



Conference Proceedings

Fourth ASPED/ISPAD Diabetes Academy Summary Report, 19th-21st April 2018, Dubai, United Arab Emirates

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Abstract

The 4th ASPED-ISPAD Diabetes Academy is an initiative by the Arab Society of Pediatric Endocrinology (ASPED) in collaboration with International Society of Pediatric and Adolescence Diabetes (ISPAD) and is exclusively sponsored by Lilly, Gulf, UAE.

The 4th ASPED/ISPAD Diabetes academy was held on 19-21 April, 2018 in Dubai, United Arab Emirates. The course was accredited with 10 CME hours, by University of Sharjah, UAE. Aimed towards young healthcare professionals in the field of pediatric endocrinology and diabetes with a primary focus on actively supporting training and education in the region. An expert faculty panel from both ISPAD and ASPED from 11 different countries selected candidates following a competitive enrollment process, announced on each organizations respective websites. This year 70 candidates were accepted (out of 115 applicants) from 13 countries (Figure).

The curriculum is delivered in three main formats; plenary, workshop and debate sessions. The plenary sessions covered novel therapeutic approaches, diabetes prevention, use of technology in diabetes management, comprehensive diabetes care, dietary challenges, monogenic and rare types of diabetes, acute and chronic diabetes complications, psychology and patient empowerment, obesity and type-2 diabetes in children. In workshops, the focus was on research methodology, nutrition, psychology and technology within smaller groups, which provided a forum for candidates to present either a diabetes research project or unique presentation of a clinical case. Winners selected by the steering committee members presented in the final plenary session. This intensive 3 days of the course has consistently aimed and successfully provided a concrete educational platform for seamless exchange of scientific information. This has contributed to improvement of care and outcome for children and youth with diabetes in the ASPED region.

Faculty Abstracts

Pathogenesis of Type 1 Diabetes – Lessons Learned from the DiViD-Study

Lars Krogvold, Oslo, University hospital, Norway

The natural history of type 1 diabetes (T1D) was described by George Eisenbarth in 1986. In recent years, his model has been slightly modified. Still, T1D is considered to be caused by one or several environmental factors in individuals genetically at risk. There is strong general evidence for influence of environmental factors in the aetiology of T1D, like variation in incidence between countries, increasing incidence over time and results from migration studies. When searching for potential aetiological factors, these factors may influence the risk for T1D early (even in utero) or later in life, acting as initiator of autoimmunity in individuals with genetic susceptibility or promoting clinical T1D in individuals with presence of autoantibodies. Many hypotheses about non-genetic etiologic factors in T1D exists; among these are infections, nutritional factors (ie. Vitamin D) and perinatal factors (ie. Caesarean section and maternal age). Through the Diabetes Virus Detection study (DiViD), we have through different methods enlightened the presence of small amounts of enterovirus, both virus proteins and viral RNA, in the islets of Langerhans in live individuals with recent onset T1D. The same viruses are not present to the same extent in non-diabetic controls. Based on this and other studies, we are now launching a randomized control trial with anti-viral medications or placebo to children aged 6-15, with the primary aim to preserve their insulin-secretion ability.

Towards Diabetes Cure

Lars Krogvold, University hospital, Norway

Ever since the milestone discovery of insulin in 1922, the treatment of type 1 diabetes (T1D) has been insulin. Primary prevention studies target very young children with increased risk for developing auto-antibodies and later T1D based on their HLA/family history. The proportion of these individuals that will develop T1D is low, therefore the intervention has to be extremely safe. All primary prevention trials to date have involved dietary interventions designed to interrupt putative environmental triggers of Type 1 diabetes. Examples of failed primary interventions include late introduction to cow's milk (the TRIGR study) and gluten (the BABYDIET study). Secondary prevention studies target individuals with the presence of one or several autoantibodies (pre-diabetes). Several attempts have been made, in fact the last 10 years more than 100 antigen or non-antigen specific immune modulating trials have been launched, unfortunately with limited or no success. The reasons for this may be several: T1D is a heterogeneous disease, too much emphasis has been put into animal models and we know too little about the underlying mechanisms for the development of T1D. Still, the future is bright: By addressing all three components of the immune system (Innate, adaptive and regulatory immunity) and thinking "out of the box" with new and combined therapies, the secrets of T1D will be revealed. To manage this, all children with recent onset T1D should be asked to participate in a clinical intervention study.

Insulin Adjustments Due to Exercise for Children with Type 1 Diabetes

Klemen Dovc, Department of Pediatric Endocrinology, University Children's Hospital, Ljubljana, Slovenia

Regular physical activity is essential for people with type 1 diabetes (T1D), with beneficial effects on cardiovascular health and physical fitness, insulin requirements, metabolic control and general wellbeing, and can diminish the risk of premature mortality. Unfortunately, the majority of people with T1D remain sedentary, exercising less than advised. Recommendations on exercise for children and adolescents living with T1D are the same as for the general population – at least one hour of physical activity each day, comprising mainly of aerobic activity, with additional higher intensity exercise a minimum of three times a week. Due to differences in type and duration of exercise, exercise-related hormonal responses, different gender and age responses to physical activity, a personalized glucose management plan should be made for each individual. This plan should include advice on glucose monitoring, exercise timing, carbohydrate intake, insulin dose modification and avoiding injecting insulin at sites involved in muscular activity.

Several strategies for insulin adjustments have been suggested and should be tailored based on factors mentioned above. Most common aerobic activities lasting more than 30 minutes are likely to require a reduction in insulin dose during exercise and up to 90 minutes before. In addition to this, the risk of nocturnal hypoglycemia is increased following afternoon exercise. With MDI treatment, a 20% basal analog dose reduction on the day of exercise, together with a reduction of meal bolus insulin by 75% one hour before exercise and a carbohydrate snack at bedtime, can improve glycemic control. For insulin pump users, a combination of pump suspension, or a temporary decrease in basal insulin infusion rate (e.g. 50%) implemented at least 90 minutes before starting exercise to give a reduced basal effect can be advised. A temporary basal reduction of approximately 20% at bedtime for 6 hours helps reduce the risk of nocturnal hypoglycemia. When exercise is planned at a time of peak insulin action (e.g. soon after a meal), a marked reduction in insulin dose should be made.

In conclusion, it is paramount that the majority of children and adolescents with T1D are engaged in regular exercise. It is the responsibility of health care professionals to be familiar with current exercise-related recommendations, as the risk of glycemic excursions can be mitigated with structured education, particularly in subpopulations prone to hypoglycemia. Novel

technologies, such as continuous glucose monitoring, smart insulin pump features (e.g. suspend before low), single- and dual-hormone closed-loop systems, could provide opportunities for better glucose management in the near future.

Technology Workshop Abstract

Nancy El Barbary, Diabetes Unit, Ain Shams University, Cairo, Egypt

Insulin delivery through continuous subcutaneous insulin pump technology aims to improve the mimicking of physiological insulin secretion. Recent literature supports potential benefits of pump therapy and its safe and effective use in all pediatric groups with T1DM. Achieving targeted glycemic control, reduction of severe hypoglycemia with no significant increase in BMI z-score, reverting hypoglycemic unawareness, decreased total daily insulin doses, episodes of DKA and glycemic variability have been reported.

Pump treatment

Basal Rate: A pump delivers programmable basal insulin which is tailored to the patient's 24 h glucose profile (insulin need). The insulin requirements may be adjusted according to the individual's physiology and lifestyle. Moreover, the advanced features of pump therapy include the ability to set temporary basal rates allow for adjustments to the usually programmed basal rate: decreasing the delivery in the case of physical activity or increasing doses for situations like inter-current illness, Similarly, different pre-programmed basal patterns can be utilized when days of differing insulin sensitivity are predictable, for example during fasting in Ramadan or menstruation.

Meal and correction bolus: Around meals and snacks extra insulin needs to be administered. These bolus doses of insulin will be calculated based on 3 parameters: the measured glucose value, the expected carb content of the meal and pre planned bolus settings

The pumps usually have a bolus wizard, which calculate the insulin dose once the three parameters are provided.

Continuous glucose monitoring: continuous glucose monitoring (CGM) provides continuous interstitial glucose values. Via a small catheter and transmitter interstitial fluid, glucose readings will be measured every 3–5 min. CGM can be blind or real time and can be used for diagnostic or therapeutic options.

The flash glucose monitoring (FCGM) systems consists of a 0.5 cm glucose sensor inserted under the skin and connected to a water resistant, plastic coin-size skin patch. It is worn for 14 days and does not require fingerstick calibrations. No alerts are given. Glucose levels are reported when the user scans the sensor by holding a reader, or a cell phone, close to the sensor. Real-time interstitial glucose levels and glucose trend arrows as well as a graph of current and stored glucose readings are provided on demand. Real-time CGMs utilize real-time alarms for thresholds and predictions of hypo- and hyperglycemia, as well as rate of change alarms for rapid glycemic excursions. Some sensors are integrated in the insulin pump. With specific algorithms in these pumps, insulin delivery may be interrupted or increased to prevent hypo or hyperglycemic values. It has been promoted as being a safety feature against hypoglycemia, especially during sleep or in patients who have hypoglycemia unawareness, while not leading to deterioration of glycemic control, as measured by HbA1c and increasing 'Time in Range'. In addition, new technological developments now enable some CGM sensors to transmit signals to the 'cloud', and allow for digital remote monitoring, through which caregivers are able to view a patient's CGM tracing and receive alerts on their own devices, including smartphones, tablets, and smart watches.

The effectiveness of CGM in children and adolescents with T1D is significantly related to the amount of sensor use. In this workshop, we will discuss an overview of pumps and CGM and their use in children and adolescents with type 1 diabetes then have a pump/sensor download slides for case discussion.

Medical Nutrition Therapy in Pediatric Obesity

Carmel Smart, John Hunter Children's Hospital, Newcastle, Australia

The prevalence of pediatric obesity is increasing worldwide and is associated with an increased risk of type 2 diabetes in young people. Medical nutrition therapy is a cornerstone of treatment and should be incorporated as part of an intensive lifestyle program targeting the whole family. Individualized interventions should aim for a sustained weight loss of at least 5-7% for adolescents and management of co-morbidities including dyslipidemia. Recommendations for dietary composition should be food rather than nutrient based. The overall aim is to decrease caloric intake and increase physical activity within the context of routine family based meals. Healthy food options are based on reducing total fat and increasing fibre and include fruit and vegetables, wholegrains, nuts and legumes. It is important to avoid sweetened beverage consumption including fruit juice and refined carbohydrates. Treatment adherence is a challenge in lifestyle interventions and frequent monitoring is needed. Current evidence indicates that one diet type is not superior to another in terms of long-term weight loss and maintenance. Studies suggest it is degree of adherence rather than the diet type that predicts positive outcomes, although very low carbohydrate diets should be avoided in children. In cases of morbid obesity in post-pubertal adolescents, ketogenic diets can be used alongside careful monitoring by the multi-disciplinary team.

It is important that all children and adolescents are screened for disordered eating, particularly binge eating. The risk of teasing, stigmatization and low self-worth are increased in childhood obesity and referral for mental health support should be considered. It is vital that the combination of diet and physical activity interventions be carried out in the family context with support provided at school if possible.

Results of ASPED Ramadan Survey

Nancy Samir Elbarbary, Diabetes Unit, Ain Shams University, Cairo, Egypt

Background: The impact of fasting during Ramadan on diabetes had been highlighted by many studies and management guidelines have been proposed by several professional groups and individuals. However, some children and adolescents with diabetes still experience some untoward events from suboptimal management during Ramadan fasting. **Aim:** The aim of this quality assurance survey is to identify any gaps in our knowledge and practice on diabetes management during Ramadan fasting in Arab countries, so we can guide our future educational activities toward improving the skills in this area. **Methods:** An electronic survey was performed using an online questionnaire of a large pool of practicing physicians. The questionnaire covered various aspects of management Ramadan fasting in patients with diabetes. Responses from 138 eligible and willing physicians were included in this survey. Majority of participants were pediatric endocrinologist/diabetologist 87 (63%), pediatrician with interest in diabetes 32(23.2%) and adult physician looking after adolescents 19(13.8%). **Results:** Most respondents (79.9%) allow their patients to fast in Ramadan if they asked for it. 75% of respondents favored conducting structured Ramadan education sessions between 2-4 weeks before Ramadan. This was reflected that 69.1% of participants responded that their patients were able to complete more than 50% of fasting days in Ramadan. Hypoglycemia unawareness was the most serious condition where the patient is categorized as being in a very high-risk group to fast during Ramadan with a response rate of 44.9%. Most of respondents (62%) reported that fasting has to be broken if blood glucose levels fall to symptomatic hypoglycemia regardless of glucose level while 49% found that the fast should be broken if blood glucose exceeds 300 mg/dl. **Conclusion:** There are still controversies regarding medical therapy and practices seen in the ASPED region, perhaps reflecting different training and affiliations. Management approaches most suitable for patients in this region are needed.

Fasting Ramadan in Young People; A Literature Review

Asma Deeb, Paediatric Endocrinology Department, Mafraq Hospital, Abu Dhabi, UAE

Fasting Ramadan is one of the five pillars of Islam. Although pre-pubertal children are exempt from fasting, many insist on observing Ramadan. In the literature, type 1 diabetes is considered a risk for fasting. As many young people with type 1 diabetes fast (regardless of medical advice), it is highly important to study the ability and the safety of fasting by young people.

Although, the literature is rich in data on fasting in the adult population, data remains sparse in adolescents and older children. Various areas of research into this topic are required. Rate of complications and safety is on top of the required data. Willingness and ability of fasting is important to study, too. Data on the best insulin regime during fasting is lacking and requirement for insulin adjustments is not consistent in various studies.

With the era of technology, well-structured studies should be feasible to produce evidence-based recommendations on this important area.

Obesity, Insulin Resistance to Type 2 Diabetes; Clinical Presentation

Sarah Ehtisham, Mediclinic City Hospital, Dubai, UAE

As the prevalence of childhood obesity has increased, type 2 diabetes has emerged in the paediatric population. Key risk factors include not just obesity, but also growth pattern, ethnicity, family history, activity level and lifestyle. Alterations in insulin sensitivity with the development of insulin resistance pre-date the onset of glycaemic dysregulation by many years, and it is apparent that impaired insulin sensitivity also mediates rises in blood pressure, triglycerides and inflammatory markers leading to features of metabolic syndrome even before hyperglycaemia develops. These changes increase the risk of macrovascular disease and it is important to be aware that children presenting with type 2 diabetes may already have macrovascular complications and comorbidities of metabolic syndrome at presentation. Type 2 diabetes usually has an indolent presentation in childhood but can also present acutely with hyperglycaemic hyperosmolar syndrome which has a significant mortality rate.

