

BUILDING A LYMPHATICS PROGRAM: THE TEAM APPROACH

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Lymphatic Disorders

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NO DISCLOSURES

STARTING POINT FOR LYMPHATIC DISEASES

- Bad Diseases with limited/no options
 - PLE
 - Plastic bronchitis
 - Chylothorax
 - Chylopericardium
 - Chyloptysis

STARTING POINT FOR PROGRAM





SYSTEMS TO PUT IN PLACE FOR LYMPHATIC IMAGING/INTERVENTION PATIENTS

- Interventionalist/Surgical team
- Imaging/Interventional Venue
- Clinical service
- Clinical pathways and protocols
- Referral/Intake
- Clinic and follow up
- Oversight/Education



PERSONNEL TO DO IMAGING/INTERVENTIONS/SURGERIES

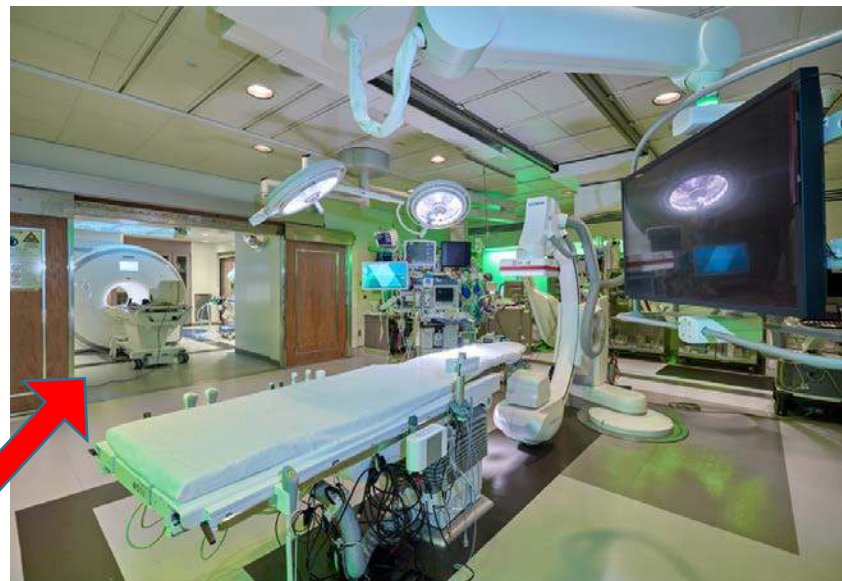
Where we started...



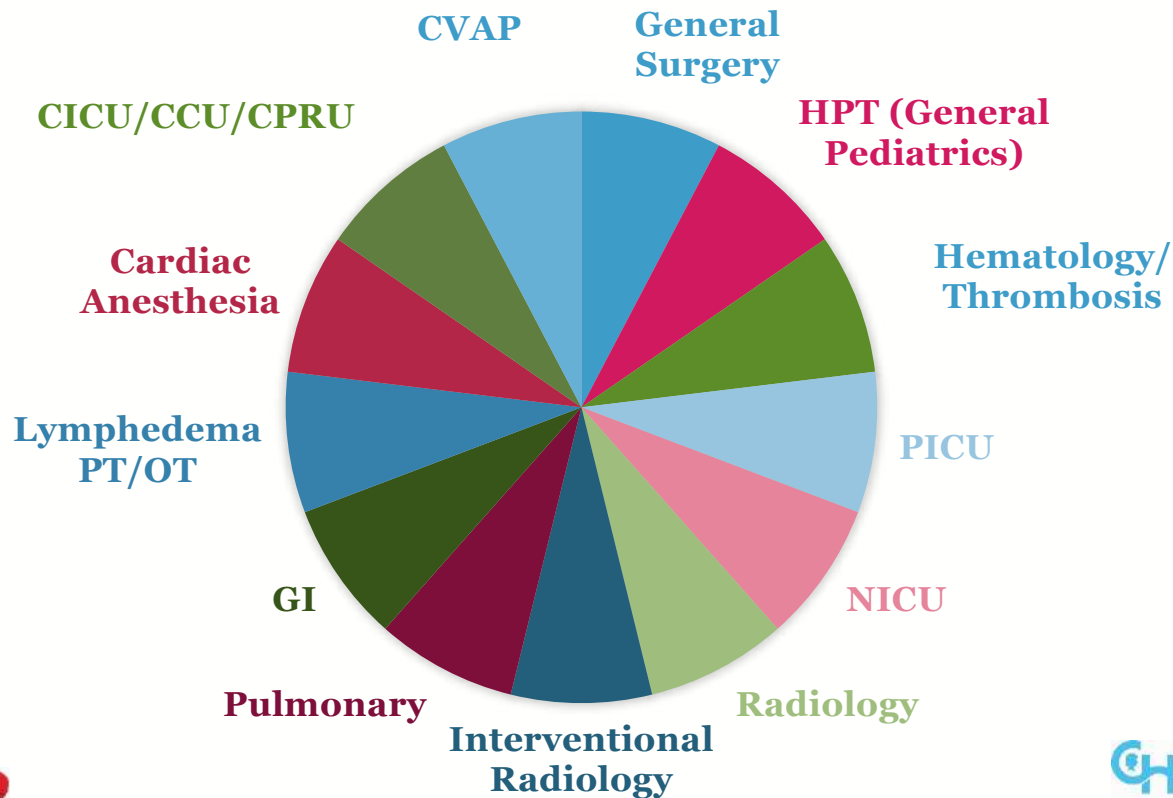
Where we are now..



