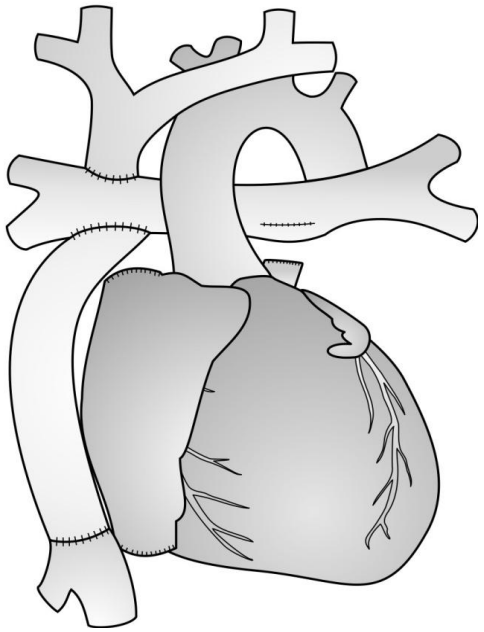


21 years follow-up of liver function in patients with Fontan circulation

(longitudinal study)

D. Jičínská, O. Materna, P. Antonová, K. Koubský, M. Jičínský,
M. Štefánek, J. Janoušek, V. Chaloupecký

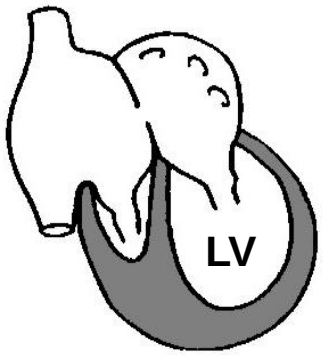
Motol University Hospital, Prague

Prague 28.11.2022

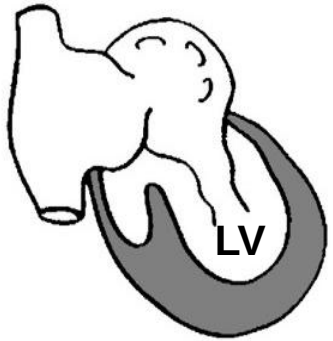
Functionally univentricular heart

Congenital heart defects that cannot be surgically divided into 2 functional ventricles → **Univentricular circulation (Fontan)**