Clinical cases will be presented to illustrate the presentation of type 2 diabetes in childhood and the complexities of distinguishing it from type 1 diabetes.

Medical Nutrition Therapy in Diabetes

Sheryl Salis, Nurture Health Solutions, Mumbai, India

There is a steady rise in the incidence of Type 1 Diabetes. Medical nutrition therapy is a main pillar for diabetes management. The goals Medical Nutrition Therapy for people with Diabetes are to attain individualized glycemic, blood pressure, and lipid goals, achieve and maintain bodyweight goals, delay or prevent complications of diabetes, address individual nutrition needs and to empower the person having diabetes and his family members with the right food choices and options to maintain the pleasure of eating.

55-60% of total energy should come from carbohydrates. While choosing carbohydrates, the glycemic index (GI) of foods and glycemic load should be considered. Low GI foods like whole grains and pulses, legumes, whole fruits vegetables should be preferred. High GI foods like sugars, processed cereals, root vegetables, fruit juices, should be avoided. The amount of carbohydrate in a meal and available insulin determine glycemic response and hence glycemic load becomes important factor to consider while developing meal plan. Monitoring total grams of carbohydrates, by use of carbohydrate counting, remains the key strategy in achieving glycemic control for people having type 1 diabetes. For people with type 2 diabetes who

are prescribed a flexible insulin therapy program, education on how to use carbohydrate counting or estimation to determine mealtime insulin dosing can improve glycemic control.

Research studies have shown that diet with greater protein content, i.e. 15-18% of total calories, may improve glucose and insulin concentrations, reduce appetite and improves satiety. In individuals with type 2 diabetes, ingested protein can increase insulin response without increasing plasma glucose concentrations. Hence, emphasis should be on first class protein to ensure renal safety. In individuals with type 2 diabetes, ingested protein appears to increase insulin response without increasing plasma glucose concentrations. Therefore, carbohydrate sources high in protein should not be used to treat or prevent hypoglycemia.

Recommendations for dietary fat remain the same as those without diabetes with a history of CVD (25% of total energy consumption) which includes 15-20 g visible fat, and saturated fatty acids less than 7%. The choice of dietary fat is important. Consumption of oil, ghee, butter all included should be limited to 0.5 kg/month/person. Diets high in PUFA appears to have effects on blood lipids similar to those diets high in MUFA. And hence, replacing saturated fatty acids and trans fats with MUFA and PUFA have shown to lower cholesterol levels.

Insulin Resistance and PCOS in an Adolescent

Hala Tfayli, American University of Beirut Medical Center, Beirut, Lebanon

Polycystic Ovary Syndrome (PCOS), a heterogeneous disorder characterized by androgen excess, irregular menses and/or cystic ovarian morphology, has peri-pubertal onset. Genetic and environmental factors that influence steroidogenesis, steroid metabolism, neuroendocrine function, insulin sensitivity and adaptation to energy excess are believed to play a role in its pathophysiology. The diagnosis of PCOS in adolescents remains challenging due to the overlap between some of the features of PCOS and the normal changes that occur frequently in the pubertal years. In addition to androgen excess and reproductive disturbances, some adolescents with PCOS are at higher risk of metabolic derangements with long-term health sequelae. Obesity, impaired glucose tolerance, diabetes and the metabolic syndrome are highly prevalent among these youth. Insulin resistance and the consequent hyperinsulinemia are believed to play a pivotal role in the development of the PCOS associated metabolic disturbances and in promoting an ongoing state of androgen excess.

Given that PCOS is not only a reproductive but also a metabolic disorder starting early in adolescence necessitates that therapeutic options target the hormonal as well as the metabolic disturbances. The conventional treatment of PCOS has been oral contraceptives (OCPs) and anti-androgenic agents. Alternative treatment modalities that target insulin resistance including insulin sensitizers such as metformin, the thiazolidinediones (TZDs), the glucagon-like peptide agonists, and metabolic surgery have been studied with variable results.

In this session we will briefly discuss some of the diagnostic dilemmas in PCOS diagnosis in adolescents, review the available data regarding the role of insulin resistance in PCOS pathophysiology and summarize some of the data regarding different treatment modalities in the adolescent age group.

Monogenic Diabetes - Palestinian Experience

Abdulsalam Abu-Libdeh, Endocrinology unit, Makassed Islamic Hospital, Palestine

Monogenic diabetes is a rare type of diabetes resulting by a single gene mutation. It accounts for about 1-2% of all diabetes cases. Inheritance may be spontaneous de novo, autosomal dominant or autosomal recessive.

Transient neonatal diabetes is usually diagnosed within the first week and resolves around 12 weeks of age.

Permanent neonatal diabetes appears within the first 6 months of life and persists throughout lifespan. Different cases with monogenic diabetes will be presented to highlight the Palestinian experience in this rare form of the disease. Special emphasis will be thrown over measures for appropriate management, genetic diagnosis and counselling. Early detection of the condition in the affected patients enables early therapeutic interventions, avoiding complications and predict the clinical course of the disease.

Research in Medical Training, the Saudi Experience

Ibrahim Al Alwan, Pediatric Endocrinology, Ministry of National Guard Health Affairs, Riyadh, KSA

Research is the systematic collection, analysis and interpretation of data to answer a certain question or solve a problem. To ensure a reliable future supply of high-qualified "Clinician-Scientists," in our region and to foster unconventional thinking and innovative research approaches. This presentation aims to promote clinical research in our region. I used the Saudi experience in clinical research which may provide valuable guidance and resources for early career physicians who are transitioning out of residency or in fellowship training, and as well as those in their early years in practice. Some of these experiences brought innovative ideas with excellent research projects, lead to strong impact in clinical practices at national and some at international levels. Obstacles facing new researchers will be discussed. Finally, a promotion and support for conducting and fostering national and regional collaborative clinical research in pediatric endocrinology.

Monogenic Diabetes; Clues to Diagnosis of Diabetes Other Than Type 1

Abdelhadi Habeb, Maternity and Children Hospital & Prince Mohammed bin Abdulaziz Hospital, Madinah, KSA

Although the majority of children with diabetes have type 1 other forms of childhood diabetes do exist. Following the rising epidemic of childhood obesity pediatricians started to see more cases of type 2 diabetes and advances in molecular genetics led to identifying some children with diabetes due to single gene defects, the so called monogenic diabetes. In addition, with the increase in the survival rate of children with cancer and other chronic illnesses cases of secondary diabetes became more prevalent.

The importance of making the correct classification of childhood diabetes are numerous: It could guide the best treatment for diabetes, define the diagnosis in other family members and explain other associated feature. However, if not sure it is safer to treat any child with diabetes as type 1.

The presentation will discuss when type 1 diabetes is unlikely and provide clinical examples of different forms of non-type 1 diabetes with more focus on monogenic diabetes.

Genetics Related Obesity and Diabetes

Abdelhadi Habeb, Maternity and Children Hospital & Prince Mohammed bin Abdulaziz Hospital, Madinah, KSA

Obesity has become one of the most public health problems worldwide, as the prevalence is increasing and the comorbidities associated with this epidemic including diabetes as part of a disease or as a consequence. Genetic factors play an important role and interact with environmental factors to produce obesity. Children with genetic syndromes associated with obesity typically have early onset obesity and characteristic findings on physical examination.

The genetic cause has been isolated in special diseases or syndromes, or attributable to a mutation in a single gene involved in regulation of body weight.

Here will discuss selected genetic syndromes associated with obesity and part of these syndromes develop diabetes mellitus.

MDI or Pump: Is That the Question?

Carine de Beaufort, DECCP/CHL Luxembourg, UZ Brussels, Belgium

Optimizing metabolic control while improving/maintaining a good quality of life, is the challenging objective of diabetes treatment in all age groups. Based on several recent reports, achievement of this good metabolic control cannot be postponed till after childhood. Intensive and ongoing education is the cornerstone of diabetes treatment in all age groups. To make optimal use of tools –different insulins and (blood) glucose control materials – is essential to meet treatment targets. Meal adjusted insulin administration or pump treatment are the current standard treatment, in combination with frequent blood glucose measurements /continuous glucose monitoring. When applying meal adjusted insulin treatment, insulin injections with pens or syringes will be given before each meal. This allows an adjustment for each meal, allowing to correct the blood glucose, calculate carbs content of the meal and evaluate what the hours after the meal will bring (calm /sports, stress etc). Three times a day to adjust and be proactive in determining the insulin dose. Before bedtime the long acting analogs will be administered, allowing Insulin coverage overnight.

The introduction of the pumps in children last century allowed a continuous insulin delivery in combination with meal boluses. This development however, is not sufficient to meet treatment targets. Although easier for the very young children, it remains questionable whether pump in youth is helpful. Continuous presence of the disease, technical problems, alarm fatigue and skin reactions are some of the drawbacks of these tools, leading to deteriorating metabolic control and quality of life. Life-long impact of the needles/Teflon catheters on the subcutaneous tissue is not yet appropriately investigated. Last but not least, the current systems still do not take over diabetes management, leaving the patient in charge and responsible for meal adjustments. These risks in combination with the costs of this treatment need to be considered before starting a child on pump treatment.

Delegates' Research Presentations Abstracts

Retrospective study: Epidemiology of Type 1 Diabetes Mellitus in Basrah, Southern Iraq

Dhaighum Almahfoodh, Faiha Specialized Diabetes, Endocrine and Metabolism Center, Basrah, Iraq

Aims: To investigate the epidemiology of type 1 diabetes mellitus (T1DM) in Basrah city, Southern Iraq, between 2012 and 2016 among people 0–40 year old. **Methods:** This was a retrospective data analysis of electronic archives for patients with T1DM registered in Faiha Specialized Diabetes, Endocrine, and Metabolism Center (FDEMC), which is a tertiary referring Center in Basrah. The data include electronic database from August 2008 to February 2016. Incidence and prevalence rates are expressed per 100,000. Population of Basrah estimates were derived from official data of The Ministry of Planning of Iraq. **Results:** There were 2536 people registered at FDEMC. Of them 53.5% were males. The overall mean age at first diagnosis was 15.3 ± 9 years and it was significantly higher in males ($P=0.0005$). The prevalence rate of T1DM in people 40 years old and younger in 2016 was 87 per 100,000. Between 1 January 2012 and 31 December 2016, there were 818 identified new cases of T1DM. Of these, 417 (50%) were males. The average annual incidence rate of T1DM was 7.4 per 100,000 (95% CI, 7.1–8.1). **Conclusions:** The incidence of T1DM in Basrah lies in the “intermediate group” according to DIAMOND project group classification. The incidence was increasing over the last three years. The data produced by this study provide a baseline for assessing future changes in the epidemiology of T1DM in Iraq.

Effect of Iron Deficiency Anemia on HbA1c

Ziyad Salman, Al Muthna medical center of diabetic and endocrine

Background: Glycated haemoglobin (HbA1c) is used in diagnosis & monitoring of diabetes mellitus is found to be affected by many factors other than blood glucose; iron deficiency anemia is one of them. **Objective:** Aim of this study to prove that HbA1c alone is not reliable for diagnosis or monitoring diabetic patient's control. **Method:** We reviewed Medline, Pubmed and NCBI, 8 studies in total and data were extracted and summarized. **Results:** After 8 studies were reviewed, we found a total of 50 controlled diabetic patients, with mean of HbA1C in controlled diabetics with and without iron deficiency anemia (IDA)were 8.81 ± 0.13 and 5.79 ± 0.01 respectively ($P < 0.05$). On the other hand, 30 non-diabetic patients with and without IDA were 6.84 ± 0.07 and 5.12 ± 0.04 respectively ($P < 0.05$). The difference between no, mild, moderate and severe anemia in both diabetics and non-diabetics was statistically significant ($p < 0.05$). Mean HbA1C% was highest in groups with severe anemia. In addition, a study of 35 patients with iron deficiency anemia was also included. All were non-diabetic based on fasting blood sugar (FBS), post prandial blood sugar (PPBS) level & history. HbA1c level, serum ferritin, serum iron level and blood cell indices were calculated before and after 3months of iron replacement. It was found that after iron therapy HbA1c level was significantly decreased & serum iron levels were significantly increased. HbA1c $6.13\% \pm 0.6\%$ before treatment and $5.12 \pm 0.5\%$ after treatment. Importantly FBS before and after treatment remained significantly positively correlated as was PPBS. HbA1c level was reduced with iron replacement in iron deficiency anemia independent of blood sugar. **Conclusion:** We found positive correlation between iron deficiency anemia and increase level of HbA1c in both diabetic and non-diabetic patients.

Epidemiology Study of Newly Diagnosed Type 1 Diabetes Mellitus, to Assessing the Awareness of Level of HbA1C in Dhi-qar Province, IRAQ

Razzaq Jameel AL-Rubae, Pediatrics Department, Dhi-Qar university, College of Medicine

Objective: A comprehensive analytical study for representative sample of newly discovered Type 1 diabetic patient aimed to assess the epidemiology and sociodemographic data in Dhi-qar province (southern of Iraq) in children aged <15 years. The present study also aimed to measure the educational level, prior to first presentation among newly diagnosed T1DM, by measuring the level of HbA1C and duration of illness. **Methods:** Prospective cross-sectional analytical study extended from February 2015 -December 2016. 103 patients newly diagnosed Type 1 DM was included and data was collected from diabetic center in Dhi-qar province. **Result:** A total 103 patients were diagnosed for the first time, during the study period of which 61 patients were males and 42 patients were females. During the months, of February to September, 40% of patients presented. Positive family history was seen only in 47 cases. HbA1c level was high >8% for all cases but significantly higher in patients who presented late and with higher glucose level at presentation. Male patients showed significant low serum sugar than those in female's patients. 30% of rural areas residents presented with very high serum sugar >500mg /dl vs. 3% in urban area. **Conclusion:** HbA1C is good indicator for poor control and delayed presentation. Rural area resident and female sex presented with high serum glucose and HbA1c at time of presentation.

