemorrhagic accidents. Currently, hemophilia continues to be diagnosed in 11% (4) of cases of post-circumcision hemorrhagic syndrome, including one case who developed an anti-FVIII inhibitor following massive exposure to FVIII to control the bleeding. All children were on plasma FVIII replacement therapy, in 67% (24) of cases, and recombinant FVIII in 33% (12). Prophylactic treatment was provided in only 36% (13) of cases, mainly due to the continuous unavailability of factor for children with poor social status, the difficulty of venous access for some, and parental refusal for others.

Conclusion: To minimize the difficulties involved in caring for children with hemophilia, it's important to take a whole-family approach. Make sure the factor is available. Envisage a therapeutic project. Establish a therapeutic educational program. Organize awareness-raising campaigns in schools and recommend suitable sports activities to strengthen muscles and flexibility and avoid accidents."

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M-O-116

Bleeding phenotype in hemophilia carriers : Tunisian center experience

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Introduction: Development of inhibitor in people with hemophilia B (PWHB) is less common than in hemophilia A

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M-P-201

Activated Partial Thromboplastin Time Clot Waveform Analysis In Hemophilia A

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Introduction: Activated partial thromboplastin time (aPTT) clot waveform analysis (CWA) is a simple global coagulation assessment representing fibrin formation kinetics. While Hemophilia A (HA) diagnosis primarily relies on measurement of factor VIII (FVIII) activity, the usefulness of the aPTT based CWA has been highlighted in recent years.

The aim of this study was to evaluate the utility of aPTT based CWA in HA patients

Methods: A retrospective study carried out from January to December 2022 including samples belonging to 81 HA patients and 32 normal control samples.

The aPTT was measured for each sample and the CWA data were retrieved from an ACLTOP® analyzer (Instrumentation Laboratory). A built-in algorithm tool generates the fibrin formation curve and its two derivatives: the first derivative (D1) corresponding to the coagulation velocity and the second derivative (D2) corresponding to the coagulation acceleration. Three CWA parameters were studied: Max1 (maximum velocity), Max2 (maximum acceleration) and Min2 (maximum deceleration) in addition to peak time of acceleration and peak time of velocity. FVIII activity was measured using a one-stage assay. Statistical analysis was performed with SPSS 25.

Results: Patients with HA showed noticeably impaired thrombin formation in CWA compared to normal control patients, with significantly longer peak times of velocity and acceleration and significantly lower heights of velocity (Max1) and acceleration (Max2, Min2) .p

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M-P-119

Inhibitor in Von Willebrand disease: Two case reports

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Introduction: Hemophilia carriers were commonly seen as healthy women who faced the sole risk of having sons with hemophilia. Nevertheless, recent research has revealed that carriers, despite having normal factor VIII levels, exhibit a high bleeding risk.

The aim of this study was to evaluate the risk of bleeding among hemophilia carriers.

Methods: A cross-sectional study was carried out in the biological hematology department over a period of 10 months from June 2021 to March 2022 including hemophilia A carriers and controls (healthy women). Bleeding assessment was performed by ISTH BAT and Pictorial Bleeding Assessment Chart (PBAC).

We considered two groups: obligate carriers and potential carriers regardless of the type of hemophilia A or B

Results: Thirty-two hemophilia carriers (26 hemophilia A carriers and 6 hemophilia B carriers) and 30 controls were included.

The ISTH BAT was positive (- 6) in five carriers, representing 16%, and negative in all control women. The Bleeding score was at least 1 in 25 out of 32 carriers, accounting for 79%, and in three controls. Carriers had an ISTH BAT score significantly higher compared to control women (2.78 ± 0.1 ; p

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M-P-202

Inhibitor in people living with hemophilia B severe : A four cases report

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Background: The activated partial thromboplastin time (APTT)-based clot waveform analysis (CWA) is a simple global hemostasis test available on optical system analyzers. The aim of this study was to evaluate the usefulness of aPTT-CWA in hemophilia A carriers.

Methods: A cross-sectional study was carried out in the biological hematology department over a period of 10 months from June 2021 to March 2022 including hemophilia A carriers and controls (healthy women). Laboratory assessment included aPTT-CWA and FVIII activity (FVIII:C).

The software program for the analysis for the APTT-CWA allows the associated first and second derivative curves (DCs) to be displayed. The first derivative curve (D1) corresponding to the velocity of clot formation and the second derivative curve (D2) corresponding to the acceleration of clot formation.

Three CWA parameters were noted:

- Max1: maximum velocity (reflecting the "thrombin burst" and bleeding risk)
- Max2: maximum acceleration (detecting any coagulation factor deficiency)
- Min2: maximum deceleration

Results: We recruited 26 hemophilia A carriers and 30 women as control group.

The medians of T max1 and T max2 were significantly higher in female carriers of hemophilia A (40.8s - 33.7s) compared to controls (37.50s - 33s; p "

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M-P-120

Insights into Philadelphia Chromosome-Positive ALL: A Retrospective Study

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Background: Acute lymphoblastic leukemia (ALL) is a hematological malignancy which is more common in children. Philadelphia chromosome-positive (Ph+) ALL is a subgroup of B-cell precursor ALL. It is characterized by Breakpoint Cluster Region-Abelson (BCR-ABL1) fusion. The development of

tyrosine kinase inhibitors (TKIs) has improved the prognosis of these patients.

The aim of this study was to describe the clinical, therapeutic and evolutionary aspects of patients with Ph+ ALL.

Methods: This is a descriptive retrospective and monocentric study including patients with Ph+ ALL treated at the Hematology Department of Monastir, Tunisia, between January 2019 and December 2022.

Results: We included 5 patients with a median age 14 years [8-51 years]. The sex-ratio was 0,6. Bone pain was the most common revealing sign (40%).

On the initial physical examination, 60% of patients had adenopathy and/or splenomegaly, while 40% of them had no tumoral mass syndrome. One patient presented with anemia-related symptoms. Two patients had concurrent infections.

In the initial complete blood count, the median white blood cells count was 62 G/L [3,8-107,4G/L]. All patients had anemia while 80% presented with thrombocytopenia. Peripheral blood smear revealed blasts in 80% of cases, with a percentage ranging between 21%-82%. The diagnosis was confirmed by bone marrow aspiration. The karyotype was normal in 60% of cases. Translocation t(9,22) was detected in only one case. BCR-ABL transcript, subtype mbcr, was detected in all cases.

Three patients were treated according to the EORTC protocol, and two according to the GRALL protocol. Corticosteroids sensitivity was observed in 60% of cases based on the day 8 smear results. Treatment with TKI was initiated with a median delay of 20 days. Only one patient was treated with second-generation TKI (Dasatinib).

