

multiple organs. PHA type 2 (Gordon syndrome) presents with hyperkalemia and hypertension. Case: Our patient is a 5 week old male who admitted for significant electrolyte abnormalities. He was followed by PCP for failure to thrive. The child was referred to ED with increased difficulty in feeding, lethargy and episodes of emesis. In the ED, the child was in a compensated shock and had a low normal BP: 76/35, HR: 169/min and fast breathing R/R: 80/minute and afebrile. P.E: showed signs of dehydration. Lab work showed: Na: 110 mEq/L, K: 9.3 mEq/L, low Chloride and Ca: 11.1 mEq/L. Endocrinology recommended IVF supplementation with 2 x NS bolus followed by IVF's at 1.5 times maintenance (D5 + NS), along with administration of Florinef 0.2 mg suspecting CAH. Renin, 17-OHP, random cortisol level and thyroid hormone levels were ordered. Results showed: TSH of 5.30 mcIU/mL and Free T4 of 2.2 ng/dL. Cortisol: 20.5 mcg/dL. He was subsequently admitted to PICU. Septic work-up was negative. He became hemodynamically stable after hydration and did not require the stress dose of hydrocortisone. Repeat Na: 133 mEq/L, K: 4.7 mEq/L, Cl: 102 mEq/L and Glucose: 90 mg/dL. ACTH stimulation test for CAH evaluation was performed, stimulated 17OHP: 88 ng/dl, cortisol: 27.1 mcg/d and normal DOC. Elevated Aldosterone: 632 ng/dl (5-80) and renin level: 351 (6.5-86). A diagnosis of PHA was made and florinef was stopped and the child was started on NaCl supplementation which normalized the electrolytes. Genetic testing was negative for *NR3C2*, *CUL3*, *KLHL3*, *SCNN1A*, *SCNN1B*, *SCNN1G*, *WNK4* and showed that the patient is a heterozygous for a variant of unknown significance, c.6276T>A (p.Ser2092Arg) in the *WNK1* gene. However, the patient did not have hypertension and urine electrolytes were also normal did not show signs of PHA 2. Conclusion: PHA can present with severe salt wasting crisis. It can be diagnosed clinically. The relationship of mutation and phenotype can be elusive. Course was uncomplicated and he was discharged from the PICU in 6 days. Sodium doses were titrated based on serum levels with eventual dose of 22.5 mEq/kg/day and sodium level was 139 mEq/L.

Bone and Mineral Metabolism

NEW INSIGHTS INTO PTH AND CALCIUM RECEPTOR SIGNALING

Is Urinary Calcium the Only Predictor of Nephrolithiasis in Patients with Asymptomatic Primary Hyperparathyroidism?

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OR07-05

The 4th International Workshop for the management of asymptomatic PHPT included, among the criteria for parathyroidectomy, the presence of hypercalciuria (dUCa>400 mg/day) and increased biochemical stone risk profile. The aim of the present study was to evaluate the biochemical stone risk profile in 176 consecutive patients (143 females and 33 males) with asymptomatic PHPT. We recorded clinical and biochemical data, including 24 hours

urinary measurements of the following parameters: volume and pH, creatinine, calcium, magnesium, sodium, potassium, ammonium, uric acid, oxalate, citrate, phosphate, inorganic sulphate and chloride and kidney ultrasound.

In our cohort dUCa>400mg/day showed a low sensitivity and positive predictive value (PPV) for nephrolithiasis with high specificity (46.2, 32.7, 73.0% respectively), while hypercalciuria by 4 mg/kg/bw (d-UCa>4mg/kg) had a high sensibility, with low PPV and specificity (79.5, 27.7, 40.1%) Daily hypomagnesuria (d-HypoMg), but not any other urinary parameter, was an independent predictor of nephrolithiasis in the univariate (OR 2.97 CI 1.27-7.09 P=0.014) and multivariate analyses adjusting for age, sex, BMI, and eGFR (OR 3.13 CI 1.17-8.42 P=0.02). d-HypoMg was relatively lower in the regression analysis with urinary calcium in patients with nephrolithiasis compared with those without. The mean ratio between (dUCa) and (dUMg) was higher in patients with nephrolithiasis compared with those without (4.6±2.0 vs 3.3±4.1; P<0.001). In the univariate and multivariate analyses the dUCa/dUMg ratio was a significant predictor of nephrolithiasis [OR 4.9 (2.3-10.5); P<0.001; OR 5.3 (2.4-11.6), P<0.001, respectively]. The AUC using the dUCa/dUMg ratio as variables was 0.69 (CI 0.60-0.79; P<0.0001). The best cut-off value, set at the highest Youden index, was equal to 4.0, with a sensitivity of 59.0% and a specificity of 77.4%.

In patients with hypercalciuria (>400 mg/24-hour) dUMg was positively correlated with dUCa in those without nephrolithiasis (r=0.50, β=0.2, P=0.002) but not in those with nephrolithiasis (r=0.05, β=0.014; P=0.8). In patients without hypercalciuria we found that hypomagnesuria remained a predictor of nephrolithiasis using either 400 mg/die (P=0.002, OR 5.12 (1.84-14.24) or 4 mg/kg bw (P=0.014, OR 6.24 (1.45-26.8). Moreover, the OR for nephrolithiasis improved using the combination of d-HypoMg with d-UCa>4mg/kg (OR 8.12, CI 1.92-34.18, P=0.004), but not with dUCa>400mg/day.

The current urinary calcium threshold of >400 mg/24-hour has a low sensitivity in detecting nephrolithiasis; our data suggest that sensitivity, specificity and positive predictive value could be improved including dUMg, dUCa/dUMg ratio and the combination of d-HypoMg with d-UCa>4mg/kg in the stone risk evaluation.

Diabetes Mellitus and Glucose Metabolism

TYPE 2 DIABETES MELLITUS

Does Short Term Intensive Insulin Therapy in Newly Diagnosed Type 2 Diabetes Mellitus Delay Eventual Insulin Dependence

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SUN-681

In patients with type 2 diabetes mellitus (T2DM), dysfunction of β-cells starts years before the diagnosis of T2DM and rapidly worsens after overt hyperglycemia. Use of short-term intensive insulin therapy (STIIT) at the time of diagnosis of overt hyperglycemia has shown clinical recovery