high salt diet affected methylation status of 11-HSD1 in the adipose tissue but not 11-HSD2 gene in the kidney in SSH. Food intake such as salt may influence the epigenesis of 11-HSD and induce hypertension.

# Diabetes Mellitus and Glucose Metabolism

## LIPIDS, OBESITY AND METABOLIC DISEASE

### *Associations of Serum and CSF Kisspeptin Levels with Metabolic and Reproductive Parameters in Men*

*Pornthira Mutirangura, MD, Chantacha Sitticharoon, MD., PhD., Thitima Chinachoti, MD,FRCAT., M.Sc.(Clinical Epidemiology), Arissara Iamaroon, MD, Namtip Triyasunant, MD, Malika Churintaraphan, MSc, Issarawan Keadkraichaiwat, BSc, Pailin Maikaew, BSc.*
Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand.

**SAT-658**

Action of kisspeptin in the central nervous system (CNS) is well known on reproductive regulation; however, its peripheral action is not well understood. Recent studies have shown that peripheral kisspeptin might be related to obesity and/or metabolic parameters in humans [1]; however, these associations are still inconclusive. This study aimed to 1) compare serum or cerebrospinal fluid (CSF) kisspeptin levels between different body mass index (BMI) groups 2) compare levels of kisspeptin between serum and CSF, and 3) determine correlations between serum or CSF kisspeptin levels with clinical, metabolic, and reproductive parameters. There were 40 male subjects who underwent an operation with lumbar puncture anesthesia. Subgroup analysis was performed to compare between the lean-normal group (n=13) which included lean and normal weight subjects, the overweight group (n=10), and the obese group (n=17) according to BMI. Blood samples were collected after at least 8-hour fasting before intravenous cannulation prior to the operation while CSF samples were obtained by lumbar puncture before administration of the spinal anesthesia. Serum kisspeptin and leptin levels were significantly higher in the obese group when compared to the lean-normal and overweight groups even after adjusted to age while CSF kisspeptin levels were comparable between different BMI groups ($p<0.05$ all). Serum kisspeptin levels were significantly higher than CSF kisspeptin levels ($p<0.001$). Serum kisspeptin was significantly positively correlated with body weight ($R= 0.351$), BMI ($R=0.549$), plasma insulin ($R=0.393$), and serum leptin ($R=0.45$) ($p<0.05$ all), and tended to have a positive correlation with the Homeostatic Model Assessment of Insulin Resistance (HOMA-IR) ($R=0.29$, $p=0.77$) but was significantly negatively correlated with plasma LH ($R=-0.37$) ($p<0.05$). CSF kisspeptin was significantly positively correlated with plasma LH ($R=0.452$, $p<0.05$). These results suggest that serum kisspeptin levels were related to increased obesity, leptin, insulin, and insulin resistance while CSF kisspeptin levels were related to reproductive parameters. In summary, central kisspeptin might have a role on reproductive regulation while peripheral kisspeptin might have a role on metabolic regulation. **Reference:** (1) Izzi-Engbeaya, C., et al., The effects of kisspeptin on beta-cell function, serum metabolites and appetite in humans. Diabetes Obes Metab, 2018. 20(12): p. 2800–2810.

# Neuroendocrinology and Pituitary

## NEUROENDOCRINE & PITUITARY PATHOLOGIES

### *Central Adrenal Insufficiency Is Rare in Adults with Prader-Willi Syndrome*

*Anna G.W. Rosenberg, BSc[1], Karlijn Pellikaan, BSc[1], Christine Poitou, MD, PhD[2], Anthony P. Goldstone, PHD,BCH,MRCP,MB[3], Charlotte Hoybye, MD,PHD[4], Tania Markovic, MBBS,PHD,FRACP[5], Graziano Grugni, MD, PhD[6], Antonino Crinò, MD, PhD[7], Assumpta Caixàs, MD, PhD[8], Muriel Coupaye, MD[2], Sjoerd A.A. van den Berg, PhD[1], Aart Jan van der Lely, MD, PhD[1], Laura C.G. de Graaff-Herder, MD, PhD[1].*
[1]Erasmus University Medical Center, Rotterdam, Netherlands, [2]Assistance Publique-Hôpitaux de Paris, Paris, France, [3]Imperial College London, London, United Kingdom, [4]Karolinska Hospital, Stockholm, Sweden, [5]Royal Prince Alfred Hospital, Bellevue Hill NSW, Australia, [6]Italian Auxological Institute, Piancavallo (VB), Italy, [7]Bambino Gesù Hospital, Palidoro (Rome), Italy, [8]Hospital Universitari Parc Taulí (UAB), Sabadell, Spain.

