ABSTRACT BOOK

Excellence in Diabetes and Endocrinology 2012; September 6-8, 2012, Rocco Forte Hotel, Abu Dhabi, UAE.

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Published: 24 December 2012
Ibnosina J Med BS 2012,4(6):258-277
Received: 12 November 2012
Accepted: 12 November 2012
This article is available from: http://www.ijmbs.org

Abstract
This conference is the second of the series of "Excellence in Diabetes and Endocrinology" organized jointly by the adult and pediatric departments of endocrinology at Sheikh Khalifa Medical City and Mafraq Hospitals in Abu Dhai, United Arab Emirates. The organizers are building on the great success of the first meeting held in 2010. The importance of adult and pediatric endocrine physicians is particularly relevant in this part of the world where the transfer of care from pediatrics to adults clinic may occur as early 13 years of age. Practicing adult physicians may find themselves faced by situations they may not have met during their formal training. Several international, regional and national speakers participated in the conference to address most topical issues in diabetes and endocrinology covering global and regional epidemiology, diagnostic strategies, and the latest good clinical care guidelines. The abstracts are grouped under the sub-headings of their respective sessions. We hope by publishing these abstracts we extend the education cause of the conference.

Key words: Diabetes, Thyroid, Phaeochromocytoma, Disorders of sex differentiation, Growth hormone deficiency, Hypoglycemia, Culturally sensitive care, Cardiovascular protection in diabetes, Vitamin D deficiency in the Middle East.

Introduction
This conference is the second of the "Excellence in Diabetes and Endocrinology" series organized jointly by the adult and pediatric departments of endocrinology at Sheikh Khalifa Medical City and Mafraq Hospitals in Abu Dhabi, United Arab Emirates. The organizers are building on the success of the first meeting held in 2010. The importance of adult and pediatric endocrine physicians getting together is particularly relevant in this part of the world where the transfer of care from pediatric to adult clinic may occur as early 13 years of age. Practicing adult

physicians trained in the west may find themselves facing clinical questions that have not been addressed during their early training days. International, regional and national speakers have been invited to participate in the conference by reviewing the current practice guidelines, latest releases in the international literature and sharing practical hands-on experiences in addition to reflecting on local and regional clinical practices, challenges and solutions. The abstracts of the presentations are published here as they were submitted by the speakers themselves par for styling and spelling. They were grouped under the subheadings of the respective sessions.

Abstract of Presentations

I. Main Program:
Plenary Lectures:

1. Genetic markers and adherence to treatment: Two important factors influencing response to Growth Hormone treatment

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Recombinant human growth hormone (rGH) is currently used to treat children and adults with GH deficiency (GHD) as well as a number of other non-GHD conditions. Children with GHD treated with the correct dose and for a sufficient period of time usually reach adult heights within the normal range. However, there is a wide variability in GH responsiveness which can be due to several factors, including the diagnosis (right or wrong), the dose of GH, severity of GHD, duration of treatment, mean parental height, genetic background, adherence to treatment, and others. The term pharmacogenomics refers to the investigation of variations in DNA and RNA characteristics as related to drug response, while pharmacogenetics is a subset of pharmacogenomics and is defined as the influence of variations in DNA sequence on drug response. A number of recent studies have shown that genetic variations in some genes (GH-receptor, IGF-I, IGFBP3) may be correlated with the clinical and biochemical response to GH treatment. The results of the PREDICT study, also indicate that IGF-I increase and stature growth during the first year may be associated to the presence of allelic variations of some genes (CDK4, KRAS, IGFBP-3, GRB10, TGFα, INPPL1, SOS1, SOS2). A validation study is underway to validate these findings. Poor adherence to treatment is also a critical factor which may result in less favorable responses to therapy in GHD children. Some recent studies have shown that the degree of poor adherence in GH-treated patients can be as high as 50%, although results vary greatly depending on the method used to record adherence. However, poor compliance with GH treatment is common and is associated with reduced linear growth. Thus, non adherent patients may not gain the physical and psychological benefits of GH treatment and may encounter later metabolic complications. In case of treatment failure due to unknown non adherence, the dose will be increased; increasing potential side effects and treatment cost, and the cost-effectiveness is decreased. The major factors influencing adherence are age (fear of injections, adolescence) the socio-economic status of the family, difficulties in handling the device used for injections, technical problems with the device, and holidays. Use of more sophisticated electronic devices has provided a useful tool to monitor adherence objectively. With these tools it will be possible to establish the long-term effect of adherence on growth and other parameters related to metabolism and body composition in GHD patients.

2. Adolescents and Young Adults: on the Edge, also in Diabetes

Ralph Ziegler, Diabetes and Endocrine Clinic for Children and Adolescents, St. Franzisku Hospital, Munster, Germany.

Adolescents and young adults face a variety of competing demands, such as balancing school duties, friendships, dating and family relationships, just coming to grips with growing up. These are very often in conflict with adherence tasks in their type 1 diabetes therapy. The purpose of the presentation is to show examples of different ways of coping with these responsibilities and address factors that impact adolescent behaviors and their decisions around self-care tasks. The presentation will review reasons underlying the impact of these factors on adolescent behavior and describe strategies to address them.

Transition of young adults with type 1 diabetes as a chronic disease to adult care is a serious and important step! To lose our patients on this way will be detrimental for their whole future diabetes related life. Therefore strategies to create a transition program have to be developed to ensure a continuous care of our young adults with type 1 diabetes. Examples will be shown and a discussion of possibilities and limitations of transition will be started.

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emerging adults: recommendations for transition from pediatric to adult Endocrine Society). Diabetes Care. 2011;34:2477-85.
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3. Garvey KC et al. Health Care Transition in Patients with Type 1 Diabetes: Young adult experiences and relationship to glycemic control. Diabetes Care. 2012;35:1716-22.

Diabetes: Size and Nature of the Problem Diabetes.

1. Diabetes in the United Arab Emirates and Gulf.
Khalid Al Jaberi, Department of Community Medicine, Health Authority Abu Dhabi. UAE.

No Abstract

2. Genetic basis for diabetes: A guide to clinicians.
Sarah Sulieman, Imperial College London Diabetes Center, Abu Dhabi, UAE.

Familial risk, pathogenesis, clinical onset, and treatment of diabetes mellitus vary according to etiology. Although Type 2 diabetes has a higher familial risk, more is known about the genetics of Type 1 diabetes. Genes contributing 60% to 65% of susceptibility to Type 1 diabetes mellitus are known. Type 1 diabetes is associated with susceptibility genes in the HLA region on chromosome 6p21 and the insulin gene on chromosome 11p15, and at least eight additional susceptibility genes are under investigation. Islet cytoplasmic antibodies provide humoral evidence of Type 1 diabetes risk. Only 10% of the genes contributing susceptibility to Type 2 diabetes mellitus are known, and they are primarily associated with uncommon subtypes of the disorder. The insulin receptor gene on chromosome 19p13 and at least five glucose transporter genes contribute to Type 2 diabetes susceptibility, and further associations may emerge from study of the glycogen synthase gene, the glucokinase gene, the MODY genes, and the leptin gene. Diabetes comorbidities may result from genetic and environmental susceptibilities independently or in combination.

3. Culturally-Sensitive "Competent" Diabetes Care.
Aus Alzaid, Department of Endocrinology and Diabetes, Riyadh Military Hospital, Riyadh, Saudi Arabia

According to the most recent publication of the International Diabetes Federation, 5 of the top 10 countries with the highest prevalence rates of diabetes in the world are located within the Gulf Region (namely, Saudi Arabia, Oman, Kuwait, Bahrain and United Arab Emirates). In Saudi Arabia for instance, one subject in four above the age of thirty is reported to have diabetes. More alarming perhaps, is the rising trend of diabetes observed in Saudi Arabia over recent years: diabetes has seen an approximately 10-fold increase over the past three decades in Saudi Arabia, (Table 1). If this trend continues, one cannot help but predict future diabetes rates in the country not different from those seen in ethnic populations such as Pima Indians whereby nearly 50% of their adult population is diabetic. We had predicted such an outcome many years ago and had urged the local community to take action against the rising tide of diabetes. Due to its general nature, diabetes is not always an easy disease to treat. Part of the difficulty lies in the fact that diabetes treatment depends almost exclusively on the part played by patients themselves. For example, to secure success of treatment, patients need to do their "homework" properly in the form of adhering to diet, maintaining a proper weight and healthy lifestyle, checking their blood glucose and taking insulin injections regularly, etc. Failing to do their homework will obviously jeopardize the quality of metabolic control achieved. However, patient performance/compliance is not only dependent on the skills and motivation of the individual patient but also on the way local culture defines an individual's convictions and attitudes towards health and disease.

