MODY 2 Case 1. 4 yo M referred for hyperglycemia in the 300s during surgery. A1c 6.4%. FBG at home 150s, asymptomatic. MGM and MGM’s siblings have diabetes. Diabetes autoantibodies (DAA) negative. C-peptide 5.4 (NL 0.78 - 5.19 ng/ml). MODY panel (GeneDx) showed heterozygous mutation in GCK gene (c.70 C>T). Patient remains off insulin, family reassured and advised to undergo genetic testing.

MODY 2 Case 2. 8 yo M diagnosed at local ED with “T1DM” after presenting with polyuria, polydipsia, and random BG 237. A1c 6.7%, C-peptide 1.9, started on basal-bolus insulin. MODY panel (sent a year later when patient was found to have low insulin requirement, negative DAA) showed pathogenic variant in GCK gene. Weaned from insulin, A1c unchanged (6.3–7%). Mother found to have same mutation. MODY 3 Case 1. 16 yo F referred by PCP who started her on insulin a year prior after an incidental finding of hyperglycemia. A1c was 7.5% at diagnosis. Mom, MGM have diabetes, unknown type (MGM thin by report). DAA neg, C-Peptide 1.74. MODY Panel showed HNF1A heterozygous gene mutation for RIS1Q. She was switched to Glyburide, blood glucose 90s. MODY 3 Case 2. 10 yo M referred from the ED for “T1DM” (weight loss, fatigue, A1c 7.6%) started on basal-bolus insulin, but lost to follow up for a year. Brother has MODY 3. DAA neg, C-Peptide 3.1. Targeted gene sequencing showed HNF1A gene mutation. He was switched to Glyburide, A1c improved to 6.7%. However, patient became noncompliant as teenager, A1c now 9.3%.

Conclusion: MODY remains underdiagnosed. A high index of suspicion should be maintained in nonobese, DAA-negative patients diagnosed with DM before 25yo. Although DAA and genetic testing can be costly, diagnosis can dramatically alter diabetes management as illustrated in all 4 cases, and overall cost of management may be lower in the end. Patients with MODY 2 do not develop vascular complications associated with diabetes, nor require pharmacotherapy. MODY 3 patients may be safely switched to sulfonylurea monotherapy, though degree of diabetes control depends on compliance with medication. Testing gives relatives previously misdiagnosed the opportunity to improve their own quality of life. More education for health care providers is warranted for prompt diagnosis and appropriate management of this condition.

Reference:1. Pihoker, et al. JCEM 2013; 98:4055–62

Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS I

Lactic Acidosis as a Rare and Unusual Presentation of Pheochromocytoma

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SUN-933

Background: Pheochromocytoma is a rare catecholamine-producing tumor of chromaffin cells in the adrenal medulla or of a paraganglion. Typically it presents with sustained or paroxysmal hypertension, severe headaches, palpitations and sweating due to hormone excess. However, the presentation can be variable and can mimic many other diseases. If left undiagnosed or untreated, it can lead to life-threatening consequences.

Case Presentation: A 35 year old female with significant past medical history of migraine headaches, poorly controlled hypertension and a recent new onset seizure, presented with progressive worsening shortness of breath and persistent abdominal pain following a gastrointestinal illness. She also reported diaphoresis, cold fingers and toes, abnormal weight gain, and orthostatic symptoms that gradually worsened for two months prior to presentation. Laboratory evaluation revealed lactic acidosis, leukocytosis, and hypokalemia. Subsequently, a CT scan of the abdomen was performed that revealed an adrenal mass with significant elevation in urine metanephrines. As a result, the patient was diagnosed with pheochromocytoma and successfully treated with laparoscopic left adrenalectomy.

Conclusion: Pheochromocytoma is a rare but can be life threatening if left undiagnosed. It is of utmost importance for clinicians to keep in mind such unusual presentation of a potentially life threatening tumor. To the best of our knowledge, this is an unusual presentation of Pheochromocytoma with severe lactic acidosis.

Thyroid

THYROID CANCER CASE REPORTS I

Mass Cord Compression: Metastasis of Insular Thyroid Cancer

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SUN-490

Intro: Insular thyroid cancer is a rare and complex form of thyroid cancer, often referred to as poorly differentiated carcinoma. The exact incidence of insular thyroid cancers is difficult to assess due to controversial classification of this thyroid cancer over the years. It is termed poorly differentiated as it falls between the well-differentiated and undifferentiated carcinomas both morphologically and biologically[1].

Case: A 41 year old Hispanic female, with a history of prolatinoma and hyperparathyroidism, presented to the hospital with 10 days of progressive lower extremity weakness and paresthesias from T4 downwards, inability to bear weight, and no bowel movement for 12 days. MRI revealed a large thoracic soft tissue mass (7x4x4cm) centered in the posterior and medial aspect of the chest wall at T4-T5 with involvement of the spinal cord and vertebral bodies. She was also found to have a right sided thyroid mass (4.5x5x4 cm) with tracheal deviation. HorThyroid function test, were normal Intact PTH was 261, Thyroglobulin over 300, and Thyroid Antibodies were negative. Patient underwent T3-T6 laminectomy, T2-T7 fusion, and T4-T5 tumor resection, which was subtotal due to vascularity. Second procedure included a right thoracotomy, chest wall resection of ribs 4 and 5 with full resection of paraspinal mass, total thyroidectomy, parathyroidectomy.
with central cervical lymphadenectomy. Pathology results of paraspinal mass showed insular thyroid carcinoma.