VENUE FOR IMAGING/INTERVENTION: XMR SUITE



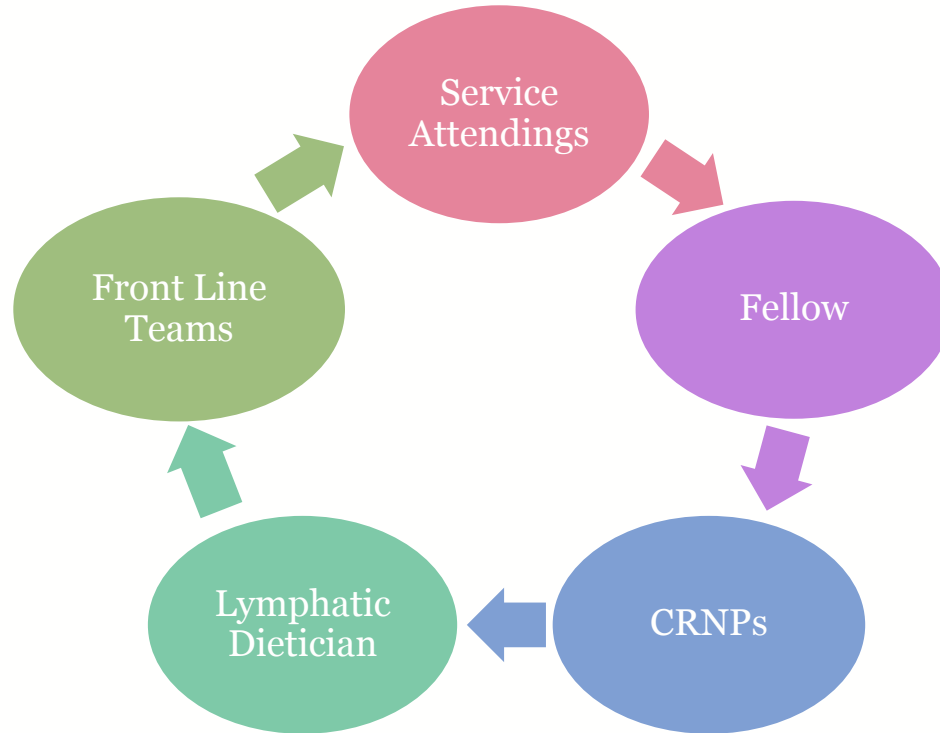
MANY PIECES OF THE PIE



TEAMS THAT PARTICIPATE IN THE CARE OF A LYMPHATICS PATIENT

- Cardiology
- General Pediatrics (HPT service)
- Cardiac Anesthesia
- CICU/CCU (Cardiac step down unit)
- CPRU
- PICU
- NICU
- GI
- Nutrition/Dieticians
- Neurology
- CVAP
- Radiology
- Interventional Radiology
- Cardiology Interventionalists
- Cardiac catheterization lab staff
- Cardiac MRI staff
- Pulmonary
- Hematology/Thrombosis
- General Surgery/CT Surgery
- Lymphedema PT/OT
- Wound Care/Ostomy team
- Intake Center/Preadmission testing

LYMPHATIC INPATIENT CONSULTATION SERVICE



LYMPHATIC SERVICE VOLUME

- For FY23 the Lymphatic service volume has been consistently >10 patients in house
- Average length of stay for a non cardiac patient is 37 days (range 4-81)
- Average LOS for PLE (cardiac) is 11 days
- Average LOS for PB (cardiac) is 6 days – 80% of those spent only one night in the ICU
- At least one non-cardiac, one neonate, and one cardiac lymphatic patient in house since the inception of the program in 2015

