The Role of the Clinical Research Nurse in a Cystinosis Study

National Institutes of Health Clinical Center Nursing & Patient Care Services

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INTRODUCTION

- Clinical Research Nurses (CRNs) at NIH practice under a model of care encompassing 5 dimensions
- This model of care provides a framework for the CRN to support the unique care requirements of research participants through care coordination, human subject protection, and specialized clinical expertise
- The role of the CRN requires a skill set and knowledge base essential to the care of patients with rare disorders
- CRNs operationalized the model of care for an child admitted with acute symptoms of Nephropathic Infantile Cystinosis (NIC) in the spring of 2011

BACKGROUND

- NIC is a rare, autosomal recessive disorder characterized by faulty transport of the amino acid, cystine across lysosomal cell walls
- Most common cause of inherited Fanconi syndrome in children, which causes the majority of clinical symptoms
- Cystine crystals accumulate and damage tissue in the kidneys, eyes, brain, bones, muscles, and pancreas
- Early diagnosis and treatment with the cystine depleting drug, cysteamine (Cystagon ®), are essential to prevent tissue damage and optimize outcomes
- Disease complications can include blindness, fractures, diabetes, hypothyroidism, life-threatening electrolyte disturbances, and kidney failure

CASE

- A 2 year-old Bolivian child was diagnosed with NIC 5 months prior to her arrival at NIH
- Parents consented child to cystinosis natural history protocol
- Patient required urgent admission to 1NW pediatric unit for stabilization and initiation of Cystagon ® treatment

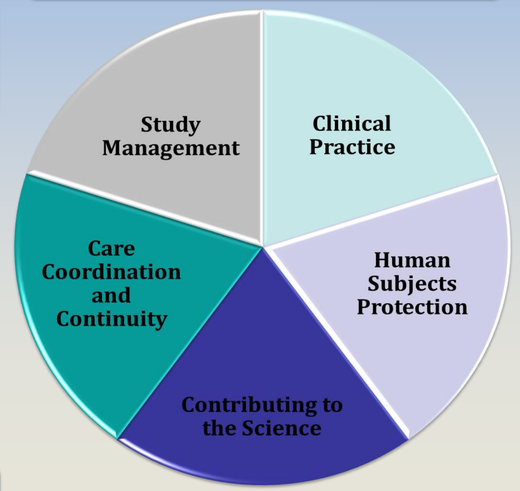
PROBLEMS

- Clinically unstable patient due to acute metabolic acidosis, hyponatremic dehydration, and hyperkalemia
- Frequent emesis, often resulting in expulsion of naso-gastric tube
- Acquired behavioral patterns: excessive water demand with refusal of other oral nutrition, resulting in inconsolable crying and vomiting if denied
- Malnourishment (<3rd percentile for weight/length for age), stunted growth, & borderline wasting
- Hypotonia and marked developmental delays- not walking or talking
- Rickets and bilateral hip dislocation

INTERVENTIONS

- CRNs assisted with the consent process & ongoing education of family related to the clinical research protocol
- Frequent laboratory monitoring and adjustment of intravenous electrolyte therapy
- In collaboration with physician and nutritionist, naso-gastric tube feeds were slowly advanced while administering intravenous sodium chloride infusion for resolution of hyponatermia associated dehydration
- Developed customized form (data collection document) and taught the parents to document water and nutrient intake as well as weighing of diapers for output estimations
- CRNs initiated a multi-disciplinary care conference which included: the research team, orthopedic surgery, endocrinology, rehabilitation medicine, nutritionist, social work, pediatric medical and nursing teams
- Taught parents proper care of newly placed percutaneous gastric tube

MODEL OF CARE



Coordinated and collected research specimens

Provided pediatric nursing care, collected clinical data, and assessed for study drug effects

Provided nursing leadership within the interdisciplinary team

Facilitated informed consent process & supported participation in the study

Coordinated interdisciplinary meetings & activities related to clinical care and the research protocol

Disseminated clinical best practices with nurse colleagues

CRN 2010 Domain of Practice Committee 2009. Building the Foundation for Clinical Research Nursing: Domain of Practice for the Specialty of Clinical Research Nursing. 2009. National Institutes of Health Clinical Center, Nursing and Patient Care Services. Available at *http://www.cc.nih.gov/nursing/crn/DOP_document.pdf*

OUTCOMES

- Patient stabilized from acute metabolic state
- Cystagon ® treatment initiated and tolerated
- Excessive water intake decreased and nutrient intake maximized
- Growth: BMI improved to 95 percentile at 6 month follow-up
- Naso-gastric tube switched to gastric tube
- Developmental advances: walking, running, playing, talking
- Attending feeding clinic
- Family knowledgeable about condition & clinical research protocol

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- Permissions obtained from family for photos and poster content