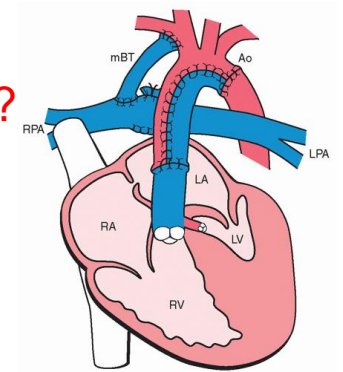
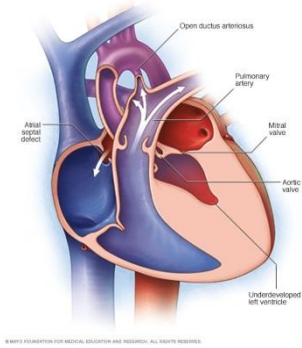




# Long-term survival in hypoplastic left heart syndrome

(How) Do the patients after St. I survive into adulthood?



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**Pediatric Cardiac Center**  
**National Institute for Cardiovascular Diseases**  
**Bratislava, Slovakia**



# Presentation goals

- What is known about survival of HLHS patients into adulthood?
- Bratislava results and experience
  - survival, physical health & complications
  - ~~– Mental health & HRQoL~~

## Thirty years and 1663 consecutive Norwood procedures: Has survival plateaued?

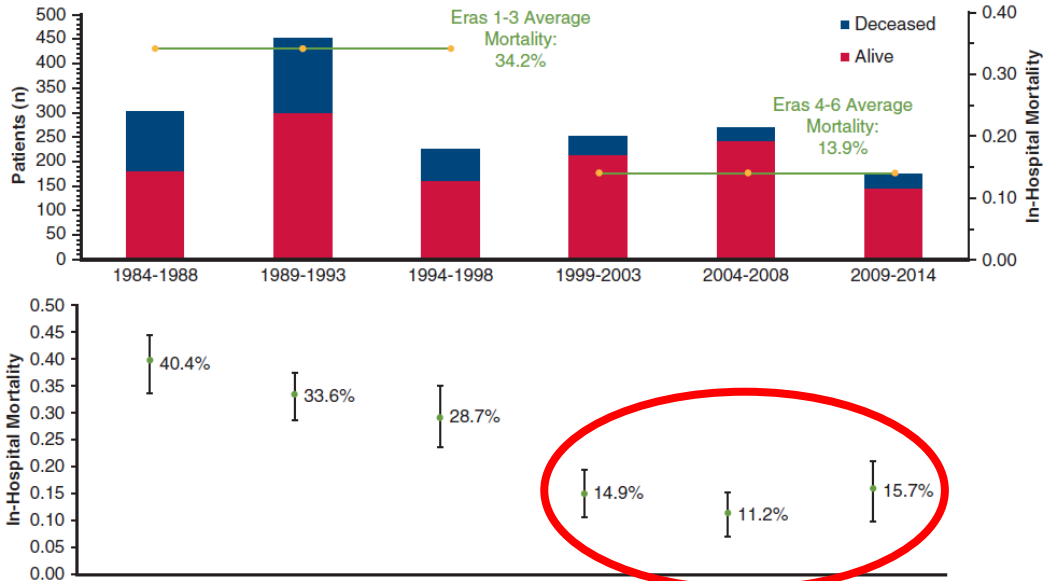
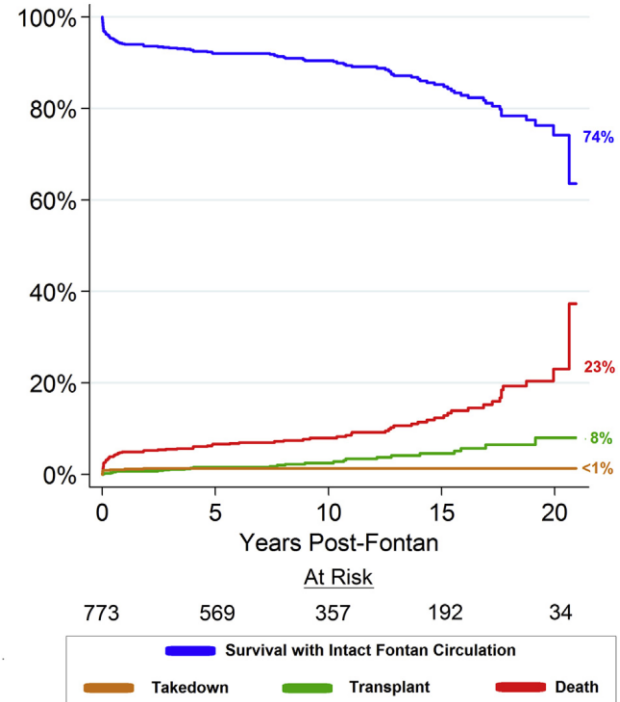


FIGURE 1. Hospital mortality has decreased after the Norwood procedure compared with earlier eras.

## Long-term survival after the Fontan operation: Twenty years of experience at a single center



# Bratislava - early results (1997-99)

## Not indicated for surgery:

- Prematurity
  - Non-cardiac congenital anomalies
  - Weight  $\leq 2500\text{g}$
  - Ascending Ao  $\leq 2\text{ mm}$
  - Severe TR
  - RV dysfunction
  - Failure to stabilize the circulation
- 
- 17 pts. not operated
  - 21 pts. Stage I + MBT