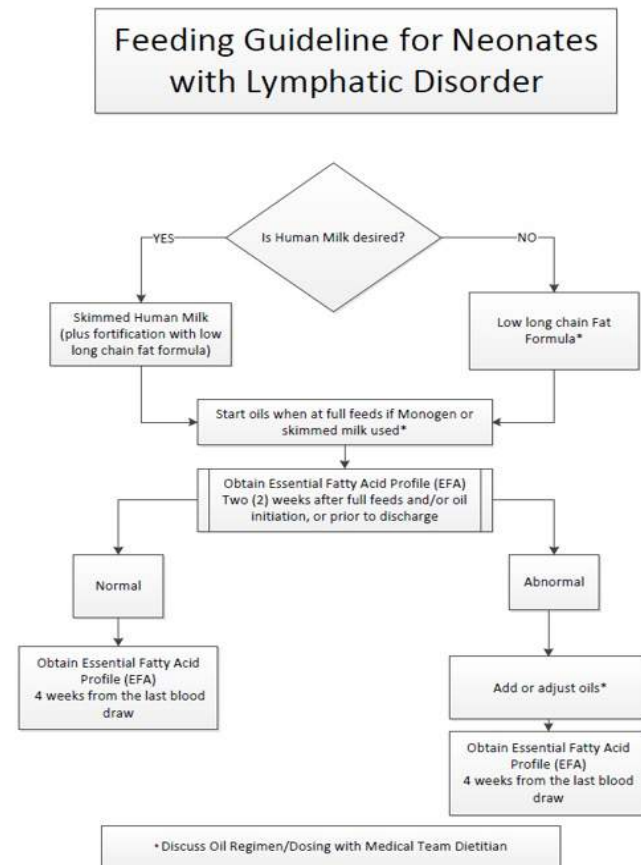
PATHWAYS AND STANDARDIZATION OF CARE



<https://www.chop.edu/clinical-pathway/management-infants-lymphatic-disorders-clinical-pathway>



GUIDELINES AND PROTOCOLS SPECIFIC TO THE LYMPHATIC PATIENT POPULATION: FEEDING GUIDELINE FOR NEONATES WITH LYMPHATIC DISORDER



ORDERS/ ORDER SETS

Order and Order Set Search

MR LYMPH

Order Sets & Panels (No results found)

During visit Medications (No results found)

During visit Procedures

Name	Type	Pref List	Px Code
MR Lymphangiogram Chest,Abd,Pel W&WO Cont	Imaging	CHOP IP FACILITY IMAGL...	74183.009
MR Lymphangiogram Abd W & W/O Contrast	Imaging	CHOP IP FACILITY IMAGL...	74183.008
MR Lymphangiogram Abdomen W Contrast	Imaging	CHOP IP FACILITY IMAGL...	74182.003
MR Lymphangiogram Abdomen WO Contrast	Imaging	CHOP IP FACILITY IMAGL...	74181.006
MR Lymphangiogram Chest W & W/O Contrast	Imaging	CHOP IP FACILITY IMAGL...	71552.003
MR Lymphangiogram Chest W Contrast	Imaging	CHOP IP FACILITY IMAGL...	71551.003
MR Lymphangiogram Chest WO Contrast	Imaging	CHOP IP FACILITY IMAGL...	71550.005
MR Lymphangiogram LT Lower Ext W Cont	Imaging	CHOP IP FACILITY IMAGL...	73719.009
MR Lymphangiogram LT Lower Ext WO Cont	Imaging	CHOP IP FACILITY IMAGL...	73718.013
MR Lymphangiogram LT LowerExt W&W/O Cont	Imaging	CHOP IP FACILITY IMAGL...	73720.015
MR Lymphangiogram LT Upper Ext W Cont	Imaging	CHOP IP FACILITY IMAGL...	73219.009
MR Lymphangiogram LT Upper Ext W&W/O Cont	Imaging	CHOP IP FACILITY IMAGL...	73220.012
MR Lymphangiogram LT Upper Ext WO Cont	Imaging	CHOP IP FACILITY IMAGL...	73218.013
MR Lymphangiogram Pelvis W Contrast	Imaging	CHOP IP FACILITY IMAGL...	72196.007
MR Lymphangiogram Pelvis W&W/O Contrast	Imaging	CHOP IP FACILITY IMAGL...	72197.011
MR Lymphangiogram Pelvis WO Contrast	Imaging	CHOP IP FACILITY IMAGL...	72195.008
MR Lymphangiogram RT Lower Ext W Cont	Imaging	CHOP IP FACILITY IMAGL...	73719.010
MR Lymphangiogram RT Lower Ext WO Cont	Imaging	CHOP IP FACILITY IMAGL...	73718.014
MR Lymphangiogram RT LowerExt W&W/O Cont	Imaging	CHOP IP FACILITY IMAGL...	73720.014
MR Lymphangiogram RT Upper Ext W Cont	Imaging	CHOP IP FACILITY IMAGL...	73219.008
MR Lymphangiogram RT Upper Ext W&W/O Cont	Imaging	CHOP IP FACILITY IMAGL...	73220.009