Post operatively, the patient reported improvement of sensation and strength in lower extremities. Genetic testing for MEN syndrome was negative.

**Discussion**: Insular thyroid carcinoma, also referred to as poorly differentiated carcinoma is a rare form of thyroid cancer. Insular carcinoma was characterized by to include the following complex histologic features, “formation of solid clusters (insulae) of tumor cells containing a variable number of small follicles; variable but consistently present mitotic activity, capsular and blood vessel invasion; and frequent necrotic foci, sometimes leading to formation of peritheliomatous patterns”[1]. The cells originate from follicular epithelium and possess the potential to concentrate radioiodine[2]. Unlike anaplastic carcinoma of the thyroid, p53 and p21 staining was negative in insular carcinomas[3]. Thryoglobulin staining is generally positive[4]. Distant metastasis occurs in about 31% of patients with insular thyroid carcinoma[5]. In cases of distant metastasis, treatment with thyroidectomy and radioiodine therapy were shown to independently improve survival[5].

The Constellation of Insular thyroid cancer, hyperparathyroidism and Prolactinoma, has not been reported before.

**References:**

[1] Am J Surg Pathol. 1984;8:655-
[2] J of Nuc Med 32(7), 1358
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[4] JCEM 99. 1167–9. 10.1210/jc:2014
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**Reproductive Endocrinology**

**CLINICAL STUDIES IN FEMALE REPRODUCTION I**

**Incidence and Predictors of Hypertension in a Cohort of Australian Women with and Without Polycystic Ovary Syndrome**

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**SAT-015**

**Background:** There is a lack of longitudinal studies exploring the relationships between polycystic ovary syndrome (PCOS) and hypertension, in population-based settings.

**Objectives:** To identify predictors of hypertension in women with and without PCOS and the relationship to body mass index (BMI).

**Methods:** We undertook a community-based cohort analysis of the Australian Longitudinal Study (ALSWH) data conducted on 9,688 young adult women, aged 21–42 years from 2000–2015. We conducted survival analysis using the Cox Proportional Hazards Model to identify predictors and person-time analysis to calculate incidence rates of hypertension.

**Results:** Overall, 9,508 women were followed for 145,159 PY (person-years) and 1,556 (16.37%) women developed hypertension during the follow-up. The incidence of hypertension was significantly higher (p = 0.001) among women with PCOS (17/1000 PY) compared to women without (11/1000 PY). There were significant differences in time to hypertension development between women +/- PCOS. Hypertension was observed among women with PCOS from early adulthood and across BMI categories. The difference in the actual number of incident hypertension cases (incidence rate difference (IRD)) between women with and without PCOS, was fourfold higher (15.8 vs. 4.3 respectively) among women who were obese at baseline, compared to age-matched lean women. PCOS was independently associated with hypertension with a 36% greater risk, adjusting for BMI and other confounders.

**Conclusion:** Our results suggest women with PCOS are more likely to develop hypertension from early adulthood, independent of BMI and with risk exacerbated by obesity.

**Neuroendocrinology and Pituitary**

**NEUROENDOCRINE & PITUITARY PATHOLOGIES**

**Flash Glucose Sensor Monitoring for Patients with Endogenous Hyperinsulinaemic Hypoglycaemia**

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**SUN-299**

Flash Glucose Sensor Monitoring for patients with endogenous hyperinsulinaemic hypoglycaemia

**Background:** Flash glucose monitoring systems (FGS) have recently been introduced and measure interstitial glucose using an amperometric electrochemical sensor assay, and are increasingly used to provide a convenient means to monitor levels on a minute-by-minute basis over two weeks in ambulatory patients with diabetes. Although continuous glucose monitoring systems have been previously used in patients with insulinoma, to our knowledge, FGS use has only been described once previously in an adult patient with an insulinoma. Here, we describe use of this system in 6 patients with confirmed endogenous hyperinsulinaemic hypoglycaemia, especially for the critical nocturnal period.

**Methods and patients:**

FGS data obtained over each 2-week monitoring was reviewed in 6 patients seen between 2018 and 2019: 5 had a biochemically proven insulinoma and 1 had Hirata’s syndrome. In 4 patients, follow-up readings were obtained after adjustment of glucose-raising medication: two on octreotide, one on diazoxide and one on diazoxide and dexamethasone.

**Results:** Median age was 63 years (range 37–83). In the 4 patients with more than one 2-week FGS assessment comparison between first and last readings demonstrated that the average duration of hypoglycaemia (<4mmol/L) 126, 171, 173 and 282 minutes improved to 46, 128, 30 and 0 minutes,