On the post-induction evaluation, none of the patients achieved cytological remission while one patient achieved cytogenetic remission, and one attained molecular remission.

All patients achieved complete remission at the end of the treatment. Only two patients underwent hematopoietic stem cell transplantation (HSCT). Two patients experienced relapse (one patient after the HSCT and the other 8 months after completing treatment), and one developed chronic pulmonary graft-versus-host disease. Actually, two patients are alive and are in complete remission.

Conclusion: Although the introduction of TKIs in the treatment, Ph + ALL is still having poor prognosis. Ongoing research and innovative therapeutic approaches are needed to improve the prognosis of such disease."

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M-P-121

Adverse effects of Imatinib in patients with chronic myeloid leukemia

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Background: Tyrosine kinase inhibitors were invented in the early 2000s and are now considered to be one of the most effective anti-cancer agents. Their discovery revolutionized the

treatment and prognosis of chronic myeloid leukemia. In this study, we aim to evaluate the adverse effects of Imatinib in our population.

Methods: a retrospective study, descriptive and analytical, about 171 patients with chronic myeloid leukemia, followed up in an internal and onco-hematology department between January 2012 and December 2022.

Results: Our study included 171 patients, with a mean age of 47.57 years [17-90], consisting of 75 men and 96 women, with an F/M sex ratio of 1.28. The myelogram classified the disease into a chronic phase in 135 patients (78.94%) and an accelerated phase in 36 patients. Ninety-seven patients (56.72%) were treated with Hydroxyurea before initiation of tyrosine kinase inhibitors. All our patients were put on Imatinib as first-line therapy.

Adverse effects of Imatinib were observed in 43 patients (25.14%). They consisted of hematological side effects in 39 patients: Thrombocytopenia in 20 patients, Neutropenia in 11, Anemia in 5, and 1 case of eosinophilia. Pancytopenia was observed in two patients. The management of hematological complications consisted of the suspension of treatment in 5 patients with platelets count $<50\,000/\text{mm}^3$, and in two patients with Neutrophils $<500/\text{mm}^3$, and the reduction of dosage in 10 patients, this resulted in the resolution of the disorder in all patients, with the restoration of treatment at a dose of 400 mg/day in 35 patients.

Dermatologically, 3 patients developed mild and transient itching on initiation of treatment, and two developed toxidermia, requiring the switch to another class of tyrosine kinase inhibitors.

A disturbance in liver function was observed in 6 patients, consisting of a slight increase in the transaminases within the two first weeks of the start of the treatment. This effect resolved after a brief cessation of treatment, with no further occurrence. Only one patient developed gynecomastia.

There was no significant association between the occurrence of an adverse event and response to Imatinib in our study ($p=0.260$).

Conclusion: Our study shows that it is not rare for adverse events to occur upon initiation of Imatinib. However, these events are often benign and can be resolved by adapting or temporarily interrupting treatment, or simply by monitoring the patient closely and observing the improvement in their condition.

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M-P-122

Polycythemia Vera and associated risk factors

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Introduction: Polycythemia Vera (PV) is a myeloproliferative disorder associated with a Janus Kinase2 (JAK2) mutation that causes the neoplastic proliferation of the hematopoietic progenitor cells. Arterial and venous thrombotic events are the most common complications in PV and the leading causes of morbidity and mortality. Therefore, risk factors of

thrombosis must be evaluated in order to facilitate the management of such complications.

Aims: The aim of this study is to evaluate the thrombosis risk factors in patients with PV.

Methods: It was a retrospective study including patients diagnosed PV and treated in the Hematology Department of Monastir (Tunisia), during the period between 2008 and 2020. Data were collected from medical files.

Results: We included 44 patients. The median age was 58 years [22-83]. Fifty-seven per cent of our patients were aged over 60 years with at least one cardiovascular risk factor in 54% of cases. We noted a slight female predominance with sex ratio at 0.91 (Table1). JAK2 V617F mutation was noted in 93% of cases and 7% had a JAK2 exon 12 mutation. In our study, 18 patients (40.9 %) had arterial or venous thrombosis at the time of diagnosis or complicated the course of the disease, all of them had positive JAK2 V617F mutation. For only 25% of our patients, a thrombotic event revealed the disease.

For the thrombotic risk, 12 patients (27.3%) were classified as low risk versus (vs) 32 (72.7%) as high risk. Cyto-reductive (Hydroxyurea) treatment was prescribed for 36 patients, 30 of whom were at high risk and 6 at low risk. Phlebotomy was recommended for 18 patients (40%) with clinical signs of hyperviscosity. Aspirin was prescribed for 28 patients (64%) at a dose of 100-250 mg per day, 14 patients (32%) had a history of thrombosis and therefore received anticoagulant therapy. Two patients (3%) had gastroduodenal ulcers that contraindicated antiaggregant and anticoagulant therapy. The univariate analysis showed an association between cardiovascular risk factors, age over 60 years and leukocytosis with the risk of thrombosis in our study population.

Overall survival at 12 months for all patients of our study was 86% vs 70% for patients with thrombotic events and at 24 months was 72% vs 50% respectively.

Conclusions: Patients with PV have good life expectancy, their treatment essentially focuses on the management of the thrombotic risk. PV patients with high risk of thrombosis should be appropriately treated including antiaggregant or anticoagulant therapy and cyto-reductive therapy."

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M-P-124

Relapse after allogeneic haematopoietic stem cell transplantation in children with acute lymphoblastic leukemia

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Background: Relapse remains the major cause of failure in children with high-risk acute lymphoblastic leukaemia (ALL) undergoing allogeneic haematopoietic stem-cell transplantation (allo-SCT). The prognosis for these patients is dismal. The identification of predictor factors of relapse is useful for the management of pre and post allo-SCT ALL.

Aims: The aim of this study was to identify factors predicting relapse after allo-SCT in children with all.

Methods: A descriptive retrospective study on pediatric patients.

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M-P-125

CD5 expression in diffuse large B-cell lymphoma

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Background: CD5+ cases of DLBCL are rare and are described as a distinct histological entity with poorer outcomes. The theory of an independent prognostic value of CD5 in DLBCL patients is widely accepted. Some studies have proposed a first-line, more aggressive therapy for these patients. In this study, we aim to identify the clinical, epidemiological, and prognostic features of our CD5+ DLBCL patients.

Methods: This is a retrospective, descriptive, and analytical study, from January 2018 to December 2021, and includes newly diagnosed patients with DLBCL during this period, and expressing CD5 marker.