After visit Orders

Select And Stay Accept Cancel

Order Sets

Order Sets

Clear All Orders

Lymphatic Intervention Admission (Age > 28 days)

Lymphatic Admission Orders

ADMIT TO:

CHOP ADMIT TO INPATIENT

Details

Pathways/Populations

Inpatient Pathways and Population - Lymphatic

Choose a pathway or appropriate patient population

Medical Infant/Child

Length Head circumference

WEEKLY First occurrence Today at 1456 Until Specified

Measure: Length Head circumference

Height Head circumference

MONTHLY First occurrence Today at 1456 Until Specified

Measure: Height Head circumference

RUE & lower extremity BP's (if patient has history of Coarctation of the Aorta or single Ventricle)

XR PORT CHEST AP OR PA

Medical Adult

Vital Signs/ Nursing

This section and below are applicable to all patients.

Continuous Monitoring

Continuous Monitoring

Schedule: Monitor at all times

P Indications: Hospitalized in an ICU, PCU, or CCU

Vital sign parameter(s): Pulse Oximetry (SPO2)

Lymphatics General Nursing Care

Activity Bedrest

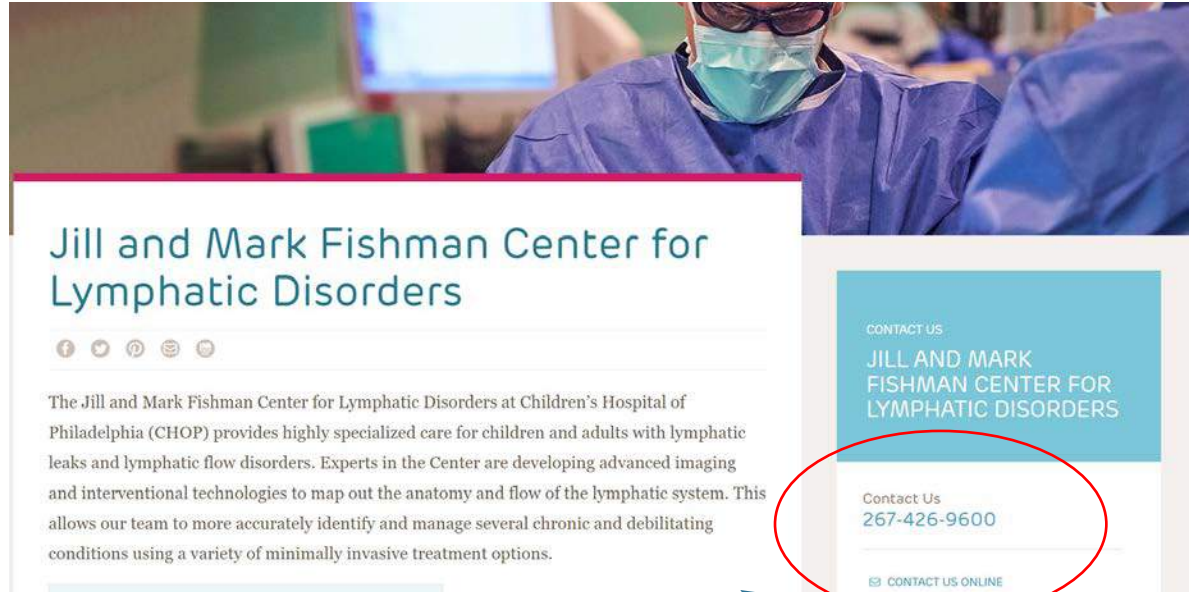
Activity: Bedrest

Bathroom privileges: No restrictions

Laps in hallway: As tolerated

REFERRAL/INTAKE

- Publications
- Social Media
- Website
- Google search
- Physician to physician



REFERRAL

- Nurse Navigator Staff and Lymphatic Nurse Coordinators:
 - Contact referring providers/families
 - Collect data/images
 - Summarize patient information
 - Point person(s) for referring providers/families



