Fontan with protein loosing enteropathy

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Martin, born 1995

- Aortic annulus and arch hypoplasia, VSD, apex-forming LV

- team opted for single ventricle palliation
- 1995 Norwood st I., 4 mm MBT
- 1997 BDG
- 2000 Fontan with lateral tunnel, fenestration 4 mm
- 2005 spontaneous closure of fenestration
- **since 2009 (14-y.-old) protein loosing enteropathy (oedema, low albumin)**
- 2014 stent into Ao isthmus (19 → 3 mmHg gradient)

Cath study 9/2020
(PLE progression)

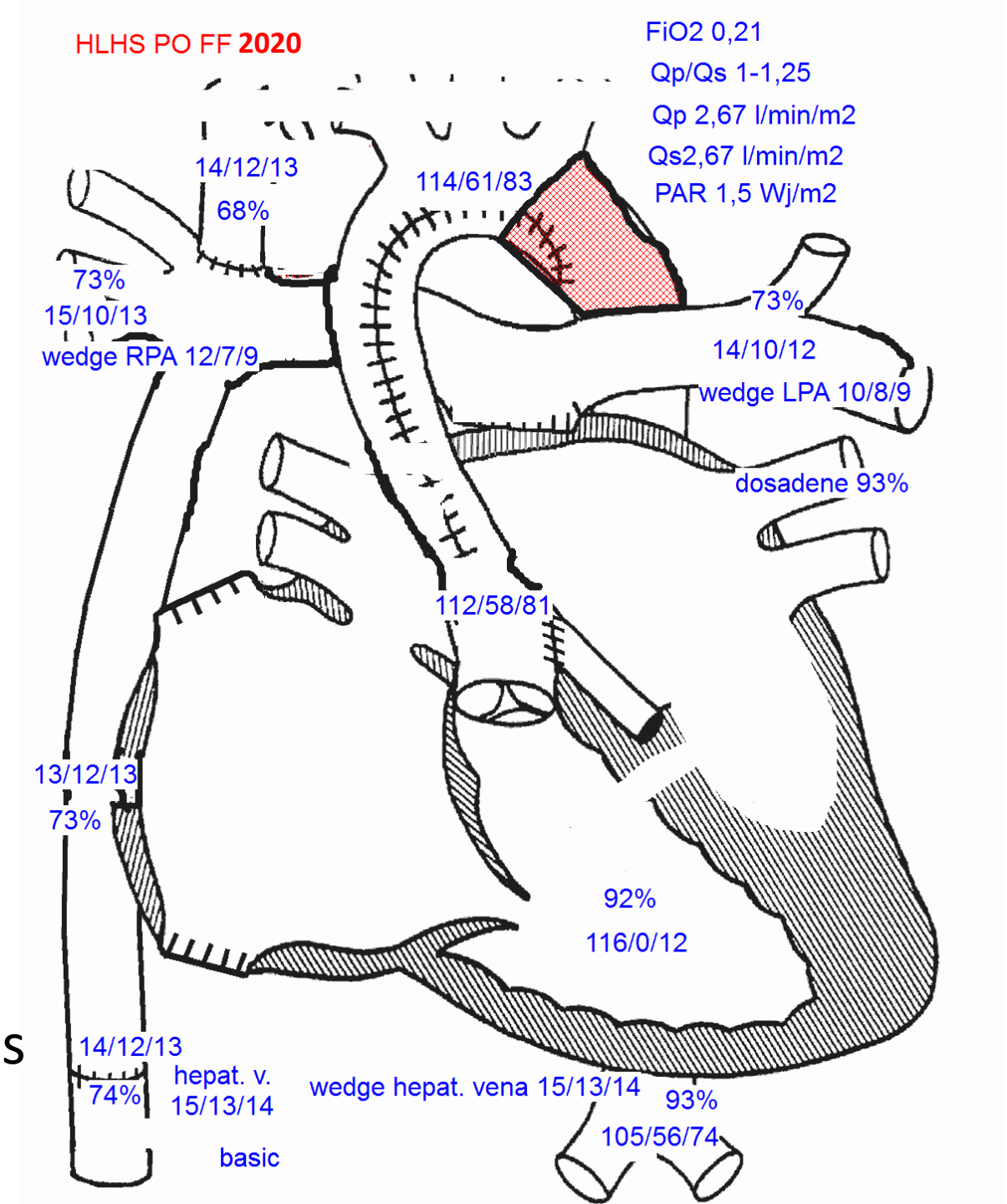
Fontan, PA´s 12-13 mmHg

TPG 4 mmHg

CO & PVR normal

no fenestration

no signif. Ao-Pulm. collaterals



Medications

Table 1
Dosing and monitoring of common medications used in the treatment of PLE.

Medication	Dose	Monitoring
Furosemide or other loop diuretics	1 mg/kg IV or PO every 6–24 h When kidney function is normal, a 40 mg dose of furosemide is approximatey equal to 1 mg of bumetanide and 20 mg of torsemide.	Electrolytes, fluid status
Albumin (25%)	1 g/kg IV as needed, as often as every 4–6 h	Fluid status
Budesonide	Starting dose 6 mg–9 mg PO per day, typically divided three times daily..	Adrenal suppression, Cushingoid features, hypertension
Spironolactone[a]	1–4 mg/kg per day (Max 200 mg per day)	Hyperkalemia, gynecomastia
Sildenafil[b]	1 mg/kg (Max 20 mg per dose) PO three times daily	Flushing, priapism, hypotension
Midodrine	1.25 mg–10 mg PO two to three times daily	Hypertension
Dopamine	Short term infusion at 3–5 mcg/kg/min	Arrhythmia and hemodynamics
Heparin (unfractionated)	3000–5000 unit/m^2/day subcutaneous	Bleeding and osteopenia
Octreotide	1-4 mcg/kg subcutaneous every 1–2 days	Gastrointestinal side effect and musculoskeletal pain