In conclusion, Arabs have a distinct lifestyle and culture that does not always work to their advantage with regards to diabetes care. The pursuit of happiness is sought and largely fulfilled by adhering to religious and social obligations. Culture and medicine may at times appear to be in conflict with each other leaving the unwary physician stranded in an unenviable position. Most issues however, can be resolved through deeper understanding of the social norms of Arab society, delicate discussions with diabetic patients, better dialogue with religious authorities, and development of better communication skills by diabetes specialists. Proper education of both diabetic patients and the public at large is of paramount importance. Concerted efforts are urgently needed at all local levels (government, public, media, and medical community) to tackle the growing problem of diabetes in Saudi Arabia and the Gulf region.

The key messages: 1) Customs and traditions represent

major priorities in life for most Arabs. 2) Tread carefully when approaching cultural and religious issues with a diabetic Moslem patient. 3) With extra care and sensitivity, most issues can be resolved with gratifying results for patient and physician and 4) Proper education is crucial to efforts to tackle the problem of diabetes in the Middle East.

4. Guidelines for Management of Type 2 Diabetes 2012: An Overview.

Ahmed Hassoun, Dubai Diabetes Center, Dubai Health Authority, Dubai, United Arab Emirates.

No abstract

5. Inpatient Management of Hyperglycemia-Evidence-based Approach in Non-Critical Care Settings

Yahya Al Zaman, Department of Medicine and Endocrinology, Military Health Services,
Manama, Bahrain.

People with diabetes are more likely to be hospitalized and to have longer duration of hospital stay than those without diabetes. Actually recent surveys have shown that at any point around 25% of all the inpatient has diabetes. This will clearly impact the morbidity and the increasing costs. Subcutaneous insulin sliding scales have been widely used for more than 70 years. Recent evidence-based data has clearly shown that using sliding scale as a sole therapy is ineffective, but actually is associated with large fluctuation in blood sugar with more risks of both hyper- and hypoglycemia. In this presentation I will try to summarize the recent AACE/ADA recommendation for inpatient glycemic control and will to reach reasonable, achievable and safe glycemic targets and protocols for inpatient (Non-ICU setting) management of hyperglycemia.

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6. Role of Glucose Control in Managing Cardiovascular Risk

Angie Bethel, Clinical Trials Unit, University of Oxford, Oxford, UK.

Patients with type 2 diabetes remain at excess risk of cardiovascular disease even after the traditional risk factors are well controlled with antihypertensive, antiplatelet, and lipid lowering agents. Recent data shows that persistent hyperglycemia may explain a portion of that excess risk. In this symposium, Dr. Bethel will review the strength of the association between hyperglycemia and cardiovascular outcomes and the evidence that improving glycemic control lowers cardiovascular risk. The talk will conclude with an overview of the rationale for and current status of ongoing cardiovascular outcomes trials in type 2 diabetes.

<u>Cardiovascular Protection in Diabetes</u>

1 Aspirin Therapy in Diabetes: Do we know all the answers?

Salah Abosnana, Rashed Center for Diabetes and Research, Ajman, UAE.

Type 2 diabetes mellitus (T2DM) confers an excess risk of cardiovascular disease (CVD). The mechanisms involved in the development of the disease are an active field of research, and prompt the development of newer and safer therapeutics with implications for cardiovascular disease. There is increasing awareness of the role of platelet dysfunction, low-grade chronic inflammation and thrombogenesis in the pathophysiology of insulin resistance, T2DM, as well as type 1 diabetes mellitus and CVD. Major scientific societies have issued guidance on CVD in diabetes. The conclusions of the two panels of experts regarding the use of aspirin for the primary prevention of CV disease in individuals with diabetes are totally divergent. Whereas the US statement recommends the use of aspirin for primary prevention in all individuals aged > 40 or with additional risk factors, in the European guidelines there is no mention of aspirin for the primary prevention of myocardial infarction or CVD death, while it is recommended for the prevention of stroke. Both recommendations seem mainly based on extrapolations from data on other high-risk groups, rather than on a comprehensive review of pertinent data. Actually, a body of evidence suggests that the efficacy of aspirin in patients with diabetes is substantially lower than in individuals without diabetes. Nevertheless, existing

knowledge is mainly derived from dated trials, including small numbers of patients, and hardly representing current strategies for the management of CV risk factors. The high level of uncertainty regarding the balance between benefits and risks of aspirin therapy has important implications for clinical practice, auditing activities, and the design and conduct of randomized clinical trials.

2 Dyslipidemia in Diabetes: A Review of the Guidelines, Targets and Drugs

Salem A Beshyah, Center for Diabetes and Endocrinology, Sheikh Khalifa Medical City, Abu Dhabi, UAE.

The world health organization (WHO) Expert Committee on Diabetes Mellitus has defined the condition formally in 1980 as 'Diabetes mellitus is a state of chronic hyperglycemia which may result from many environmental and genetic factors, often acting jointly'. A very interesting and innovative definition was proposed by Miles Fisher of Glasgow in 1996 to be "'Diabetes is a state of premature cardiovascular death which is associated with chronic hyperglycemia and may also be associated with blindness and renal failure'. The presentation has three areas. Firstly, to make the case for "Diabetes as a cardiovascular disease" in favor of the new definition of an old condition by demonstrating the "vascular event-equivalent status of diabetes". The potential cardiovascular risk factors in diabetes both non-modifiable and non-modifiable will be reiterated. Hyperglycemia and hypertension would have already been discussed by other speakers. A more emphasis will be put on the discussion if the classical pattern, standard of care evaluation and management diabetic dyslipidemia will be furnished. Patients with T2DM have an increased prevalence of lipid abnormalities, contributing to the high risk of CVD. Lipid lowering mainly by statins reduces CVS events and CV deaths in diabetes. LDL Targeted strategy is the prime strategy. The General LDL Target: LDL <100 (2.6) though even lower target LDL<70 mg/dl (2.0 moll/l) remain an option in higher risk groups.

3 Rational Management of Hypertension in Diabetes: A Review of the Guidelines, Targets and Drugs.

Sarah Sulaiman, ICLDC, Abu Dhabi, UAE.

Both essential hypertension and diabetes mellitus affect the same major target organs. The common denominator of hypertensive/diabetic target organ-disease is the vascular tree. Left ventricular hypertrophy and coronary artery disease are much more common in diabetic hypertensive patients than in patients suffering from hypertension or diabetes alone. The combined presence of hypertension and diabetes concomitantly accelerates the decrease in renal function, the development of diabetic retinopathy and the development of cerebral diseases. Lowering blood pressure to less than 130/80mm Hg is the primary goal in the management of the hypertensive diabetic patients. Beta-blockers have been reported to adversely affect the overall risk factor profile in the diabetic patient. In contrast, calcium antagonists, angiotensin converting enzyme inhibitors (ACE-I's) and angiotensin receptor blockers (ARB's) have been reported to be either neutral or beneficial with regard to the overall metabolic risk factor profile. Combination therapy is usually required to achieve blood pressure goal in diabetic patients. The addition of aldosterone antagonists may be beneficial in patients with resistant hypertension and low levels of serum potassium. Aggressive control of blood pressure, cholesterol and glucose levels should be attempted to reduce the cardiovascular risk of diabetic hypertensive patients.

Insulin Pump Therapy and Advanced Therapeutics and Technology.

1. Insulin pumps; basics of therapy

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The first insulin pump was used in 1963, with insulin and glucagon being infused together. It was the size of a large rucksack. Technology has come a long way over the ensuing 50 years. From a simple insulin-filled syringe driver, the pump has moved to a mini computer, which can integrate a blood glucose meter, built-in bolus calculator, and even an integrated continuous glucose monitor. The latest technology automatically and wirelessly records insulin dose, blood glucose, eating and physical activity, saves it online and allows instant access to clinicians and family members, enabling them to call or text whenever needed with guidance and advice! These advances have enabled over half a million people around the world to benefit from using insulin pumps. Over the last decade, insulin pump therapy (continuous subcutaneous insulin infusion or CSII) has gained increasing popularity among patients with diabetes. It is the most physiological method of insulin delivery, closely able to simulate the normal pattern of pancreatic insulin secretion: continuous 24-hour adjustable 'basal' insulin delivery with superimposed prandial 'boluses'. In this 45-minute session, we will briefly look at the current

pump technology and the basic settings for a pump-starter; focus on the pros and cons of pump technology; and review the comprehensive pre-pump training which is vital for obtaining the maximum benefits of using this technology. Finally, we will cover the challenges associated with being attached to a piece of technology 24/7, recognizing that it is not a cure or even 'smart'.

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2. Insulin Pump Therapy; an update

Ralph Ziegler, Diabetes and Endocrine Clinic for Children and Adolescents, St. Franzisku Hospital, Munster, Germany

Thirty years after its introduction, the use of continuous subcutaneous insulin infusion (CSII) appears to be the most physiologic method currently available to deliver insulin. This data keeps increasing, especially among children and adolescents. When properly used, the technique is safe and effective in patients with type 1 diabetes as published in a consensus statement that summarizes the recommendations for CSII in pediatric and adolescent patients with type 1 diabetes (1). Newer insulin pumps with several features as multiple and temporary basal rates in very small steps, bolus calculators or automated shut-off at the occurrence of hypoglycemia, when used together with continuous glucose monitoring (CGM), make the therapy even more effective, easier and saver. Downloads and analysis of stored information, e.g. on self-measurement blood glucose (SMBG) and boluses increase efficiency of diabetes management and motivation of the patients.