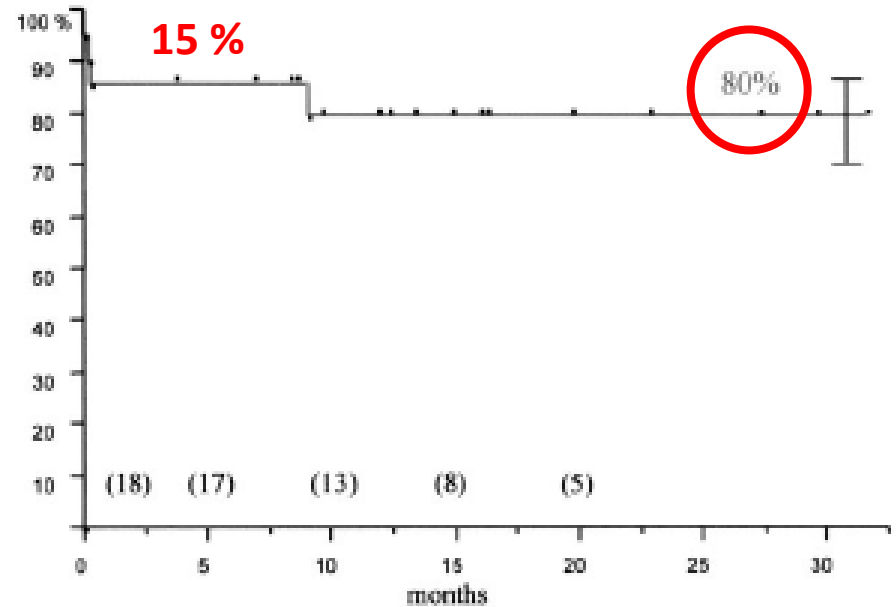
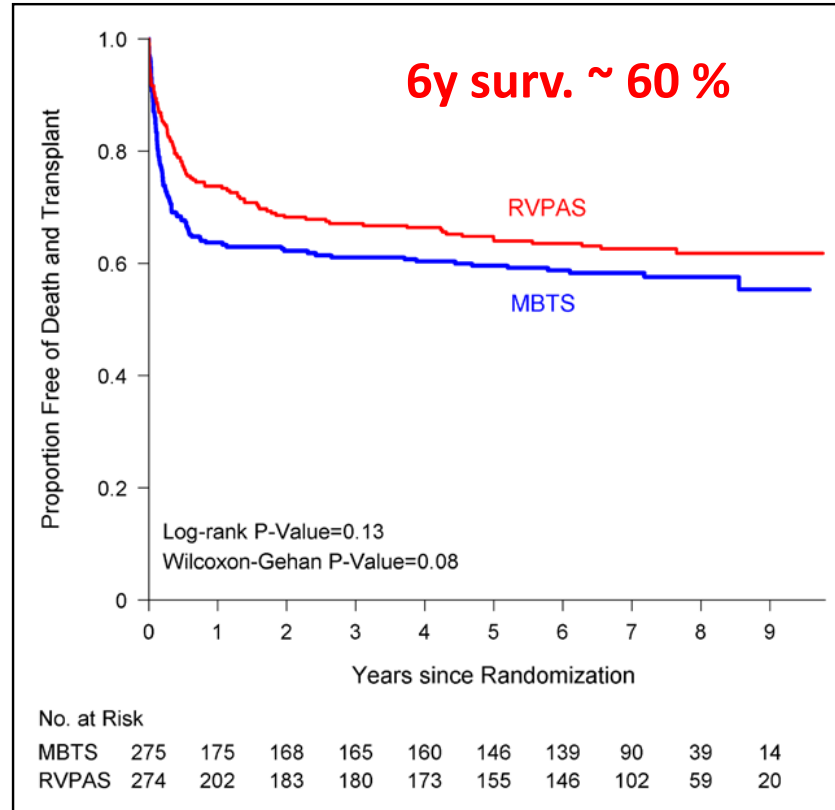


Fig. 4. Actuarial survival, in months, for the entire group of operated patients after stage I and II palliative surgery for hypoplastic left heart syndrome. Error bar indicates 70% confidence interval. Numbers of patients at risk are in parentheses.

# SVR trial (2005-2008 st.I)

- ~ 550 pts.



# Adults after Norwood procedure ?

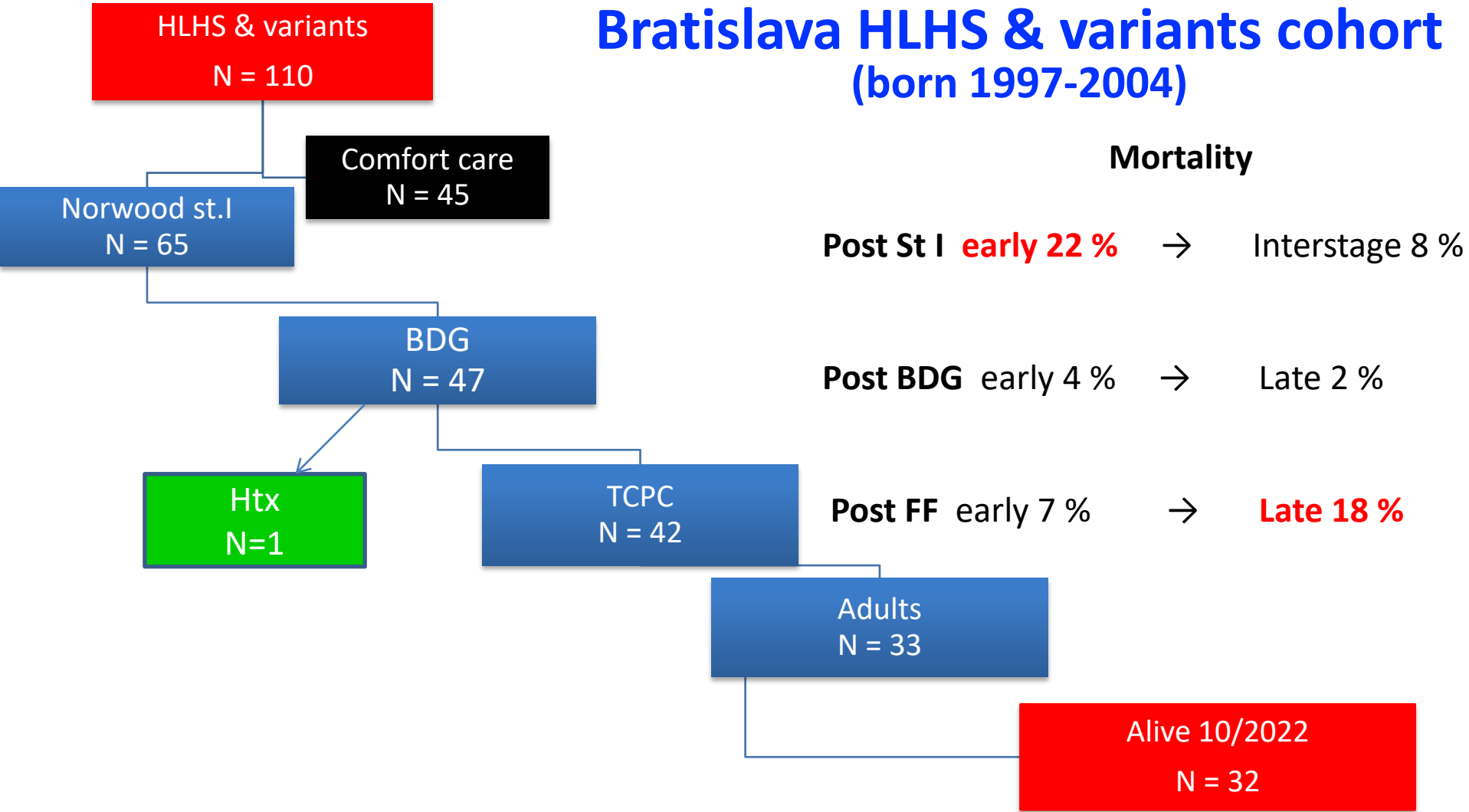
- **1980-90**: adulthood < 20 % pts.

