Clinical Presentation/Diagnosis: S.J. has a significant past history of neurofibromatosis Type 1 (NF1) and presented with signs and symptoms of early puberty manifesting as rapid linear growth, height above the 97% for age, and deepening of his voice, which had been noted by his primary care physician.
Past History: S.J. was diagnosed with NF1 at 4 years of age with multiple café-au-lait spots and freckling throughout his trunk and neck areas. The initial magnetic resonance imaging (MRI) of his brain and spinal cord revealed an optic-nerve glioma. Subsequent MRIs demonstrated extension of the neurofibromatosis into his hypothalamus. There is no family history of NF.
Evaluation: S.J. presented to the endocrine clinic at 8 years of age with his height 148.9 cm, and weight 45.6 kg (both >95%). Physical examination findings included Tanner Stage III to IV pubic hair, a pubertal phallus with testicular volume of 12 to 15 ml. Bone age at 13 years. Review of linear growth data revealed that he had been above the 95th% from age 4. Laboratories included a normal Free T4, TSH, and Prolactin. Measurement of IGF-1 was elevated for age at 411 ng/ml (113–261 ng/ml) but normal range for his bone age. LH 4.42 μIU/ml (0–2.3 μIU/ml), FSH 3.65 μIU/ml (0.26–3.0 μIU/ml), and testosterone 185 ng/dl (3–10 ng/dl) were consistent with central pubertal activation.
Interventions: After confirmation of central precocious puberty, S.J. began a GNRH analog (15 mg every 28 days) to suppress his puberty. Despite evidence of a suppressed gonadal axis, his growth velocity continued to be increased. Repeat IGF-1 of 827 ng/ml (113–261) was suggestive of growth hormone (GH) excess. A GH suppression test showed elevated GH levels confirming the diagnosis of gigantism. A somatostatin analog was added to his medical therapy.
Discussion/Recommendations: S.J. has both central precocious puberty and gigantism of hypothalamic origin. Gigantism is a rare complication of neurofibromatosis. The combination of both diagnoses involves a complicated medical management and creates challenges for the endocrine team. The patient and family require education about his condition and the necessity for close follow-up.

doi:10.1016/j.pedn.2009.02.008

008 – An 11-Year-Old Male With Type 1 Diabetes was Diagnosed With Celiac Disease Despite Being Negative for Serological Markers for the Disease

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Patient Demographics: Eleven-year-old Caucasian boy.
Clinical Presentation/Diagnosis: J.H. when 6 years old was diagnosed with Type 1 diabetes. He did well on Novolog insulin via insulin pump. His serum glucose levels for the most part remained in the target range, and he never required hospitalization for diabetic ketoacidosis. His HbA1c's ran between 6.0% and 7.4%. At 11 years of age, he presented in the diabetes clinic for routine follow-up. He complained of intermittent epigastric discomfort and also occasional loose stools.
Past history: He has Type 1 diabetes and a first-generation relative who was diagnosed with celiac disease.
Evaluation (Studies/Assessment): Physical examination revealed a healthy-appearing boy. The weight was 55.2 kg (95th percentile). The height was 158.3 cm (95th percentile). Vital signs were normal. A complete physical examination was normal. With the incidence of celiac disease up to 10% for children with diabetes, blood was obtained for celiac disease and genetic markers. The antigliadin IgG was 6.2 U/ml (nl <10.0), antigliadin IgA was 0.7 U/ml (<5.0), antihuman tissue transglutaminase IgA was 0.5 U/ml (nl <4.0), antiendomysial IgA was negative, total serum IgA 149 mg/dl (41–395). The patient was positive for DQ8 heterozygocity, which increased the risk factor two times over the general population (moderate risk). Because of continued symptoms unresponsive to medical therapy, upper endoscopy with small bowel biopsy was undertaken, which revealed villous blunting and intraepithelial lymphocytes consistent with celiac disease.
Interventions: He has been placed on a gluten-free diet.
Discussions/Recommendations: Children with Type 1 diabetes have a higher incidence of celiac disease than the general population. Our child had negative serological markers for the disease, but his GI symptoms continued and DQ8 testing suggested that he was at moderate risk. In addition, he had a relative with celiac disease. Although serological markers should be adequate screening for children with Type 1 diabetes, those who have continued gastrointestinal symptoms, or relatives with the disease should be furthered evaluated by a pediatric gastroenterologist for endoscopy and biopsy, the gold standard for diagnosis.

doi:10.1016/j.pedn.2009.02.009

009 – Injection Force Dynamics of Improved NordiFlex [somatropin (rDNA origin)] Versus the Current Version of Norditropin NordiFlex