In Germany, nearly 35% of the pediatric diabetes population is being treated with CSII (in our center 68%), in children <6yrs of age up to 70% (2) (in our center 100%). Even if there are several advantages to pump therapy, barriers to success still exist especially in the pediatric and adolescent population, which have to be addressed to avoid misconceptions and exaggerated expectations. Intensive education and training of a multidisciplinary diabetes-teams, patients and caretakers are necessary to realize the advantages of this nearly physiologic, though costly, therapy alternative.

In this presentation the current possibilities of CSII in children and young adults will be discussed.

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3. Clinical Examples of Use of Technology in Management of Diabetes in Children.

Asma Deeb, Department of Pediatric Endocrinology, Mafraq Hospital, Abu Dhabi, UAE.

The use of technology in management of diabetes underwent a dramatic evolution over the recent years with millions of affected children around the world benefiting from these advances. Use of technology in the diabetes field centers on two main areas namely monitoring of plasma glucose and delivery of insulin. Starting with the use of urine strips to check for glucose, patients now can use various smart devices to self-monitor blood glucose including those glucometers equipped with built-in insulin bolus dose calculators. A major revolution in technology is the use of continuous glucose monitoring systems (CGMS) which can either be blind that are mostly used for diagnostic purposes or the real time which patients can utilize to monitor and instantly correct glucose level variation. The other main category of technology is the use of insulin pump devices for continuous subcutaneous insulin infusion. Various models of insulin pumps have been designed and used in many age group populations. A combination technology is the

use of insulin pump with integrated CGMS which provide a tool for proper pump programming and adjustment and ensure availability of reliable blood glucose readings for patients. More recently, advanced pump were designed in which automatic pump switch off is induced by a certain level of hypoglycemia which is the first step in devising the artificial pancreas. The talk will be an interactive session showing clinical examples of real patients with various clinical problems and their solutions using CGMS and insulin pumps.

4. Integrated Technology for Better Diabetes Care
Ghassan Nabulsi, Medtronic MENA Diabetes Division, Beirut, Lebanon.

Continuous Glucose Monitoring (CGM) has been shown in multiple studies to help diabetes patients achieve better glucose control without increasing hypoglycemia (1). Medtronic iPro™ 2, the only FDA approved Professional CGM, provides a more complete picture of patients' glycemic profile by collecting blinded glucose data over 6 days; It identifies fluctuations unrevealed by fingersticks alone and acts as behavior modification in diabetes self-management. Ultimately, iPro™ 2 allows health care professionals (HCPs) to make confident treatment decisions that can lead to better management of Diabetes. In the Minimed Paradigm® VeoTM Pump, the Real-Time CGM is integrated within the system and enables automatic shut-off of insulin infusion if glucose levels fall below a predetermined limit to protect against severe hypoglycemia. Recently, a new version of CareLink® Pro Therapy Management Software for diabetes has been approved by FDA. It the first software program to offer advanced decision support to HCPs. The software analyzes data from a patient's insulin pump, CGM device and blood glucose meter to identify the most important patient information in 2 easy-to-use reports. First, the "Therapy Management Dashboard" provides a snapshot of the patient's key insulin delivery and glucose information on one page. In addition, it pinpoints the exact times the patient experienced a hypoglycemic or hyperglycemic glucose pattern and prioritizes these patterns so that clinicians know which times of day are the biggest challenges for their patients. Second, the new "Episode Summary" highlights key events that occur prior to hypoglycemic and hyperglycemic episodes, providing clinicians with quick insights on potential areas that may need to be addressed (2). Furthermore, the "Episode Summary" offers therapy considerations so that clinicians can make the most informed treatment decisions possible. By reducing the amount of time it takes to interpret patient data, clinicians may have more time to spend with patients fine tuning and making adjustments to therapy and behavior.

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Growth Disorders Assessment & Treatment

1. The Clinical Approach to the Short Child
Abdulla Al Herbish, Dr. Suliman Al Habib Medical Group, Riyadh, Saudi Arabia

Short stature is not an uncommon clinical problem encountered in the clinic of every general pediatrician or pediatric endocrinologist. The overall prevalence of moderate and severe short stature, based on a community based study done in Saudi Arabia, was 11.3% and 1.8% for boys and 10.5% and 1.2% for girls (1). Short stature is defined as the length or height which lies below the 3rd percentile (2). There are many growth charts developed in the last decades based on data derived from various sources. The most logical to be used is the one reflecting the national background of the assessed child. These growth charts are available in some countries (3). Growth velocity is a very vital tool in assessment of short children. Assessing a short child requires careful history which aims to collect all relevant details pertaining to the possible etiologies of this common clinical problem. These details include but not limited to peri-natal data e.g. birth weight, gestation age, significant neonatal symptoms and illnesses, detailed family history inclusive of parents heights and pattern of their growth and puberty, and a thorough review of systems looking for evidence of chronic illnesses or malabsorption. Performing a thorough clinical examination is also very important. This needs to focus on body proportion, midline defects, evidence of chronic illnesses or syndromes and evidence of endocrinopathy like goiter…etc. An important diagnostic tool in assessment of short children is the bone age which needs to be interpreted in view of the chronological age using the international standards (4).

The most common forms of short stature are genetic and constitutional. Further investigations needs to aim to rule out or in some important etiologies like chronic illnesses, anemias, rickets, malabsorption e.g. celiac disease, and renal insufficiency. Karyotyping is also essential in short girls. Endocrine tests needs to include thyroid functions and growth hormone. The latter is essential if suboptimal growth velocity is documented. Measurements of IGF1 and IGFBP3 are useful. Growth hormone assessment is a difficult task taking into consideration the pulsatile fashion of its secretion. The type and the number of growth hormone tests needed to label a short child to be growth hormone deficient are different from one country to another. They are however essential for the diagnosis of GH deficiency (5,6). Once a short child is diagnosed, neuroimaging is indicated to study the morphology of the pituitary and the hypothalamus and exclude tumors like craniopharyngioma. Pure congenital forms of growth hormone deficiency, growth hormone resistance, and IGF1 deficiency are rare and deserve their specific tests.

Growth hormone is indicated for children with growth hormone deficiency, children born small for gestation demonstrating sub-normal growth velocity, children with chronic renal insufficiency and girls with Turners syndrome. Idiopathic short stature is also an indication to use growth hormone in some countries. This however may lead to growth hormone use in any short child something which certainly needs an objective thought (7). Response to growth hormone is very important issue for any clinician to observe (8). Compliance and adherence to growth hormone needs to be followed up very closely (9).

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Clinical Practice Debate:

1. Clinical Practice Debate: For the motion "This house believes that growth hormone stimulation tests are essential for assessment of growth hormone deficiency states!"

Ibrahim Alwan, Department of Pediatric Endocrinology, King Abdulaziz Medical City
and King Saud Bin Abdulaziz University, Riyadh, Saudi Arabia.

The spectrum of growth hormone deficiency (GHD) in children ranges from a partial deficiency to a complete absence of growth hormone (GH), resulting in slightly short stature or severe growth retardation. In spite of the abundant availability of GH, the accurate conclusion of GHD is essential for the diagnosis of the underlying disorders and for treatment decisions. Both clinical and biochemical evaluation are pivotal to diagnose GHD. No single test or set of tests can define it. The pulsatile nature of GH secretion mandates a need for stimulation tests, hence 34 provocative tests have been developed and 189 different combination protocols were found. Such stimulation tests have become standard in both endocrine practice and for health care insurance coverage. Bright et al found that the growth hormone stimulation test (GHSTs) had 82% sensitivity but only 25% specificity to detect GHD. Low specificity may contribute for continuous debate for the need of GHSTs to diagnose GHD. However, GHSTs are still essential instrument in diagnosis GHD for the following reasons:

1) Absence of alternative tests with higher specificity and sensitivity more than GHSTs. 2) Inter-individual differences in GH handling, methodological variability and test standardization, led to the low predictive power of GHSTs. 3) In spite of low specificity, there is a good correlation between low growth hormone levels in GHSTs with GH therapy in children with GHD. 4) Based on the Australian clinical experience, only clinical and minimal investigation led to financial burden on the health system.

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2. Clinical Practice Debate: Against the motion "This house believes that growth hormone stimulation tests are essential for assessment of growth hormone deficiency states!"

Walid Kaplan, Department of Pediatric Endocrinology, Tawam Hospital, Al Ain, UAE.