Vanessa Bustard,
BSN, RN, CPN



Diane Garofalo



Melissa Lyons, BSN,
RN

Lymphatic Intake Team

STREAMLINED REVIEW OF DATA FOR REFERRAL PATIENTS

Nurse
Navigators
and
Lymphatic
Nurse
Coordinators



Presentation of
case at weekly
Multidisciplinary
Conference



Plan of care
is made



Letter sent out
to referring
team; +/-
conference call

OUTPATIENT CLINIC

- In FY23 we saw 117 patients in Lymphatic outpatient clinic
- Lymphatic Dietician with 0.5 FTE dedicated to clinic appointments, outpatient needs and inpatient rounding
- Participate in Comprehensive Vascular Anomalies Multi-Disciplinary Clinic

EDUCATION



LE&RN Online Symposium:

New Diagnostic Procedures and Interventions for Patients with Systemic Lymphatic Disease

Presented by Christopher Smith, MD, PhD



Lymphatic Education
& Research Network

MR Lymphangiography and Beyond



EDUCATION: GRAND ROUNDS/CONFERENCES

- Grand Rounds
- Invited to speak at 25+ conferences both nationally and internationally.
- Some examples of the meetings are:
 - PICS
 - World Congress of Pediatric Cardiology and Cardiac Surgery
 - Chicago Lymphatic Forum
 - Brazilian World Congress of Cardiology and World Congress of Cardiology
 - CHOP Cardiology Conference
 - LE&RN Symposium



IN-HOUSE EDUCATION

- CHOP Pediatric Grand Rounds
- CHOP Surgical Grand Rounds
- CHOP Physician and Nursing Conferences
- Fellow lectures
- Resident lectures
- APP/RN orientation
- NICU Educational Rounds
- Lunch and Learn Unit based Education Sessions

Third Annual CHOP Lymphatic Disorder Conference

June 2, 2023 | 8 a.m. – 5:30 p.m.

Children's Hospital of Philadelphia
HUB for Clinical Collaboration

Learn more: chop.cloud-cme.com

For more information about
the Lymphatic program:
<https://chop.edu/lymphatic>


Children's Hospital
of Philadelphia
Jill & Mark Fishman
Center for Lymphatic Disorders



NURSING STANDARDS/PATIENT FAMILY EDUCATION

- Nursing Standard: Care of the Lymphatic Embolization Patient
- Lymphatic Embolization: Patient Family Education Manual
- Lymphatic Disorders Feeding Guidelines: Neo-Surgical Clinical Care
- Discharge Instructions Following Lymphatic Intervention for Plastic bronchitis
- Discharge Instructions Following Lymphatic Intervention for Protein Losing Enteropathy with a Cardiac History (Child)
- Patient Instructions: Discharge Instructions Following Lymphatic Intervention for Chylothorax (Child)

FOLLOW UP

- Outpatient Lymphatic Clinic
- Lymphatic Dietician Outpatient Clinic
- Automated emails sent out at dedicated timelines
- Lymphatic Nurse Coordinators
- Telemedicine visits

