Primary aldosteronism (PA) usually presents with recalcitrant hypertension, hypokalemia and an elevated aldosterone: renin ratio. It is commonly attributed to adrenal adenomas or hyperplasia with familial hyperaldosteronism (FH) remaining a rare etiology. FH is sub-divided into glucocorticoid remediable, type I, and non-glucocorticoid remediable, types II – IV. The initial diagnosis of such a condition during pregnancy and in the setting of worsening kidney disease presents a diagnostic and management challenge as this precludes adrenal vein sampling and contrast imaging. Our case highlights the importance of early screening for PA and illustrates the need for updated guidelines on aldosteronism workup in the setting of ESRD and pregnancy.

Tumor Biology
ENDOCRINE NEOPLASIA CASE REPORTS I

Carney Complex: A Case of a Rare Multiple Endocrine Neoplasia Misdiagnosed as Peutz-Jeghers Syndrome
Sonali Vaid, BS BME, MD¹, Faizan Hasnain, MS⁴, Rachel Morgan, MD⁷.
¹St. Vincent Hospital, Carmel, IN, USA, ²Indiana University School of Medicine, Indianapolis, IN, USA.

SUN-940
Carney Complex (CNC) is an extremely rare multiple endocrine neoplasia caused by germline inactivating mutation in protein kinase A type I-alpha regulatory subunit (PRKAR1A gene). Mode of inheritance is mostly autosomal dominant; 25% of cases are due to de novo mutations. Only 750 world-wide cases have been reported. Most patients are diagnosed in the second or third decade. Clinical features include cutaneous myxomas, angiomyxoid nodules, lentiginous skin pigmentation, cardiac myxomas, and benign and rare malignant endocrine tumors. These endocrine tumors include and are not limited to prolactinomas, thyroid tumors, primary pigmented nodular adrenocortical disease (PPNAD), and large cell-calciﬁying Sertoli cell tumors (LCCSCT). Diagnosis is often challenging as disease manifestations can occur sporadically over a large span of time, and patients may present with various conditions such as Cushing syndrome, like our case. We present a case that demonstrates the importance of early recognition of this rare disorder.

A 28-year-old Caucasian male with PMH of HFrEF, HTN, Sertoli cell tumor status post orchietomy, vertebral fractures, and surgical removal of lip angiomyxomas presented to clinic for hypogonadism. Physical examination revealed marked Cushingoid features and facial lentigines above his eyes and on his lips. His eclectic medical history and unique exam ﬁndings lead to ﬁnding of a unifying diagnosis. His labs revealed severe Cushing syndrome, and computed tomography (CT) of his abdomen was performed due to ACTH independent hypercortisolism, demonstrating a bilateral lobular appearance of the adrenal glands. Combination of labs and physical exam ﬁndings of lentigines, skin myxomas, cushingoid features, rare angiomyxoma, LCCSCT and hypercortisolism lead to diagnosis of Carney Complex. He was misdiagnosed with Peutz-Jeghers in his adolescence due to LCCSCT and mucosal lentigines; therefore, hormonal screening was not routinely performed. Untreated Cushing led to severe osteoporosis with vertebral fractures and heart failure. Treatment included bilateral adrenalectomy. Pathology report conﬁrmed rare PPNAD. PPNAD and LCCSCT are extremely rare tumors almost exclusively linked to Carney Complex. Interestingly, family history did not reveal endocrine disorders, cancers, or severe illnesses. Genetic testing returned positive for the PRKAR1A gene mutation. Given the consequences of untreated hormonal aberrations seen in this disorder, an early and accurate diagnosis is imperative.

Thyroid
THYROID CANCER CASE REPORTS II

Papillary Thyroid Carcinoma Arising in a Thyroglossal Duct Cyst: A Case Report
Gabriela Zuniga, MD, Glicelda Galarza Fortuna, MD, Alejandro Guzman-Duvant, MD, Juan Paramo, MD, Michael Pagacz, MD.
Mount Sinai Medical Center Florida, Miami Beach, FL, USA.

