Surgical Strategies to Treat Lymphatic Insufficiency in Congenital Heart Disease

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26th Annual Update on Pediatric and Congenital Cardiovascular Disease

From left to right: Viktor Hraska, Vibeke E. Hjortdal, Yoav Dori, Christian Kreutzer

"The Lymphomaniacs"


mark
wahlberg
mila
kunis
seth
macfarlane
ted

Single Ventricle lesions in 2023

- LT/ECC Fontan K > 25-year follow-up
- Not a “failed strategy”.
  - Excellent Survival= 90 % at 30 years. (1)
  - QOL is reasonable
- Fontan K Failure 10/20 %
  - Preventing late failure is the new challenge.
  - Identifying patients at risk for late M/M with normal Ventricular Fx
    - Identifying patients at risk for lymphatic failure.
    - Treatment for patients with lymphatic failure.

(1)AUS/NZ Fontan Registry Report 2019

Lymp ulation

Fontan K circulat of the lymphatic

- Increased lymph
- Lymph drainage
  - No diastole.
  - High CVP (12-15
  - Stasis in thoracic
- "Lymph will find
  - Early Lymph Co
    - Pleural effusions
  - Late Complicati
    - Effusions, Ascite

ce MA, Am J Physiol 258, 1990

etence & pumping failure.

Lymphedema=organ fibrosis

Late complications on Fontan circulation

Ectasi

Musc
dysfun

Open access

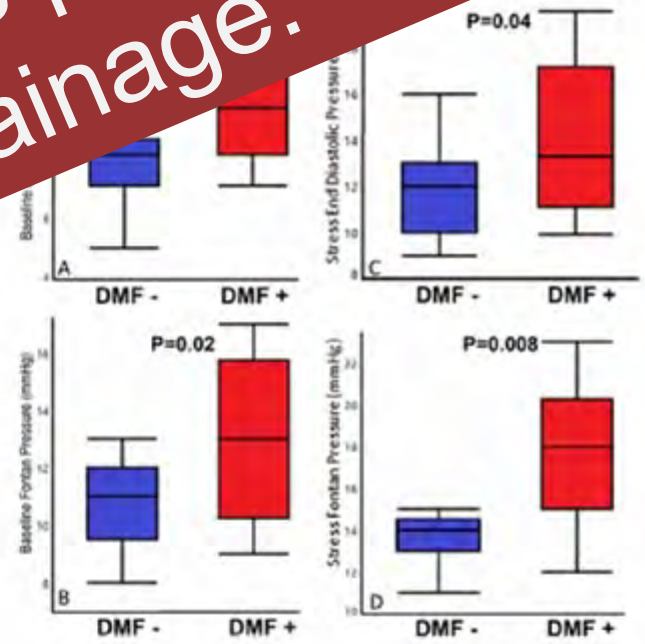
openheart

**Myocardial fibrosis, dias
dysfunction and el
in the Fonta**

Figure 5 Gr e results and the potential relati n hepatic and cardiac fibrosis. MMPs, matrix r proteinases; TIMPs, tissue inhibitor of metalloproteinases.

Sourc
Venous and Lymphatic Diseases, www.cardiology.mhmedical.com

We must do something!
1. Reduce Central Venous pressure.
2. Improve Lymphatic Drainage.



na
coma

When does it start?

Journal of the American Heart Association

ORIGINAL RESEARCH

Prevalence and Cause of Early Fontan Complications: Does the Lymphatic Circulation Play a Role?

Reena M. Ghosh, MD; Heather M. Griffis, PhD; Andrew C. Glatz, MD, MSCE; Jonat
Christopher L. Smith, MD, PhD; Matthew J. Gillespie, MD; Kevin K. Whitehead, M
Michael L. O'Byrne, MD, MSCE; David M. Biko, MD; Chitra Ravishankar, MD
Yoav Dori, MD, PhD

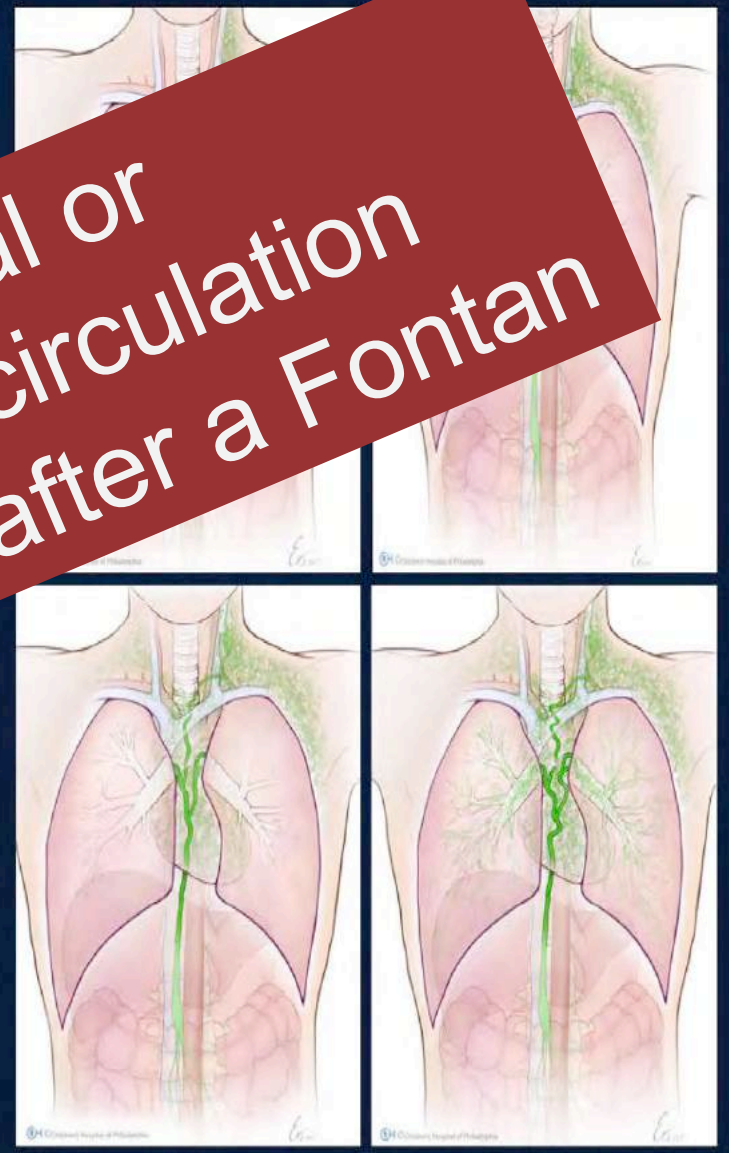
METHODS AND RESULTS: This was a retrospective, single-center study of pati
2012 to 2017. The primary outcome was early *Fontan* complic
takedown, extracorporeal membrane oxygenation
transplant. Complication causes were a
atrioventricular valve regurgitation, (3) pe
tion, and (4) chylothorax or plastic bronch
lymphatic perfusion abnormality. The coho
of 58 (34.5%) in group 1, 8 of 58 (14%) in grou
imaging was available for 126 (53%) patients. F
developing early complications (*P*=0.001).