Results: We report 6 cases of CD5+ DLBCL among a total of 184 DLBCL patients (3.26%), consisting of 3 women and 3 men. The median age at diagnosis was 60.5 years [44-71 years], and the average diagnostic delay was 6.3 months. Three patients had B signs at diagnosis, and five had a tumoral syndrome (adenopathy and/or splenomegaly). Performans status was -1 in 5 patients. Histological examination showed a proliferation index greater than 80% in 4 patients, CD45 positivity in one patient, and BCL2 and BCL6 positivity in two and three patients respectively. Extension workup showed extensive involvement (stage III/IV) in 5 patients. A bulky mass was described in four of the six patients. The IPI score was greater than three in 4 patients, and only one patient developed venous thrombosis.

Complete blood count revealed anemia in 5 patients, lymphopenia in 2 patients, and thrombocytopenia in one patient. ESR was elevated in 4 patients, and we observed high levels of LDH in 5 patients.

Five patients were placed on the RCHOP protocol, three patients achieved complete remission after first-line treatment, while two were reported to be refractory. One patient died from severe disease before the start of treatment; he had been diagnosed with a voluminous gastric mass measuring 15 cm.

We found a significant correlation between CD5 expression and adrenal involvement ($p=0.033$) and anemia ($p=0.01$).

Conclusion: Our study, despite the small number of patients, confirms the rarity of this entity and its poor outcome. Larger studies are needed to properly characterize these patients so that appropriate action can be taken as soon as they are diagnosed. Some studies propose dose-adjusted R-EPOCH and autologous stem cell transplantation, but evidence of their effectiveness is still lacking in these situations.

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M-P-127

The Bulky mass in diffuse large B Cell Lymphoma: Prognostic impact? A treatment guiding factor?

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Background: In DLBCL, multiple studies demonstrated the negative effect of bulky mass on the prognosis, particularly in localized stages, as well as its impact on first-line response and relapse. In this study, we aim to evaluate the prognostic impact of the bulky mass in our patients with DLBCL.

Methods: retrospective, descriptive and analytical study of newly diagnosed DLBCL patients followed up in an internal medicine and onco-hematology department between January 2018 and December 2021. A Bulky was defined as a mass exceeding 7 cm in large diameter.

Results: We analyzed 184 DLBCL patients treated in our hospital between 2018 and 2021. A bulky tumor was detected in 66 patients (35.86%). The sex ratio F/M is 1. Age ranged from 16 to 87 years, mean age at diagnosis 56.01 years. Sixty per cent of patients had B-signs at diagnosis, and 74.24% presented with clinically evident tumor syndrome.

Blood count revealed anemia in 34.84% of patients, lymphopenia in 30.30% of patients, and thrombocytopenia in 3 cases. Inflammatory markers were elevated in 39.3% of patients, LDH was up to normal in 78.78% of cases. The CT-scan showed that 20 patients had limited stage disease (Ann Arbor I or II). Low IPI was found in 48.48% of patients and thrombosis was observed in 14 patients (21.21%).

Among our patients with bulky mass, 12 patients passed away before the start or completion of 4 cycles of treatment, and three were lost to follow-up.

A total of 48 patients received the R-CHOP regimen as first-line treatment, while 3 cases of mediastinal Bulky received DA-R-EPOCH protocol. After first-line, 33 patients achieved complete remission, while 18 required second-line therapy, of whom 13 patients were deemed refractory. We have lost 8 of them. In total, 76% of our patients with Bulky mass, who completed their therapy, responded to the treatments initiated. Bulky mass was associated with skin involvement ($p=0.037$), lymphopenia ($p=0.035$), high CRP ($p=0.018$), high LDH ($p=0.001$), CD45 positivity ($p=0.009$), a proliferation index above 80% ($p=0.002$), high IPI ($p=0.026$), thrombosis ($P=0.001$) and death ($p=0.0001$).

The 4-year survival of patients with a bulky mass was 40.5% versus 78.2% of patients without a bulky mass ($p=0.0001$).

Conclusion: Our study highlights the aggressive form of the DLBCL in patients with a bulky mass. We need to improve the therapeutic management in this group of patients. As reported in other studies, consolidative radiotherapy can be used for bulky and residual mass after immunochemotherapy."

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M-P-128

Factor VIII Level in Mothers of Patients with Severe Hemophilia A and its Impact on Joint Status

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Background: Hemophilia A; is an X-linked bleeding disorder. The characteristic bleeding phenotype in patients with severe hemophilia is recurrent spontaneous hemarthrosis with development of hemophilic arthropathy. Bleeding phenotype of heterozygous female carriers with factor VIII (FVIII) activity level lower than 40% is variable and may be comparable to males with mild hemophilia.

Aim of the Work: To assess factor VIII level in mothers of patients of hemophilia A correlating it with the occurrence of clinically and radiologically documented arthropathy.

Patients and Methods: A cross sectional study included 40 mothers for known patients with severe hemophilia A. Studied participants were subjected to detailed analysis of bleeding history, six major joints range of motion (ROM) examination, functional independence score in hemophilia (FISH), FVIII level one stage assay and magnetic resonant imaging (MRI) of the reported affected joints with calculation of radiologic joint scores.

Results: The mean age of studied mothers was 42.3 (range 20-59) years, mean weight 78.9 (range 59-95) kg and mean BMI 28.97 (range 22.04-35.65). Mean total FISH score was 23 (range 20-24) and mean total MRI score 4.36 (range 0-8); 40% of studied participants had one bleeding episodes, while 32.5% had 2 or more bleeding episodes; 10% of them had epistaxis, 10% bleeding from minor wounds, 17% bleeding post teeth extraction, 15% had postpartum hemorrhage and 12.5% had menorrhagia. ROM assessment showed knees to be the most affected joints with decreased mean ROM in right and left knees of (123.25 ±12.06) and 120.00 ±15.81) respectively. Mean FVIII activity level was 43.33 IU (± 22.79) with 51 % of studied participants had factor VIII level less than 40 while 48.7% had their level more than 40. By comparison of participants from both groups, there were no differences in age, weight, BMI, MRI scores between groups of factor VIII less and more than 40%. There is negative correlation between FVIII level and pain score ($r=-0.409$; $P=0.009$)

Conclusion: Potential female hemophilia A carriers are at risk of spontaneous bleeding and early joint affection, yet it is not related to specific cut-off level of factor VIII."