2009 as needed (also HCT) →

2009 infusions; also IVIG substitutions →

2022 – 1 month trial – no effect

2009 (eplerenone) →

2014 →

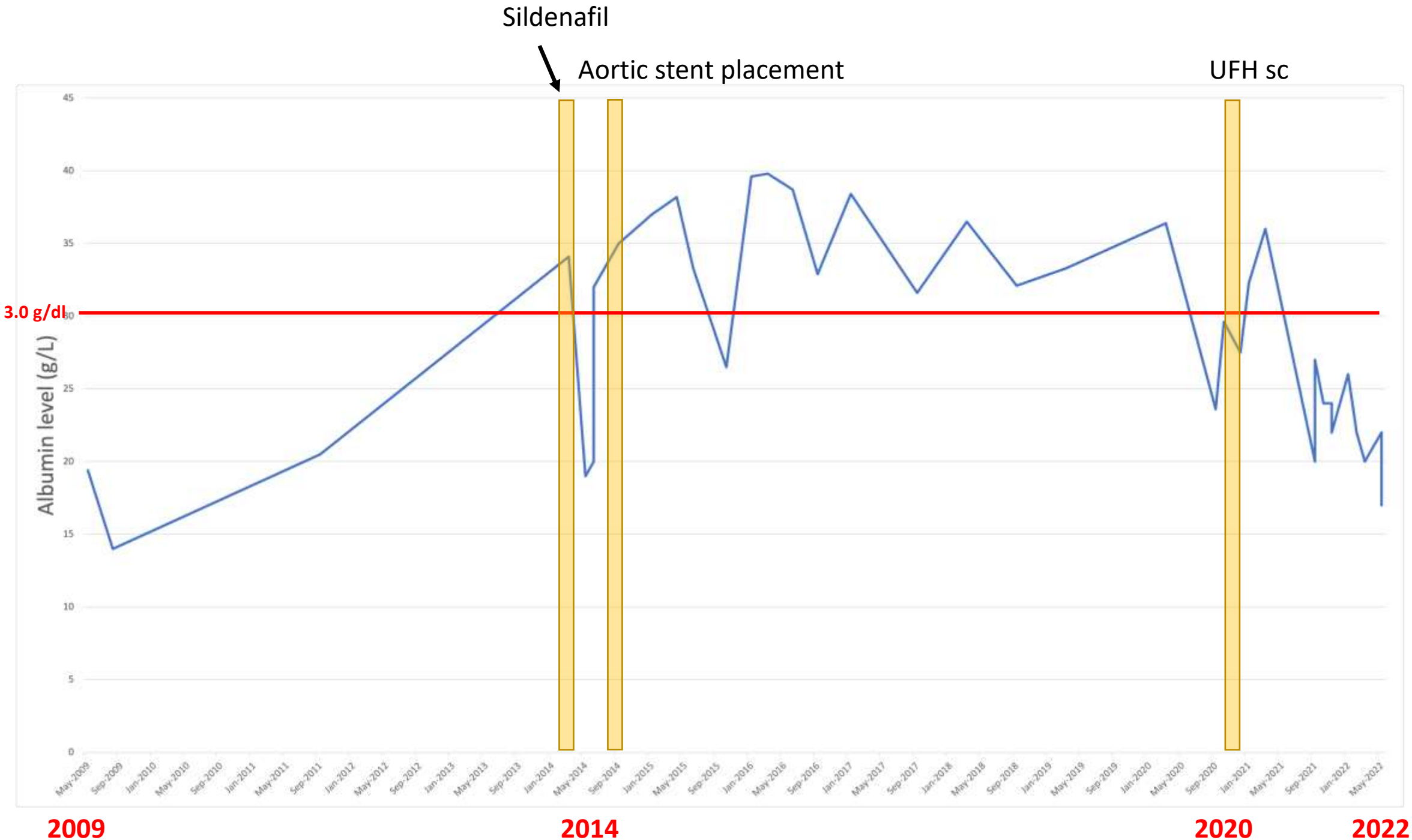
2021-22 trial – no effect

2020-21 trial – temporary effect

2009 → loperamid - for diarrhea

2009 → : ↓ fat, ↑ protein diet

Alsaied T et al., Int J Cardiol 2022


Sildenafil
Aortic stent placement
UFH sc
3.0 g/dl
Albumin level (g/L)
45
40
35
30
25
20
15
10
5
0
May-2009
Sep-2009
Jan-2010
May-2010
Sep-2010
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Sep-2011
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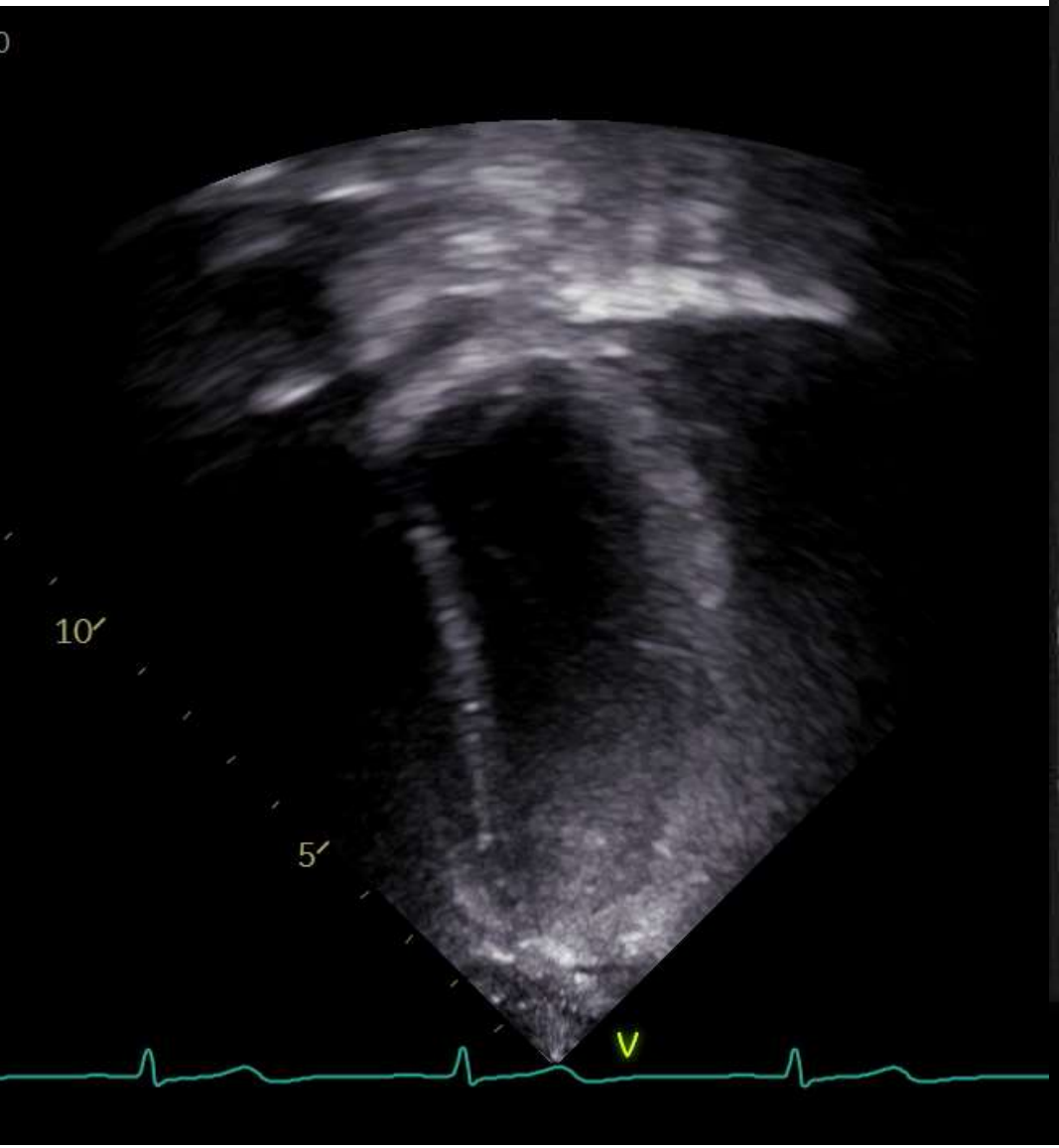
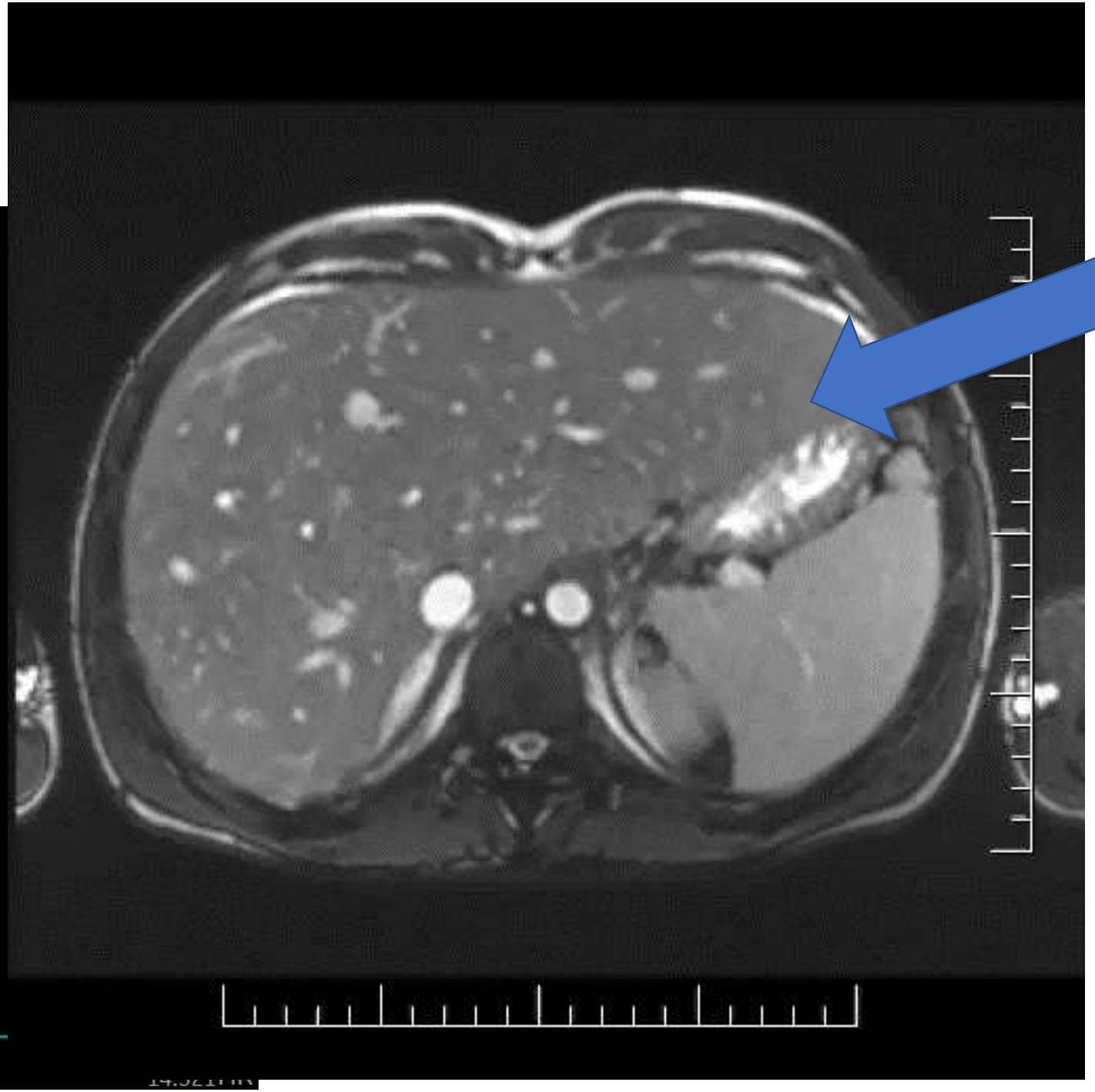
May-2021
Sep-2021
Jan-2022
May-2022
2009
2014
2020
2022