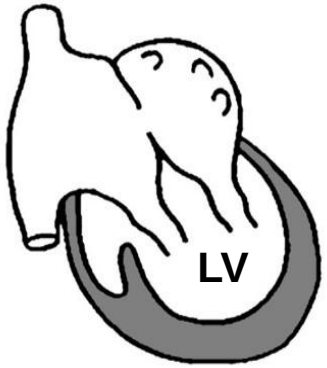
Hypoplastic right ventricle



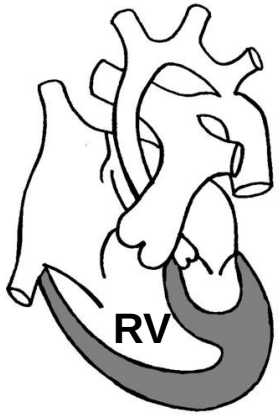
Trikuspid atresia



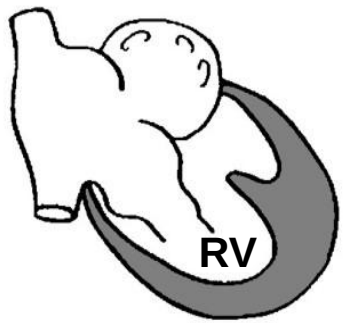
Double inlet left ventricle



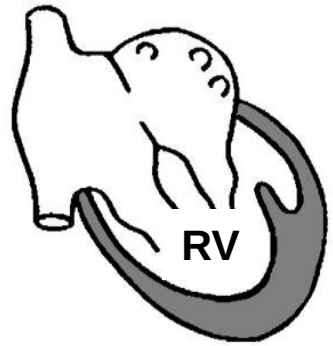
Hypoplastic left ventricle



Mitral atresia



Double inlet right ventricle



Functionally Univentricular Heart – Staged surgery

1st phase
Newborn

Newborn

Pulmonary blood flow

Decreased

Hypoxemia

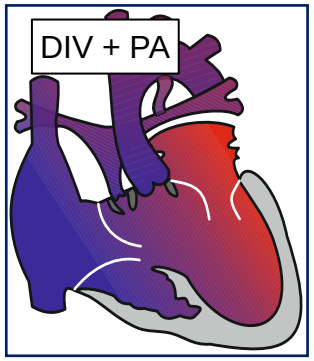