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M-P-129

High incidence of Intracranial haemorrhage in Egyptian children with hereditary afibrinogenaemia: A single center study

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Inherited FVII deficiency is the most common of the rare autosomal recessive bleeding disorders and the prevalence is higher in those countries where consanguineous marriages are

frequent, especially with respect to the severe forms of the disease. It is second in distribution amongst our Egyptian pediatric cohort with rare coagulation disorders following up in Hematology clinic, Cairo University Pediatric Hospital. Patients have a wide phenotypic variability ranging from asymptomatic to life-threatening bleeding, including intracranial hemorrhage (ICH) with significant mortality and morbidity. We studied the clinical spectrum of our cohort to see if their bleeding phenotype behave differently in a population with multiethnic historical background. We correlated FVII activity with the severity of the bleeding phenotype. F VII deficiency was diagnosed when factor assay was below normal ($n = 70-120\%$) and patients were classified into asymptomatic, minor and major bleeders. 50/170(29.4%) of our rare coagulation disorders pediatric cohort were diagnosed with inherited FVII Deficiency with equal sex prevalence and were usually of a consanguineous marriage. 48% are major (cerebral bleeding, hemarthrosis, gastro-intestinal) bleeders, 40% minor bleeders and 12 % asymptomatic diagnosed after a hemostatic screen prior to surgery. The bleeding phenotype showed wide variability and mostly correlated with FVII activity. Cerebral bleeding was reported in 24% of patients and they were all major bleeders (FVII activity

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M-P-130

Immunodeficiency in Children with Diamond Blackfan Anemia

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Background: Inherited bone marrow failure syndromes are a diverse group of disorders characterized by BM failure, with more than 1 extra-hematopoietic abnormalities. Diamond Blackfan anemia typically presents with erythroid aplasia in the first year of life. Recent associations of DBA with immunodeficiency started to be reported. We aimed to study the frequency of recurrent infections in children and adolescents with DAB with an immunodeficiency related score and whether it is related to humoral immunodeficiency.

Methods: A cross-sectional study including patients with Diamond-Blackfan anemia following at Ain Shams University, Cairo, Egypt. Revision of records for disease presentation, association with congenital anomalies, transfusion dependency and steroids response were analyzed. An immunodeficiency related score was applied to all patients since birth with reviewing of the clinical histories. Immunoglobulin assays were performed to studied patients by nephelometry

Results: Thirty patients with DBA were recruited, the mean age at diagnosis was 6.1 ± 3.9 months, male: female ratio was 2.1:1. Consanguinity was found among 59.1% parents, family history of DBA and malignancy in 4 18.2% patients each. Cardiac, renal and bone anomalies were found in 22.7%, 13.6% and 4.5% of patients respectively. 28.6% of patients showed response to steroid therapy while 68% patients were transfusion dependent. IDR score more than 3 was reported in 13 (43%) and immunoglobulin deficiency in 4 (14%) and were maintained on regular intravenous immunoglobulin replacement therapy.

Conclusion: The combination between Diamond Blackfan anemia and immunodeficiency is a recently under-reported association. Some of the immunodeficiency entails humoral immunodeficiency which necessitates infection preventive approaches.

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M-P-131

A descriptive study of patients with myelodysplastic syndrome: experience of the clinical hematology department of Monastir-Tunisia

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A descriptive study of patients with myelodysplastic syndrome: experience of the clinical hematology department of Monastir-Tunisia

Introduction: Myelodysplastic syndrome (MDS) is a group of clonal diseases characterized by cytopenias resulting from inefficient hematopoiesis. It predominates in the elderly and represents the most frequent pre-leukemic state in adults.

The aim of this study was to describe the epidemiological, clinical, prognostic and therapeutic characteristics of patients diagnosed MDS in our department.

Patients and methods: This is a retrospective study conducted in the hematology department of Monastir-Tunisia and including patients followed up for MDS between January 2014 and February 2023.

Results: A total of 53 patients was included in our study. The sex ratio was 1.4. The mean age at diagnosis was 72.1 ± 11.9 years. The circumstances of discovery were an anemic syndrome (72.6%), a fortuitous discovery (20.8%) or a hemorrhagic syndrome (6.6%). A bone marrow aspiration was performed for all patients. Two patients (3.8%) underwent bone marrow biopsy because of inconclusive myelograms. A karyotype was performed in 33 patients (62.3%), with the following results: normal in 26 cases, no cell growth in 3 cases and chromosomal abnormalities in 4 cases. According to the World Health Organization 2016 classification, patients were classified as MDS with unilineage dysplasia in 47.2%, MDS with multilineage dysplasia in 22.6%, MDS with ring sideroblasts (RS) with unilineage dysplasia in 7.5%, MDS with RS with multilineage dysplasia in 5.7%, MDS with excess blasts-1 in 9.4% and MDS with excess blasts-2 in 7.5%. According to their Revised International Prognostic Scoring System (R-IPSS), 3.8% of our patients were at very low risk, 58.8% at low risk, 24.5% at intermediate risk, 9.4% at high risk and 3.8% at very high risk.

Therapeutic management consisted of transfusions of red blood cells and platelets in 94.3% and 20.7% of our patients, respectively, prescription of androgens in 15.1%, Azacitidine in 3.8%, cyclophosphamide in 1.9% and filgrastim in 1.9% of patients.

The mean follow-up time was 23.6 months. Complications included febrile neutropenia in 5.7%, hemorrhagic syndrome in 17% and transformation to acute myeloid leukemia in

7.5%. At the latest updates of analysis, 28.3% of patients were alive.

Conclusion: In accordance with the literature reports, MDS is a disease of the elderly, predominantly men. Approved treatment options for MDS are limited, and management depends essentially on risk, as assessed by the R-IPSS score.

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M-P-132

Epidemiological Pattern Of Childhood Cancer Treated At Private Dr Suliman Fakeeh Hospital, Jeddah City, Saudi Arabia And Their Outcome

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Background and aims: Increasing role of private hospital at KSA supported by implementing medical insurance involvement in medical care and charity society to support treatment of children with cancer.

Frequent surveillance of cancer outcome is important to improve modes of treatment.

Cure rate of acute lymphoblastic leukaemia reach 85% and 70% for solid tumour

Methods: From October 2017 to December 2022 all paediatric cases diagnosed with cancer will be reviewed to assess even free survival, EFS and overall survival, OS

Results: Total cases 63 with male 42 and female 21. Acute lymphoblastic leukaemia ALL 21, Acute myeloid leukaemia AML 2, Hodgkin lymphoma HL 4, Non-Hodgkin lymphoma NHL 4, Wilms tumour WT 8, Retinoblastoma RET 7, Neuroblastoma NBL 4, Brain tumour 4, bone tumour 2, and 1 for each; synovial sarcoma, LCH, HLH, ovarian teratoma, CML, hepatoblastoma HBL and abdominal inflammatory Myo fibroblastic tumour.

Disease recurrence 13/63 with EFS 80%.

9 cases died, 1 from toxicities (infant HL) and 8 recurrent diseases (3 ALL, 2 NBL, 1 RET, 1 HBL and 1 brain stem glioma). 4 cases salvaged after disease recurrence; ALL with early combined relapse after allogeneic SCT, stage 4 HL received 2nd line chemotherapy then autologous SCT, RET received 2nd line chemotherapy and synovial sarcoma local relapse received 2nd line chemotherapy with safe margin 2nd surgery and local radiotherapy with OS 86%.

