lung disease, failure to thrive. Patient was found to have hyponatremia, hyperkalemia, high FeNa of 1.3% (intrinsinc renal disease) and elevated BUN/Cr (92/1.15). Renal US found echogenic kidneys with poor cortical medullary differentiation suggesting renal disease. Further evaluation noted high aldosterone (1700 ng/dL) and renin (400 ng/mL/hr) levels. He was placed on low protein formula to help optimize BUN level. Baby was diagnosed with secondary PHA1 due to renal disease and started on NaCl supplementation. This led to normalization of BUN, creatinine and improvement in electrolytes. Patient also had high serum calcium ranging from 11.1 to 12.0 mg/dL. Hyponatremia, hyperkalemia, hypercalcemia could be attributed to possible CAH, however state screen and ACTH stimulation testing were normal. Further workup showed high 25-OH-vitamin D > 99 ng/mL, PTH 46.9 pg/mL, phosphorous 5.4 mg/dL and 1,25-OH-vitamin D 63.1 pg/mL. Urine Ca/cr ratio was 0.522. Vitamin D supplementation was stopped and daily total fluids increased. Subsequently, there was improvement in serum Ca at 10.9 mg/dL and 25-OH Vitamin D of 74 pg/mL. Next Generation Sequencing (NGS) was carried out, with a focus on the etiology of persisting hypercalcemia, including familial forms of hypercalcemia and Williams Syndrome. NGS revealed a likely pathogenic variant, c.2365 + 2T>C (p.?), in NR3C2, consistent with a diagnosis of AD PHA 1. Conclusion: This is a case of AD PHA1, marked by renal mineralocorticoid receptor resistance associated with persisting hypercalcemia. Initial hypercalcemia could be explained by hypercalcitroninosis D. It is important to note that electrolyte abnormalities, including persistent hypercalcemia, could be also secondary to the kidney disease found on renal US. There are only few reports of hypercalcemia in patients with PHA1 in the literature. In children with electrolyte abnormalities and failure to thrive, monitoring of serum and urine electrolytes would facilitate early accurate diagnosis and timely treatment.

Addison’s disease is a rare clinical entity that requires a high index of suspicion. It may rarely present with weakness and/or back pain. Early recognition and treatment of Addison’s disease adequately resolves hyperkalemic paralysis. It is good clinical practice to keep an open-ended differential list to help reduce diagnostic suspicion bias.

### Adrenal

#### ADRENAL CASE REPORTS

### Hypertension and Hirsutism in a Young Female: A Rare Form of Congenital Adrenal Hyperplasia?

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**Background:** Rare forms of congenital adrenal hyperplasia (CAH) especially can be a diagnostic and management challenge. We present a case of hypertension and hirsutism with elevated mineralocorticoid and sex steroid precursors concerning for non-classic rare form of CAH.

**Clinical Case:** A 22 y/o female was evaluated in our endocrinology clinic for primary hypertension and hirsutism (modified Ferriman-Gallwey score of 12) but without menstrual irregularity, clitoromegaly or deepening of voice. Familial history was significant for hypertension and hirsutism in her mother. Renal and pelvic ultrasonographies were normal. CT abdomen showed thickened bilateral adrenal glands. Case detection testing was positive with PAC/PRA of 19.4/0.4 however intravenous saline suppression test suppressed aldosterone to 5 pg/dL, hence ruled out primary aldosteronism. Cortisol suppressed to 0.6 mcg/dL after 1 mg overnight Dexamethasone, hence ruled out Cushings’s syndrome; prolactin and TSH were normal, serum HCG was undetectable. This prompted work up for other endocrine causes of hypertension and hirsutism which revealed follicular phase elevated dehydroepiandrosterone-sulfate of 772 mcg/dL (35–430), pregnenolone of 603 ng/dL (15–132), 17-hydroxyprogrenenolone of 1516 ng/dL (<226), 11-deoxycortisol of 39.4 ng/dL (<32); but normal 17-hydroxyprogesterone of 36.8 ng/dL (15–70), androstenedione of 1.12 ng/mL (0.26–2.14) and free testosterone of 26 ng/dL (9–44). 250-mcg ACTH administration stimulated cortisol to 24.6 mcg/dL, pregnenolone to 1478 ng/dL, 17-hydroxyprogrenolone to 1716, 11-deoxycortisol to 64.2 ng/dL but 17-hydroxyprogesterone only stimulated to 59.9 ng/dL. We initiated spironolactone at 12.5 mg increased to 25 mg daily, which normalized her blood pressure. She was counseled regarding its teratogenicity but is she declined birth control, as she was not engaging in heterossexual intercourse. Urinary steroid analysis and genetic testing were pending.

**Conclusions:** Hypertension and hirsutism accompanied by elevated pregnenolone, 17-hydroxyprogrenenolone, 11-deoxycortisol and dehydroepiandrosterone-sulfate, however, normal baseline as well as stimulated 17-hydroxyprogesterone, raise concern for co-occurrence of partial 3-betahydroxysteroid dehydrogenase deficiency in addition to non-classic 11-betahydroxylase deficiency. Management with mineralocorticoid receptor antagonist helped control hypertension.

### Adrenal

#### ADRENAL CASE REPORTS

### Hyperkalemic Paralysis Due to Addison’s Disease

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A 60-year-old woman with type 1 diabetes mellitus, past history of breast cancer, degenerative disc disease, hypothyroidism and asthma gave a 5-day history of flaccid paralysis and a 5-month history of back pain and worsening mobility. Investigations revealed hypercalcemia, hyperkalemia and hyponatremia which were initially thought to be due to hypercalcemia of malignancy on the assumption that her breast cancer had recurred. Imaging of the spine did not reveal cauda equina syndrome. There was no clinical and biochemical improvement in spite of fluid resuscitation.

Hypoaldrenism was suspected and confirmed three days later. Adrenal replacement therapy fully resolved her paralysis within 24 hours.