CONCLUSIONS: There is substantial morbidity in the e
post-Fontan period. Half of those who developed early complications had lymphatic failure or persistent effusions unrelated to structural or functional abnormalities. Preoperative T2 imaging demonstrated that patients with higher-grade lymphatic perfusion abnormalities were significantly more likely to develop early complications. This has implications for risk stratification and optimization of patients before Fontan palliation.

Patients with an abnormal or "exhausted" Lymphatic circulation are going to do poorly after a Fontan

Type 1:
Little or no
abn

Type 2:
Abnormal perfusion in supra-clavicular region

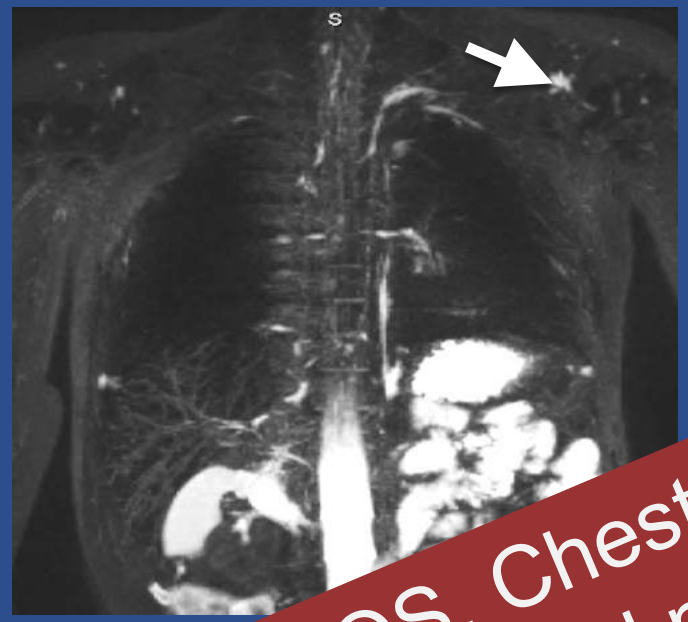


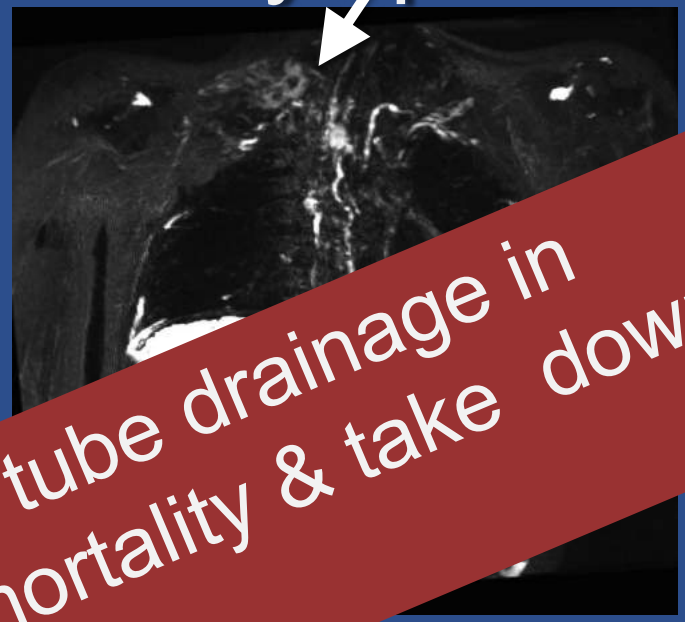
Type 3:
Abnormalities extending into mediastinum

