

## ABSTRACTS BOOK

**Second ASPED/ISPAD Diabetes Postgraduate Course;  
12<sup>th</sup>-14<sup>th</sup> May, 2016, Abu Dhabi, United Arab Emirates.****Guest Editors: Nancy El Barbary<sup>1</sup>, Carine DeBeaufort<sup>2</sup>, Asma Deeb<sup>3</sup>**

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**Abstract**

The second ASPED/ISPAD Diabetes postgraduate course is an intensive course initiated by the Arab Society of Paediatric Endocrinology and Diabetes (ASPED) in collaboration with the International Society of Pediatric and Adolescence Diabetes (ISPAD). The course is exclusively sponsored by Lilly, Gulf. The course was held in Abu Dhabi over 3 days. It was run by an expert group of faculty from 12 different countries. The faculty group consisted of 4 speakers from ISPAD, 13 speakers and mentors from ASPED. Candidates were selected following open competitive applications which was advertised by both ASPED and ISPAD. Strict enrolment criteria were agreed on by the ASPED/ISPAD course committee. 64 candidates (out of 142 applicants) from 12 countries were enrolled and attended the course. Course curriculum was delivered in the form of lectures, interactive sessions, case scenarios and research presentation in small group sessions. The themes of the curriculum featured; comprehensive diabetes care,

diabetes registry (international and regional experience), dietary challenges, monogenic diabetes (international and regional experience), diabetes management in special circumstances (school, sick days and Ramadan fasting), diabetes complications; acute and chronic, psychology and patient empowerment, obesity and type 2 diabetes in children, novel therapeutic approaches and diabetes prevention and use of technology in diabetes management. The course ended with tasks to follow and recommendations. A task force of 7 members from 6 countries was nominated to initiate and follow up on the execution of the tasks. The main task was to start building up an ASPED diabetes registry amongst different Arab countries. Plans regarding curriculum, venue and course format for future annual courses will be implemented based on faculty and attendees feedback reports.

**Key words:** Diabetes, Thyroid, Pituitary, Growth, Pediatric, ASPED, ISPAD.

## I. Executive Report

### Context

Diabetes is increasing globally, the rates of diabetes, both type 1 and type 2, are escalating rapidly around the world at alarming rates. Most of the children and adolescents with diabetes develop type 1 (autoimmune) diabetes but increasing numbers in the developed world present with type 2 (non-autoimmune) diabetes often associated with obesity. Training physicians to take care of this increasing population is one of the aims of the scientific societies. The second ASPED/ISPAD Diabetes postgraduate course is an intensive course initiated by the Arab Society of Pediatric Endocrinology and Diabetes (ASPED) in collaboration with the International Society of Pediatric and Adolescence Diabetes (ISPAD). The course is exclusively sponsored by Lilly, Gulf.

### The Arab Society for Pediatric Endocrinology and Diabetes (ASPED)

ASPED was launched in Abu Dhabi, United Arab Emirates in September 2012 upon the initiative of a group of Pediatric Endocrinologists from the Middle East and North Africa. The society is a non-profit scientific organization and is registered under the Dubai Association Center (DAC) under License number DAC-0001. Its aim is to ensure a high standard of care and development in the field of pediatric endocrinology and diabetes in the Arab region extending from the Gulf through the Northern African countries.

### The International Society for Pediatric and Adolescent Diabetes (ISPAD)

ISPAD is a professional organization that brings together doctors, nurses, dieticians, psychologists, scientists and other professionals who are driven to improve the well-being of children and families afflicted by diabetes throughout the world. ISPAD accomplishes this by improving understanding of the etiology and epidemiology of diabetes, and providing education to physicians and other health care professionals as to the proper care of children and adolescents with diabetes, as well as developing guidelines for appropriate diabetes care.

ISPAD is the only worldwide organization concentrating on all aspects of diabetes in children, adolescents and young adults focusing upon the underlying science and quality of care that these young people receive. It has

published, and updates every 4-5 years, the ISPAD Clinical Practice Consensus Guidelines (Pediatric Diabetes 2014,15(S20) freely accessible. Currently an e-learning program is developed based on these guidelines. Membership offers -among others- free access to the Journal of the Society: Pediatric Diabetes (IF 3.488), the possibility to apply for participation in healthcare professional or physician science schools, an annual Conference and participation in the SWEET Registry. A continuous open discussion on cases and other topics is accessible through the Forum on the ISPAD website ([www.ispad.org](http://www.ispad.org)).

### The second ASPED/ISPAD Diabetes Postgraduate Course

The course was held at the Sofitel hotel, Abu Dhabi between the 12<sup>th</sup> and the 14<sup>th</sup> of May 2016. It was an initiative from ASPED in collaboration with ISPAD. It is a continuation from the first course of June 2015 and is aiming to empower and update physicians practicing in the Arab countries who are involved in the care of young people with diabetes.

The course is intended to be a platform to share expertise, research and development in the field of diabetes. It was advertised for open competition in ASPED and ISPAD sites and strict enrolment criteria were enforced by the ASPED/ISPAD course committee. The curriculum covered the main issues related to diabetes management in young people and the latest updates and insights to physicians involved in managing children and young people with diabetes. The meeting highlighted the interests and the needs of pediatric endocrinologists in the region.

In addition it was the ideal setting for networking and brainstorming about research ideas, collaboration with international societies and programs and available resources. With prominent international and regional speakers from ISPAD and ASPED, the course gathered 79 participants from 12 countries (one honorary place was offered to a candidate from Pakistan) and was run by experienced pediatric endocrinologists from ASPED and ISPAD.

The course showed a diverse gathering of paediatricians and diabetologists who participated effectively in the course curriculum.

<b>Table 1.</b> Curriculum of the course sessions divided to plenary sessions and small group discussions.	
Plenary sessions	Small group discussion/presentations
Diabetes comprehensive care	Diabetes unusual Presentations
Diabetes registry and bench marking	Challenges of diabetes management
Technology applications in Diabetes	Type 1 diabetes complications
Diabetes management in special circumstances (school, sick days and Ramadan fasting)	Diabetes and psychology
Diabetes complications; acute and chronic	Diabetes education
Psychology and patient empowerment in diabetes	Monogenic diabetes; MODY and neonatal diabetes
Newer therapy modalities and prevention in diabetes	Hyperinsulinemic hypoglycemia
Obesity and type 2 diabetes in children and adolescents	Obesity and type 2 diabetes
Monogenic diabetes (international and regional experience)	Diabetes and associated syndromes

### Course sessions

Various formats of presentations and specialized sessions were run during the course (Table 1). The plenary sessions covered the main topics in diabetes and the small group discussion/presentations aligned in 5 parallel sessions, featuring various interesting issues in diabetes. There were hands-on sessions on how to establish a diabetes registry in various countries. In addition, there has been a lot of discussion about diabetes service provision in less developed countries where resources are sparse.

### Diabetes technology

The participants were split into 5 small groups with 2 moderators each. During these session, detailed discussion/presentations were run on use of technology in diabetes. Advances in use of insulin pumps and continuous glucose monitoring were discussed and interesting case scenarios utilizing technology were presented.

### Course special features and highlights

Amongst all the interesting topics discussed in the course, there was an obvious common interest in presenting topics markedly related to the regional features of diabetes. In particular, there was special emphasis on dietary challenges and various dietary habits/routines practiced in countries within the region. Similar to the 2015 course, neonatal diabetes, metabolic complications of childhood obesity and type 2 diabetes were highly popular topics chosen by candidates for presentation. Enlightening lectures were presented on new advances in treating and preventing diabetes.

### Course tasks-to-follow and recommendations:

The conference enabled participants to meet and link up with senior ISPAD and ASPED researchers and clinical experts, as well as with regional experts and fellow clinicians in a collegial environment encouraging active discussions and exchange of ideas. Inspired by the course success, ASPED has set various goals among its top priorities for the near future. A task force of 7 members from 6 countries was formed. The main task with priority is to establish diabetes registry amongst various countries from the region.

## II. Abstracts of Case Presentations

### 1. An Infant with Diabetes and Insulin Requirement of 13 unit/kg/day

**Ahmed Yousif Ibrahim, Sheikh Khalifa Medical City, Abu Dhabi, United Arab Emirates.**

**Case Report:** A full term female patient of consanguineous parents born with a low birth weight (1.17 Kg) presented with neonatal diabetes at age of 5 weeks. The neonate was treated first with IV insulin, shifted afterwards to SC insulin (Detemir) then Glargine. Notably, large doses of Glargine 16 unit twice / day at age of 7 weeks was needed to achieve good glycemic control (weight 2.44 kg, TDD= 13.1 unit/kg/day). Family history revealed a previous sib death with a similar clinical picture at age of 26 months due to febrile illness and the mother had a history of abortion at age of 7 weeks. On examination, she had dysmorphic features (broad and prominent nose, long philtrum, prominent lips, hypertelorism, low set ears and increased body hair mainly on shoulders).

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### 2. An Omani Child with MODY Type 3 (heterozygous for HNF1A) Aida Alshahi, Dibba Hospital, Dibba, Oman

**Case Report:** An 11 years old male child of non-consanguineous parents presented at the age of 9 years after head trauma. During admission, he had persistent hyperglycaemia without ketoacidosis. Family history revealed a mother with gestational diabetes that persisted after delivery and is currently on oral hypoglycaemic

drugs. His grandparents are diagnosed with type 2 diabetes. Physical examination showed that he was obese (BMI of 28.3 m<sup>2</sup>) with mild acanthosis nigricans, otherwise his systemic examination was normal. He was started on glargine and pre-meal short acting insulin. During follow up, his readings were still high with HbA1C of 11%. Laboratory investigations revealed negative auto- antibodies with normal C-peptide. Therefore, metformin was added suspecting the diagnosis of type 2 DM. In view of his uncontrolled diabetes, absence of severe obesity, positive maternal history of gestational diabetes, family history of type 2 DM without obesity and negative antibodies, genetic study for MODY genes was performed which revealed positive heterozygous pathogenic variant in HNF1A. Currently, he is on insulin glargine 19 units/day and started on gliclazide 20 mg once daily. His blood glucose readings started improving and HbA1c dropped to 9%.

**Conclusion:** MODY is a genetically and clinically heterogeneous group of conditions, which is diagnosed through suspected gene sequencing. Although the genetic testing is expensive, the diagnosis has important implications for patient management and prognosis. A targeted selection of individuals for genetic testing is necessary to improve the yield of diagnosis. Regarding our patient, the confirmed mutation necessitate the need for close follow up for the micro and macro complications, adjustment in sulphonylurea doses and possibility of insulin discontinuation without risk of ketoacidosis.

### 3. Diabetic Ketoacidosis Misdiagnosed As Acute Abdomen

**Ali Hasan Dhari Al-Jumaili, Central Teaching Hospital for Pediatrics, Baghdad, Iraq.**

**Background:** Diabetic ketoacidosis (DKA) represents the most severe derangement of insulin-regulated metabolism. The acidosis and ketosis creates an ileus causing abdominal pain, nausea and vomiting. Occasionally the ileus will produce pain severe enough to raise concern for acute abdomen. **Aim:** To present a case of DKA in a known diabetic patient misdiagnosed as acute appendicitis and compare that with a similar case report. **Case Report:** An 8- year old child who is a known case of type 1 diabetes mellitus presented with abdominal pain and vomiting and was referred to pediatric surgery in a private hospital as a case of acute appendicitis. Prior to surgery, his blood sugar was high and he was referred for endocrinology consultation. He

was treated according to DKA protocol and made a full recovery. The patient was discharged on basal-bolus insulin regimen and scheduled for follow up. **Conclusions:** This case report highlights that acute abdomen might mimic DKA. We recommend that capillary blood glucose check to be part of management of children presenting with dehydration and symptoms suggestive of acute abdomen. Raising awareness of this differential diagnosis among medical staff is necessary.