*Feinstein, JACC 2012*

- **Born before 1996**: multicenter study (Toronto, Boston, Melbourne...)
  - Stage I (n=543): survival to Fontan 27 % pts.
  - **Transplant-free survival to adulthood: 14 % pts. (n=76)**
  - **24 % of adult pts. major complication** (death, Htx listing, admission for cardiac failure, PLE, VTE, VT) **during ~ 3,3 y. of follow-up** (age 21,6 y.)

*Wilson, Circulation 2018*

# Bratislava HLHS & variants cohort (born 1997-2004)

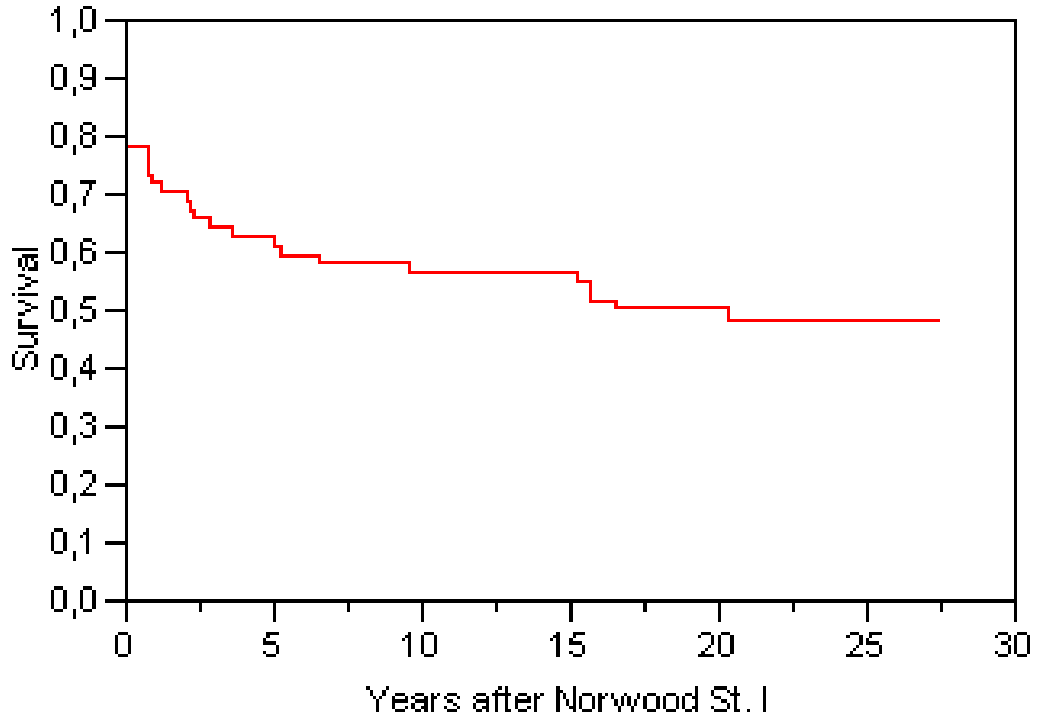




# Survival after Norwood St. I

## 1997-2004

- 30d. 78 %
  - 1y. 71 %
  - 6y. 58 %
  - 10y. 55 %
- 
- 18y. 51 %



Years	0	1	5	10	15	20	25
No. at risk	65	47	42	37	37	26	2

# Aortic stenosis vs. Aortic atresia

(1997 – 2004)

„Preselected“ pts. cohort

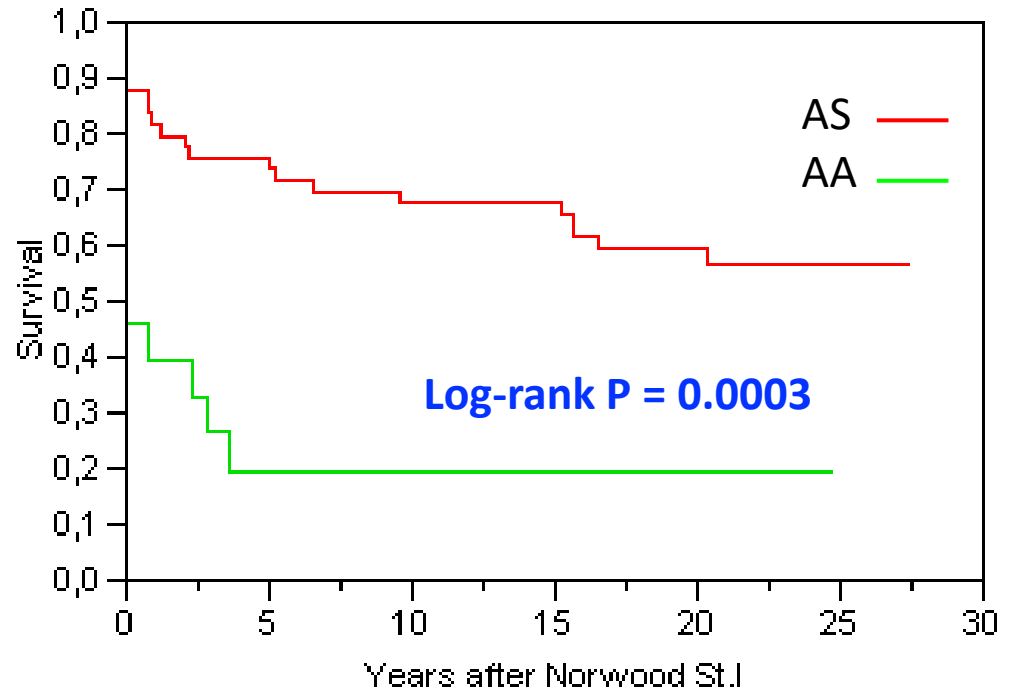
- **Ao Atresia - 23 %**  
(in SVR trial - 62 %)

