syndrome such as tall stature, hypogonadism, congenital malformations and neurocognitive issues. Hypogonadism may cause abdominal adiposity hence increasing risk of insulin resistance and type 2 diabetes mellitus (DM). Klinefelter syndrome has been associated with certain autoimmune diseases however there is no autoimmune disease link described with XXYY syndrome. We present a patient with XXYY polisomy with insulin requiring DM.**Clinical Case:** A 26-year-old intellectually disabled male with history of XXYY polisomy presented with seizures and was evaluated for DM management. At age 18, he was noted to have hyperglycemia while undergoing dental extractions for brittle enamel leading to the diagnosis of DM. He was initiated on oral medications for a short period of time and transitioned to insulin due to significant hyperglycemia. Endocrine evaluation at the time also revealed hypergonadotropic hypogonadism and evaluation for Klinefelter syndrome revealed 48 XXYY aneuploidy. Patient’s history was significant for poorly controlled hyperglycemia with HbA1C ranging in 12–14% range in the last few years. History was negative for diabetic ketoacidosis (DKA) as per his primary endocrinologist, despite being noncompliant with his insulin therapy. History was negative for retinopathy, nephropathy or macrovascular complications, although he did have distal extremity paresthesia. On exam, he was noted to be edentulous with tall stature, BMI 21.7, facial dysmorphism, pes planus and 5th-digit clinodactyly bilaterally. Family history was positive for type 2 DM in father and prediabetes in mother. He presented with seizures and was diagnosed with brain abscess. Hyperglycemia was initially managed with IV insulin, followed by basal/bolus therapy. Fasting labs 20 hours after receiving insulin glargine showed glucose of 284 mg/dL, C-peptide 0.6 ng/mL (ref range: 0.8–3.5), proinsulin less than 1.6 pmol/L (less than 8.0), insulin 4 uIU/mL (3–19) suggestive of type 1 DM. Anti-GAD65 and anti-ICA antibody levels were negative. Additional antibody evaluation for type 1 DM (islet antigen 2, insulin autoantibody, micro-insulin and zinc transporter 8) is currently pending at the time of writing. **Conclusion:** Prevalence of XXYY syndrome is 1:18 000–1:40 000 in males. There are few case reports describing the association of type 2 DM with this syndrome. Our patient’s low BMI, low insulin and C-peptide with hyperglycemia indicate type 1 DM, although the absence of DKA in the setting of noncompliance suggests residual beta cell function or maturity onset diabetes of the young (MODY). Neither T1DM nor MODY has been reported with 48XXYY previously. Clinicians should be aware of this association as it has implications in terms of management of DM.

**Diabetes Mellitus and Glucose Metabolism**

**ISLETS AND INSULIN SECRETION**

**A Novel Population of FOXO1-Expressing Cells in the Stomach Controls Cell Plasticity by Regulating the Cyclin CCNE1**

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**SUN-646**

A promising new therapy for type 1 diabetes is the reprogramming of gut enteroendocrine cells into cells that produce insulin. The mechanism by which gut epithelial cells are converted into cells that make insulin remains unknown. We have previously found that elimination of Foxo1 in neurogenin3 (Ngn3)-expressing cells of the intestine generates glucose-sensing, insulin-producing cells that are capable of reversing streptozotocin-induced diabetes. Others have reported that stomach cells have a similar property when made to express β-cell factors Ngn3, Pdx1, and MafA. Using mice bearing a Foxo1-GFP knock-in allele, we traced Foxo1-expressing cells in the gut to subpopulations of Ngn3+, as well as acid-secreting parietal stomach cells. To study these cells, we established a 2D co-culture method in which primary stomach cells are isolated from mice and cultured with embryonic fibroblasts. Deletion of Foxo1 in this system generated cells immunoreactive for insulin and C-peptide. Interestingly, Foxo1 ablation also altered the abundance of other gastric cell populations, including more parietal cells and decreased expression of stem cell marker, Lgr5. Tissue-specific elimination of Foxo1 in vivo in either Ngn3+ or parietal cells also resulted in the appearance of insulin+ cells, increased parietal cells, and reduced Lgr5 mRNA. To determine how Foxo1 regulated these changes, we used cells isolated from reporter mice that change from red to green after genetic recombination to collect Foxo1-deleted primary stomach cells using FACS. While the mRNA levels of many known Foxo1 targets did not change, cyclin E1 (CCNE1), which regulates G1 to S-phase progression of the cell cycle, was significantly decreased. Conversely, primary stomach cells overexpressing Foxo1 had increased levels of CCNE1. Finally, using ChIP-seq, we found that Foxo1 binds directly to the CCNE promoter in a nutrient-dependent manner. In summary, we show that Foxo1 is expressed in a subpopulation of stomach parietal cells and that it regulates their function through the cell cycle regulator, CCNE1.