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Background: An improved pen device with soft push technology for administering growth hormone (GH) NordiFlex (trademark of Novo Nordisk Health Care AG) has been developed.
Aims: The aim of this study is to evaluate dose force improvements of NordiFlex (5, 10, and 15 mg) compared with the current version of Norditropin NordiFlex (registered trademark of Novo Nordisk Health Care AG; 5, 10, and 15 mg).
Methods: Because the viscosity of the 5, 10, and 15 mg Norditropin formulations are similar and the mechanics of the three injection pens (NordiFlex 5, 10, and 15 mg) are the same, the injection force dynamics were evaluated in the improved NordiFlex 10 mg/1.5 ml versus the current version of Norditropin NordiFlex 10 mg/1.5 ml. A NovoFine (registered trademark of Novo Nordisk A/S) 30G 8-mm or a NovoFine 32G 6-mm needle was used. A maximum dose of 3.0 mg (0.45 ml) was delivered at speeds of 4, 6, and 8 mm/seconds and repeated three times per pen using 36 pen samples of each pen device tested.
Results: Dose force was significantly lower for the improved NordiFlex than for the current version of Norditropin NordiFlex at

each dose speed tested. The reduction in dose force for the improved NordiFlex versus the current version of Norditropin NordiFlex was between 29% and 42% with NovoFine 30G 8-mm needle and between 29% and 39% with NovoFine 32G 6-mm needle. The greatest reduction in dose force (42%) was observed at the lowest push button speed 4 mm/seconds and NovoFine 30G 8-mm needle.

Conclusion: Compared with the current version of Norditropin NordiFlex, the improved NordiFlex showed a 29% to 42% reduction in dose force, which was similar for both the NovoFine 30G and 32G needles. The greatest reduction in dose force (42%) was observed at the lowest push button speed of 4 mm/seconds.

Clinical Implications: The lower dose force of the improved NordiFlex may make injections easier, thus potentially improving adherence with the prescribed GH treatment regimen.

doi:10.1016/j.pedn.2009.02.010

010 – NordiCare NOW: Effects on Communication and Efficiency—Nurses Perspectives on its Impact from Four Pediatric Endocrinology Sites

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As insurers tighten requirements to prove medical necessity and increasingly mandate product switching, health care providers carry the burden of assuring that patients receive prescribed care. This requires providers to support and track patients through an arduous and ongoing process to obtain and maintain access to Norditropin [somatropin (rDNA origin)]. The complexity is compounded by increasing patient loads and frequently shifting formularies. Convenient and effective support for this process is valued, especially when it can be personalized to fit the needs of individual health care providers. Norditropin requires a long-term commitment by the patient, caregiver(s), and health care providers, beginning with prior authorization and extending throughout the treatment process. This process involves many challenges, from the initial authorization of the medication, to providing injection training, to the need for reauthorizations when approvals have expired. For patients prescribed Norditropin, NordiCare is a comprehensive support program available to health care providers, patients, and caregivers that address these challenges. Continuous and clear communication is needed between NordiCare and health care providers to assure that relevant information is shared. To address this need, in June 2008, the NordiCare NOW Web site was launched. NordiCare NOW is a secure yet convenient Web site for health care providers that assist them to stay up-to-date on case status through an online/Web-based communication avenue with NordiCare. It is specifically designed to protect personal health information while providing real-time 24/7, on-demand access to a log of all communications related to the patient and to the patients' reimbursement status. Many features are available to help health care providers save time and stay in control of their patients' cases in a personalized manner, from the initial prior authorization throughout the entire duration of the patient's treatment. This poster will review this new service, providing an overview of the Web site. Specific features of the Web site will be presented including how these features have facilitated the overall process at four pediatric endocrinology sites, in both academic and private practice settings.

doi:10.1016/j.pedn.2009.02.011

011 – Experience at Texas Children's Hospital: Transitioning Provocative Stimulation Testing From The Outpatient Clinic to an Infusion Center

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Provocative stimulation is an important tool used to evaluate patients for certain endocrinopathies. Although there are many testing protocols and stimulating agents, the basic testing principle remains consistent. Most stimulation testing protocols require baseline blood sampling of hormones. The patient receives a stimulating agent by subcutaneous, intramuscular, or intravenous injection; and additional blood samples are collected at specified time intervals to measure the patient's hormonal response to the stimulus given. Stimulation testing is useful in diagnosing growth hormone deficiency, adrenal disorders, and pubertal disorders. Performing stimulation testing in the outpatient clinic has become increasingly difficult. The testing procedure has not changed, but other challenges have arisen. Stimulating agents have been intermittently, and sometimes become permanently, unavailable from pharmaceutical manufacturers. Some protocols have been revised to use alternate stimulating agents; however, many protocols now require additional blood samples and a longer evaluation period for the patient. Our clinic recognized that the increased length of stimulation testing visits impacted staffing and decreased the number of tests completed daily. Occasional medication errors were reported; possibly related to staff/patient ratios. Performing fewer tests increased patients' wait time once scheduled. In addition, we noted that declining insurance reimbursement and increasing office visit length diminished the profitability of performing these tests in the outpatient clinic. Many clinic offices look to infusion centers to perform stimulation testing. Infusion center staff generally possess the technical and observational skills necessary to efficiently initiate intravenous catheter placement and monitor patients throughout their procedure. Staff must be instructed on specific test preparation (i.e., fasting, medication cessation, etc.) and potential stimulating agent side effects or contraindications. We found that compiling a stimulation test protocol manual was helpful. Staff must also have access to a pharmacy, emergency medical equipment and additional personnel if they should be needed. Since performing stimulation tests in our hospital's infusion center, there have been no reported medication errors. When staffed by knowledgeable, trained personnel, ambulatory infusion centers provide an excellent alternative location for patients to have provocative stimulation testing performed.

doi:10.1016/j.pedn.2009.02.012