Short stature is one of the most common conditions seen in Paediatric Endocrinology practice. The majority of such cases are due to non-endocrinology causes, and growth hormone deficiency, in particular, is responsible for fraction of these cases. In spite of that, confirming the status of growth hormone deficiency (GHD) is considered one of the most important challenges in dealing with short stature. While growth hormone provocative test has been considered as the gold standard of diagnosing GHD over the decades, the weight of evidence in the past 10-15 years has shown many limitations against the routine use if this test. Besides being time consuming, labor and cost intensive, and, fairly, unsafe, growth hormone stimulation test (GHST) is non-reproducible, and is widely affected by many variables such as BMI, pubertal stage, stimulants, and type of assay. Additionally, the diagnostic cut-off value is arbitrary, and highly controversial, this is why the majority of the pediatric endocrinologists in the US do not consider the GHST as the gold standard test to diagnose GHD. Clinical evaluation of growth velocity, supported by much simpler laboratory studies, such as IGF-I and IGF-BP3 levels, and imaging studies (when indicated) provide a better alternative to the GHST in the many cases. The aim of the presentation is to highlight the multiple limitations of using the GHST in its current way, and the available alternatives that can be considered in making the difficult diagnosis of GHD.

<u>Advancing Frontiers in Clinical Endocrinology</u>

1. Seamless Transition of Growth Hormone Treatment from Paediatric to Adult Clinics

Sandro Loche, Pediatric Endocrinology Unit, Microcitemico Hospital, Cagliari, Italy

According to the 2005 consensus, the transition age refers to a broad set of physical and psychological changes, arbitrarily defined as starting in late puberty and ending with full adult maturation. This usually implies a period from mid to late teens until 6-7 years after achievement of final height. Regarding patients with growth hormone deficiency (GHD), transition patients are GHD subjects with childhood onset GHD (COGHD) who have been treated with GH and have completed their linear growth. Treatment with GH should be continued during the transition age to obtain full maturation of skeleton and muscle and to prevent the metabolic and cardiovascular complications of adult GHD. A number of studies have shown that discontinuation of GH therapy is followed by a reduced acquisition of bone mineral content, which is reversed by treatment. Reinstitution of treatment also improves body composition, lipids and quality of life. The great majority of patients with isolated idiopathic GHD have normal GH secretion when retested ad the attainment of final height. Thus, confirmation of GHD is mandatory in all adolescents with COGHD. Retesting is usually not necessary in patients with transcription factor mutations, those with isolated GHD due to known mutations, and in patients with more than three pituitary hormone deficits. All the remaining patients should be retested for confirmation of permanent GHD. The insulin tolerance test (ITT) is the preferred test

for GH reserve in the transition patients followed by the GHRH plus arginine test and the glucagon test. The cutoff limits for these stimulation tests in the transition age have not been clearly established but seems to be somewhat higher than those used in the adult patients. GH doses should be individualized rather than weight-based starting with low doses which should be titrated according to IGF-I levels. Periodic monitoring is needed to look for possible side-effects and long-term benefits.

2. The Challenges of Diagnosis and Management of the XY Female

Ieuan Hughes, Department of Pediatrics, University of Cambridge, UK

The term, XY female, generally refers to complete sex reversal whereby the phenotype is a normal female despite the presence of a Y chromosome, testis determination but coupled with some defect in androgen synthesis or action. In reality, the differential diagnosis includes complete gonadal digenesis (Sawyer syndrome), defects in androgen production (such as LH receptor mutation and 17β-hydroxysteroid dehydrogenase deficiency) and total resistance to androgen action (complete androgen insensitivity syndrome). An understanding of normal fetal sex development underpins the classification of the causes of an XY female and logical pathway for investigation.
Management is focussed on presentational issues occurring either in infancy (for example, prenatal genotype / postnatal phenotype mismatch; bilateral inguinal swellings) or at puberty with the onset of virilisation in a hitherto normal prepubertal girl. Occasionally, the latter clinical scenario results in female to male gender reassignment. Gonadal tumor risk is highest in the XY female with gonadal dysgenesis and less than 5% in recent studies of CAIS. The genetics of the XY female is becoming well characterized, including progress with understanding the etiology of gonadal dysgenesis. Pooling of resources utilizing international DSD databases is the way forward to make further progress in understanding the XY female.

3. Current Perspectives in Hypogonadotrophic Hypogonadism

Pierre-Marc Bouloux, Centre for Neuroendocrinology, Royal Free Campus, University College Medical School, London UK

Hypogonadotrophic hypogonadism (HH) is a condition wherein sex steroid concentrations are reduced in the presence of inappropriately normal or frankly low LH and FSH levels. The abnormality is therefore of hypothalami-pituitary origin, and may be congenital or acquired. In this presentation, I shall focus on the congenital causes of gonadotrophin deficiency, where clinical investigation, backed up by experimental work, has identified numerous genetic lesions that affect either the ontogeny of GnRH neurones, or secretion of GnRH , or where there are abnormalities in the responsivity of gonadotrophs to endogenous pulsatile GnRH . Although frequently an isolated abnormality, HH may also be accompanied by deficiencies of other anterior pituitary hormones, usually in the context of developmental abnormalities of the anterior pituitary (e.g. PROP 1 mutations). Gongenital GnRH deficiency (with or without anosmia) can be inherited as an autosomal dominant , recessive or X-linked condition. Increasingly, it is becoming evident that oligogenic inheritance underpins the phenotype, characterised by incomplete penetrance and variable expressivity within families sharing at least one mutation in genes recognised to be critical for GnRH secretion. I shall be discussing the biology of KAL1 (anosmin 1), FgFR!, FGF8, prokinecitin 2, prokinecetin receptor 2, Kisspeptin 1 receipt, Kisspeptin 1, GnRH receptor mutations, GnRH 1 , tachykinin 3 (TAC 3) and tachykinin 3 receptor mutations (TACR3) as well as the concept of polygenicity.
GnRH neurone are born in the nasal placed from where they multiply and then migrate in an axonophilic manner along the axons of the vomeronasal nerve into the olfactory bulb area, and thence into the septobasal hypothalamus where they disperse. There are altogether some 2000 GnRH neurone in man, and they send axons to the portal capillaries of the median eminence where pulsatile GnRH neurosecretion occurs. KAL 1 is expressed particularly on the outer layer of the olfactory bulb, where our data suggest that anosmin 1, its encoded protein, acts as a modulator of FDGFR1 signalling. FGF8 may be one of the principal ligands for FGFR1. Mutations in FGF8 cause a broad spectrum of disorders of pubertal development ranging from absent to partial to complete puberty. The associated non reproductive phenotypes include hearing loss, and a range of skeletal features (high arched palate, cleft lip/palate, severe osteoporosis, camptodactyly, and hyperlaxity of the digits) and variable expressivity among family members harbouring the same mutation. It is becoming evident that the KISSR1/KISS1 system is a major gatekeeper of the timing of puberty. TAC3/TAC3R mutations are associated with a relatively unique phenotype wherein males have a high incidence of

micro phallus and cryptorchidism, as well as reversibility during adulthood in both males and females, implying that the Beurokinin B pathway may well be more critical during the neonatal period and less critical during adolescence and adult periods of development and functioning of the GnRH neurons.

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Contemporary Management of Pituitary Tumors

1. Hyperprolactinemia: The good the Bad and the Ugly

Ibrahim H Sherif, Al Aria Clinic, Tripoli Libya.

In 1932 Oscar Riddle coined the name prolactin for an anterior pituitary hormone because of its ability to stimulate crop milk production in pigeons and in 1933 developed the pigeon crop sac assay which was the only test till radioimmunoassay measurement of prolactin became available in the seventies when the link to amenorrhea galactorrhea syndrome was established. We now know that prolactin is a hormone and a cytokine of many functions that include effects on mammary gland development, lactation, pregnancy and the immune system. Hyperprolactinemia is common in endocrine practice and is said to occur in about 0.4% of normal adult populations and up to 5% of family planning clinic patients and 9% of adult onset amenorrhea but the majority are due to causes other than prolactinoma. Prolactinomas account for 40% of pituitary adenomas and therefore are the commonest. Prolactinomas can be divided into three main forms which is micro, macro or invasive prolactinoma. It can be generally said that the higher the level of prolactin the bigger is the prolactinoma except in certain situations when the disparity between the size of the adenoma and the prolactin level may be due to the hook effect. Hyperprolactinemia is an abnormality that is almost always correctable with correcting the underlying cause or with dopamine agonist therapy which not only normalizes prolactin but also shrinks the adenoma either partially or fully making chances of cure possible and the use of the word tumor unwarranted. Surgery enjoys less success than medical therapy and is often used for debulking or in patients intolerant or resistant to medical therapy whilst radiotherapy is only recommended in special circumstances because of slow onset, low cure and high complication rates. Once dopamine agonist therapy is started pregnancy can occur and the drug should be stopped the safety profile for bromocriptine and cabergoline in pregnancy is good and the fear for pressure symptoms for micradenoma are extremely small but significantly more for macroprolactinoma hence it may be wise to delay pregnancy till significant shrinkage occurs but overall results are reassuring and very few patients need emergency surgery to decompress during pregnancy. Hyperprolactinemia is a treatable disorder compatible with good quality of life and may even be curable in significant number of cases.