LV apex-forming

LV & RV systolic function preserved





2022 – 27 y. old man after Fontan

- Multicompartment lymphatic failure

(oedema, diarrhea, pleural effusions, ascites) → ↑diuretics, ↓ QoL

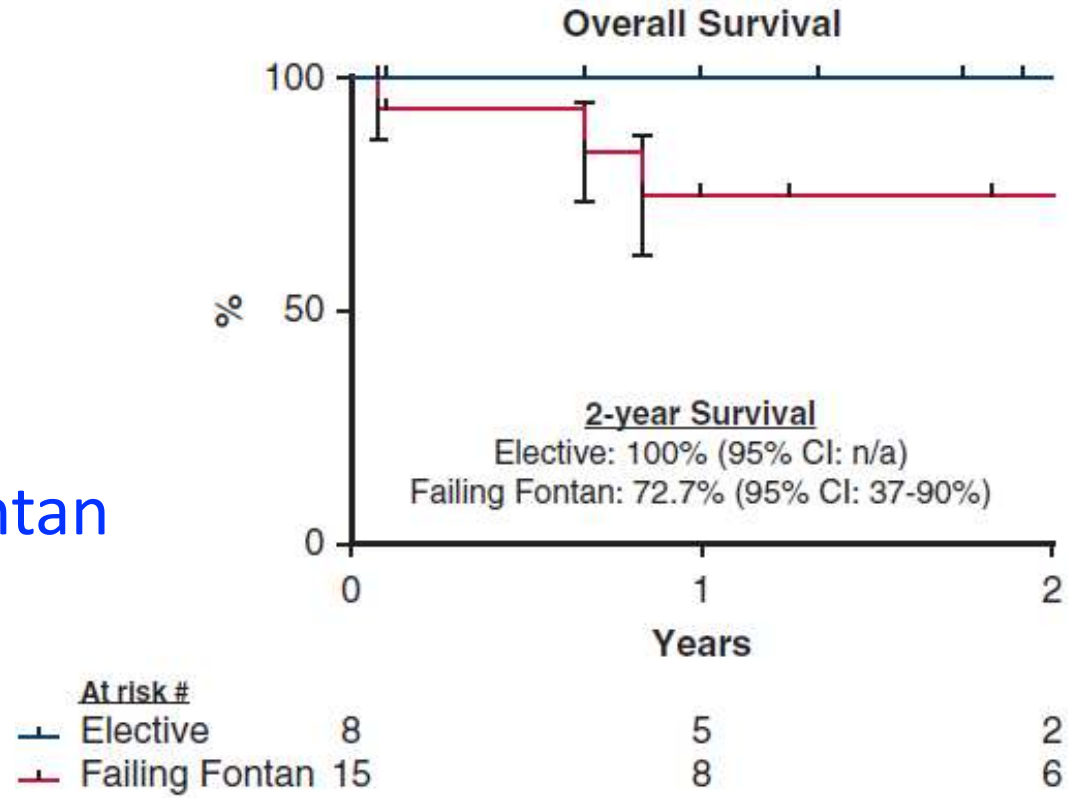
- Fenestration creation
- Biventricular conversion
- Heart transplantation
- Lymphatic procedure

What are the treatment options ??

Biventricular conversion after Fontan completion: A preliminary experience

Ilias P. Doulamis, MD, PhD,[a] Supreet P. Marathe, MD,[a] Breanna Piekarski, MPH,[a] Rebecca S. Beroukhim, MD,[b] Gerald R. Marx, MD,[b] Pedro J. del Nido, MD,[a] and Sitaram M. Emani, MD[a]

- Boston experience – 23 children
- age 10 y. (7,5-13 y.)
- elective conversion superior to failing Fontan
- mean LVEDP 10 → 15 mmHg



Heart transplantation ?

- 12 US centers, 68 pts. listed, 52 pts. Htx
- median 14.2 y. (4.7-26.2 y.)
- 25% patients - PRA > 10%
- 17% positive retrospective cross-match

Fontan-associated protein-losing enteropathy and post–heart transplant outcomes: A multicenter study

Kurt R. Schumacher, MD, MS,[a] Sunkyung Yu, MS,[a] Ryan Butts, MD, MS,[b]

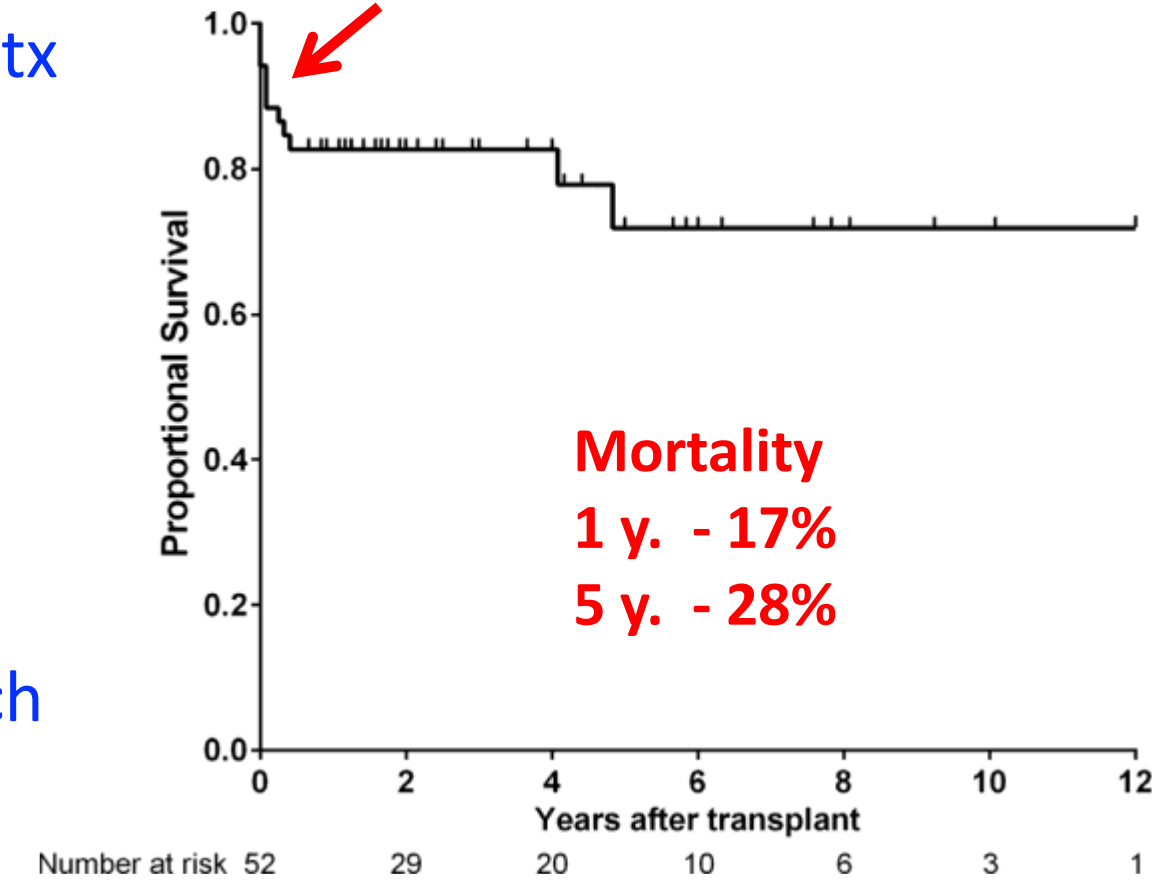


Figure 1 Post-HTx survival.