MON-438
Introduction: Thyroglossal duct cysts (TGDCs) are uncommon benign congenital entities. Rarely, thyroid carcinoma can arise from a TGDC; the most common being papillary thyroid carcinoma (PTC). Similar to TGDC, carcinomas originating within them can present as an asymptomatic midline neck mass. Signs of malignancy include dysphagia, dysphonia, weight loss, and rapid growth. Given the rarity of TGDC carcinomas, clinical management remains controversial, particularly regarding the requirement for total thyroidectomy.

Case: A 52-year-old female with history of an anterior central neck mass initially noted in 2017. A 0.3-cm left lobe mid-segment cyst and a complex thyroglossal avascular simple cyst measuring 2.4 × 1.1 × 1.8 cm was observed during ultrasound (US). She presented to the endocrinology clinic in April 2019 due to progressive enlargement of the mass. Repeat thyroid US revealed that the cystic structure had become complex with a peripheral solid component and measured 3.3 × 2.1 × 2.2 cm. FNA was performed and found to be suspicious for PTC (Bethesda category V) and positive for the BRAF V600E mutation.

Patient was referred for surgical evaluation. Physical examination revealed a midline anterior 10-cm, painless, and fixed mass above the thyroid that moved with deglutition and tongue protrusion. Contrast computed tomography scan showed a large multiloculated cystic structure measuring 4.1 × 4.4 × 5.9 cm. A lobulated soft tissue mass measuring 2.2 × 2.4 × 3.0 cm was noted internally along the inferior margin of the cyst. She underwent en-block resection of the TGDC in addition to a total thyroidectomy. Histopathological examination identiﬁed a 7.5 × 5.5 × 5.0 cm cystic mass with inﬁltration of the fat and ﬁbrous stroma. Solid area in the lower portion measuring 2.6 × 2.4 cm. Thyroid gland examination was otherwise unremarkable. No areas of extension of the mass into the thyroid tissue were clearly identiﬁed and no other gross lesions were observed. The solid area within TGDC contained a tumor with ﬁndings characteristic of PTC. Postoperatively, she was placed on thyroid hormone replacement therapy.
Conclusion: The main difficulty encountered with cancer developing from TGDC is that the diagnosis is usually made during surgery and from definitive pathological samples. The most common surgical procedure used is the Sistrunk procedure. Some studies have suggested that this procedure alone is an adequate therapy, but others advocate the need for total thyroideotomy. The Sistrunk procedure is considered to be appropriate for low-risk patients, but high-risk patients must undergo total thyroideotomy. The decision to perform a total thyroideotomy in this patient was based on her high-risk classification due to: age, sex, cyst size, and a positive FNA for malignancy. Follow-up includes an annual physical examination, thyroglobulin levels, and an US every 6 months during the first year and annually thereafter.

Pediatric Endocrinology
PEDIATRIC SEXUAL DIFFERENTIATION, PUBERTY, AND BONE BIOLOGY

Screening of Vitamin D and Calcium Concentrations in Neonates of Mothers at High Risk of Vitamin D Deficiency
Sheikh Arif Maqbool Kozgar, FRACP1, Paul Chay, FRACP2, Craig Frank Munns, MBBS, PhD, FRACP3.
1Latrobe Regional Hospital, Traralgon, Australia, 2Liverpool Hospital, Liverpool, NSW, Australia, 3The Children’s Hospital at Westmead, Westmead, Australia.