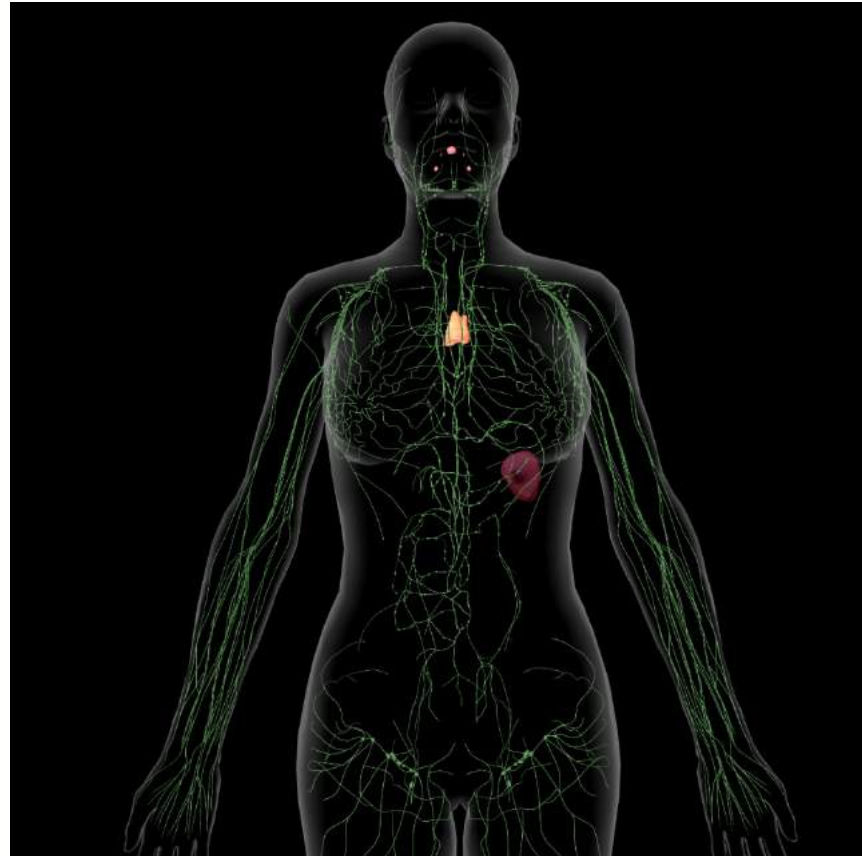


Please use this QR code to fill out our lymphatic follow up form after discharge.
Complete at: 2 weeks, 2 months, 6 months, 12 months.

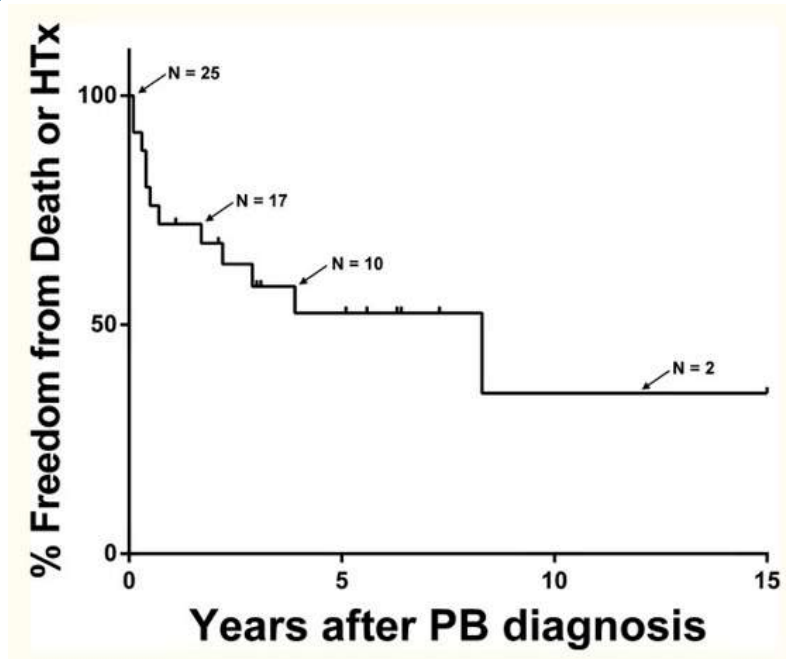
POSSIBILITIES TO DEFINE THE FIELD OF LYMPHATICS ARE ENDLESS



BRINGING TO LIGHT A CIRCULATORY SYSTEM THAT WAS PREVIOUSLY IN THE DARK..



PLASTIC BRONCHITIS SURVIVAL CURVE (PUBLISHED 2014)

