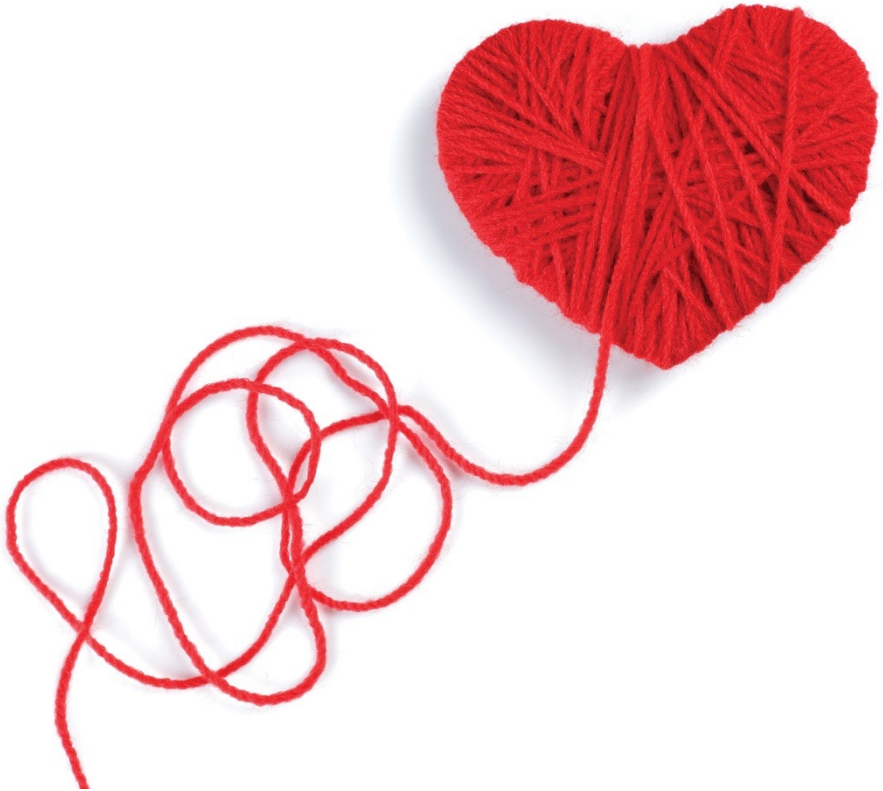
CARDIOLOGY 2023

COLLABORATION BETWEEN TWO ADVANCED PRACTICE NURSE TEAMS CARING FOR THE PULMONARY VEIN STENOSIS PATIENTS WAITING FOR LUNG TRANSPLANT



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PULMONARTY VEIN STENOSIS (PVS)

- Rare and serious condition caused by an abnormal thickening of the pulmonary vein (PV) walls; most often affects more than one pulmonary vein
- Lung transplant is introduced as an option upon initial diagnosis with PVS, but medical and surgical strategies are first sought out due to our expertise in managing these patients.
- Patients with aggressive disease poorly responsive to medical or surgical therapies and/or disproportionate pulmonary hypertension are encouraged to meet with lung transplant coordinator to discuss transplant and evaluation.

LUNG TRANSPLANT EVALUATION

- The evaluation process involves meetings with multidisciplinary transplant team members, diagnostic testing, and subspecialty involvement as needed to help determine potential barriers to lung transplant candidacy
  - *Multidisciplinary transplant team members:* pulmonologist, lung transplant surgeon, social worker, dietician, pharmacist, and physical therapist
  - *Diagnostic testing:* chest CTA, gastric emptying, swallow study, DEXA
  - *Subspecialty involvement:* GI, feeding team, infectious disease
- After evaluation completion, a selection committee meeting of multidisciplinary team members review the evaluation and determine if patient is appropriate for lung transplant listing.
- Some patients are not eligible for transplant due to both medical and psychosocial contraindications.

