Provide Hereditary Tyrosinemia Type I (HIT) Caregiver Education based on Nursing Research and Evidence based Practice to Improve Patient Outcomes

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**Project Title:** Provide Hereditary Tyrosinemia Type I (HTI) caregiver education based on nursing research and evidence based practice to improve patient outcomes

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**Project Description:** This research project aims to understand the caregiver’s role in providing care to children with Hereditary Tyrosinemia Type I (HTI) and the importance of advocacy for this vulnerable aggregate population in the United States.

Hereditary Tyrosinemia Type I (HTI) is a genetic disorder caused by a deficiency of the funarlylacetate hydrolase (FAH) enzyme that results in an elevation of tyrosine, and other toxic metabolites without a clear genotype-phenotype correlation. The only available pharmacotherapy for HTI is nitisinone and it is a life-saving therapy for children with HTI. Prior to the use of nitisinone some children with HTI did not survive past early childhood, but now outcomes are improved. Although children are now surviving longer, researchers are finding changes in cognition and social function. The deficiency of FAH enzyme is associated with the accumulation of toxic metabolites and hypermyelination in an animal model of HTI studied at the University of Alabama in Huntsville (UAH).

Early treatment with low protein diet and nitisinone that is consistent decreases adverse effects by limiting cellular changes. Evidence based care for children with HTI can decrease morbidity and mortality. Literature reviews of the evidence and development of educational materials for families and caregivers is essential to improve health-related quality of life (HRQoL). The Revised Wilson and Cleary model for health-related quality of life (HRQoL) describes how an alteration in the environment can change outcomes for a child with HTI. Development of these educational materials is an environmental change that can improve outcomes.

Caregiver education units (CEUs) for HTI has the potential to significantly impact patient outcomes and is not currently available. The goal of this project is to design evidence based educational materials for caregivers of children and adults with HTI. Caregivers that understand their role in caring for children with HTI can improve patient outcomes and quality of life for both. Strengths based nursing that empowers the patient and family to gain greater control over their health should be used to educate families using an online platform for advocacy throughout the United States.
This proposed study is part of a research program regarding HTI in collaboration with Dr. Elizabeth Barnby DNP, ACNP-BC, FNP-BC, CRNP. This proposed study will create research based products that can be utilized to educate caregivers regarding HTI and promote advocacy for this vulnerable aggregate.

**Student Duties, Contributions and Outcomes:** The student researcher’s specific duties will begin with a thorough literature review related to the topic, the caregiver’s role providing care to children and adults with HTI. The literature review should consist of a minimum of 15-20 peer-reviewed articles outlining the caregiver’s role. The student will then assist with the development of evidence-based education materials which may include but are not limited to brochures, PowerPoint presentations, and other electronic word documents. The tangible contributions made by the student would include an increased knowledge of the rare pediatric disease HTI, a review and analysis of the caregiver’s role within the aggregate population of children and adults with this diagnosis, and the ability to disseminate their new knowledge. The student researcher will have the opportunity to explore this topic within the United States and benefit from the expertise of nurse scientists by working with College of Nursing researchers and in consultation with clinical experts throughout the southeastern region of the United States by presenting at the annual conference at Southeastern Regional Genetics Group (SERGG) in Asheville, North Carolina. Specific outcomes the student would obtain from this experience would include an improved ability to complete scholarly literature reviews, increased knowledge and understanding of HTI, and a deeper knowledge of caregiver’s roles in the population with HTI.

**Faculty Requirements and Mentorship:** The student must be accepted into the College of Nursing and be in academic good standing. Ideally, the student will be currently enrolled in NUR 307 or have completed this course. The student must have a basic understanding of scholarly writing and be capable to create brochures, PowerPoint presentations, and electronic word documents. During this 10 Week RCEU experience Dr. Mark Reynolds and/or Dr. Elizabeth Barnby will supervise all research activities. Dr. Barnby will assist with mentoring the student and reviewing student designed documents for researched evidence-based best practices and correctness. The student will maintain weekly sessions for the purpose of reviewing and synthesizing literature, discussion of knowledge obtained through information gathered during the literature review, and later in the semester creating evidence based educational documents. Written and verbal feedback related to all aspects of the research process will be provided to the student researcher.