Variables Associated with Persistence of C-peptide Secretion among Patients with Type 1 Diabetes Mellitus

Ahmed Jaffar Hendi Al Ali, Paediatric Endocrinology, Al-Faiha Specialized Diabetes, Endocrine, and Metabolism Center, Iraq

Background: C-peptide is a reliable method for estimating the beta cell residual function. The objective of this study: to assess the variables associated with persistence of C-peptide secretion among patients with type 1 diabetes mellitus (T1DM). **Methods:** This was cross-sectional study. Enrolled patients with T1DM with at least one year or more duration. Random C-peptide with concomitant plasma glucose at least 144 mg/dl (8mmol/l) was measured and at this cut-off considered as a stimulated value. Variables that was assessed were age at the time of enrollment, age at diagnosis of diabetes, gender, family history of diabetes, duration of diabetes, frequency of insulin injection per day, insulin dose (units/kg/day), type of insulin, devices delivery, BMI at enrollment, blood pressure, glucose (plasma), lipid profile, HbA1c, TSH and antibodies to glutamic acid decarboxylase (GAD65), thyroid peroxidase antibodies (anti-TPO) and tissue transglutaminase antibodies-IgA (anti-TTG-IgA). **Results:** A total 324 patients were included in the study. A Higher level of C-peptide has been seen if the disease acquired at age after 18 years with detectable C-peptide seen among 17.7% compared to 31.7% in those diagnosed below 18 years. The longer the duration of diabetes, the more the loss of C-peptide. On logistic regression analysis, only duration of diabetes less than six years, and insulin dose <1 U/kg/day were statistically significantly associated with the detectable level of C-peptide in this cohort of T1DM. **Conclusion:** Diagnosis of T1DM at a late age, lower insulin requirements were associated with higher/detectable C-peptide.

Risk Factors that Influence Diabetic Retinopathy in Children and Adolescents with Type 1 Diabetes Mellitus

Wasnaa Hadi, Department of Pediatric, Al-Mustansyria University, Baghdad-Iraq

Background: Diabetic retinopathy is a highly specific vascular complication of diabetes mellitus. **Objective:** To estimate the association between the various risk factors implicated in the occurrence of diabetic retinopathy in children with type 1 diabetes mellitus. **Methods:** A prospective study was performed on 126 patients with type 1 diabetes mellitus attending the Pediatric Endocrinology consulting clinic at Central child Teaching Hospital in Baghdad city over a period of 4 months (from the 1st of November 2014 to the 1st of March 2015), all patients had a diabetes duration ≥ 1 year. Each patient had been screened with a simplified questionnaire. Study variables include: age, gender, diabetes duration, doses of insulin, blood pressure components, glycated haemoglobin levels, and lipid profile. All patients were referred to ophthalmology department for detection of presence of diabetic retinopathy. Data were statistically analyzed using Pearson's chi square (X²) test and Pearson's independent sample t-test, P-value of ≤ 0.05 was considered as significant. **Results:** A total of 126 patients with T1DM were screened; their ages ranged from 3.3-18 years. Retinopathy was detected in 22.22% patients. The study showed a significantly high incidence of diabetic retinopathy in patients who had diabetes duration for 10 years and above. The mean of insulin doses was significantly higher in patients with retinopathy. The risk of patients with retinopathy was significantly more for those with systolic and diastolic blood pressure $\geq 90^{\text{th}}$ centile. There was no significant association between glycated haemoglobin, total cholesterol levels ≥ 5.1 mmol/l and triglycerides level ≥ 1.7 mmol/l and the presence of diabetic retinopathy. **Conclusion:** Older age, longer diabetes duration, high insulin requirement, obesity and high blood pressure are associated with a higher risk of retinopathy in this cohort.

Prevalence of Celiac Disease among Type1 Diabetes Children in the City of Mosul

Farah Sameer yahya, department of pediatrics, Mosul college of medicine, Mosul, Iraq

Background: Celiac disease is an autoimmune disease that affects GIT that triggered by gluten, a protein which is found in wheat, barely and rye. Celiac disease is found in 4 to 9% of children with type 1 diabetes but, in 60 to 70% of these children, the disease is asymptomatic ('silent' celiac disease). Children with type 1 diabetes are at increased risk for celiac

disease during the first 10 years of diabetes. **Method:** 75 children between age 5-12 years with type 1 diabetes who attend endocrinology and diabetes clinic at our hospital where screened for celiac disease by anti-tissue transglutaminase IgG Ab (ATT-Ab) ELISA results more than 12 considered positive. **Results:** Five out of 75 patients (6.6%) were seropositive for ATT-Ab. Short stature, chronic diarrhea and abdominal distension were the main association in affected patients. **Conclusion:** The prevalence of celiac disease among type 1 diabetes children is 6.6% in our cohort and screening for celiac disease is recommended.

Eye Complications in Children and Adolescent with Type 1 Diabetes Mellitus

Munib Ahmed Alzubaidi, *Department of Pediatrics, College of medicine, University of Baghdad*

Background: The most common type of diabetes occurring in childhood is type 1 DM, which is accompanied by chronic micro vascular changes affecting most body systems, especially the eye, leading to cataract and diabetic retinopathy. Diabetic retinopathy without appropriate management is emerging as one of the leading causes of blindness. **Aim of the study:** To estimate the presence of eye complications (cataract and retinopathy) among children and adolescent with type 1 diabetes mellitus and to study the effect of various factors on their occurrence. **Patients and methods:** This study was carried out over nine months' period (from first of May till the end of January 2014) at Children Welfare Teaching hospital/Medical City. All patients were referred for full ophthalmology assessment. Statistical analysis done using T test, Chi square, P value <0.05 regarded as statistically significant. **Results:** This study included 150 patients with type 1 diabetes mellitus, their age ranges from 4.5-19 years, with duration ranges from 2-18 years. Female to male ratio was 1.8:1. Out of 150 diabetic patients, 24/150 (16%) had eye complications, 9 (6%) of them had retinopathy while other 15 (10%) had cataract. The age of all patients who had eye complication was >10 years which is highly significant (P=0.009) and the incidence of eye complications increases with increasing duration of diabetes (P=0.04). Twenty two (14.7%) female and two (1.3%) male had eye complications, which is highly significant (P=0.002). HbA1c>10 in 17/24 (70.8%) patients with eye complications. There was a significant association between the presence of eye complications and macroalbuminuria (P=0.02). **Conclusions and recommendations:** The incidence of eye complications (cataract and diabetic retinopathy) increases with increasing age and duration of diabetes and it is more common in female and it is associated with the presence of other chronic complications of nephropathy.

Early Detection of Diabetes Mellitus in Paediatric Beta-Thalassemia Major Patients Using Continuous Blood Glucose Monitoring

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Background: Pancreatic iron overload and diabetes mellitus (DM) are common in β -thalassemia major (β -TM). Continuous glucose monitoring system (CGMS) enables more diagnostic accuracy and a better achievement of an optimal glycemic control. **Objectives:** To detect early alterations in glucose homeostasis in patients with β -TM using CGM system compared with oral glucose tolerance test (OGTT). **Methods:** This cross-sectional study was conducted on 200 patients β -TM patients. Patients were studied focusing on transfusion history, transfusion index, iron chelation therapy and compliance to chelation. Complete blood picture, serum ferritin and random blood glucose (RBG) were measured. Patients with RBG \geq 140 mg/dL were subjected to OGTT, insertion of CGMS for 3 days and assessment of HbA1c. **Results:** Screening with RBG revealed that 20 patients (10%) had RBG \geq 140 mg/dL. Using OGTT, 7(3.5%) patients were in the diabetic range, 7(3.5%) had normal OGTT while 6 (3%) had impaired glucose tolerance. The CGMS showed that 7(3.5%) patients had IGTT (6.5%) and 13 patients had diabetes mellitus. The percentage of diabetic patients diagnosed by CGMS was significantly higher than that with OGTT (p=0.012). According to CGMS readings, 10 of the 13 patients with diabetes had abnormal HbA1c readings

of diabetic range (6.5-9.9%) while 5 of the 7 patients with impaired glucose tolerance had HbA1c readings in the prediabetic range (5.5-6.1%). Serum ferritin were significantly higher among patients with RBG \geq 140 mg/dL ($p=0.001$). HbA1c was positively correlated to maximum blood glucose, average blood glucose, SDS blood glucose and area under the curve \geq 140 mg/dL. **Conclusions:** The use of CGMS in the diagnosis of early glycemic abnormalities (prediabetes) among patients with β -TM appears to be promising and superior to other known diagnostic modalities namely OGTT and HbA1c.

Effect of Metformin on Hyperandrogenism in Adolescent Girls with Type 1 Diabetes

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Background: Hyperandrogenism with or without polycystic ovarian syndrome is seen in adolescents with type 1 diabetes (T1D), especially those with suboptimal control. **Objective:** To assess the effect of metformin on hyperandrogenism and ovarian function in adolescents with T1D. **Methodology:** It was a prospective study in the outpatient clinic of Diabetic Endocrine Metabolic Pediatric Unit (DEMPU), Children Hospital, Cairo University where 28 T1D females showing signs of hyperandrogenism were included. History taking and anthropometric measures were performed, then the patients were assessed for the manifestations of hyperandrogenism. Measurement of estradiol, testosterone, dehydroepiandrosterone sulfate, androstenedione, prolactin, HbA1c, FSH and LH was done during the follicular phase of the cycle with assessment of ovarian morphology by transabdominal ultrasound, progesterone was measured during the luteal phase. Patients were subjected to 500 mg metformin twice daily orally for one year, then re-evaluated regarding clinical and biochemical parameters. **Results:** Study included 28 girls with mean age of 15.62 years and mean diabetes duration of 5.75 years. Acne was found in 75%, mean hirsutism score (using Ferriman-Gallway score) of 16.5 and 84.6% had polycystic ovarian morphology. Metformin therapy for a year produced a significant reduction in weight ($p=0.001$), body mass index (BMI) ($p=0.002$), acne ($p=0.008$), hirsutism score (0.007), LH ($p=0.008$), testosterone ($p<0.001$) and androstenedione ($p=0.028$) in adolescent girls with T1D. There was significant reduction of the patients with oligohypomenorrhea ($p=0.000$). There were no significant reduction in the daily insulin requirements or HbA1c. Nausea and abdominal pain were the commonly reported complications of metformin (64%). **Conclusion:** Metformin improved BMI and cycle regularity and the clinical manifestations of hyperandrogenism in T1D adolescent girls. However it didn't improve the glycemic control significantly.

Hypoglycemia in Children with Type 1 Diabetes

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Background: Hypoglycemia is a major complication of insulin treatment in patients with type 1 diabetes (T1D), limiting full realization of glycemic control. **Objective:** The aim of our study was to describe the characteristics of hypoglycemia in children followed in our consultation for T1D. **Methods:** We conducted our study (2015-2017) on parents of children with T1D who were asked, at each consultation, about episodes of hypoglycemia. The following characteristics were studied: age, sex, parental education level, hemoglobin A1c (HbA1c) level, insulin regimen, self-monitoring of blood glucose, hypoglycemic events: causes, severity and treatments. **Results:** 330 children (44.2% males), mean age was 10.6 ± 4.2 , diabetes duration was 6.7 ± 2.2 years. 70% of the children used multiple daily insulin injections. HbA1c \leq 8% in 53% of cases. The mean number of glycemic control was 4 ± 2.1 /day. Hypoglycemia was severe in 20% of cases and its overall incidence was 2 events/100 patients/year. The cause was unknown in 54.8% of cases and the treatment was done in the hospital in 40% of cases. The glucagon was used at home in only 14.8% of cases. 33.9% of patients had repeated hypoglycemic episodes. Age of children, HbA1c and parental education level did not play a role in the occurrence of severe hypoglycemia. Duration of diabetes and insulin regimens were factors related to episodes of severe

hypoglycemia. **Conclusions:** Hypoglycemia in our population of children with T1D is not associated with HA1c level or insulin regimens. In half of cases of severe hypoglycemia, the cause is unknown and the treatment is done in the hospital. These results undergo the importance of a therapeutic education program for improving the management to prevent or reduce the occurrence of hypoglycemia for our diabetic children.

Characteristics of Children Presenting with Newly Diagnosed Type 1 Diabetes Mellitus in South Africa

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Background: The clinical presentation of Type 1 Diabetes in children can be acute or insidious, and symptoms may be subtle and frequently misinterpreted. Presentation with diabetic keto-acidosis (DKA) may be associated with significant morbidity and mortality in the paediatric population. **Aim:** To review the characteristics of children presenting with DKA at the time of diagnosis, and to determine the frequency of missed diagnoses in the previous month. **Methods:** A retrospective review of children with newly diagnosed Type 1 Diabetes between January- December 2015. Children presenting with DKA were compared with those who presented without DKA. **Results:** Sixty-three children were diagnosed with Type 1 Diabetes over the study period. Of these, 44 (69.8%) presented with DKA at diagnosis. The median duration of symptoms preceding diagnosis in the DKA group was 2 weeks, versus 4 weeks in the non-DKA group ($p=0.002$). There was no significant difference between the groups with respect to ethnicity, gender and age at presentation. 27/42 (64.3%) of patients presented to healthcare facilities in the month preceding diagnosis and were misdiagnosed. **Conclusion:** Patients who presented with DKA had a shorter duration of symptoms than the non-DKA group. Ethnicity had no effect on characteristics at presentation. There was an unacceptable rate of missed diagnoses of Type 1 diabetes in both private and public healthcare facilities.

