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.

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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.

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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.

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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.