

Informational

011–X-Linked Hypophosphatemia (XLH): New Knowledge for Nurses Regarding Genetics, Pathophysiology and Clinical Presentation

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Background: Over 80 years ago Fuller Albright reported a vitamin D-resistant form of rickets, which is likely the initial description of X-linked hypophosphatemia (XLH). Recently the genetic basis of the disease has been identified and a more complete understanding of the pathophysiology has emerged.

Purpose: To increase awareness of the latest published data on the genetics, clinical manifestations, differential diagnoses, biochemical findings, as well as growth and development of patients with XLH.

Description of Topic: XLH is a rare, progressive, life-long disorder and the most common form of heritable rickets. The estimated prevalence of XLH is 1:20,000 to 1:25,000. XLH is caused by loss-of-function mutations in the phosphate regulating endopeptidases on the X-chromosome (PHEX) gene, leading to high circulating levels of fibroblast growth factor 23 (FGF23). Over 300 PHEX mutations have been reported. An X-linked dominant inheritance pattern is typical; however, de novo PHEX mutations are reported in up to 20–30% of the cases. Excess FGF23 increases urinary phosphate losses with consequent hypophosphatemia, resulting in rickets and osteomalacia. Clinical manifestations may include lower limb deformities, short stature, bone and joint pain, dental abscesses, delayed walking, and gait abnormalities. Neurological features may include Chiari 1 malformation and craniosynostosis. Low serum phosphate levels, a low renal tubular threshold for phosphate reabsorption (TmP/GFR), and low or normal circulating 1,25(OH)₂D are characteristic biochemical findings in patients with XLH. The symptoms of XLH vary among individuals and while there is similar pathophysiology for children and adults, clinical manifestations can differ. Debilitating consequences in adults include osteoarthritis, enthesopathy, spinal stenosis and pseudofractures. In addition, complications of conventional medical therapy (phosphate salts and active vitamin D analogs) include nephrocalcinosis and hyperparathyroidism.

Clinical Implications: Previously, XLH was considered a disorder that manifests only during growth; however, adolescent patients require attention along with a smooth transition to adult care. In addition, recognition of the complex disease features of XLH is essential for accurate diagnosis and management. Pediatric endocrine nurses are well qualified to provide the latest disease state education to patients and families, and to encourage routine clinical evaluation to assess treatment response, disease progression, and therapeutic complications.

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012–Food Literacy: Can Pediatric Endocrinology Nurses Focus Upstream?

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Background: American families are spending less time procuring, preparing and cooking meals at home. As well, there is less time dedicated to nutrition education in schools. Food literacy is defined as “a collection of inter-related knowledge, skills, and behaviors required to plan, manage, select, prepare, and eat foods to meet needs and determine food intake, as well as, the scaffolding that empowers individuals, households, communities or nations to protect diet quality through change and support dietary resilience over time.” Adolescents generally lack food literacy skills and increasingly consume food away from home with fast food meals and processed snacks. Approximately 1 in 3 adolescents in the U.S. are obese, increasing their risk for diabetes and cardiovascular disease. Upstream efforts continue to be explored to combat rising obesity rates. Programs that promote food literacy in adolescents have the potential to influence healthy lifestyles.

Purpose: To inform pediatric endocrinology nurses about food literacy, its components, and existing food literacy education programs ready for implementation.

Description of Topic: A literature review on food literacy was completed. High food literacy has been associated with improved eating of a healthy diet, including more fruits and vegetables and less fast food. Food literacy programs should address adolescents’ knowledge, skills, and attitudes to make healthy diet choices. Potential topics include gardening, recipe reading, food label reading, learning about healthy vs. unhealthy foods, food preparation and cooking, animal welfare, and farm to table slow food concepts. The USDA SNAP-Ed website provides well developed, user-friendly food literacy teaching ideas with accompanying learning objectives, lesson plans, and teaching strategies that could readily be implemented by pediatric endocrinology nurses.

Clinical Implications: Pediatric endocrinology nurses typically provide care to adolescents with obesity and its related comorbidities. Pediatric endocrinology nurses have an opportunity to expand their practice to primary and secondary prevention of obesity, using upstream health education to promote adolescents’ food literacy. By partnering with schools, churches, Boys and Girls clubs, and Scouts troops, pediatric endocrinology nurses may offer health education, building food literacy skills in whole communities with the ultimate goal of slowing the rise in obesity rates.

Food literacy has been described as “the ability of an individual to understand food in a way that they develop a positive relationship with it, including food skills and practices across the lifespan in order to navigate, engage, and participate within a complex food system. It’s the ability to make decisions to support the achievement of personal health and a sustainable food system considering environmental, social, economic, cultural, and political components.

The core of food literacy is the adolescent’s ability to use food knowledge and skills to make healthy dietary choices and encompasses aspects of planning and managing, selecting, preparing, and eating healthy foods.

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013–Delivering High Quality Pediatric Diabetes Care Beyond Hospital Limits

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Background: Pediatric diabetes is a complex yet delicate condition that requires well-orchestrated interdisciplinary collaboration usually found within facilities providing high level acute care. As part of a large tertiary care centers community expansion, we were

tasked to develop a high functioning pediatric diabetes clinic providing the same level of care utilizing resources outside the hospital setting. Challenges included integration of new, complex workflows among other sub-specialty services, coordination of limited interdisciplinary support, and staff development in providing higher levels of care.