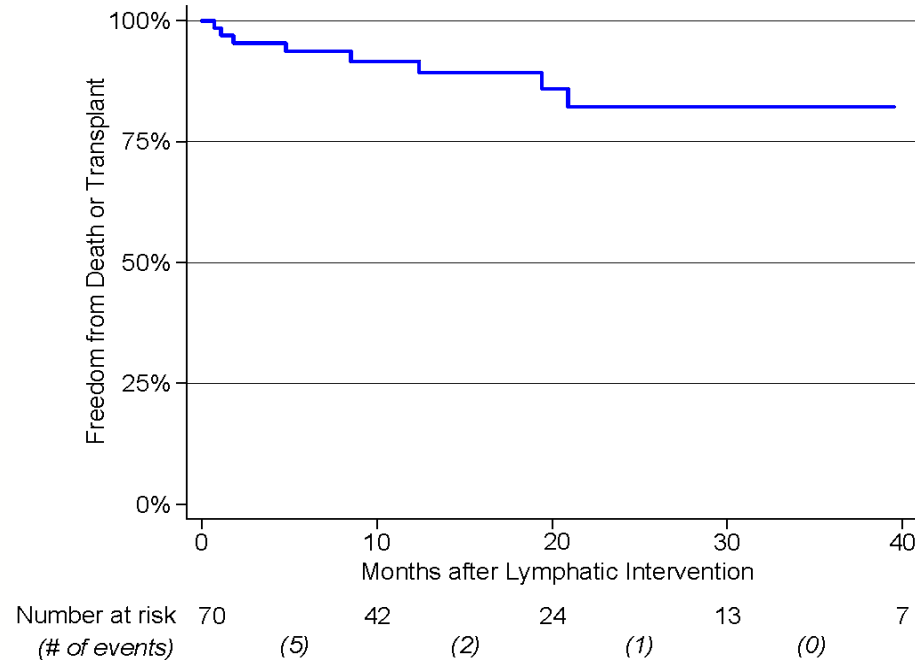


J Am Heart Assoc. 2014 Apr; 3(2): e000865. **Risk Factors and Outcome of Fontan-Associated Plastic Bronchitis: A Case-Control Study**

[Kurt R. Schumacher, MD, MS, Tajinder P. Singh, MD, MS, Joseph Kuebler, MD, Kayla Aprile, BS, Molly O'Brien, MPH, and Elizabeth D. Blume, MD](#)



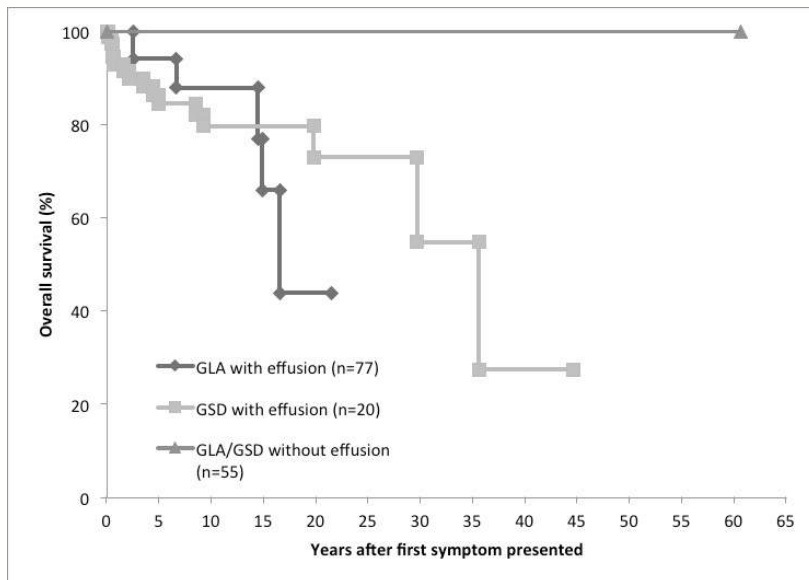
OPPORTUNITIES TO CHANGE THOSE CURVES



Median follow up 19 months (IQR 8 - 30)
→ 8/70 (11%) with death or transplant

IN FACT MOST OF THESE LYMPHATIC DISEASES HAD SIMILAR SURVIVAL CURVES...

**Gorham Stout
and
Generalized
Lymphatic
Anomaly**



	n	5 year overall survival \pm SE (%)
GSD patients complicated by effusion	20	94 \pm 6
GLA patients complicated by effusion	77	84 \pm 5
GSD/GLA patients without effusion	55	100 (N/A)

THERE ARE NOW OPTIONS FOR THE OPTIONLESS...

- Previously life-threatening diseases now have options:
 - PLE
 - Plastic bronchitis
 - Chylothorax
 - Chylopericardium
 - Chyloptysis
 - Multicompartment lymphatic failure



CHOP Researchers Use Precision Medicine to Reverse Severe Lymphatic Disorder in a Patient with KLA

Using genetic sequencing and targeted treatment, CHOP researchers resolved a

LETTERS

<https://doi.org/10.1038/s41591-019-0479-2>

nature
medicine

ARAF recurrent mutation causes central conducting lymphatic anomaly treatable with a MEK inhibitor

Dong Li¹, Michael E. March¹, Alvaro Gutierrez-Uzquiza^{1,2}, Charly Kao¹, Christoph Seiler², Erin Pinto³, Leticia S. Matsuo⁴, Mark R. Battig¹, Elizabeth J. Bhoj¹, Tara L. Wenger⁴, Lifeng Tian¹, Nora Robinson¹, Tiancheng Wang¹, Yichuan Liu¹, Brant M. Weinstein⁵, Matthew Swift⁴, Hyun Min Jung⁶, Courtney N. Kaminski¹, Rosetta Chiavacci¹, Jonathan A. Perkins¹, Michael A. Levine^{6,9}, Patrick M. A. Sleiman^{1,9}, Patricia J. Hicks⁹, Janet T. Strausbaugh⁸, Jean B. Belasco^{1,10}, Yoav Dori⁹ and Hakon Hakonarson^{1,5,11*}

From Portland to Philly: Bubba's Journey with Chylopericardium



Lymphatic Microsurgery: Daniel's Story

Reversing Severe Lymphatic Disorder Caused by Noonan Syndrome:
Maria's Story



Facing a future of recurring internal bleeding, Maria tried an experimental drug. It completely remodeled her lymphatic channels and stopped the bleeding.