1 y. survival

- **AA 33 % vs. AS 80 %**

20 y. survival

- **AA 20 % vs. AS 57 %**

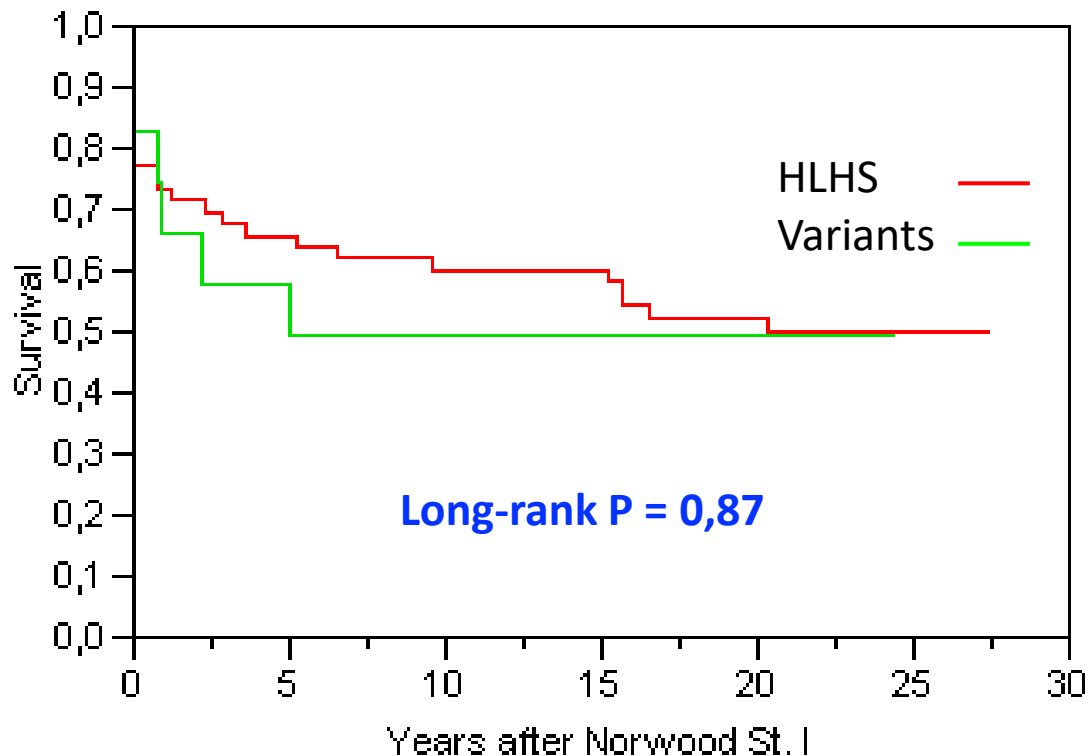


Years	0	1	5	10	15	20	25
AS	50	41	38	33	33	23	2
AA	15	6	3	3	3	3	0

# HLHS vs. Variants

## Variants of HLH (n=12, 18 %)

- hypo LV/AoV/AA+VSD (n=6)
- rdAVSDC (n=4)
- single RV+SVOTO (n=2)



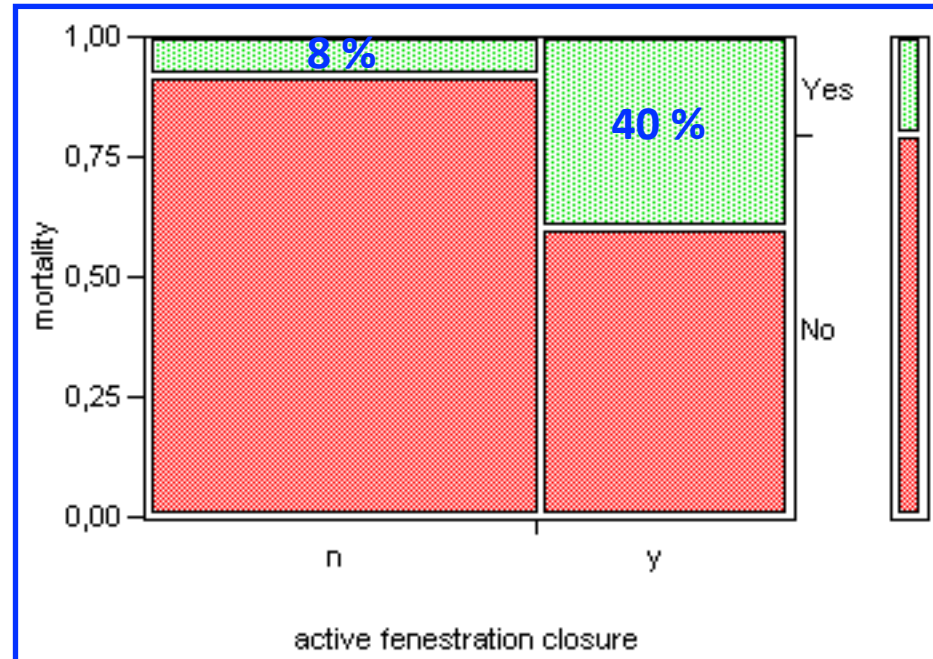
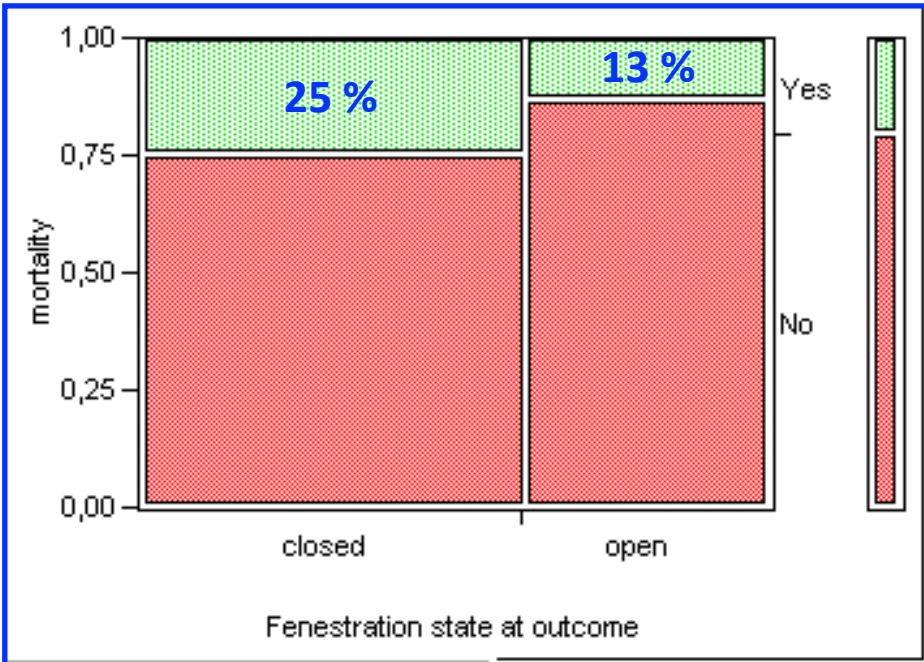
Years	0	1	5	10	15	20	25
HLHS	53	39	34	30	30	22	2
Variants	12	8	7	7	7	4	0