Insulin Therapy Regimes, Glycemic Control, Knowledge and Quality of Life in Children with Type 1 Diabetes

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Objectives: To further describe the changes in insulin therapy regimens and HbA1c in children and adolescents with type 1 diabetes, and their associations with diabetes knowledge and quality of life (QOL). **Research design and methods:** The study included 4293 children and adolescents (12.9 ± 2.6 yr, more than one year of diabetes) attending AJD (Aide aux Jeunes Diabétiques) summer camps between 2009 and 2014. The distribution of insulin regimens and the associations between HbA1c, therapeutic regimens, diabetes knowledge (AJD questionnaire) and Quality of Life (QOL, Ingersoll et Marrero, Hvidoere Study Group short version) were assessed. **Results:** The percentage of youth treated with the insulin pump increased up to about 45%, basal bolus stabilized around 40%, and other regimens decreased majorly. HbA1c was higher with regimens using premixed insulins only ($9.05 \pm 2.43\%$), but there was no difference between pump ($8.12 \pm 1.09\%$), basal bolus ($8.32 \pm 1.33\%$) and 2-3 injections ($8.18 \pm 1.28\%$). Mean HbA1c decreased by 0.014% per year. The percentage of HbA1c < 7.5% increased by 1.5% per year, and the percentages of HbA1c > 9% or > 10% decreased by 4% and 5.5%, the changes being greater with the pump. HbA1c was weakly associated with diabetes knowledge, and strongly with general health perception and perception about diabetes. **Conclusion:** The percentage of T1D children and adolescents with the highest risk of complications decreased markedly. The distribution of HbA1cs better depicts the glycemic control in a population than the mean or the percentage of patients reaching the target (7.5%). HbA1c was more strongly associated with general health perception than with therapeutic regimens and diabetes knowledge.

Diabetic Ketoacidosis at Onset of Type 1 Diabetes (Study of 173 Cases)

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Background: Diabetic Ketoacidosis (DKA) at onset of type 1 diabetes (T1D) is still a fairly common finding, particularly in young children, and is the leading cause of morbidity and mortality. **Aim:** To study the epidemiological, clinical, biological, immunological, therapeutic and evolutive features of DKA at onset of T1D in order to better understand the factors associated with its severity. **Methods:** A retrospective, descriptive and comparative study was conducted among children who were hospitalized for DKA at onset of T1D between January 2000 and December 2015 at the Pediatric Department of Hedi Chaker University Hospital of Sfax, Tunisia. **Results:** We collected 173 new cases of type 1 diabetic children admitted for DKA at onset of T1D; the hospital frequency was 28 per 10 000 hospitalizations with an average annual incidence of 11 new cases per year. During the study period, DKA revealed diabetes in 173 of 450 new T1D cases (38.4%). Thirty six (group 1), fifty five (group 2) and eighty two (group 3) children were in mild, moderate and severe DKA respectively. The mean age of the children was 7.13 years \pm 3.98. There were 81 boys (46.82%) and 92 girls (53.18%) (sex ratio: 0.88). There were more children with familial T1D in group 1 (25%) and group 2 (21.8%) than in group 3 (13.4%) ($p=0.24$). Polyuria and polydipsia were reported in 99.4% of cases, its duration was not correlated with the severity of the DKA ($P=0.079$). At admission, 20.2% of children had an altered level of consciousness. The mean venous glucose was 26.17 mmol/ l \pm 6.91. Hypokalemia was more common in group 3 ($p<0.001$). Among our children, 127 children were treated according to the Lestradet protocol and 34 children were treated according to the ISPAD protocol. The decrease in capillary as well as venous glucose, the decrease in ketonuria and the increase in pH were significantly greater in children treated according to the ISPAD protocol compared to those treated according to the Lestradet protocol. The linear regression study showed that during treatment, the ISPAD protocol was more contributive to decrease blood glucose levels, to increase pH and to correct sodium and potassium levels; compared to the Lestradet protocol. The most common complication in the acute phase was hypokalemia (80.12%). After stopping the IV insulin therapy protocol, all children were put on a conventional insulin therapy regimen with 2 injections at an average dose of 0.85 IU/kg/day \pm 0.23. The mean follow-up was 4.07 years \pm 3.15. 2 children who presented with severe DKA died at the acute phase. **Conclusion:** DKA at onset of T1D is a leading cause of death in children with T1D in our cohort. Education of diabetes and DKA as a presentation of new diagnosis of diabetes is crucial amongst primary health care providers.

Predictors of Glycemic Control in Patients with Type 1 Diabetes: A Study from Jordan

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Background: Poor metabolic control in children with type 1 diabetes leads to future microvascular and macrovascular complications. **Objectives:** Identifying children with risk factors for poor metabolic control is important to implement certain strategies in management of these patients in order to decrease the progression of future complications. **Methods:** A retrospective study conducted by chart review of children with type 1 diabetes. The main outcome was metabolic control and it was assessed by HbA1c levels. Linear logistic regression analysis was used to evaluate possible predictors of metabolic control. One-way ANOVA analysis was used to detect significant differences in HbA1c between categories. **Results:** Significant predictors of metabolic control were: age, number of clinic visits, dietary compliance at home and at school, and the person who decided the insulin dose. One-year increase in age lead to a significant increase in HbA1c level. An increase in HbA1c level in children who attended more than four clinic visits was observed compared to those who had attended the usual recommended number of clinic visits. A decline in HbA1c levels was predicted in children who counted carbohydrates

precisely or who did not eat more than 15-20 grams of carbohydrates at school without insulin. When family members (other than the mothers or the patients themselves) decided the insulin dose, the HbA1c level increased significantly. **Conclusion:** Poor metabolic control was associated with certain clinical characteristics including lack of carbohydrate counting and absence of direct mother care. Diabetes medical teams should focus on children with these risk factors to optimize their metabolic control and provide appropriate clinical care plans.

The Prevalence of Celiac Disease in Palestinian Children with Type 1 Diabetes Mellitus

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Background: Celiac disease is an immune-mediated disorder triggered by gluten exposure in genetically predisposed individuals, causing small bowel mucosa atrophy with a more or less evident malabsorption syndrome. The negative impact of CD on health such as growth failure, osteoporosis and intestinal lymphoma may be reduced with dietary treatment. Early diagnosis and prompt management is, therefore, important in reducing both its morbidity and mortality. High prevalence of CD has been reported in patients with type 1 diabetes mellitus (T1DM) with reported prevalence rates ranging from 1-16% in screening studies published all over the world. **Objectives:** To report the prevalence of celiac disease in Palestinian children with type 1 diabetes mellitus. **Search methods:** A cross sectional study was conducted on 237 diabetes mellitus type 1 children attending Palestine Diabetes Institute in Ramallah. They were tested for the presence of anti-tissue transglutaminase immunoglobulin An antibody and total immunoglobulin A level. The patients testing immunoglobulin An anti-tissue transglutaminase-positive were offered small bowel biopsy. **Main results:** A total of 238 children with type 1 DM were initially included, age of participant's ranged from 2 to 28 years. 84 children lost follow up. Of 152 children with T1D who screened for anti- TTG, 27 (17.7%) were positive for anti- TTG. A total of 27 patients underwent upper endoscopies and duodenal biopsies with 15 patients showing histological features consistent with celiac disease (9.8%). There was no statistically significant differences in relation of celiac disease with thyroid abnormality ($p=0.253$) nor body mass index ($P=0.824$). In addition, There was no statistically significant differences in the level of HbA1C in relation to celiac disease among patients with DM1 ($P=0.059$). **Conclusion:** The prevalence of celiac disease among Palestinian children with type 1 diabetes mellitus is about 9.8% one of the higher reported prevalence in the Arab world.

Permanent Neonatal Diabetes Due to kATP Channel Mutation with Switching from Insulin to Sulphonylurea

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Neonatal diabetes mellitus (NDM) is rare, with a prevalence of approximately 1 in 500,000 infants worldwide and might either be transient (TNDM) or permanent. Several genetic mutations might cause NDM and until now, most permanent types of NDM required life-long insulin therapy. Recently, heterozygous activating mutations in the genes forming the ATP-sensitive K⁺ channel (KATP channel), KCNJ11 and ABCC8, have been shown to cause neonatal diabetes. Depending on the facts that sulphonylurea binds to the same channel and would open the KATP channel by a non-ATP dependent route, around 150 cases of permanent NDM - until now – have come off insulin and gone onto sulphonylurea successfully.

We report clinical aspects of the successful transfer to oral treatment in two cases (sisters) of young children who are homozygous for an ABCC8 missense mutation, confirming a diagnosis of recessively inherited neonatal diabetes due to mutations in the SUR1 subunit of KATP channel.

Type I diabetes and autoimmune hepatitis

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Background: Type 1 diabetes (T1D) is often associated with other autoimmune diseases especially autoimmune thyroiditis and celiac disease but may present others rare autoimmune form. **Case report:** We report the case of a 13 years old girl with 7 years of (T1 D). She presented cholestatic jaundice with abdominal pain and nausea, without hepatomegaly. The biological explorations showed poor glycemic control with hemoglobin A1C at 11%, and very high serum transaminases. Anti-muscle Antibodies were positives. The liver biopsy and histology study revealed autoimmune A 1F2 origin. Management was and supportive with multidisciplinary care and she made a full clinical and biochemical recovery. Few cases have been reported and described the association diabetes and autoimmune hepatitis. There is characteristically no progression to fibrosis and cirrhosis and the symptoms are reversible with improved glycemic control and treatment.

Structured Therapeutic Education in the Management of Diabetes: Casablanca Experience

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Patient education is an important aspect of care to young diabetic children. It is proposed not only to help the patient to understand the illness and the treatment but also to help becoming self-sufficient. In 2015 we have created HAYAT, which is a support association for children with type 1 diabetes. This work presents a descriptive study of the therapeutic education program followed in our association. We also evaluate the metabolic balance of patients adhering to this program. The association enrolled up to date 746 young diabetic patients aged 1-18, of which 21% comes from outside Casablanca. 52% of patients are on conventional insulin regimen. Many specialists participates in our education program; diabetologists, dieticians, pharmacists, diabetes educators, art and craft specialist and a dental surgeon. The school of diabetes of the association HAYAT is considered as a first of its kind in Casablanca and allows a better management of the diabetes and makes it possible to promote the autonomy of the patient to live optimally, in order to achieve his personal and professional projects, despite the constraints of disease and treatment.

Children and Adolescents with Type 1 Diabetes at School: Student and Parents Perceptions of Support for Self-Management and Teachers Attitude.

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Background: There are more than 1,106,500 children and adolescents with type 1 diabetes globally with annual 132,600 new cases (IDF 2017). In Saudi Arabia type 1 diabetic below 20 years of age exceeds 35000 patients; most of them are enrolled in public or private schools. The daily self-management of children and adolescents with type 1 diabetes is complex and dynamic. It requires frequent self-monitoring of blood glucose (SMBG), insulin injections, and individual meal plans. Long school hours, changes in daily schedule make diabetes management even more complex. In addition, children are continually growing during each academic year, requiring frequent changes to their individualized treatment plans. Decisions concerning adjustments in insulin doses occur often while children are in school and frequently must be made by a school nurse, teacher, parent, or guardian following protocols in the comprehensive diabetes care plan prescribed by the children's physicians. **Aim of the study:** To improve the outcome of type 1 diabetes management at school. **Method:** Cross sectional study surveyed electronically school aged children and adolescents with type 1 diabetes, their parents, and the school personnel. The investigators electronically approached all children and adolescents and their parents or school teachers

through the National School Health Program, Diabetes/Endocrine Centers, and primary care centers about their interest in participation. A series of questionnaires were translated by the researchers to identify the diabetes-related experiences of the children and adolescents, their parents, and their school personnel. This was not a randomized trial, and all patients were asked to participate in the survey. Thus, a convenience sample was used. **Results and conclusion:** Overall experience dealing with diabetes in school was positive although discrimination still present. Of note the limitations to diabetes self-management at school including supportive nutritional services, adequacy of school personnel training and the school preparedness for dealing with emergencies. Also, a major defects in policies for care and rights of diabetic students exists.

Type 1 diabetes mellitus (T1DM) in toddlers and schoolchildren in Najran region, Southwestern Saudi Arabia–Correlation with osteocalcin and vitamin D

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Background: There is an ongoing interest in the relationship between vitamin D status and diabetes control and complications. However, data from Saudi Arabia are limited. Human studies demonstrated the positive association between insulin secretion, insulin sensitivity and serum levels of osteocalcin and vitamin D. **Aims:** This study aimed to explore the possible role of circulating osteocalcin in the pathogenesis of T1DM and its correlation with fasting plasma glucose (FPG) and vitamin D levels in Saudi Arabia children with T1DM. **Methods:** This cross-sectional observational hospital based- case control study that included 132 Saudi children clinically diagnosed as type 1 diabetes mellitus and 72 apparently healthy children. **Results:** There were statistically weak negative correlations for FPG with UCOC and TOC in diabetic children ($r = -0.294$, $p < 0.01$ and -0.358 , $p < 0.0001$ respectively). A moderately significant positive correlation between FPG and BMI and a significantly negative correlation between 25(OH)D₃ and FPG ($r = -0.62$, $p < 0.0001$) were found. Moreover, there was a moderately significant positive correlation between levels of vitamin D and UCOC in healthy control group ($r = 0.457$, $p < 0.0001$). Such significance was not present in diabetic group ($r = -0.077$, $p < 0.38$). **Conclusion:** Levels of 25(OH)D₃, TOC, and UCOC were significantly diminished in children with T1DM suggesting the bidirectional influence of β cells on vitamin D and osteocalcin and vice versa. The reverse correlation between UCOC and HbA1c may indicate the probable prognostic value of UCOC as the lower UCOC, the worse glycaemic control in diabetic children.

Delegates Case Presentations

IPEX Syndrome; An Interesting Presentation

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IPEX syndrome, namely, immune-dysregulation with autoimmune polyendocrinopathy and enteropathy X-linked recessive syndrome. It develops due to gene mutation responsible for the synthesis of a specific protein forkhead box protein 3 (FOXP3).

IPEX syndrome is a rare disorder that affects an estimated 1 in 1.6 million people. T1DM is a common feature and can be seen as early as in the 1st month of life. Other symptoms due to profound immune dysregulation that can be seen in males with IPEX syndrome include enteropathy, dermatitis, failure to thrive, thyroiditis and recurrent infections Many other autoimmune phenomena like nephritis, pneumonitis, hepatitis, vasculitis, arthritis, myositis, alopecia, and autoimmune cytopenias can be presented. Management for immunodysregulation polyendocrinopathy enteropathy X-linked syndrome has

seen limited success in treating the syndrome by bone marrow transplantation. We report a child with IPEX syndrome who had an interesting presentation of the disease.

