NATIONAL INSTITUTE OF NURSING RESEARCH
Spring Science Workgroup

Increasing Nursing Research Opportunities in Cystic Fibrosis
May 1-2, 2001

EXECUTIVE SUMMARY

A meeting of the National Institute of Nursing Research (NINR) Spring Science Workgroup on Increasing Nursing Research Opportunities in Cystic Fibrosis was convened on May 1-2, 2001, in Bethesda Maryland. In accordance with Public Law 92-463, the meeting was open to the public from 5:00 to 7:00 P.M. on May 1 and from 8:00 A.M. to the 3:00 P.M. adjournment on May 2. Dr. Patricia A. Grady and Dr. Hilary D. Sigmon presided as chairs.

WORK GROUP MEMBERS PRESENT

Denise B. Angst, DNSc; Lois A. Brass, RN, BSN; Frank J. Cerny, PhD; Becky Christian, PhD, RN; Stanley M. Finkelstein, PhD; Margaret Grey, PhD, CPNP, FAAN; Dana S. Hardin, MD; Leslie Hoffman, PhD, RN, FAAN; Larry G. Johnson, MD; Kathleen Knafl, PhD; Mary Jo McCracken, RN, MS, CPN; David Orenstein, MD; Michael S. Schechter, MD, MPH; Janet Williams PhD, RN, FAAN

FEDERAL EMPLOYEE WORKGROUP MEMBERS PRESENT

Patricia A. Grady, PhD, RN, FAAN; Hilary D. Sigmon, PhD, RN; Carole Hudgings, Ph.D., RN, FAAN; Robin L. Gruber, MPS; Muriel Battle

OPEN MEETING SUMMARY

I. Call to Order, charge to the workgroup

Dr. Grady called the meeting to order and welcomed the participants. She gave the workgroup its charge, which was to identify ways to increase the number of nurse researchers engaged in nursing research in the area of cystic fibrosis (CF). Dr. Grady noted that the National Institute of Nursing Research (NINR) is especially interested in encouraging and supporting interdisciplinary work; accordingly, this workshop brings together professionals from several disciplines and varied research interests. Dr. Grady and Dr. Sigmon acknowledged the support of the NIH Office of Rare Diseases for the workshop.

II. Discussion of Nursing Research Opportunities in Cystic Fibrosis

Cystic fibrosis is the most common inherited life-shortening disease in white populations, occurring in about 1 in 3300 live white births. The patient registry of the Cystic Fibrosis Foundation lists some 21,000 persons now living with the disease. An abnormal gene causes cell defects related to salt and water transport, leading to

progressive difficulties with breathing and exercise tolerance as well as gastrointestinal and other problems, including a heightened risk for diabetes and short stature. The gene that causes CF was identified in 1989, but while that has made possible carrier and prenatal testing, it has not yet resulted in clinical breakthroughs for people with the disease.

The average life expectancy of persons with CF has increased markedly as a result of biomedical advances—from 14 years in 1969 to more than 30 years in 1995. These advances (which include organ transplantation and more effective approaches to management) have increased not only life expectancy, however, but also the number of complications to be handled by patients, families, and clinicians. Some complications are physical, such as diabetes and the sequellae of lung or liver transplantation; others stem from the challenges of coping with the disease and with the stringent nutritional and exercise regimens it makes necessary. As with any illness, the physical and psychosocial impacts compound each other.

There are biological, psychological and social dimensions to managing and treating CF—a broad spectrum of issues to which nursing research is well suited. The NINR is seeking ways to increase nursing research in this area, aiming both to bring new nurse researchers into ongoing CF research and to encourage those already engaged in nursing research in other areas to widen their focus to include CF. After reviewing what is known about the etiology and impacts of CF, the May 1-2 workgroup identified gaps in the science about which research is needed and a variety of approaches to increasing research and strengthening NINR's CF portfolio. The recommendations are summarized below.