# Late post Fontan mortality

- 8/39 pts. (20 %)
  - Lymphatic failure, n = 3
  - Sudden cardiac death, n = 1
  - Heart failure, n = 1
  - Portal hypertension, n = 1
  - Hemoptysis, n = 1
  - Infection, n = 1

# HLHS - Fontan survivors (N = 39)

## Fenestration



• Mortality P = NS

P = 0.037

# Adult survivors after Norwood St. I.

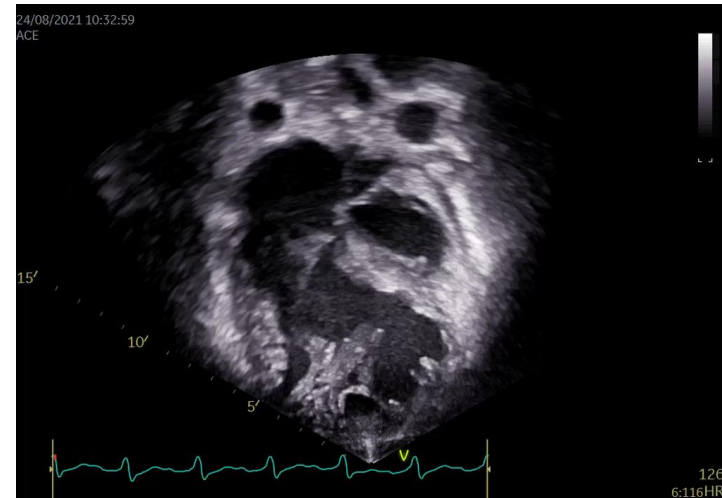
(N = 33; mean f-up 21 years)

## Fontan-specific major adverse cardiovascular events

- Thromboembolic event / stroke (24 %)
- Arrhythmia (19 %)
- Heart failure with RV/TV dysfunction (9 %)
- Hemoptysis (9 %)
- Lymphatic failure (9 %)
- **Free of any adverse event (45 %)**

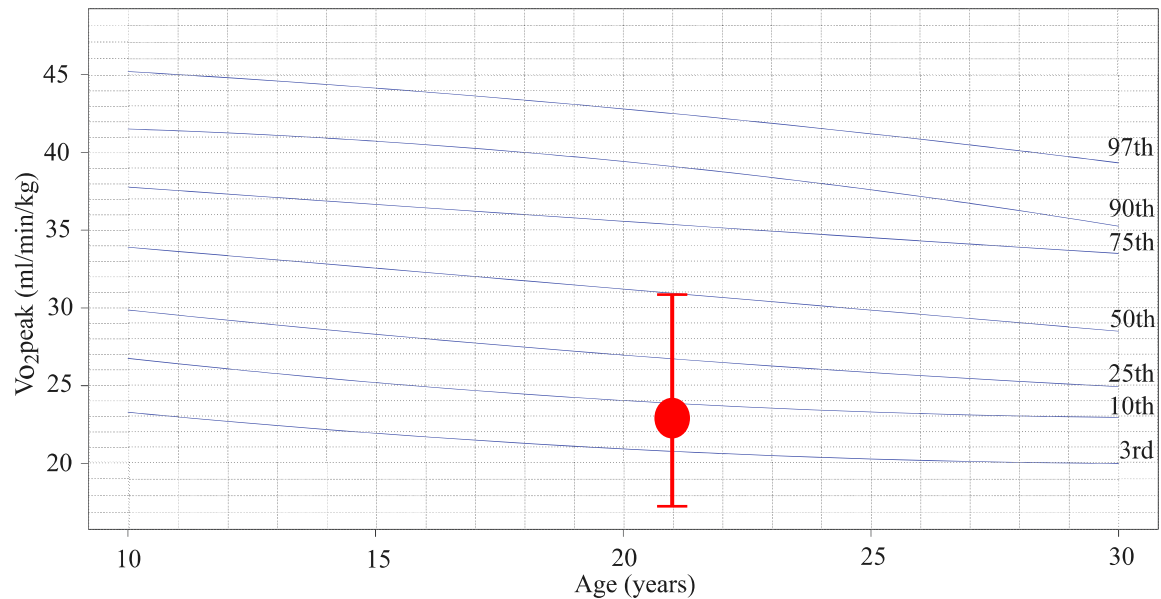
# Heart failure

- RV systolic dysfunction (MRI EF < 40 %) – 4/33
- TV failure (severe regurg.) – 1/33
- ↑ NT-proBNP (> 150 ng/l) – 10/33
- Exercise tolerance (CPET):
  - $VO_2$ max 22,1 ml/min/kg (16,5 – 30,1)
  - Max. Workload 1,5 W/kg (0,97-2,0)



# Fontan VO<sub>2</sub> max nomograms

Figure 1 VO<sub>2</sub>peak quantile regression in men with percentiles, the total number of tests (N = 434).



