

Past History: Patient A was diagnosed with GHD and hypothyroidism at age 18 months. Patient B was diagnosed with GHD at age 7 1/2 years; other medical conditions include fetal alcohol syndrome with failure to thrive, global developmental delay, attention-deficit/hyperactivity disorder, and gastroesophageal reflux with fundoplication.

Evaluation: Patient A had laboratory assessments as follows: IGF-1 70 (201–609 ng/mL), thyroid function studies normal, fasting glucose 80 (56–145 mg/dL), and fasting cortisol 21 (6.0–23.0 µg/dL). Insulin tolerance test with GH maximum 0.9 ng/mL. Repeat MRI showed empty sella. Patient B had laboratory assessments as follows: IGF-1 129 (209–602 ng/mL), thyroid function studies normal, and fasting cortisol 21.5 (4.2–38.4 µg/dL). Insulin tolerance test with GH maximum 1.5 ng/mL. Repeat MRI was normal.

Interventions: Patient A was restarted on GH therapy at a transition dose of 0.03 mg/kg per day. He achieved an 18-lb weight gain with 2 months of therapy and improvement in energy level. Patient B will restart on GH therapy at a dose of 0.01 mg/kg per day.

Discussion/Recommendations: The etiology of weight loss in these adolescent males is not understood. Metabolic changes in adipose tissue result in weight gain with increased adiposity and reduced muscle mass in GH-deficient young adults. This phenomenon is opposite of the usual presentation.

doi:10.1016/j.pedn.2012.03.011

The Need for Assessing Cortisol-Binding Globulin in Evaluation for Cushing's Syndrome in a Young Girl

Patty Graves RN, CPNP, CDE

Central Ohio Pediatric Endocrinology and Diabetes Services, Columbus, OH

Patient Demographics: 16-year 7-month-old Caucasian female.

Clinical Presentation: Referred by a neurosurgeon for evaluation of her endocrine status. She had a pituitary lesion and polyuria and polydipsia suggestive of diabetes insipidus (DI).

Past History: She had a few months' history of increased thirst and urinary frequency. She experienced headaches twice a week, regular menstrual cycles, and no significant changes in her weight or energy level. Her urine was "like water."

Evaluation: Height was at the 25th percentile and weight was at the 50th percentile. Specific gravity on urinalysis was 1.010. An MRI showed an enlarged pituitary gland, a lesion on the pineal gland (a cyst or mass), and a pituitary lesion that could be interpreted as a Rathke's cleft cyst or macroadenoma. Follow-up MRI was recommended. Pituitary testing included a prolactin of 29 ng/mL (normal [nl] <24), IGF-1 of 346 ng/mL (nl range), TSH of 15 µU/mL (nl <5.5), FT4 of 1.03 ng/dL (nl 0.89–1.76), elevated antiperoxidase antibodies, and normal LH and FSH levels. AM cortisol was 47 µg/dL (nl 7–20), ACTH of 19 pg/mL (nl 6–48), PM cortisol of 25 µg/dL (nl 4–11), urinary free cortisol level of 56 µg/24 hours (nl 2–38), and a cortisol of 4 µg/dL after suppression with dexamethasone. A corticosteroid binding globulin (CBG) was 6.2 mg/dL (nl 2.3–3.9).

Interventions: She started at a low dose of desmopressin after an overnight fast both as a diagnostic study and for clinical therapy. Repeat electrolytes were normal. The thyroid abnormality, unrelated to her pituitary issue, showed Hashimoto thyroiditis. She was started on 75 µg thyroid supplementation. Because the elevated cortisol level resulted from CBG excess, she did not require treatment.

Discussion/Recommendations: Differential diagnoses included hypopituitarism because of the abnormal MRI and symptoms of DI. Elevated cortisol levels were unexpected because she lacked symptoms or physical characteristics of elevated cortisol levels. Approximately 75% of the cortisol in circulation is bound to CBG. The cortisol is thought to be biologically active only when it is not bound to CBG. Health care providers need to consider differential diagnoses and not narrow their focus on expected findings and make an inaccurate diagnosis. The patient/family must understand that CBG excess caused the elevated cortisol levels and does not require treatment.

doi:10.1016/j.pedn.2012.03.012

Failure to Thrive Because of Inherited Congenital Isolated Growth Hormone Deficiency

Lisa Michele Pincham MSN, RN

Children's National Medical Center, Washington, DC

Patient Demographics: A 22-month-old female, severe failure to thrive.

Clinical Presentation: Length was 66 cm (–5.1 SD), and weight was 6.6 kg (–6.7 SD). Prominent forehead and midfacial hypoplasia were noted. Muscle mass was decreased.

Past History: Birth weight 5 lb 11 oz at term, grew well for 4 months and then progressively deviated below the curve in length and weight. Mother's height was 5 ft 3 in., with menarche at age 13 years. Father, –4 SD, was diagnosed with isolated growth hormone deficiency at 7 years of age, and treated (5 ft 4 in.). Siblings included a 6-year-old brother who was very small at age 22 months during an endocrine evaluation and a 3-year-old sister with height and weight at both –4 SD.

Evaluation: Free T4 was 1.28 ng/dL (normal 1.1–1.7), TSH 1.8 µU/mL (normal range). IGF-1 less than 25 ng/mL (44–174) and IGFBP-3 less than 0.5 µg/mL (1.3–3.5) were both very low. Growth hormone stimulation testing peak of 1.1 ng/mL. DNA sequencing of the GH-1 gene found a heterozygous sequence variance.

Interventions: Growth hormone (GH) therapy was started at 0.27 mg/kg per week. Headaches began 5 days later, likely because of increased intracranial pressure, so GH was stopped and the dose reduced by one third, which was tolerated. She has grown about 12 cm during the first 10 months but is still –3.5 SD.

Discussion/Recommendations: Failure to thrive in the first 2 years of life rarely has an endocrine etiology. In this case, recognizing the importance of the family history and better compliance with follow-up care of the older siblings might have resulted in earlier diagnosis and treatment. The headaches, likely due to benign intracranial hypertension, suggests that this complication of GH therapy might be more common in children with this rare and severe form of GH deficiency, so starting GH at lower doses than usual would be prudent.

doi:10.1016/j.pedn.2012.03.013

Standardization of Endocrine Nursing Practice: Establishment of a Special Interest Group

Lisa Michele Pincham MSN, RN, Isabel Couto MSN, RN, CPN

Children's National Medical Center, Washington, DC

In October 2010, a Special Interest Group (SIG) was initiated to support endocrine nursing practice at an urban medical facility. A