



# ABSTRACTS OF THE PEDIATRIC ENDOCRINOLOGY NURSING SOCIETY

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## A Survey of Knowledge Related to Cystic Fibrosis-Related Diabetes

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**Background:** Cystic fibrosis (CF) is the most common life-threatening autosomal recessive genetic disease in the United States. Cystic fibrosis-related diabetes (CFRD) is the most common comorbidity of CF, and as patients age, the likelihood of developing diabetes increases. CFRD leads to decreased lung function, poor nutritional status, and decreased survival rates. Symptoms can be subtle and may be overlooked. The extent to which individuals with CF know about CFRD is unknown.

**Aims:** Assess knowledge of CFRD in adults with CF. Describe where information related to their disease is obtained. Examine relationships between measures.

**Methods:** A cross-sectional descriptive study was used. Adults (>18 yrs) with CF were recruited during an outpatient CF visit and asked to complete a 15-item CFRD Knowledge Survey. The survey included 10 items on knowledge of CFRD, 2 items on obtaining information about CF, and 3 items on experience with diabetes.

**Results:** Twenty-six individuals with CF participated, 70% were male, mean (SD) age was 27.73 (10.23). Regarding knowledge, 92% had heard of CFRD, 65% knew symptoms of CFRD, and 58% knew how the diagnosis was made. A majority (65%) reported they seek information related to CF only when necessary. The three most common sources for obtaining information were internet, physician, and clinic. The total knowledge score was significantly correlated with the experience score ( $r = 0.50, p = 0.009$ ), and understanding the importance of knowing that you have CFRD ( $r = 0.80, p < 0.001$ ). The experience score was also significantly correlated with understanding importance of knowing if you have CFRD ( $r = 0.60, p = 0.001$ ). Understanding the importance of knowing if you have CFRD was also significantly correlated with being diagnosed ( $r = 0.43, p = 0.028$ ).

**Conclusions/Clinical Implications:** As the number of patients diagnosed with CFRD continues to increase, both pediatric and adult endocrine nurses need to be educated and equipped to successfully manage this patient population.

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## How Much Is Enough? The Usefulness of Peak Cortisol Levels in Identifying Adrenal Insufficiency in Children Undergoing Insulin Tolerance Test for Short Stature

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**Background:** Identification of asymptomatic patients with subtle dysfunction of the hypothalamic–pituitary–adrenal (HPA) axis is a diagnostic challenge. HPA axis response to hypoglycemia during an insulin tolerance test (ITT) is considered the gold standard in the evaluation of suspected adrenal insufficiency, especially in the setting of growth hormone deficiency (GHD). The short 1  $\mu\text{g}$  cortrosyn stimulation test (CST) is also an accurate and practical screening test for the adequacy of ACTH reserve.

**Aim:** The aim of this study was to determine the utility of ITT and CST in determining adrenal insufficiency in children undergoing evaluation for GHD.

**Methods:** Retrospective analysis of the integrity of the HPA axis in subjects evaluated for potential GHD during the 5-year period from January 2005 to December 2009 was performed. A total of 520 children underwent ITT for potential GHD, of which 388 (75%) were boys. The mean age was 10.9 years ( $SD = 3.6$  years), and the mean maximum cortisol was 18.3  $\mu\text{g}/\text{dL}$  ( $SD = 5.6$ ).

**Results:** Of the 520 children who underwent ITT, 240 (46%) had a peak cortisol less than 18  $\mu\text{g}/\text{dL}$  and were considered positive for possible adrenal insufficiency. Of those with a positive ITT, 131 underwent a CST, of which 44 (34%) continued to remain positive. Of these 44 children, 17 had a second CST, and 12 (71%) remained positive. Fourteen children who had cortisol levels less than 18  $\mu\text{g}/\text{dL}$  following the initial ITT underwent a metyrapone test, all of whom proved adrenally sufficient.

**Conclusions:** In participants with a peak cortisol level less than 18  $\mu\text{g}/\text{dL}$  during ITT, a subsequent 1  $\mu\text{g}$  CST excluded adrenal insufficiency in two thirds of the subjects. Furthermore, a second CST in participants with a peak cortisol level of less than 18  $\mu\text{g}/\text{dL}$  during the first CST was unlikely to exclude adrenal insufficiency.

**Clinical Implications:** The ITT is useful in the evaluation of participants with potential GHD because it assesses the adequacy of both the growth hormone and HPA axis. In those children with

subnormal cortisol response during ITT, only a single 1 µg CST is needed for further assessment of adrenal insufficiency.

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#### Concept Analysis of Fear: Focus on Childhood Fear and Implications for Endocrine Nursing

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**Background:** Clinicians often underestimate the fear that children have with frequent needlesticks required in the care of many endocrine disorders (Bracha, 2004). While fear lessened, 28% of young children with Type 1 diabetes mellitus continued to report needle fear 6 months after diagnosis (Howe et al., 2011).

