

# STS/EACTS Latin America Cardiovascular Surgery Conference

November 15-17, 2018

Hilton Cartagena | Cartagena, Colombia



## Surgical Management of Lymphatic Complications After Univentricular Heart Repair

Christian Kreutzer MD

Head, Pediatric and congenital Heart Surgery

Hospital Universitario Austral.

Buenos Aires. Argentina

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The Society  
of Thoracic  
Surgeons



EACTS  
European Association for Cardio-Thoracic Surgery

**No disclosures**



# 50 yrs of Evolution



- Optimized Fontan candidacy
  - Normal pulmonary anatomy and Low PVR.
  - No pulmonary venous obstruction
  - Preserved ventricular function.
  - Preserved AVV function
  - Staged palliation.
- Optimized Fontan surgical Technique: Extr. Conduit
  - Refinement of CPB management and End organ protection at Fontan
  - Technical perfection of Fontan Pathway pathway
  - Prevention of atrial arrhythmia (no-touch Atrium)
  - Preservation phrenic nerve

# Fontan Kreutzer in 2018



- Extra cardiac conduit Fontan (ECC) 20 year follow up
- Survival of 92 %. (Aus NZ Fontan Registry)
- Not a “failed strategy”
- Excellent Survival (better than many Bivent Repairs)
- QOL is reasonable
- Dark clouds on the horizon
  - Fontan failure w preserved V. Fx.
  - Fontan failure w S Ventricle Failure

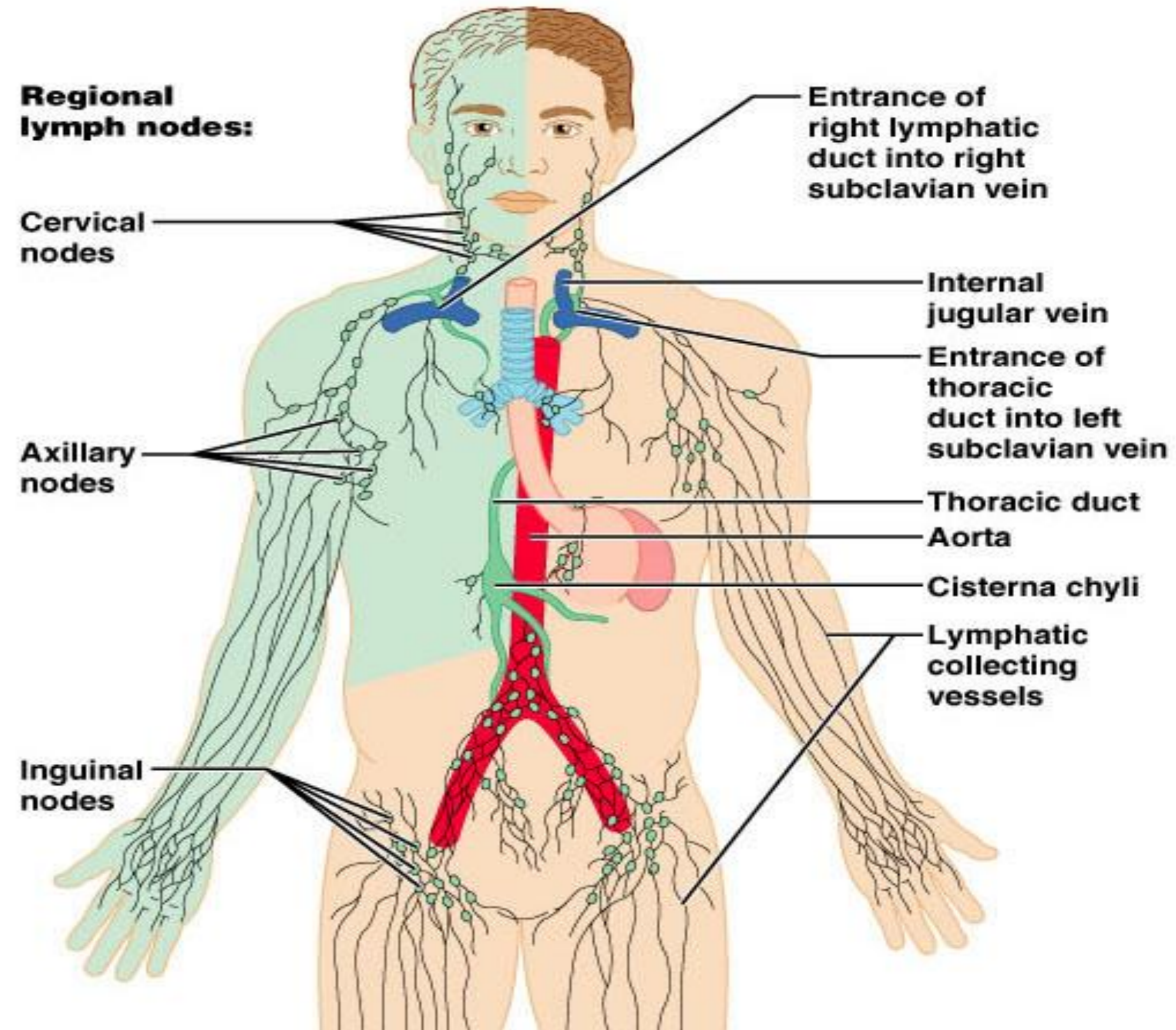
# Fontan Kreutzer in 2018



- What is a Fontan failure? Let's be honest.
- A suboptimal management is ***our*** failure not Fontan's
  - Most SV CHD have normal pulmonary arteries at birth.
  - Sub aortic stenosis can and should be avoided since birth.
  - Chronic volume overload (long standing loose bands, large BT shunts)
  - Phrenic nerve palsy is always iatrogenic.
  - Technical perfection of the Fontan Pathway can and should be always present.
  - Optimal CPB management. TCA, Organ protection @ Fontan Kreutzer
  - Lymphatic failure can be identified pre Fontan and may be prevented.

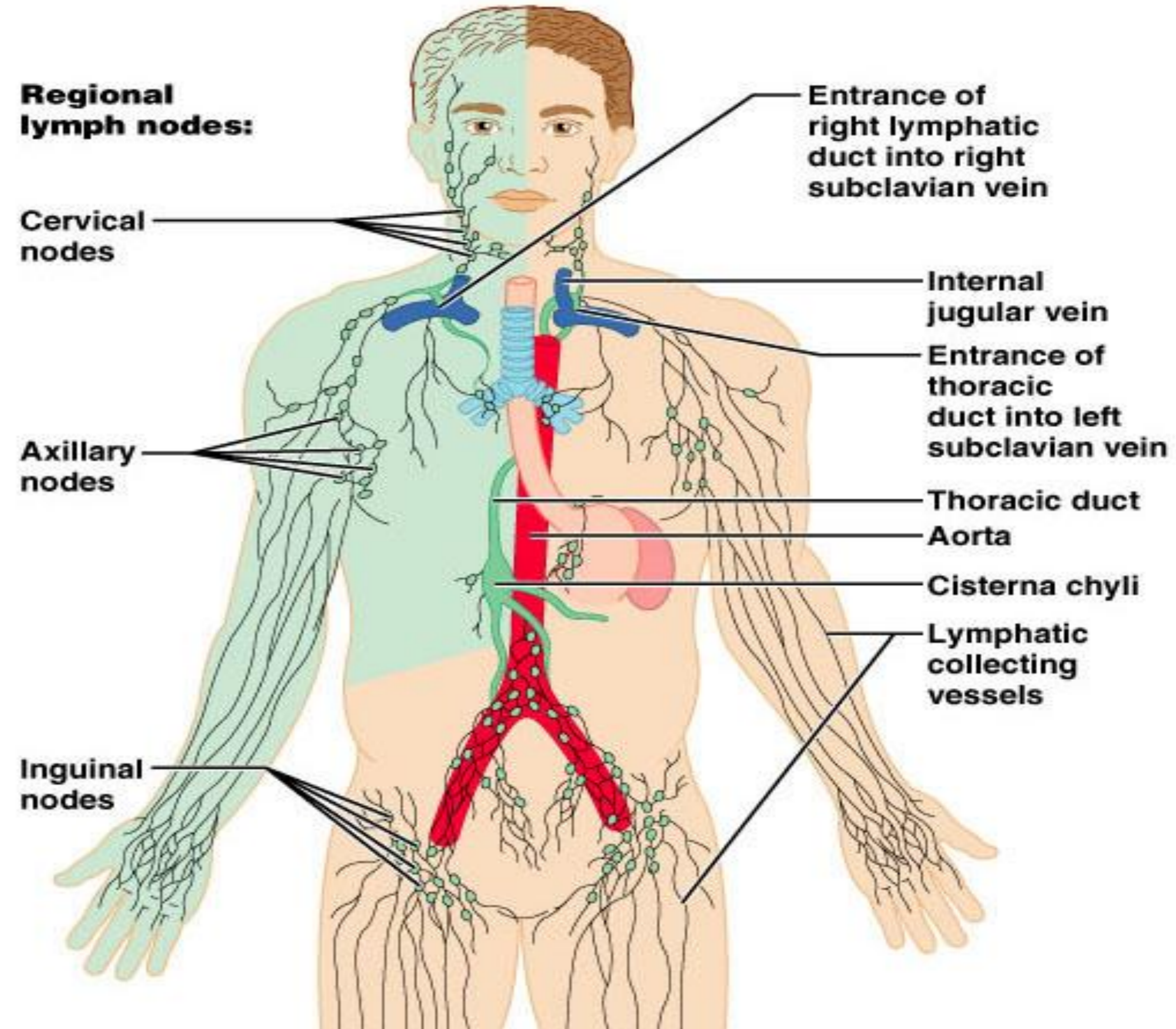
# The lymphatic Circulation

- Perfect system favouring forward flow
- Contraction of Lymphatic Vessels.
  - Pacemaker cells.
  - Valves every 1/3 mm
  - Pulsatile flow.
  - “Twist” of lymphatic vessels
- A lymphatic “Heart”
  - Rate
  - Contractility.
  - Preload
  - Afterload