**Pediatric Endocrinology**

**PEDIATRIC ENDOCRINE CASE REPORTS I**

**Maternal Transmission of Pituitary Stalk Interruption Syndrome (PSIS)**

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**SAT-067**

**Background:** Pituitary stalk interruption syndrome (PSIS) is a rare entity characterized by thin or absent pituitary stalk, hypoplastic/aplastic anterior pituitary and ectopic posterior pituitary (EPP) on magnetic resonance imaging (MRI). PSIS can be associated with variable degrees of pituitary insufficiency 1. Most cases of combined pituitary hormone deficiency are sporadic, however in familial cases, there can be AD or AR inheritance with more than 30 genes identified in association with combined pituitary hormone deficiency (CPHD). We describe how diagnosis of 2 children with PSIS led to the discovery of the condition in their mother.
Clinical Case: Child 1 presented at age 3yrs with growth failure in 2003 with ht z score -4.24 SD. Subsequent work up revealed low IGF-1 (< 25 ng/mL) and MRI showed EPP, small anterior pituitary gland and absent pituitary stalk. No GH stim test was performed. He was started on GH supplementation and later was diagnosed with central hypothyroidism, central adrenal insufficiency and hypogonadotropic hypogonadism and is doing well on multiple hormone replacement at age 19 yrs. Child 2, a half-brother to child 1 (same mother), presented at age 1yr with growth failure in 2017 with ht z score -2.06. GH stimulation test with glucagon was abnormal and resulted in a very low GH response (peak GH 0.52 ng/mL). MRI showed EPP with small anterior pituitary gland and interruption of the stalk. Later he was found to have central hypothyroidism and mild central adrenal insufficiency. He is receiving standard hormone replacement at 3 yrs of age. Mother of above 2 patients presented 6 mos postpartum in 2017 after her 7th and last pregnancy with fatigue and amenorrhea. Laboratory evaluation revealed central hypothyroidism (FT4 0.76 ng/dL) and she was prescribed levothyroxine followed by resumption of her menses. She was unable to breastfeed her children due to lack of supply. There were no concerns for DI, amenorrhea or infertility. She was referred to Endocrinology in 2019 for persistent fatigue with a question of GH deficiency. IGF-1 level was normal 114 ng/mL(z score -0.39) and GH stimulation test (clonidine + glucagon) was abnormal with peak GH 1.85 ng/mL. MRI showed EPP with hypoplastic pituitary stalk. Genetic testing was done for CPHD Sequencing Panel at Prevention Genetics which includes GL12, HESX1, LHX3, LHX4, OTX2, POU1F1, PROP1F1, PROP1, SOX2, SOX3 genes and results were negative. She has 4 other children (21, 12, 11, 10yrs) who are currently being investigated for hormone deficiencies. One child died at 3 months of age due to SIDS. Conclusion: We present 3 family members with PSIS. This family highlights the variable clinical phenotype of PSIS and importance of careful family history when evaluating children with congenital pituitary abnormalities and supports the need for more extensive gene panels for evaluation of CPHD. Reference: Acta Endocrinologica, 2017. 13(1):96–105

Bone and Mineral Metabolism

BONE AND MINERAL CASE REPORTS I

An Unusual Case of Brown Tumor in the Left Femur Associated with Secondary Hyperparathyroidism

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SAT-338

Background: Brown tumor of hyperparathyroidism (BTHPT) are non-neoplastic bony lesions that arise from increased osteoclastic activity in the setting of uncontrolled hyperparathyroidism, (HPT). The prevalence is low, with a frequency of 3% in primary HPT and 2% in secondary HPT. It is rare as earlier detection and treatment of HPT prevents the progression and development of BTHPT. BTHPT is more common when HPT is untreated. BTHPT are benign tumors which usually show high 18Fluorodeoxyglucose-PET/CT (FDG) uptake. The mechanism of elevated FDG uptake has been suggested to be the presence of giant cells, and intracellular glucose metabolism of the macrophages may also play a role. We describe a case of a patient who presents with FDG uptake-negative BTHPT of the left femur.

Clinical Case: 62-year-old male with end-stage renal disease (ESRD) and secondary HPT presented with severe left hip pain for many months. The patient had poor access to health care and had not received routine medical treatments at an outside facility prior to presentation at our facility. Laboratory studies showed PTH 1942 pg/ml (14–72), Calcium 8.7 ng/dl (8.4–10.2), phosphorus 3.6 mg/dl (2.5–4.9), Vitamin D total 19.7 ng/ml (30–140), Vit D 1,25 9 pg/ml (14–72), Calcium 8.7 ng/dl (8.4–10.2), phosphorus 3.6 mg/dl (2.5–4.9), alkaline phosphatase 57 u/L (33–94). FDG scan revealed several lucencies in the left proximal femur, with no uptake. The patient underwent left total hip arthroplasty due to impending pathological fracture. Surgical pathology revealed fragments of fibrous tissue with fibroblasts, hemosiderin and interstitial hemorrhage, consistent with brown tumor.

Conclusion: We describe a unique case of BTHPT of the left femur which was the unrecognized cause of hip pain. BTHPT occurs late in the setting of HPT and is considered as a sign of poorly controlled disease. This is the first case of BTHPT to our knowledge that is not FDG-avid. This is unusual given the vascularity seen in these tumors and highlights that glucose utilization may not necessarily reflect the degree of osteoclastic activity in these tumors. Medical treatment has been shown to be sufficient in helping resolve these lesions, although the process is usually slow. Conversely, in cases of refractory disease, subtotal parathyroidectomy may be required. Our case describes an FDG-negative BTHPT, and highlights that although rare, this should be considered in the management of patients with ESRD, secondary HPT and hip pain. Patients can present with various imaging characteristics. Despite being rare, this case underscores the importance of recognition of BTHPT, so that intervention may be rendered early.

Pediatric Endocrinology

PEDIATRIC OBESITY, THYROID, AND CANCER

A Case Report of Neonatal Hyperinsulinism in an Infant with Turner Syndrome Successfully Treated with Diazoxide

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MON-105

Background: Only four previous cases of hyperinsulinemic hypoglycemia have been reported in association with Turner Syndrome. All of these cases involved young females with a mosaic form of Turner Syndrome involving a ring X chromosome; an abnormality found in just 16% of Turner Syndrome karyotypes. Two of these cases showed responsiveness to diazoxide which stabilized their blood glucose levels and allowed for a longer fasting tolerance.

Reference: doi: 10.1210/jendso/bvaa046 | Journal of the Endocrine Society | A269