**SUN-308**

**Introduction:** Prader-Willi syndrome (PWS) is associated with several hypothalamic-pituitary hormone deficiencies. There is no agreement on the prevalence of central adrenal insufficiency (CAI) in adults with PWS. This is partly due to the variable results of the synacthen test, compared with the more robust metyrapone test (MTP) and insulin tolerance test (ITT). In some countries, patients with PWS receive stress-dose corticosteroids during physical or psychological stress. Side effects of frequent corticosteroids use are weight gain, osteoporosis, diabetes mellitus and hypertension, already major problems in adults with PWS. However, undertreatment of CAI can cause significant morbidity or even mortality. To prevent over- and undertreatment with corticosteroids, we assessed the prevalence of CAI in a large international cohort of adults with this rare disorder.
**Methods:** The hypothalamic-pituitary-adrenal axis was tested in 81 adult subjects (55 Dutch, 10 British, 10 French, 6 Swedish) with genetically confirmed PWS. For multiple-dose MTP, 11-deoxycortisol >230 nmol/L (7.6 g/dL) was considered sufficient. For Dutch, French and Swedish patients who underwent ITT, cortisol >500 nmol/L (18.1 µg/dL) was considered sufficient. For British patients cortisol >450 nmol/L (16.3 µg/dL) was considered sufficient, as this center used a different assay. Additionally, we reviewed medical files of 645 adults with PWS from Italy (240), France (110), the Netherlands (110), Australia (60), Spain (45), Sweden (38) and the United Kingdom (42) for symptoms of hypocortisolism/adrenal crisis during surgery.
**Results:** Data on 81 adult subjects (46 males and 35 females), median age (range) 25.2 yr (18.0 – 55.5), median BMI (range) 29.1 kg/m$^2$ (20.0 – 62.0), with genetically confirmed PWS were collected. 33 subjects (41%) were using GH treatment since childhood. Multiple-dose MTP was performed in 45 subjects and ITT in 36 subjects. Both tests

were well tolerated by all individuals. CAI was excluded in 80 of 81 patients. One patient with a peak cortisol level of 494 nmol/L (just below cut-off level of 500 nmol/L) was prescribed hydrocortisone for use during physical stress. There was no relation between baseline cortisol and ITT/multiple-dose MTP results. Even patients with a low baseline cortisol level (lowest: 102.0 nmol/L) had normal responses. Among the 645 patients whose medical files were reviewed, 200 had undergone surgery without perioperative corticosteroids treatment. None of them displayed any features of hypocortisolism/adrenal crisis.
**Conclusions:** CAI is rare (1.2%) in adults with PWS. Based on these results, we recommend against routinely prescribing corticosteroids stress-doses in adults with PWS.
**Funding:** CZ foundation.

# Diabetes Mellitus and Glucose Metabolism

## TYPE 2 DIABETES MELLITUS

### *Management of Hyperglycemia in an Adult Patient with Glycogen Storage Disease Type 1b*

*Alexandra Mikhael, MD, Robert S. Zimmerman, MD.*
Cleveland Clinic, Cleveland, OH, USA.