#### **4. Permanent Neonatal Diabetes Mellitus: with a Mutation in the KCNJ11 Gene on Oral Sulfonylurea Treatment**

**Amal Al Hakami, King Khalid University Hospital, Riyadh, Saudi Arabia.**

Neonatal diabetes mellitus (NDM) is considered a rare disease, affecting 1:500,000 births. It is diagnosed in the first six months of life, with persistent hyperglycemia requiring insulin treatment and can be either transient or permanent. There has been major progress in recent years uncovering the genetic causes of diabetes presenting in the first year of life, which increased awareness and knowledge about the long-term management of NDM. Our patient is 2 ½ years old who presented at the age of 2 month with an acute illness in the form of fever, vomiting and increase work of breathing. He was born at 38-weeks of gestation, with a birth weight of 2.3 kg. His parents were consanguineous with no history of diabetes in the family. He was found to have hyperglycemia (blood glucose 38 mmol/L) with clinical and biochemical evidence of ketoacidosis. Patient was started on insulin infusion and then shifted to subcutaneous insulin (NPH and regular insulin) after DKA resolution. Genetic testing was performed and showed mutation in the KCNJ11 gene in which patients with this mutation may respond to oral sulfonylureas. At age of one year, patient was transferred successfully from subcutaneous insulin to oral glibenclamide with good glycemic control.

#### **5. Permanent Neonatal Diabetes due to KCNJ11 Gene Mutation.**

**Amer Al Ali, King Fahd Hospital, Riyadh, Saudi Arabia.**

We report a new case with neonatal diabetes who presented at the age of 7 weeks with diabetic ketoacidosis and referred to hospital. The patient was treated with subcutaneous injection of basal insulin in the form of NPH (1 unit twice daily). His gene testing study

revealed K channel mutation, so he was shifted to sulfonylurea at age of 4 months with gradual tapering of insulin dose. Finally, the patient was successfully weaned off insulin and continued on oral sulfonylurea only. Currently, he is 18 months old with HbA1c of 7%.

#### **6. Glycogenic Hepatopathy Complicating Poorly Controlled Adolescent's Type 1 Diabetes Mellitus: a Case Study**

**Anas Mohammed Ahmed Al-Shoomi, Children Specialized Hospital, King Fahad Medical City, Saudi Arabia.**

**Introduction:** Glycogenic hepatopathy (GH) is an uncommon cause of liver dysfunction that occurs in patients with poorly controlled type 1 diabetes mellitus (T1DM). **Case presentation:** A 14-year-old boy with T1DM diagnosed at 7years presented with recurrent diabetic ketoacidosis (DKA) because of non-compliance due to family issues. He also had complained of vague abdominal pain several times although his systemic examination was unremarkable. His medications included insulin glargine and aspart. Family history was negative for liver disease. Systemic exam showed height and weight at 25<sup>th</sup> percentile and hepatomegaly. His laboratory exam showed a HbA1c of 15%, ALT:541 umol/L, AST:418 umol/L, total bilirubin 2.5umol/L, Gamma GT 505umol/L and ALP 2305umol/L. Further investigations such as prothrombin time, INR, serum levels of albumin, ceruloplasmin, copper, bile acids and alpha-fetoprotein were within normal ranges. Autoimmune and viral hepatitis panel were negative. Ultrasound revealed hepatomegaly without focal lesion. The patient was managed for intensive glycemic. Two months later, transaminases were normalized and liver size decreased to the normal level. **Conclusion:** Glycogenic hepatopathy is uncommon but it should be considered in children with T1DM and hepatomegaly. Treatment of this condition is limited to improvement of the glycemic control.

#### **7. Hypertriglyceridemia with Eruptive Xanthomas and Lipemia Retinalis in a Newly Diagnosed Diabetes Mellitus**

**Azad A. Haleem, College of Medicine, University of Duhok, Kurdistan, Iraq.**

**Case Study:** A 5-year-old previously healthy girl presented with a 4-day history of progressive epigastric abdominal pain, polydipsia, secondary nocturnal enuresis and history of weight loss. Her initial assessment

revealed tachypnea with Kussmaul type of respiration, tachycardia and moderate dehydration. The girl was hyperglycemic (plasma glucose level more than 600 mg/dl) and acidotic (pH 7.14, bicarbonate level 3.9 mmol/L), with urinalysis revealing ketonuria and glucosuria. After admission, appropriate fluid resuscitation and insulin treatment were started. The patient's diabetic ketoacidosis resolved after 24 hours then shifted to subcutaneous insulin therapy (Glargine and Asparte as a basal bolus regime) along with proper nutritional therapy. On examination: Skin lesions were observed; non-tender yellow papules with creamy-colored centers on extensor surfaces of the arms, hands and feet. Ophthalmoscopic examination showed creamy white retinal vessels with a faded pinkish white retinal background both in the periphery and posterior pole of the retina. Laboratory findings showed a grossly lipemic serum with elevated serum levels of triglycerides 2869 mg/dl, cholesterol 498mg/dl, Amylase 45 and HbA1c was 14.8%. Given the extent of her hyperlipidemia and hyperglycemia, the patient continued on a diabetic diet along with subcutaneous insulin therapy. After one month, all her investigations were repeated revealing normal serum glucose and lipid profile. Now the patient has neither skin lesions (eruptive xanthomas) clinically nor lipemia retinalis on ophthalmoscopic examination. **Conclusion:** This case illustrates a young person with hypertriglyceridemia associated with eruptive xanthomas and lipemia retinalis in a newly diagnosed diabetes mellitus. Previous reports suggested that this phenomenon is due to genetic abnormalities of lipoprotein lipase, or a transient decrease in lipoprotein lipase activity secondary to insulin deficiency.

### 8. Lost in the Middle!

**Badi Al Enazi, Alyammama Hospital, Riyadh, Saudi Arabia.**

**Case study:** An 8 year old known boy with diabetes for 2 year present frequently with DKA and had multiple admission to the intensive care units. He lived in a small peripheral city with his father who is a drug addict. Parents are divorced and father did not supervise his diabetes management. On the last admission, his HbA1c was 14%. Social service moved to protection house and was separated from his father. In 6 months' time, his HbA1c dropped to 8%. **Conclusion:** Unstable social life affects the diabetic child care and management and there is a strong role of the treating doctor and social

service that can save the patient and help in diabetes management and glycemic control.

### 9. Glycogen Hepatopathy in Type 1 Diabetes Mellitus

**Basheer El Naeem, Madinah Maternity and Children Hospital, Saudi Arabia.**

**Case study:** A 9 years old boy was diagnosed with DKA and was maintained on 0.8unit kg/day of glargine and rapid acting insulin. He was admitted 3 times to hospital because of DKA since diagnosis. In the last admission, he presented with increasing abdominal distension, constipation, weight loss and constipation. His mother stated that he frequently refuse to take his medication regularly. On examination, he looked cachectic, pale with distended abdomen and hepatomegaly. His investigation showed severe acidosis, high random glucose, AST, ALT and GTT. Hepatitis screening was within normal range. Wilson disease and autoimmune hepatitis were excluded. Ultrasound showed homogenous enlarged liver with no focal lesion. He continued to have high transaminases and developed lower limb edema and ascites. His insulin requirement remained high at 2.5 unit kg/day. **Conclusion:** Diagnosis of glycogen hepatopathy was suggested.

### 10. Cystic Fibrosis-Related Diabetes

**Bushra Barakat, Dubai Hospital, Dubai, United Arab Emirates.**

A 15 years old girl is known to have cystic fibrosis, congenital hypothyroidism and B-thalassemia since infancy. She developed diabetes mellitus and her glycemic control was poor with HbA1c of 15.0%. Later on, she presented with symptomatic multinodular goiter with neck mass, voice change, difficulty of swallowing and had a total thyroidectomy. A differential diagnosis of her type of diabetes will be presented.

### 11. Metabolic and Psychological Aspects of Obesity Deepti Chaturvedi, Burjeel hospital, Abu Dhabi, United Arab Emirates.

**Introduction:** Childhood obesity is one of the most rampant issues in the present pediatric endocrine practice. Unfortunately, it remains one of the most difficult problems to manage and treat in a holistic way and has many aspects to its management. Aim: To discuss the challenges in the management while dealing

with cases of simple obesity along with its metabolic and psychological implications. **Case history:** A 12 old male referred to pediatric endocrine clinic with history of increased weight gain since the last 5 years. There is a positive family of obesity in mother who performed gastric banding in 2010. The patient is followed up over a period of three years annually and his psycho-metabolic profile is discussed. **Evaluation:** Weight is 123 Kg (>95<sup>th</sup> centile), height is 165 Cm (BMI: 45Kg/M<sup>2</sup> (>97<sup>th</sup> centile), waist circumference 126 Cm, BP-146/78 mmHg. He had acanthosis nigricans, gynecomastia and the rest of the systemic examination was normal. Laboratory investigations: SGOT: 35.8, SGPT: 48.6, FT4-16.7 pmol/L (12.6-21), TSH-3.20m IU/L, cortisol: 72, HbA1C: 6.4%; S.insulin-2024 pmol/L(18-172)S.cholesterol-6.26 mml/l,tg-1.44mmol/l, HDL.1.09 mmol/L. **Management and outcome:** Patient was started on metformin along with management for hyperlipidemia. Both mother and son were referred for nutritional and psychological counseling.

## 12. Initial Management of Newly Diagnosed Child with Diabetes!

**Farah Al Mutawa, Al-Amiri Hospital, Kuwait City, Kuwait.**

An 8 years-old boy previously healthy presented with 3 days history of polyuria, polydipsia, one-day history of generalized lethargy and history of weight loss. There was no history of fever, abdominal pain, nor history of contact with sick patient. He was seen initially in the polyclinic where random blood glucose was tested and showed high levels, and then he was referred urgently to the hospital. He had unremarkable past medical history. He was a product of full term spontaneous vaginal delivery with birth weight of 3.45 kg. There was neither perinatal or postnatal complications nor any history of gestational diabetes. There was a positive history (maternal side) of type 1 diabetes mellitus in two uncles; his grandfather and aunt suffered from hypothyroidism. The patient was diagnosed with diabetes and was started on insulin treatment. Initial management of newly diagnoses children with diabetes and choice of insulin regime is discussed.

## 13. Rogers Syndrome; Thiamine-Responsive Megaloblastic Anemia

**Hamed Suliman, Nizwa Hospital, Nizwa, Oman.**

A 10 year old girl presented at age of 18 months with pancytopenia and DKA. Elder sister was deaf and died at

age of 2 years with severe transfusion-dependent anemia and DKA. Parents were distant relatives. Her diabetes was better controlled and anemia corrected after being started on thiamine 100mg/day. Her condition was deteriorating with interruption of thiamine. At the age of 3.5 years old, thiamine was increased to 200 mg/day and she was better controlled. On examination she was underweight (< 3<sup>rd</sup> centile), height (< 3<sup>rd</sup> centile) and had severe pallor. Investigations revealed a blood glucose of 28.5mmol/l, plasma C-peptide of <265 pmol/L (265-1324), Hb chromatography was normal. Her peripheral blood smear showed hypochromia, anisopoikilocytosis with macrocytosis. Bone marrow examination revealed ring sideroblasts with megaloblastic changes. ICA Abs were negative, RBC folate was 162 pmol/L (130-520). She had bilateral sensorineural hearing loss on audiogram. Her optic fundi were normal. On the basis of history and investigations, she was diagnosed to have thiamine- responsive anemia with diabetes mellitus and sensorineural deafness (Rogers' syndrome). She was treated with 100 mg/day thiamine orally daily together with insulin split-mixed regimen at 1.5 units/kg/day.