Increased

Heart failure
Pulmonary hypertension

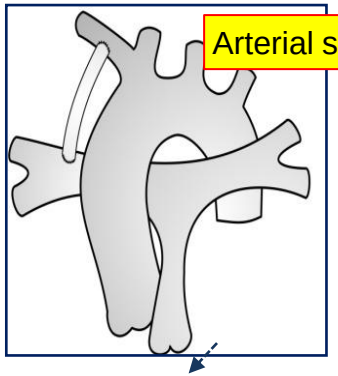
Extremely increased

Circulatory failure
Shock

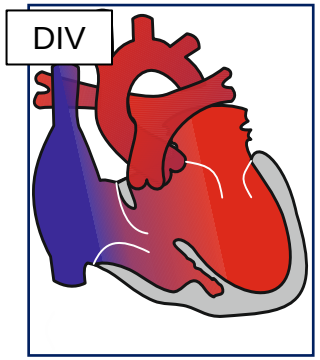
DIV + PA



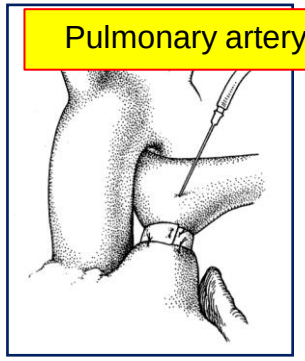
Arterial shunt



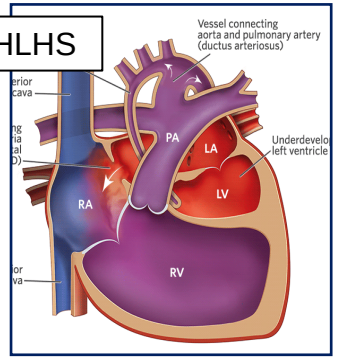
DIV

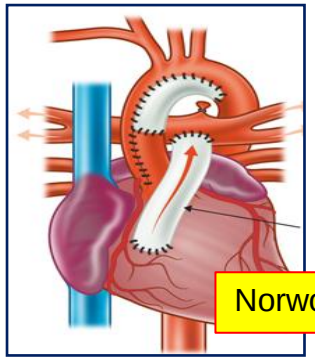


Pulmonary artery banding



HLHS



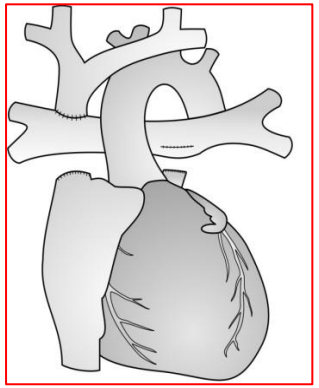


Norwood

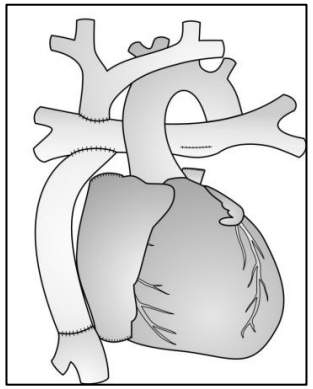
