Clinical Implications: Using a three-bag system within a DKA order set creates a streamlined process that reduces frequency of verbal orders and creates efficiency for nurses. Further study with a larger sample size is warranted to verify additional benefits of using a three-bag system.

doi:10.1016/j.pedn.2012.03.008

A Case of Type 1 Diabetes in a Toddler With a Family History of Neonatal Diabetes

Diane DiFazio MSN, CRNP
Children's Hospital of Philadelphia, Philadelphia, PA

Patient Demographics: The patient is a 33-month-old Caucasian female diagnosed with Type 1 diabetes mellitus (T1D).
Clinical Presentation: L. K. presented with 9-day history of polydipsia and polyuria. At home, the urine revealed positive ketones, and a blood glucose checked on home glucometer was 542 mg/dL. L. K. was transferred to a children's hospital and admitted with hyperglycemia and ketonuria without acidosis. Glucose management in the hospital was difficult because of hyperglycemia during the day and hypoglycemia at night without the initiation of insulin.
Past History: L. K. was born to a mother who was treated with insulin since the newborn period and said she was "born without a pancreas." L. K.'s mother is one of six children born to parents without diabetes. Two of L. K.'s mother's siblings have T1D, and two other siblings without diabetes have children with T1D. L. K. has a 4-year-old sibling without diabetes.
Evaluation: A HgbA1C was 9.1%, and a diabetes autoimmune panel revealed positive insulin antibodies of 24 uU/mL and positive ICA512 antibodies of 3.6 U/mL. Mutational analysis for PDX1 (encodes IPF1/associated with MODY Type 4) was performed and was negative.
Interventions: L. K.'s ketosis resolved, and she was initially treated with 2 U of lantus in the morning. She was discharged on 2 U of Lantus in the morning and 0.5 U of Humalog for blood glucose levels greater than 350 mg/dL.
Discussion/Recommendations: It is unclear whether L. K.'s mother had neonatal diabetes or a defect in PDX1, a very rare condition that results in pancreatic agenesis when homozygous for the mutation. Heterozygous mutations result in MODY Type 4. There are little data regarding the risk of T1D in children of mothers with neonatal diabetes, whereas children of mothers with MODY 4 are at risk for Type 2 diabetes. Family history is vital in newly diagnosed patients with T1D, and although neonatal diabetes and pancreatic agenesis are rare conditions, when it is present in a family member, those patients should be observed to see if there is transmission of a monogenic form of diabetes.

doi:10.1016/j.pedn.2012.03.009

Congenital Hyperinsulinism Associated With Beckwith Wiedemann Syndrome

Enyo Dzata MSN, CRNP
Congenital Hyperinsulinism Center, The Children's Hospital of Philadelphia, Philadelphia, PA

Patient Demographics: J. D. is a 9-month-old Caucasian male with hyperinsulinism (HI).
Clinical Presentation: J. D. reportedly had hypoglycemia at birth (exact data unavailable). He was discharged day of life (DOL) 3, when blood glucose (BG) levels "stabilized." On DOL 4, he had a seizure, was taken to the emergency room, and had a dangerously low BG level of 6 mg/dL (70–110). His medical evaluation was consistent with HI, but he failed medication therapy. He was discharged home on continuous feeds, but over the next several months had persistent hypoglycemia with BGs less than 50 mg/dL. Therefore, he was readmitted to his local hospital. Without additional treatment options, he was transferred to a hyperinsulinism center for further evaluation.
Past History: J. D. was the product of a pregnancy complicated by maternal lupus. He was full term and weighed 4.6 kg (large for gestational age).
Evaluation: HI was confirmed when laboratory values revealed a detectable insulin level, suppressed serum beta-hydroxybutyrate, and an inappropriate glycemic response to glucagon at the time of hypoglycemia. HI genetic testing was negative. An 18 F-DOPA PET scan of the pancreas suggested a possible focal lesion in the pancreatic head causing HI. During exploratory surgery, there was no evidence of focal or diffuse HI. The pathology of J. D.'s pancreatic biopsies indicated diffuse islet cell hyperplasia. J. D. had hemihypertrophy, raising the suspicion of Beckwith Wiedemann syndrome (BWS). Mosaic BWS was diagnosed, with a mutation on chromosome 11 (11p15.5p11.11).
Interventions: A gastrostomy tube was placed for continuous enteral administration of dextrose. J. D. maintained BGs greater than 70 mg/dL for 12 hours on a glucose infusion rate of 5 mg/kg per minute. He was discharged home on this regimen.
Discussion/Recommendations: BWS is frequently associated with HI. The clinical course and response to treatment have been variable, and medical therapy may not be successful. Further research is needed to analyze this association and treatment modalities. The genetic mutations in both HI and BWS are on chromosome 11, raising the question of a genetic link. Alternatively, because BWS is an overgrowth syndrome, perhaps the HI is simply an effect of islet cell hyperplasia.

doi:10.1016/j.pedn.2012.03.010

Unexplained Weight Loss in Two Growth Hormone-Deficient Adolescent Males

Cynthia Gordner BS, RN
Penn State Hershey Children's Hospital, Hershey, PA

Patient Demographics: Patient A is a 15 ½-year-old Caucasian male. Patient B is a 17-year-old Caucasian male.
Clinical Presentation: Patient A has been followed in an endocrine clinic since the age of 18 months with growth hormone (GH) and thyroid deficiencies. GH was discontinued 4 months prior because of growth completion (bone age 16y 6 m @ 15y 1 m). He had an appendectomy 1 month ago and reported diminished energy level and a 15-lb weight loss despite adequate oral intake and absence of gastrointestinal symptoms. No acute illness was noted. Patient B has been followed in endocrine clinic since age 7 years with growth hormone deficiency (GHD). GH was discontinued 6 months prior because of poor compliance (bone age 14y @ 15y 5 m). He reported a 20-lb weight loss and diminished energy levels. No changes had occurred in his medical regimen, and no other acute illness was present.

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 hypopitiutarism 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