## 14. Sodium Bicarbonate in Severe Diabetic Ketoacidosis

**Jasem Daowd, Prince Hamzah Hospital, Amman, Jordan.**

**Introduction:** Sodium bicarbonate (NaHCO<sub>3</sub>) is not recommended in the management of diabetic ketoacidosis (DKA) unless there is severe hyperkalemia or cardiovascular compromise because it might increase the risk of brain edema. Treatment with IV fluids and insulin should improve the acidosis. **Case Study:** Here in we report a 13 year old female child who presented with shortness of breath and decrease level of consciousness of one day duration. Physical examination revealed a sick looking child; she was severely dehydrated with Kussmaul breathing, confused with GCS of 8. Respiratory rate was 45/min, heart rate 150/min, BP 130/80 mmHg. Chest, cardiovascular and abdominal examinations were normal. **Initial investigations:** ABG showed pH 6.82, HCO<sub>3</sub> 2.6, pCO<sub>2</sub> 18, blood sugar 32mmol/l, Urea 4.9mmol/l, creatinine 102 umol/l, Na 142meq/l, K 4.8meq/l and the urine was positive for ketones. **Management and outcome:** A diagnosis of severe DKA was made and she was treated as per the standard DKA protocol. She deteriorated and her level of consciousness started to reduce. Twenty four hours later, she was still severely acidotic with blood pH 6.89, HCO<sub>3</sub> 5.8, pCO<sub>2</sub>28 and

GCS of 6. Brain MRI showed no brain edema. Sodium bicarbonate was administered and resulted in dramatic improvement of her clinical and laboratory condition. **Conclusion:** Treatment of dehydration and hyperglycemia with IV fluid and insulin may not be sufficient to improve acidosis in DKA even after 24 hours. Sodium bicarbonate may be needed even without severe hyperkalemia or cardiovascular compromise.

#### **15. A Challenging Case of Neonatal Diabetes**

**Khaled Ateeq Alghamdi, King Abdullah bin Abdulaziz University Hospital, Saudi Arabia.**

Neonatal diabetes mellitus (NDM) is not uncommon in Arab countries. Some of these cases are due to KATP channels mutation. Our patient is 5 months old boy a product of full term normal vertex delivery and symmetrical IUGR. He had double aortic arch with a vascular ring. He presented at age of 5 months with severe DKA and negative autoantibodies. Few weeks later, he developed milestone regression, visual and hearing loss, epileptic encephalopathy, leukopenia and neutropenia followed by death at age of 18 months due to chest infection. His parents were first degree cousins. He had two siblings (boys) who died at age of 12 and 14 months due to NDM, epilepsy, white matter disease, spastic quadriplegia with no conclusive diagnosis. He has a healthy adolescent sister. There was a family history of abortion at two and five months of gestation (boys). MRI brain showed white matter disease of the entire cerebral white matter as well as brainstem and cerebellum. Extensive workup was done including genetic study for most of the NDM genes as metabolic and neurological workup were negative. Awaiting for whole genome sequences.

#### **16. A Case of Neonatal Diabetes**

**Khalid Hassan Alkandari, Mubark Al Kabeer Hospital, City of Kuwait, Kuwait.**

The patient is a full term 11 months old Kuwaiti male infant with birth weight of 3 Kg with no prenatal complication. At age of 24 day, he was admitted with history of fever, irritability and lethargy of two days duration. He was not distressed and fairly hydrated but with poor reflex, and was lethargic. His weight was 2.7 kg. Incidentally, blood sugar level was found to be high of 63 mmol/l. Patient was investigated to rule out neonatal sepsis. His urinalysis revealed ketones and glucose +4. He was started on IVF (0.9 % normal saline)

and insulin 0.1 unit/kg. After few days, oral hypoglycemia drug was initiated (Glibenclamide) for several days but with no improvement. Insulin injections 2-3 doses daily were resumed. Genetic testing revealed a homozygous mutation in ABCC8 gene. He was switched to treatment with oral Sulphonylurea (Glibenclamide) after that his blood sugar was well controlled. Proper following of a protocol for switching this type of neonatal diabetes to oral hypoglycemic agent is necessary.

#### **17. Diabetic Ketoacidosis Complicated by Venous Thrombosis**

**Khawla Al Blooshi, Sheikh Khalifa Maternity and Pediatric Hospital, Ajman, United Arab Emirates.**

Venous thrombosis is a rare complication of diabetic ketoacidosis (DKA) in pediatric age group. Here in, we describe a 3 years old boy with Down's syndrome who was admitted with DKA as a first presentation for type 1 diabetes mellitus. The patient presented to the emergency room with altered sensorium and signs of hypovolemic shock. Femoral central line was inserted for monitoring. Investigation showed hyperglycemia and high anion gap with metabolic acidosis and features of acute renal failure. He was treated according to DKA guidelines where clinical and laboratory data were improving. On the 2<sup>nd</sup> day of admission, patient became hypotensive treated with fluids and inotropic support. Repeated laboratory investigations showed high sodium, urea and creatinine. In view of his clinical condition and results, there was a high suspicion of renal vein thrombosis. US abdomen with Doppler revealed no flow in the main right renal vein with reverse diastolic flow in the right renal artery. Hypercoagulability work-up revealed no abnormality. On the 5<sup>th</sup> day of admission, patient developed scrotal and lower limb edema. Repeated US abdomen with Doppler showed extensive inferior vena cava thrombosis extending up to the right renal vein and right renal enlargement. There was blood flow around the thrombus. The patient was subsequently anticoagulated with heparin. After 2 days repeated US abdomen with Doppler showed complete resolution of thrombosis and patient was discharged home in a good condition.

#### **18. A Case of Patient with Type 1 Diabetes Mellitus**

**Mariem HH Alqattan, Farwaniya Health District, Kuwait.**



A 10 year old Kuwaiti boy was admitted to the emergency department with hyperglycemia without ketosis or acidosis. He received full education by the diabetes team. He was discharged after 5 days with basal and rapid acting insulin and regular follow up in pediatric diabetic outpatient department. He has a positive family history of type 1 diabetes mellitus and type 2 diabetes in his paternal grand parent. Investigations showed a HbA1c of 12%. Celiac screen was negative, anti GAD, anti insulin antibodies were negative and C peptide level was < 0.4 U/ml. After 1 year of diagnosis, insulin pump was used but was removed after 6 months. He had repeated pump occlusion resulting in repeated episodes of hyperglycemia particularly after waking up from sleep. After removal of the pump, he has been shifted to basal (glargine) and rapid acting insulin (novorapid). He is doing fine with carbohydrate counting method with a recent HbA1c of 6.1%. This is an example that insulin pump therapy might not be suitable for every patient and various challenges are faced when using such technology.

#### **19. Mucormycosis in a 12 Years Old Patient with Diabetic Ketoacidosis**

**Muneera Fadhil Ridha Alhusseiny, Medical College, Baghdad University, Iraq.**

**Case Study:** A 12 years old female patient was diagnosed with type 1 diabetes and started on mixtard insulin. She complained of swelling of the right side of face with pain in the right upper back teeth region of two weeks duration. The pain was severe of throbbing nature and continuous. It was aggravated by taking foods with inability to brush. Intraoral examination revealed maxillary palatal necrosis of about 4.5×3 cm. There was gingival and mucosal denudation exposing the necrotic bone with pus exudation. Extreme tenderness was elicited on palpation. Based on these findings, a diagnosis of mucormycosis involving the palate was suspected with a differential diagnosis of Wegener's granulomatosis being considered. An array of investigations was carried out. Her blood sugar level was 499 mg/dl. CT of Paranasal Sinuses depicted mucosal thickening in left frontal, posterior ethmoid, sphenoid and floor of right maxillary sinus. Right maxillary sinus showed fluid level with bubbly appearance and mucosal thickening obliterating right osteomeatal opening. CT of brain was normal. Tissue biopsy with histopathology showed non septate mucormycotic hyphae and candida were detected in pus culture. Under General anesthesia,

palatal debridement was performed. Topical rinsing with hydrogen peroxide and povidone iodine was prescribed with local amphotericin B instillation of 3 mL and the patient was advised to continue injection insulin and amphotericin B. **Conclusion:** Mucormycosis of oral cavity could be seen in patients with uncontrolled diabetes mellitus as in our case. Hence, a proper understanding of such a fulminate entity is needed as the underlying systemic conditions may be quiescent and dentists may be the first to identify the debilitated status of the patient.

#### **20. Hypoglycemia: The Cost of Ineffective Diabetes Management!**

**Mai Hajjat, Princess Rahma Teaching Hospital, Amman, Jordan.**

A 7 year old female was diagnosed with type 1 diabetes and put on twice daily standard insulin regimen (pre-mixed insulin 30 regular/70 NPH) at 1.2 unit/kg/day. She experienced frequent episodes of symptomatic and documented nocturnal and diurnal hypoglycemia. She also had frequent emergency visits for hypoglycemia and complained of day time somnolence and poor school performance. Her initial HbA1c at the time of first visit was 4.6%. The family had been misinformed that her HbA1c was within normal range and meant adequate control. In the diabetes clinic, the patient and her family were counseled on how monitor blood glucose at time of insulin dosing, documenting hypoglycemia and management of hypoglycemic episodes. They were also given dietary and exercise advice and provided with a daily blood sugar log books. Two weeks later, the patterns of blood sugar readings were studied. She was found to have severe blood sugar fluctuations, alternating between very high readings (400+) and low ones (range between 35-70). She was kept on three times daily mixtard insulin but her doses were altered and tailored according to her needs. Since her first presentation, she has been regularly followed up in the clinic. She has maintained at least 3 to 4 times daily blood sugar checks and her doses were altered accordingly. She experienced less frequent episodes of hypoglycemia, reported less disturbed sleep pattern and less anxiety for her family. Her HbA1c has been maintained in the range of 7.2 to 7.8%.

## 21. Wolfram Syndrome: A Rare Cause of Monogenic Diabetes Mellitus

**Maimoona Juma Al Qanoobi, Royal Hospital, National Center of Diabetes and Endocrinology, Oman.**

A 12 year old boy presented to the emergency department at age of 5 years with 6 weeks history of classic symptoms of hyperglycemia. His blood glucose high (28.8 mmol/L). He had no ketones in urine and his arterial blood gas was normal. His HbA1c was 12.5%. He was previously healthy. Parents are first degree cousins and his grandfather is known to have diabetes mellitus. His anti-GAD and anti-islet cell antibodies were negative. He received basal bolus therapy in the form of NPH and regular insulin with a low total daily dose (0.66 IU/Kg/d). His HbA1c dropped gradually with frequent hypoglycemia which required adjustment of his doses regularly. At age of 8 years his insulin regimen was changed to glargine and lispro. Upon follow up, he continued to have frequent hypoglycemia (HbA1c <7%) with low insulin requirement. Genetic study was performed and confirmed Wolfram syndrome WFS1 mutation. He did not have hearing or visual problems.

## 22. Hyperinsulinemia in Rabson-Mendenhall Syndrome

**Maria Thomas, Al-Amiri Hospital, City of Kuwait, Kuwait.**

**Introduction:** Rabson Mendenhall syndrome is a rare form of severe insulin resistance due to a recessive mutation of insulin receptor. Other associated clinical manifestations include renal abnormalities, facial dysmorphism, skin abnormalities and dental dysplasias. **Case presentation:** Here, we report a 14 year old Lebanese female with Rabson- Mendenhall Syndrome with severe insulin resistance and acanthosis nigricans along with cluster of renal pathologies including nephrocalcinosis and dental dysplasia. **Conclusion:** We highlight the strong association between high insulin resistance with renal anomalies in Rabson Mendenhall Syndrome.