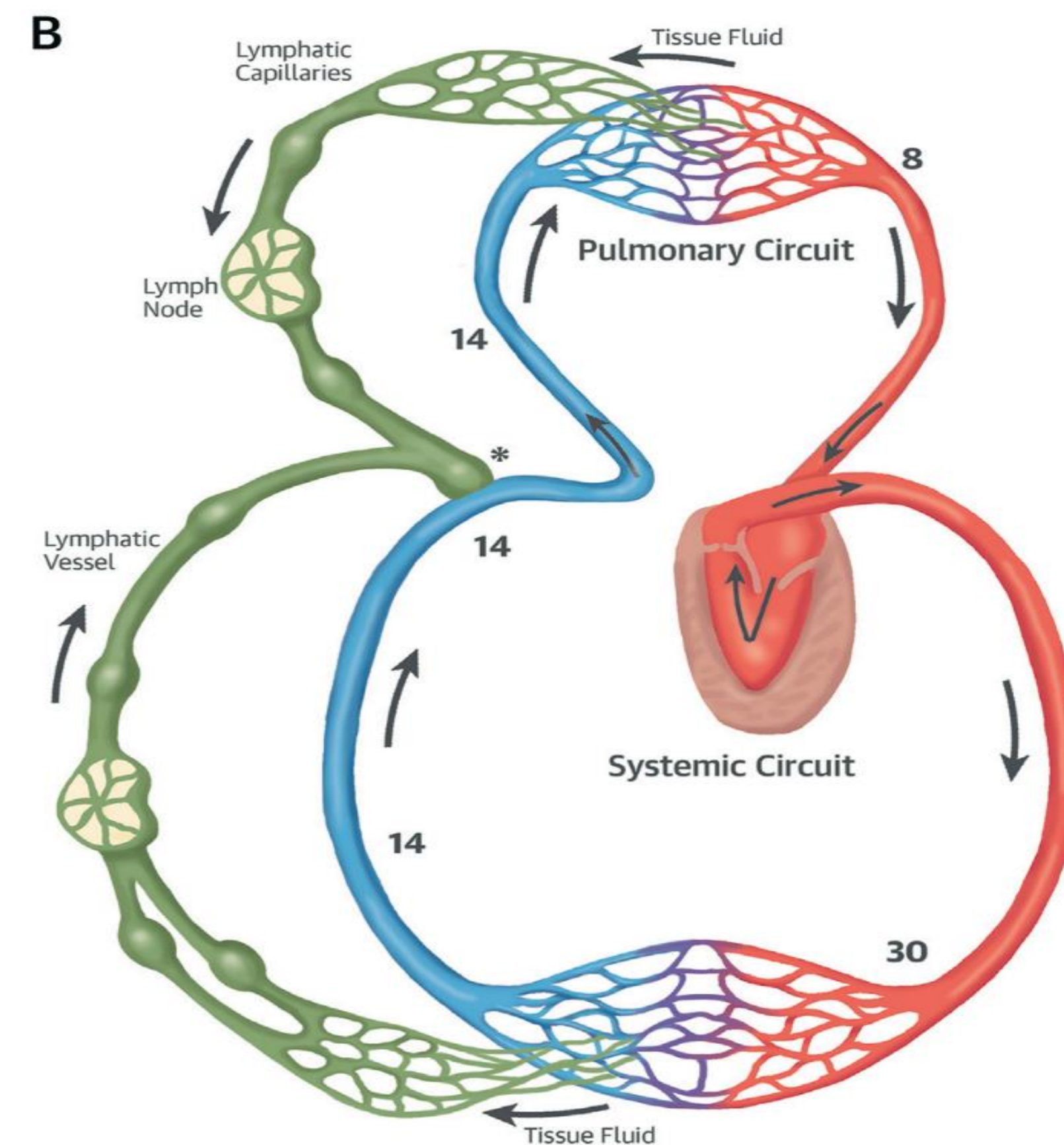
# the lymphatic circulation

- Thoracic duct carries 85% of the total lymph flow
- Right Lymphatic Duct.
- Dormant Lympho venous Communications.
  - To systemic veins
  - To pulmonary veins on left lung (rare)
- Lymph = 1 % of Venous Return.
  - 10/20 x increase in right heart Failure
  - Increased Preload.
- Cessation of Drainage at CVP of 22 mm Hg.
  - Increased Afterload



# Fontan Kreutzer Lymphodynamics

- Normally lymphatic circulation drains to the lowest pressure site
  - To intrathoracic veins.
  - Drainage increases with inspiration
  - Drainage increases with diastole.
  - “Suction” of Lymph.
- Another True Fontan paradox
  - Lymph is required to drain at a similar or higher pressure than it is produced by hydrostatic pressure.
  - Increased “afterload” for the Lymphatic Circ
- In Fontan physiology, lymphatic circulation drains to high pressure site
  - No diastole
  - Only Inspiration to increase thoracic duct flow



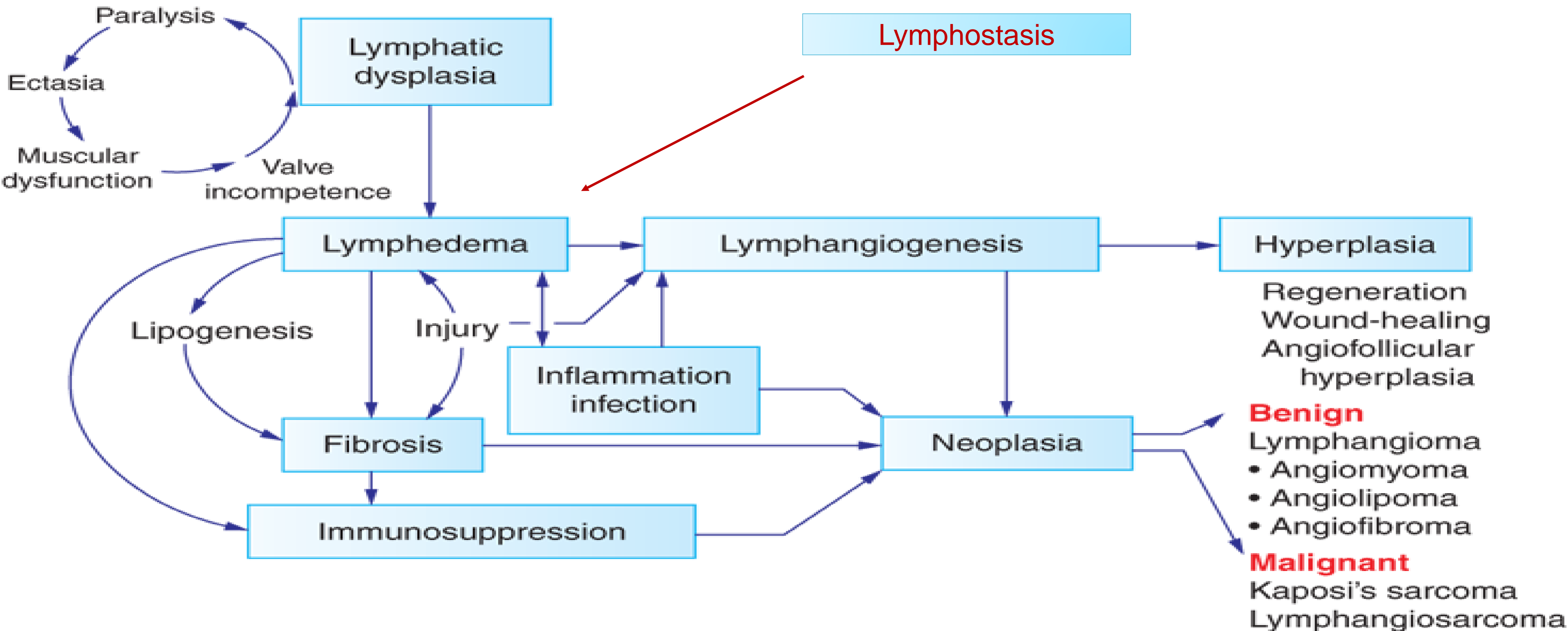


# Lymphatics and Fontan circulation

Fontan circulation operates at or above the functional limits of the lymphatic circulation

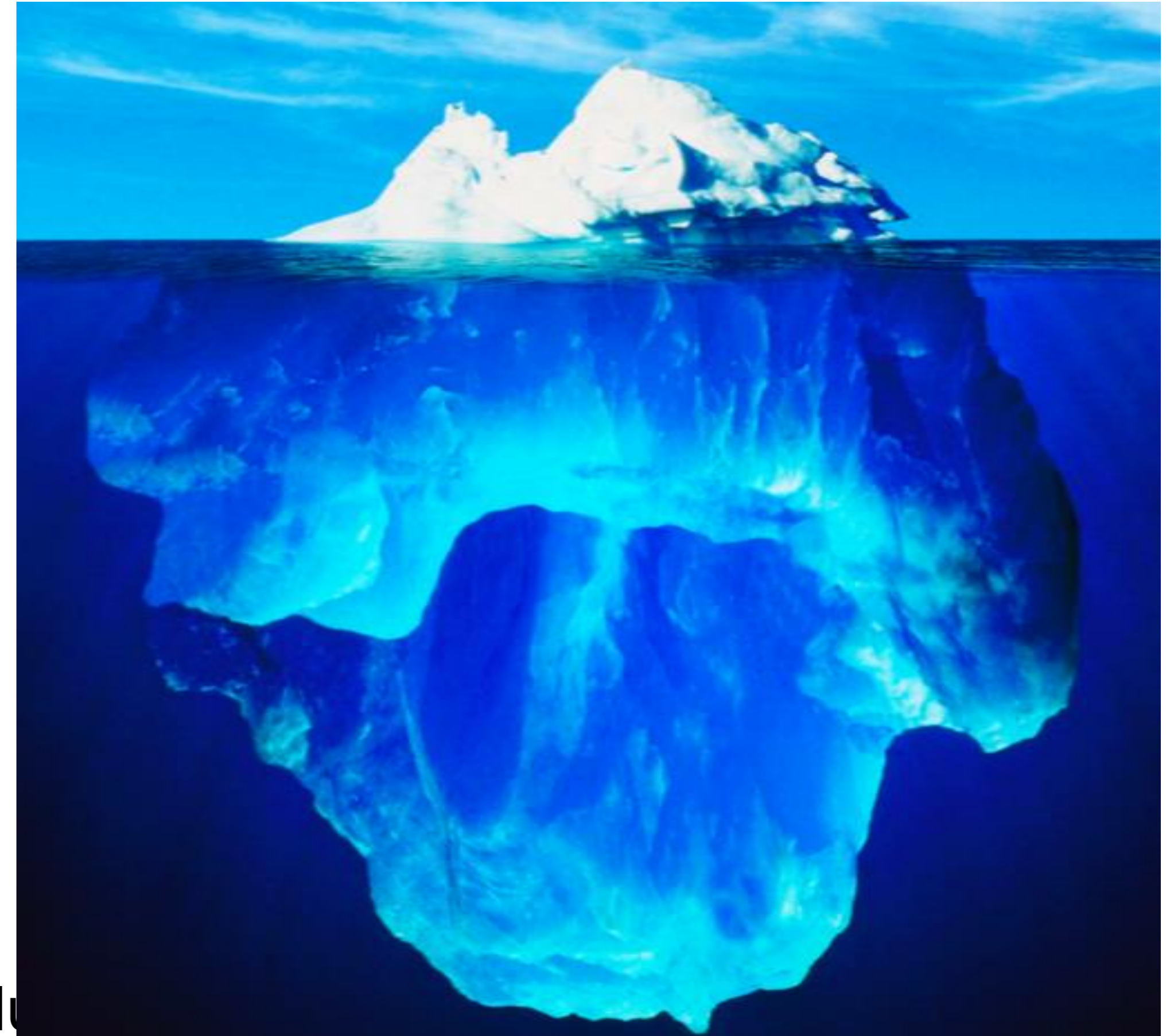
- Lymph drainage compromised
  - High CVP (12-15 mm Hg) Increased Afterload. Cessation of TD flow at 20-25 mmHG
  - Mechanical obstruction (Innom Vein thrombosis)
  - Stasis in thoracic duct. Thoracic duct dilation and valve incompetence
  - No opening of dormant communications.
- Early Lymph Complications
  - Pleural effusions, Pulmonary lymphatic edema, Ascites.
- Late Complications
  - Effusions, Ascites, PLE, plastic bronchitis.
  - Chronic Lymphostasis. Liver fibrosis, Renal Failure, Lung Fibrosis, Myocardial Fibrosis?