2. The Surgical Aspects of Pituitary Tumors

Dominic Venne, Department of Neurosurgery, Sheikh Khalifa Medical City, Abu Dhabi, UAE.

With the exception of micro and some macoprolactinomas, most other pituitary tumors would need consideration of surgery as the definitive management. These include acromegaly, pituitary Cushing's disease and acromegaly. The pre-surgical assessment nd perioperative standards of care will be presented relying heavily on the illustrating these with clinical photographic and imaging technology. An evidence-based argument will be made for the multidisciplinary team work. Expected results in good hand and illustration of the concept of "critical volume" of clinical work load will be put forward. Acute and long term complications of surgery will be discussed.

3. Update on the Management of Cushing's Disease

Pierre-Marc Bouloux, Centre for Neuroendocrinology, Royal Free Campus , University College Medical School, London, UK

Cushing's Disease is rare, with a calculated incidence of 5 -7 per million of population. It is caused by a basophil adenoma of the pituitary, with uncontrolled release of ACTH causing cortisol hypersecretion from hyperplastic adrenal glands. Management of this condition requires first and foremost clinical suspicion of its presence. In an era of obesity and PCOS, this is not always obvious and early disease is easily missed. Nonetheless, the presence of weight gain, with a

central distribution, easy bruising, thin skin, myopathy and hypertension should increase the index of suspicion of the clinician. The biochemical diagnosis is confirmed by the presence of 2-3 raised urinary free cortisols, and the failure of a 09.00 cortisol to be suppressed following a 1mg overnight (23.00) dexamethasone suppression test. In a small series of 100 consecutive obese adults (BMI>30), a false positive rate of morning cortisol >1.8mcg/dL was found in 8% of individuals receiving 1mg overnight dexamethasone, while it was only 2% in those receiving 2mg overnight dexamethasone.In this presentation, I shall discuss the value of the various tests (standard low dose and high dose dexamethasone suppression tests, midnight salivary sortisol, CRH testing, dexamethasone + CRH) performed to identify to confirm the presence and establish the cause of the hypercortisolism- i.e. ACTH dependent and non ACTH dependent. Imaging plays an important role in diagnosis, and the current sensitivity of pituitary MRI scanning in identifying a pituitary lesion (usually <1cm diameter) is in the order of 70%. Final confirmation of the diagnosis requires the performance of a bilateral inferior petrosal ACTH sampling before and after CRH administration: basophil adenomas are expected to hypersecrete ACTH, creating a central to peripheral ACTH gradient >2-3:1. failure to do so may indicate the presence of an ectopic ACTH producing source. I shall describe a recent case in our department that underpins the difficulty of making a diagnosis of CS. Confirmed pituitary lesions can be expected to be cured in 70-80% cases if the transphenoidal adenomectomy is performed by an experienced neurosurgeon, with preservation of normal anterior pituitary function. The recovery of normal corticotrophin function may take several months in these cases to occur, during which time the patient usually requires hydrocortisone replacement. Other treatments for Cushing's Disease include medical therapy, including ketoconazole, meytapone, O'P'ddd, etomidate, cabergoline and the more recent pasireotide. These are not long term treatments however, although they are useful as an interim measure. If pituitary adenomectomy has been unsuccessful , bilateral adrenalectomy (laparoscopic) is curative of the CS, but requires lifelong hydrocortisone and fludrocortisone replacement , and usually a course of pituitary irradiation to reduce the risk of future Nelson's syndrome developing.

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Molecular Genetics of Pediatric Endocrinology

1. Excellence in Endocrinology 2012 Special lecture: Advances in Genetics of Hyperinsulinaemic Hypoglycaemia

Khalid Hussain, Great Ormond Street Hospital and UCL Medical School, University of London, London, UK.

Hyperinsulinaemic hypoglycaemia (HH) is a cause of severe and persistent hypoglycaemia in the newborn and infancy period. It is an extremely heterogeneous disorder with respect to clinical presentation, pancreatic histology and molecular biology. The clinical severity of HH varies with age at onset of hypoglycemia (severe hypoglycemia in neonates) and has major consequences in terms of therapeutic outcome and genetic counseling. The commonest genetic cause of persistent HH are autosomal recessive mutations in the genes ABCC8 and KCNJ11 (encoding the two subunits SUR1 and KIR6.2 respectively) of the pancreatic ATP-sensitive potassium channel (KATP). Histologically there are two major subtypes of the disease, namely focal and diffuse. Both the diffuse and focal forms share a similar clinical presentation, but result from different pathophysiological and molecular mechanisms. In addition, diffuse HH usually presents as an autosomal recessive disorder, whereas focal HH is sporadic. Differentiation of diffuse from focal disease is important in terms of management as focal disease requires a limited pancreatectomy (curing the patient from the hypoglycaemia) whereas diffuse disease will require a near total pancreatectomy. Imaging with 18F-DOPA-PET/CT is now the gold standard for differentiating diffuse from focal disease. Other rare genetic causes of HH include mutations in the GCK (glucokinase), GLUD1 (glutamate dehydrogenase), HAHD (Short Chain 3-hydroxyacyl-CoA), HNF4A (hepatocyte nuclear factor 4-alpha), and SLC16A1 (monocarboxylate transporter 1) genes. HH may also be part of an underlying syndrome (such as Beckwith-Wiedemann, Costello and Kabuki) and multisystem disorders such as congenital disorders of glycosylation (CDG). HH following gastric bypass surgery for morbid

obesity has been reported in adults with pancreatic histological changes similar to infants with persistent hyperinsulinism. The rapid and accurate diagnosis of HH is very important, as a delay and inappropriate management can lead to brain damage. During the talk I will discuss the clinical presentation and diagnosis of HH and review the underlying molecular mechanisms that lead to dysregulated insulin secretion.

2. Genetic and Clinical Spectrum of Permanent Neonatal Diabetes in Arabs

Abdelhadi Habeb, The Pediatric and Maternity Hospital, Al Madina Al Monawarah, Saudi Arabia.

Permanent neonatal diabetes (PND) is a monogenic form of diabetes and its highest incidence rate was reported in the Arab populations of northwest Saudi Arabia (NWSA) followed by Oman. So far mutations in at least 14 genes have been identified in around two third of patients. In Europe, Japan and the USA mutations in KCNJ11 and ABCC8 genes encoding for the pancreatic KATP channel are the commonest cause of PND. Most patients with these mutations have isolated diabetes and achieve better diabetes control when switched from insulin to oral sulphonylurea. However, we have recently found no KATP channel mutations in our PND cohort in NWSA and that most patients have syndromic rather than isolated PND. This finding was further explored in a large cohort of Arab patients in whom EIF2AK3 gene mutations were the commonest followed by INS and KATP gene mutations. Our recent work indicated that PNDM in the Arabs has a different genetic spectrum compared to other populations and it is more likely to present as part of recessive syndrome rather than in isolation, possibly due to the higher rate of consanguinity. The presentation will discuss the genetics of PND and highlight our recent studies on the genotype and phenotype of this condition in Arab population.

3. KATP Channel Mutations; when Life with Needles is Not Always a Destiny!

Dina G Ramadan, Pediatric Endocrinology, Sabah Hospital, Kuwait

Neonatal diabetes mellitus (NDM) is a rare monogenic disorder and is defined as diabetes with onset in the first 6 months of life. In about 50% of cases, it is life-long (Permanent NDM). In the rest of cases, it disappears during first 18 months of life, but can reappear later (Transient NDM) (1). The KATP channel is a key for glucose-stimulated insulin release from the pancreatic beta cell. Activating mutations in the KCNJ11 and ABCC8 genes, that encode the two pancreatic KATP channel subunits, Kir6.2 and SUR1 respectively, are the commonest cause of permanent NMD. In a recent publication, it was found that other causes are more common in Arabs (2). Sulphonylurea, a KATP channel inhibitor, can restore insulin secretion and in some patients with ABCC8 mutations, transfer from insulin to oral sulphonylurea therapy has been successful and has resulted in improved glycemic control (3). We present two brothers with permanent NDM, born to healthy Syrian cousin parents. Both presented with severe DKA very early in life. The cause was found to be due to a homozygous novel missense mutation, W688R, in exon 15 of the ABCC8 gene. Both were successfully switched, very early in life, from insulin to oral sulphonylurea (glibenclamide) with excellent glycemic control. They are on normal diet and have normal physical and developmental milestones with a current follow-up period of up to 5 years and 1.5 years respectively. We stress the need for awareness for NDM and highlight that genetic testing is available such that a genetic diagnosis is possible for about 60% of permanent NDM cases. All patients diagnosed with diabetes before the age of 6 months should be referred for genetic testing irrespective of their current age in order to identify the cases likely to benefit from treatment with sulphonylureas.