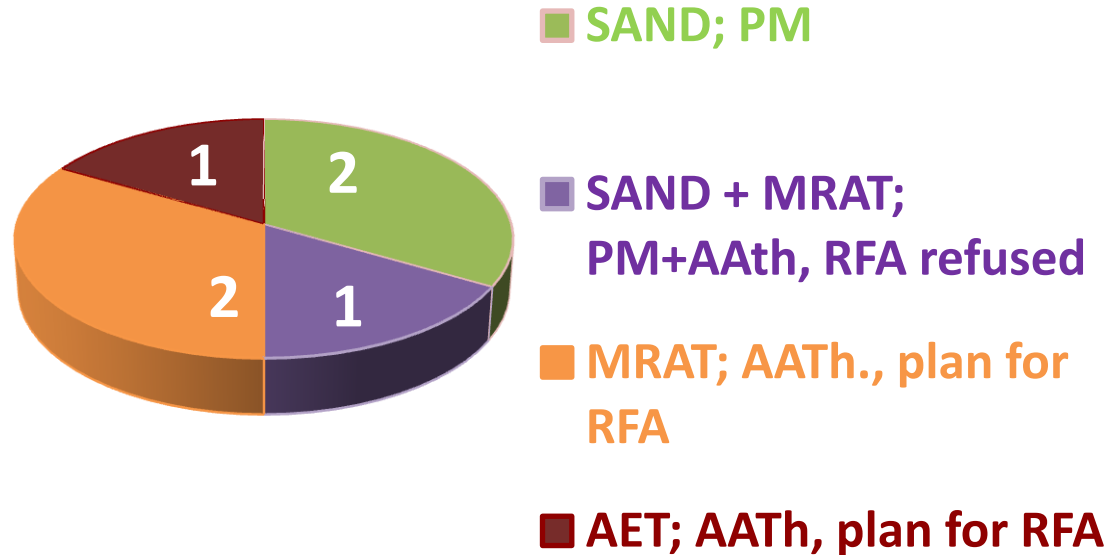


# Arrhythmias in adults after Norwood St.I

## None / Insignificant Rhythm Disturbance (26 pts.)

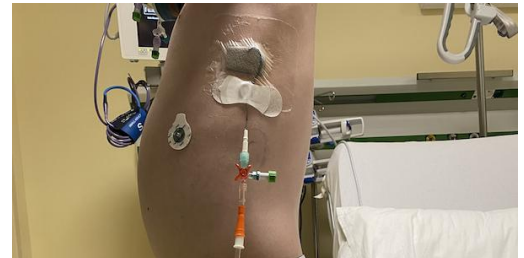
- SVES / PVC
- SAND w/o clinical relevance
- Non-sinus suprav. rhythm
- undefined SVT – single short run

## Clinically significant arrhythmia (6 pts. – 19 %)




# Lymphatic failure

- Protein losing enteropathy / multicompartiment lymphatic failure (3 pts)
  - 1 died (20 y.)
  - 1 lymphatic procedure + TDD
  - 1 Htx evaluation
- Plastic bronchitis (1 pt.)



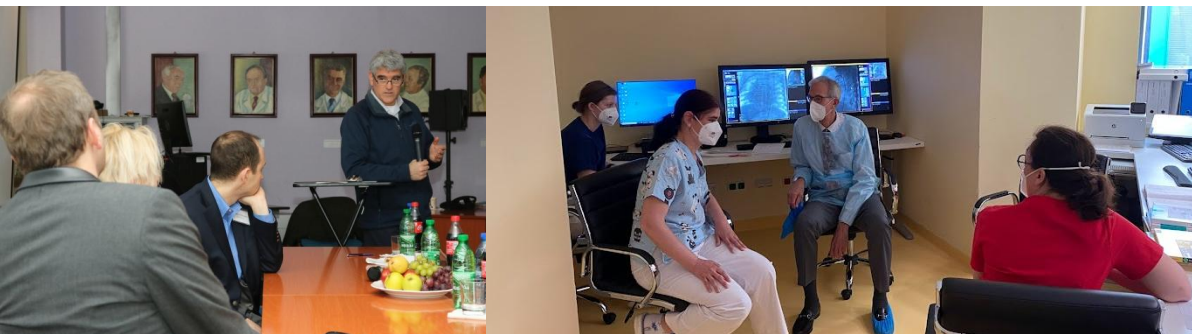
## Transatlantic medical consultation and second opinion in pediatric cardiology has benefit past patient care: A case study in videoconferencing

Lubica Kovacikova, MD, PhD<sup>1</sup>  | Martin Zahorec, MD, PhD<sup>1</sup> |  
Peter Skrak, MD, PhD<sup>1</sup> | Brian D. Hanna, MDCM, PhD<sup>2</sup> | R. Lee Vogel, MD<sup>2</sup>