**SUN-698**

Background: Glycogen storage disease type 1b (GSD1b) is caused by a deficiency of glucose 6 phosphatase leading to glycogen deposition. The hallmark findings of GSD1b are hypoglycemia and lactic acidosis. GSD1b can be associated with hepatomegaly, hypertriglyceridemia, hematologic abnormalities, hypothyroidism, inflammatory bowel disease, proteinuria, and hypoglycemic seizures. Main stay of therapy is to avoid fasting and to ingest frequent feeds high in complex CHO. Raw cornstarch (CS) has been used for the treatment of hypoglycemia in GSD1b since the early 1980s. CS is digested slowly, providing a steady release of glucose allowing for more stable glucose levels over a longer period of time as compared with other sources of CHO. Adults may require more CS to maintain BG >70 mg/dL and lactate <2mmol/L during the night. We report a case of an adult patient with GSD1b admitted with inability to tolerate oral intake and found to have persistent hyperglycemia on admission requiring insulin therapy.
Clinical Case: A 31 year old female with a history of GSD1b complicated by hypoglycemia, Crohn's disease, chronic pancreatitis and neutropenia was admitted for abdominal pain, emesis and inability to tolerate CS. She was made NPO and was started on a dextrose drip to avoid hypoglycemia and hyperlactatemia. The rate of dextrose infusion was adjusted to maintain lactate levels <2 mmol/L. She developed persistent hyperglycemia with glucose values of 250–350 mg/dL. Laboratory evaluation revealed an HbA1c of 7.6% (reference [ref] 4.3–5.6%), C-peptide of 1.3 ng/mL (ref 0.8–3.9 ng/mL), lactate of 2.3–3.1 mmol/L (ref 0.5–2.2 mmol/L). BHB levels were normal (WNL). Anti-GAD, insulin and islet cell antibodies were negative. The main goal was to avoid hypoglycemia while keeping ketone and lactate levels WNL. The decision was made to start a regular insulin infusion at a constant rate of 1 u/hr to keep BG <180 mg/dL while on the dextrose infusion with close monitoring of lactate and BHB. Insulin and dextrose drip rates were adjusted based on BG. The ultimate goal was to determine total daily insulin requirements while ingesting her home CS doses and to transition to long acting insulin. Short acting insulin boluses were not used given risk of hypoglycemia. The patient's unpredictable tolerance to CS made determining a fixed insulin dose challenging. She eventually managed to tolerate CS and was transitioned to 10 units of insulin Detemir twice daily. She was discharged with plans to get a CGM and a ketone meter. BG readings at home were between 140–170 mg/dL.
Conclusion: We report a rare case of GSD1b and diabetes. The pathogenesis may be related to effects of chronic pancreatitis or metabolic syndrome. Treatment of hyperglycemia in patients with GSD1b is challenging given the heightened risk of fasting hypoglycemia. Treatment options are limited, and there are no data regarding the safe use of insulin in this patient population.

# Thyroid

## THYROID DISORDERS CASE REPORTS III

### *Dual Ectopic Thyroid with Normally Located Thyroid Gland in a Patient Presenting with Subclinical Hypothyroidism: A Case Report*

*Stephen M Rueda, MD[1], Mark H Joven, MD[1], Nixon See, MD[2], Hossein Gharib, MD[3].*
[1]University of Santo Tomas Faculty of Medicine and Surgery, Manila, Philippines, [2]Our Lady of Fatima University College of Medicine, Valenzuela, Philippines, [3]Mayo Clinic, Rochester, MN, USA.

**MON-466**

**Background:** Ectopic thyroid involves the presence of thyroid tissue in sites other than its normal anatomic location. Dual ectopic thyroid with normally located thyroid gland is extremely rare with only a few cases reported in the literature.
**Clinical Case:** A 24-year-old female presented to the clinic with subclinical hypothyroidism (TSH of 10.54 uIU/ml [ref, 0.27–4.2] and free T4 of 12.96 pmol/L [ref, 12–22]). She noted a gradually enlarging submental mass during the preceding year, which had been present since her adolescent years.
Examination was notable for a 2-cm firm, non-tender mass at the submental region that slightly moved with deglutition. The thyroid was not palpable in its normal location. The relaxation phase of the deep tendon reflex was prolonged. Repeat laboratory testing showed persistently elevated TSH of 13.3 uIU/ml and normal free T4 of 13.39 pmol/L. TPO antibody was normal. Thyroid ultrasonography showed a hypoplastic thyroid measuring 2.7 x 0.7 x 0.4 cm on the right and 3.0 x 0.7 x 0.4 cm on the left. A well-circumscribed complex nodule measuring 2.7 x 3.2 x 2.1 cm was noted on the submental region corresponding to the patient's submental mass. Computed tomography of the neck with contrast showed two hyperdense foci - measuring 3.0 x 2.4 cm at the submental region and 1.4 x 1.2 cm at the base of the tongue. No enhancing thyroid tissue was seen anterior to the thyroid cartilage. $^{99m}$Technetium-pertechnetate scan showed absent focal tracer uptake in the anterior neck and thorax. There were foci of increased tracer activity in the submental and lingual regions.