The Journal of Heart and Lung Transplantation, Vol 38, No 1, January 2019

HLA class II antibodies		
Date	Max. MFI	
16.9.2021	10 750,70	only DR
3.1.2022	10 298	only DR
7.2.2022	13 297,80	only DR
9.3.2022	12 075,99	only DR
12.4.2022	13 055	DR,DQ
5.5.2022	4 312,70	only DR

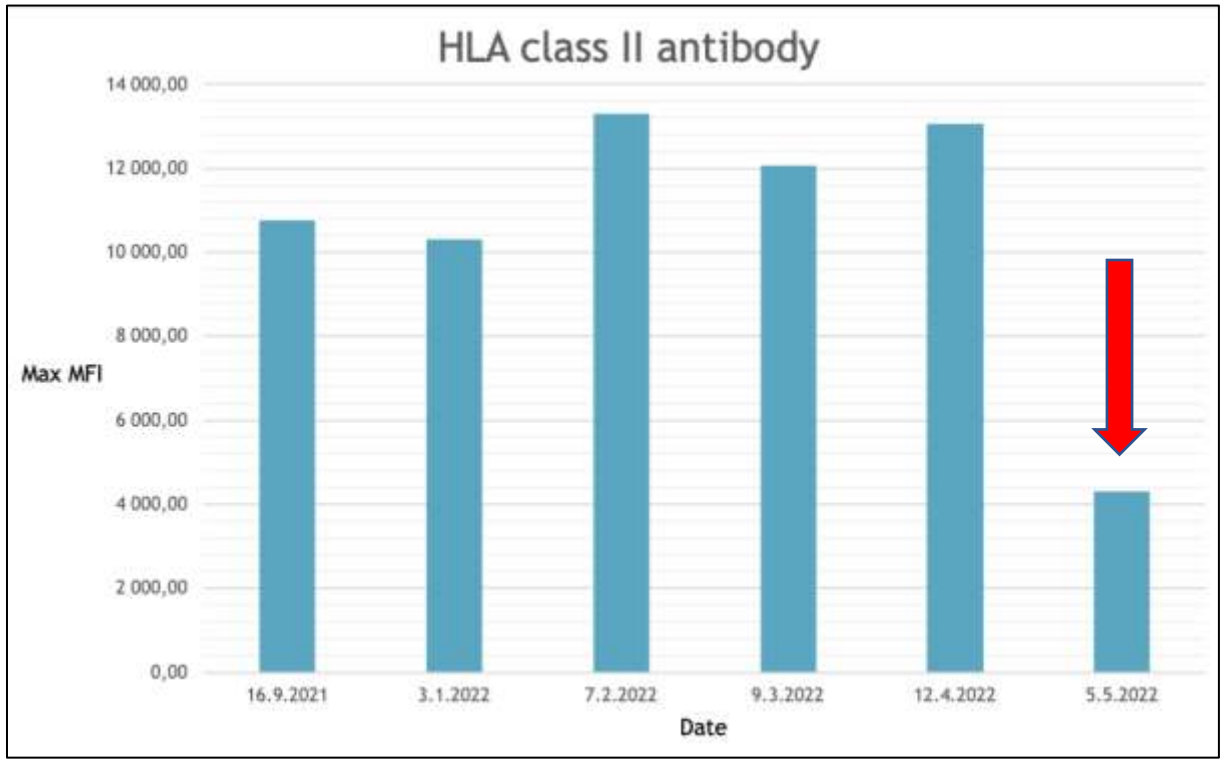
HLA class I antibodies		
Date	Max MFI	
16.9.2021	negative	
3.1.2022	negative	
7.2.2022	1409,3	A1
9.3.2022	1181,1	A1
12.4.2022	negative	
5.5.2022	negative	

PRA 2021 negat.

Martin, 27 y. – sensitized

6 months of desensitization therapy

- IVIG monthly + rituximab



Lymphatic intervention ?

