

At reevaluation one year later, RS had a decreased bone density, low estrogen and normal gonadotropins. These data, combined with her persistent irregular scant menses, raised the suspicion of negative effects of her inhaled steroids. Unusual fatty deposits have been reported with Dulera.

Interventions: After we consulted with her Allergist, RS began a Dulera taper. Three weeks after stopping Dulera she experienced exhaustion and fatigue. Cortrosyn stimulation testing revealed suppressed cortisol levels with no rise to stimulation. ACTH was normal. She began replacement hydrocortisone.

Five months later at age 16 she had been weaned to very low dose hydrocortisone replacement, had grown one inch, and had normal menses. The fatty deposits in her legs were completely resolved.

RS was weaned to stress dose steroids only. A follow up Cortrosyn stimulation test revealed an improved, but still low, baseline level with an appropriate rise to stimulation. We advised her to continue the stress dosing with plans for further reevaluation.

Discussion/Recommendations: Chronic use of inhaled steroids is increasingly common. Endocrinology nurses must be aware of the potential consequences of inhaled steroid use in children, including suppression of the pituitary-adrenal axis, impaired linear growth, possible irregular menses and low bone density. Iatrogenic adrenal insufficiency can be life threatening. Watching for it and avoiding it are imperative.

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008–17-year-3-Month-Old Female with CF and Hypoglycemia

Shayne Dougherty CRNP

Nurse Practitioner, The Children's Hospital of Philadelphia, Philadelphia, PA

Patient Demographics: Patient is a 17-year-3-month-old Caucasian female

Clinical Presentation: Patient presented to clinic with post prandial hypoglycemia without the use of exogenous insulin.

Relevant Past History: She has a history of hypothyroidism, impaired glucose tolerance, cystic fibrosis (CF) diagnosed at birth due to meconium ileus, and celiac disease. Weight 45.6 kg (42%) and height 1.48m (1%). Family history includes brother with CF, mother with hypothyroidism, and father with hypertension.

Evaluation: Workup included oral glucose tolerance test (OGTT), HgbA1c, diabetes autoimmune panel, and thyroid studies. OGTT was remarkable for fasting BG=95, 1 hr BG= 309, 2 hr= 248. HgbA1c 6.1%. All other labs were normal. Continuous glucose monitoring (CGM) revealed postprandial hyperglycemia after meals with overcorrection causing lows (60's) without exogenous insulin use.

Interventions: Management of CFRD is best with a team approach including patient, family, pulmonary and endocrine teams. Management includes blood glucose monitoring, nutritional counseling, and insulin. Patient and family completed diabetes education training with a CDE and RD, and were taught to monitor postprandial blood sugar 1-2 hours after meals and to carbohydrate count. Patient was started on 2 units Lantus insulin daily and Novolog insulin prior to all meals/snacks. Patient returned 3 months later and postprandial hypoglycemia was eliminated with initiation of pre-meal insulin.

Discussion/Recommendations: CFRD is the most common comorbidity in CF. The average age of onset is 18-21 years, affecting 20% of adolescents and 40-50% of adults. With a combination of insulin deficiency and insulin resistance, CFRD worsens the

prognosis of CF and is associated with worsening pulmonary function, increased infections, nutritional decline, and increased mortality. Postprandial and OGTT-related hypoglycemia is common in CF. Postprandial hypoglycemia is associated with early glucose dysregulation (higher peak plasma glucose) and arises from late secretion of endogenous insulin. Administration of insulin at meals may prevent hyperglycemia excursions and blunt post-prandial hypoglycemia events. CFRD should always be a differential diagnosis for any patient who presents to endocrine clinic with postprandial hypoglycemia. Endocrine nurses are in an important position to educate patients regarding symptomatology, pathophysiology, and treatment of CFRD.

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009–Family History Plays an Important Role in the Diagnosis of Multiple Endocrine Neoplasia Type 2A

Courtney Kivel MS, CRNP

Children's Hospital of Philadelphia Division of Endocrinology and Diabetes, Philadelphia, PA

Patient Demographics: Patient is a 13-year-old African American male.

Clinical Presentation: Patient presented with bilateral thyroid nodules incidentally discovered on head and neck imaging for occipital skull fracture with subarachnoid hemorrhage sustained while playing football. On dedicated thyroid ultrasound, there were sub-centimeter nodules in the left and right lobes.

Relevant Past History: Patient has no significant past medical history. Detailed review of family history revealed that maternal grandmother had Multiple Endocrine Neoplasia Type 2A (MEN2A), medullary thyroid carcinoma (MTC), and pheochromocytoma. Patient's mother and 4 siblings, age 11, 15, and 23, had undergone no testing at time of presentation.

Evaluation: The vital signs and physical exam were normal, without palpable nodules or abnormal lymphadenopathy. On review of thyroid ultrasound images, nodules were low risk based on size. The right sided nodule was hypoechoic with irregular borders. There were no calcifications and no abnormal lymph nodes. Patient was referred for fine needle aspiration which revealed MTC in both nodules. Calcitonin was elevated at 58.8 pg/mL (0.0-7.5 pg/mL). Carcinoembryonic antigen (CEA) was elevated at 6.1 ng/mL (0.0- 3.0 ng/mL). Plasma metanephrine panel, calcium, and phosphorus were normal. Staging was completed with neck CT with contrast re-demonstrating bilateral thyroid nodules and no abnormal lymph nodes, and chest and abdomen CT with contrast normal.

Interventions: Patient was referred to otolaryngology and is scheduled for total thyroidectomy with bilateral central neck dissection. He and his siblings were also referred to the Tumor Predisposition Clinic and genetic counseling.

Discussion/Recommendations: Thyroid cancer is most often asymptomatic and may be found incidentally on head and neck imaging. This case illustrates the importance of a detailed family history in the diagnosis of medullary thyroid carcinoma. The American Thyroid Association has published updated guidelines on the diagnosis and treatment of MEN2A and MTC. Early detection based on family history and available genetic testing is essential for successful treatment including prophylactic thyroidectomy and ongoing surveillance for known comorbidities.

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