# Lymphostasis



# PLE is the tip of the iceberg

- Fontan Kreutzer: a vicious cycle for GI tract
  - Elevated CVP and mild Portal Hypertension
  - Decreased mesenteric perfusion
    - Elevated Mesenteric Vascular Resistance.
  - Lymphostasis & Chronic splachnic inflammation
    - Increased Lymph production.
    - Impaired lymphatic drainage
    - Lymphostais, Inflammation & Fibrosis
  - PLE.
    - Lymphatic decompression into low pressured gut l



# Lung “lymphodynamics”

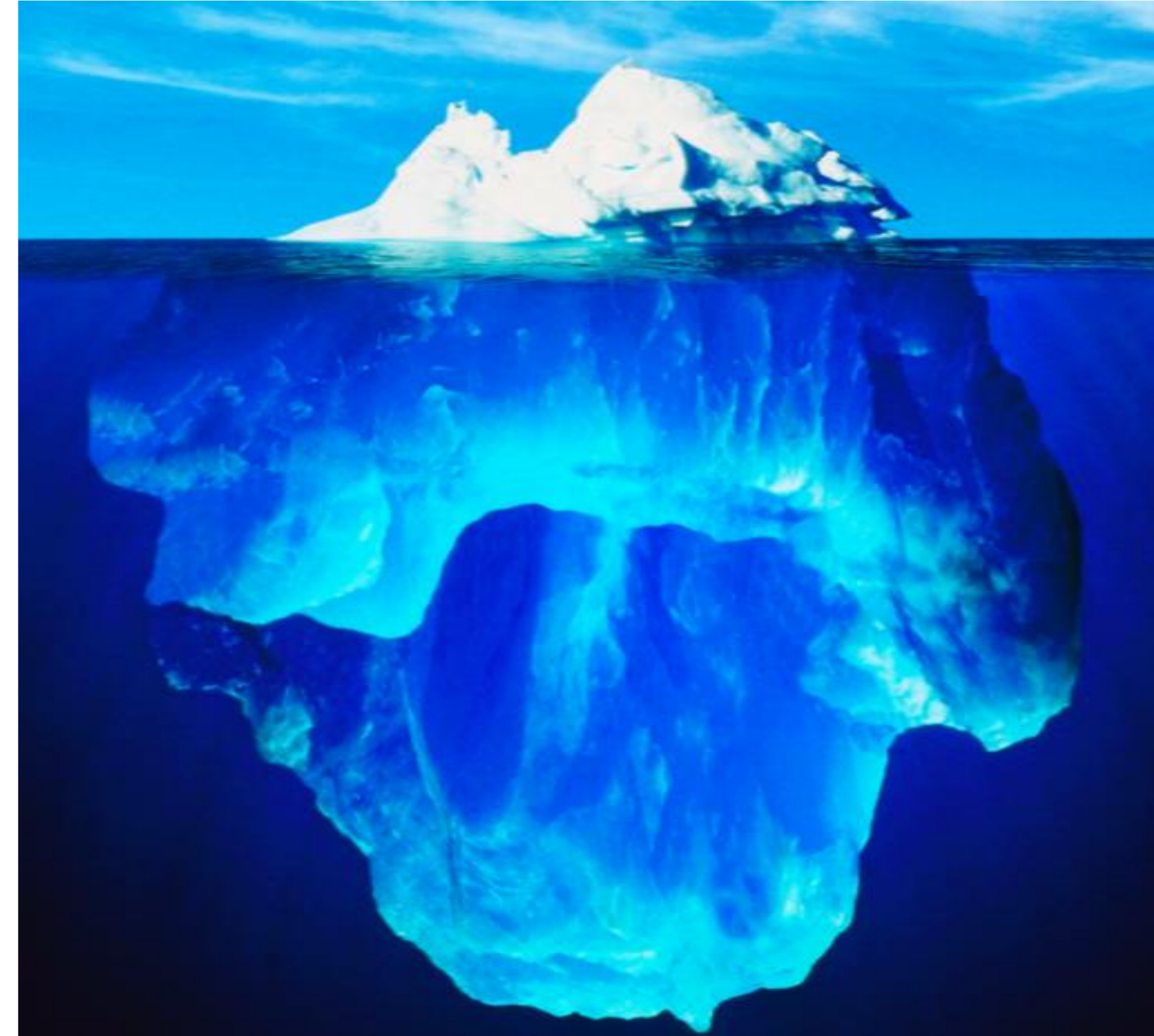
- “you can’t have pulmonary edema in a Glenn or Fontan”
  - You can’t have Hydrostatic pulmonary edema.
  - Constant tendency to fluid accumulation in the lung in the Glenn and Fontan

Typical values of pulmonary pressures (mmHg)			
	PAP (mean)	PCWP	Thoracic Duct pressure =CVP
Normal Circulation	15	8/12	6/8
Fontan Kreutzer or Glenn	12/15	5/8	12/15

- Early: Pulmonary congestion /Effusions.
- Late: Lung fibrosis & Plastic Bronchitis.

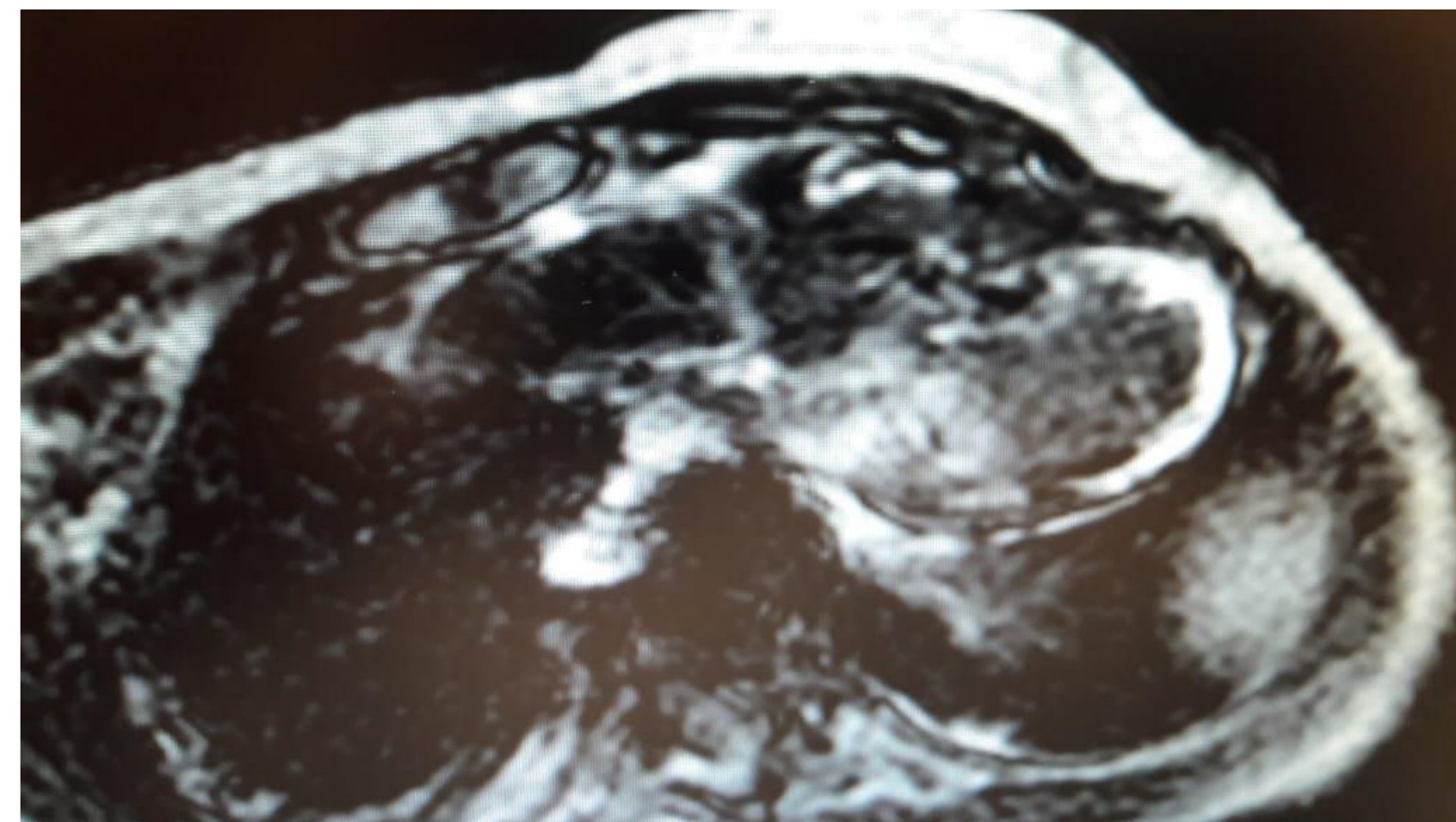
# Plastic Bronchitis: another tip of the iceberg

*“Plastic bronchitis airway casts from children with Fontan physiology are composed of fibrin and are **cellular and inflammatory** in nature, providing evidence that their formation cannot be explained simply by lymph leak into the airways. A derangement in **inflammation** resolution likely contributes to cast formation.”*



# Heart lymphatics

- Extensive Lymphatic network. Drains into Thoracic Ducts
- FX
  - Fluid Balance
  - Removal of Inflammatory mediators
  - Healing.
- Disruption of the lymphatic circulation results in the development of Myocardial fibrosis in dogs
- Fontan circulation
  - Coronary venous drainage @ normal pressure.
- 30 % of adult Fontans show Myocardial Fibrosis by MRI.
- Stiff hearts w diastolic dysfunction.
- A longlife process of Lymphostasis, and chronic inflammation resulting in fibrosis?