AWAITING TRANSPLANT

- Average wait time varies by age and blood type
- Many younger patients deteriorate medically prior to lung transplant with frequent hospitalizations, and may die waiting for transplant
- Monitoring and care while waiting includes:
  - Monthly sedated echocardiograms, lung perfusion scans
  - Bimonthly Avastin infusions, when indicated
  - Frequent cardiac catheterizations (every 4-6 weeks)
  - Monitoring of Panel Reactive Antibodies for sensitization

STATISTICS

	Alive	Deceased
s/p lung transplant	3	3
s/p 2nd lung transplant	1	1
s/p heart / lung transplant	0	1
Listed for lung transplant	2	6
Delisted related to clinical improvement	3	-
Transplant evaluation in process	1	-

Table 1. PVS patients listed for lung transplantation (n=18) from 2009 to 2022

BENEFITS OF APRN COLLABORATION

- Frequent communication between cardiology and pulmonary teams.
- Attendance at each others' weekly team meetings when appropriate
- Ability to provide continuity of care and goal planning for patient and family
- Lab management
- Consultation with other specialists, including Pediatric Advanced Care Team
- Nutritional monitoring and management as an outpatient
- Communication with local providers, visiting nurse agencies, and schools
- Seamless transfers between local acute medical rehabilitation hospital
- Emotional support for the patient and family as they await transplant.
- Expanding knowledge base of each subspecialty

CASE STUDY

12 year old (ex-25 weeker) with history of chromosomal duplication of 16p11.2, chronic lung disease and PVS with initially left-sided involvement diagnosed at 5 months of age.

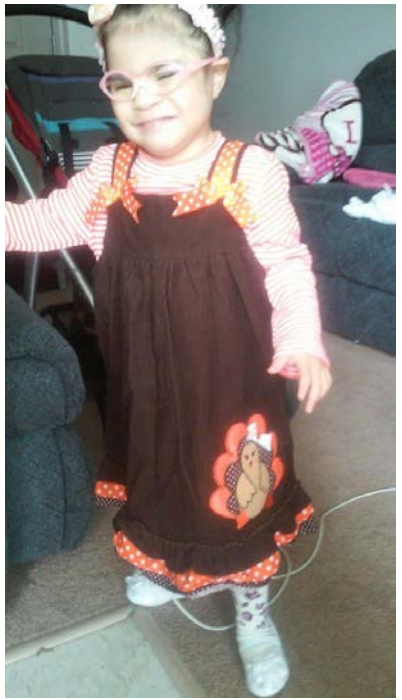
Clinical course:

July 2012 (14 months of age)

- Bilateral PVS on echocardiogram. Referred to Boston Children's Hospital for further medical and surgical intervention
- Sutureless surgical repair
- Enrolled in the Gleevec and Avastin protocol
- Listed for lung transplant, went to Franciscan's Hospital while awaiting transplant.

February 2013 (21 months of age)

- Bilateral lung transplant
- Did well post-transplant with close monitoring of graft function and immunosuppression



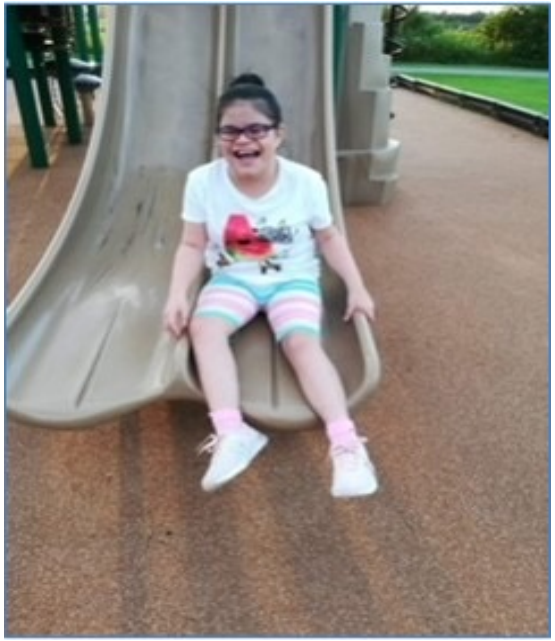
CASE STUDY CONTINUE

April 2021 (10 years old)

- Presented with decreased oxygen saturations, from post-transplant baseline of 99-100% awake/ 98% asleep down to 94-98% awake, 98% asleep and with activity (dance) as low as 86-90%.
- Full work up including bronchoscopy with biopsy, chest CTA, 6-minute walk, and lab studies for thyroid function and iron studies completed, all normal.

January 2022 (11 years old)

- Cardiology reengaged, echocardiogram and Holter obtained which were normal. Supported pulmonary rationale that this may be a new baseline. Encouraged monitoring weight and structured physical activity program.



THANK YOU!

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