Type 4:
Abnormal perfusion extending into the lung

CHOP Pre Fontan K T2 MRI Lymphatic evaluation

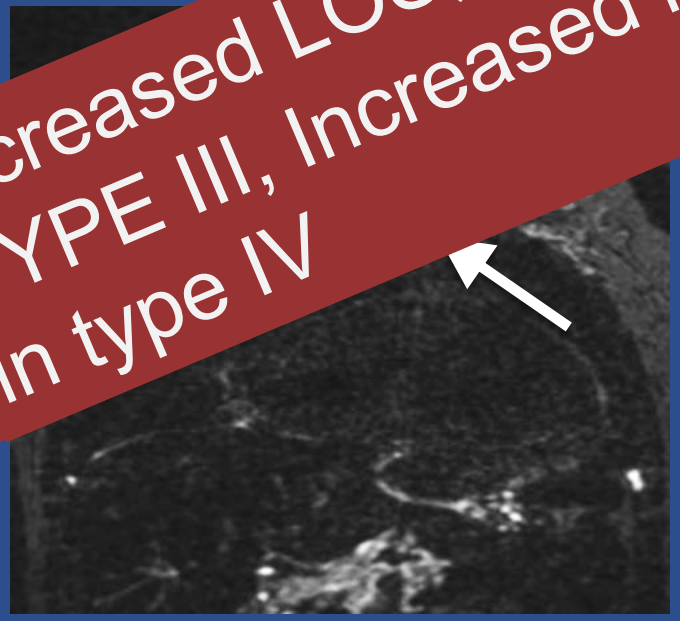
Type 1
little or no abnormalities

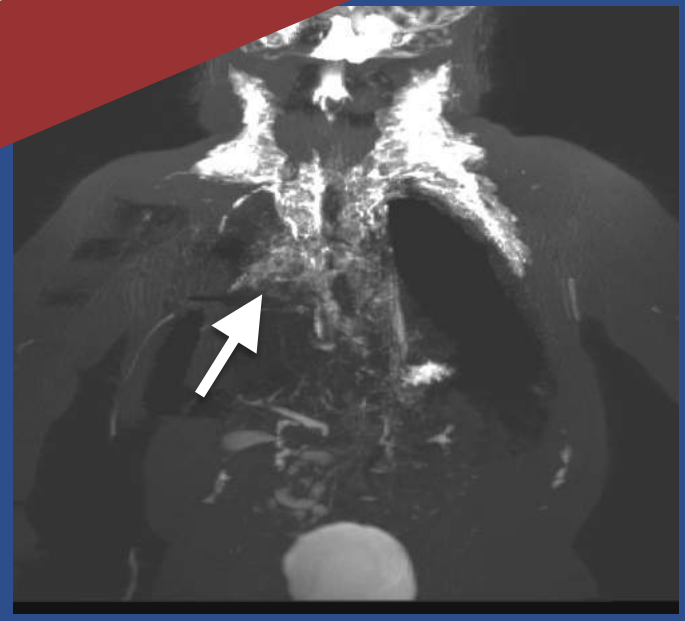




Type 2
abnormalities in supraclavicular region

Type 3
abnormalities extending into the mediastinum



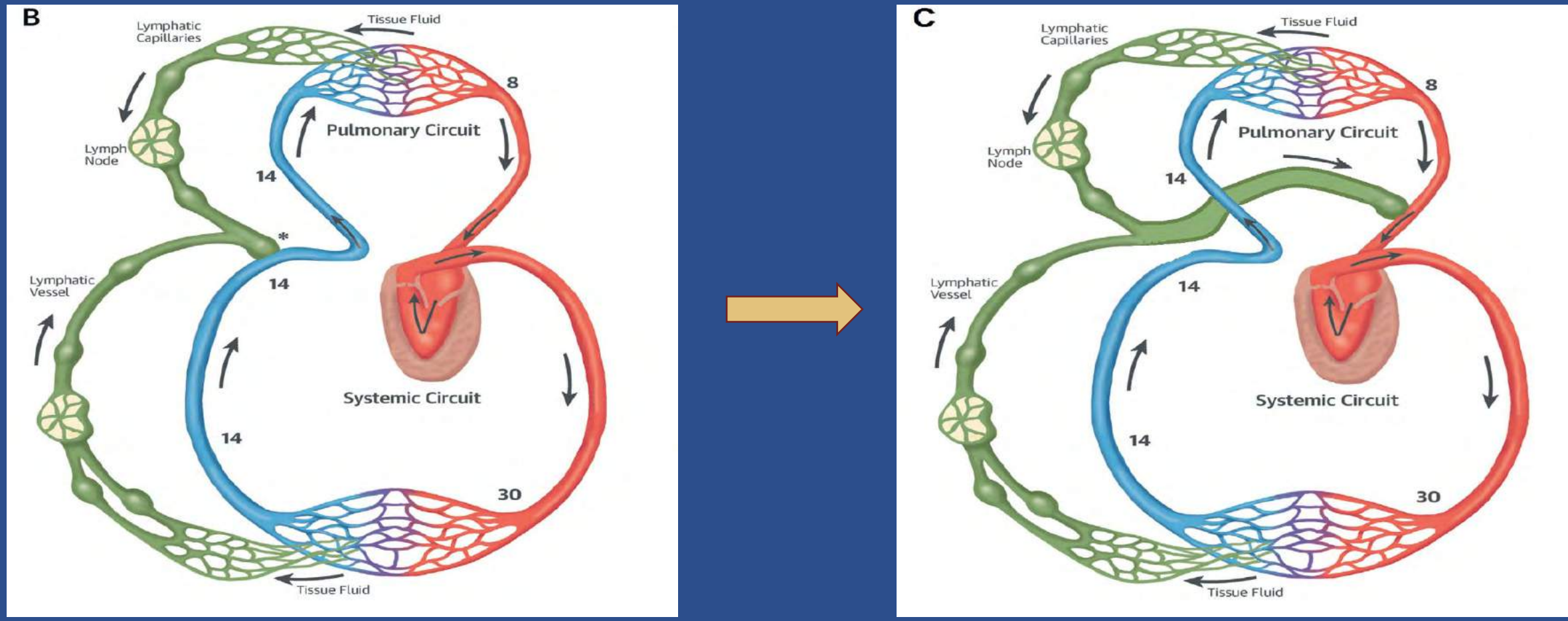


Type 4
abnormalities extending into the lung

Increased LOS, Chest tube drainage in TYPE III, Increased mortality & take down in type IV

