

WSA Increases Research Funding for 2017 - Supports Critical Research efforts into Cardiovascular and Anesthesia Issues.

The WSA has awarded research grants to Boston Children's Hospital and Columbia University Medical Center (NYC) for important research into cardiovascular and anesthesia issues in Williams syndrome.

The Cardiac Anesthesia team at Morgan Stanley Children's Hospital at Columbia is conducting a study aimed at "Improving Clinical Outcome in Children with Williams-Beuren Syndrome." The proposed study will summarize an extensive clinical experience in children with Williams syndrome who had anesthesia care over a ten-year period at Columbia University Medical Center. The results will provide the necessary framework to design a robust clinical database for the anesthesia/sedation care for these children, which will be critically important to inform future practices and improve outcomes.

The study at Boston Children's Hospital involves four separate but integrated projects over a three year period. Project 1 aims to create a simulation-based program to train surgical teams around the successful management of anesthesia-based emergencies for the patient with Williams syndrome.

Project 2 will utilize intravascular ultrasound imaging to understand the arterial wall and its response to catheter intervention. The vascular abnormalities that create obstruction (stenoses) in the pulmonary arteries of patients with Williams syndrome are poorly characterized. An improved understanding of the underlying nature of the vessel wall, and its response to intervention could improve the safety profile of balloon angioplasty for peripheral pulmonary artery stenosis.

Project 3 will examine the current state of healthcare delivery for patients with Williams syndrome and use this information to develop optimal models of care deliveries. The goal of these analyses will be to have an enhanced understanding of the current state of healthcare delivery, and the healthcare needs of people with Williams Syndrome across the lifespan, as well as to identify populations of patients with Williams syndrome with lower access to critical services.

Project 4 will perform a cross-sectional retrospective study to assess the relationship of medical and family factors to health-related quality of life, adaptive functioning, and neurodevelopmental function in children with Williams syndrome followed at Boston Children's Hospital. BCH will work with the WS Registry to allow for inclusion of comparable data, facilitate coordinated future studies and enhance general awareness, specifically focusing on cardiovascular aspects of WS. Following the retrospective study, a prospective analysis and study of DNA samples will be used to determine if genetic modifiers of the varied vascular complications of WS can be identified based on general knowledge of the genetic controls of vascular development that are either predictive of severe vascular disease or protective from severe vascular disease.

WSA has also extended funding for the behavioral study by Dr. Bonnie Klein-Tasman and her team at the University of Wisconsin, and the Longitudinal study by Dr. Carolyn Mervis and her team at the University of Louisville.

Future website blog posts from each research team will provide more detail on each study.