## 23. Diabetic Ketoacidosis Complicated with Critical Care Neuropathy

**Nabila Zinaty, Adan Hospital, Kuwait.**

An 11 year old girl presented with hyperglycemia and severe metabolic acidosis. Four hours from presentation,

she had deteriorated level of consciousness with an arterial PH of 6.8 and HCO<sub>3</sub> of 2.1. Hence, she was intubated and mechanically ventilated. Later on, she developed persistent hypokalemia, needed high ventilation settings, developed renal function impairment and echocardiogram revealed depressed ventricular function (LVEF=30%). On the eighth day, she was extubated but found to be unable to get out of bed with weakness in both lower limbs and bilateral foot drop, brisk reflexes on both side but more on left side with no cranial nerve dysfunction or sensory loss. MRI revealed right thalamic area infarction and nerve conduction report showed no response from right and left peroneal nerve. However, both upper limbs were normal. The patient made a gradual recovery with no neurological sequelae.

## 24. A Toddler with Early Onset Morbid Obesity Ohoud Saleh AL-Zahrani, King Faisal Specialist Hospital, Riyadh, Saudi Arabia

**Case study:** A 2 years old boy was referred at age of 16 months for evaluation of obesity. He was full term with a birth weight of 2.2 kg. At age of 4 months, the patient started to have rapid weight gain of approximately 1.5-2 kg/month. At age of 9 months, his weight was 16 kg. He is reported to have a normal appetite compared to siblings. During the first year of life, he was on demand bottle feeding. He seems to be satisfied after his meal and no history of excessive crying for milk. Mother delayed the introduction of soft diet until 1 year as a trial to control the child's weight. He was put on low fat diet with restriction of carbohydrate intake. He drinks fresh juice only with no added sugar. He is reported to have snoring, interrupted sleep difficulty to breath at night. Parents are first degree cousins; they have 2 other kids who are normal. Mother's BMI is 46 kg/m<sup>2</sup>, she has no medical disease and not on any type of medications. Father is healthy and has normal BMI with no family history of obesity. His physical examination, his height is at the 30 percentile, weigh above the 97 percentile with an SDS of +7 SD. General examination showed generalized obesity, almond shape eyes, short fingers with no polydactyly, no other dysmorphic features, no acanthosis nigricans, no Cushing features and no tonsillar hypertrophy. Genitalia confirmed presence of micropenis with normal descended testes bilaterally. **Work up:** Early onset of obesity genetic testing panel

showed that the patient has homozygous splice site mutation in Leptin Receptor gene (LEPR).

**Management:** Multidisciplinary approach, including pediatric endocrinologist, dietitian and genetic counselor was started and bariatric surgery was considered.

## 25. Brittle Diabetes of Mauriac Syndrome Picture

**Ola Zekry, Alsabah Hospital, MOH, State of Kuwait.**

**Case Study:** A 15-year-old male with an 11 years history of type 1 diabetes was referred for evaluation of growth retardation, abdominal distension and poor diabetic control. The patient was on Lantus and Novorapid in a dose of 1u/kg/day. There were previous attacks of diabetic ketoacidosis, hyperglycemia with ketonuria and recurrent episodes of documented hypoglycemia. HbA1C was 11.25 % on several visits over the last one year. Urine albumin creatinine ratio was high with blood pressure on the 97<sup>th</sup> centile for his gender and height. Captopril was started to improve his proteinuria. Anthropometric data revealed height 137 cm (<3<sup>rd</sup> percentile), weight 33 kg (<3<sup>rd</sup> percentile), body mass index 17.58 kg/m<sup>2</sup>, height age 10.5 years, weight age 11 years, bone age 7.3 years, testes both were 5ml.

**Conclusion:** Mauriac syndrome is still seen amongst poorly controlled adolescents with diabetes. It affects growth adversely and results in delayed puberty.

## 26. Challenging Case of Persistent Acidosis in Type 1 Diabetes Mellitus

**Suha Atyani, Mubarak Al Kabeer Hospital, Kuwait.**

We report a 7 year old Kuwaiti boy who was admitted with a picture of severe diabetic ketoacidosis as a first presentation of type 1 diabetes mellitus. He presented with abdominal pain, polyuria, polydipsia and increased work of breathing. His initial investigations showed high blood glucose of 40 mmol/l, with severe metabolic acidosis. He was treated as per DKA protocol in PICU and showed improvement in terms of his hydration, neurological examination and glycemic control, so he was shifted to MDI insulin regimen. Despite this, the child continued to have persistent metabolic acidosis. On further evaluation, he was found to have normal anion gap metabolic acidosis and alkaline urine along with hypercalciuria. His metabolic workup was negative. However, his family history was positive for type 1 renal tubular acidosis. Based on these data, the child was suspected to have renal tubular acidosis type 1 along with type 1 diabetes mellitus. So, he was commenced on oral sodium bicarbonate therapy with resultant

alleviation of his metabolic acidosis. The management of diabetic ketoacidosis was challenging in this case as we know that bicarbonate therapy is not the standard practice yet this case may implicate that bicarbonate therapy can be of use in such association.

## 27. Insulin pump therapy in a case of neonatal diabetes with severe low birth weight

**Omer Ahmed, Royal Hospital, Muscat, Oman.**

**Introduction:** Neonatal diabetes mellitus (NDM) is defined as diabetes before 6 months of age, which is a rare disease. Nearly half of individuals with NDM are affected by permanent neonatal diabetes mellitus (PNDM). The common causes of PNDM are mutations in KIP channel genes (KCNJ11, ABCC8) and the insulin gene (INS). However, there are other causes of PNDM like inactivation of glucokinase gene (GCK).

**Case study:** Here in, we present a case of PNDM due to homozygous nonsense mutation of GCK. A 2 year old boy who was born at 37 weeks gestation via emergency cesarean section with a birth weight of 1.54 kg. He presented with hyperglycemia on day 2 of life. He was given initially regular and NPH insulin with frequent hypoglycemia that necessitates to be shifted to insulin pump therapy with marked improvement of his glycemic control on regular follow up. **Conclusion:** The purpose of this case presentation is to confirm the possibility of the safe use of insulin pump therapy in very small babies.

## 28. Diabetic Ketoacidosis with Rhabdomyolysis

**Rafik Fathy Abdulaziz, Adan Hospital, Kuwait.**

A 5 year old boy previously healthy presented with history of lethargy, vomiting and altered level of consciousness. He was drowsy, clinically dehydrated with rapid deep respiration and Glasgow coma scale (GCS) of 10/15. Blood glucose (BG) level was 35 mmol/L, blood PH 7.06, bicarbonates 8.5 mmol/L. The patient received IV normal saline bolus (10ml/kg) then started on IV insulin infusion, but his level of consciousness deteriorated with GCS of 8/15. So, he was admitted to the PICU, cerebral oedema was suspected both IV fluid rate decreased and IV mannitol was given. One day after admission, his consciousness deteriorated (GCS 5/15), developed hypotension, poor peripheral perfusion and decreased O2 saturation. He was intubated and mechanically ventilated then he was given inotropic support. BG and BGA were normal, but CK was highly elevated. However, CK MB, troponin, ECG and

echocardiography all were normal and cardiac EF 65%. Thirty six hours later, he started to pass reddish brown urine. CK was 85000 then 120000, so he was diagnosed to have rhabdomyolysis. After replacement of fluid deficit guarded by CVP and correction of electrolytes disturbances, inotropes was tapered and stopped then patient was successfully extubated. CK levels and renal functions have gradually improved and the patient was discharged from PICU to the general ward fully conscious and hemodynamically stable.

### **29. Role of Dietary Interventions in Management of Type 1 Diabetes Mellitus**

**Radia Rezak, EHS Pediatric Canastel, Oran, Algeria.**

Dietary control has a very important role in diabetes management particularly in children. The glycemic imbalance can result from dietetic error or poor compliance to the given regimen. We present 3 case scenarios highlighting dietary issues in diabetes. Case 1: 14 years old teenager hospitalized for diabetic ketoacidosis (two previous hospitalization for the same diagnosis). Case 2: 13 years old teenager for hypoglycemia (teenage has been following for 6 years in our unit with previous hospitalization for ketoacidosis). Case 3: 7 years old child hospitalized for uncontrolled diabetes. All patients had uncontrolled diabetes. The 3 cases show lack of dietary control and crave for high GI foods. Detailed description of the challenges of diabetes management in the 3 patients will be presented.

### **30. Fanconi-Bickel Syndrome: Typical versus Atypical Presentation**

**Salwa Musa, Gaffar Ibn Auf Pediatric Hospital, Khartoum, Sudan.**

Fanconi Bickel syndrome is a rare autosomal Recessive disorder of CHO metabolism due to inactivating mutation of glucose transporter 2 (GLUT 2). GLUT 2 transports glucose in and out of hepatocytes, pancreatic B cells and the basolateral membranes of intestinal and renal epithelial cells. We present 3 cases, 2 of them with typical presentation of FBS with growth failure, features of rickets, metabolic acidosis and impaired glucose tolerance. On examination, they showed typical features of rickets, stunted growth and acidosis. Investigation confirmed renal rickets and liver biopsy showed typical glycogen accumulation in the liver. Diagnosis of FBS was confirmed and genetics came to confirm SLC2A2 mutation. The 3<sup>rd</sup> one, 2 months old child with polyuria

and chest deformity. Random blood sugar was high and neonatal diabetes was suspected. Genetics came to confirm SLC2A2 and FBS. Neonatal Diabetes as a presentation of FBS is well known and highlights the role of GLUT 2 in human beta cells. **Conclusion:** We conclude that FBS should be considered as a cause of neonatal diabetes ones other origins are excluded, particularly when patients are related or when diabetes is transient, even in the absence of features of FBS.

### **31. Diabetes, Obesity, Convulsions and Abnormal Sleep Pattern**

**Samah Al Hassan, Gafaar Ibn Auf Hospital, Khartoum, Sudan.**

A 4 year old female known to have diabetes for 2 years on premixed insulin presented with excessive weight gain, increased appetite, fatigability and abnormal tendency to frequent sleeping. This is associated with several attacks of convulsions and abnormal behavior especially during sleep. There are no symptoms suggestive of other systemic involvement nor significant family history. She is intellectually poor and has limited social interaction. On examination, the patient is obese, not interactive but not dysmorphic. BMI above 95<sup>th</sup> percentile with acanthosis nigricans, lipomastia and waddling gait. Systemic examination is unremarkable with no neurological deficit. She has poorly controlled diabetes with readings between 300-475mg /dl. Her thyroid and liver functions are normal. EEG report is normal and has negative ANA profile. Brain imaging was done and showed no abnormalities. I shall present this challenging case of diabetes and associated psychological impairment and will discuss the possible hypothalamic involvement related to her disturbed sleep and behavior.

### **32. A Very Challenging Case of Recurrent Hypoglycemia in Type 1 Diabetic Child**

**Hanan Al Hassan, Mubarak Al-Kabeer Hospital, City of Kuwait, Kuwait.**

A 10- year old girl was diagnosed as type 1 diabetes mellitus. She presented with diabetic ketosis without acidosis. She received full education by diabetic team and started on MDI insulin. She lost follow up and then experienced recurrent hypoglycemia and loss of consciousness. Full workup of hypoglycemia in type 1

DM was done. Critical sample was withdrawn during hypoglycemic attacks and metabolic workup was done. A thorough investigation in the hospital disclosed that in each attack of hypoglycemia she had, high insulin level and lower normal c-peptide. Accordingly, diagnosis of exogenous injection of insulin was made. Social workers and psychiatrist were informed as there was difficulty dealing with the mother accepting the idea that her daughter is having diabetes and accusing all the medical staff that they wrongly diagnosed her and she was refusing to give insulin. The aim of this presentation is to discuss the challenges, difficult cases of recurrent hypoglycemia in type 1 diabetes and to alert doctors to the possibility of the diagnosis of Munchhausen's syndrome, a proof of it would exhaust resources. Our conclusion is a high index of suspicion for factitious illness is raised when confronted with history and clinical findings that contradict laboratory findings. Psychological wellbeing of patients with diabetes is a cornerstone in their care.