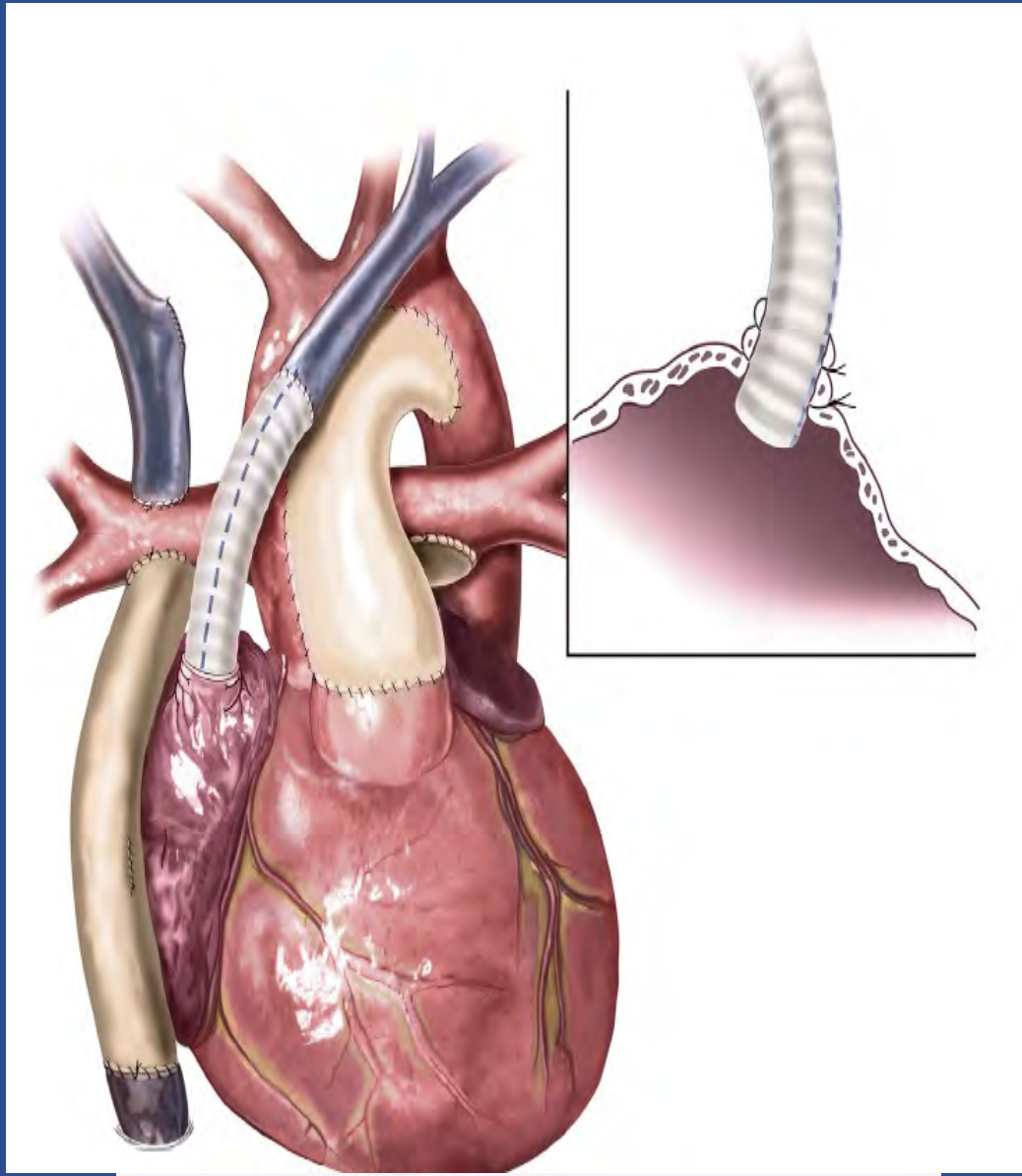
LYMPHATIC DECOMPRESSION IN FONTAN

Restore a “normal” Lymphatic Drainage



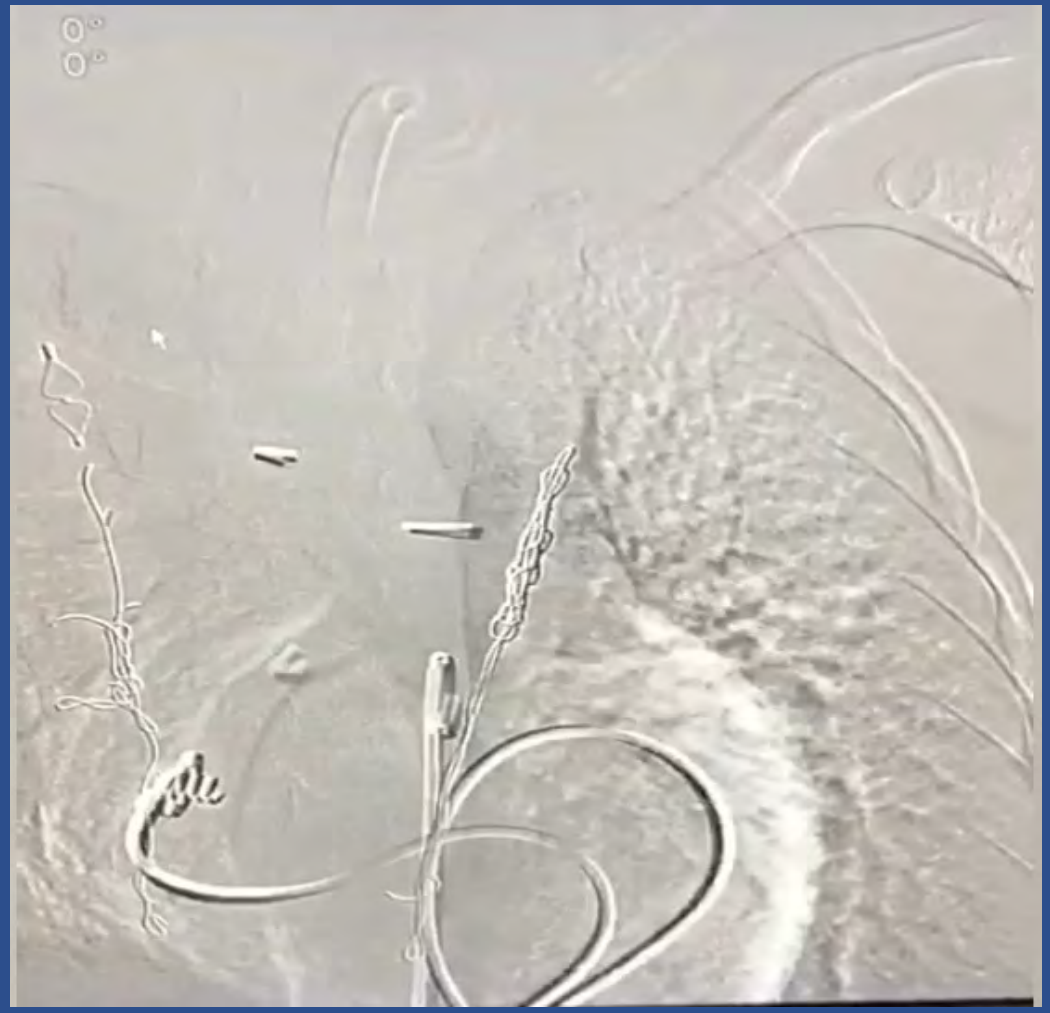