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

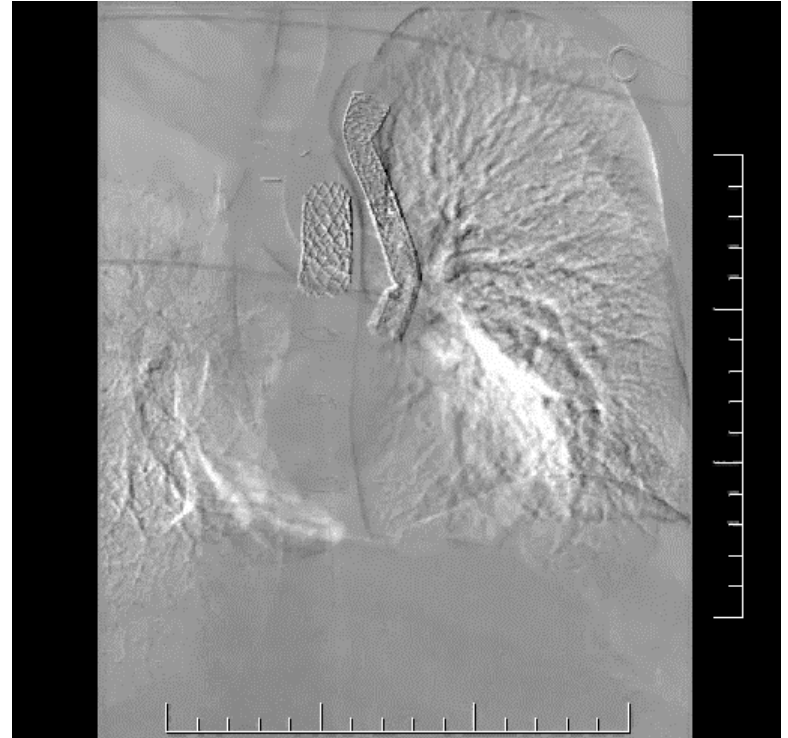
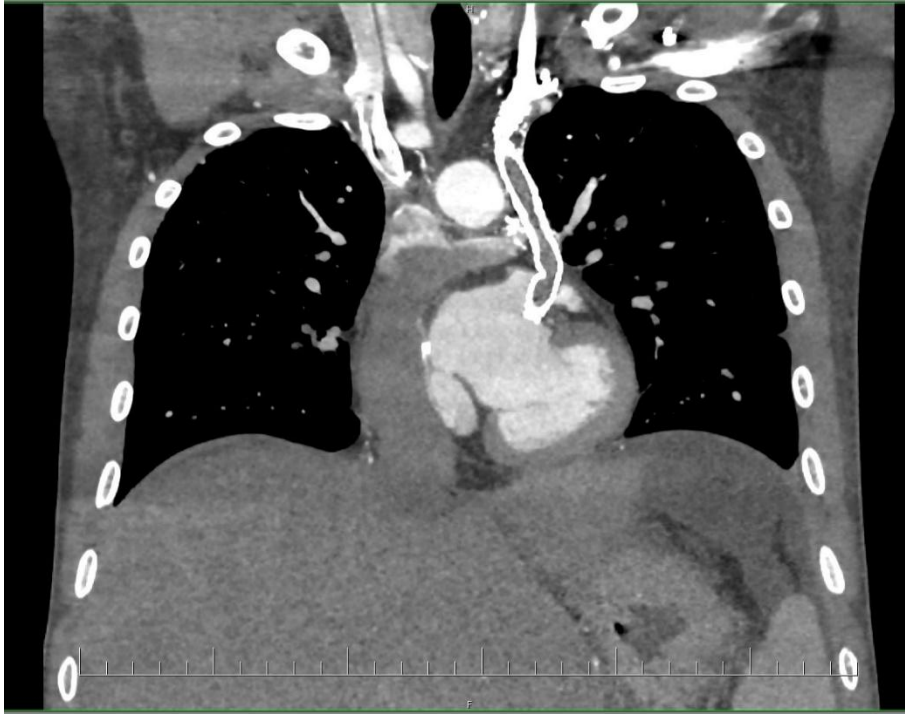


*Courtesy of the patient*



# Lymphatic embolization & Thoracic duct decompression

- 2 months post proc.: leg and **arms oedema**, **satO<sub>2</sub> 97%**
- **thrombosis of the stentgraft** despite stable INR 2,5-3,0 during f-up