13/63 cases indicated radiotherapy

Conclusion: Private hospital could support health care system in diagnosis and management of childhood cancer. Using risk directed therapy for all types of tumours would have impact on outcome.

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M-P-135

Charity Medical Care For Hemoglobinopathies At Madinah, Saudi Arabia

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Background: Charity health care is defined as medical care provided free or discounted to people who cannot afford to pay for the cost of medical services. There are 1050 charity societies in Saudi Arabia, out of which 230 health charities. Madinah Hereditary Blood Disorders Charity Society [MHBDCS] is a charity health society established in 2010 with the aim to provide social and medical services for patients with hemoglobinopathies in Madinah region, Saudi Arabia. In 2020, MHBDCS launched the Initiative of Charity Medical Care for Hemoglobinopathies. The aim of this study is to demonstrate the experience of this health charity initiative in Madinah, Saudi Arabia.

Methods: This is a retrospective observational study with the aim to review the initiative of charity health care for hemoglobinopathies patients over 3 years from September 2020 to September 2023. The comprehensive medical care provided by the MHBDCS started on the first of September 2020. 3 hematology clinics per week were opened at a private medical complex; the clinics provide the services free of charge. 2 clinics were for thalassemia patients, and one clinic per week was for sickle cell disease (SCD). The charity medical services include Regular outpatient follow-up, free-of-charge laboratory tests and medication supply for all patients, and on-demand free medical staff consultation

Results: The total number of hemoglobinopathies patients registered in the MHBDCS was 1085. 339 [31%] were non-eligible for governmental health services. The total number registered for charity medical care was 157 patients; 84 were thalassemia, and 75 were SCD. Total visits to the clinics were 2234 visits to the Thalassemia clinics and 1542 visits to the SCD clinic. The cost of medications was SAR 1,274,361. The cost of laboratory tests was SAR 30,8617. The cost of staff was SAR 823,200. The total cost of all medical charity services was SAR 2,406,178. The outcomes of the initiative chronic medical care were:

- 1 Reduction of hospital admission and emergency visits.
- 2 Reduction of frequency of SCD painful crisis.
- 3 Provide social and financial support to patients and their families.
- 4 Improve the overall quality of life

Conclusions: Comprehensive medical charity care is vital for non-eligible and eligible patients with Hemoglobinopathies. It improves the quality of life and reduces social and financial burdens to the patients and their families. We recommend having this service in all health charity societies in Saudia Arabia. We recommend providing acute charity care too.

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M-P-139

Hydroxyurea utilization and dose optimization in children with sickle cell disease (SCD)

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Background : Sickle cell disease (SCD) is quite prevalent genetic disorder in Saudi Arabia and the disease is more endemic in eastern and southwestern regions of the country. Hydroxyurea (HU) is the only approved drug to ameliorate some of the clinical problems of SCD with myelosuppression

is the main HU dose limiting effect. Data showed that, patients who received HU titrated to the maximum tolerated dose (MTD) have greater clinical effects. Noteworthy, optimized HU utilization in Saudi Arabia remains underused. This study examines the utilization of HU to MTD in children with SCD.

Methods: This retrospective observational study was conducted in King Abdulaziz Medical City-Jeddah. All children aged less than 15 years with SCD and receiving hydroxyurea in inpatient and outpatient between January 2018 and December 2020 were included. Those with missing follow up data were excluded. Data was collected from the patients' electronic medical records that include demographics, HU starting dose, MTD achieved, neutrophil count, platelet counts, total hemoglobin and hemoglobin F pre and after starting HU. The primary outcome is to examine MTD achieved. Secondary outcome is to address reasons for failing to achieve MTD. The chi-square was applied through SPSS version 23 for assessing any association with outcome variables and demographic characteristics.

Results: The total patients included initially were 123 children, 47 children were excluded because of missing data and the remaining 76 patients were eligible for the study. Male to female ratio was 1:1 with a mean age of 7.6 years. The mean HU starting dose was 17.6 mg/kg (SD± 6.09). Majority of included patients 65.8% (50/76) never able continue receiving HU beyond 25 mg/kg with a mean MTD was 25.07 mg/kg (SD± 6.5). The laboratory investigation showed significant reduction in absolute neutrophil count, platelet counts and rise in total hemoglobin and hemoglobin F from baseline after starting hydroxyurea. Emergency department visit more than three times per year to control SCD related complications were documented in 40.8% (31/76). Of those, the median HU MTD was 27 mg/kg (IQR 20-30). Poor compliance (20%) to the prescribed dose and significant severe myelosuppression (31.6%) were the main reasons for not achieving MTD.

Conclusion: The majority of pediatric with SCD were not able to reach stable MTD of HU beyond 25 mg/kg. Our findings support the need to reconsider the HU MTD goal in our patient population."

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M-P-140

Prevalence of Transfusion Transmissible Diseases Between Madinah Blood Bank Donors, Saudi Arabia

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Background: The possible risk of TTDs caused by blood-borne pathogens is one of the most undesirable risks of blood transfusion worldwide.

Objective: This study aimed to screen TTDs carriage and prevalence in Madinah blood bank donors, as well as to describe

the dominant TTDs in the study area within Al-Madinah city. The study also aimed to provide an examination over time of the prevalence of TTDs for three years 2020, 2021 and 2022, at Al-Madinah blood banks and compare validated results obtained from previous studies with our study results.

Methods: The testing method included screening serological tests for TTDs and confirmation assays according to each pathogen. The samples were collected and tested according to guidelines approved by the Association for Advancement of Blood & Biotherapies (AABB), Central Board for Accreditation of Healthcare Institutions (CBAHI) and Saudi Food & Drug Authority (FDA). The study subjects were 104538 male and female volunteer and replacement donors aged 18 to 60 years who donated blood from 2020 to 2022 at five blood banks within Al-Madinah city. Donor samples were tested, abnormal TTDs results were further tested for confirmation, and all results were documented according to the following policy.

Results: A total of 104538 donors, 1061 donors giving abnormal serology results for at least one marker, had an overall carriage rate of 1.01%, and only 416 were confirmed by the approved confirmation method. Out of 1061 serology-positive donors, 334, 305, 214, 105 and 103 donors were positive for Hepatitis B Virus (HBV), Hepatitis C Virus (HCV), syphilis, Human Immunodeficiency Virus (HIV) and Human T-Lymphotropic Virus (HTLV) infection, respectively.

Conclusion: TTDs positive donors tested by serological methods among Madinah blood bank donor males and females were found to have the same local TTDs rate. The rate of positive TTDs tests was low out of total screened donors. Inspection of the TTDs pattern among TTDs positive donors by serological methods revealed that HBV infection was high compared with other tested pathogens. The high percentage of study subjects with HBV infection indicates a need more efforts to explain high HBV prevalence.