Bidirectional cavopulmonary anastomosis

Total cavopulmonary connection

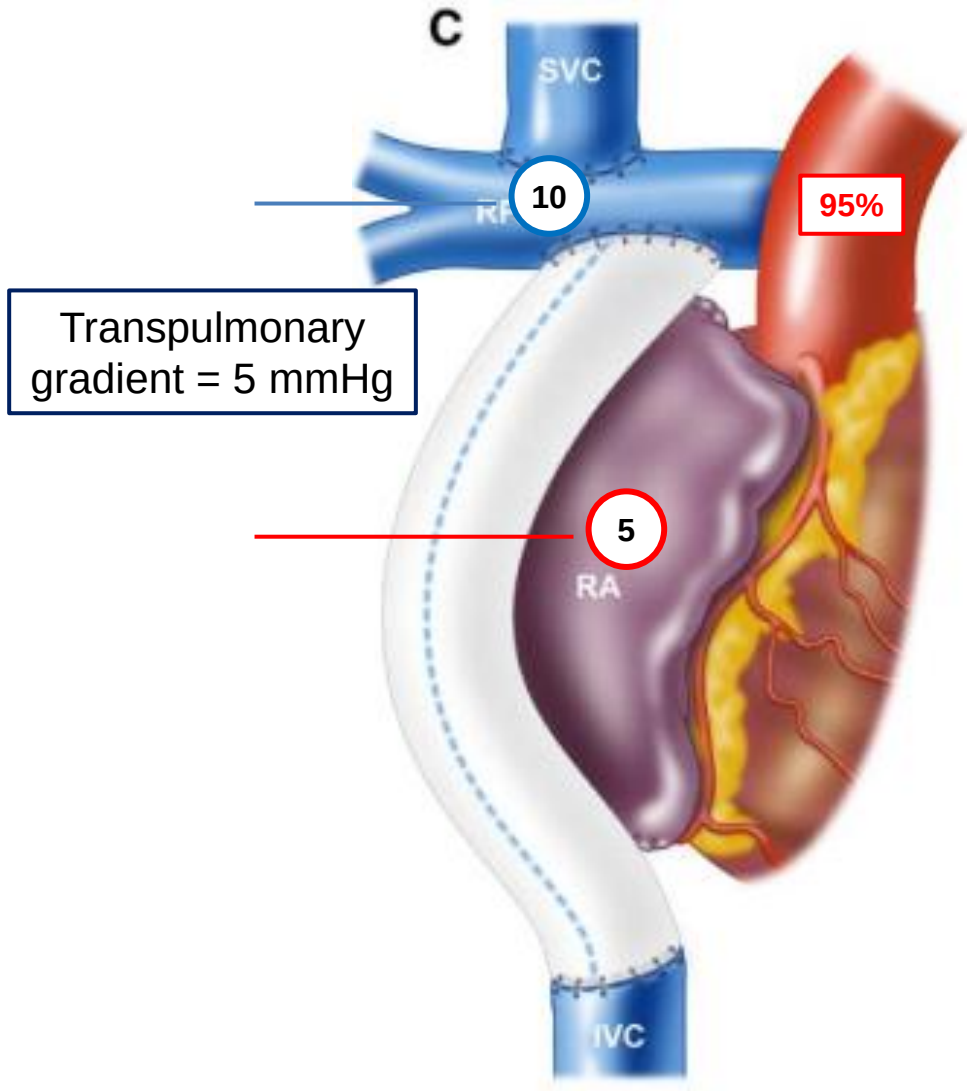
2nd phase
Age ~ 4 months



3rd phase
Age ~ 4 yrs

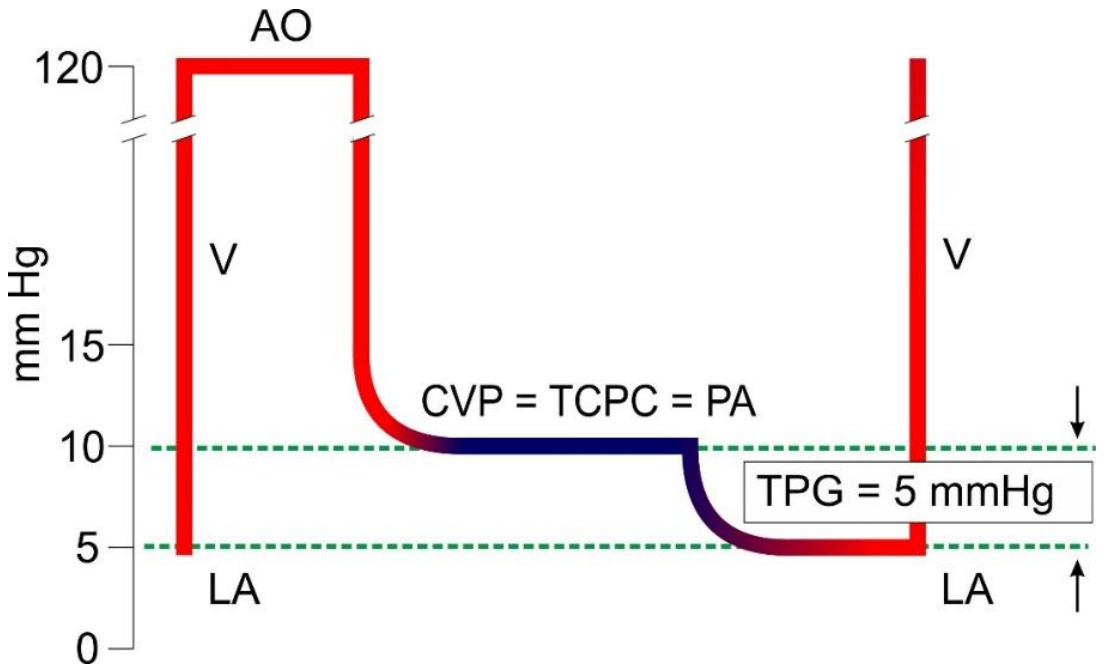


Total cavopulmonary connection

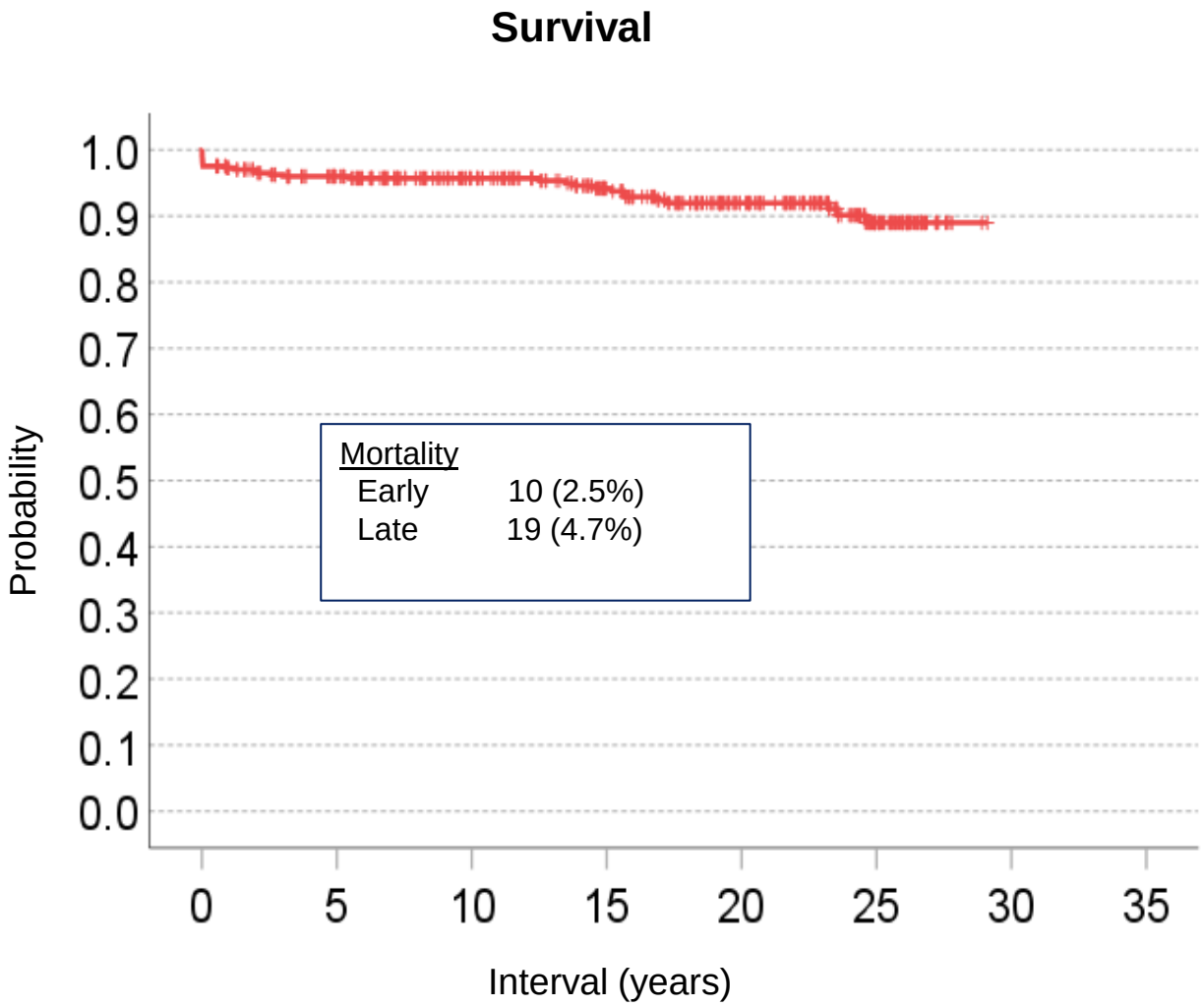


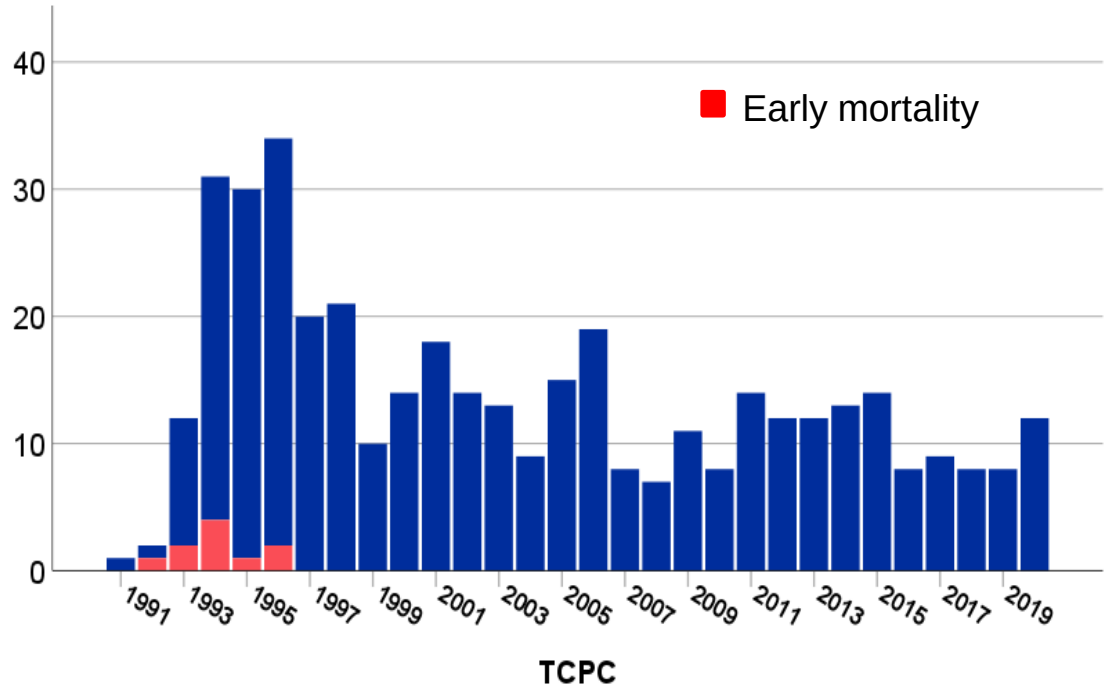
Effects

- normal ventricular volume load
- SaO_2
- increased systemic venous pressure



Total cavopulmonary connection in Children´s Heart Centre Prague (1991 - 2020) N= 407