### **33. Experience with Insulin Pump Therapy in a 10 year Old Type 1 Diabetes Mellitus Patient with Poor Metabolic Control: A Case Report**

**Suja Mathew, Al Jahra Hospital, Al Jahra, Kuwait**

**Case study:** A 10 year old male was admitted to the hospital with classic symptoms of polydipsia, polyuria and weight loss with a random plasma glucose of 25mmol/L. He was diagnosed as type 1 diabetes mellitus after preliminary workup and was started on multiple daily injections of insulin. There was no history of diabetes, other endocrine or autoimmune disorders in the family. His HbA1C at presentation was 13% and pancreatic auto antibodies were positive (GAD & IAA). The patient was started on MDI with total daily dose of 0.6 U/Kg of insulin, with glargine and three times daily aspart insulin pre-meals. The blood glucose normalized with occasional hyperglycemia reaching 14-16mmol/L, 2-3 times per week, related mainly to eating out and consumption of sugary drinks. The child and parents were counseled and started on carbohydrate counting, which resulted in better BG readings and HbA1C improved to 8%. After 5 months of diagnosis, his glucose profile showed marked fluctuation with marked hypoglycemia A1c was 8.5%. The parents were especially worried about the hypoglycemic events. According to the child and parents, there was no change in lifestyle including diet or exercise. He was started on sensor-augmented insulin pump therapy. This resulted in improvement of his glucose readings. HbA1C improved

to 8%. The parents are happy about the flexibility or meal timings and improved control of blood glucose readings.

**Conclusion:** Fluctuations in blood glucose control has been considered one of the indications of insulin pump therapy, irrespective of HbA1C levels. In our patient, this proved not only to stabilize the blood glucose readings, but also improvement in HbA1C and better parent satisfaction.

### **34. Mauriac Syndrome**

**Tara Hussain, Shar hospital, Sulaymania, Iraq.**

A 14 years old boy lives in rural area with very poor controlled type 1 diabetes mellitus since the age of 5 years. He was on twice premixed insulin doses but with poor compliance and infrequent glucose monitoring by glucometer. He presented with extreme short stature. On examination his height and weight were well below the 3<sup>rd</sup> percentile. He had moon facies, waxy skin, high pitched voice, abdominal distention, poor vision, no signs of puberty and testicular size was 3ml and hepatomegaly. The patient was noted to have hypertension with blood pressure of 170/120 documented by multiple examiners in different settings and with different instruments. He also had a grade 2-3 systolic murmur. His investigations showed a HbA1c of 19%, low IGF1. Thyroid function and celiac screen and 24 hours urine protein were normal. Treatment consisted of multiple daily injections of insulin glargine and insulin aspart. The aim was to gradually reduce his HbA1c to avoid any adverse effect on his vision if sudden reduction of glucose readings were attempted. Hypertension was treated with antihypertensive medications. Conclusion: It is important to assess if hypertension is secondary to nephropathy or is it a macrovascular complication of diabetes. This complication can occur early in disease course in very poorly controlled patients.

### **35. Diabetes Mellitus Associated with Interesting Skin Lesions**

**Wafaa Laymoun, Faculty of Medicine, Mansoura University, Mansoura, Egypt.**

**Background:** The H syndrome (OMIM 612391) is a recently described as an autosomal recessive disorder characterized by cutaneous hyperpigmentation, hypertrichosis, hepatosplenomegaly, heart anomalies, hearing loss, hypogonadism, short stature, hyperglycaemia /diabetes mellitus, hallux valgus, and fixed flexion contractures of the toe and finger joints.

Histologically, there is an inflammatory infiltrate consisting mainly of histiocytes, later replaced by fibrosis of the deep dermis and subcutis. Case report: A 6 year old female presented with hyperglycemia, skin hyperpigmentation and sensorineural hearing loss. Clinical, laboratory, histopathological, and radiological evaluation were in favor of the diagnosis of H and/or PHID syndrome. Moreover, she presented with previously un-described features; absent pectoralis major muscle and a supracondylar bony spur in left humerus. Mutation analysis of SLC29A3 gene was performed for all members of the family. Homozygous mutation previously reported in literature c.1279G>A [p.G427S] was found in the patient and unexpectedly in her asymptomatic brother.

### **36. Neonatal Diabetes with INS Gene Mutation Yasser Alghanmi, Maternity and Children Hospital, Jeddah, Saudi Arabia.**

We report homozygous mutation in the human insulin gene in one patient with neonatal diabetes. The parents are reported to have heterozygous INS gene mutation. This mutation is inherited as an autosomal recessive manner in this family. However, this mutation inheritance was autosomal dominant in other familial cases and the majority had de novo mutation. Diabetes presented in neonatal period with marked hyperglycemia and without diabetic ketoacidosis was not associated with  $\beta$  cell autoantibodies and was treated since diagnosis with insulin. The mutation is in critical regions of the proinsulin molecule, and we predict that they prevent normal folding and progression of proinsulin in the insulin secretory pathway. The abnormally folded proinsulin molecule may induce the unfolded protein response and undergo degradation in the endoplasmic reticulum, leading to severe endoplasmic reticulum stress and potentially  $\beta$  cell death by apoptosis. This process has been described in both the Akita and Munich mouse models that have dominant-acting missense mutations in the Ins2 gene, leading to loss of  $\beta$  cell function and mass. The identification of insulin mutations as a cause of neonatal diabetes will facilitate the diagnosis and possibly, in time, treatment of this disorder.

### **37. Wolcott-Rallison Syndrome Mohsina Noor Ibrahim, National Institute of Child Health, Karachi, Pakistan.**

**Introduction:** Wolcott-Rallison syndrome (WRS) is a very rare genetic disease, characterized by permanent neonatal diabetes mellitus (PNDM) with multiple epiphyseal dysplasia and other clinical manifestations, including recurrent episodes of acute liver failure. **Case report:** Three months old female infant presented with fever, vomiting and polyuria for one week duration. She is a product of consanguineous marriage of first cousin parents. Her three siblings had history of diabetes diagnosed during infancy and all died with complications of diabetes and severe infections. Her father was also diabetic. Our patient had developed respiratory distress and uncontrolled blood sugar. She had history of previous multiple admissions and work up at primary care center and tertiary care center where she was managed for fever and recurrent urinary tract infection. She is immunized up to date. She was born by normal vaginal delivery and pregnancy was also uneventful with a birth weight of 3kg. Her blood sugar was persistently high with HbA1c 14.8%. Skeletal survey showed both hands with irregular carpal bones. Coxa vera deformity was seen in pelvic x-ray. There was flattening of proximal metaphysis of right femur. Skull, spine and long bones appeared normal. Liver function test, renal function test and echocardiography were normal. Genetic analysis of neonatal diabetes and sample were analyzed for common mutations of NDM like ABCC8, KCNJ11, INS and EIF2AK3 genes. She was positive for EIF2AK3 mutation and final diagnosis was confirmed of Wolcott-Rallison Syndrome. Our patient is doing well on insulin therapy and is 18 months old currently. **Conclusion:** The genetic etiology could be determined in cases of neonatal diabetes mellitus as it guides to management, systemic involvement and genetic counseling.

### **38. Neonatal Diabetes; How Rapid Genetic Analysis Alters Clinical Management Sarah Ehtisham, Royal Manchester Children's Hospital, Manchester, UK.**

Neonatal Diabetes mellitus is a rare, monogenic form of diabetes, presenting in the first 6 months of life. It can be permanent or transient and can remit in infancy but return later in life. In the UK the commonest causes are mutations in the K-ATP channel. We present the case of a small for gestational age baby who presented at 7 weeks of age in DKA, He was stabilized on standard DKA management and re-established breast feeding. He was initially managed on IV sliding scale insulin to establish his insulin requirements and subsequently

transferred onto insulin pump therapy and required approximately 1u/kg/day of Novorapid. Genetic analysis demonstrated a heterozygous ABCC8 mutation associated with a transient form of neonatal diabetes and opened the possibility of oral sulphonylurea treatment instead of sc insulin. He was transferred onto glibenclamide over a week, slowly weaning down the insulin as the glibenclamide dose was increased. He went home at 11 weeks of age on a tds dose of glibenclamide with excellent diabetes control. Over the next few months, his glibenclamide dose was gradually weaned and he came off treatment completely by 1 year of age when his diabetes remitted. This case highlights some practical management issues with pump therapy in neonatal diabetes and strategies to manage a breast-fed infant, and also demonstrates the improved glycaemic control on sulphonylurea treatment in this condition. It demonstrates the importance of early genetic analysis as it allowed us to simplify his management and take him off insulin completely.

### **39. A Child with Type 2 Diabetes Mellitus**

#### **A. Brahim, Military Hospital of Algiers, Algeria.**

The incidence of type 2 diabetes affecting children is increasing worldwide following the increased incidence of risk factors predisposing to this chronic disease. Here, we present a male child aged 14 years who has three risk factors for predisposition to type 2 diabetes: family history of type 2 diabetes including both parents, obesity and exposure in utero to hyperglycemia. These risk factors were associated with abnormalities of the fasting glycaemia (blood glucose level > 10 mmol/L) and symptoms of insulinopenia. However, there were no metabolic complications. Despite the absence of insulin resistance symptoms (acanthosis nigricans, hypertension, dyslipidemia, nonalcoholic fatty liver disease), the diagnosis of type 2 diabetes was suspected in absence of anti GAD antibodies while levels of C-peptide and insulin were normal. Insulin therapy was introduced in presence of high HbA1c levels (12.1%) with gradual intake of oral treatment (Metformin 500mg twice daily) associated with dietary rules and physical activity. The outcome was favorable with control of blood glucose and HbA1c, without metabolic or microangiopathic complications. Monogenic diabetes in such a scenario should be considered.

### **40. Mauriac Syndrome; A Diagnosis That Still Exists**

#### **Layla Al Romaihi, Latifa Hospital, Dubai, United Arab Emirates.**

A 6 year old boy was admitted for further evaluation and management of poorly controlled type 1 diabetes mellitus. He was diagnosed with type 1 diabetes at the age of 2.5 month and was treated initially with insulin isophan. Since then he had a total of 27 admissions for blood sugar control due to poor compliance. He had also one elder diabetic sibling who died with severe hypoglycemia during sleep. At the age of 4 years he was admitted with hypoglycemia and jaundice. Physical examination showed hepatomegaly. His differential diagnosis at that time was infectious hepatitis. He had mild hepatomegaly and normal liver texture. He was treated symptomatically and was discharged home. Later his Hepatitis profile came to be negative. At the age of 5years, he was admitted with fever and abdominal pain. He was seen initially in a private clinic and treated with Ceftriaxone. During admission, he was found to have low blood glucose of 45mg/dl, febrile, jaundiced with no edema or respiratory distress. Physical examination revealed congested throat, tender abdomen with hepatomegaly of 8 cm below the costal margin, with no splenomegaly. No stigmata of chronic liver disease. His growth parameters, showed a weight and a height below the 3<sup>rd</sup> centile. The main differential diagnosis were as the following: Diabetes mellitus with hypoglycemia, URTI, viral hepatitis, drug induced hepatitis, biliary tract disease and a form of diabetic complication. Celiac screen which was negative and IGF was low. Thyroid function test, cortisol levels were normal and his bone age read at 5-6years. His hepatitis profile, CMV and EBV were all negative. Also, he had negative autoimmune hepatitis antibody panel, normal ceruloplasmin and copper levels. He was treated with vitamin K and fresh frozen plasma due to disturb coagulation profile. His insulin dosage were adjusted for better glycemic control. Ceftriaxone was discontinued. Later, he started to have edema with disturbed renal functions. He also started to have abdominal distension so he was treated with Frusemide. With the above therapy, the patient improved slowly and his renal and liver function tests improved. His latest investigation showed: HbA1C 10.4 Alt 0.2, ALP 600, total bilirubin/direct bilirubin 2/2.3, urea of 59 and creatinine of 0.5. Clinically, his liver regressed in size to 2-3 cm below the costal margin. Mother was instructed on the importance of strict blood glucose control, but this continued to be a real struggle with this patient. The case highlights the challenge of multisystem involvement with diabetes and lack of firm diagnosis of polyendocrinopathy syndrome.