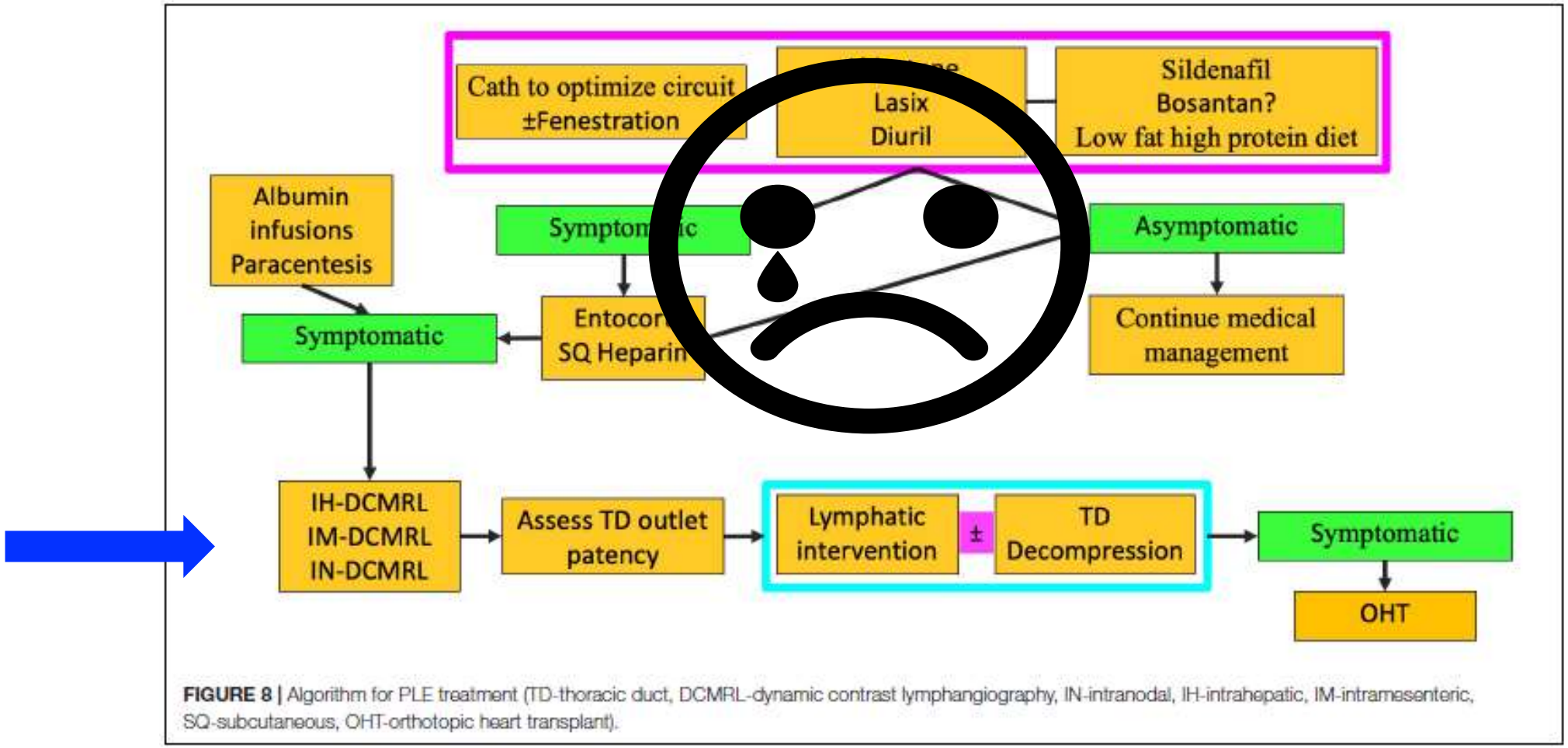


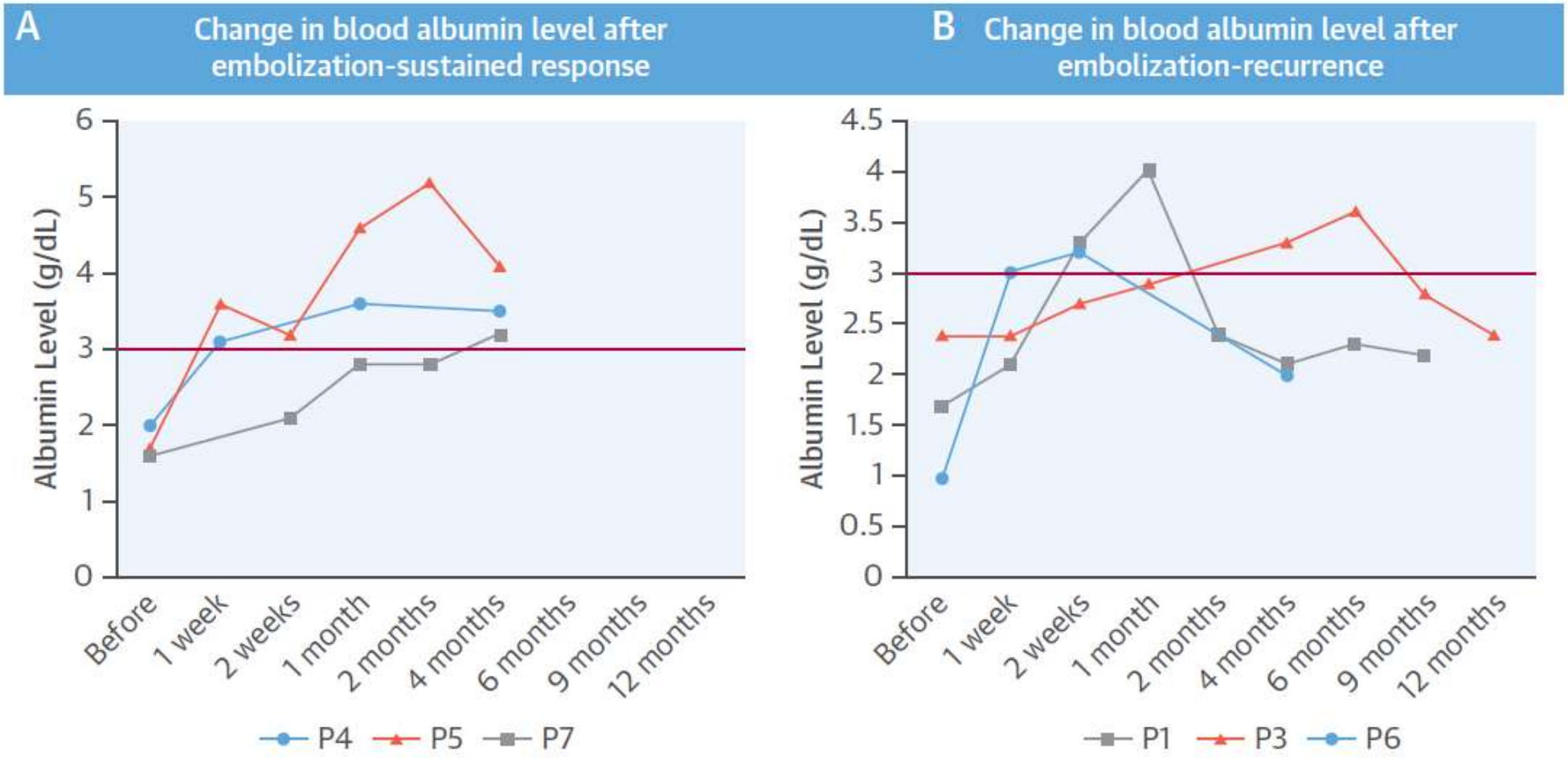
FIGURE 8 | Algorithm for PLE treatment (TD-thoracic duct, DCMRL-dynamic contrast lymphangiography, IN-intranodal, IH-intrahepatic, IM-intramesenteric, SQ-subcutaneous, OHT-orthotopic heart transplant).

1. Direct lymphatic interventions – closing the "channels"

2. Anatomic interventions targeting the increased lymphatic pressures/afterload

Dori Y and Smith CL (2022) Lymphatic Disorders in Patients With Single Ventricle Heart Disease. Front. Pediatr. 10:828107.

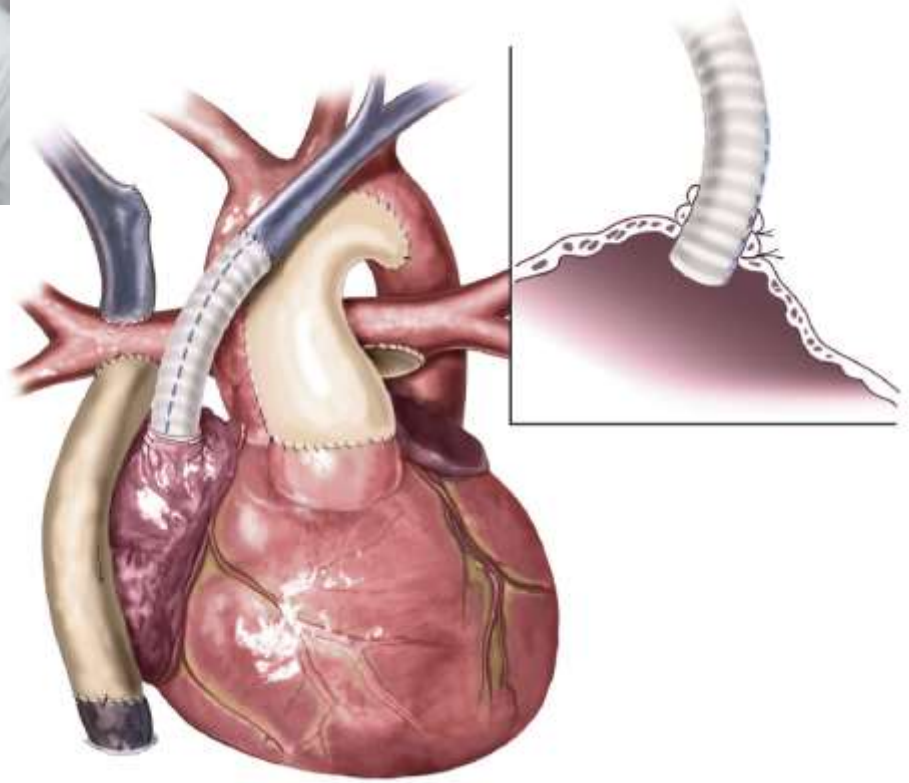
Hepatic and periduodenal Lymphatic Embolization

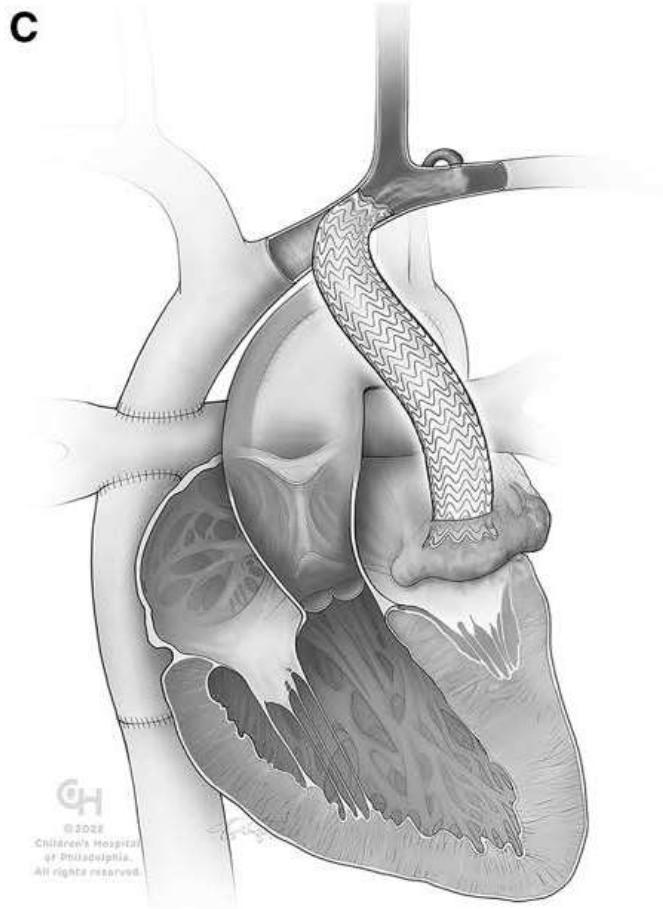


Itkin, M. et al. J Am Coll Cardiol. 2017;69(24):2929-37.