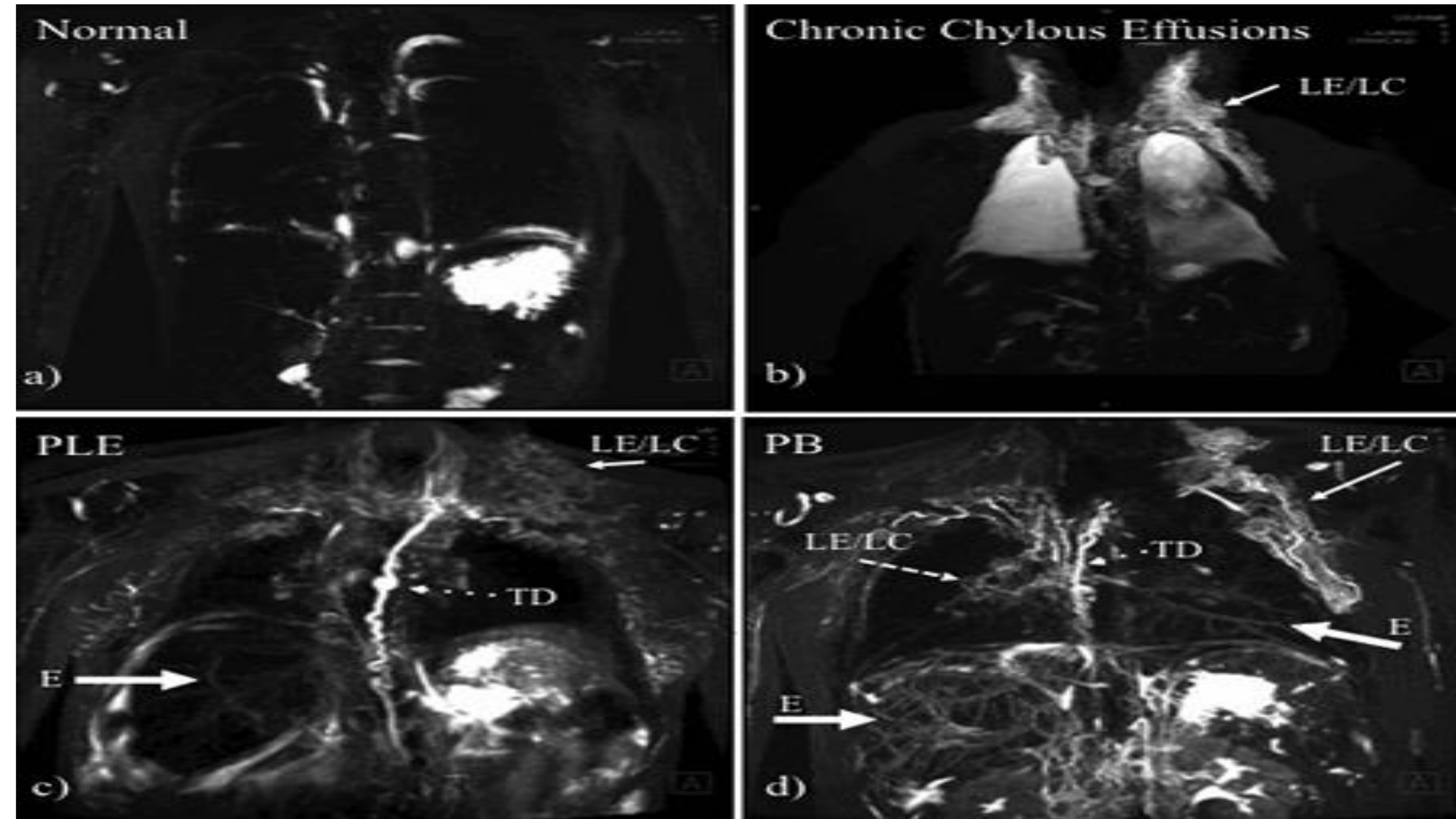
Arch Patol 1963 Oct;76:424-33.

KLINE IK, MILLER AJ, KATZ LN.

# Fontan Lymphatic complications Diagnosis & intervention

M Itkin & Y Dori U. Penn

- Non contrast MRI (T2 weighted)
  - Classification of Abnormalities.
  - Identification of TD and abn.
- CDMRL
  - Circulation. 2013; 128: A16061. Ann Thorac Surg. 2014 Aug;98(2):634-4
  - Pediatrics. 2014 Aug;134(2):e590-5
  - Circulation. 2016 Mar 22;133(12):1160-70
  - Itkin et al. JACC 2017
- Transabdominal Catheterization of TD.
- Lymphangiography and Lymphodynamics
- Intervention on PLE and PB



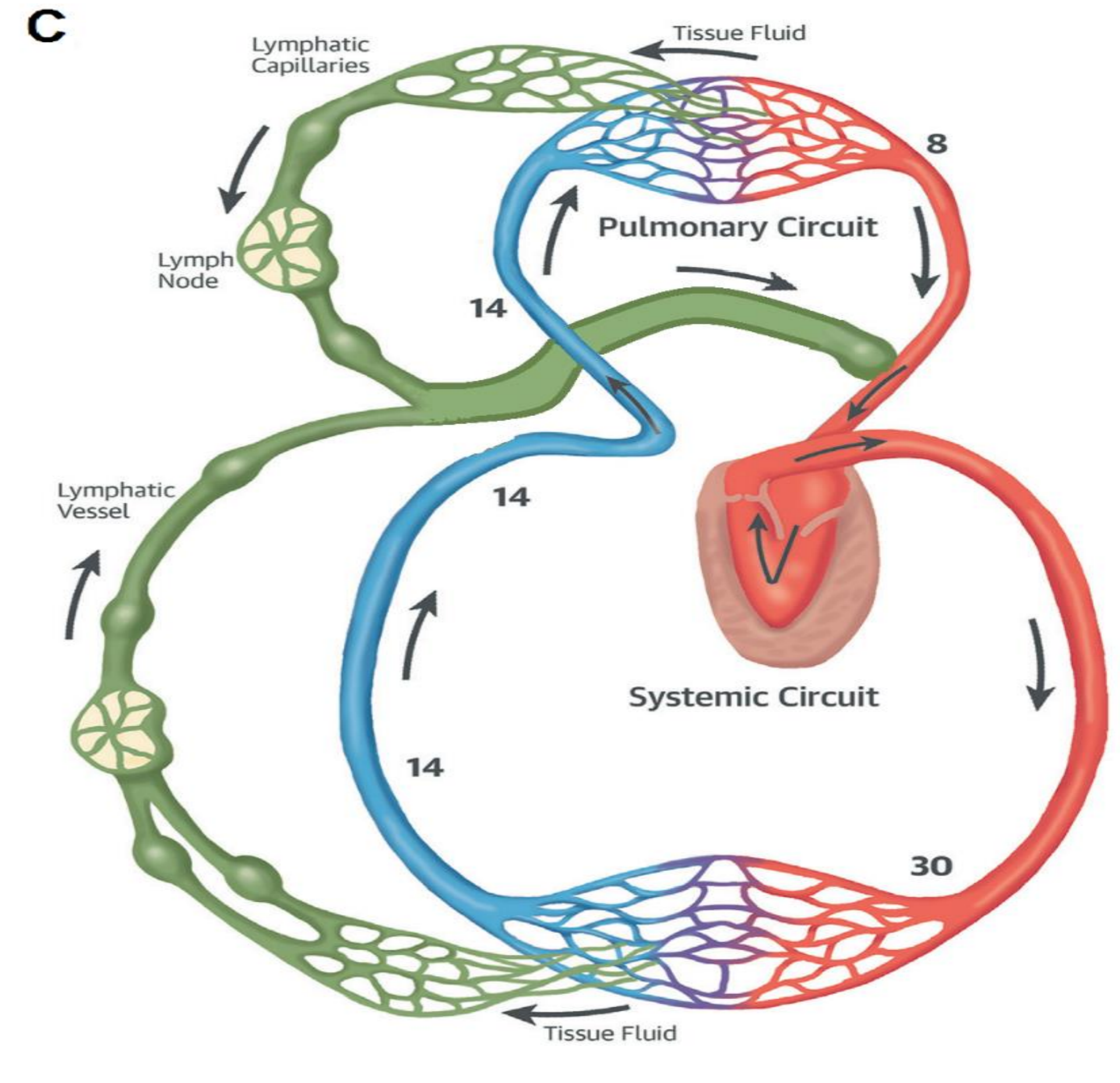
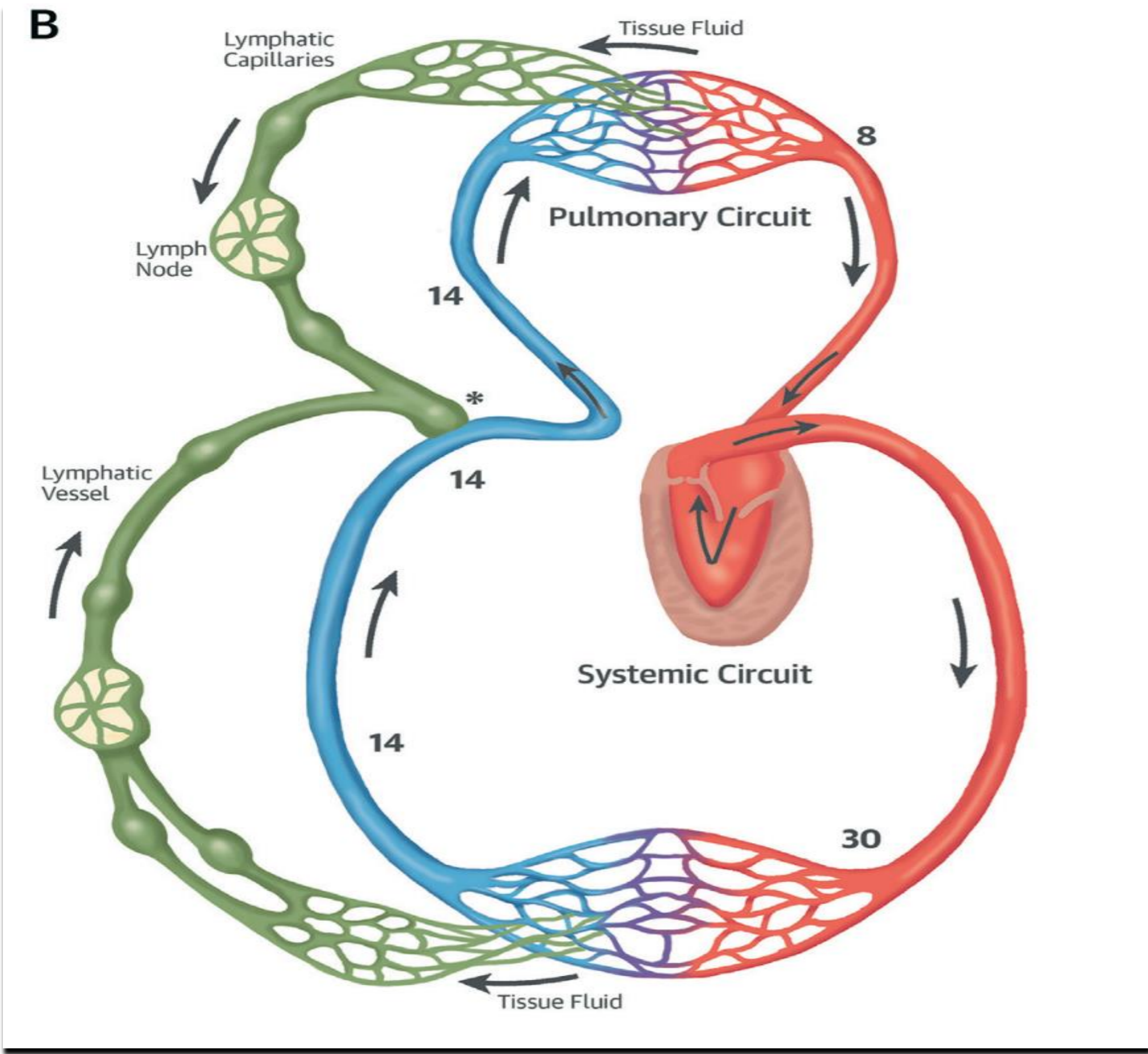
Maximal Intensity Projections (MIP) of MRI Lymphangiograms in 4 patients. Diagnosis listed in top left corner. (List of abbreviations: PLE-protein losing enteropathy, PB-plastic bronchitis, LE-lymphangiectasia, LC-lymphatic collaterals, TD-thoracic duct, E-edema)