#### 41. Factitious Hypoglycemia

**Sawsan Ghanem, Amiri Hospital, Kuwait city, Kuwait.**

An 8 Years old child presented with severe hypoglycemia needing intensive care admission. Detailed history revealed that the child was given more than 10 times the assigned dose of insulin. The case highlights the importance of thorough history taking in cases of hypoglycemia. Factitious disease is not uncommon in diabetes with hypoglycemia being the commonest form. The presentation could be inflicted by the patient himself in an attempt of attention seeking or for the purpose of getting sweets to treat the hypo. More dangerously, it could be a feature of Münchhausen syndrome by proxy.

Detailed management of this particular case and psychological approach and management will be presented.

#### 42. A Case with Diabetes and Celiac Disease; A Double Trouble Situation

**Khawla Abdulla, Dubai, United Arab Emirates.**

This boy diagnosed at the age of 9 years with type 1 DM following admission to pediatric ward with DKA. HbA1C at time of diagnosis was 13%. He was started on MDI regimen of insulin with a dose of around one unit per kg per day. He did well on this dose and he experienced a honey moon period when his dose was changed slightly. His serial HbA1C were 5.3%, 6.1%, 7.6% and 8.6%. A year later, he started getting fluctuant blood glucose and unpredictable hypoglycemia. Further follow up revealed strong positive Anti-endomysial antibodies (EMA). He had an endoscopy and the result of histopathology confirmed Celiac disease. Hence, he was started on Gluten free diet and his follow up showed marked improvement.

#### 43. A Novel Mutation in the FOXP3 Gene in a Palestinian Infant Affected with Immune Dysregulation, Polyendocrinopathy, Enteropathy, X-linked (IPEX) syndrome

**Abdulsalam Abu-Libdeh, Makassed Islamic Hospital, Jerusalem, Palestine.**

**Background:** Neonatal Diabetes Mellitus (NDM) presents as uncontrolled hyperglycemia during the first 6 months of life. The majority presents with IUGR, failure

to thrive, decreased subcutaneous fat and low or undetectable C-peptide levels. NDM is classified into permanent (PNDM) and transient (TNDM). IPEX (Immune dysregulation, Polyendocrinopathy, Enteropathy, X-linked) syndrome results in approximately 4% of cases of males with PNDM. It is caused by mutations in the FOXP3 gene that codes for the scurf in protein which is a member of the forkhead/winged helix domain family of DNA-binding proteins. Here we describe a novel mutation in the FOXP3 gene in a Palestinian infant with IPEX syndrome. **Clinical Data:** A male infant was born at term to healthy Palestinian parents with birth weight of 1700 gm. He presented shortly after birth with hyperglycemia, consistent with neonatal DM, persistent diarrhea, FTT and decreased subcutaneous fat. He died later with klebsiella sepsis. IPEX syndrome was suspected and confirmed by molecular testing.

**Molecular Data:** DNA Sequencing of the FOXP3 gene in the deceased infant and his mother showed that he is hemizygous for a novel missense mutation (C424Y) in exon 12 of the gene, predicting substitution of tyrosine for cysteine at codon 424, while his mother is heterozygous for the same mutation. **Conclusion:** This is the first report of IPEX syndrome in a family of Middle Eastern origin. FOXP3 gene sequencing should be performed in any male patient diagnosed with PNDM who develops other possible autoimmune associated conditions, even in the absence of the full IPEX syndrome.

### III. Abstracts of Research Work

#### 1. Debate: Old and New Insulin

**Abdulsalam Abu-Libdeh and Ghaleb Al Zughayer, Makassed Islamic Hospital, Jerusalem, Palestine.**

Insulin is the oldest of diabetes medications and is widely recognized as being the most effective at lowering glycemic levels and maintain its control. There are new formulations, new concentrations that have been brought to the market and there are other formulations still under development. In addition, old insulin have been used widely and efficiently for a long period of time for many patients. We have different insulin types now to use, they offer more options to achieve better management and control than has been described before. The problem is not a lack of variety; the real problem is the staggering price increases for new types of insulin in recent years. Newer formulations of insulin promise to



be effective but also more costly than the insulin already on the market.

## 2. A Descriptive Study of the Epidemiological Criteria of Children and Adolescents with Diabetes in Both the Clinical Pediatric Diabetes Center and the Pediatrics Department of King Fahd Hospital

**Ahmed H. Alghamdi<sup>1</sup> King Fahad Hospital and Pediatric Diabetes Center, Alabha, Saudi Arabia.**

**Objectives:** This study describes and compares the epidemiological criteria of children with diabetes followed in the pediatric clinic, the pediatric endocrinology and the diabetes clinic of the pediatric department from year 2007 to 2014. The results of this study are compared with other studies in Saudi Arabia and relevant international studies. **Patients and Methods:** This study is a descriptive study of the epidemiological criteria of diabetes in children followed in the diabetes center and the diabetes clinic in pediatric department of KFH. The children were diagnosed as diabetic ketoacidosis or having polyuria and polydipsia with blood sugar levels of more than 200 mg/dl. In the above settings, 372 cases were seen. Five of which were excluded, as they did not fulfill the inclusion criteria. An access program was used for data collection and SPSS 17 was used to analyze the results. **Results:** A total of 372 cases with diabetes were studied and were divided in groups according to their age. The first age group was between one day and six years and comprised 128 (34.4%) patients. The second group aged from seven years to 12 years and comprised 174 (46.7%) of patients in addition to the third group which comprised 58 (15.59%) of patients. The average age of the included patients was 8.32 years and females constituted 52 % (194 cases) of all cases. Type 1 diabetes constituted 95.4% (n= 355 patients) and type 2 DM constituted 1.88% (n=7). Monogenic diabetes affected three patients (0.79%), with one case of monogenic neonatal diabetes confirmed by genetic study and two other cases suspected to be MODY. The highest incidence was in the autumn months with 105 children affected constituting 27.8 % of all cases. The year 2012 had the highest rate of diabetes occurrence, with 59 child and adolescent affected at an incidence rate of 25.48 / 100000. The Al-Baha region had the highest number of cases constituting 37.7% (n=140) of cases. Diabetic ketoacidosis as the first presentation constituted 44.2% (n=167 patients). **Conclusion:** Diabetes in children is increasing significantly in the recent years, requiring more detailed analysis of its epidemiological factors to find any

significant correlations, which may help in its prevention.

## 3. Prevalence of Celiac Disease in Children with Type 1 Diabetes Mellitus in Port Sudan

**Amel Aziz Malik, Red Sea University, Port Sudan, Sudan.**

**Objective:** To determine the prevalence of celiac disease (CD) in children with type 1 diabetes mellitus (T1DM) who follow-up in the pediatric diabetic clinic in Port Sudan eastern Sudan in year 2015 and to describe the clinical features indicative of CD in the screened patients. **Study Design:** In this observational cross-sectional study, 52 children who were diagnosed with T1DM were subjected to screening for CD with tissue transglutaminase antibody testing. Those who tested positive were tested for anti-endomysial antibody (anti-EMA). Four patients were offered intestinal biopsy for the confirmation of diagnosis. Clinical profiles and manifestations of CD in the confirmed patients were compared with age matched who have no celiac cases in the clinic. **Results:** The study revealed the prevalence of CD (based on serology) with tissue transglutaminase antibody testing in children with T1DM was 14.46%. Of the seven diagnosed CD patients, more were symptomatic at the time of screening while 2 were asymptomatic. The major clinical features indicative of CD were fatigability, intestinal symptoms, distention, anemia and short stature. **Conclusions:** The high prevalence of CD in children with T1DM emphasizes the need for routine relatively expensive screening programs to be in place for these high-risk populations.

## 4. Regimens of Insulin Therapy and Their Impact on Glycemic Control

**Radhouene Attia, Pediatric Diabetes & Endocrinology Department, Necker Hospital, Paris, France.**

**Objectives:** To describe the changes in insulin therapy regimens and HbA1c in children and adolescents with type 1 diabetes (T1DM) and their associations with diabetes knowledge and quality of life (QOL). **Methods:** The study included 4293 children and adolescents (12.9±2.6 yr) with 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 insulin only ( $9.1 \pm 2.4\%$ ), but there was no difference between pump ( $8.1 \pm 1.1\%$ ), basal bolus ( $8.3 \pm 1.3\%$ ) and 2-3 injections ( $8.2 \pm 1.3\%$ ). 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. **Conclusions:** The percentage of T1DM 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.

#### **5. Prevalence of Thyroid Autoimmunity in Children and Adolescents with Type 1 Diabetes Mellitus** **Basheer Al Naem, Gezira State, Sudan.**

**Objective:** Prevalence of thyroid autoimmunity is reported to be significantly higher among young patients with type 1 diabetes mellitus (T1DM) than in the age matched general population. This condition is characterized by the presence of thyroid specific auto antibodies in patient's serum and by varying degrees of thyroid dysfunction. The aim of this study was to determine the prevalence of autoimmune thyroiditis among children with T1DM in Gezira state (Sudan). **Methods:** This is a prospective cross sectional hospital based study. One hundred and forty five diabetic children and adolescent and 50 normal children who served as a control group after obtaining informed consent. Both history and examination were reviewed. Blood was taken for thyroid peroxidase antibody levels. Those who were positive for TPO Abs, TSH and T4 levels were included. **Result :** Out of 145 diabetic children and adolescent who were studied, 11 patients (7.3%) had positive antibody. One patient had biochemical evidence of hypothyroidism. Four of those with positive TPO had subclinical hypothyroidism (2.8%). The remaining 6 patients were euthyroid. Female to male ratio was 1.75:1. All the control have negative

result for thyroid antibodies. **Conclusions:** These results show high prevalence of autoimmune thyroiditis among children with T1DM so this can justify the recommendation of screening diabetic children and adolescent for thyroid disease by analyzing circulating TSH and antibodies for TPO at diabetes onset.

#### **6. Prevalence of Celiac Disease in Type 1 Diabetes Mellitus in Children and Adolescents Attending Children Welfare Teaching Hospital** **Hana A. Abduljabbar, Children Welfare Teaching Hospital, Baghdad, Iraq.**

**Background:** The association of celiac disease and type 1 diabetes mellitus (T1DM) is known worldwide due to shared auto immunological background. Celiac disease could present in diabetic patients with non specific symptoms or remains asymptomatic. Periodic serological screening is necessary for early diagnosis. **Objectives:** To estimate the prevalence of celiac disease in children with T1DM. **Patients and methods:** A total of 152 children with type 1 diabetes attending the Children Welfare Teaching Hospital consists of 85 girls with a mean age of  $10.3 \pm 3.7$  years and mean duration of diabetes of  $3.5 \pm 2.5$  years in the duration from May 2010-May 2011. Those patients were screened for celiac disease using immunoglobulin A and G tissue transglutaminase (tTG) antibodies, immunoglobulin A endomysium antibody (EmA), and antigitamic acid decarboxylase (Anti GAD) antibodies estimation. **Results:** Anti tissue transglutaminase antibody was positive in 25 patients with higher ratio in girls (68%). Duodenal biopsy was performed in 15 patients, 13 had histological changes of celiac disease, making its prevalence of 8.6%. The classical presentation of the disease was lacking in most patients but they presented with short stature, which was below the third percentile in 79% of patient with celiac disease. In most cases, the disease was diagnosed within the first year of the diagnosis of diabetes. **Conclusion:** Annual autoantibody screening is recommended for early diagnosis and management of patients with T1DM

#### **7. Assessment of HLA Profile and Environmental Factors in Egyptian Children with Type 1 Diabetes.** **Hoda Atwa, Suez Canal University, Cairo, Egypt.**

Type 1 diabetes mellitus (T1DM) is a complex disease involving a combination of multiple factors, such as genetic susceptibility, immunologic dysregulation and

exposure to environmental triggers. The human leukocyte antigen (HLA) region is the single most important genetic determinant of T1DM susceptibility, yet variability in the HLA region has been estimated to explain only approximately 60% of the genetic influence of the disease. Over 50 identified non-HLA genetic polymorphisms support the notion that genetics alone cannot explain T1DM. A series of evidence supports a critical role of exogenous factors in the development of T1DM. The fact that <10% of individuals with HLA-conferred diabetes susceptibility do progress to clinical disease. In addition, a pairwise concordance of T1DM is of <40% among monozygotic twins. A more than 10-fold difference in the disease incidence among Caucasians living in Europe were found. Accordingly, progression to clinical T1DM typically requires the unfortunate combination of genetic disease susceptibility, a diabetogenic trigger, and a high exposure to a driving antigen. Enteroviruses are proposed candidate triggers due to temporal correlations between infection and T1DM autoimmunity and to detection of viral proteins in diseased islets. Furthermore, evidence of a cause-effect relationship is lacking.