**Aims:** A concept analysis of fear with a focus on childhood fear was undertaken to define the antecedents, criteria, and consequences of fear to develop a theoretical definition of fear that informs nursing interventions to help children cope with fear.

**Methods:** A literature review was conducted on fear theories, neurophysiology of fear, and childhood fear. A brief review of animal fear was also included because it offers an interesting perspective. The methods of Walker and Avant (2005) were used to describe the criteria, antecedents, and consequences of fear.

**Results:** Two leading theories include fear as a conditioned response or fear as an evolutionarily adaptive response to imminent threat. There are two key criteria for fear: 1) a neurophysiologic response that triggers an autonomic physiologic response; 2) a behavioral response: freeze, flight, fight, fright, and faint (Bracha, 2004). The *antecedent* to fear is the presence of a real or perceived threat (Forsner et al., 2009). While the range of threats seem infinite, the subjective experience of what is feared and the fear itself are influenced by developmental age, vulnerability, past experiences, and genetic factors. The *consequences* of fear include conditioning and avoidance.

**Conclusion:** Fear is a conditioned or evolutionary adaptive, neurophysiologic and behavioral response that is part of the normal developmental experience in childhood to real or perceived threat. A model and contrary case will be presented.

**Clinical Implications:** Needle fear begins as an evolutionary adaptive response but with negative experiences, the child's fear response escalates. Desensitizing interventions with incremental exposure to needles help children cope. A nurse can turn a negative moment for a child fearful of needles into a powerful, empowering experience.

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#### A Prospective Study of Growth and Development of Children Recently Adopted From Orphanage Care

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**Background:** More than 200,000 international adoptions by U.S. families occurred between 1999 and 2010. Prior studies suggest that the effects of institutionalized care on growth and development may not be fully reversible.

**Aims:** The exact mechanisms through which early life stress affect biobehavioral outcomes have yet to be determined, but environ-

mental influences could regulate both biological and psychosocial development through an effect on the hypothalamic–pituitary–adrenal (HPA) axis. Children were evaluated shortly after adoption for baseline HPA axis function and its association with biobehavioral measures.

**Methods:** This is a prospective study of 10 recently adopted children (19–40 months) with an average time spent in orphanage care of 23.6 ± 9 months. Eligible participants had no history of significant medical, developmental, or behavioral problems. Anthropometric measurements, physical examination, HPA axis tests, bone age, neurocognitive testing, and behavioral questionnaires were evaluated.

**Results:** Shortly after adoption by a U.S. family (1.8 ± 1 months), height standard deviation unit (Ht SDU) was  $-1.6 \pm 0.8$ ; weight SDU was  $-0.9 \pm 1.2$ ; and head circumference SDU (HC SDU) was  $-1.8 \pm 1$  (WHO growth standards). Bone age was consistent with chronological age in four, advanced in three, and delayed in three children. Time in orphanage care was positively associated with serum cortisol ( $r = .64, p < .06$ ) and negatively associated with Ht SDU ( $r = -.63, p < .05$ ). Neurocognitive testing (Bayley-III) showed significant delays in all scores. HC SDU was positively associated with cognitive and receptive language subscales on the Bayley III ( $r = .62$  and  $.69$ , respectively). Child Behavior Checklist response endorsed one child with attention/withdrawal symptoms. However, response on the Behavior Rating Inventory of Executive Function endorsed clinically significant inhibitory control in half the children, and subscale scores for behavioral regulation were positively associated with HC SDU ( $r = .9, p < .05$ ). HPA axis testing revealed no significant abnormality.

**Conclusion:** Children adopted from orphanage care experience a negative impact on linear growth, HC, cognitive, and behavioral development. Prenatal factors and time in orphanage care were associated with negative effects on linear growth, serum cortisol, cognitive, and behavioral outcomes.

**Clinical Implications:** Careful assessment of prenatal and environmental risk factors will help to identify children at risk for untoward effects on growth, cognitive, and behavioral outcomes and target early interventions.

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#### Social–Demographic Factors Associated With Pediatric Diabetic Ketoacidosis Admissions in Southern West Virginia

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**Background:** Diabetic ketoacidosis (DKA) is a well-known complication in children with Type 1 diabetes, with a mortality rate estimated at 2%. Sparse data are available from the literature describing the sociodemographic factors associated with DKA admissions in children. A previous study identified that children with Type 1 diabetes who are of non-Caucasian race and those with Medicaid had increased incidence of DKA admissions.

**Aims:** The aim of this study was to identify the sociodemographic factors associated with DKA admissions including type of insurance coverage, income by county, race, gender, and HbA1c in West Virginia, a primarily rural part of Appalachia.

**Methods:** A retrospective chart review was conducted of patients, aged 1 to 18 years, with known Type 1 diabetes with DKA admitted