THE NEED IS THERE AND THE TIME IS NOW...

129 total procedures done in FY23

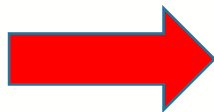
- PLE interventions
- Chylothorax interventions
- Plastic bronchitis interventions
- Ascites (chylous and non chylous) interventions
- Chylopericardium interventions
- CLFD (anasarca) procedures
- Lymphedema procedures
- Lymphovenous Anastomoses
- Hybrid pleurodesis with embolizations
- Thoracic duct externalization

GLOBAL COLLABORATION

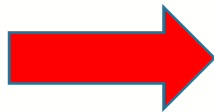


ULTIMATELY... IT'S ABOUT THE PATIENTS

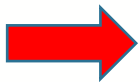
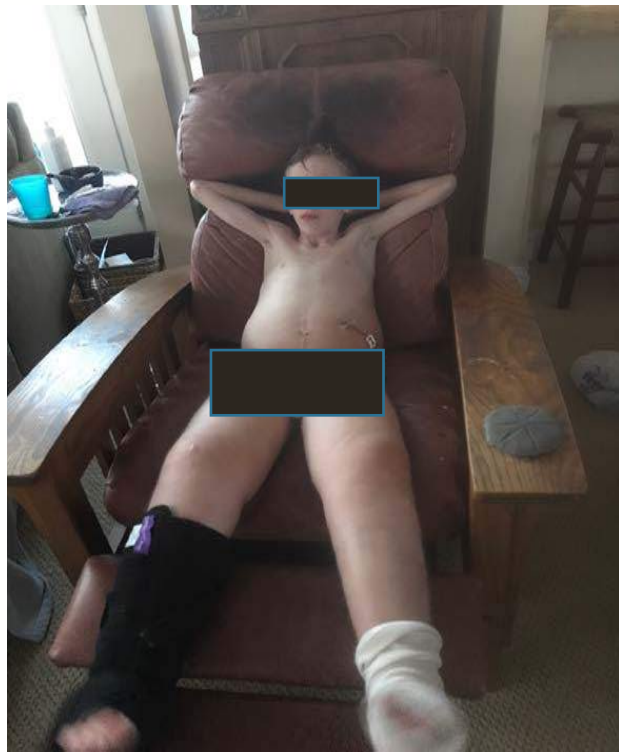
MULTICOMPARTMENT LYMPHATIC FAILURE



CONGENITAL LYMPHATIC FLOW DISORDERS



LYMPHATIC CONDUCTION DISORDERS



PLASTIC BRONCHITIS



MULTICOMPARTMENT LYMPHATIC FAILURE (PLE, ASCITES, PLEURAL EFFUSIONS)

“Dirk Dares to Dream Again”



IN SUMMARY

- Change and Innovation is crucial for the advancement of medicine
- Patients with lymphatic dysfunction require multidisciplinary care
- Collaboration as a team is vital in a multidisciplinary program
- Setting up strong support systems are crucial to the success of any new program
- Systems put in place to improve patient outcomes and decrease patient morbidity and mortality
- We have taken the first steps in unraveling this new frontier but much more needs to be done

THANK YOU

- Dr. Yoav Dori
- Dr. Christopher Smith
- Dr. Aaron DeWitt
- Dr. David Goldberg
- Dr. David Biko
- Dr. Chitra Ravishankar
- Dr. Dalal Taha
- Dr. Jonathan Rome
- Dr. Pablo Laje
- Dr. Katsuhide Maeda
- Dr. Jefferson Brownell
- Dr. Ganesh Krishnamurthy
- Dr. Finn Srinivasan
- Dr. Fernando Escobar
- Lauren Biroc CRNP
- Melissa Lyons RN
- Vanessa Bustard RN
- Diane Garofalo



<https://www.chop.edu/centers-programs/jill-and-mark-fishman-center-lymphatic-disorders>