### **8. Sudan Childhood Diabetes Program**

**Ilham Mohammed Omer, University of Khartoum, Sudan.**

**Background:** The Sudanese Childhood Diabetes Association, as part of its many objectives towards helping diabetic children, decided to raise the awareness among the medical personnel and establishing new diabetes clinics in different states of Sudan. This goal was achieved through the Sudan Childhood Diabetes Program in collaboration with the World Diabetes Federation. **Objectives:** The aim of the program was training doctors, dieticians and diabetic educators from different states of Sudan. Moreover, establishing new 25 diabetes clinics in different states, providing the clinics with protocols of management and educational materials. Lastly, having branches of Sudanese Childhood Diabetes Association in different states of Sudan. **Methods:** This was a 2 year program conducted in Jabir abuliz Center (the main Sudan diabetic center). The training program was a one year course for doctors, dieticians and educators. A 6 day course of theoretical and practical sessions. **Results:** We were able to train 85 pediatricians, 89 Dieticians and 89 educators. Twenty five diabetic clinics were established. Awareness was raised among other medical personnel through the trained doctors

.Awareness was raised among population as some of the trainee started to talk to the media as local radios and TV. Also, simple advices can be done over mobile phones. **Conclusion:** Childhood Diabetes Association have branches in most states now. Reduction in the cost of diabetes management to the families as the services were offered to them in different states.

### **9. Care of DKA in a Pediatric Hospital**

**Jamila Lahrach, Sanae Abourazzak, Service de Pédiatrie, CHU Hassan II, Morocco.**

Diabetic ketoacidosis (DKA) is a metabolic emergency and a major cause of morbidity and mortality of a diabetic child. It represents the extreme manifestation of insulin deficiency, and it can be either inaugural or related to rapid decompensating in a known diabetic patient. The aim of our study is to assess the quality of care for children with DKA by monitoring the recovery of consciousness, the correction of electrolytes disorders and also by establishing short and medium-term prognosis. DKA is a medical emergency. Prevention of complications, particularly brain edema, is based on analysis of a pre-treatment protocol which take into account the pathophysiological knowledge, and also, on a very regular clinical and laboratory monitoring. The best prevention is early diagnosis of diabetes, since 90% of diabetes occur in the absence of family history of diabetes. This prevention can be done by raising awareness of the general population and doctors upon the warning signs of the disease. Preventing recurrence is summarized in educating parents and children who need to know the signs of DKA warning, while insisting on the correct home monitoring of capillary blood glucose and ketonuria. Emphasis is placed on the importance of early diagnosis, awareness of doctors and nursing staff, improving the management and enforce the implementation of protocols supported.

### **10. Fasting the Holy Month of Ramadan in Older Children and Adolescents with Type 1 Diabetes in Kuwait**

**Kholoud Mohamed, Faculty of Medicine, Kuwait University, Kuwait.**

**Background:** Ramadan is the holy month of fasting for Muslims. New evolving technology in the treatment of type 1 diabetes (T1DM) had encouraged Muslim diabetics to pursuit the practice of fasting. There are limited data on fasting of children and adolescence with T1DM during the Holy Month. Objective and

hypotheses: Our aim is to investigate the ability, effect and safety of children and adolescence with T1DM to fast the Holy month of Ramadan 2014. **Methods:** This was a prospective observation study of children and adolescence with T1DM for at least one year who intended to fast the Holy month of Ramadan 2014. Pre Ramadan, children and their families were evaluated and educated about diabetes management during Ramadan. The following clinical outcomes were investigated before, during and after the Holy month including glycosylated hemoglobin A1C (HbA1C), number of days fasted, number of hypoglycemic and hyperglycemic episodes, and number of emergency hospital visits. **Results:** A total of 50 children and adolescence were recruited with a mean age of  $12.7 \pm 2.1$  years, 23 (46%) were males and 27 (54%) were females. 27 (54%) of cases were on multiple daily injections (MDI) insulin regimen and 23 (46.0) were on pump therapy and there was no significant difference between the two groups as regards mean age, gender, duration of diabetes, and HbA1C prior to Ramadan. The children fast a mean of  $20.0 \pm 9.9$  days. Most common cause for breaking the fast was mild hypoglycemia (mean blood sugar during the attacks  $3.0 \pm 0.3$  mmol). Two patients had one episode of DKA during fasting due to lower respiratory tract infection and pump failure respectively. HbA1C after Ramadan was predicted by pre-Ramadan HbA1C level ( $r=0.533$ ). **Conclusion:** Fasting in children with T1DM above the age of 10 years is feasible and safe in both pump and non-pump users. Pre-Ramadan education of the families and their children along with intensive monitoring of those children during the fasting month is crucial.

#### 11. Autoantibodies and Random C-peptide Level in Pakistani Children with Type 1 Diabetes Mellitus

**Mohsina Noor Ibrahim, National Institute of Child Health, Karachi, Pakistan.**

**Background:** Diabetes mellitus affects large number of Pakistani children and its incidence has increased over the last decade. Data about incidence and types is quite scarce. No work has been provided until now regarding its etiology and causative factors/or types of diabetes mellitus in Pakistani children. Consanguinity and interfamily marriages make the situation more vague and complex. **Objective:** To do the autoantibodies and random C-peptide level in Pakistani children with type-1 diabetes mellitus and to determine different phenotypes of pediatric diabetes in our population. **Material and**

**Methods:** The study will be conducted to all children attending diabetes clinic and admitted in diabetic ward at National Institute of Child Health, Karachi. Autoantibody positivity will be defined as  $\geq 1$  positive out of glutamic acid decarboxylase antibodies (GADA), insulin autoantibodies (IAA), and Islet cell cytoplasmic antibodies (ICA) and C-peptide level  $\geq 0.9$  ng/mL (Preserved  $\beta$  cell function ( $\beta+$ ). Moreover gender, race, ethnicity, weight, height, standard deviation scores (SDS), body mass index (BMI), family history of consanguinity and diabetes will also be recorded. HbA1C, blood sugar levels, diabetic ketoacidosis at the time of admission will also be recorded. Inclusion Criteria are all children with type 1 diabetes less than  $\leq 19$  years of age. Exclusion criteria are: children with thalassemia, drug-induced diabetes and other chronic diseases. Available interim results of the study will be presented.

#### 12. Association of CTLA-4 gene polymorphism with Type1 Diabetes among Palestinian Population

**Abdulsalam Abu-Libdeh, Makassed Islamic Hospital, Jerusalem, Palestine.**

**Background:** Type 1 diabetes (T1DM) is one of the common multifactorial autoimmune disorders caused by destruction of pancreatic beta cells by auto-reactive T-lymphocytes. It is thought to be a result of genetic susceptibility combined with environmental factors. Cytotoxic T-lymphocyte antigen-4 (CTLA-4) gene plays an important role in down regulating T cell activation and proliferation. The CTLA-4+49G>A polymorphism is one of the most commonly studied polymorphisms in this gene and reported to be correlated with a higher risk of various autoimmune diseases including type 1 diabetes. **Objective:** This study was conducted to determine the incidence of polymorphism in the CTLA-4 gene associated with T1DM in the Palestinian population. Blood samples were collected from 37 patients with T1DM (age ranges from 4-27 years) who have been followed at the national institute of diabetes in Ramallah, Palestine. Genotyping for the CTLA-4 gene was carried out by performing polymerase chain reaction (PCR). **Results:** The results showed that 30 patients (81% of all samples) had a mutation at A/G49 polymorphism in exon 1 of the CTLA-4 gene. Statistical analysis revealed that there is significant correlation between diabetes and the incidence of A/G49 polymorphism in exon 1 of CTLA-4 gene ( $P>0.05$ ). **Conclusion:** We conclude that there is a strong

association between CTLA-4+49G>A polymorphism and T1DM in the Palestinian population. This allows genetic diagnosis and can help understanding the pathophysiology of T1DM, which can lead and pave the way to the development of new therapies for T1DM.

### 13. Survey Study on Incidence of Hypoglycemia and its Relation to Fear of Hypoglycemia in Patients with Diabetes

**Rima Tahhan, Al\_Zahra Hospital, Dubai, United Arab Emirates.**

**Background:** To identify the frequency of self-reporting moderate and severe hypoglycemia and its relation to demographic, clinical variables, fear of hypoglycemia (FoH) among patients with diabetes in outpatients' clinics in United Arab Emirates. **Methods:** The study used hypoglycemia patient questionnaire by ADA/Endocrine society 2013 workshop on hypoglycemia. After obtaining approval of hospital ethical committee patient verbal consent, patients with diabetes attending outpatient specialty clinics were encouraged to participate. FoH was assessed by Hypoglycemia Fear Survey HFS-II. Hypoglycemia incidence was grouped according to its impact on patient's function into two groups; mild/no hypo and moderate/severe hypo group. Analysis was done using SPSS 21 for windows. **Results:** Total study group included 104 patients with diabetes. Type 2 DM 92.3%(96), Type 1 DM 7.7%(8). Mean age ,BMI, diabetes duration and HbA1c were 45.88±9.6, 30.88±6.04, 7.68±5.89 and 7.60±1.33 respectively. 56.7% (59) reported mild/no hypo versus 43.3% (45) moderate/severe hypo. Diabetes duration increased the incidence of moderate/sever hypo; 6.4±5.7 (59) versus 9.36±5.75 (45) for no/mild hypo and moderate/severe hypo respectively p=0.002. The calculated hypo rate per patient-year 4.375 events for T1DM patients and 0.468 for T2DM. In the total sample 20.2% (21) reported severe hypo, 43.3% (45) moderate/severe hypo and 56.7% (59) no/mild hypo. Total HFS-II and its sub-scale worry and behavior scores were significantly higher among those subjects reporting moderate/severe hypo 26.5±22.7,16.9±15.1,9.4±9.4 respectively versus 9.8±11.6, 6.2±7.7 and 3.6±6.8 for the group of no/mild hypo( p<0.001). Type 1DM sub-group 7.7% (8), 4 males and 4 females. Mean age, BMI, diabetes duration and HbA1c; 32 y, 27.6, 12 y and 8.32% respectively. All Type 1 DM patients reported hypo in the last one year; 50% of the episodes were severe, 25% moderate and 25% are mild. Mean (SD) of self-reported hypo per week

is 2.3(2), reported mean (SD) of severe hypo in 3 months and one year; 1.3(1.7) and 3.8(7.7) and for moderate hypo in 3M and one year; 7.7(10.3) and 15.3(21.6) respectively. FoH were higher among DM1 patients as reflected in higher scores on Total HFS-II and its worry and behavior subscale;41.2(28.7),33.1(21.3) and 13.1(11.7) respectively. **Conclusion:** Clinically significant hypo are common among patients with diabetes on medication especially Type 1 DM. FoH is significant among patients with moderate/severe hypo. FoH is augmented among Type1 DM. HFS-II is an easy and useful tool to assess FoH in clinical setting.