TD drainage to a Low pressured atrium with Diastole and Inspiration.

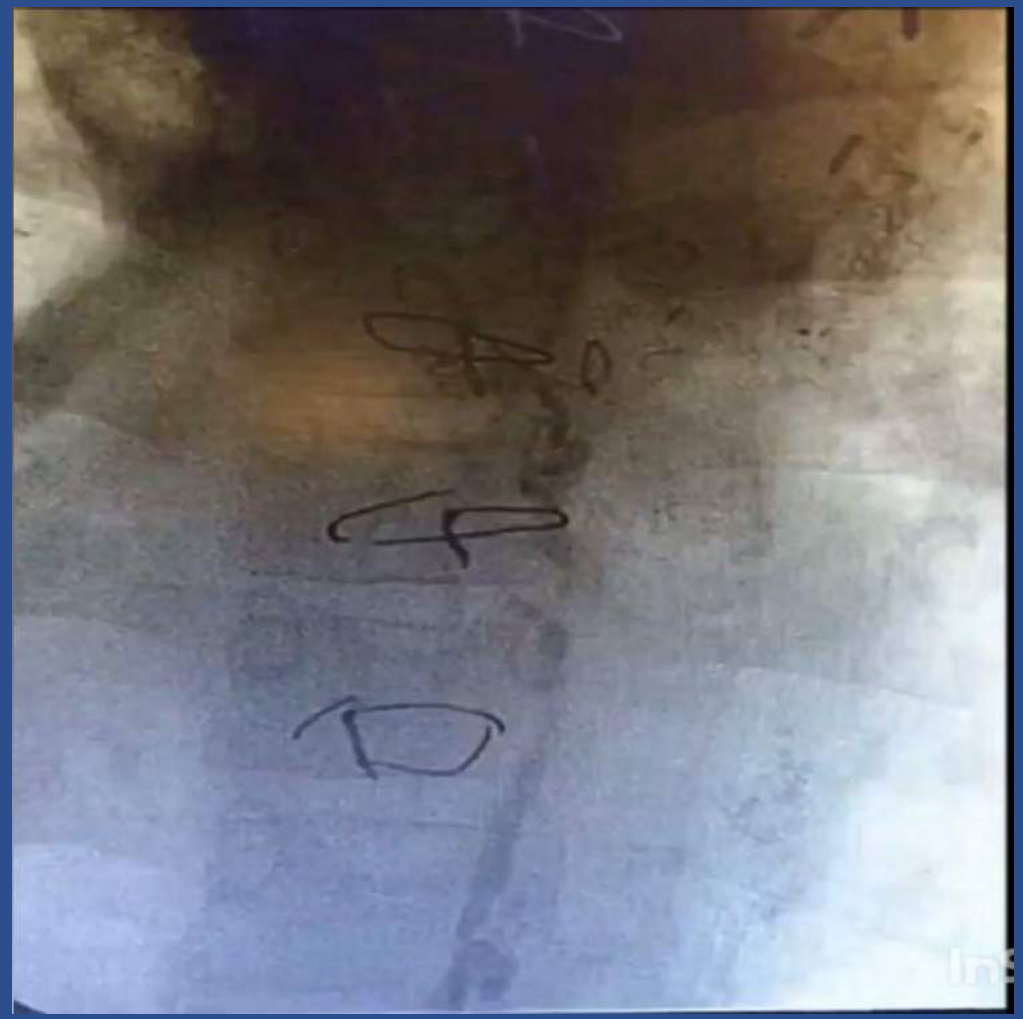
TD decompression: Indications & Techique