Neonatal Wolfram Syndrome: Novel De-novo Dominant Mutation Presenting as an Unusual Clinical Phenotype

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Background: Wolfram, known also as DIDMOAD (Diabetes Insipidus, Diabetes Mellitus, Optic Atrophy and Deafness) syndrome (WS) is a rare neurodegenerative disorder resulting usually from biallelic WFS1 gene mutations. Diabetes mellitus usually its first symptom, rarely presents prior to 1 y of age. **Clinical and molecular studies:** our patient is a 5 years and 10 months old boy from a consanguineous Palestinian family. He presented neonatally at 40 days of age with persistent neonatal diabetes, bilateral cataracts, congenital (prelingual) deafness and left hydronephrosis. During follow up he developed Failure to thrive, microcephaly, severe psychomotor retardation, seizures, severe scoliosis and bilateral lower limbs contractures. Laboratory investigations revealed normal serum electrolytes, lipase and thyroid function tests, normal urine osmolality, low serum insulin levels, negative anti insulin antibodies, normal pancreas by sonogram, 46,XY karyotype, and normal sequencing of the KiR6.2 gene. Whole exome sequencing revealed a heterozygous, c.923 C>T (p. S308F) novel, de-novo, missense mutation in an evolutionary conserved amino acid of WFS1; that was defined damaging by predicting software. Although wolframin 1's function has not been established its known formation as an oligomer suggests, that a dominant negative effect may cause the severe phenotype. **Conclusion:** A novel de-novo heterozygous WFS1 mutation causes a unique and severe WS with cataracts, deafness and diabetes mellitus presenting neonatally. The clinical application of next-generation sequencing technology enhanced the diagnosis of a rare genetic disorder in a patient with atypical presentation and may have a role in defining new clinical manifestations of rare syndromes, such as WS.

Severe DKA in a patient with Acquired insulinoma

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Patient 13 and 1/2 years old female child diagnosed to have insulinoma in June 2015 with history of recurrent attacks of hypoglycaemia. Patient underwent embolization of insulinoma in July 2015 by interventional radiologist. After embolization, patient improved then stopped treatment. In November 2017 patient was admitted with history of recurrent vomiting and recurrent attacks of hypoglycemia. Pancreatic MRI showed Neuro-Endocrine pancreatic tumor and was started on Then started on Diazoxide. In 20-12-2017 patient was admitted as a picture of DKA (pH of 7, s. ketonemia and glucose 750 mg/DL). Patient was treated for severe DKA. Insulin was stopped a week after. This is a rare condition in which acquired Insulinoma complicated with DKA. **Conclusion:** Autonomous insulin secretion from insulinoma might result following embolization. Transient insulin deficiency leading to DKA can complicate the insulinoma embolization.

Glutamate Dehydrogenase Hyperinsulinism; An Interesting Case Scenario

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Background: Mutations in the human HNF4A gene encoding the hepatocyte nuclear factor (HNF)-4 are known to cause maturity-onset diabetes of the young (MODY1). heterozygous mutations in this gene were reported to cause transient an persistent hyperinsulinemic hypoglycemia. We report two patients who presented with persistent, frequent attacks of hyperinsulinemic hypoglycemia. **Case report:** the first patient was born at full term following an uneventful pregnancy to a 25 years old mother. His milestones were normal. After his first year started to have attacks of loss of consciousness and seizure

associated with hypoglycemia. patient was admitted in the hospital and critical samples results showed hyperinsulinemia. Diazoxide started with Dextruse12% IV fluid but patient continued to have frequent hypoglycemia. Octreotide started up to maximum dose and diazoxide stopped. PET scan done showed generalized increased uptake of pancreas suggestive diffuse nesidioblastosis. He underwent near total pancreatectomy which was done laparoscopically. Afterwards, his blood glucose became stable and controlled with octreotide and some frequent hypoglycemia. Molecular genetic report for congenital hyperinsulinemia Next Gen Sequencing Panel result Heterozygous for c.1321A>F in HNF4A. There is positive consanguinity between parent with strong family history of diabetes mellitus in grandfather and aunts also history of similar presentation in one sibling of their cousin but was transient resolved at school age. family history of diabetes mellitus in a one year old sibling in the other cousin. The other patient is a four years old girl with history of frequent hyperinsulinemic hypoglycemia but responsive to octreotide. His gene study result showed heterozygous mutations of HNF4A gene. **Conclusion:** Heterozygous HNF4A mutations can therefore cause both transient and persistent (mild and sever) hyperinsulinemia hypoglycemia

Persistent of Hyperinsulinism Hypoglycemia in Two Brothers

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Nesidioblastosis is a congenital condition that affects newborns and young children. It has two broad categories which are focal adenomatous hyperplasia and diffuse abnormality of the islets.

Case 1: is a 4 years old child admitted for sudden loss of consciousness and abnormal movement. The blood sugar was 15 mg/dl; critical samples showed high serum insulin and c-peptide. Other tests were unremarkable. Central vein line used for the management with poor control. Medical imaging for the pancreas was normal. Pet scan revealed a pancreatic tumor and he had subtotal pancreatectomy. Histopathology showed features of Nesidioblastosis with characteristic diffuse hyperplasia of Langerhans islets with predominantly of beta cells subtype with beta cells metaplastic transformation of ductal basal multipotential epithelial cells with slight enlargement and hyperchromasia. Two months postoperatively, blood sugar was normal without any supportive measures.

Case2: is the older brother who is eight years old. He had a similar presentation to his sibling. Blood sugar was 20 mg/dl with a high serum insulin and c-peptide. Pancreatic MRI showed a pancreatic mass which was confirmed to be a focal form of hyperplasia. His course was milder than his brother and he was managed with medical treatment.

Conclusion: although rare, persistent hyperinsulinemic hypoglycemia of infancy should be considered in early evaluation of hypoglycemia of young infants, or even older children, especially if parents are consanguineous.

Case Report of Diabetic Ketoacidosis Misdiagnosed as Acute Abdomen

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Introduction: Diabetic ketoacidosis represents the most sever derangement of insulin regulated metabolism. The acidosis and ketosis create an ileus causing abdominal pain, nausea and vomiting. Occasionally the ileus will produce pain sever enough to raise concerned for acute abdomen. **Case report:** A 10 years old child, who is a known case of type 1 diabetes mellitus presented with abdominal pain and vomiting. He was referred to pediatric surgery as acute appendicitis. at the theater when the surgeon revealed that his blood sugar high. Blood tests showed a high blood glucose and ketonemia. His blood pH was 7.1. Diagnosis of DKA was made and started intravenous fluids and insulin. The patient improved and abdominal pain subsided. He was discharged home the second day on his original insulin regime. **Discussion:** clinically and biochemically the results were in keeping with diabetic ketoacidosis rather than acute appendicitis. Ileus, secondary to hypokalemia can cause

abdominal pain, nausea, vomiting which might be severe enough to mimic acute abdomen. **Conclusion:** This case report highlights that ileus in DKA might mimic acute abdomen of a surgical origin.

Steroid-induced Diabetes in a Patient with Multiple Autoimmune Disease

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Background: Steroid are widely used anti-inflammatory and immunosuppressive medication, they have many and serious side effects including hyperglycemia. Steroid induced diabetes mellitus (SIDM) is defined as hyperglycemia associated with the use of glucocorticoids in a patient without a prior history of diabetes mellitus. The criteria for diagnosis are the same criteria for type 1 DM. Risk factors include higher doses and prolonged use of steroids. **Case report:** A 13-year-old girl, known case of Hashimoto thyroiditis since the age of 3 years on thyroxin therapy. She was diagnosed to have SLE at the age of 13 years after which she presented in encephalopathic state. She was started on intensive steroid regimens after which she developed hyper glycemia. She has a positive family history of autoimmune diseases.

Examination revealed goiter beside other features of SLE. Investigation confirmed diagnosis of mixed connective tissue disorders. She was put on insulin therapy to control her blood sugar while she was on steroid and she was followed up until steroid was weaned off. **Conclusion:** Different steroids used in different regimens and routes can subsequently impact glucose profile adversely. Diagnosis of steroid induced diabetes is similar to diabetes mellitus and Hyperglycemia usually normalized after stopping the steroid although it may persist, so follow up is important.

Primary Amenorrhea

Eman Abdulla Ali Enaw, Gaafar Ibn Auf Tertiary Hospital, Sudan

Background: Delayed puberty is one of the common problems associated with diabetes. The mean age of menarche in Sudanese diabetic girls is 15 years.

17 years old female presented with primary amenorrhea. She is a known case of diabetes for one year on mixed insulin. Her last HbA1c was 6.7. Breast enlargement started by the age of 11 years and progressed normally. Shortly after that pubic and axillary hair started to grow but there had no menstruation or cyclic pain. No history suggestive of thyroid disease, mother and sisters has their menarche by the age of 14 years. Her BMI was above the 95th centile. She had extensive acanthosis nigricans over the neck, axillae and cubital fossae, no acne or hirsutism, patient has adult type pubic and axillary hair with stage 5 breasts.

Her HbA1c was 6.7. She had a normal thyroid function, negative celiac screening Her FSH is 5.7, LH 7.1 mIU, Prolactin 13.8 (4-28 and testosterone of 0.2(0.1-0.8). Her abdominopelvic ultrasound showed normal uterine size with large ovaries (20 ml bilaterally) with multiple cysts.

Diagnosis: Primary amenorrhea secondary to polycystic ovary disease. **Conclusion:** Polycystic ovarian syndrome should be considered in girls with primary amenorrhea with diabetes particularly if they are obese and have a good diabetes control.

H Syndrome

Samar Abu Samra, Gaafar Ibn Auf Tertiary Hospital, Khartoum, Sudan

An 18 years old male who has type 1 diabetes since the age of 12 years. He was diagnosed with deafness since the age of 8 months. Over the years, he developed other clinical symptoms including hyperpigmentation of the skin, hypertrichosis and lower limb oedema.

On examination, patient appeared wasted. He had exophthalmous, normotensive, with no cardiac or liver anomalies were detected. Investigations showed a HbA1c 10.8%. ESR 90 mmHg. Normal U/E, LFT. Genetic studies confirmed diagnosis of H syndrome with both parents being carriers.

Uncommon Cause of Diabetes; An Interesting Case

Background: Fibrocalculous Pancreatic Diabetes is a unique and severe form of diabetes mellitus which show insulin resistance, it occurs secondary to chronic non-alcoholic calcific pancreatitis. The exact etiology is obscure, but it is likely to be a multifactorial disease with both genetic and environmental components. **Case report:** 10 years old female, from west Sudan, presented with a 2-year history of steatorrhea and intermittent epigastric abdominal pain and weight loss. Two months following these symptoms, she developed polyuria, polydipsia and generalized fatigability. The condition was not preceded by trauma or infection. She had no significant past history and is on no medications. **Clinically:** She looked ill. Her BMI was below 3rd percentile. There was a bilateral parotid swelling, abdominal distention and a palpable liver. She had glucosuria and her HbA1c was 18%. She had high amylase and fat globules in stool. CT abdomen showed findings of chronic pancreatitis with scattered pancreatic calcification. **Conclusion:** Although rare, fibrocalculous pancreatic diabetes should be considered when both endocrine and exocrine pancreatic deficiency is seen.

An Uncommon Cause of Neonatal Diabetes

Areej Ahmed Bashier Ibrahim, Gafar Ibn Auf Tertiary Hospital, Khartoum, Sudan

Background: Neonatal diabetes mellitus with hypoinsulinism is an uncommon condition with an estimated incidence from 1 in 300,000 to 400,000 live births. Diabetes mellitus with hyperinsulinism due to congenital generalized lipodystrophy (CGL) presenting in the neonatal period is even rarer. Incidence of CGL varies between regions from 1 in 10,000,000 to 1 in 500 000. Individuals are insulin resistant and 25–35% develop diabetes mellitus between ages 15 and 20 years. **Case report:** A 4 month male presented with two weeks history of polyuria and documented hyperglycemia twice. Physical examination revealed an apparently muscular infant, absent subcutaneous fat more marked at buccal and gluteal areas, umbilical hernia, hepatosplenomegaly and macropenis. Investigations revealed hypertriglyceridemia and hepatic steatosis. Gene testing showed mutation at AGPAT2 gene in a homozygous status for the patient and a heterozygous for the parents. His diabetes was controlled by multiple daily injections of insulin at a dose of 2 units/kg/day. **Conclusion:** Neonatal diabetes with dysmorphic features and organomegaly raises the rare possibility of congenital lipodystrophy. This must lead one to consider genetic investigations and appropriate genetic counselling.

Munchausen Syndrome by Proxy

Mohammad Hosny, Mansoura University Children's Hospital, Egypt

Treatment of diabetes in young children is quite challenging. The caregiver plays an essential role in the course of treatment.

Munchausen syndrome by Proxy (MSBP) is the production or feigning of physical or psychological symptoms in another person, usually a child under the care of the individual with the disorder. MSBP is usually challenging and difficult to detect. We present a two years old girl with type 1 diabetes (T1DM) on multiple daily injection regimen. She presented with severe unexplained diabetic complications leading to difficult treatment. Despite prolonged admission and changing lines of treatment, her blood sugar was not controlled. Diagnosis of MSBP was suspected. Mother was confronted and admitted the inappropriate act.

Conclusion: In settings of treating T1DM in children MSBP is not common but it should be considered especially in cases which does not follow the usual course of the disease or not responding to usual treatment.

A Challenging Case of type 2 Diabetes Mellitus

Mohammed Helmy, Pediatric Endocrinology, Tanta University, Egypt

Background: The frequency of type 2 diabetes has markedly increased in the paediatric age group since the end of the 20th century. Metabolic syndrome is defined as a group of metabolically related risk factors that are usually associated with the condition of insulin resistance, systemic hypertension, and adiposity. **Case report:** A female child aged 13 years presented with nocturnal enuresis. Random blood glucose was 280 gm/dl with a fasting glucose of 269 mg. HbA1c was 13%. She was started on insulin therapy at a total daily dose of 1.2 u/kg/day. On examination, there was acanthosis nigricans, BMI>95th waist circumference also >95th, with average blood pressure for age. There was a strong family history of type 2 diabetes. The patient experienced repeated attacks of hypoglycaemia, and the dose of insulin was decreased gradually till 0.5 u/kg/day. Her fasting C was normal and Anti-GAD was negative. Diagnosis of type 2 diabetes was confirmed and she was started on Metformin. Gradually, insulin was withdrawn with instructions for weight reduction and exercise. **Conclusion:** Obese youth presenting with diabetes with rapid reduction of insulin requirement should be considered the possibility of having type 2 diabetes.