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4. Familial Glucocorticoid Deficiency; phenotype genotype correlation

Hessa Al Kendari, The Pediatric Endocrine Unit, Farwania Hospital, Kuwait City, Kuwait

Context: Familial glucocorticoid deficiency type 1 (FGD1) is a rare autosomal recessive disorder resulting from defective ACTH receptor (melanocortin receptor type 2, MC2R). Individuals with this condition usually present in infancy or early childhood with the signs and symptoms of isolated glucocorticoid deficiency. Prolonged jaundice,

neonatal hepatitis, recurrent hypoglycemia, convulsions and profound skin hyperpigmentation can be the first signs of the disease. To date, hypothyroidism has been reported as an associated feature in a few cases. Objectives: The clinical features along with MC2R and TSH receptor (TSHR) genetic analysis of five Arab kindreds from four Bedouin families are described. Patients: The subjects were children with the clinical and biochemical features of FGD1 associated with a resistance to TSH (RTSH) phenotype. Three patients had hyperferritinemia at presentation and two developed growth hormone deficiency.
Methods: Mutation analysis of MC2R and TSHR was performed by direct gene sequencing. Results: Analysis of the MC2R gene revealed a homozygous insertion of a cytosine nucleotide between codons 153 and 154 in all of the patients. This mutation would be expected to cause a translation frame shift after codon 154 and a premature termination codon at 248 of the MC2R mRNA. No TSHR gene mutations were identified.

Occasional Lecture:

Thyroid disease in children

Suliman Abusrewil, Department of Pediatric Endocrinology, University of Tripoli, Tripoli, Libya.

Thyroid gland is a crucial organ, with very important function for physical growth, development, puberty, organ function, metabolism, fertility, and body temperature regulation. These functions are carried out with the interaction of only two chemicals, thyroxine (T4) and tri-iodothyronine (T3). Thyroid diseases are common and can have serious complications among adults; however these adverse effects can be more pronounced in children, because they are often harder to identify early enough, and affected children experience the most harmful outcomes. Screening, early diagnosis, and treatment for congenital hypothyroidism in the developed world has eliminated the devastating sequel of thyroid hypofunction in this age group. Children diagnosed with thyroid problems need the support of their families to ensure that they are taking their medication regularly and understand their conditions. It is also recommended that schools/nurseries are informed so that they are aware of the child's diagnosis and medication requirements. It is also important to remember that treatment is available and early intervention will help to avoid any long term sequel or complications.

Disorders of Sex development (DSD)

1. State of the Art Lecture II: Gender Assignment in DSD

Ieuan Hughes, Department of Paediatrics, University of Cambridge, UK

Disorders of sex development (DSD) encompass a large group of conditions which present with a wide spectrum of genital phenotypes. A common, isolated disorder like hypospadias poses no difficulty in gender assignment. Others, such as mixed gonadal dysgenesis, partial androgen insensitivity syndrome (PAIS) and some disorders of androgen biosynthesis require a multitude of biological, cultural and social considerations to be taken into account before a consensus can be reached on an appropriate gender assignment and sex of rearing. Members of a DSD multi-disciplinary team must be familiar with the concepts of gender role versus gender identity, their postnatal development and the influence of prenatal androgens. Certain conditions such as complete androgen insensitivity syndrome (CAIS) and congenital adrenal hyperplasia (infancy diagnosis), are usually irrefutably gender assigned female. In contrast, PAIS, mixed gonadal dysgenesis and 5α-reductase deficiency can be gender dimorphically assigned for which the parameters for collective decision-making will be discussed

2. Meet the Expert Session: Case-Based Discussion on Management of DSD

Fawzia Mandani, Elham Al Amiri, Rasha Tarif, Ieuan Hughes, Sleiman Gebran. Ieuan Hughes, Departments of Paediatric at Mafraq Hospital, Cairo University, Kuwait University and University of Cambridge, UK

Three cases were presented and they were followed by deep analysis and discussion:

a. **9 years old 46 XY girl with pure gonadal dysgenesis (Swyer syndrome)**
   A 9 year old Egyptian girl was noticed at birth to have mild ambiguity of genitalia (fusion of labia minora). She is the 3rd child born to a non-consanguineous healthy parents. A boy and a girl siblings are reported healthy. She is product of full term normal vaginal delivery, born after uneventful pregnancy and no had perinatal problems. Birth weight was 4.4 Kg and all other growth parameters were normal and she had no dysmorphic features. At day 3 of life, she developed urinary retention with a supra pubic mass felt just below the umbilicus.

This started a whole battery of investigations that ended by confirming the diagnosis of DSD 46 XY pure gonadal dysgenesis. The girl underwent gonadectomy at 3 years of age. She is under follow up for future hormone replacement therapy. Images and further clinical features were be discussed in the presentation and expert opinion will be sought for discussion.

b. An uncommon Case of Ambiguous Genitalia
The patient presented at the age of 1.6 years with ambiguous genitalia with palpable gonads. Genital examination revealed a microphallus, single opening with penoscrotal hypospadias, penile chordee, bifid scrotum, and bilateral undescended testes that were felt in inguinal areas. Laboratory profile was extensively studied and pelviabdominal ultrasound revealed testes which was confirmed by biopsy. There were no Mullerian structures and adrenals were normal. Phallic response to DHT was adequate and surgical correction was successfully done at the age of 1.8 years. The karyotype was conclusive and DNA analysis established the final diagnosis. The dilemma of contradiction between the Karyotype results with the clinical, radiological and the biochemical findings will be discussed.

c. A 46, XY fetus with female external genitalia
A 46,XY fetus with female external genitalia suggests different conditions, some very rare. A complete evaluation is mandatory after delivery to reach a correct diagnosis. Interpretation of hormone levels can be, in many times, difficult and inconclusive. Specific genetic investigations should be performed when possible. We present here a rare case of XY female baby with congenital adrenal hyperplasia, detected by the UAE national screening program to have high 17OHProgesterone. Later on, the baby developed salt loss and hyperkalemia. Genetic study helped to reveal the enzymatic defect but could not explain the complete normal female appearance of the external genitalia

Thyroid Update in Adults

1. Special Issues in the Management of Hypothyroidism in Adults
Salem A Beshyah, Center for Diabetes and Endocrinology, Sheikh Khalifa Medical City, Abu Dhabi, UAE.

Management of hypothyroidism encompasses three different levels. Recognition of hypothyroidism; finding the possible causes of hypothyroidism, confirming the diagnosis biochemically and deciding on the need for replacement therapy. It has been stated that in modern endocrine practice, we take no pride in parading features of gross endocrine conditions such as hypothyroidism. Overt hypothyroidism should be confirmed biochemically readily by measuring serum TSH and serum Thyroxine. Initiation and maintenance of Thyroxine replacement therapy should take place under the supervision of the primary care physicians by default, it may be possible to include it under the medical list if a patients is attending a general internal medical clinic for other reason. However, under special circumstances patient ought to be referred to endocrine specialist. The classical method of starting smaller doses and increasing it t slowly continue however, there are Compelling Scenarios when starting Thyroxine dose nearer to the average replacement dose: (1.6 ug/kg B Wt) such as pregnancy, soon post radioiodine and immediately post total thyroidectomy. Transient therapy for symptomatic thyroiditis should be clearly documented and explained to patients with clear documentation and communication to the patient regarding the need to stop the medication at some mutually agreeable timing with the patient. Extra care should be taken to avoid over replacement in elderly patients to avoid cardiovascular and possibly bone. Concomitant use of some medications particularly calcium and iron may interfere with absorption. It is now accepted that different bioavailability between brands may interfere with the consistency of replacement therapy. TSH measurements have no role in assessment of secondary hypothyroidism. In general, with sound knowledge, good clinical practice most patients with primary and secondary hypothyroidism may enjoy good and smooth replacement regimen. The small proportion that may encounter difficulties with replacement therapy should benefit from the expertise of the endocrinologists.

2. Controversies in Management of Graves' Disease
Pierre-Marc Bouloux, Centre for Neuroendocrinology, Royal Free Campus, University College Medical School, London UK.

Interactive Case-based Discussion - No abstract

3. Management of Thyroid Dysfunction During

Pregnancy and Post partum

Bashir Salih, Department of Obstetric Medicine, Corniche Hospital, Abu Dhabi, UAE.