SUN-083
Objective: The aim of this study was to determine, retrospectively, the serum 25OHD and calcium concentrations of screened neonates of mothers at high risk of 25OHD deficiency (maternal 25OHD < 25 nmol/L or unknown vitamin D concentrations and risk factors for vitamin D deficiency) and critically analyse whether their measurements contribute to the management of these neonates.
Methods: Serum 25OHD and calcium concentrations from 600 samples of umbilical cord blood or venous blood collected from neonates over a 12-month period were analysed. 25OHD concentrations were reported for all while both the corrected calcium concentrations and vitamin D concentrations were available for 569 samples.
Results: There was little or no evidence of association between neonatal 25OHD concentrations and gender, gestational age or birth weight. There was a high prevalence of vitamin D insufficiency (27.6%, 30–50 nmol/L) and deficiency (21.3%, < 30 nmol/L) in neonates from high-risk maternal groups. There was a statistically positive but weak correlation (\( r = 0.22, P < 0.0001 \)) between serum calcium and 25OHD concentrations. Only 7 neonates out of 569 (1.2%) had calcium levels in the hypocalcaemic range; however, a significant number (47.6%) were reported to be in the hypercalcaemic range. Nearly all of these were venous samples collected in first 24 hours after birth. We calculated the reference interval for corrected calcium from our data of venous samples in first 24 hours and the upper limit was significantly higher (2.38–3.04 mmol/L) than the standard reference range used.
Conclusion: Vitamin D deficiency is prevalent in neonates of high-risk mothers but the risk of hypocalcaemia due to vitamin D deficiency at birth is low. Screening neonates entails blood testing which can cause distress to neonates and their parents, substantial impost on staff and financial burden on the health care system. 25OHD deficiency is corrected relatively easily in neonates with supplementation and vitamin D supplementation of neonates from birth without routine screening appears to offer better value of care. Also, the data from this study suggest that the paediatric reference range for corrected calcium concentrations in neonates is higher and the paediatric reference range should be reconsidered.

Bone and Mineral Metabolism
BONE AND MINERAL CASE REPORTS II

Bone and Mineral Metabolism

CHARGE Syndrome: Unusual Cause of Hypogonadism Leading to Osteoporosis
Ankur Modi, MD1, Veronica Piziak, MS, MD, PHD2.
1Baylor Scott & White, Temple, TX, USA, 2Scott & White, Temple, TX, USA.

MON-355
CHARGE syndrome is an unusual cause of hypogonadism; it is characterized by coloboma, heart defect, ateresia choanae, retarded growth and development, genital hypoplasia and ear anomalies. Two-thirds of affected patients have a mutation within the chromodomain helicase DNA-binding protein-7 gene, which is involved in embryonic development. The involvement of this gene in the pathogenesis of isolated idiopathic hypogonadotrophic hypogonadism (HH) has been postulated. The reported incidence of this syndrome ranges from 0.1–0.2/10000 (1).
A 24 year old female presented to our facility for further management of her HH and osteoporosis in the setting of her CHARGE syndrome. She was born full-term and diagnosed with this condition at the age of 6. Formal genetic testing as an adult demonstrated mutation within the CHD7 gene (chr 8:61,757,970). She had delayed puberty secondary to her hypogonadism; she was not treated with HRT as benefits were not considered significantly sufficient. She subsequently developed osteoporosis at the age of 20 which was treated at an outside facility with pamidronate IV Q4 months along with calcium and Vitamin D supplementation. Her initial Dual-energy X-ray absorptiometry (DXA) showed scoliosis in the lumbar spine (bone mass density (BMD): 0.514,osteoporosis by Z-score) with total hip showing BMD: 0.738,osteopenia by Z-score. Follow-up DXA after 3 years showed statistically significant improvement in bone mineralization of her L-spine [BMD: 0.595, +16%] and total hip [BMD: 0.777, +13.5%]. She presented to our facility in 2018 with labs showing normal calcium and 25-OH D; treatment with pamidronate was continued. She had a repeat DXA in 2019 which showed Z-scores of 0 in the left and right femoral necks. She was given the option of continued treatment for her osteoporosis versus monitoring and chose the latter with follow-up DXA.
Hypogonadotrophic hypogonadism is associated with delays in puberty or pubertal arrest. Lateinizing Hormone Releasing Hormone and HCG tests should be performed within the four months of life or at puberty in cases of hypogonitalism. GH deficiency should be investigated as a cause for growth retardation with GH stimulation levels. Hormone replacement is often required at puberty for