Autoimmune Polyendocrinopathy

Mona Hosny, Health Insurance National Authority, Alexandria Egypt

A 4 years old male child with diabetes presented with short stature and recurrent diarrhea. On examination, he had proportionate short stature (Ht, - 3.8 SD), abdominal distention. Investigation revealed microcytic hypochromic anemia. He had features of severe autoimmune thyroiditis with TSH 1151 IU/ ml, positive thyroid anti peroxidase antibodies. HbA1c was 7.1%. He was started on levothyroxine and continued the MDI of insulin.

On follow up of growth velocity, it remained poor despite the adequate glycemic control and the euthyroid state with treatment. Biopsy on upper gastrointestinal endoscopy confirmed diagnosis of celiac disease and he was commenced on gluten free diet. The child developed vitiligo and failed to gain linear height. Growth hormone provocation with clonidine was done which showed a growth hormone peak less than 7 ng/ml.

Autoimmune polyendocrinopathy type 3B (autoimmune thyroiditis, IDDM, celiac disease, vitiligo) with isolated growth hormone deficiency is a recognized form of polyendocrinopathy. It carries a management dilemma of the possibility of impairing diabetes control on growth hormone treatment.

A Case of Type 1 Diabetes with Hepatomegaly

Omneya Magdy, Pediatric Department, Alexandria University, Egypt

Background: Glycemic control is important in children with type 1 diabetes to prevent microvascular and macrovascular complications. Poor control of type 1 diabetes mellitus (T1DM) in adolescents may lead to Mauriac syndrome, which is characterized by dwarfism, obesity and hepatomegaly and elevated transaminases. Here we report a 12 years old boy with poorly controlled type 1 diabetes and hepatomegaly. **Case report:** 11.5 years old boy was diagnosed with type 1 diabetes since the age of 3. His diabetes control is poor. The patient was on basal bolus 1.2 unit/kg/day.

His height was height 134.5 cm (-1.8. SD). He had hepatomegaly with ALT of 128 U/L (up to 40), and AST of 146 U/L (up to 32). Serum triglycerides was 190 mg/dl (0-150).

Ultrasound examination revealed fatty change and liver biopsy revealed moderate glycogenesis and mild macrovascular steatosis. Mauriac syndrome is suspected. **Conclusion:** Mauriac syndrome is a rare complication of poorly controlled diabetes mellitus in adolescence, but it should be suspected when poor diabetes control is associated with poor growth, hepatomegaly and abnormal lipid profile.

Management Challenges in a Child with Hyperinsulinemic Hypoglycemia

Rana Abd El Hakim Ahmed, Children's hospital, Ain Shams University, Cairo, Egypt

Background: Hyperinsulinemic hypoglycemia is characterized by inappropriate secretion of insulin. Medical treatment may not be effective. Subtotal pancreatectomy is a resort to avoid neurological complications, but it carries the risk of developing diabetes mellitus or repeated surgical intervention. **Case report:** A 2 and ½ year old boy was referred with sweating and dizziness. He was the third child of non-consanguineous parents. His elder brother died at the age of four. He was growing at the 10th percentile for height and the 90th percentile for weight. His investigations revealed blood glucose of 21 mg/dl, serum insulin >320 µU/mL (normal 4-23) serum beta-hydroxybutyrate 0.1mmole/l (normal 0.02-0.2), serum GH 8.2 ng/ml and serum cortisol 2.7 ug/dl (6-16 a.m.).

Sequence analysis of the GCK, HADH, ABCC8 and KCNJ11 genes did not identify a mutation. Glucose infusion rate was 15 mg/kg/min but hypoglycemia persisted and was complicated by convulsions. Octereotide was started and increased up to 15 microgram/kg/6 hrs with failure to control hypoglycemia. Subtotal pancreatectomy was performed with initial stabilization, then hypoglycemia recurred necessitating IV glucose infusion with a glucose infusion rate at 18 mg/kg/min, SC Octereotide and IV hydrocortisone. Oral diazoxide was administered but failed to achieve normalization of blood glucose. Gastrostomy was done and enteral feeding commenced. Normoglycemia was achieved and he had a normal neurological development. **Conclusion:** Resistant hyperinsulinemic hypoglycemia presents a challenge to the treating physician. A number of unidentified mutations might be existing to warrant further genetic studies to explore new treatment strategies.

Macrocytic Anemia in a Newly-diagnosed 12-year-old Diabetic

Yasmeen Ashraf, Alexandria University, Egypt

Background: Thiamine-responsive megaloblastic anemia syndrome (TRMA) is an autosomal recessive disorder with features including megaloblastic anemia, sensorineural hearing loss (SNHL) and non-type 1 diabetes mellitus (DM). The disease onset is in infancy or early childhood. It is caused by mutations in SLC19A2 gene encoding a high-affinity thiamine transporter protein, which is responsible for effective utilization of thiamine in various tissues, including β-cells of pancreas. **Case report:** A 12-year-old female, born to consanguineous parents, presented with diabetic ketoacidosis with HbA1c 9.9%, low C-peptide. She improved on intravenous fluids and insulin. She was pale with weight and height at -2.5SD. She had no organomegaly, purpura, ecchymotic patches or arthritis. Investigations showed macrocytic anemia, thrombocytopenia, mild leucopenia (Hb 8.8 g/dl, MCV 122.8 fl, reticulocyte 0.8%). She was initially diagnosed with vitamin B12 deficiency but RBC folate and serum vitamin B12 levels were normal.

Her BM biopsy was hypocellular for age, with depressed megakaryopoiesis, megaloblastic erythropoiesis, depressed granulopoiesis, no abnormal infiltrates and adequate iron stores. Anti-GAD, anti-islet cell antibodies were negative. Her audiogram revealed high-frequency SNHL of left ear. Patient was diagnosed with TRMA and received a therapeutic trial with oral thiamine, upon which her Hb, platelets, WBCs, reticulocyte % improved, but still macrocytosis persisted (MCV 116.7fl). Her insulin requirements had also decreased. **Conclusion:** Audiogram, normal vitamin B12 and folate levels are the key to diagnosis of TRMA in any patient with DM and megaloblastic anemia. Treatment with pharmacological doses of thiamine

ameliorates megaloblastic anemia, increases reticulocytic count, but the red cells remain macrocytic. It can also improve DM. Therefore, confirms the diagnosis. Follow-up audiogram is yet recommended to detect if treatment prevents progression of SNHL or improves it.

A Mystery in a Diabetic Case in Pediatrics

Ahimaa Raafat Elsayed, Alexandria University Children Hospital, Alexandria, Egypt

Background: Patients with diabetes who develop cerebral disorders have been demonstrated at an electrophysiological, structural and cognitive level. However, the pathogenesis is still not clear. Episodes of hypoglycemia and poor metabolic control may be some of the factors affecting the cerebral function. Moreover, peripheral neuropathy is a common long-term complication of diabetes. However, there is a growing appreciation to the presence of peripheral neuropathy with the onset of diabetes or even in the pre-diabetes. Besides, Peripheral neuropathy is one of a long list of the causes of cachexia. **Case report:** We report a case of a rare presentation of diabetes associated with cerebral dysfunction, peripheral neuropathy and cachexia. 8.5 years old female child diagnosed with diabetes one month before referral to Alexandria University Children Hospital by uncontrolled diabetes. During this period, she developed abnormal behavior and cachexia. Tumor, an infectious, endocrinological or gastrointestinal disorders were excluded. CT brain raised the possibility of brain atrophy while MRI brain was unremarkable. Nerve conduction concluded that there was cerebral dysfunction besides peripheral neuropathy. **Conclusion:** Peripheral neuropathy and cerebral dysfunction might present acutely rather than as late complications of diabetes.

A Case of Type 1 Diabetes Complicated with Mauriac Syndrome

Dina Khedr, Diabetes Endocrine and Metabolism Pediatric Unit (DEMPU), Cairo University, Egypt

Background: Mauriac syndrome is still a complication of non-adherent children with type 1 diabetes mellitus. **Case report:** A 13 years old female, known with diabetes for 8 years. She had challenging social issues. She was treated with multiple daily injection of insulin and was provided with a comprehensive social support. Despite the attempts for diabetes control, she presented with growth failure, delayed puberty, hepatomegaly and dyslipidemia. She progressed to have autonomic neuropathy and limited joint mobility. **Conclusion:** In spite of the great advance in diabetes management, Mauriac syndrome still exists in real life particularly in those with poor glycemic control and adverse social circumstances. An outreach program targeting those children can be effective and is recommended.

Mauriac Syndrome after Islet Cell Transplantation

Marian Fares Nashed, Galaa Military Hospital, Cairo, Egypt

Background: Mauriac syndrome is characterized by dwarfism, obesity, hepatomegaly and elevated transaminases in patients with T1DM. **Case report:** A 12-year-old girl, known case of T1DM since the age of 5 years, presented with a complaint of distended abdomen and pain in right hypochondrium for 2 months. She has a history of islet cell transplantation 3 years ago, after which she was only on long acting insulin for 1 month then short acting insulin was added due to hyperglycemia.

Examination showed short stature (height 139 cm, -2.8SD), weight was 35 kg (-1.9 SD). Liver was palpable clinically 7 cm below costal margin, with no splenomegaly or free fluid. Extremities were diffusely atrophied with a positive prayer sign. She was prepubertal. Initial laboratory tests showed HbA1C 9%, C- Peptide \leq 0.1 ng/mL, AST 63 U/L, ALT 100 U/L, GGT

99 U/L. Abdominal ultrasound showed hepatomegaly with increased echogenicity. Marker for autoimmune hepatitis and viral hepatitis were negative.

Nerve conduction velocity showed right and left median nerve demyelinating mononeuropathy at wrist. Based on the clinical history and investigations, the final diagnosis of Mauriac syndrome was made and the patient was advised tight control of diabetes. She was switched to basal bolus insulin regime. She showed diabetes improvement and her hepatomegaly regressed. **Conclusion:** Despite modern insulin regimens and islet cell transplantation in children with type 1 diabetes, Mauriac syndrome still exists because the success of treatment is so dependent on patient compliance.

Diabetes Ketoacidosis Complicated by Cerebral Oedema

Rania Haddad, Elshatby hospital, Alexandria, Egypt

Background: Diabetic ketoacidosis is the most common cause of morbidity and mortality of Type 1 diabetes mellitus. Cerebral edema is one of its complications, so appropriate management and early decisions are needed to prevent this complication. **Case report:** A six years old boy presented to the emergency department with one week's history of vomiting, fatigue, polyurea, and polydipsia. On admission, he had a Glasgow comma scale of 8/15. Blood glucose was 450 mg/dl, pH of 6.82 and serum bicarbonate of 4.8 mmol/l. Urine dipstick for ketones was strongly positive. He was treated for severe diabetic ketoacidosis. Gradually, his comma scales fell to 3/15. He was admitted to the ICU, mechanically- ventilated and given intravenous mannitol 1 gram/kg. With the intensive treatment, he regained full consciousness and his metabolic parameters returned to normal levels. **Conclusion:** in DKA, rehydration fluid should not be overestimated. Initial management of cerebral oedema should include intravenous mannitol or hypertonic saline.

A Case Report of Klinefelter's Syndrome with Type 2 Diabetes

Sakina kherra, CHU-H DEY Hospital Pr-N Hamoud, Algeria

Background: Klinefelter's syndrome is the commonest form of sex chromosome aneuploidy as well as the most common chromosomal cause of male infertility. Typically, patients are tall and slim, with gynaecomastia, small testes and hypergonadotrophic hypogonadism. Recently a new phenotype has been described with increased risk of metabolic syndrome and diabetes. We report the case of Klinefelter's syndrome in an adolescent with type 2 diabetes associated with insulin resistance. **Case report:** A nine years old child was referred to our department for evaluation of obesity. He has learning disability as does a first cousin, and the latter was also treated for central precocious puberty. Aged 12.8 years, physical examination revealed height 170 cm (+2.12SD). He was noted to have long arms and legs, with gynaecomastia, testes 2 ml volume, penis 4 cm in length. Acanthosis nigricans was present located in the neck area.

Chromosome analysis showed a 47,XXY karyotype. Hormonal profile showed hypergonadotrophic hypogonadism with LH of 14 UI/L (<4.6 UI/L), FSH of 30 UI/L (<3.6 UI/L) and testosterone of 0.1 ng/L (0.2–0.8). Fasting plasma glucose was elevated at 2.3 g/l (<1.24 g/L), HbA1C 8% (<6%), insulin 50 μ UI/ml (<15). GAD antibodies were negative.

Klinefelter's syndrome with hypogonadism, obesity and type 2 diabetes was diagnosed and treatment was started with metformin and insulin. **Discussion:** The mechanism of diabetes in this syndrome is related to obesity compounded by the hypogonadism of Klinefelter's syndrome. Low levels of testosterone may be predictive of future central adiposity and metabolic syndrome.

Wolfram Syndrome: An Interesting Case Scenario

Ahlem Laabed, Department of Pediatrics, University of Batna, Algeria

Background: Wolfram syndrome is a rare neurodegenerative genetic disorder. Patients present with diabetes followed by optic atrophy in the first decade. Patients progress to develop diabetes insipidus, deafness, urinary tract, and neurological abnormalities. **Case report:** We report an Algerian 13-year-old female patient, born to a consanguineous marriage, who acquired diabetes mellitus at the age of 2 years and has been receiving insulin since then. She has suffered progressive visual deterioration since the age of 7 years. Fundus examination showed pale disc in both eyes, indicative of optic atrophy. There was no evidence of diabetic retinopathy. Over a year, she presented with nocturia and high urine output (8 liter per day) with later development of incontinence and voiding difficulty. Renal sonography showed pelvicalyceal dilatation in both kidneys and bladder enlargement.

Hypothalamic pituitary MRI was showed absent posterior pituitary signal. Treatment with desmopressin was started. With the combination of symptoms, wolfram syndrome was suspected. Molecular analysis of WFS1 gene showed that she is homozygous of pathogenic variant (c.1775_1776delTG(p.Leu592Tyrfs*13) in WFS1 gene.