Thyroid disorders are common in pregnancy. Both hypo and hyperthyroidism are associated with adverse pregnancy outcomes. Thyrotoxicosis has been implicated in infertility, miscarriage, still birth, pre- term delivery, IUGR & pre eclampsia. β-blockers and anti-thyroid drugs (Methimazole and Propylthiouracil) are the drug of choice in treating hyperthyroidism. Radioactive iodine is contra indicated due to fetal transfer. There is no role for block & replacement therapy. Thyroidectomy if indicated (no responsive to medical treatment, stridor, dysphasia, and carcinoma) should be done optimally in the 2nd trimester. Overt hypothyroidism is associated with increased rate of miscarriage, anemia, fetal loss, preeclampsia, low birth weight and reduced IQ in infants. Hence the need to replace with thyroxine aiming to normalize maternal serum TSH values within the trimester specific range. (First trimester 0.1 - 2.5, 2nd trimester 0.2 - 3 and 3rd trimester 0.3 - 3 mIU/L the ATA guidelines 2011) Postpartum thyroiditis is caused by destructive autoimmune lymphocytic thyroiditis and is associated with TPO antibodies. If TPO antibodies are present risk of thyroiditis is 50 – 80 %.Recurrence risk is about 70 % in future pregnancy. The initial thyrotoxic phase might be followed by euthyroid and hypothyroid phase .Most patients recover spontaneously. If diagnosed early and treated adequately, thyroid disorders in general are associated with good pregnancy outcome.

Clinical Management Challenges Symposium

1. Investigation and Perioperative care of Phaeochromocytoma

Pierre-Marc Bouloux, Centre for Neuroendocrinology, Royal Free Campus, University College Medical School, London UK.

Phaeochromocytoma is a rare catecholamine secreting neoplasm, occurring in less than 0.2% of patients with hypertension. When extra-adrenal, the term paraganglioma is used. The distinction between the two is important because of implications for associated neoplasms, risk of malignancy, and genetic testing. Hypertension associated with the classical triad of episodic headache, sweating and palpitations should evoke suspicion of an underlying diagnosis of catecholamine secreting tumor. About 50% of patients have episodic hypertension. The clinician should also suspect the diagnosis in patients with resistant hypertension, an associated familial syndrome (e.g. MEN2, NF1, VHL), an incidentally discovered adrenal mass, hypertension and new onset or atypical diabetes mellitus, a presser response during anesthesia, surgery or angiography, onset of hypertension at a young age, idiopathic dilated cardiomyopathy, and a history of gastric stromal tumor or pulmonary chondroma (Carney triad).

Diagnosis is best confirmed by measurements of urinary and fractionated metanephrines and catecholamines. The majority of metabolism of catecholamines is intratumoral, with formation of metanephrine and normetanephrine. These may be measured in the urine using 24 hour collection, corrected for completeness by measuring the creatinine. A sensitivity and specificity of 98% has been reported with this diagnostic approach. If clinical suspicion is high, fractionated plasma metanephrines should be estimated, since the test is simple and the predictive value of a negative test is very high. However, the specificity is relatively poor. Tricyclic antidepressants interfere most commonly with the interpretation of urinary catecholamines, and ideally measurements should only be carried out in such patients after 2 weeks discontinuation of the drug. It should be noted that catecholamine secretion can be appropriately increased in patients following stroke, myocardial infarction congestive heart failure and obstructive sleep apnoea. Levodopa interferes with the measurement of urinary dopamine. Biochemical confirmation should be followed by imaging. 10% tumours are extra adrenal , but 95% within the abdomen and pelvis. paragangliomas are in 75% cases in the superior or inferior paraaortic areas. Both CT and MRI have a 95% + sensitivity, but given the high incidence of adrenal incidentaloma, only 70% specific, and low osmolar iv contrast can be given safely with CT, even in the absence of alpha and beta blockade. 111-In-pentreotide Octreoscan) may occasionally reveal an unusually sited lesion (e.g. cardiac). 123-I mIBG scanning is useful in patients with large (>10cm) adrenal tumors (high malignancy risk) and FDG PET is useful in identifying metastatic activity. Preoperative medical therapy is aimed at controlling hypertension, and volume expansion. Alpha blockade is given several weeks before surgery, and the non- competitive inhibitor phenoxybenzamine the preferred agent, at an initial dose of 10mg bid, the final dose typically 20-100mg per daub in divided doses. beta blockade may be commenced 3 days after the start of alpha blockade. Treatment consolidation with 3 days of preoperative IV phenoxybenzamine (0.5mg/Kg) is recommended by some.

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2. Vitamin D Deficiency and Bone Health in the Middle East:

Hussein F Saadi, Department of Medical Subspecialties, Cleveland Clinic Abu Dhabi, Abu Dhabi, UAE.

Vitamin D plays an essential role in maintaining a healthy mineralized skeleton for most land vertebrates including humans. Sunlight causes the photoproduction of vitamin D3 in the skin. Once formed, vitamin D3 is metabolized sequentially in the liver and kidney to 1,25-dihydroxyvitamin D. The major biological function of 1,25-dihydroxyvitamin D is to keep the serum calcium and phosphorus concentrations within the normal range to maintain essential cellular functions and to promote mineralization of the skeleton. Most foods do not contain any vitamin D. Foods fortified with vitamin D have a variable amount present and cannot be depended on as a sole source of vitamin D nutrition. Exposure to sunlight provides most humans with their vitamin D requirement. Aging, sunscreen use and the change in the zenith angle of the sun can dramatically affect the cutaneous production of vitamin D3. Vitamin D insufficiency and vitamin D deficiency is now being recognized as a major cause of metabolic bone disease in the elderly. Vitamin D deficiency not only causes osteomalacia but can exacerbate osteoporosis. It is generally accepted that an increase in calcium intake to 1000-1500 mg/d along with an adequate source of vitamin D of at least 400 IU/d is important for maintaining good bone health. Low levels of vitamin D have been linked to many chronic diseases. Decreased muscle function and increased fall risk in elderly people; prostate, breast and colorectal cancers; diabetes mellitus; and other health problems have been associated to low circulating levels of 25-hydroxyvitamin D. This paper presents an overview of the available scientific evidence for the non-calcemic actions of vitamin D in humans. There are lots of important non-skeletal systemic consequence of vitamin D deficiency, which was often, was underestimated. The purpose of this review is to discuss the non-skeletal systemic manifestations of vitamin D deficiency. These include effects on the cardiovascular system, immune system, reduction of malignancy etc.

3. Investigation of the Non-diabetic Hypoglycemia in Children and Adults; An Overview.

Khalid Hussain, Great Ormond Street, London, UK.

Hypoglycemia is one of the most common biochemical abnormalities observed in clinical practice. Thus an understanding of the mechanisms that lead to hypoglycemia is important for patient management. Over the last a few years there have been tremendous advances in understanding the molecular and biochemical basis of some types of hemia. In the neonatal, infancy and childhood periods the most common cause of severe and persistent hypoglycemia is congenital hyperinsulinism (CHI). Hypoglycemia may also be a manifestation of ACTH, cortisol and growth hormone deficiency either in isolation or as part of a broader picture of hypopituitarism. A large number of metabolic diseases will lead to hypoglycemia but in these cases there will always be clues form the biochemistry. Although an insulinoma is the commonest cause of hyperinsulinemic hypoglyc emia in adults, postprandial hyperinsulinemic hypoglycemia has been reported in patients with non-insulinoma pancreatogenous hypoglycemia, following gastric bypass surgery for morbid obesity and in patients with mutations in the insulin receptor gene. Insulin autoimmune syndrome is the commonest cause of hypoglycemia in Japan. Some mesenchymal tumours produce "big IGFII" which has insulin like action leading to severe hypoglycaemia. During this talk I will outline the causes of hypoglycemia children and adults and discuss the investigations.

II. Special Symposia

1. Multi-Hormonal Therapy of Type 1 Diabetes.

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Insulin deficiency is generally considered the sine qua non of type 1 diabetes and insulin replacement is the fundamental therapy for type 1 diabetes. Recent studies have evaluated the roles of other hormones in type 1 diabetes. For example, there is inappropriate glucagon secretion with meals, and lack of glucagon response to hypoglycemia. Also, in addition to insulin deficiency, there is deficiency of C-peptide and amylin secretion, since C-peptide and amylin are produced in the pancreatic islet beta cell and secreted along with

insulin. Therefore, simple insulin replacement therapy is not sufficient to correct all of the metabolic defects. Amylin replacement may be desirable. In addition, C-peptide may have beneficial effects including limiting the development of diabetic complications. Inhibition of inappropriate glucagon secretion also may be desirable. It turns out that the adipocyte hormone leptin suppresses inappropriate glucagon secretion and stabilizes glycemic control in type 1 diabetes. Several studies also have demonstrated that the GLP-1 receptor agonists exenatide and liraglutide may also improve glycemic control in type 1 diabetes, in part by modulating glucagon secretion. Attention to other hormones may improve both glycemic control and the lives of people with type 1 diabetes

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2. Acromegaly and Gigantism in History and Culture

Salem A Beshyah, Center for Diabetes and Endocrinology, Sheikh Khalifa Medical City, Abu Dhabi, UAE.