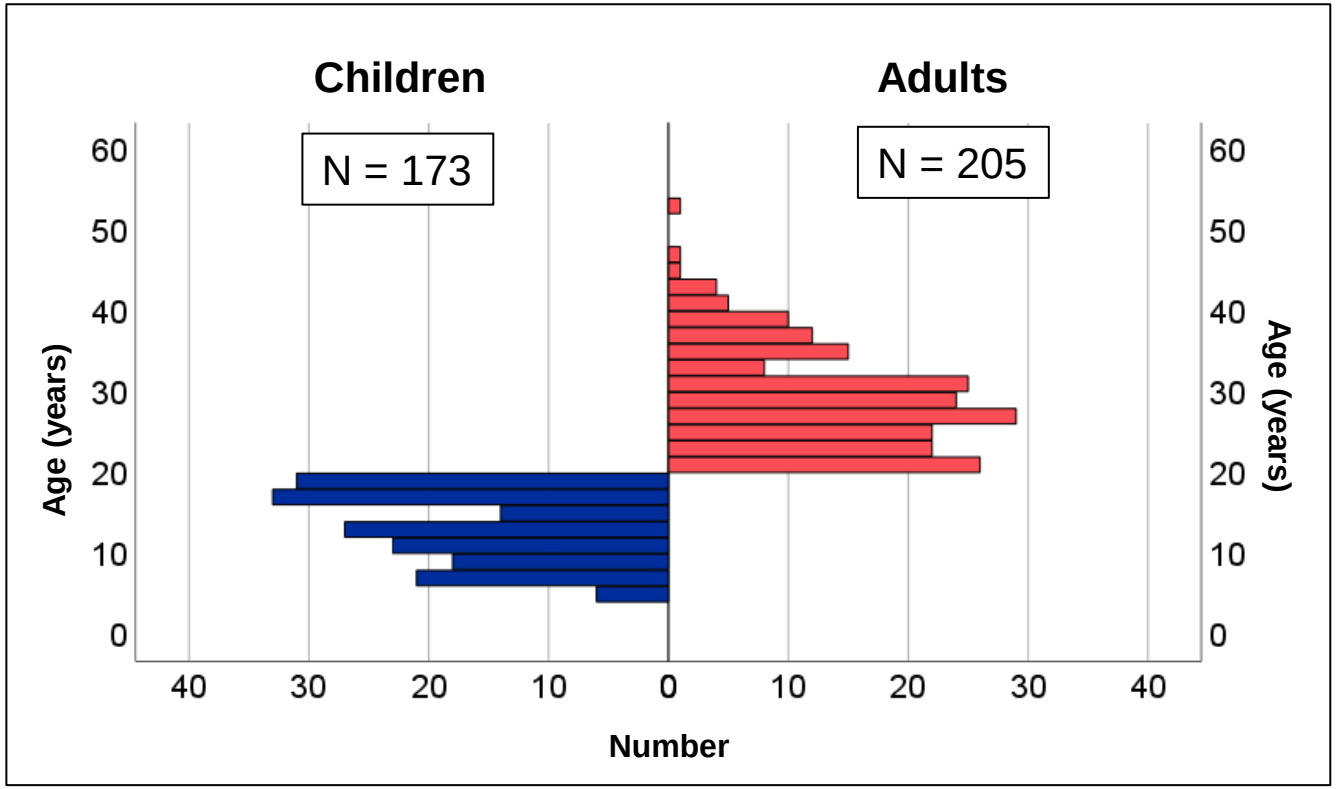




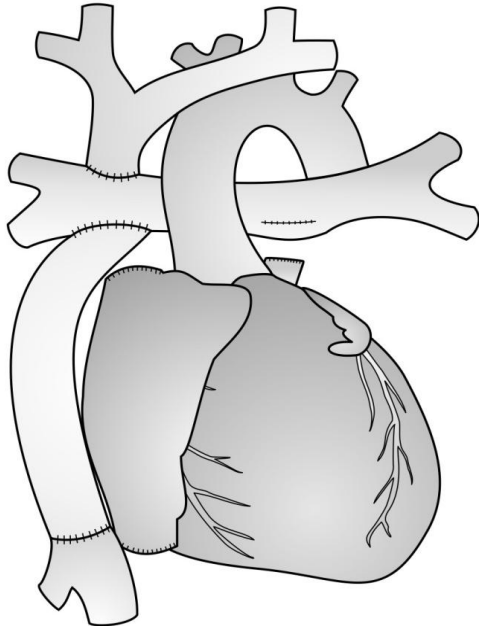
HTX 9 patients

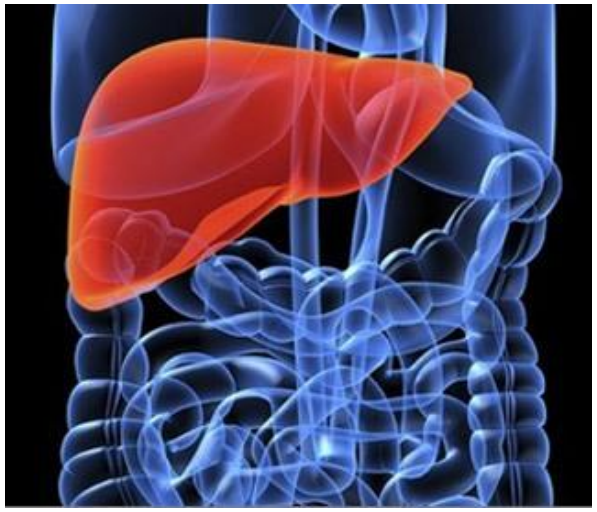
Total cavopulmonary connection in Children´s Heart Centre Prague (1991 - 2020)

378 survivals



Impact of Fontan circulation on end-organs



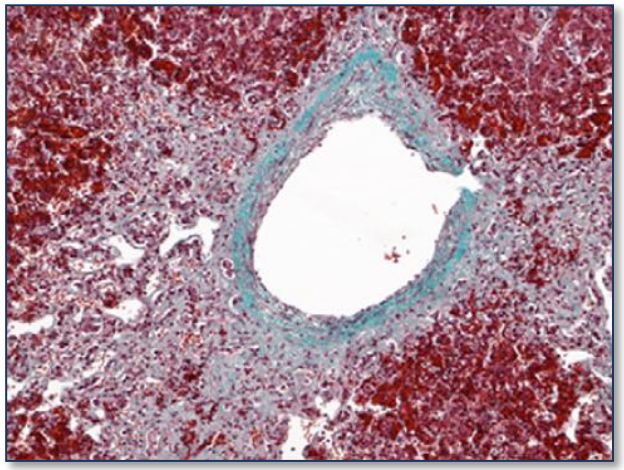


Fontan associated liver disease (FALD)