# Late Fontan end Organ Fibrosis

- Mandatory liver/renal/lung surveillance.
- Shall we wait for the inevitable? Organ Fibrosis
- Therapeutic options are limited and primarily used for failing pts.
- An elevated CVP is inherent to the Fontan circulation
- Increased lymph production is inherent to the Fontan circulation
- Impaired lymphatic drainage may NOT be inherent to Fontan circulation



# LYMPHATIC DECOMPRESSION IN FONTAN





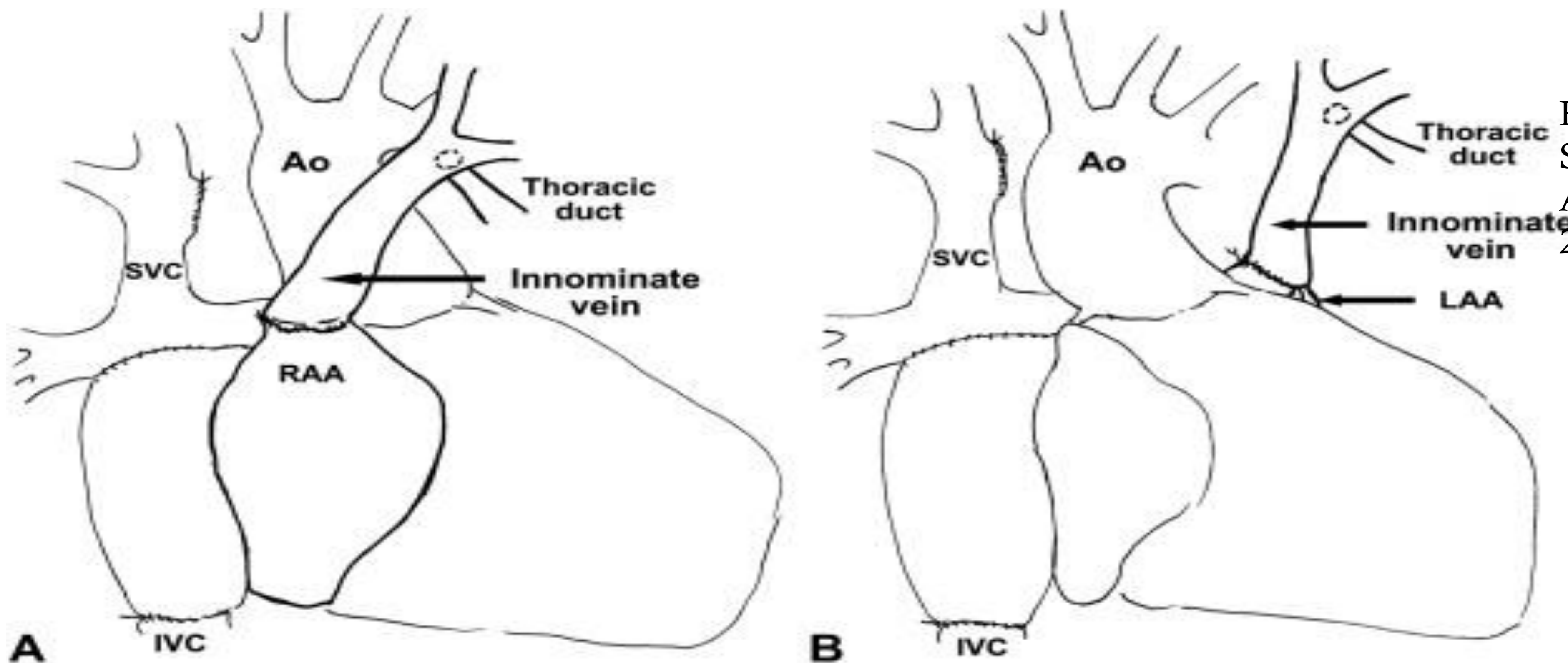
# The holy grail?

Cole, Witte MH Circulation, 1967; 36(4):539-43

- 40 dogs with TR and PS “Fontan” reproduction. (High CVP, low LAP)
- 20 x Increased production of Lymph (ascites, heart failure)
- Increased CVP w Cessation of TD flow.
- @ Day 3 Thoracic Duct to Pulmonary Vein shunt
  - Increased lymph drainage
  - Complete Resolution of ascites
  - Increased Na and Water Excretion.
  - Reduction of CVP from 15 to 10 mmHg
    - Increased Cardiac Index?
- *“Experimental evidence that the manifestations of circulatory congestion can be relieved in dogs with isolated right-heart failure when the TD is anastomosed to a normotensive pulmonary vein”*

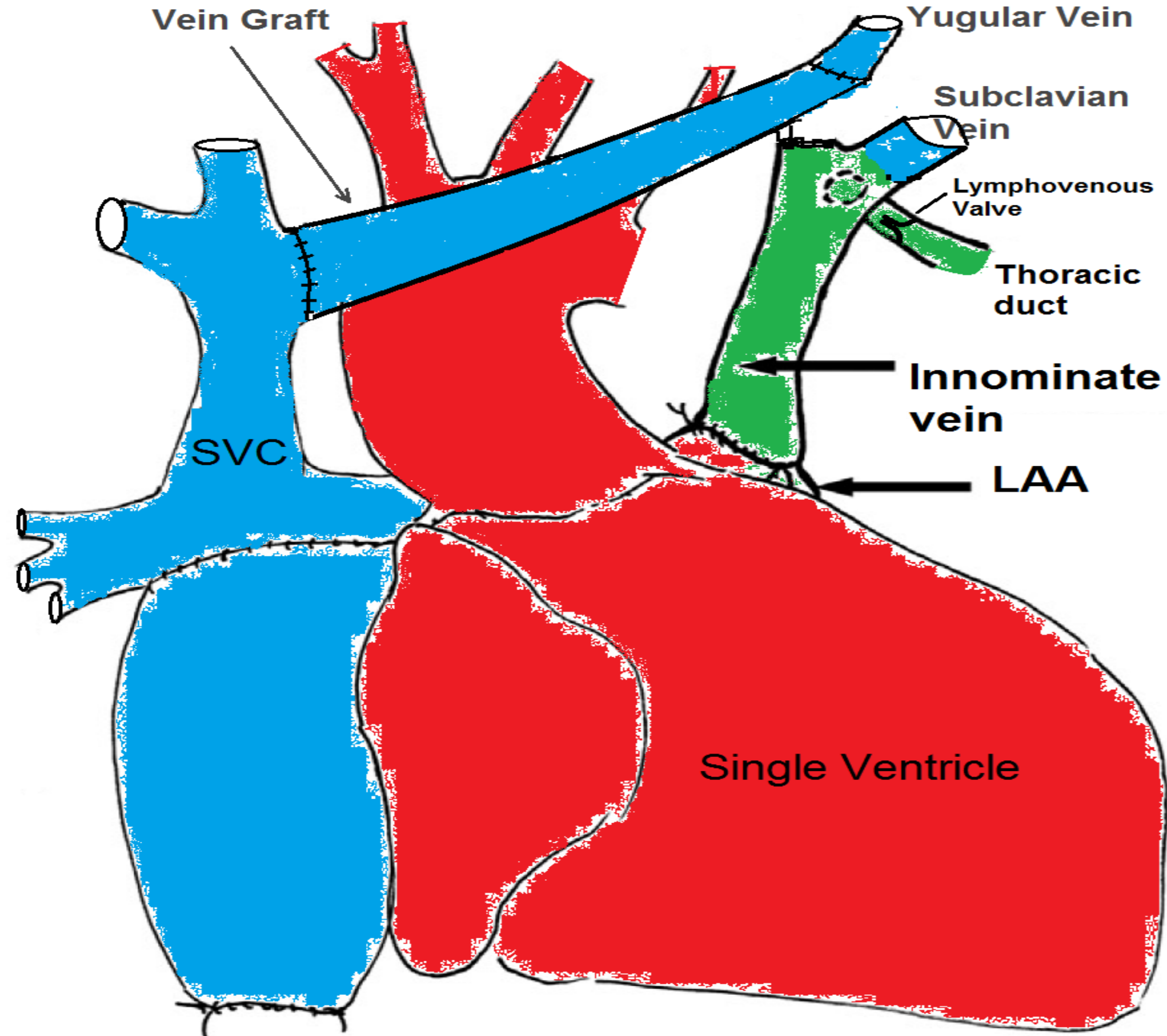
# Hraska procedure

- TD decompression via Innominate vein-left atrium anastomosis for PLE.
  - Inn vein take down.
  - Fenestration
    - Decompression of the Fontan through the brain veins/transverse sinus



Hraška, V Ann Thorac Surg. 2013; 96(2):709-11  
António, Ann Thorac Surg. 2016;101(6):2370-3