- Failing Fontan with PB/PLE/Effusions.
- Concomitant to Fontan procedure for high-risk patients (Prophylactic)
  - Thoracic Lymphangiectasia types 3 and 4.
- Early failure, Ascites and Hidrothorax.
- Inn Vein Turn Down: Hraska Procedure.
  - Direct Innominate vein w TD turn down to LAA
    - Anatomy of LAA
    - Wide patent anastomosis, low risk of thrombosis
  - Long distance between Inn Vein and LA?
    - Ringed PTFE graft
    - Dunked in LA cavity. (NOT IDEAL, AC)
  - diminutive LAA?
    - Ringed PTFE graft
    - Dunked in RAA. (NOT IDEAL, AC)

Hraska procedure Venous & Lymph Catheterization





Videos courtesy of Marcelo D. Rivarola & Alejandro R Peirone

Pre Fontan Completion MRI Lymphatic Screening

- Since 1/2017 to 1/2023, 37 pts were included in the Cohort.
  - Dx: HLHS (n=10), Heterotaxy Synd (n=10), DILV (n=5), TA (n=5), DORV (n=5), PA-IVS(n=1) Ebstein's Anomaly (n=1).
- MRI Analysis, CHOP Classification & Pathway:
  - Group A: Types I and II (n=26): "Classic" extracardiac Fenestrated Fontan
  - Group B: Types III and IV (n=11): Extracardiac Fontan with prophylactic Lymphatic Decompression
- 1 Early mortality in Group A.
- 1 inn Vein Turn Down Thrombosis & occlusion.
- Less volume of effusions in LD (group B). *p=.03*

Early Experience with Lymphatic Decompression Concomitant to Fontan Kreutzer Procedure. World J Pediatr Congenit Heart Surg . 2020 May;11(3):284-292

Lymphatic Decompression Concomitant to Fontan Kreutzer Procedure

- Follow up: (med 30 m).
- Patency of Inn Vein turn down assessed by Echo in 10 surv.
  - One partial occlusion
- No PLE or PB.
- One late death in LD group.
  - progressive right PV stenosis.
- No differences between groups
  - In survival
  - O_2 sat= Group A=92%, Group B= 91.4%
  - Functional status

Prophylactic Lymphatic decompression going bad

- **4 y/old:**
  - **HLHS, AA,MA.**
  - **Status post:(HUA)**
    - Norwood. (neonate)
    - Bidirectional Glenn @ 5 m. (Chylothorax)
- **@ 3 years of age. Fontan Completion.**
  - **Pre Fontan Cath. Normal PA pressure & EDP. Good PA anatomy. Multiple arterial embolizations.**
  - **Pre Fontan T2 MRI: Type IV abnormalities.**
  - **Fontan Completion with Inn Vein turn down**
  - **Uneventful procedure, extubated in the OR.**
  - **Chest tubes removed @ PO day #4. Sats in the low 90's.**

Prophylactic Lymphatic decompression going bad

- **But…**
- **6 # PO day, Sats in high 90's. 98%, shortness of breath.**
- **Increasing bilateral Effusions. (Left chest predominance)**
  - **POD 6, 180 ml**
  - **POD 7, 450 ml.**
  - **POD 8, 780 ml.**
  - **Left Cervical venous ingurgitation**
  - **Catheterization:**
    - Normal Fontan Pathway.
    - Complete occlusion of Inn Vein.