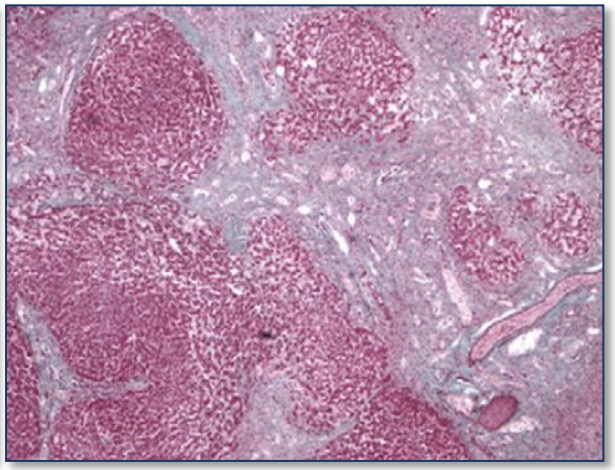
Liver biopsy

Author	Year	N		Age (yrs)		Interval (yrs)	Fibrosis Sinus	Fibrosis Portal	Cirr-hosis	Fontan
Kutty	2014	10	**14**	± 6	**11**	± 6	80%	50%	20%	"good"
Evans	2015	14	**15**	(7-24)	**12**	(2-21)	100%			"good"
Bryant	2011	16	**24**	(16-40)	**17**	(7-29)	100%	80%	13%	"stable"
Wu	2017	75	**28**	(13-59)	**21**	(11-35)	97%	90%	33%	"stable"

