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March 1, 2010

Cystinosis Research Foundation
18802 Bardeen Avenue
Irvine, CA 92615-1521

Dear Cystinosis Research Foundation:

Enclosed please find the 6-month progress report for “Psychosocial Functioning in Cystinosis: Stress and Coping with a Chronic Illness”, (Doris Trauner, Principal Investigator).

We thank you for the opportunity to conduct this research.

Sincerely,

Doris Trauner, Ph.D.
UCSD Department of Neurosciences

Psychosocial Functioning in Cystinosis:
Stress and Coping with a Chronic Illness

Doris A. Trauner, M.D. Principal-Investigator
Amy M. Spilkin, Ph.D., Co-Investigator
Angela Ballantyne, Ph.D., Co-Investigator

Six-Month Progress Report - Cystinosis Research Foundation
9/1/09 – 3/1/10

A. SPECIFIC AIMS

The specific aims for this project are:

1) To examine which chronic illness-related symptoms lead to psychological distress in adolescents and adults with cystinosis.

2) To determine which coping strategies are most commonly used by individuals with cystinosis and their caretakers.

3) To determine whether individuals with cystinosis and their caretakers experience decreased quality of life compared with participants in another chronic disease group and a healthy control group.

4) To determine which variables aid or impede adherence to medical regimen in cystinosis patients. Given the life-saving nature of the prescribed medical regimen, it is important to identify variables associated with greater adherence.

B. PROGRESS TO DATE

This progress report covers the period from 9/1/09 – 3/1/10. Since this study was funded, one of the investigators, Jennifer Williams, left UCSD to pursue graduate studies elsewhere. Dr. Angela Ballantyne was thus added as a co-investigator. As we reviewed the proposed battery of tasks we had initially proposed to administer to our study participants, it became apparent that the proposed measures were not as comprehensive as we had initially hoped they would be at probing the relevant psychosocial areas we wanted to explore. After a detailed search of the literature, we have developed a comprehensive questionnaire battery that will more appropriately address the specific aims of this study. This test battery is listed below.

We made another change in the initial protocol. We had proposed to use individuals with chronic renal disease from other causes as a “disease control” group. However, we decided that recruiting individuals with chronic renal disease in the short period of time allotted for this study (12 months) would be very difficult based on numbers of CRD patients at our institution who were within the age range of our study. We thus decided to use adolescents with another chronic illness, juvenile diabetes mellitus, as a “disease control” group. The rationale for selecting this population was that children and adolescents with diabetes must take daily medication in order to survive, require frequent blood tests and doctor visits, have late complications of the disease, and have a lifelong condition, all very similar to the issues individuals with cystinosis face. We have the added benefit of a large clinical population of juvenile diabetics due to a Pediatric Diabetes Research Center and an active clinical diabetes group at our institution. Dr. Michael Gottschalk, Director of the Pediatric Diabetes Clinic, is now a co-investigator on this project. He and his staff are handing out questionnaire packets to parents and adolescents between the ages of 12-18 years with diabetes. Our goal is to obtain completed questionnaires from 30 parents and 30 adolescents with nephropathic cystinosis, 30 each from parents and children with juvenile diabetes, and 30 each from parents and children with no chronic medical condition (healthy controls). In order to accomplish this goal within the 12-month time frame and with limited funding, we have engaged undergraduate students to assist with the study instead of hiring a research assistant. The costs of the questionnaires, mailing, and subject payments will utilize most of the budget allotted for the study. Furthermore, due to the limited funding and duration of this study, participants will no longer receive IQ testing. Instead, we are sending the questionnaire packets to adolescents and their parents and asking them to return them by mail. Every individual who completes the questionnaire packet receives $50 for their participation.

Over the first 6 months of the study we obtained IRB approval for the study, ordered testing supplies, and compiled and mailed packets to be sent to participants.

We have sent out questionnaire packets to33 individuals with cystinosis and their parents, distributed 50 packets to the Diabetes Clinic at UCSD, and sent out 30 packets to control participants and their parents. We have obtained completed questionnaire packets from 10 adolescents and 11 parents of adolescents with cystinosis, and from 5 individuals and 3 parents of an adolescent with diabetes. Hence, we have made substantial progress toward the goals of this study.

C. RESULTS

We are in the process of scoring, reliability checking, and data entering the questionnaire data collected so far. We are contacting families by email or telephone who have received packets but who have not yet returned them to encourage their completion of the questionnaires. We have not yet analyzed any of the data.

D. FUTURE PLANS

In the next 6-month period, we plan to finish collecting the questionnaire data on individuals with cystinosis, diabetes, and control participants. We also plan to have all the data scored and data entered, and data analyses to be completed. We hope to be ready to prepare a manuscript for submission by the end of the year.

Table 1. Protocol for questionnaires to be completed by adolescents with cystinosis, diabetes, or no chronic medical condition, and by their parents.

Adolescent	Parent
Family Environment	**Family Environment**
• Family Environment Scale *20 min*	• Family Environment Scale *20 min*
Quality of Life/Health Status	**Quality of Life/Health Status**
• PedsQL *10 min*	• Quality of Life Inventory *5 min*
Coping and Resiliency	**Coping and Resiliency**
• A-COPES *15 min*	• F-COPES *15 min*
• RSCA (9 – 18) *5 min*	
Stressors and Social Resources	**Stressors and Social Resources**
• LSSRI-Y *30-60 min*	• LSSRI-A *30-60 min*
	• Impact on Family *10 min*
Parent-Adolescent Relationship	**Parent-Adolescent Relationship**
• PARQ *35 min*	• PARQ *35 min*
Independence	**Independence**
	• SIB-R *20 min*
Anxiety/Depression	**Anxiety/Depression**
• Beck Youth Inventories (Depression, Anxiety) *15 min*	• Beck Anxiety Inventory *10 min*
	• Beck Depression Inventory – II *5 min*
Demographics/Adherence	**Demographics/Adherence**
• Adolescent Medication Adherence Questionnaire *10 min*	• Parent Report Child Medication Adherence *10 min*
	• Demographic Information *10 min*
2 ½ hrs.	3 hrs.