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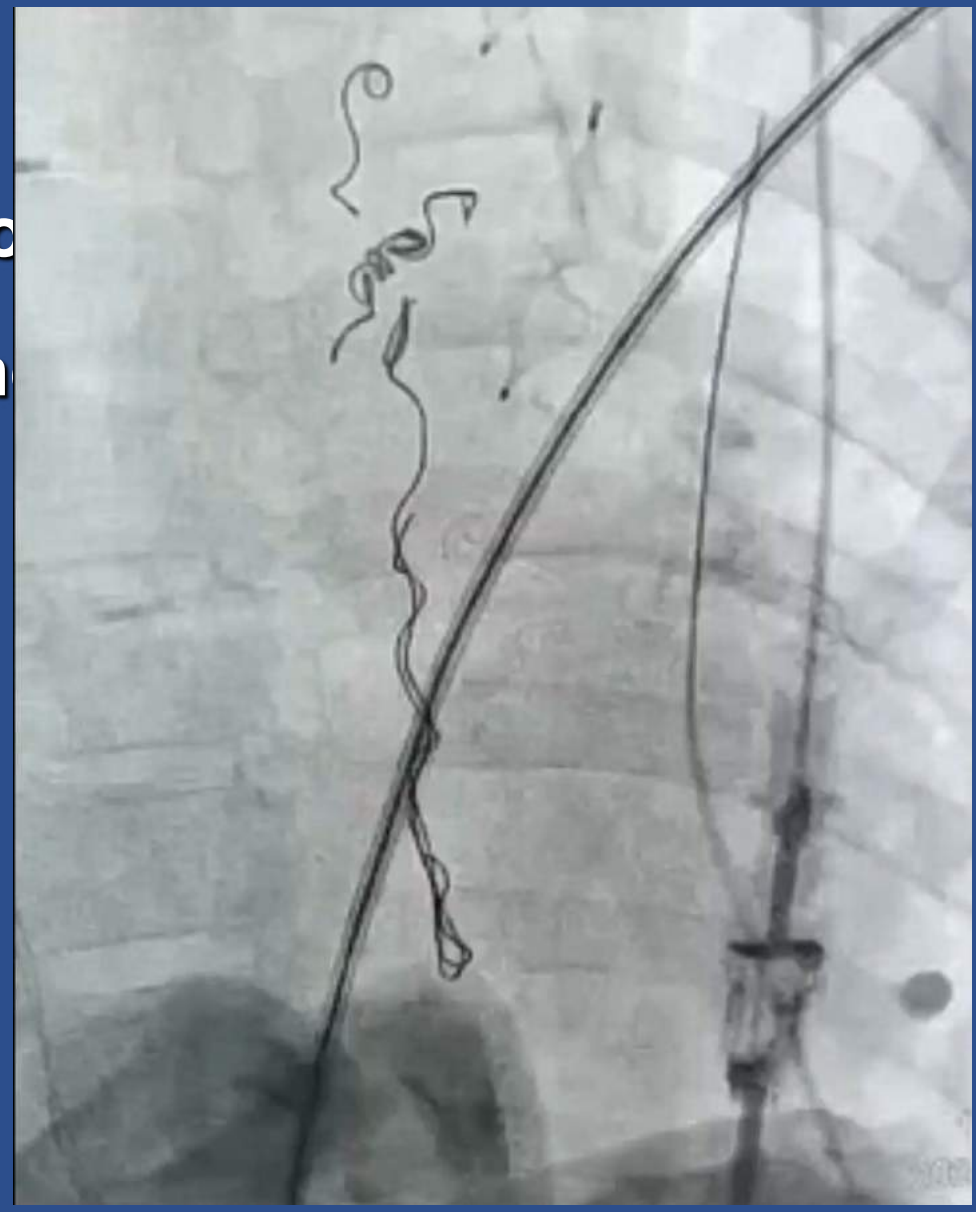
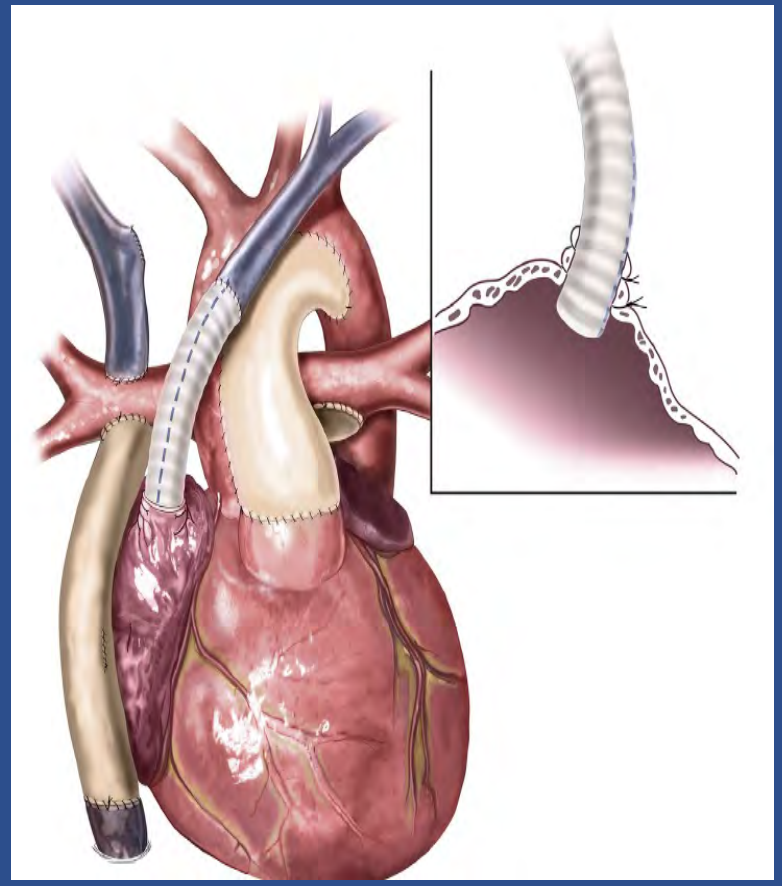


Prophylactic Lymphatic decompression going bad

- **4th Sternotomy**
  - **Inn Vein Turn down revision.**
  - **DHCA, LAA "neck"**
  - **PTFE 12 mm ringed conduit dunked in RAA.**
  - **Discharged POD 7th. Strict AC regime**
  - **Asymptomatic @1 year of F/up**
- **But…**
  - **CT Angio: Partial occlusion of PTFE conduit.**
  - **Ascending neo aorta Compression.**



Failing Fontan with PLE/PB/Effusions (n=9)

- Criteria for TD decompression surgery:
  - Preserved or mildly depressed Single Ventricle Fx (crucial) & normal EDP.
  - Patent Thoracic duct and upper systemic veins.
  - Ideally to the Left.
    - Right TD with a Right Glenn requires conduit interposition.
- Rationale: restore a normal Lymphatic drainage.
  - But… Once the leak is present, probably it wont stop after decompression.
    - For PLE: Duodenal pressure= -2/10 mmHg
    - For PB: Airway pressure is negative.
- Lymphatic Intervention:
  - Secondary to TD decompression.
    - Embolization of Lung Lymphatic Collaterals for PB
    - Embolization of Lymphatic collaterals for PLE.