Conclusion: Wolfram syndrome is a rare neurodegenerative disorder which should be suspected in patients with diabetes mellitus and optic atrophy. Genetic analysis is useful to confirm the diagnosis.

Diabetes Mellitus and Other Gastrointestinal Autoimmune Association

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Background: Type 1 diabetes is well known autoimmune disorder and is frequently associated to other autoimmune conditions. **Case presentation:** We report a young male teenager who was diagnosed with type 1 diabetes from 7 years of age. He developed refractory diarrhea and was diagnosed with Celiac disease and commenced on gluten free diet. His gastrointestinal symptoms persisted and developed persistent dyspepsia and melena. An upper gastrointestinal fibroscopy and biopsy confirmed the diagnosis of Crohn's disease. HLA typing showed related loci of HLA DQ 2 and HLA DR4. **Conclusion:** Multiple autoimmune diseases can be associated with diabetes. HLA subtyping confirm the shared autoimmune link.

Insulin Resistance in an Adolescent Girl

Kiran Choudhry, Paediatric Endocrinology Department, Tawam hospital, Al Ain, UAE

Background: The prevalence of type 2 diabetes mellitus is increasing in adolescents around the world in all ethnicities. **Case Report:** 13 years old female with irregular periods and candida vaginitis was found to have 4+glycosuria, no ketonuria. Fasting blood sugar 128, High insulin and c-peptide, HbA1c 9.8% with mixed dyslipidemia. Diabetes antibodies were negative. Positive family history for metabolic syndrome. Weight gain of 7 kg in the last 1 ½ year. Only other symptoms were fatigue and nocturia. She was started on long acting insulin at 0.6 units/kg/hour. Later on, sliding scale with meals was added. She was started on meal plans based on 1800 Calorie diet with lower carbs (33% vs 45%). She was also started on an exercise regimen of brisk walking 45 minutes per day. The HbA1c dropped to 8.2% at 6 weeks. At that time Metformin was added and weaning of insulin started. Four months after the diagnosis she was weaned off insulin and maintained on Metformin only. 1 year after the diagnosis she had lost 7% of her initial weight, HbA1c was 5.4% on low dose Metformin and continued healthier lifestyle. Lipid profile normalized without intervention. **Conclusion:** Increased insulin resistance due to puberty hormones obesity and poor life style choices is increasing the rates of type 2 diabetes in children. Many of these children can be managed without insulin with vigorous life style modification and oral medications.

Insulin Resistance and Type 2 Diabetes in an Adolescent

Dr Deepti Chaturvedi, Burjeel hospital, Abu Dhabi, UAE

Background: Insulin resistance and type 2 Diabetes is becoming increasingly common in childhood and adolescence. Disease evolution can be rapid with multiple challenges associated with its management. **Case report:** A 12 year old girl, Bangladeshi in ethnicity, presented with primary concern of progressive weight gain and body aches since last 4 years. Mother had Type 2 Diabetes following diagnosis of gestational diabetes. Her BMI was 24.36 kg/m² at >>97th centile. There was accompanying severe acanthosis and striae around the abdominal area.

Investigations showed a HbA1C of 6.6% with a high serum insulin levels of 999.20 pmol/L (17.8-173). Glucose tolerance test revealed a fasting serum glucose of 5.27 mmol/L and 2 hours post prandial of 13.38 H mmol/L. Lipid profile showed high levels of total Cholesterol 5.20 mmol/L, serum triglycerides 1.85 H mmol/L and serum LDL cholesterol 3.49 mmol/L. Hormonal profile showed high Thyroid Stimulating Hormone of 10.48 H mIU/L (0.6 - 4.84) with normal FT4-6.12 pmol/L. Serum SGPT was high at 60.90 H U/L and ultrasound scan of abdomen showed fatty liver changes. Thus, the patient was diagnosed with type 2 Diabetes with hypothyroidism, mixed hyperlipidemia with non -alcoholic fatty liver disease. Appropriate medical management including metformin and thyroxin was started. Patient was advised lifestyle changes in form of diet and exercise. **Conclusion:** In the present milieu of increasing obesity and insulin resistance in children and adolescents, it is imperative to diagnose Type 2 diabetes proactively and manage these cases appropriately.

Chronic Inflammatory Demyelinating Polyneuropathy in Type 1 Diabetes

Omer Ahmed Omer, National Diabetes and Endocrine Centre, Royal Hospital, Muscat, Oman

Introduction: Diabetic neuropathy is a prevalent chronic complication of diabetes mellitus. The term diabetic neuropathy is normally used to refer to the forms of neuropathy that are most prevalent among patients with diabetes, which are distal symmetric polyneuropathy (DSPN) and diabetic autonomic neuropathies. However, not all patients with diabetes and neuropathy symptoms have these typical diabetic neuropathies and patients with diabetes can develop inflammatory neuropathies. Inflammatory neuropathies include diabetic radiculo-plexus neuropathies and vasculitic multiple mononeuropathies, but the most common and most treatable is chronic inflammatory demyelinating polyneuropathy (CIDP). **Case:** A 13 years old boy who was diagnosed to have type 1 diabetes March 2015 with positive anti GAD and anti ICA antibodies. He was on regular follow up with poor glycemic control. He was recently noted to have progressive weakness of upper and lower limbs associated with pain and wasting of the limbs. This was initially thought to be due peripheral neuropathy related to glycemic control. However, the short duration of diabetic history in this patient was unusual, so other causes of neuropathies were considered. Nerve conduction studies suggested demyelinating polyneuropathy. Lumbar puncture was done and CSF showed slight elevation of protein with normal white blood cells and glucose level. Treatment with intravenous immunoglobulin was commenced with marked improvement. **Conclusion:** In patients with type 1 diabetes presenting with peripheral neuropathy, chronic inflammatory demyelinating polyneuropathy needs to be considered as it is treatable condition.

Severe Hypertriglyceridemia Causing Severe Acute Pancreatitis in a Child with Type 1 Diabetes Presenting in Ketoacidosis

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Introduction: Poorly controlled diabetes as a cause of acute pancreatitis has been reported in patients with type 1 diabetes. Severe hypertriglyceridemia due to excessive lipolysis from lack of insulin effect during diabetic ketoacidosis can

trigger acute pancreatitis, which has been reported rarely in children. **Case presentation:** A 12-year-old girl, followed for 1 year for type 1 diabetes, presented for vomiting and abdominal pain associated with diabetic ketoacidosis. On admission, she was dehydrated and had tachypnea. She had an epigastric abdominal and left hypochondrial pain. Her arterial blood gas measurements showed a pH of 7.26, PCO₂ 26 mm/Hg and bicarbonate 10 mmol/L. Her serum sodium was 127 mmol/L, potassium 3.1 mmol/L and glucose 12 mmol/L. Urinalysis showed 3+ glucose and 4+. She was treated intensively for diabetes ketoacidosis. Her abdominal pain worsened. Abdominal CT scan showed signs of severe acute pancreatic with necrosis. Acute pancreatitis secondary to hypertriglyceridemia associated with the severe insulin deficiency was suspected. The serum triglyceride value was 64 mmol/L, total cholesterol 13 mmol/L. A serum lipase value was measured and found to be 414 units/L (reference range 6–51 units/L). The child was put on low-fat enteral feeds after a digestive rest for 4 days and she was given Xylocaine infusions for 5 days. Her abdominal pain resolved after 10 days. **Conclusion:** The recognition of acute pancreatitis in any patient with ketoacidosis has important implications in the management of the patient as insulin requirements and the tempo of recovery can be altered. In addition, a higher suspicion for sequelae of pancreatitis, such as necrosis or pseudocyst formation is needed if symptoms are slow to resolve or the pancreatitis is severe.

Diabetes Challenging Diagnosis

Sinan Abuleil, Hadassah University Hospital, Palestine

A 12 y old female presented with clinical picture of diabetic ketoacidosis after a history of polyuria, polydipsia and weight loss. She was treated for DKA and commenced on basal bolus regime. Mother had gestational diabetes and grandmother had diabetes on insulin since the age of 35y of age. HbA_{1c} was 14.5% and Anti GAD negative. She improved dramatically on insulin treatment and required minimal doses of insulin. Considering high BMI and lack of antibodies, the possibility of T2DM has been raised. OGTT and C peptide were done with results more supportive of type 2. She was commenced on metformin with significant improvement in her condition.

Differentiating T1DM & T2DM is not an easy task always, especially in Anti GAD negative patients, so combining clinical presentation, physical examination, blood tests and regular follow-up can guide you to the right diagnosis.

Neonatal Diabetes and GLIS3 Gene Mutation

Dawoud Ayyad, Jerusalem, Makassad Hospital, Palestine

Background: Neonatal diabetes mellitus with congenital hypothyroidism has been reported. Other association of congenital glaucoma, cholestasis, hepatic fibrosis and polycystic kidneys can also be seen. **Case report:** A female neonate was delivered by section at 35 weeks had a birth weight of 1850 gm. She was admitted to NICU with respiratory distress. She was found to have direct hyperbilirubinemia, elevated liver enzymes, anemia, thrombocytopenia, fluctuated hyperglycemia, and hypothyroidism. Extensive investigations confirmed cholestasis, hypothyroidism, and hyperglycemia responding to insulin. GLIS3 gene abnormality was suspected, and was confirmed by whole exome sequencing. **Conclusions:** GLIS3 mutation should be suspected when neonatal diabetes is diagnosed with multiple hematological and gastrointestinal associations. Genetic diagnosis confirms the diagnosis.

Donohue syndrome

Aseel Hamarshi, Makassad Hospital, Jerusalem, Palestine

Background: Diabetes mellitus due to insulin resistance, apart from type 2 diabetes, is rare. Mutation of insulin receptor is a genetic disorder that results in leberechism. Donohue syndrome is an extremely rare autosomal recessive genetic

disorder resulting from homozygous mutations in the insulin receptor gene. Donohue syndrome is associated with a fatal congenital form of dwarfism with features of intrauterine and postnatal growth retardation. It is characterized by fasting hypoglycemia and postprandial exaggerated hyperglycemia with hyperinsulinism and dysmorphic features. **Case report:** We present a case of Donohue Syndrome encountered in a female born to consanguineous parents. She was born at 37 weeks by elective section due to severe intrauterine growth restriction. The patient was admitted to the Neonatal Intensive Care Unit due to low birth weight. She was noted to have dysmorphic features, abdominal distension, poor weight gain and hypertrichosis. Laboratory examinations showed fasting hypoglycemia, postprandial persistent hyperglycemia and anemia. The diagnosis of Donohue syndrome was diagnosis based on the clinical features. Genetic diagnosis confirmed the presence of homozygous mutations in the insulin receptor gene. Treatment of Donohue Syndrome is supportive and requires the combined efforts of a multidisciplinary team. **Conclusion:** There is a scarce genetic information on Donohue syndrome among the Arab population. Consanguinity is one of underlying reasons for the appearance of rare this genetic disorder. Genetic counseling is important in such families.

Cerebral Edema During Treatment of Diabetic Ketoacidosis: A case report

Abderrazaq Abu Mayaleh, Pediatric department, Palestine Red Crescent Society Hospital, Hebron, Palestine

Background: Cerebral edema remains the leading cause of death in children with Type 1 diabetes mellitus. It occurs in 0.5-1% of paediatric DKA episodes, and one-quarter of those children will die from it. The causes of cerebral edema are still poorly understood. Possible contributing factors may be younger age, new-onset diabetes, high glucose levels, severe dehydration, severe acidosis, and treatment with bicarbonate. Furthermore, cerebral edema can still occur even when the management of DKA follows current 'best practice' guidelines. **Case report:** A 10 years old boy, known case of type 1 DM since age of 5 years presented acutely ill with abdominal pain, recurrent vomiting and hypoactivity. The patient is known for poor compliance and poor diabetes control. He had Kussmaul breathing but was hemodynamically stable. Blood glucose was 615 mg/dl, blood gas showed severe metabolic acidosis (PH=7.06, CO₂=21, HCO₃=6), Urea and creatinine were elevated, 62 mg/dl and 1.3 mg/dl, respectively. He was treated with intravenous fluids and insulin infusion. Deterioration in the patient's level of consciousness (GCS 6/15) occurred 12 hours after commencing management. Brain CT showed mild cerebral edema, so treated with I.V Mannitol twice and hypertonic saline 3%. Fluids were changed to normal saline instead of half normal saline. The patient regained his full consciousness and his DKA resolved 16 hours after the diagnosis and starting treatment of cerebral edema. **Conclusions:** Awareness and early intervention in treating children with suspected clinical cerebral edema during DKA management are crucial to prevent morbidity and mortality of such a devastating complication.

Growth Deceleration as a Red Flag of Celiac Disease in a Child with Diabetes

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Background: Celiac Disease (CD) occurs in patients with Type 1 Diabetes (T1D) with a prevalence of 4.4-11.1% versus 0.5% of the general population. The mechanism of association of these two diseases involves a shared genetic background: HLA genotype DR3-DQ2 and DR4-DQ8 are strongly associated with T1D, DR3-DQ2 with CD. The classical severe presentation of CD rarely occurs in T1D patients, but more often patients have few/mild symptoms of CD or are completely asymptomatic (silent CD). **Case report:** A 5½ year old boy, diagnosed to have diabetes mellitus since age of 2 years. Anti-GAD antibodies at time of diagnosis were negative. On routine physical exam, a heart murmur was detected and echocardiography revealed interrupted aortic arch for which he underwent a cardiac surgery one month after diagnosis of diabetes. Screening tests for thyroid function and celiac antibodies were performed annually since time of diagnosis of DM as

a routine and were normal. Growth assessment was documented in each visit to the clinic and remained within normal till the age of five, when his growth velocity started to decelerate (~3 cm/y). Therefore, a new anti-tissue transglutaminase antibodies levels were done and came highly positive. Upper endoscopy and duodenal biopsy were performed and confirmed severe villous atrophy consistent with severe CD. **Conclusions:** it is important to consider celiac disease as a cause of growth deceleration in asymptomatic children with type 1 diabetes.