Symptomatic th. → recurrences

Thoracic duct decompression





Innominate vein turn-down, Hraska V, Ann Thorac Surg 2013

Transcatheter TDD, Smith C. Circ Cardiovasc Interv 2022

Thoracic duct decompression
Results

Innominate vein turn-down procedure: Killing two birds with one stone

Viktor Hraska, MD, PhD,[a,b] Vibeke E. Hjortdal, MD, PhD,[c] Yoav Dori, MD, PhD,[d] and Christian Kreutzer, MD[e]

JTCVS Techniques • June 2021

Transcatheter Thoracic Duct Decompression for Multicompartment Lymphatic Failure After Fontan Palliation

Christopher L. Smith, MD, PhD; Yoav Dori, MD, PhD; Michael L. O'Byrne, MD, MSCE; Andrew C. Glatz, MD, MSCE; Matthew J. Gillespie, MD; Jonathan J. Rome, MD

Circ Cardiovasc Interv. 2022;15:e011733.

Surgery

- 6/12 critically ill patients with PLE
- no procedural mortality
- Follow-up 0-36 months
- 50% no residual symptoms of PLE
- 50% died during f-up

Catheterization

- 10/12 frail patients with PLE
- no procedural mortality
- Follow-up 1-20 months
- 50% no symptoms of PLE
- 2 pts. improved, 3 pts not
- Albumin 2.5 → 3.8 (median)

SPECIAL ISSUE ARTICLE

WILEY Congenital Heart Disease

Congenital Heart Disease. 2017;1–6.

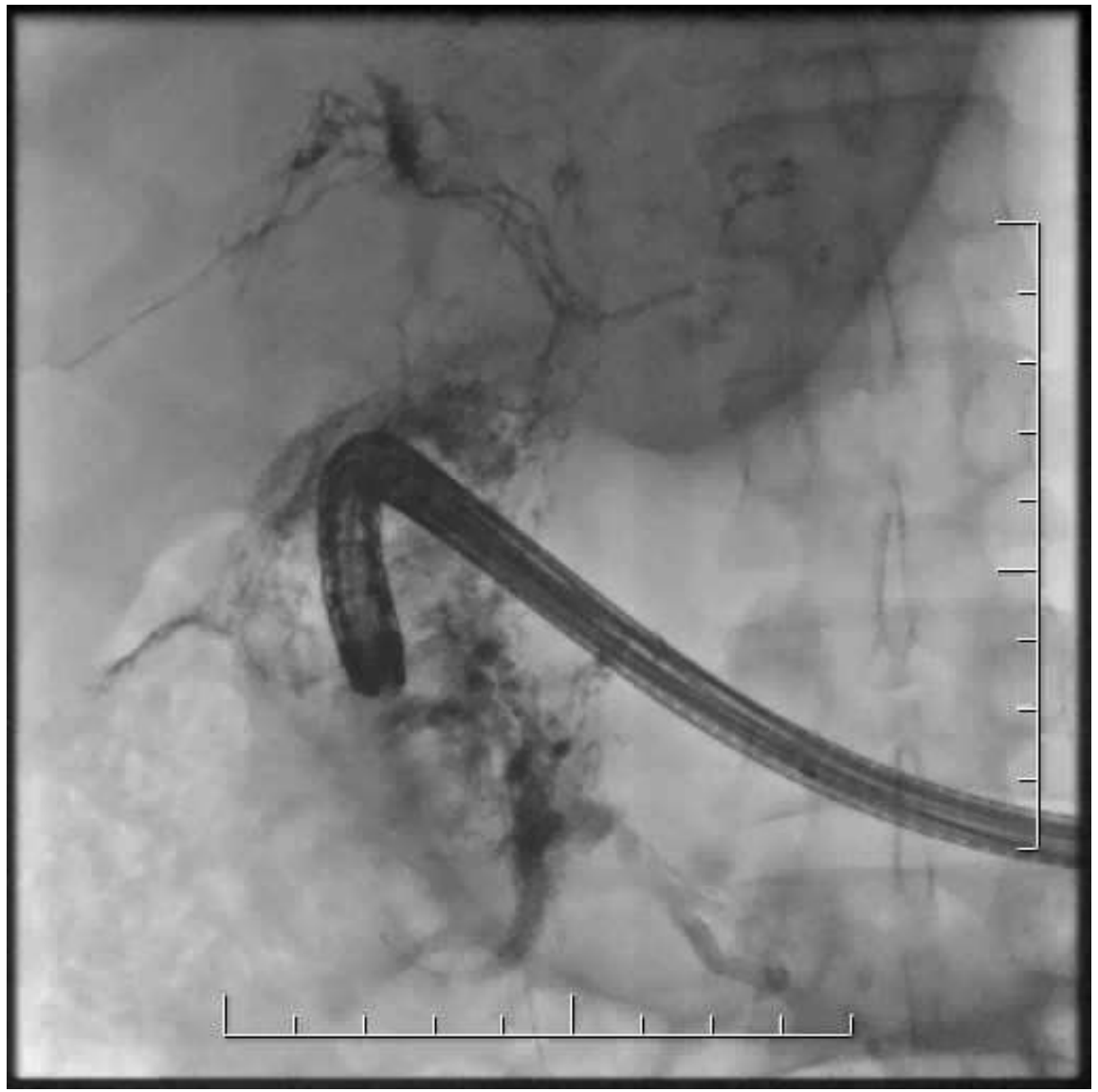
Transatlantic medical consultation and second opinion in pediatric cardiology has benefit past patient care: A case study in videoconferencing

Lubica Kovacikova, MD, PhD[1] | Martin Zahorec, MD, PhD[1] | Peter Skrak, MD, PhD[1] | Brian D. Hanna, MDCM, PhD[2] | R. Lee Vogel, MD[2]

- 54 teleconferences since 2013
- 5 AAF satellite symposia
- 6 professorships by CHOP experts