Clinical experience in Failing Fontans: PLE

Age (yrs)	Weight (kg)	Diagnosis	Time since Fontan	Ascites	Effusions	Procedure	Outcome
5	15	Heterotaxy syndrome, asplenia, common AVVR,	2y	Yes, massive	Yes, Bilat	R Glenn take down, AVVr.	Normal Albumin Late death, acute Pulm Hemorrhage 6m f/up .
19	55	Heterotaxy Syndrome Asplenia	8 y	Yes	Yes	Hraska, RJSC–RA PTFE conduit	Alive 5 yr f/up, normal albumin
53	56	Tricuspid Atresia II	35 y	Yes, massive	No	Fontan conversion + Hraska	Normal Albumin. Late Death, 2 yrs f/up acute Hepatic Failure.
18	55	Mitral Atresia DORV	14 y	Yes	No	Hraska + ECC change	Initial Mild Improv. PLE relapse, Intervention X2, Partial Hraska Connection occlusion

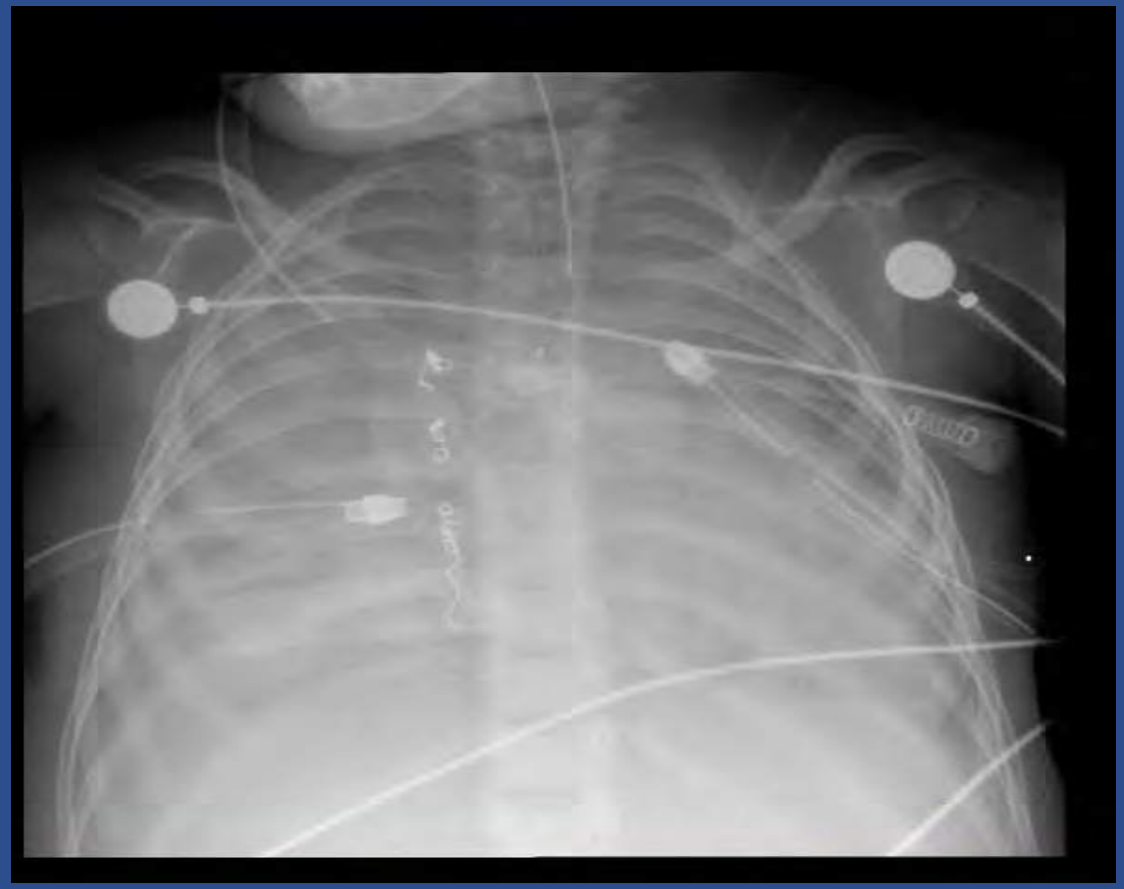
Clinical experience in Failing Fontans: PB & intractable effussions

Age (yrs)	Weight (kg)	Diagnosis	Time since Fontan	Plastic Bronc.	Ascites	Pleural effusions	Procedure	Outcome
6	21	PA IVS Stenotic BDG	2 y	Yes	No	Yes, right	Hraska + PA plasty	Alive. 2y No PB after Cath Intervention.
6	16	Heterotaxy síndrome	1,5 yrs	Yes	No	No	Hraska	Alive, 2 yr Asymptomatic
18	57	TA 1B ECC stenosis	14	No	Yes	no	Hraska + ECC change	Alive, asymptomatic
4	15	Heterotaxy syndrome, asplenia	2m	No	Yes	Yes, Bilat	Hraska, TAPVR correction.	Early death, Vent Dysfx
3	8	HLHS	4m	No	Yes, massive	Yes, Bilat	Hraska	Late death 2 yrs f/up, viral pneumonia

Case III

- 3 y/old. 4 m post Fontan (another Institution)
- Unbalanced AV Canal status post Norwood and Glenn
- 4 m after Fontan, Cachectic patient, 8 kg, ventilated.
- Fontan Failure with chronic ascites and Hidrothorax (600/800 ml/d)
- TD decompression
  - “Classic Hraska” Inn Vein Turn Down
  - Complete cessation of Effusions and Ascites @POD #4
- Discharged home on PO Day #27

Chest x-ray
Pre/Post Hraska 4th PO day



Take home message

- Prophylactive Lymphatic decompression is possible @ Fontan completion.
  - Promising results for patients with mod/severe Thoracic Lymphangectasia.
  - Preventing failure seems to be the game. Thrombosis & Occlusion!
- For early Fontan Lymphatic failure, it may have role.
- For late Lymphatic Failure: “First restore normal drainage & phisiology, then intervention”, (Normal Vent Fx) Thrombosis & Occlusion!
- Lymphatic intervention has a key role.
  - Dx and Understanding the unique physiology of the Lymphatic circulation.
  - Intervention for PLE, PB, Chylothorax, etc.

Questions?
Comments?

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