

PHI Grant Awarded to Johns Hopkins Team to Study Early Use of Noninvasive Ventilation

Post-Polio Health International, of which International Ventilator Users Network is an affiliate, announced that it has awarded a \$25,000 research grant to a team from Johns Hopkins University. The researchers propose to determine whether early use of noninvasive positive pressure ventilation (NIPPV) prolongs survival in patients with amyotrophic lateral sclerosis (ALS) and to relate their findings to other neuromuscular diseases including polio and its late effects.

ALS, also known as Lou Gehrig's disease, is a progressive neurodegenerative disease that attacks nerve cells in the brain and spinal cord resulting in muscle weakness, atrophy and, eventually, death. The respiratory muscles are invariably affected, and respiratory failure is the most common cause of death in ALS patients.

NIPPV is becoming an increasingly standard treatment for patients with chronic respiratory failure, and observational studies suggest that NIPPV prolongs survival, but it is not known whether it modifies disease progression or simply provides support in the terminal stages of the disease. Currently, guidelines recommend starting NIPPV when a patient's forced vital capacity – a measure of breath exhalation – falls below 50 percent of the predicted value.

The Johns Hopkins study aims to determine whether NIPPV is more beneficial in prolonging survival if it is started earlier in the course of ALS. The study will also attempt to determine if a point can be identified at which survival is maximized.

"If our study demonstrates prolonged survival in patients who begin using NIPPV earlier than currently recommended, it will have profound implications for how patients with ALS and other neuromuscular diseases are managed," said Principal Investigator Dr. Noah Lechtzin, instructor of medicine in the Division of Pulmonary and Critical Care Medicine at Johns Hopkins University. "It will suggest that a more proactive approach is beneficial. This study could alter this passive approach to patient care."

"Although the patients in the present study have ALS, we believe that the results will apply to a majority of the people we serve with post-polio syndrome, as well as to those with other neuromuscular diseases such as muscular dystrophy, spinal muscular atrophy and multiple sclerosis," said Joan L. Headley, executive director of Post-Polio Health International.

Others on the Johns Hopkins team in addition to Dr. Lechtzin include Nurse Coordinator Lora Clawson, director of clinical research in the ALS Center, and Research Assistant Anne M. Lang, in the Division of Pulmonary and Critical Care Medicine. Dr. Gregory B. Diette, assistant professor of medicine, and Dr. Charles M. Wiener, associate professor of medicine, both also with the Division of Pulmonary and Critical Care Medicine, will act as consultants to the study. ▲