Pierre Marie described acromegaly as a medical syndrome in 1886. The causative role of the pituitary gland was not recognized at that time, however, and the enlarged pituitaries found at autopsy were thought by many to be just another feature of the hypertrophy that affected many parts of the body. By 1900, pathophysiologic correlations by Benda and others had suggested that the hypertrophy or hyperplasia of the pituitary might be the cause of acromegaly, and the adenomatous nature of pituitary enlargement was ultimately documented. Acromegalic persons and giants have been described in literature and art throughout history. The unfortunate people afflicted with this condition have been held in awe, and enormous strength and physical powers have been attributed to them, although these are rarely present, at least for any extended period of time. When looking at endocrinology in international art work, one remembers the endocrinologist' interpretation of David victorious over Goliath. In modern days, years many giants and acromegalics were either ridiculed or made a fortune by virtue of their physical features attributed to giagantism and acromegaly. A few of these examples will be discussed with view to enhancing the audience knowledge of clinical features of acromegaly and remembering the clinical feature when they face the next patients with acrpmegaly and they will make that first step to early diagnosis and timely management with (hopefully) a better outcome.

3. Acromegaly 2012: Timely Clinical Recognition and Medical Management

Ali B Khalil, Center for Diabetes and Endocrinology, Sheikh Khalifa Medical City, Abu Dhabi, UAE.

Acromegaly is a long-term condition in which there is too much growth hormone (GH) and the body tissues get larger over time. Acromegaly occurs in about 6 of every 100,000 adults. The cause of the increased GH release is usually a noncancerous (benign) tumor of the pituitary gland but ectopic production may also occur. The presenting symptoms reflect the effects on the skeletal and non-skeletal tissues and metabolic effects of GH excess. Early recognition is very important. The diagnosis is suggested by positive screening tests such as very high serum IGF-1 and is established by lack of suppression of GH secretion during oral glucose tolerance. Source of excessive GH secretion is established by the usual pituitary imaging methods mainly MRI scanning with contrast. The primary therapy is by pituitary surgery. Radiotherapy has a role when surgery could not remove the whole tumor. Medical therapies mainly by Octreotide or Bromocriptine work by mainly control of GH release. Pegvisomant directly blocks the effects of GH and has been shown to improve symptoms of acromegaly. These medications may be used before surgery, after surgery, or when surgery is not possible. Patients with treated acromegaly needs lifelong follow up.

4. Transcriptional regulation of pituitary development: Clinical implications.

Jan Lebl, Department of Pediatrics, Faculty of Medicine, University of Prague, Hungary

Embryonic and fetal development of human anterior pituitary is governed by a cascade of transcription factors. At the early stage, they orchestrate morphogenesis not only of pituitary, but also of eyes, optic nerves, and both facial and cerebral mid-line structures. Later, transcriptional factors regulate the differentiation of pluripotent pituitary cells into five specific cell lineages of the anterior pituitary – corticotrophs, gonadotrophs, thyreotrophs, somatotrophs, and lactotrophs, allowing the life-long physiological hormonal production. Therefore, defects in the "early factors" result mostly in dysmorphic syndromes with hypopituitarism, whereas defects in the "late factors" lead to a phenotype of combined pituitary hormone deficiency. The "sonic hedgehog cascade" (genes SHH, GLI2, PATCH) is expressed already in gestational weeks 3-5, at the stage of morphogenesis of human forbrain. Defects in this cascade result in holoprosencephaly, a spectrum of forebrain malformations characterized by failure of the prosencephalon to form two lateral hemispheres. Holoprosencephaly is associated with variable endocrine phenotype. OTX2 gene defects are typically linked to anophthalmia and hypopituitarism. Septo-optic dysplasia (mid-line defects, optic nerve hypoplasia and hypopituitarism) may result from HESX1 defects, but also from defects in the FGFR1/FGF8/PROKR2 network, and, rarely, from OTX2, SOX2 or SOX3 mutations. Furthermore, mutations in the "late" transcriptional factors PROP1, POU1F1, LHX3 and LHX4 underlie combined pituitary hormone deficiency (CPHD) without major dysmorphic features, allowing etiological explanation of about 25% cases of "idiopathic" CPHD. The identification of genetic cause of hypopituitarism in a pediatric patient allows not only genetic counselling in affected family and prenatal diagnosis in most severe defects, but also clinical prediction of risk of hormonal deficiencies (in holoprosencephaly and septo-optic dysplasia) or of evolving hormonal phenotype within the life-span (e.g. ACTH deficiency in PROP1 defect in adolescence or early adulthood). Besides, genetic finding of a PROP1 defect may confirm benign nature of a pituitary mass that may be described as a large pituitary adenoma by a radiologist and might lead to unnecessary neurosurgical intervention.

5. How to Properly Count Carbs

Laila King, London Medical, 49 Marylebone High Street London, UK.

Carbohydrate counting is a meal-planning approach that focuses on carbohydrate as the main nutrient resulting in post-meal hyperglycaemia. Since the 1980s, training courses, such as DAFNE (Dose Adjustment for Normal Eating), have been developed with the hope of teaching people with diabetes how to estimate carbohydrate grams in their meals and snacks, and adjust their meal insulin doses accordingly aiming 'to think like a pancreas'. In the next 60 minutes, an attempt is made to briefly describe the main methods of carbohydrate counting, including (1) carbohydrate exchange method, (2) nutritional labels, (3) carb factors and weighing scales, (4) carbohydrate reference lists, and (5) eyeballing or visualising carbohydrate portions. Carbohydrate counting can be broken down into BASIC and ADVANCED carb counting. The latter involves using algorithmic Insulin to Carbohydrate Ratios (ICRs) and Insulin Sensitivity Factors (ISFs) tailored to the individual's target glucose levels. Disappointingly, this kind of time-consuming and labour-intensive approach has not been shown to result in any significant reduction in HbA1c, hypo- or hyperglycaemia, or improved quality of life. This could be simply explained by the inadequacy of such a simplistic one-hormone-based strategy to only one of the macronutrients, ignoring the effect of fat and protein in mixed meals, whilst forgetting the plethora of secondary factors that influence diurnal blood glucose levels.

Even the most determined and conscientious carb counters have found their attempts of matching insulin to every mouthful of food in real life unsuccessful and recognised that simply lowering the amount of the nutrient, mainly responsible for unpredictable glucose spikes, makes their life easier and gives them better glycaemic control. Paradoxically, American Diabetes Association, Diabetes UK, and various national diabetes nutrition guidelines continue to recommend that people with diabetes 'base all their meals on starchy carbohydrates'. When you understand the complexities of carbohydrate counting, ask yourself if you need to learn it or not, and whether perpetuating the still-prevalent message of high-carbohydrate diet is in the best interests of your pancreatically challenged patients. "Everything should be made as simple as possible, but no simpler.' (AEinstein)

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6. Day-to-Day Self-Management of Type 1 Diabetes (Friday PM)

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Diabetes is both a life-long and a 24/7 condition. Glucose control and its short- and long-term effects are almost entirely in the hands of the person who lives with it. The individual's motivation to eat healthily, exercise, take insulin, test glucose levels and respond to any deviations from target, and maintain a normal body weight, all compete with life's other demands and motivations. It has been calculated that a person with Type 1 diabetes would be required to carry out 33 discrete self-care episodes, taking two and half hours (142 minutes) daily. This does not even include exercise for at least 30 minutes, daily foot check, extra blood glucose testing before driving, carrying supplies of hypo treatments and diabetes ID, and ketone testing equipment just in case of emergency. Not surprisingly, few people with Type 1 diabetes manage to adhere to such rigors of daily self-care. According to the National Diabetes Audit 2010 in the UK, only 16% of Type 1 diabetes children and adolescents achieve HbA1c level of ≤ 7.5% (58mmol/mol) whereas in Germany over 72% manage that level. Burn-out happens inevitably when the individual and/or their family member realize the unattainability of tight glycemic control regardless of the daily sacrifice of time and effort.

Self-management of a chronic illness develops into an "illness career" in which the person learns to respond to health changes, relationships with health care professionals, and psychosocial aspects of the disease. The terms "self-care", "self-management" and "decision-making" are used increasingly by health care professionals – reflecting a healthy shift towards valuing the patient's responsibility and autonomy in their own care. However, this must not be interpreted to mean that every chronically ill person is strong, competent and empowered, ignoring the continuing need many people have for professional expertise, empathy and support.

The space between being acutely ill and being well is where most people with Type 1 diabetes dwell, trying to "find balance" and "minimize the intrusiveness of the disease". To be "normal" within the abnormality of the disease includes learning what works best, dealing constantly with a perceived threat – not infrequently leading to a conscious, rational decision to keep blood glucose level higher than recommended. Mastering diabetes day-to-day self-management has been aptly likened to the relationship between a dog and his master: if you can discipline it, keeping it balanced in order to let it follow you, without being scared, without letting it control you, then you feel better. Those who manage this have mastered their disease, adding "life to years" and not just "years to life".

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Final Remarks

We hope by publishing these abstracts we extend the education cause of the conference.

We would like to take the opportunity to thank all faculty of the conference (speakers and moderators) and all the sponsors whose unconditional educational sponsorship made the conference possible.