# TD decompression: How and when?

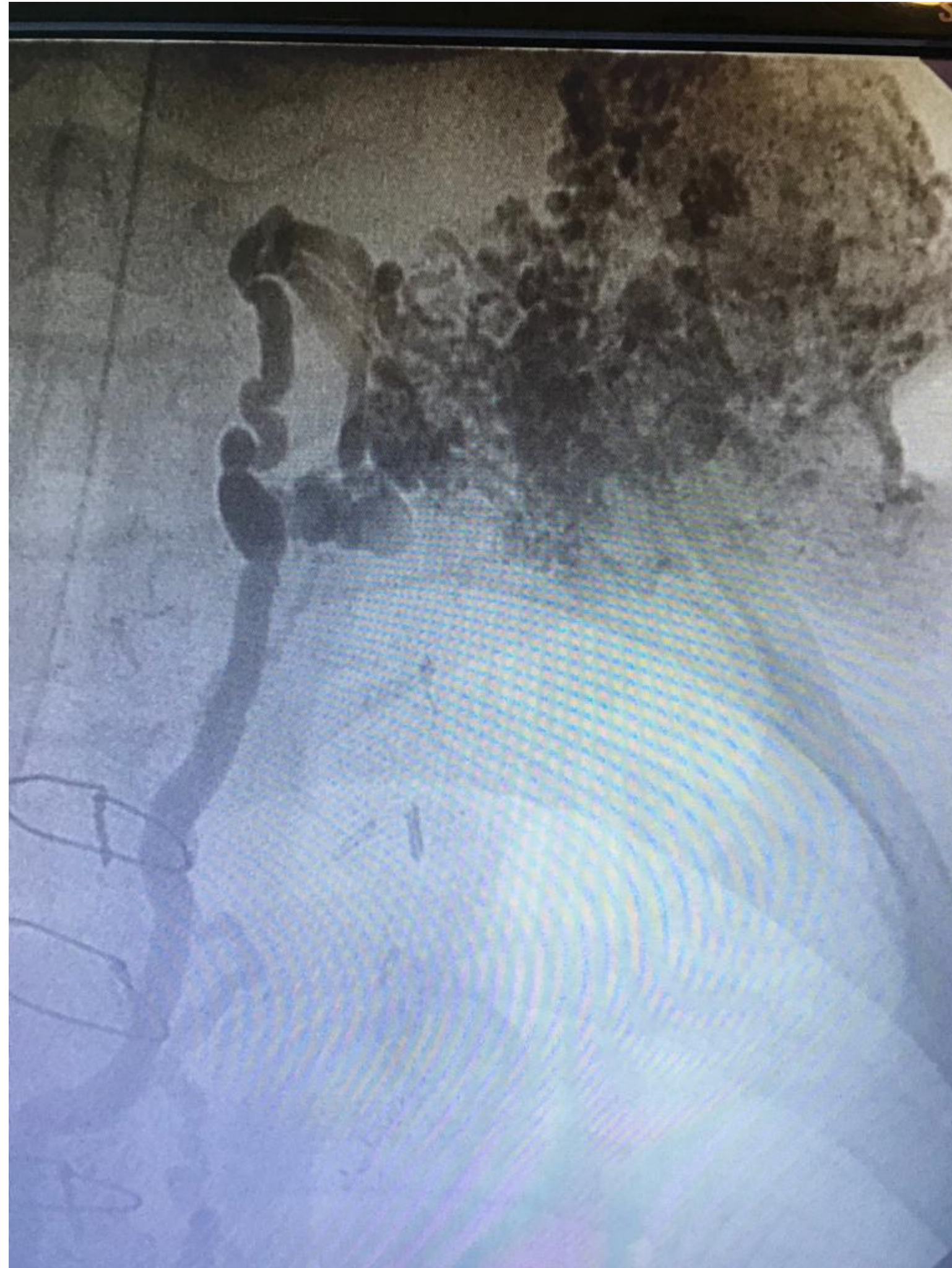


- Modified Hraska Procedure.
  - PTFE graft
  - Vein homograft
  - Subclavian vein flow to “flush” the system
- Failing Fontan with PLE/PB +/- Organ Failure
- W
- Early failure, Ascites and Hydrothorax
- Concomitant to Fontan procedure for high risk patients?
  - Thoracic Lymphangiectasia by MRI.
  - Elevated PVR
  - Systolic/diastolic dysfunction

# Clinical experience in Failing Fontans

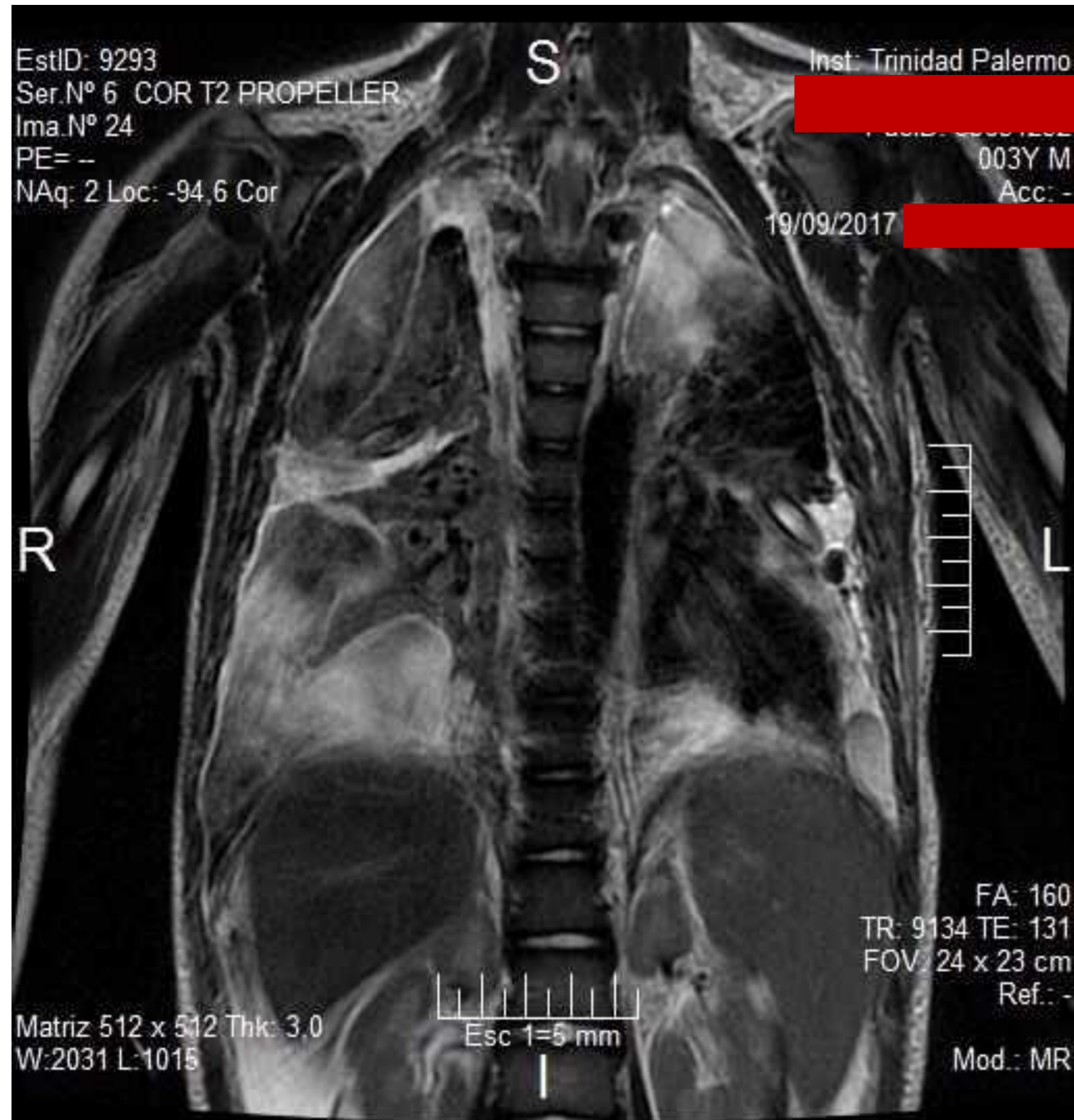
Age (y)	Weight (kg)	Diagnosis	Time since Fontan	PLE	Ascites	Effusions	Plastic Bronc.	Procedure	Outcome
5	15	Heterotaxy syndrome, asplenia, common AVVR,	2y	yes	Yes, massive	Yes, Bilat	No	R Glenn take down, AVVR.	Late death, Pulm Hemorrhage 3m.
3	8	HLHS	4m	yes	Yes, massive	Yes, Bilat	No	Hraska	Alive, 8m, Class II
4	15	Heterotaxy syndrome, asplenia, <i>depressed RV function.</i>	1m	no	Yes	Yes, Bilat	No	Hraska	Early death, Vent Dysfx
6	21	<b>PA IVS Stenotic BDG</b>	2 y	No	no	Yes, right	yes	Hraska	Alive recurrent PB