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M-P-141

Glanzmann thrombasthenia: Genotype / phenotype correlation

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Introduction: One of the rare bleeding disorders is Glanzmann thrombasthenia (GT), with an incidence of 1:1,000,000. However, the case differs in the Middle East, where GT has a higher rate; in Madinah, Saudi Arabia, GT prevalence was 1:10,000. Few studies have looked into the correlation of Genotype/phenotype. In this study, we are looking into the correlation between genotype and clinical presentation.

Methods: This is a report from the AlMadinah GT registry in the Madinah Hemophilia Comprehensive Care Center. The registry covers a period of 20 years and includes all patients with a confirmed diagnosis of GT. 136 GT patients were identified. 11 patients had no data available. Disease severity is defined based on the severity of the clinical course, where the severe clinical course is defined as bleeding continued for more than 3 days, a drop in hemoglobin for more than 2 grams / L from the baseline line, or needing admission to the intensive

care unit. The moderate clinical course is defined as bleeding that needs hospital admission, while the mild course is bleeding that can be stopped at a home setting management. 15 mutations were carried by 1-9 GT patients per each mutation. Here, we are comparing only three mutations carried by 83 GT patients. The research and ethical committee approved the study.

Results: One hundred twenty-five GT patients were studied: 67 (54%) females and 58 (46%) males. The median age was 16.5 (10 days to 56 years). 98% of GT patients were the product of consanguineous marriage. Family history of GT was reported in 107 (86%) cases. Twenty-five families have more than one sibling (2-5) with GT. Eighteen mutations have been identified: 15 mutations in the ITGA2B gene and 3 mutations in the ITGB3 gene. 42 GT patients carry the mutation ITGA2B:exon20:c.1879-2A>G (Mutation 1). 28 GT patients carry the mutation ITGB3:exon13:c.2112delC (Mutation 2). 13 GT patients carry the mutation ITGA2B:exon13:c.1210+5G>A (Mutation 3). 8 mutations carried by one GT patient only. The other 7 mutations are carried by 3-9 patients. The disease course in Mutation 2 was mild in 20 patients, moderate in 46 patients, and severe in 25 patients. While mutation 1 was mild in 48, moderate in 36, and severe in 9 patients. Mutation 3 was mild in 50, moderate in 25, and severe in 25 patients. The P-value was 0.00001 when comparing mutation 2 and 1 and 0.03 in comparing mutation 2 to 3 but was 0.6 when comparing mutation 1 to 3. The bleeding frequency in Mutation 2 was once/year in 16 patients, 2-4/year in 25 patients, and once/month in 46 patients. While mutation 1 was once/year in 53 patients, 2-4/year in 28 patients, and once/month in 7 patients. Mutation 3 was once/year in 29 patients, 2-4/year in 43 patients, and once/month in 21 patients. The P-value was <0.0000001 when comparing mutation 2 and 1 and 0.03 in comparing mutation 2 to 3 but was 0.003 when comparing mutation 1 to 3.

Conclusion: Genotype can affect the phenotype in GT patients, where the mutation ITGB3:exon13:c.2112delC (Mutation 2) has a more severe disease course and more frequent bleeding.

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M-P-142

Labor And Delivery Outcomes Among Ladies With Glanzmann Thrombasthenia

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Introduction: The high prevalence of Glanzmann's thrombasthenia (GT) in Madinah (was calculated as high as 1:10,000) makes the rate of pregnancies higher too. GT is an autosomal recessive bleeding disorder due to functional platelet defects impairing platelet aggregation.

Methods: This is a report from the AlMadinah GT registry in the Madinah Hemophilia Comprehensive Care Center. The registry covers a period of 20 years and includes all patients with a confirmed diagnosis of GT. 136 GT patients were identified. 11 patients had no data available. The research and ethical committee approved the study.

Results: Among the 28 pregnancies identified, the Maternal age at the time of pregnancy was 20 - 42 years (median 30 ± 5.7 years). Mothers' Gravidity was 1-7 (median 2.5 ± 1.6), and parity was 1-6 (median 2 ± 1.4). Among the 23 term pregnancies, Labor took place at the gestational age of 34-40 weeks (median 38 ± 2 weeks). Sixteen (70%) deliveries were vaginal, and 7(30%) were cesarean sections. Four GT ladies requested cesarean section; the 4 of them had concerns about GT consequences. Fetal distress has been reported in two instances. One case cesarean section has elected by medical attendance as this case was one of early GT cases. Prophylactic platelets transfusion had given pre-delivery on four occasions, rFVIIa on 4, and a combination of rFVIIa and platelets transfusion on six occasions. Tranexamic acid (TA) was used in all cases. Ten deliveries were conducted with no prophylaxis. Primary postpartum hemorrhage (PPH) occurred in 13 (72%) cases, while 5 (28%) GT ladies had secondary PPH. The Twenty-three deliveries produced 24 babies (13 girls and 11 boys). Four babies had bleeding (2 intracranial hemorrhages, one subdural hematoma, and one gum oozing), and 9 had alloimmunization with mild to severe thrombocytopenia. Of all babies tested for GT, 6 of them were GT. Five miscarriages (18%) identified. Four had managed with prophylactic therapy, two had platelets and TA and two rFVIIa and TA. All five cases need post delivery platelets transfusion, TA continued till no more bleeding.

Conclusions: Labor and Delivery among ladies with GT carries a higher risk of bleeding. Management of Labor and Delivery in women with GT has to be managed promptly. The high rate of PPH among GT ladies makes it essential to treat it promptly. Vaginal delivery considered safe if managed early and prophylaxis conducted.

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M-P-144

Hospitalizations for sickle cell disease in Qatif central hospital pediatric population

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Background: Sickle cell disease (SCD) is well known to be as one of the most common diseases that are inherited in a monogenic pattern worldwide. Cases of sickle cell disease is increasing global health dilemma. According to local cross-sectional study done in 2005, The prevalence of sickle cell disease of children and adolescents in Saudi Arabia is 24 per 10,000. Although the prevalence of SCD has been decreasing in all regions of Saudi Arabia, the prevalence remains high in comparison with other countries. In eastern province, SCD phenotype found to be benign and milder clinically. Numerous reasons have been proposed for SCD pediatric age hospitalization.

Objective: the aim of this study was studying the reason, rate and factors affecting Length of stay for SCD patient hospitalizations.

Materials and methods: This study was a single center retrospective electronic chart review study intended to study pediatric age population with SCD, aged 6 months to 14 years, who have been hospitalized in Qatif Central hospital, Saudi Arabia, between 1 January 2015 and 13 march 2019.