Research Opportunities

In the **psychosocial** area, compliance is a crucial issue because of the challenging nutritional and exercise demands of managing the disease. In addition, several research topics fall in the general area of "living well with uncertainty," relating to the development of strategies to help children build strong coping skills, plan for adulthood as life expectancy increases, and prepare for the end of life. Middle childhood and adolescence are ages of particular interest to researchers because of the important adaptations that need to happen at those developmental stages.

Exercise and nutrition were major and closely related discussion themes in the workshop. Efforts to promote fitness are crucial interventions that can improve and lengthen the lives of persons with cystic fibrosis. Among other things, management of exercise and nutrition can mitigate the effects of diabetes. The workgroup noted, these are areas where serious compliance issues arise, and more research is needed to determine better ways to persuade children to adhere to their regimens. Also, participants noted that no practical tools currently exist to measure dyspnea or exercise tolerance in children.

In the areas of **biology, genetics, and physiology**, the workgroup called attention to research questions related to genetics, the treatment of diabetes, and the prediction of severity and outcomes. Longitudinal and outcome data are needed to identify the most effective regimens for different individuals. In the areas of biology, genetics,

physiology, and the psychosocial area, workgroup members noted the many synergies with the research on children with diabetes.

Genetic testing raises a host of questions relating to ethics, privacy, the potential for discrimination, voluntariness, and population-based screening. Research is also needed to prepare nurses and others for the optimum methods to educate patients, families, and the general public about genetic testing.

The workgroup reported that early research on **SES-related risk factors** for CF reveals that low-income persons with CF are at greater risk for adverse outcomes such as growth stunting and for earlier age of death. Participants stated that research has not found differences in access to care among income groups, suggesting that environmental exposures may be causing the disparities. More research is needed to determine the risks and to identify ways of eliminating the disparities among income groups.

On the **uses of technology,** the workgroup cited that research is needed on further uses of technology to support self-care and home care, and on using communication resources to link children with CF with each other and to connect researchers and potential subjects.

Strategies for Increasing Nursing Research and Research Training

The workgroup then turned to strategies for strengthening the CF portfolio at NINR and supplementing nursing research expertise in this area. The discussion centered on four general approaches: networking nursing research into the CF community; collaborating with outside agencies; engaging more RNs as researchers; and capitalizing on existing NINR programs. All of the suggestions shared the theme of increasing collaboration and developing interdisciplinary teams.

Ideas for **networking into the CF community** centered on using the Cystic Fibrosis Foundation (CFF), including its Registry and Therapeutic Development Network trials, as a source of and conduit to research and funding opportunities. Possible crossovers between CF and other areas such as diabetes, nutrition, and exercise were noted. Other ways of expanding research include secondary data analysis on data collected for other purposes, and the possible use of non-categorical, non-disease approaches.

The agencies and organizations besides the CFF that were mentioned as **possible collaborators** include industry, the National Heart, Lung and Blood Institute, the National Institute of Diabetes and Digestive Diseases, and the Juvenile Diabetes Foundation.

Suggestions for **increasing the numbers of doctorally prepared nurses into CF research** included more active mentoring of registered nurses. The workgroup also suggested that registered nurse clinicians be added to multidisciplinary CF research teams as active collaborators. It was stressed that more support should be given to help non-Ph.D. nurses with an interest in research to transition into the Ph.D. track.

On **capitalizing on NINR programs**, those mentioned as appropriate mechanisms for nursing research on cystic fibrosis were the Institutional National Research Service Award (T32), the Request for Applications (RFA) for clinical trials, the NINR Center mechanism, and traditional research grants (R01s).

Several **research training needs and opportunities** were identified in the workshop discussions. In the clinical research area, these included training nurse researchers and other health professional researchers to test the best ways to: teach children about exercise and nutrition, to provide genetic counseling, to incorporate technology into nursing models for research, and to invest research dollars in areas of home monitoring and telehealth care. In the research training area, there were suggestions to assist doctorally prepared nurses to write effective research proposals in the area of CF, encouraging pre-Ph.D. nurses with the "research bug" to continue their education towards a doctoral degree, and encouraging interdisciplinary and inter-disease collaborations in research training.

The meeting was adjourned at 3:00 p.m. on Wednesday, May 2.