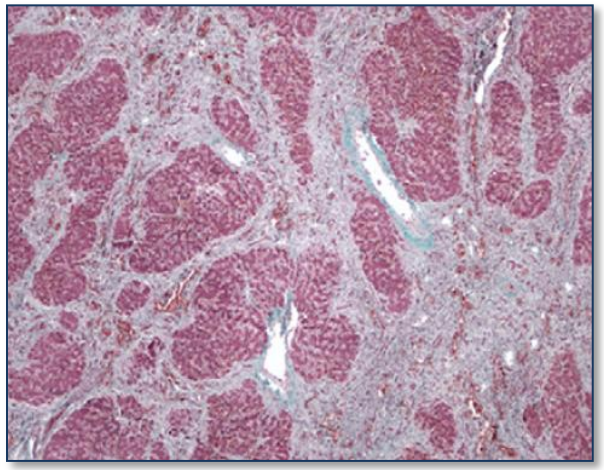
Sinusoidal fibrosis



Portal fibrosis



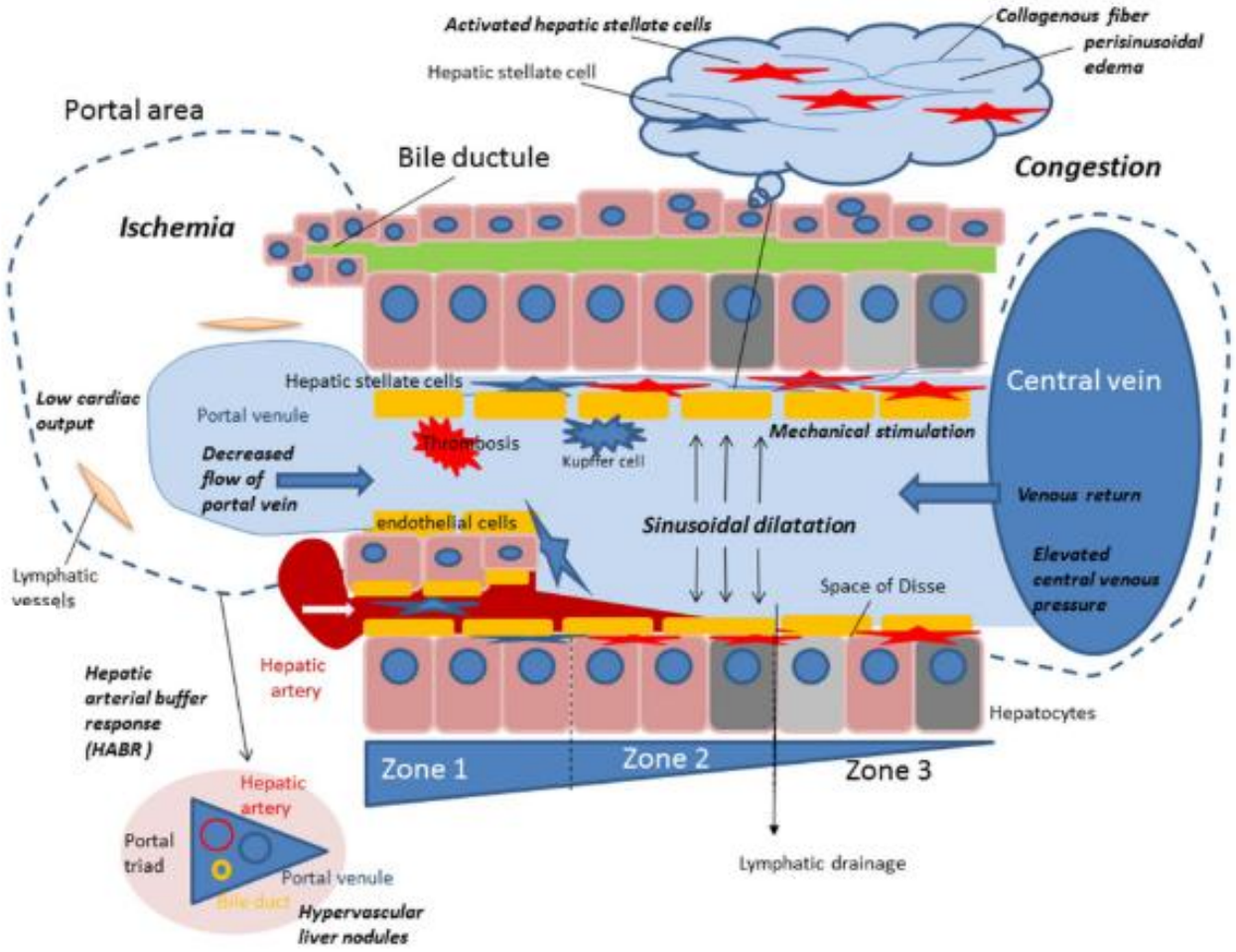
Cirrhosis



Fontan associated liver disease (FALD)

Patophysiology

- increased venous pressure
- borderline cardiac output
- change in blood supply
- hypoxia (rest, exercise)
- duration of the Fontan procedure
- pre / perioperative liver impairment



Kogiso 2020

> Front Cardiovasc Med. 2022 Sep 29;9:986653. doi: 10.3389/fcvm.2022.986653. eCollection 2022.

Impact of liver fibrosis and nodules formation on hemodynamics in young adults after total cavopulmonary connection. A magnetic resonance study

Václav Chaloupecký [1], Denisa Jičínská [1], Viktor Tomek [1], Ondřej Materna [1], Roman Gebauer [1], Rudolf Poruban [1], Petra Antonová [2], Theodor Adla [3], Matěj Štefánek [3], Vojtěch Illinger [4], Karel Kotaška [5], Jan Janoušek [1]

TCPC longitudinal study in Children‘s Heart Centre Prague

Coagulation profile and liver function in 102 patients after total cavopulmonary connection at mid term follow up

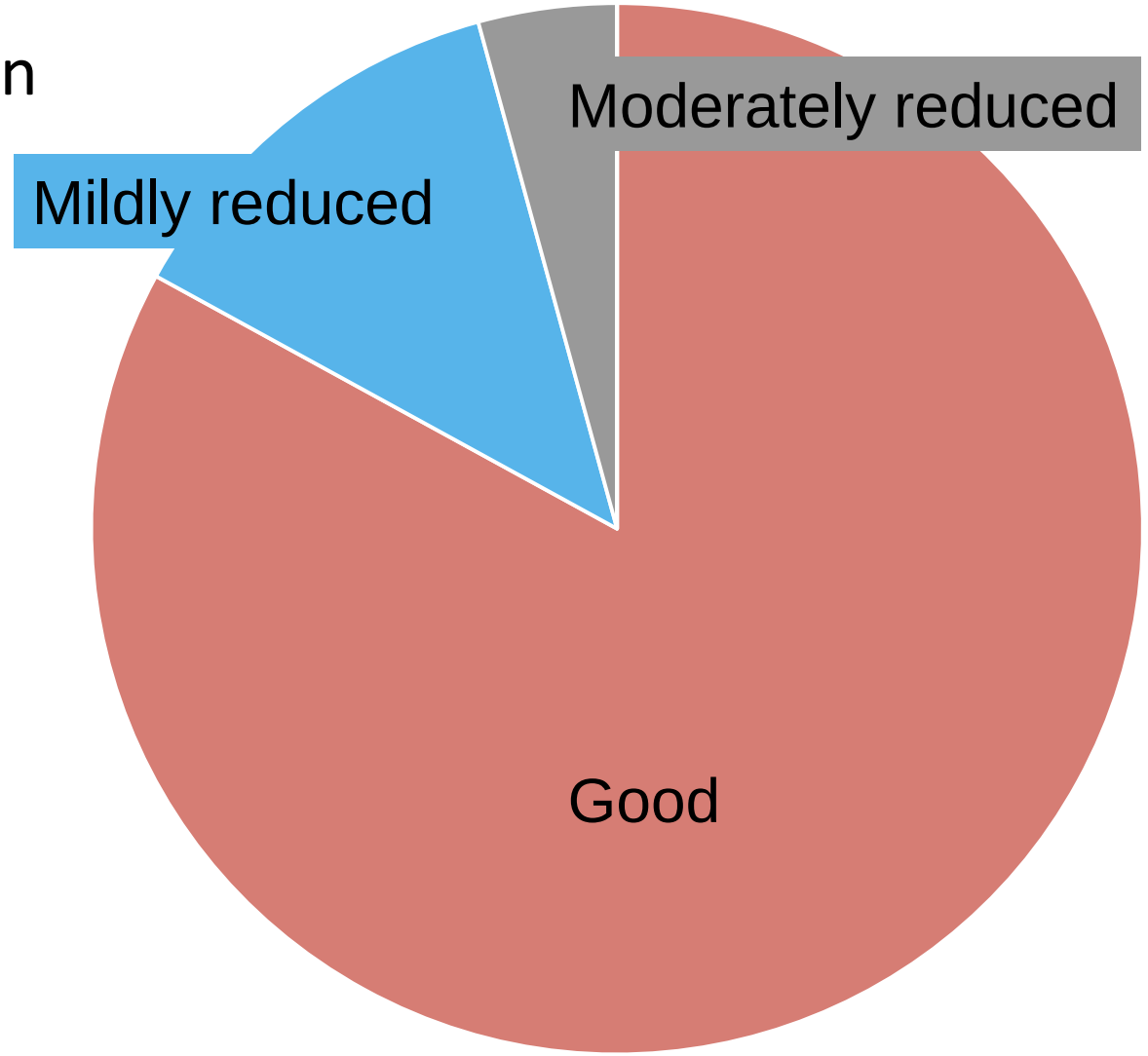
V Chaloupecký, I Svobodová, I Hadačová, V Tomek, B Hučín, T Tláskal, J Janoušek, O Reich, J Škovránek