# Case IV



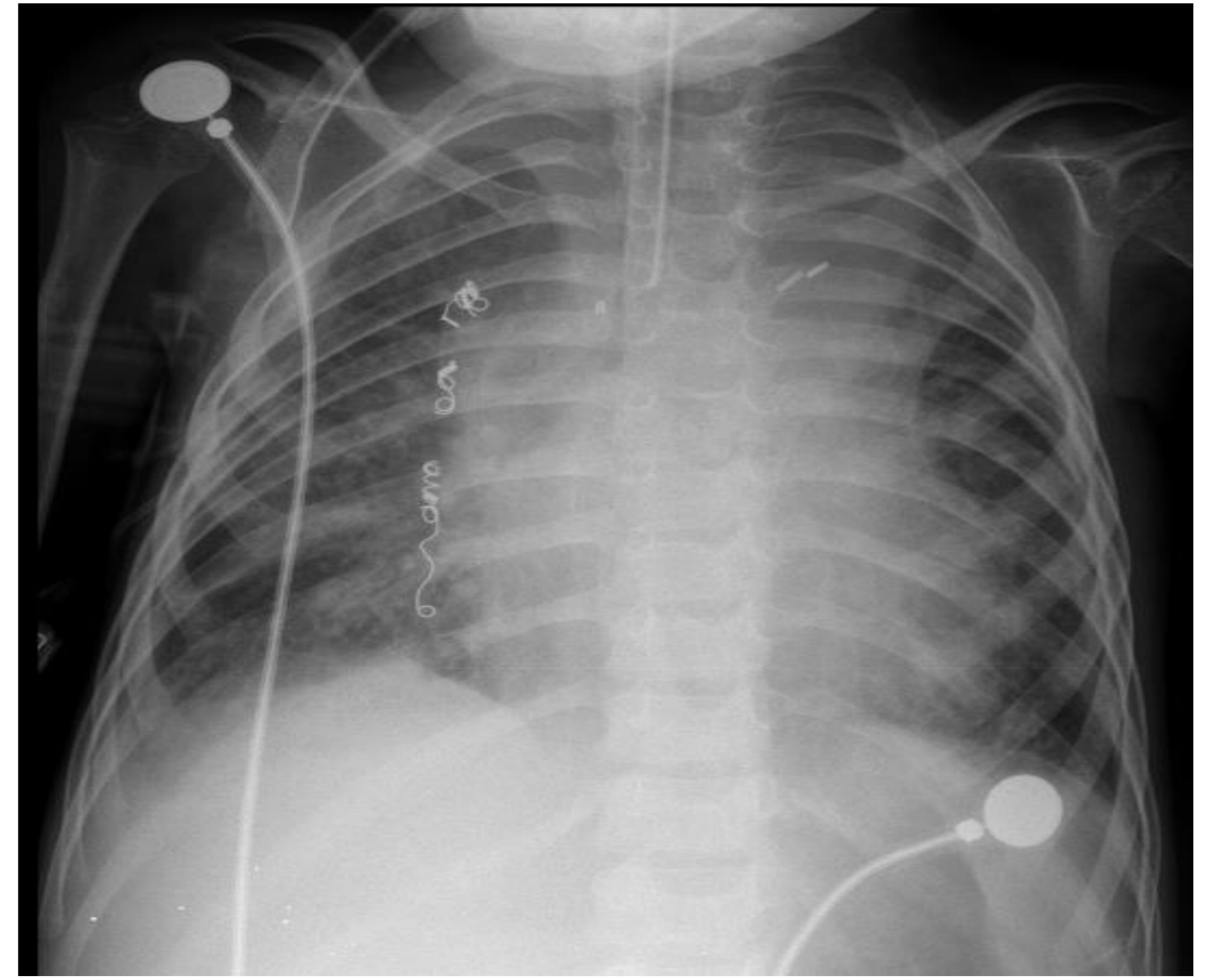
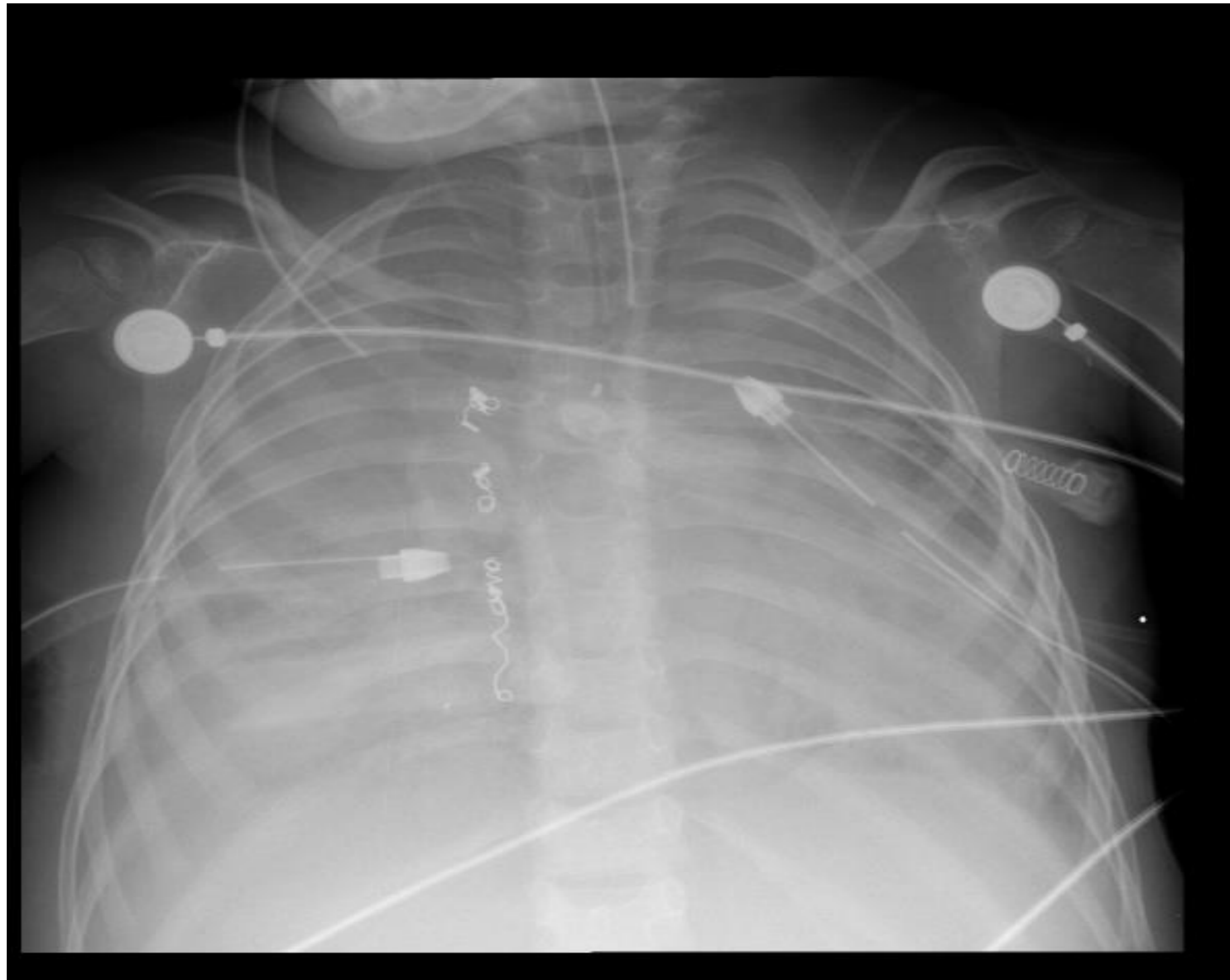
- 6 y/old. 2 yrs post Fontan
- Plastic Bronchitis. 2 episodes per month
- Hraska procedure with LIJ ligation.
- Recurrent PB.
- Embolization and complete cessation.
- First case of combined Lymphatic decompression and intervention for Lymphatic complications

# Case II



- 3 y/old. 4 m post Fontan
- Unbalanced AV Canal status post Norwood and Glenn
- Cachectic patient, 8 kg, ventilated 45 d.
- Fontan Failure with chronic ascites and Hydrothorax (600/800 ml/d)
- TD decompression
  - “Classic Hraska”
- Complete cessation of Effusions and Ascites in 6 days.
- Nutritional support, discharged home on Oxygen.
- Fup 8m , 10.5 Kg, Right effusions.

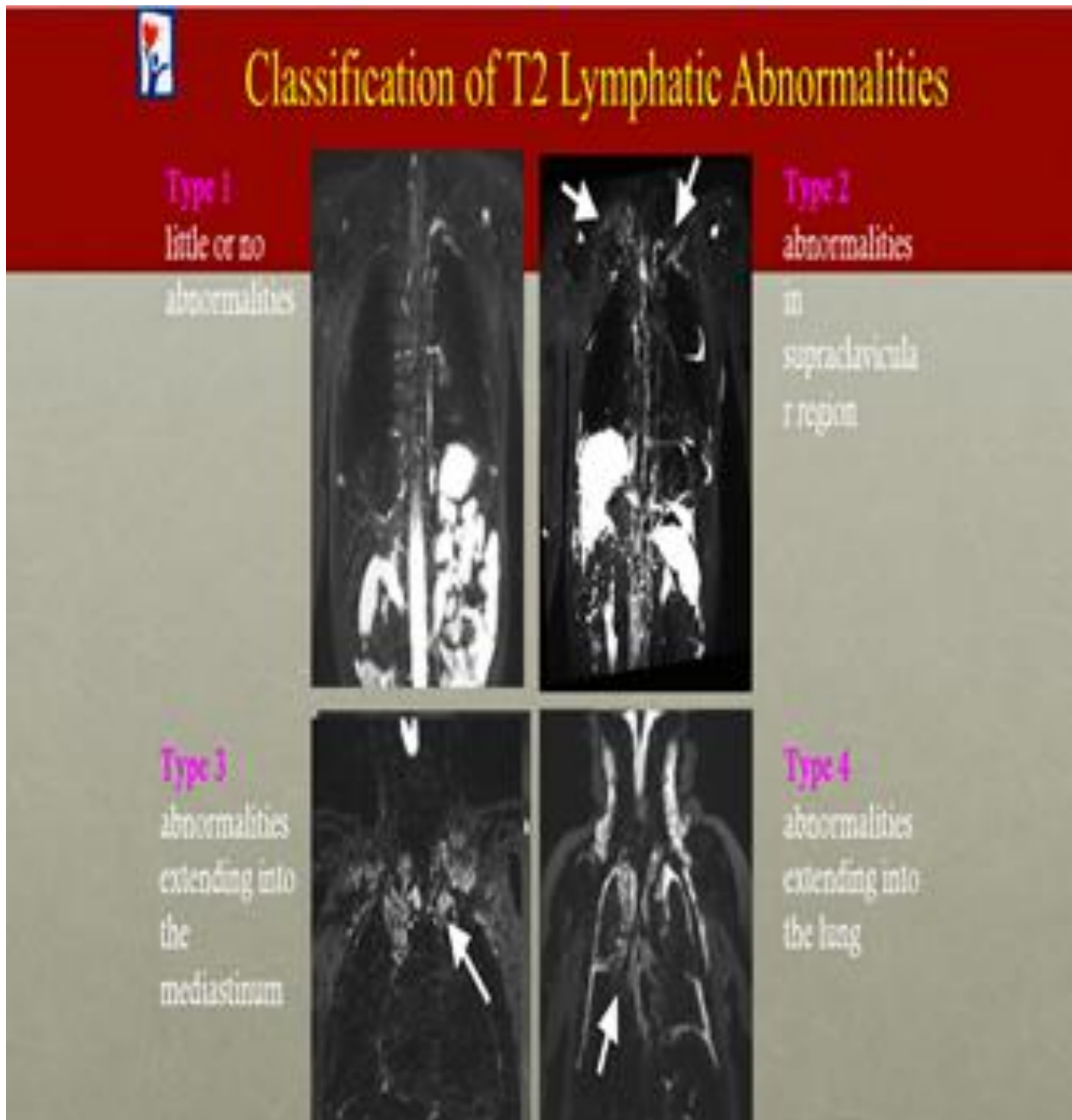
# chest x-ray Pre/Post Hraska 4th PO day







