ABSTRACTS OF THE PEDIATRIC ENDOCRINOLOGY NURSING SOCIETY

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 μg cortroysn 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 μg/dL ($SD = 5.6$).
Results: Of the 520 children who underwent ITT, 240 (46%) had a peak cortisol less than 18 μg/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 μg/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 μg/dL during ITT, a subsequent 1 μg 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 μg/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

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