Results: Total number of studied patients was 464. Total number of admissions to pediatric medical ward were 6026 admissions, out of these 1449 admissions were SCD related complications (24%). Reason of admission to SCD patients in our center found to be, starting from the most common to the least common: PVOC (69.27%), Viral infection (21.22%), Acute chest syndrome (5.04%), Splenic sequestration (3.19%), Others (2.56%), Hemolytic crisis (1.99%), Aplastic crisis (1.21%), hepatic 1.14%), Sepsis (0.78%), Abdominal crisis(0.71%), Osteomyelitis (0.71%), Headache (0.57%), Septic arthritis (0.43%), CNS crisis(0.35%), Elective admission for MRI (0.28%). Results of our study showed significant impact on length of stay between different reasons of admissions ($P=0.000$). Additionally, number of comorbidities patient admitted with had a significant impact on length of stay ($P=0.000$).

Conclusion: as based on the results of our study, it is clearly demonstrated the significant percentage of admissions from the total population, that it reflects the burden in term of hospital bed capacity utilization and fundal recourses usage. Accordingly, this highlights the significance of affordability of cost effective SCD modifying agents."

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M-P-145

Neurological Complications in Children with Sickle Cell Disease at Qatif Central Hospital in Saudi Arabia

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QCH

Abstract Background: Neurological complications significantly burden children with sickle cell disease (SCD). SCD is the most common hemoglobinopathy; however, its effect on neurological development in children remains unclear. This study aimed to investigate the range of neurological complications in children with SCD and predict the modifiable risk factors in children with SCD in the Qatif region of Saudi Arabia.

Methods: A retrospective cohort study including 944 children aged.

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M-P-146

The Role of Hydroxyurea on Growth in Children with SCD

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Background: Sickle cell diseases (SCD) has been proven to alter the development of children on many axes. Growth velocity evaluation, encompassing weight and height dynamics, is pivotal in deciphering the overarching growth and developmental trends within paediatric cohorts. Although growth pattern in SCD has been studied, no data are available in arabian phenotype at Saudi Arabia. Objective: This longitudinal and retrospective study aimed to investigate the role of hydroxyurea on growth in children with sickle cell disease (SCD) at Qatif Health Network, Qatif, Saudi Arabia. The objectives were to study the growth velocity in SCD children on hydroxyurea, and compare local data with published literature. Methods: This study is a retrospective study where

patients were receiving hydroxyurea treatment and were following up at the Qatif Health Network between March 2016 and March 2022. A total of 266 pediatric patients with SCD on hydroxyurea, aged 1 to 14 years, 205 patients were included in the study. Baseline and recent weight and height measurements were collected and analyzed. Weight and height percentiles were determined, and growth velocity curves were plotted based on Saudi growth chart. Data were statistically analyzed using the Wilcoxon Signed Ranks Test, and a p-value

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M-P-149

Prevalence Of Peripherally Inserted Central Catheter Thrombosis In Acute Leukemia Patients

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Peripherally inserted central catheter (PICC) is commonly used in the management and treatment of cancer patients for delivery of chemotherapy and blood transfusion. In addition to administration of supportive care such as nutritive fluids and pain medications. However, the use of PICC line was found to be an independent risk of developing thrombosis in cancer patients, most commonly thrombosis of upper extremities.

PICC line related thrombosis is associated with significant morbidity and mortality in acute leukemia patients. Therefore, it is important to estimate the prevalence of thrombosis following PICC line insertion and the investigate the associated risk factors.

Aim: This study aims to estimate the prevalence of catheter-associated thrombosis in patients with hematological malignancies in a single center.

Method: We conducted a single-center, cross-sectional study of 95 newly diagnosed, hospitalized, adult, acute leukemia patients with a PICC line, consecutively selected between May 2015 to December 2020. Demographics, co-morbidities, and thrombotic events following first PICC line placement were collected from electronic records retrospectively.

Results: 95 patients newly diagnosed with acute hematological malignancies were included in our study. The gender distribution among patients was equitable, with an average BMI of 26.2 ± 7.9 kg/m². 51% of patients had acute myeloid leukemia and 49% had acute lymphoid leukemia.

Out of 95 patients, 14(14.7%) were diagnosed with PICC line thrombosis using upper limb ultrasound. Clots were observed in several locations most common was the basilic vein (31%), followed by cephalic, subclavian, and axillary, all at 18% of observed thrombi locations related to PICC line. We found no significant association between patient-related factors and the occurrence of PICC thrombosis.

Conclusion: The prevalence of PICC line-related thrombosis in acute leukemia patients in our center is comparable to other retrospective studies. We found no association with patient related factors contrary to other studies which reported an

association with older age and obesity. We recommend further studies to estimate the incidence of PICC line-related thrombosis in a larger sample size and/or a multi-centric approach."

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M-P-203

Overview of Sickle-cell disease management in the Pediatric Hematology and Oncology Center of Rabat (Morocco)

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Introduction: Sickle-cell disease (SCD) is the most common genetic disease with around 312,000 newborns every year with sickle cell disease, 80% of them in Africa. SCD is characterized by multiple-system morbidity and an increased risk of mortality.

In the absence of a national register and neonatal screening, data on sickle cell disease in Morocco are approximate. The WHO estimates the rate of haemoglobinopathy carriers in Morocco at 6.5%, which would suggest the existence of 30,000 cases of major forms of thalassaemia and sickle cell disease (3,5 of the population thought to be carriers).

A cohort of children with SCD is being followed at the Pediatric Hematology and Oncology Center (CHOP) in Rabat, Morocco and was the subject of our study. The main aim of our study was to describe the epidemiological, haematological profiles and patient follow-up.

Materials and methods: we conducted a retrospective, descriptive and analytical study of all patients aged less than 15 years at diagnosis, with SCD, followed in a Pediatric Hematology and Oncology Center (CHOP) in Rabat between January 1, 2007 and December 31, 2022.

Results: During the study period, 280 children with sickle cell disease were followed up at CHOP of Rabat. The median age of patients was 3 years (3 months-13 years), only 35% of our patients are under 2 years old, with a sex ratio of 1.2 (149 boys and 131 girls). 95% of our sickle cell patients were from the north and north-west of Morocco, and particularly from 2 towns: Kenitra in the north-west and Larache in the north of Morocco. The majority of patients (249 or 89%) were homozygous SS, 21 patients with sickle β -thalassaemia ($S\beta^0:8$, ($S\beta^+ :13$) and 10 patients with sickle haemoglobin C disease (SC). Half of our patients was transfused at least once (49.6%) and 10 patients in our cohort are immune (3.5%).