# Fontan Outcome of 83 Glenn Patients With Pre Fontan T2 MRI Imaging (mandatory!!)

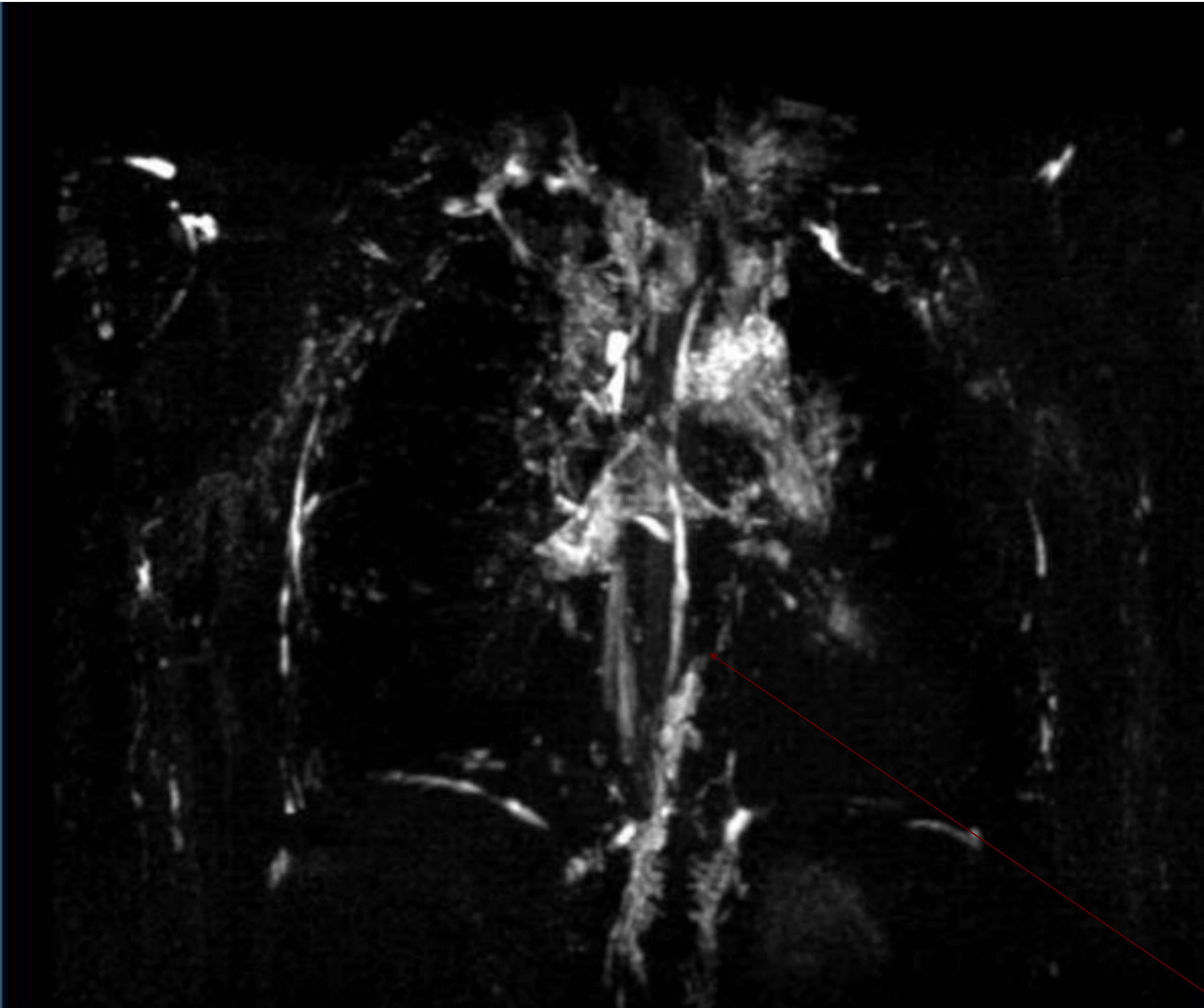


	Type 1/2	Type 3	Type 4	<i>p</i>	Type 1/2 vs Type 3	Type 1/2 vs Type 4	Type 3 vs Type 4
<b>TCPC Completion</b>							
Yes	52 (98.1%)	16 (94.1%)	6 (46.2%)	<0.001	0.429	<0.001	0.009
No	1 (1.9%)	1 (5.9%)	7 (53.8%)				
<b>TCPC Takedown</b>							
Yes	0 (0.0%)	0 (0.0%)	1 (7.7%)	0.066	n/a	n/a	n/a
No	53 (100%)	17 (100%)	12 (92.3%)				
<b>Transplant</b>							
Yes	0 (0.0%)	0 (0.0%)	3 (23.1%)	0.023	n/a	0.036	0.179
No	53 (100%)	17 (100%)	10 (76.9%)				
<b>Mortality</b>							
Alive	53 (100%)	17 (100%)	8 (61.5%)	<0.001	n/a	<0.001	0.009
Deceased	0 (0.0%)	0 (0.0%)	5 (38.5%)				
<b>Mean Duration of Effusions (d)</b>	6.88 (3.45)	12.13 (9.99)	14.75 (6.85)	<0.001	0.006	0.029	1
<b>Mean Duration of Hospital Stay (d)</b>	9.31 (3.23)	24.44 (29.12)	25.5 (7.05)	<0.001	0.001	0.085	1

# Lymphatic decompression @Fontan

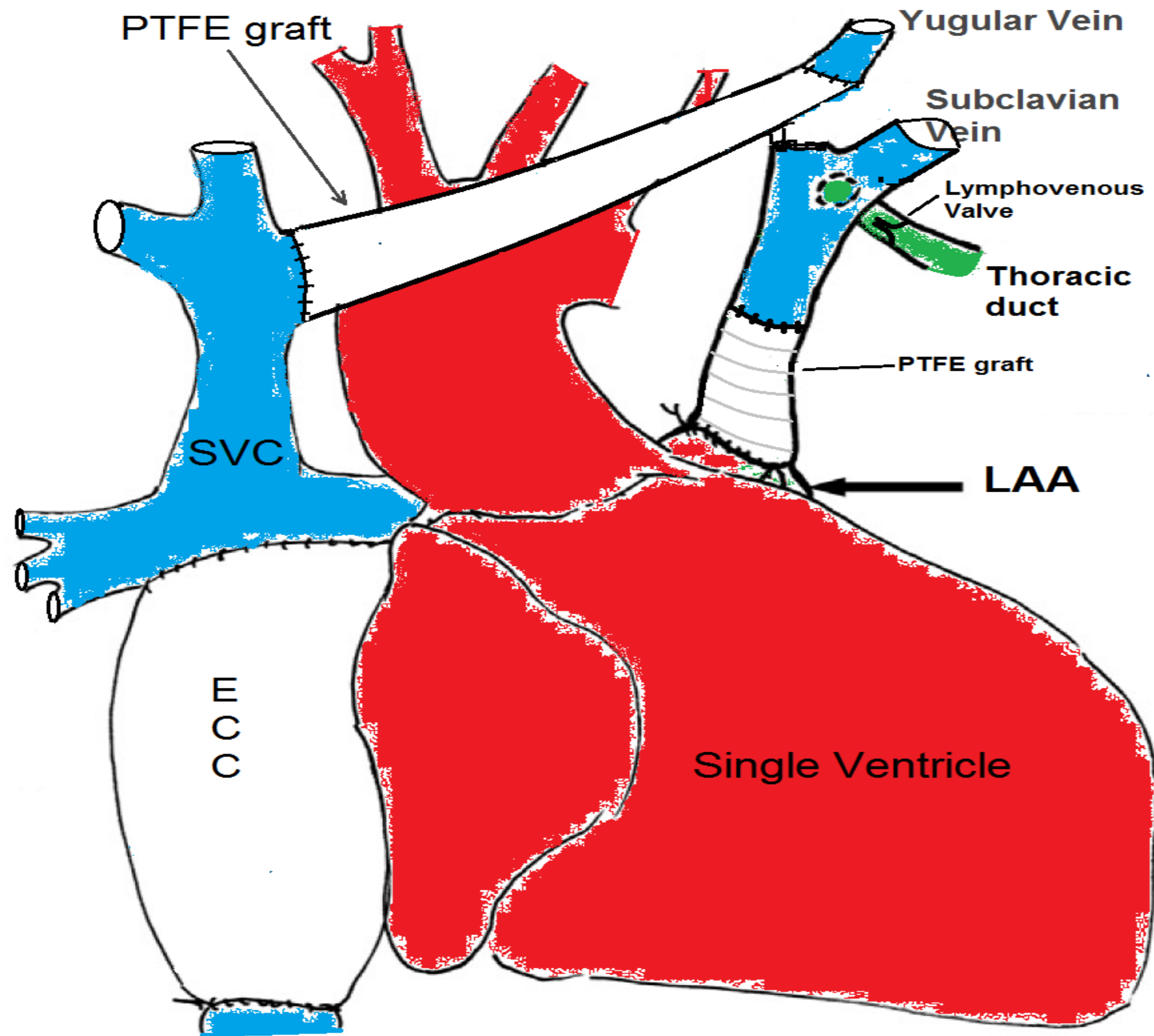
Age (y)	Weight (kg)	Diagnosis	Pre Fontan MRI classification type	Chest tube duration (days)	Procedure	outcome	Follow UP
22	55	DILV, SLL, Rest BVF, S/P Banding and Glenn	4	2	Modified Hraska. LIJV to RSVC graft	Alive	20 m, Class I
3	12	HLHS S/P Norwood and Glenn	4	5	Hraska	Alive	6 m. Class I
3	11	DORV MA, S/P banding atrial septectomy and Glenn	3	7, Right Hydrothorax.	Hraska	Alive	2m Class 1.
2	13	HLHS	4	3	Hraska	Alive	2 m Class I

# Case I



- 22 y/o criss cross heart, S/P Glenn and PA banding @ 4 yrs.
- Severe cyanosis
  - Sat 70/75 %
  - PA pressure 15 mm Hg.
  - Massive Lymphangiectasia by MRI
  - MRI CHOP class:
    - Type 4 Abnormalities
    - TD dilatation and Tortuosity
    - Pulmonary Lymphangiectasia

# Case I



- TD decompression
  - Extracardiac conduit Fontan
  - Fenestration via subclavian vein
  - PTFE graft LIJ-SVC
- Extubated POD #1, No effusions
- Alive 20 m after FK
- Class I

# Lymphatic decompression in Fontan Kreutzer

- Potential benefit
  - Improved lymphatic drainage (decreased Afterload)
  - Lower CVP: Lymph flow outside Fontan flow. (Decreased Preload)
  - Ideal Fenestration: no systemic desaturation
  - Decreased hepatic congestion. Fibrosis and cirrhosis?
  - Prevention of PB and PLE
  - Resolution of PB and PLE post Fontan?
    - Word of Caution. Concomitant intervention.
  - Resolution of ascites, pleural effusions.
  - Increased cardiac index.
- Potential detriment
  - Unknown effects of lymphatic flow (Inflammatory mediators, Bacteria, etc) to the left sided circulation without pulmonary first pass.
  - Technical difficulties, leaks and blockade of lymphatic drainage.

# Summary

- **Lymphatic Intervention is here and it may benefit outcome**
  - **Interventional Cath for PLE and PB.**
  - **Surgical Lymphatic decompression may**
    - **Amielorate fibrosis and End Organ Failure**
    - **Improve cardiac output.**
    - **Prevent or treat Fontan Lymphatic Failure.**
      - **Early**
      - **Late**