Courtesy of the patient

Step 1 Hepatic and periduodenal Lymphatic Embolization



Histoacryl glue embolization

2 channels in liver

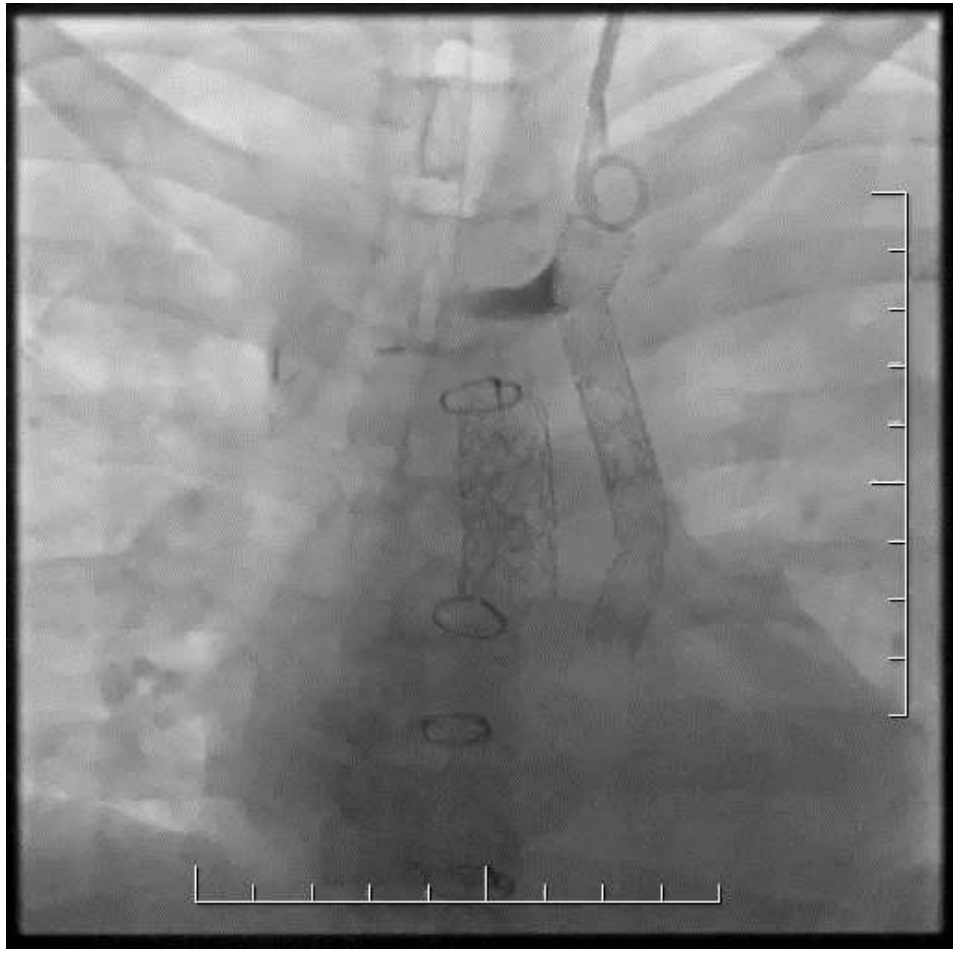
5 channels in periduodenum

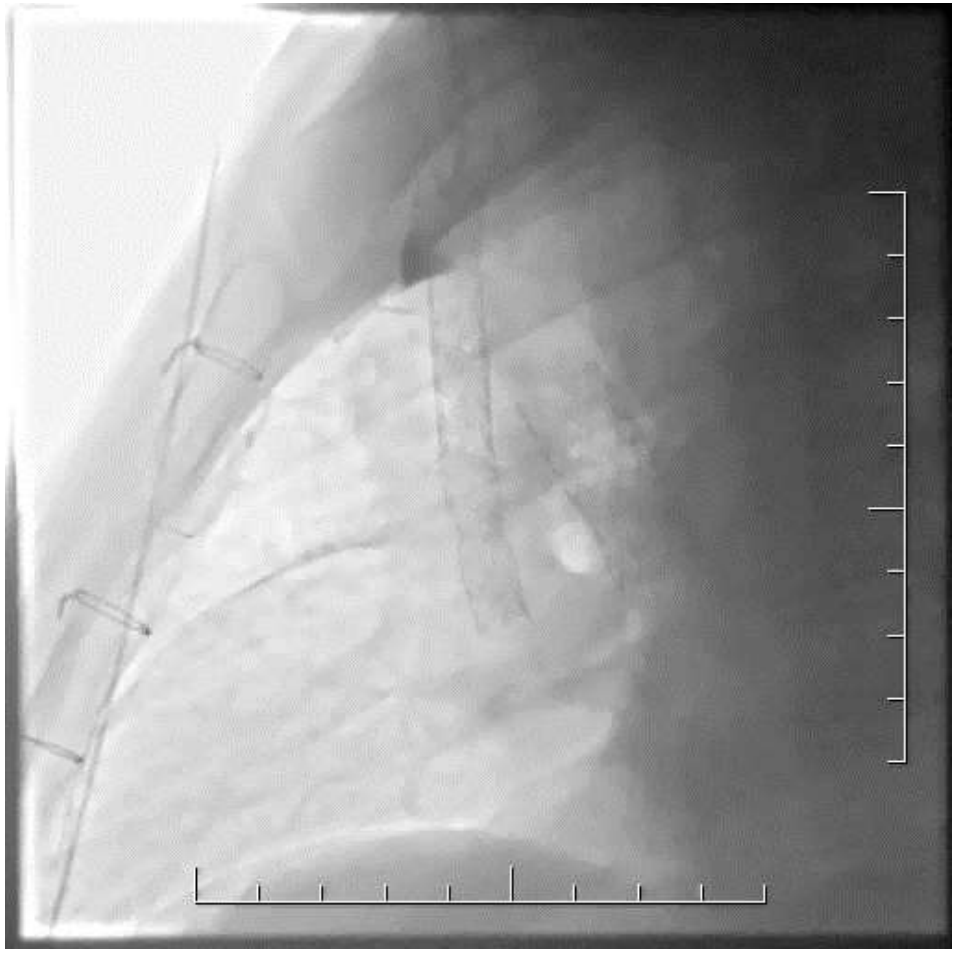
- transient laboratory pancreatitis

(Lipase: 8x increase)

- oedema & effusions progression

Step 2 - Thoracic duct decompression





2 stentgrafts 9 x 57mm implanted

Step 2: Thoracic duct decompression

- Day 6 after lymphatic embolization
- 7 days ICU, 11 days hospital stay
- Bivalirudine 24 h → Warfarin (INR 2-3)
- Albumin 2.3 → 3.4 (in a week)
- $satO_2$ 97% → 88-91%
- small residual ascites, no effusions

Courtesy of the patient

…not the happy-end yet

- 2 months post proc.: leg and arms oedema, satO_2 97%
- thrombosis of the stentgraft despite stable INR 2.5-3.0 during f-up

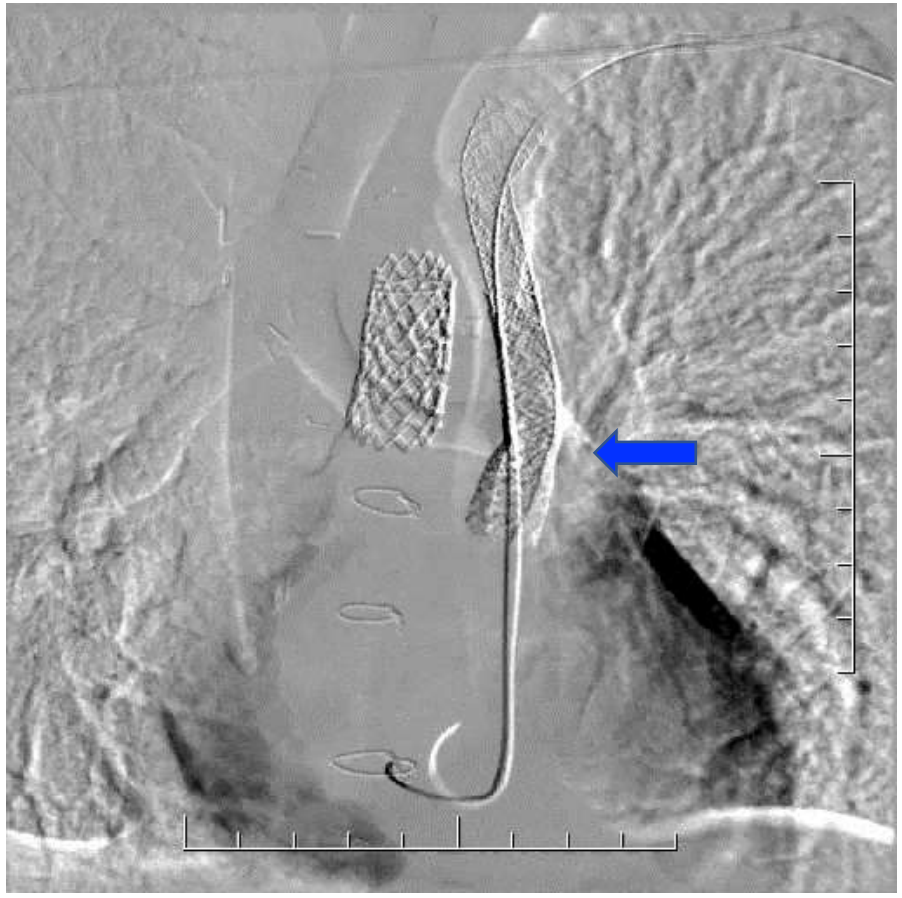
…not the happy-end yet

- 2 months post proc.: leg and arms oedema, satO_2 97%
- thrombosis of the stentgraft despite stable INR 2.5-3.0 during f-up

