



Advancing Research and Clinical Care For Children with Pulmonary Vein Stenosis

The Evolving PVS Network

The PVS Network is a rapidly evolving multi-institutional collaborative community that is dedicated to advancing research and clinical care for children with pulmonary vein stenosis (PVS).

In late 2015, the PVS Network was informally assembled by committed clinicians dealing with the frustrating outcomes of PVS. Participation in the Network is built around a commitment to advancing care of children with PVS through sharing of clinical data. Creation of a shared prospective and retrospective database using a centralized data collection and analysis model will facilitate the analysis of important research questions and provide the necessary infrastructure required to continue advancing care for this rare disease.

***Through collaboration, we hope to build a strong
committed community focused on improving
outcomes for children with PVS***

The mission of the PVS Network is to advance research and clinical care for children with PVS.

To achieve this mission, we have the following aims:

Aim 1: Create a common clinical surveillance protocol to harmonize follow-up for children with PVS and facilitate data sharing.

Aim 2: Create a collaborative multi-institutional prospective and retrospective database to define progression variables, risk factors and outcomes in children with PVS.

Aim 3: Create a collaborative infrastructure for furthering PVS clinical studies and interventional clinical trials.

Aim 4: Create a central resource for knowledge translation for both the medical community and parents facing a new diagnosis of PVS.

Participating Institutions

The Hospital for Sick Children	Christopher Caldarone & Rachel Vanderlaan
Children's Hospital of Philadelphia	William Gaynor & Jonathan Rome
Cincinnati Children's Hospital	Russel Hirsch & Andrew Redington
C.S Mott Children's Hospital	Adam Dorfman & Richard Ohye
Children's Clinics and Centers of Minnesota	David Overman
Stollery Children's Hospital	Ian Adatia & Ivan Rebeyka
Morgan Stanley Children's Hospital	David M.Kalfa & Emile Bacha
University of Iowa Children's Hospital	Peter J. Gruber
Emory Children's Hospital	Bahaaldin Alsoufi
Texas Children's Hospital	Henri Justino

Other institutions expressing interest include: Boston Children's and Utah's Primary Children's Hospital

Thanks to everyone for their commitment!

Update on PPHNet collaboration

Steve Abman invited RV and CC to make a presentation describing the goals, structure, and objectives of the PVS Network with the membership of the PPHNet (<http://pphnet.wpengine.com/>).

Although the two groups have different areas of focus, there is considerable overlap and potential for collaboration. Whereas the PPHNet has focused on all modes of pulmonary hypertension, the PVS Network is focusing on pulmonary hypertension limited to the subset of patients in whom anatomic narrowing of the pulmonary veins is the primary initiator of pathophysiology. Despite this difference in focus, the two groups share a common objective in developing better understanding and treatment for patients with pulmonary hypertension. The call was extremely successful and the members of the PPHNet voted to fully support the development of the PVS Network and work in a collaborative manner.

Clinical Surveillance: Early Detection, Early Intervention

At the Hospital for Sick Children, we developed and implemented a simple 3-dimensional (3D) imaging-based Clinical Surveillance Protocol (CSP) aimed at harmonizing individual clinical practices and improving detection of PVS progression. **Diagnosis of PVS is confirmed with 3D imaging and follow-up 3D imaging occurs at 1, 6 and 12 months following any major intervention, in addition to echocardiography studies.**

Introduction and adherence of the CSP was associated with early identification of PVS progression in 3 out of 8 patients and decreased time to first 3D imaging study (131 vs 54 days, $p < 0.05$) suggesting that the CSP improved detection of disease progression.

The rationale for developing a CSP is based on the hypothesis that **early detection of disease progression may lead to windows of opportunity for intervention**. Based on clinical and lab studies, early detection is likely to be associated with better upstream calibre of veins, a decrease in the number of veins involved, and greater likelihood of reversibility in the pulmonary vascular lesions - all of which may improve outcomes.

Adoption or formalization of a CSP by participating institutions, in addition to their standard of care, leads to harmonization of elements of follow-up and facilitates data sharing and creation of an infrastructure to evaluate interventions (medical, interventional and surgical) aimed at slowing the progression of this disease.

Ongoing evaluation of institutional CSPs and refinement will be the foundation for consolidating current clinical practices and inform discussion around generating consensus guidelines.

Meetings and Presentations

CHSS Annual Meeting October 23-24, 2016

Oral presentation: Rationale for development of a pulmonary vein stenosis (PVS) clinical surveillance protocol and establishing a PVS collaborative network

AHA Scientific Sessions November 12-17, 2016

Oral presentation: Session Monday Nov 14, 10:00-11:45am
“Proliferation and Endothelial to Mesenchymal Transition Contribute to Progressive Lesions in a Surgical Model of Pulmonary Vein Stenosis”

AHA Scientific Sessions November 12-17, 2016

There will be a special session devoted to PVS
Monday, November 14, 3:45- 5:00 pm. Topics will include:

- a. Pulmonary Venous Development and Disease Mechanisms (**K Degenhardt**)
- b. Detecting Pulmonary Venous Anomalies and Stenosis: Role of Echo, MRI, Catheterization (**SJ Yoo**)
- c. Pulmonary Venous Anomalies with Other CHD: Impact upon Care and Outcome in the Fetus and Newborn (**V Yarlagadda**)
- d. Surgical and Interventional Management for Pulmonary Vein Stenosis (**C Caldarone**)
- e. Novel Medical Therapies for Pulmonary Vein Stenosis (**K Jenkins**)

PVS Network AHA Dinner

At the AHA, we have a great opportunity to meet as a group. After the PVS session on November 14th, we will have a dinner for all members of the PVS Network. Additionally, we have extended an invitation to members of the PPHNet.

The dinner will include a presentation and discussion on the strategic directions for the PVS Network.

Time: 6:30pm, Location TBD. Official email invitation to follow shortly.
RSVP leslie.jones@sickkids.ca

Save the Date!

10th International Conference Neonatal & Childhood Pulmonary Vascular Disease
March 9-11, 2017
San Francisco

Special PVS Symposium and Consensus Guideline Working Group Session

<http://www.ucsfcmc.com/2017/MPD17002/info.html>

Research Protocols Available

Pulmonary vein stenosis: disease mechanisms and outcomes

Prospective and retrospective clinical PVS database
Option for participation in genetics/whole genome sequencing project

Disease progression and outcomes in pulmonary vein stenosis

Retrospective clinical data collection only

Contact Info and Requests for Protocols

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