### 14. Obesity in Childhood

**Nadia Shaukat, Dubai Hospital, Duabi, United Arab Emirates.**

An exploratory study to assess the effectiveness of structured interventions to reduce and sustain the achieved BMI in children with simple childhood obesity attending the out-patient department in Dubai Hospital. Obesity and overweight are 2 conditions that are non-communicable in nature and very easily preventable by non-pharmacological methods. Failure to prevent these conditions results in a host of diseases that are difficult and expensive to treat, and have a whole lot of complications, some of which are even fatal. In days of yore, one very rarely came across these conditions in children, except for pathological reasons. Now, however, it is reaching alarming proportions and it is high time we, as healthcare providers, took measures to contain it. In the United Arab Emirates., even now, this is reaching a very vulnerable group of people, the children's population. "In 2007, an estimated 22 million children under the age of 5 years were overweight throughout the world." (WHO, 2009). There has been a substantial increase in the incidence of cases of obesity in children as a result of copying unhealthy lifestyles from the Western world. The State Indicator Report on Physical Activity, 2014, CDC presents state level information on physical activities in various states in America. The long term benefits of regular physical activities help maintain healthy bodies & mind & the consequences of inactivity have a definite increase in health risks & a shorter life span. The relationship between physical activity, diet and BMI is well established in children who have no disease condition.

### 15. Evaluation of Puberty and Menstrual Cycle Disorders in Type 1 Diabetes: A Case-Control Study of 30 Girls

### **Naila Aicha, Belfort Hospital, Algiers, Algeria.**

**Background:** In developing countries, children with diabetes is insufficiently treated and morbidity remains high particularly in pubertal development. Cycle disorders are cardiovascular and osteoporosis risk factors. **Objective:** To assess the percentage of girls with delayed puberty, delayed age of menarche and cycle disorders in our population of diabetic girls and look at the risk factors such as glycemic control and age of onset of diabetes before 10 years. **Patients and methods:** This is a case-control survey consists of 30 diabetic girls and 30 controls matched for age and BMI who were included according to inclusion criteria: age  $\geq$  11 years with type 1 diabetes and receiving basal bolus regimen. Data collection was performed on two questionnaires for witnesses' girls and girls with diabetes. Their clinical features, the characteristics of the menstrual cycle (menstrual cycle length, dysmenorrhea, and premenstrual syndrome) and the characteristics of diabetes all were recorded. **Results:** Delayed puberty is significantly greater in diabetic women: 25% vs. 3.3% in controls. Furthermore, no significant difference in the age of menarche and cycle disorders were observed (9.5% in diabetics vs. 10.7% in controls). Quality of glycemic control, the age of onset of diabetes as a risk factor for delayed puberty and disorders of cycles were non-significant in our survey. **Conclusions:** Our study found delayed puberty but not more cycle disorders in girls with diabetes. Glycemic control and age of onset of diabetes did not affect puberty and cycle disorders. Studies of larger sizes are necessary for more conclusive results. Raising awareness of the need to review pubertal development at each consultation must be undertaken. We also have to record the dates of cycles in a log book which would allow us to detect any cycle disorders in adolescents with type1 diabetes.

### **16. Prevalence and Predictors for Development of Microalbuminuria in a Sub-set of Sudanese Children with Type 1 Diabetes**

**Sahar Mirghani, Madinah Maternity and Children Hospital, Saudi Arabia.**

**Objective:** To determine the prevalence and the risk factors for development of microalbuminuria among diabetic children with T1DM receiving care in a diabetes center in Khartoum. **Methods:** This is a prospective cross sectional analytic study. A total of 84 patients aged 11-19 years who had been diagnosed as having T1DM

and attending a single clinic with duration of diabetes of 2 years or more were included in the present analysis. Data about their present age, age at diagnosis, sex, duration of the disease, family history of type 1 or 2 diabetes were collected. Body mass index, blood pressure and Tanner staging were assessed. Collected blood and spot urine samples were analyzed for glycated hemoglobin and urinary albumin. **Results:** Nephropathy was classified as normal in 50 (59.5%), microalbuminuric in 21(25%), macroalbuminuric in 13 (15.5%). **Conclusions:** There is a high prevalence of microalbuminuria in our cohort. Early diagnosis and prompt treatment of diabetic nephropathy is mandatory in patients with T1DM.

### **17. Prevalence of Dyslipidemia in Iraqi Children** **Saif Al Bayti, Central Children Teaching Hospital, Baghdad, Iraq.**

**Background:** Coronary heart disease is a leading cause of death worldwide. Because the atherosclerotic process begins in childhood, it is prudent to minimize adult coronary risk factors in younger people. For this reason, American Academy of Pediatrics (AAP) recommended that routine screening program for blood lipid levels to be performed in all children. **Objectives:** To investigate the prevalence of dyslipidemia among Iraqi children and adolescents. **Methods:** Overall, 465 children and adolescents (270 boys, 195 girls) aged between 5-15 years, residing in urban and rural areas of Baghdad, Iraq were enrolled in this randomized cross sectional study. **Results:** The overall prevalence of dyslipidemia was encountered in 21% of participants. The most common form of dyslipidemia was decreased HDL-C (11.6%), followed by hypertriglyceridemia 8.1%, hypercholesterolemia 6.2% and high LDL in 3.4% of patients. High non HDL level was found to be elevated in 6.8%. Dyslipidemia was more common in girls. The mean TC, LDL-C and non HDL-C was significantly different according to the age being higher in the younger age group (5-10 years old). Whereas TG levels were found to be higher in older age group (11-15 years old). Mean lipid and lipoprotein levels were significantly higher in participants residing in rural area; except for HDL-C mean levels which were lower. Family history of cardiovascular diseases had a sensitivity of only 30% in detecting participants with dyslipidemia. **Conclusions:** gender, age, and area of residence are important factors for serum lipids and lipoproteins levels in Iraqi children and adolescents. Also, serum non-HDL-C levels could

be used as an appropriate tool for detecting dyslipidemia in childhood.

### **18. Elevation of Serum Alanine Aminotransferase and Aspartate Aminotransferase Enzymes Levels as a Predictor of Non-Alcoholic Fatty Liver Disease in Type 2 Diabetics.**

**Samer Nema Yassen, University of Kufa, Kufa, Iraq.**

**Background:** The studies of liver function tests in type two diabetic patients reveal that abnormal tests are not uncommon encountered in those with poorly controlled diabetes and those tests are considered as predictor for non-alcoholic fatty liver disease. **Aim of the study:** To detect the correlation between liver transaminases (alanine aminotransferase (ALT), aspartate aminotransferase (AST) and non-alcoholic fatty liver disease (NAFLD) in type 2 diabetic patients. **Patients and Methods:** This cross sectional descriptive study was conducted at diabetes and endocrine center in A-Sadr medical city in Najaf, from February 2014 to March 2015. A total of 120 type 2 diabetic patients were included (male 39 and female 81). Subjects were recruited according to simple random sampling method meeting the selection criteria. Several different factors were studied including age, sex, type of treatment, body mass index and control of diabetes by measuring glycated hemoglobin. Liver functions and Ultrasonography were performed to all the patients. **Results:** A total of 120 patients participated in this study. Their mean age was  $52 \pm 8$  years with a range from 40 years to 65 years. Mean BMI was 25.8 (range 22.5 to 30.5) and 50 patients were treated with insulin therapy while the rest were treated with OHDs. The mean levels of ALT & AST was 43 and 30 IU/L respectively. Regarding clinical characteristics, only 18.4% of the patients had increased liver size, 23.4% had increased liver echogenicity. The AST & ALT levels were significantly higher among the patients with poor glycemic control compared to those with good glycemic control ( $p = 0.001$  &  $0.003$  respectively). Significant association also found in level of ALT & AST with the increase in BMI ( $P=0.024$  and  $0.014$  respectively). Increase liver size and echogenicity were significantly present among the patients with poor compared to those with good glycemic control ( $p = 0.018$  &  $0.002$  respectively). Significant association also found with the increase in BMI ( $P=0.044$  &  $0.013$  respectively). The mean values of liver function tests had no significant correlation with age, sex, mode of therapy or type of diabetes. **Conclusions:** There was significant correlation

between increase level of ALT & AST and the U/S evidence of NAFLD in type 2 diabetic patients. Significant correlation was also found between increase BMI and raised level of liver transaminase as there was significant association between control of DM and their levels. Based on the findings of this study, raised ALT and AST are more common among diabetic patients with higher BMI and those with poorly controlled diabetes. Derangement of liver enzymes correlated statistically significantly with fatty liver on ultrasound. Therefore, abnormal liver function tests among poorly controlled diabetic patients can be used as indicator of associated non-alcoholic fatty liver disease.

### **19. Genetics of Diabetes in Egyptian Children below 5 Years and their Impact on Management Strategy: A Study Protocol**

**Yumna Shaalan, Cairo University, Cairo, Egypt.**

**Background:** Incidence of diabetes in children below 5 years is rising disproportionately to the rising incidence of diabetes in all age groups. HLA genes are the most incriminated, accounting for approximately 40% of familial clustering of type 1 DM. Different combinations at the DRB1, DQA1 and DQB1 loci are recognized to have strong linkage to type 1 DM. Monogenic diabetes is about five times more prevalent in pediatric population in communities with low prevalence of obesity than type 2 DM. The clinical presentation of NDM is almost as that of type 1 DM and management with insulin is almost always indicated at presentation. However, some cases would require different treatment regimens to achieve good glycemic control owing to their different etiology. Diagnostic genetic procedures are then highly justified despite the high cost. **Aims:** This project aims to do a genetic map for Egyptian children under 5 years with T1DM and NDM so as to develop a diagnostic approach to the exact etiology among this vulnerable group. Moreover, to correlate each type with the clinical outcome most importantly the glycemic control, dosage of insulin adjustment and diabetic complications. **Methods:** the study will include 300 patients with diabetes onset below 5 years of age, 100 of them with onset below 6 months. The study will be conducted over children attending the diabetes care clinic in DEMPU in Children's Hospital. The team will include 5 pediatric endocrinologists, 5 chemical pathologists, and 3 nurses. DNA samples will be tested for genetic mutations using different molecular techniques as DNA sequencing, real time PCR and PCR-RFLP according to the mutation tested for. ELISA technique will be used to detect islet

cell autoantibodies, autoantibodies to insulin, autoantibodies to GAD (GAD65), and autoantibodies to the tyrosine phosphatases IA-2 and IA-2 $\beta$ .

## **20. Genetics of Type 1 DM**

**Bassam Y. Abu-Libdeh, Makassed Islamic Hospital and Al-Quds University, Jerusalem, Palestine**

Type 1 diabetes mellitus results from autoimmune destruction of the insulin-producing beta cells. This process occurs in genetically-susceptible subjects and is probably triggered by one or more environmental agents. It usually progresses over many months or years during which the subject is asymptomatic and euglycemic. This long latent period is a reflection of the large number of functioning beta cells that must be lost before hyperglycemia occurs. Polymorphisms of multiple genes are known to influence the risk of type 1A diabetes. These genes are divided into MHC-Genes (HLA-DQ  $\alpha$ ; HLA-DQ  $\beta$  & HLA-DR) and Non-MHC genes (mainly preproinsulin & PTPN22). These genes influence risk, but only human leukocyte antigen (HLA) alleles have a large effect. Environmental factors are believed to play important factors in pathogenesis. There are no definite ones but studies have pointed to possible association with some pregnancy-related and perinatal influences, viruses and ingestion of cow's milk and cereals.

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