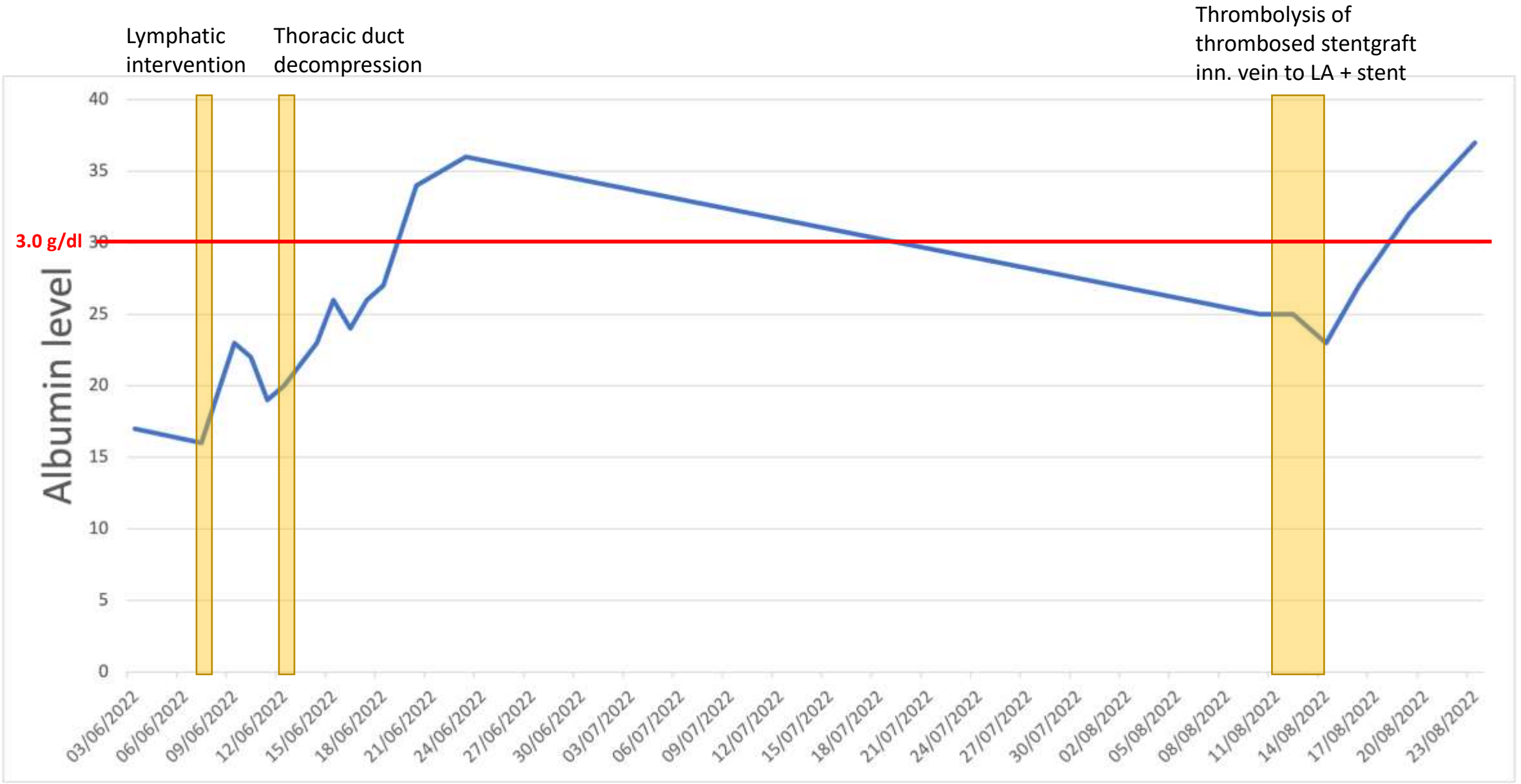
Site-directed thrombolysis

- Valved infusion catheter provides endhole occlusion
- low dose rt-PA 3 days with q 24 h angio – thrombus resolution
- stent to distal narrowing
- clinical improvement → Warfarin + clopidogrel

https://healthmanagement.org/

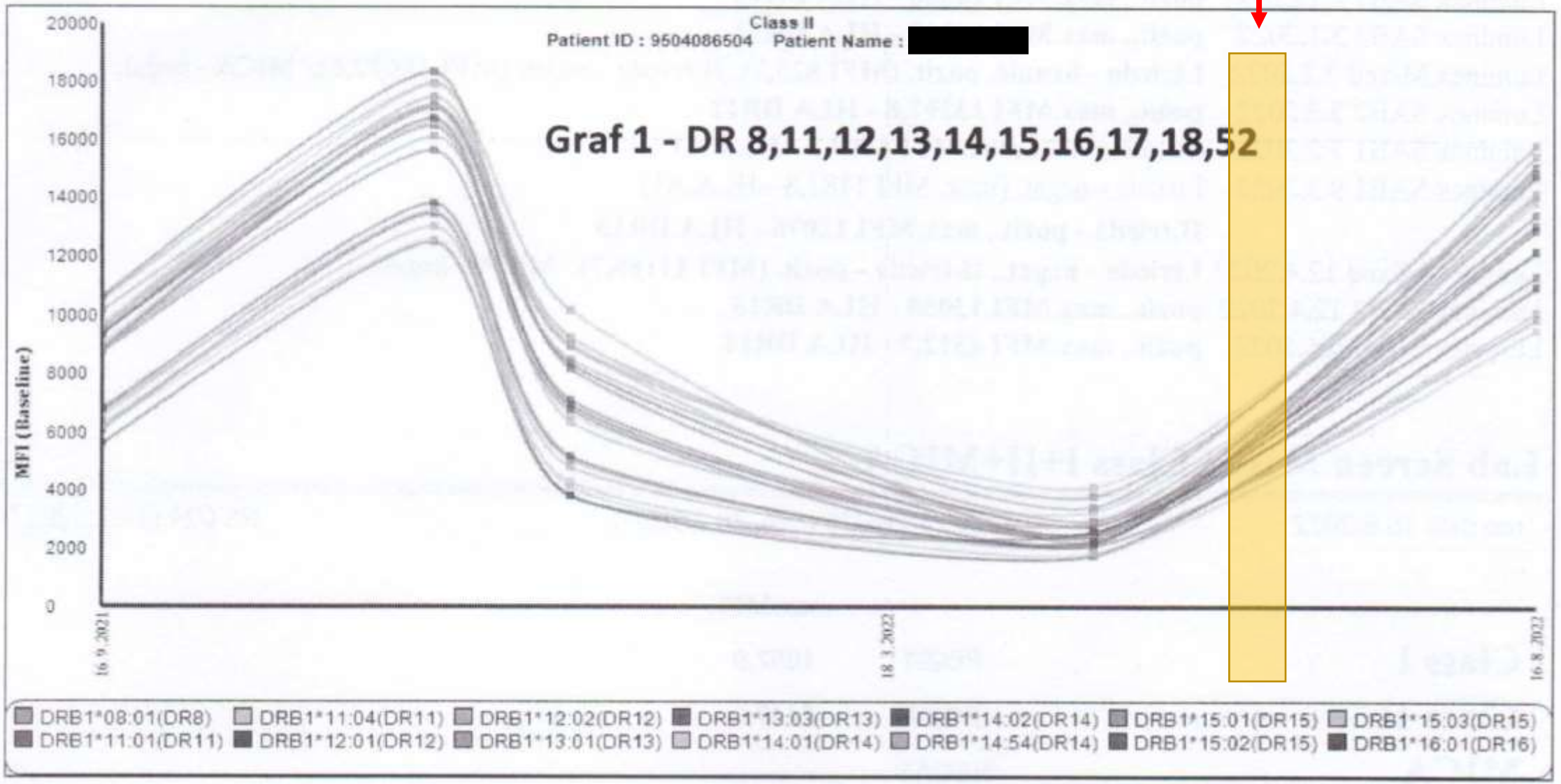


Trend of albumin blood levels



Sensitization status

Lymphatic intervention



Conclusions

- PLE - progressive Fontan complication
- PLE - complex decision making process
- Innovation - brings new options (lymphatic interventions)
- „Learning, sharing, networking“.... our responsibility to our patients
- Interdisciplinary & Interinstitutional (& International) collaboration inevitable to achieve improved outcomes (despite COVID, war etc...)

Thanks to

- CHOP – Cardiac Center & “Lymphomaniacs”
- American Austrian Foundation
- Fulbright Commission


DO NOT ENTER
IF YOU HAVE
SYMPTOMS OF
— COVID19
OR
IF YOU THINK
PUTIN IS COOL
NOLA
KITCHEN