SCD-related deaths (n = 10) occurred at a median age of 8 years, due to acute anaemia (n = 5), infection (n = 2), stroke (n = 2) and Acute chest syndrome (n = 1).

Conclusion: Sickle cell disease is common in Africa. In Morocco, this hemoglobinopathy is considered as a public

health problem. Its management requires the implementation of a newborn screening to avoid acute complications associated with significant morbidity and mortality.

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M-P-154

Iron Deficiency Anemia among Patients with Glanzmann's Thrombasthenia: Single Center Experience

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Introduction: Iron Deficiency anemia (IDA) is a common side problem among patients with Hereditary bleeding disorders. Glanzmann's Thrombasthenia (GT) is a defective platelet aggregation due to missing or non-functioning glycoprotein IIb / IIIa. IDA among patients with GT is common, representing a challenging situation, particularly in patients with frequent bleeding that needs medical attention. GT is considered a common bleeding disorder in AlMadinah, Saudi Arabia, with a rate of 1 in 10,000.

Aim: Cross-sectional study describing IDA among GT patients.

Methods: Cross-sectional retrospective study extrapolated from AlMadinah GT registry. The patient's biodata was obtained from the registry, and medical records were reviewed. Only those patients with completed iron status data were included.

Results: Among the 125 GT patients, biodata obtained from the AlMadinah GT registry, 101 (80%) patient medical data were retrieved from electronic medical records, including laboratory data. IDA was diagnosed in 69 patients (68%). Median hemoglobin was 8.3 g/l (3.4-10.6), Median mean corpuscular volume (MCV) was 66 fl (53-72), while red cells distribution width (RDW) median was 21% (15-33), and Mentzer index 20 (16-36). Median serum iron level was 4 umol/l (2-7), median serum ferritin was 16 ng/l (8-30), median total iron binding capacity (TIBC) was 38 umol/l (28-55), and Transferrin saturation was 10.6 % (4-20). Apart from pallor, decreased activities, and fatigability, 3 patients had suffered from pica, 2 had koilonychia, and 2 had smooth tongue. Oral iron therapy alone has been tried in 35 (50%) patients with good response (in terms of IDA correction) in all 35 patients. 9 (13%) patients tried intravenous iron therapy alone, where all responded. Alternate Oral and intravenous iron therapy was tried in 25 (36%) patients, where 17 (68%) patients could correct their IDA. 8 patients were refractory to iron therapy. IDA among GT patients was more common among female patients, with a male-to-female ratio of 1:1.4. 28 (75%) ladies at reproductive age had IDA. 33 (70%) children under the age of 14 years had IDA.

Conclusion: IDA is a common problem among patients with GT and should be considered, monitored, and managed promptly. Children and women of reproductive age are more vulnerable to IDA among GT patients.

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M-P-153

Iron Intake Among Women with Eating Disorders

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Background: Eating disorders (EDs) involve persistently disturbed eating and related behaviors that result in altered consumption or absorption of food with potential physical or psychological function impairment. According to the Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5), there are eight major types of EDs: anorexia nervosa, bulimia nervosa, binge eating disorder, pica, rumination disorder, avoidant/restrictive food intake disorder, eating disorder not otherwise specified, and unspecified feeding or eating disorder. This investigation aimed to study the prevalence of EDs and assess the iron intake of nutrition students and dietitians from multiple provinces in Saudi Arabia.

Methods: This study included 163 female nutrition students and dietitians. EDs were diagnosed using the Eating Attitudes Test – 26 item (EAT-26) and Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition. Iron intake was estimated using repeated 24-hour recall records. Diets were analyzed using food processor nutrition and fitness software. The intakes were compared with the recommended dietary allowances (RDA).

Result: According to EAT-26 diagnostic criteria, 25 participants had ED; accordingly, the prevalence of EDs was 15.4%. One participant (4%) had anorexia nervosa, one (4%) had bulimia nervosa, two (8%) had binge eating disorder, seven (28%) had an eating disorder not otherwise specified, and 14 (56%) had unspecified feeding or eating disorder. Nutrition students were nine (36%) subjects, while 16 (64%) were dietitians. The diet analysis showed that these subjects had a higher iron intake, 38 ± 154 mg, than the RDA of 18 mg for female adults, indicating they had 211% of the RDA.

Conclusions: eating disorders were more prevalent among dietitians than students. The EDs included other specified feeding or eating disorders and unspecified cases. Subjects with ED had an iron intake more than the RDA by 211%.

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H-P-155

Factor V Deficiency And Geroderma Osteodysplastic : In Case Report

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Introduction: Geroderma Osteodysplastic (GO) is a rare autosomal recessive connective tissue disorder. The etiology remains unknown, However in the *GORAB* gene (1q24) have

been described. The association with a constitutional deficiency in factor V was not found. Congenital factor V deficiency is a rare autosomal recessive bleeding disorder caused by mutation of the F5 gene located on chromosome 1q23. The association of the two and the genetic site opens up a reflection.

Objective: Describe an exceptional clinical case of factor V deficiency associated with GO, this patient must be operated for a congenital hip dislocation, illustrating the preparation for surgery as well as the postoperative monitoring of the risk of bleeding.

Observation: The case of a 4 year old girl. Who, her sister had the same phenotype but was not diagnosed and died of a cerebral hemorrhage

The first symptoms were noted at birth with hypotrophy, then at the age of 1 year, instability when walking and at the age of 3, the patient presented episodes of epistaxis without any other history of hemorrhage, reason for his consultation. The examination finds failure to thrive, drooping cheeks, a wrinkled face, wrinkled hands and feet, hyperextensibility and dry skin especially in the extremities, ligamentous hyperlaxity and joint instability, dislocation of both hips and dorsal hyperkyphosis without cognitive deficit.

Preoperative assessment shows the prothrombin time at 25%, a Cephalin Kaolin time of 91.1 seconds versus control at 32.2 seconds (3xTime). The factor V dosage was less than 1%, while the other factors were normal, the liver test was normal. The family assessment is normal. The analysis of the clinical exome returns in favor of GO with presence in the homozygous state and with a mutation (describe the type of mutation) clinical exome returns in favor of GO with presence in the homozygous state and with a mutation in the GORAB gene located at chromosome 1q24. The patient was put under reducing traction for her dislocation and then underwent surgery. The factor V level after fresh frozen plasma infusion (30 IU/Kg) was 20%. The surgery is carried out under fresh frozen plasma coverage at a dose of 30 IU/kg once per 24 hours, without hemorrhagic incidents.

Conclusion: Early diagnosis allows the prevention of bleeding and the monitoring of the patient on the hematological and orthopedic level. The contiguity of the two genes favored an altered molecular rearrangement due to an altered mutational mechanism that could be studied at a later time. However, genetic counseling is necessary in these cases.

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