Purpose: To transition pediatric diabetes care to outreach locations while maintaining high quality, efficient family-centered care in a multidisciplinary format.

Description of Topic: The overall objective was to transform standard practices into best practices with limited resources without any negative effects in patient outcomes. The first step was to conduct a needs assessment to identify all components required to meet patient outcomes. Needs identified required collaboration among nursing leaders, physicians, educators, pharmacists, social workers, Information Technology personnel, drug/Durable Medical Equipment representatives, Child Life, local school districts, and families.

In collaboration with the aforementioned, we successfully developed a pediatric diabetes clinic through the following achievements: Provided patient access to social work, diabetes educator, and dietician via telemedicine; Created efficient clinic workflows through the development of electronic nursing assessment tools, known as “smart-sets” specific to the diabetes patient; Provided same level of in-person interpreter services through use of video interpreting system; Enhanced and implemented new school medication form for school nurses to more effectively care for diabetes patients; Created staff development tools that provides training and education necessary for care team members managing complex and acute conditions related to pediatric diabetes

Clinical Implications: We were successful and deemed a best practice clinic with many of our new processes adopted by other locations. Our newly created “smart-sets” provide care team members with efficient and comprehensive assessment tools that produce optimal patient outcomes while providing more opportunities for patient education and reinforcement of compliance measures. Additionally, our innovative approach allows us to provide remote or in-person access to services needed to provide comprehensive, holistic pediatric diabetes care.

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014–ENDO Handbook: Endocrine Nurse Detailed Orientation Handbook

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Background: Orientation has been an essential part of new nurses training in the inpatient setting for decades. This training helps mold the new nurse's confidence, job satisfaction, and retention. Often this orientation in the outpatient setting falls short in comparison. However the benefit of a structured usually held for inpatient nurses only is also applicable for outpatient clinic nurses. In addition to confidence, job satisfaction and retention, many outpatient clinics are specialized so additional training is necessary. Having a structured orientation will also ensure consistency with training.

Purpose: To provide a structured outpatient clinic orientation manual to ensure the new clinic nurse is fully prepared to function in an endocrine specialty clinic setting. The manual will also serve as a resource for experienced clinic nurses.

Description of Topic: This project is to help improve the orientation process for an outpatient endocrine clinic nurse. Information and educational materials are collected and structured into one orientation manual for the trainee to utilize. It will include a list of department members and their roles, daily tasks including policies

and procedures associated with these tasks, documentation of clinic tasks, stimulation testing procedure and protocol, patient and nursing education, as well as task competency lists for the orientee to complete. This orientation manual will remain in the clinic setting so all nursing staff may reference. It will be updated accordingly as tasks and policies change.

Clinical Implications: A structured orientation handbook will provide reference materials, educational materials, and task checklists for the nurse to follow and utilize during their training. Structured orientation programs have been found to improve a nurse's confidence in their new role. When nurses are confident in their roles, stress is reduced and retention and turnover rates are decreased. Having one orientation manual will provide consistency with training, therefore all patient education will be consistent and all clinic tasks will be completed in the same manner.

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015–Adrenal Insufficiency in Children: Teaching Nurses to Teach Caregivers

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Background: Adrenal insufficiency (AI) is the impaired synthesis and release of adrenocortical hormones due to various mechanisms (e.g., autoimmunity, genetic, ACTH deficiency, and corticosteroid-induced). AI affects quality of life and is associated with increased morbidity and mortality, especially in younger patients. Children are at highest risk during times of physical stress. Many children do not receive stress doses of medication when needed. Relatively novice endocrine nurses do not feel adequately prepared to educate children, adolescents, and families about AI and its treatment.

Purpose: The purpose is to increase nursing knowledge, skills, and competency in teaching caregivers about disease process and management of adrenal insufficiency day to day and during times of physical stress.

Description of Topic: The poster will describe the action steps used to prepare endocrine nurses to teach caregivers about adrenal insufficiency and its management. The PRECEDE-PROCEED model will provide a framework for education. The project leader will secure organizational support, work with endocrinology providers to standardize stress steroid dosing, identify or modify evidence-based teaching resources, and develop charting templates and smart phrases for documentation. Components of the education plan include an overview of AI, pathophysiology (including congenital adrenal hypoplasia, Addison's disease, hypopituitarism, and corticosteroid-induced adrenal insufficiency), daily medications, and prevention of adrenal crises (use of medical alerts; emergency hydrocortisone; and when to call clinic, go to ED, or call 911). Nurses will be mentored by an experienced pediatric endocrinology nurse.

Clinical Implications: A major role of pediatric nurses is teaching children, adolescents, and families. Ineffective education of caregivers about AI places children at risk for adrenal crises. Nurses need to be able to teach caregivers of newly diagnosed children with AI and properly triage phone calls from caregivers during illness or injury or prior to procedures. They also need to be knowledgeable about school emergency plans. Investing in the training of novice endocrine nurses about AI can empower caregivers to better manage this disease. During times of physical stress, adherence to stress dosing can reduce the need for emergency department visits and hospital admissions.

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