Diabetes Mellitus and Muscular Dystrophy

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Background: Common metabolic and endocrine alterations exist across a wide range of muscular dystrophies. Skeletal muscle plays an important role in glucose metabolism and is a major contributor in different signaling pathways. Therefore, its damage may lead to different metabolic disruptions, Two of the most important metabolic alterations in muscular dystrophies maybe insulin resistance and obesity. **Case Report:** An 8-years old female patient, who is a known case of muscular dystrophy, presented with polyuria, polydipsia and polyphagia. Clinical examination revealed positive Gower sign and proximal muscle weakness. Lab tests revealed HbA1c of 8.7%. Random blood glucose was 23.6 mmol/L. Her autoantibodies were negative. She was started on basal bolus regime of insulin. Her insulin requirement came down rapidly and she was shifted to metformin. Her glycemic control remained good on Metformin. **Conclusion:** Diabetes mellitus with features suggestive of type 2 diabetes can be seen in association of muscular dystrophy in the absence of obesity.

A novel mutation in the Pancreatic duodenal homeobox-1(PDX-1) gene in a Palestinian family resulting in Neonatal Diabetes

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Background: PDX-1 gene is involved in the early development of the pancreas and plays a major role in glucose-dependent regulation of insulin gene expression. Defects in this gene causes pancreatic agenesis, which can lead to early-onset insulin-dependent diabetes mellitus. **Clinical data:** A 1-day-old male newborn, admitted to NICU due to antenatal diagnosis of duodenal atresia, polyhydramnios and IUGR. Laparotomy revealed duodenal web, resection was done with duodeno-duodenal anastomosis. He was noticed to have hyperglycemia since admission, C-peptide: <0.02, Insulin level <0.5, normal thyroid function tests, abdomen CT was suggestive of dorsal pancreatic agenesis. **Molecular data:** DNA sequencing of the PDX-1 gene for the patient revealed a novel homozygous mutation Leu166Pro in exon 2 of the PDX1 gene. Parents were heterozygous for the same mutation. **Conclusion:** Congenital absence of the pancreas is an extremely rare condition. To our knowledge, this is the first description of this disease in a Palestinian family. Molecular confirmation allows accurate genetic counseling, early diagnosis of affected kindreds, early therapeutic interventions and avoiding complications.

Severe Diabetic Ketoacidosis with Distal Hypoperfusion: Case Report

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Background: Ketoacidosis is the most frequent acute metabolic complication of mellitus diabetes. **Case report:** A 15 years old girl presented with diabetes ketoacidosis. She was admitted for management and was commenced on intravenous fluid and insulin infusion. Her course was complicated with urinary infection and was treated by antibiotics. On the fourth day of admission, she was noted to have a cyanosis of toes. After treatment of the infection and normalization of hyperglycemia, the cyanosis of the toes disappeared. The hypoperfusion due to the severe ketoacidosis may explain the distal cyanosis. **Conclusion:** The distal hypoperfusion of extremities is a rare complication of severe diabetic ketoacidosis.

DEND syndrome A Moroccan case

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Background: DEND syndrome, defined as developmental delay with epilepsy, muscle weakness and neonatal diabetes is rare. Many mutations have been described in the KCNJ11 encoding Kir 6.2 and ABCC8 encoding SUR1. This ABCC8 mutation can account for both neonatal diabetes and the neurological phenotype. Sulfonylurea (Glibenclamide) treatment led to both improved glucose homeostasis and an increase in mental and motor function. Early recognition of patients with DEND syndrome may have considerable therapeutic benefit for the patient. **Case report:** We report a case of a full-term male infant with neonatal diabetes mellitus born to consanguineous parents. The baby was born at term with a weight of 3200g. The patient presented on the 15th days with severe dehydration and a tonic clonic seizure. He had metabolic acidosis, glucosuria and ketonuria. The diagnosis of neonatal diabetes mellitus was made, and we started insulin on a 0.4 units/kg/day dose. Investigation showed c-peptide of 0.26 ug/l and a negative anti-GAD antibodies. Molecular biology showed ABCC8 mutation. Neurologically, the patient has epilepsy treated with 25 mg of vigabatrin, delayed psychomotor acquisitions and strabismus. The evolution was marked by a gradual decrease in his insulin requirements with the introduction of Glibenclamide at a 6 months age on a dose of 0.5 mg/kg/day. After 20 months of treatment, glycemic control is satisfactory with a 6.1% A1C without severe hypoglycemia. Moreover, the patient is well controlled by vigabatrin with disappearance of seizures. **Conclusion:** The most severe clinical form of permanent neonatal diabetes mellitus presents as Developmental delay, Epilepsy and Neonatal Diabetes (DEND) syndrome. Diagnosis is confirmed by genetic mutation testing. Oral sulfonylurea therapy improves neurological outcome.

Unusual Presentation of B-ketothiolase Deficiency Mimicking Type 1 Diabetes Mellitus: Case Report

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Background: Beta-ketothiolase (BKT) deficiency is a disorder of ketone body metabolism and isoleucine catabolism. It is a rare autosomal recessive condition, estimated to affect 1 in 1 million newborns. Patients with BKT deficiency have intermittent ketoacidotic attacks and are usually asymptomatic between episodes. **Clinical case:** our patient is a male presented at age of one year with clinical and biochemical evidence of diabetes ketoacidosis, blood glucose level 22 mmol/l. He was managed according to DKA guidelines and started on multi-dose insulin regimen insulin at an initial dose 0.5 u/kg/day. Insulin requirements decline over few weeks to 0.3 u/kg/day and due to normalization of blood glucose, insulin therapy was stopped. Afterward, the patient, had two further episodes of DKA requiring insulin with a short period of insulin requirement after the events. In between DKA attacks blood glucose level range from 4.4-8.8 mmol/l, occasional episodes of hyperglycemia especially with periods of stress reaches 15 mmol/l and managed with one to two units of short-acting insulin. At two years of age, he presented with ketoacidosis and hypoglycemia, blood glucose level 2.2 mmol/l. The unusual presentation of hyperglycemia and hypoglycaemia accompanied with ketoacidosis prompted genetic testing. Biochemical testing revealed increased urinary 2-methyl-3-hydroxybutyric acid which is consistent with BKT deficiency diagnosis and, genetic testing revealed a novel homozygous mutation in ACAT1 gene c.592G>A p.(Glu198Lys). The patient was advised to avoid prolonged fasting and started on a low protein diet. Since then he had developed mild episodes of ketosis with illness required intravenous hydration. **Conclusion:** mild hyperglycaemia, reaching up to 14.1 mmol/L, not requiring insulin treatment has been reported in patients with BKT deficiency. Our patient presented with significant hyperglycemia 22 mmol/l mimicking type 1 diabetes and required insulin treatment intermittently and treated as diabetic ketoacidosis, which represents unusual presentation for patients with BKT deficiency.

Insulin Resistance Syndrome with Precocious Puberty; An Interesting Case Report

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Insulin resistance defined as state of a tissue in which greater than normal amount of insulin is required to elicit quantitatively normal response. Pathogenic variant in INSR gene are the most common cause of monogenic insulin resistance, we prescribe a 9 years old girl with class 1 pathogenic variant INSR and precious puberty with family history of same gene mutations

Case report: A nine years old girl presented with excessive body hair, acanthosis nigricans in the neck, body dryness increased gradually since five year. She had history of symptomatic hypoglycemia. There is a positive family history of consanguineous marriage. Three cousins had the same condition. On Physical examination she has Subtle dysmorphic features (coarse facial feature, small upper lips, long philtrum, prominent cheek and nose). Sever acanthosis nigricans in the axilla bilaterally, knees, elbows and neck. She had dry skin with hypertrichosis and loss of subcutaneous fat. Her weight and height 50th centile with normal growth velocity and was at Tanner stage 3 for puberty. Investigations showed an insulin level range 148-114 mIU/l, HbA1C 5.6%. LHRH test showed pubertal range. Genetic diagnosis whole exome sequencing was performed and showed INSR(NM_000208.2, sequencing /homozygous variant c.433C >Tp.(arg145Cys). The genetic diagnosis of INSR related insulin resistance was confirmed. **Conclusions:** Insulin resistance syndrome is a genetic disorder, the treatment of which still remains unsatisfactory. Our plan to start subcutaneous leptin therapy, which was tried in few reported cases and promises good results. Newer research targeting gene therapy may help improve the clinical outcome in these patients.

MODY4 in an 8-year-old Saudi Girl

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Background: Maturity onset diabetes of the young (MODY) is autosomal dominantly inherited diabetes that, despite a young age of onset, is not insulin dependent. It results from B-cell dysfunction rather than insulin resistance. It is rare, accounting for just 1–2% of all diabetes. It is often misdiagnosed as type 1 or type 2 diabetes. MODY 4 is a Mutation of insulin promoter factor-1 (IPF-1) also known PDX1, first defined in 1997 and is a very rare cause of MODY. **Case report:** Our patient is an 8-year-old boy. He was diagnosed as T1DM 3 years ago and started on basal bolus regime of insulin. His diabetes was poorly-controlled but he was sensitive to insulin and had multiple episodes of hypoglycemia following small doses of insulin.

Clinical examination confirmed that no specific dysmorphic features, patient's height was between 10th-25th percentile, weight on 25th percentile and his BMI was between 10th-25th percentile, there was no acanthosis nigricans. His lab results showed HbA1C was ranging from 13.6 to 16.1, Celiac screening was negative, Islet cells and GAD AB were negative. Genetic study showed heterozygous mutation for INSR related type of diabetes. **Conclusion:** Confirming the diagnosis of monogenic diabetes enables the most appropriate treatment for them can be determined. As it is familial in origin, it is important to advise other family members of its risk of inheritance.

Acute Kidney Injury Association with DKA

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Background: Diabetic ketoacidosis (DKA) occurs in 10 to 70% of children with type 1 diabetes mellitus (DM1) across different study populations. It has a significant risk of mortality, mostly due to cerebral edema. Acute kidney injury (AKI) is a common event in hospitalized children and implies a sudden worsening of the kidney's ability to function. The clinical manifestations of pediatric AKI range from a mild increase in serum creatinine to anuric renal failure that requires dialysis.

The most common risk factor for pediatric AKI is prerenal disease or volume-responsive AKI, which caused by hypovolemia and reduced renal perfusion. **Case report:** A 13 years old boy known case of DM1 for 1 year presented to emergency with history of vomiting for 1 day after missed insulin dose in last night. He is on multiple daily injection of insulin at 1.1 units/kg/day. His diabetes is poorly-uncontrolled diabetes with high level of HbA1C (8.5 – 10%). He presented with severe dehydration and severe hyperglycemia. He was anuric and his venous pH was 7.22 with bicarbonate level of 12.6. Management of DKA initiated. Initial labs work showed normal CBC, electrolyte. In addition, BUN 7.6 mmol/L, Creatinine 116 umol/L and HbA1C 11.6%. He has recovered from the DKA but his renal function remained impaired with a BUN 9.9 mmol/L, creatinine 128 umol/L and Microalbumin/ Creatinine ratio 1.2. On intensified management, renal function normalized on day 3 and patient was discharged home. **Conclusion:** Severe AKI was associated with worsening markers of volume depletion and acidosis. Clinicians should consider AKI as a frequent complication that accompanies pediatric DKA.

MODY Type 4 in Pediatric Patient Presentation and Management

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Background: MODY is a clinically heterogeneous autosomal dominantly inherited diabetes that, despite a young age of onset, it is not insulin dependent. It results from B-cell dysfunction rather than insulin resistance. **Case report:** 4 years old Saudi boy presented with high blood glucose. He had no osmotic symptoms or weight loss. There was a strong family history of diabetes. On examination, he had no dysmorphic features and had a normal growth parameters. Investigations showed a HbA1c of 6.5% and a negative anti GAD antibodies. Genetic study revealed a positive heterozygous variant on PDX1 gene. Family screening was done and showed the same gene defect. The patient started on insulin then shifted to sulfonylurea. **Conclusion:** MODY 4 is rare in paediatrics. Molecular diagnosis of monogenic diabetes alters management and identifies affected and at-risk family members. Thus, genetic testing should be pursued in all patients meeting a clinical diagnosis of maturity-onset diabetes of the young.

Extensive Management of Generalized Lipoatrophy in Type 1 DM

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We report a 9 years old boy who presented with multiple lipoatrophy at insulin injection sites. He was diagnosed with diabetes at the age of 6 years. His lipoatrophy was noted early since diagnosis and did not improve with changing the insulin type. Resting the injection site improved the lipoatrophy but only slightly. Use of injection through an iPort over buttocks showed no lipotrophy and subsequently improvement in glucose profile. Examination showed severe lipoatrophy in the arms, abdomen and thighs.

Investigations showed positive insulin auto antibodies (IAA) 973.3 U/ml (normal <0.4). His HbA1C was 9.3% initially but fell down to 8.3 on the use of iPort. Later on, Dexamethasone at 0.04 mg was injected subcutaneously at the hypotrophied area with satisfactory results. **Conclusion:** use of subcutaneous cannula might be helpful in preventing lipoatrophy in susceptible patients.

Monogenic Obesity; ROHHAD Syndrome as an Example

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Introduction: ROHHAD syndrome (rapid-onset obesity with hypothalamic dysregulation, hypoventilation, and autonomic dysregulation) is a rare and complex disease, presenting in previously healthy children at the preschool age. Up to 40% of cases are associated with neural crest tumours. **Case report:** we report a 5-year-old boy with symptoms of rapidly

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progressing obesity. Few months of the rapid weight gain, he developed hypothalamic dysfunction (central hypothyroidism, hyperprolactinemia) with severe electrolyte imbalance, hypoventilation followed by cardiopulmonary arrest that required resuscitation and intubation at a time) and severe autonomic dysregulation (unexplained fever, recurrent episodes of diarrhea alternating with constipation). He was diagnosed with ROHHAD syndrome based on the classical clinical presentation. Although the pathophysiology of this syndrome remains unclear, the medical associations and complications are described in the literature and need to be anticipated and screened for at early stage. At present there is no genetic testing available to diagnose ROHHAD, so the diagnosis is based on the clinical presentation and clinical course which should include collaborative consultation by experts in a multidisciplinary approach. **Conclusion:** Multidisciplinary care is crucial to the successful management of these patients. Ideally, this team needs to include primary care physicians, pulmonologists, endocrinologists, cardiologists, intensivists, otolaryngologists, surgeons, gastroenterologists, neurologists, ophthalmologists, psychologists, and respiratory therapists.



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