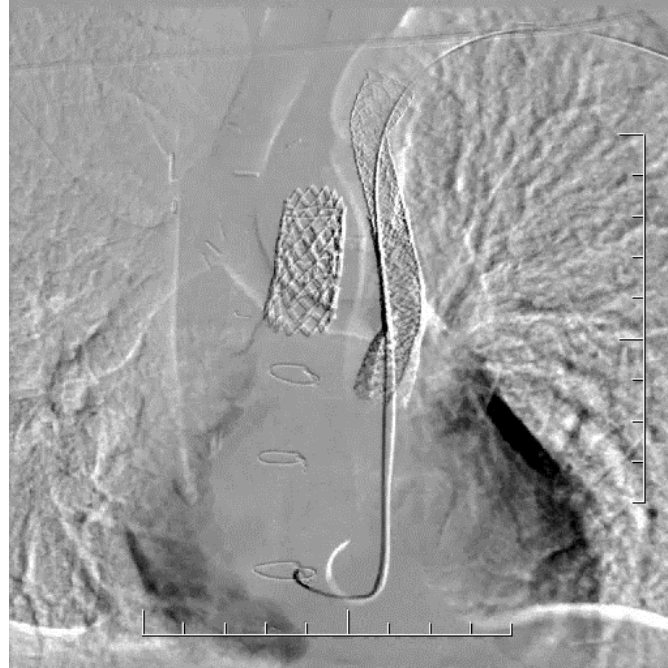


# Site-directed thrombolysis

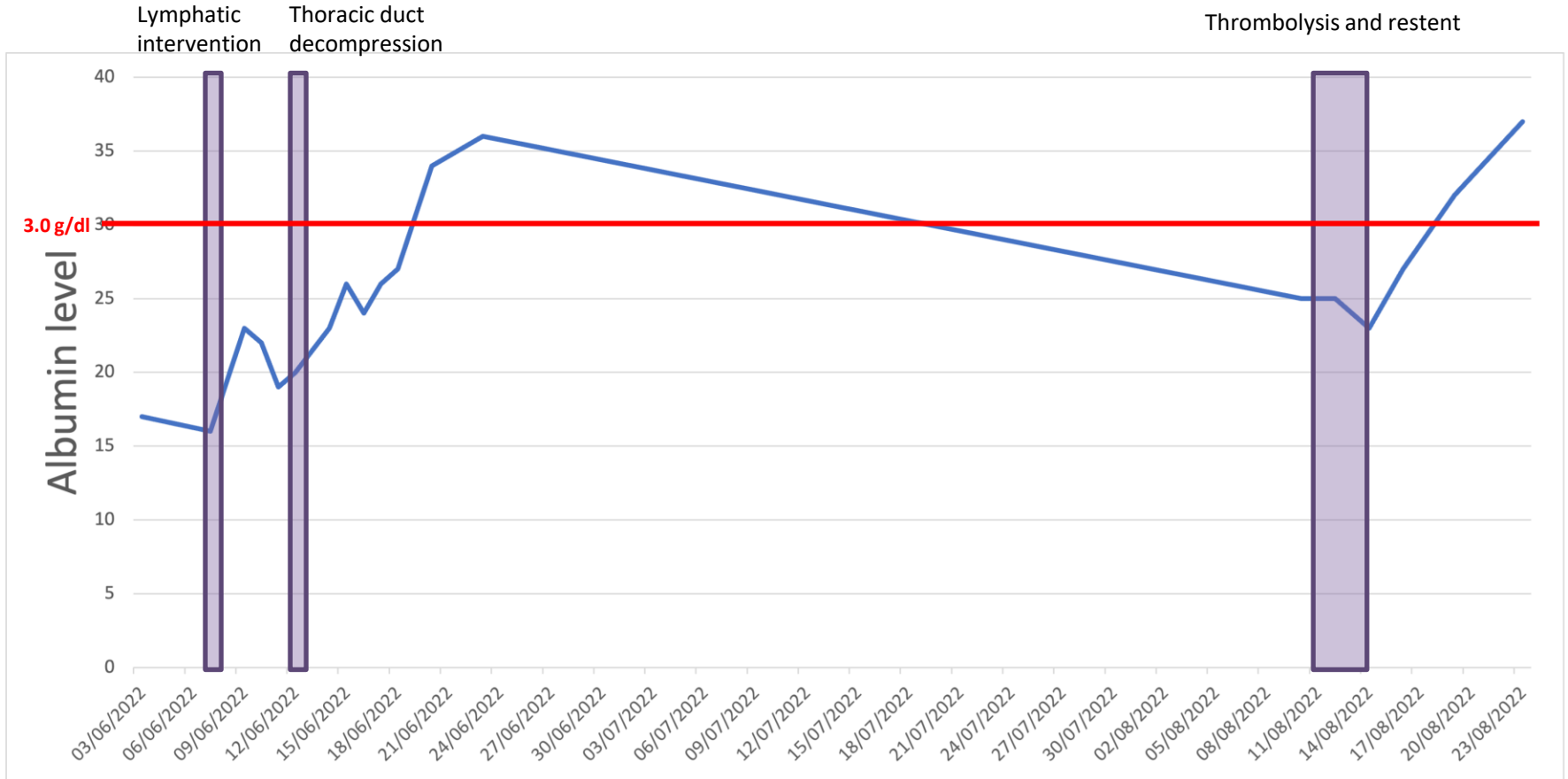
- Valved infusion catheter provides endhole occlusion
- low dose rt-PA 3 days with q 24 h angio – thrombus resolution
- stent to distal narrowing
- clinical improvement → Warfarin + ASA



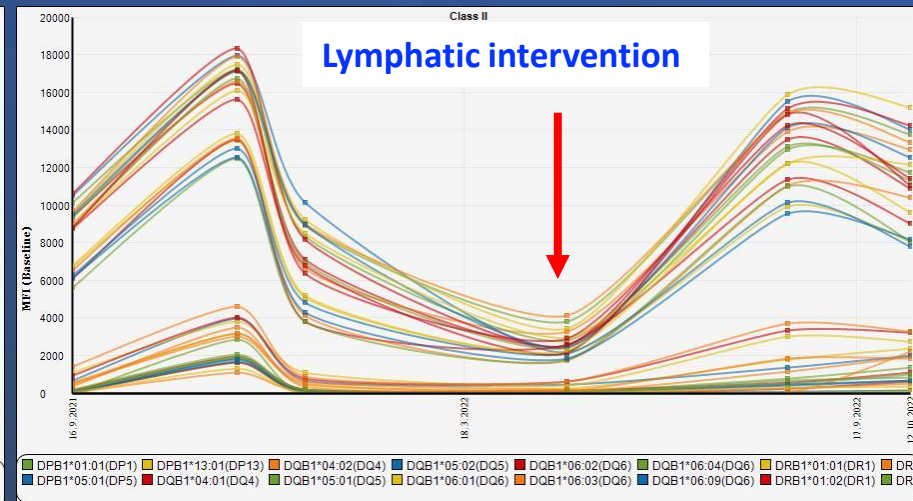
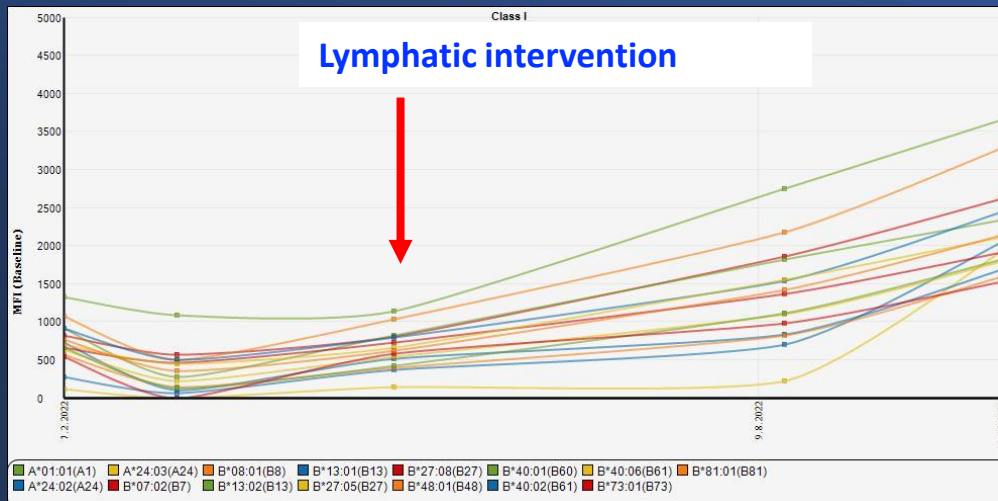
<https://healthmanagement.org/>



# Trend of albumin blood levels



# Sensitization status



# Conclusions

- 50 % of (selected) pts. after Norwood St. I survived into adulthood
- Aortic atresia poses a high risk of early mortality after St.I
- Interventional fenestration closure is associated with mortality
- Close to ½ of HLHS survivors reaching adulthood is free of any major adverse CV event
- New advancements (lymphatic procedures...) might improve the long-term outcome



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Klinika detskej kardiológie LFUK a DKC

Vás pozývajú na

## XXVII. Memoriál prof. Ireny Jakubcovej



Odborný program

10. – 11. november 2022

Detské kardiocentrum, NÚSCH a.s.



Lekárska fakulta  
Ul' v Bratislave



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

NOLA  
KITCHEN

*Porto, Portugal, August 2022*