Heart 2005

	Study 1	Study 2	Study 3
Period	2000 – 2001	2009 - 2010	2017 - 2018
Number of patients	102	69	47
Age at study	9 (4 - 22)	17 (12 - 31)	26 (21 - 38)
Interval from TCPC (yrs)	5 (1 - 8)	13 (9 - 17)	21 (18 - 24)

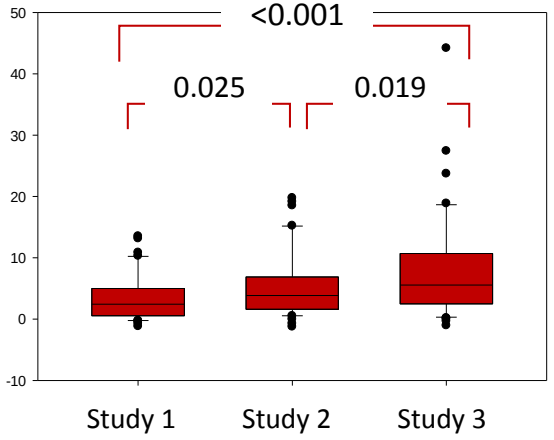
ECHO findings

Ventricular function

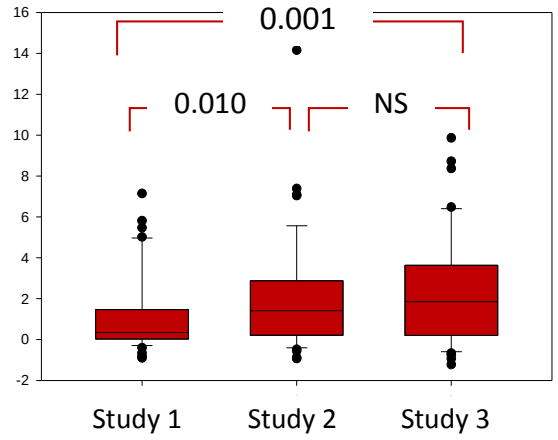


Liver test

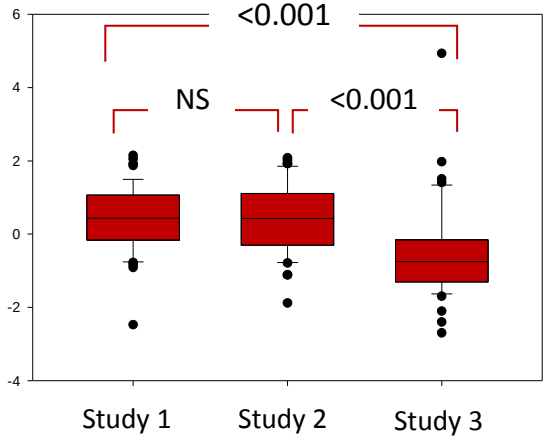
GGT (Z-score)



Bilirubin (Z-score)

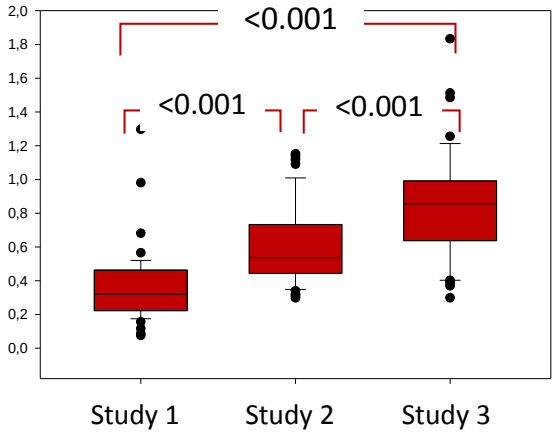


Prealbumin (Z-score)

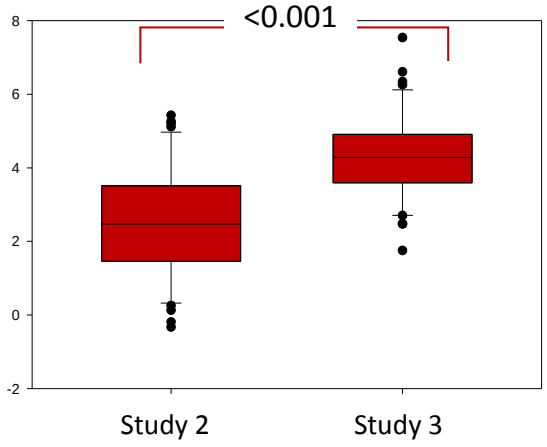


FIB 4

FIB-4 = age*AST/(PLT*(ALT)^0.5)



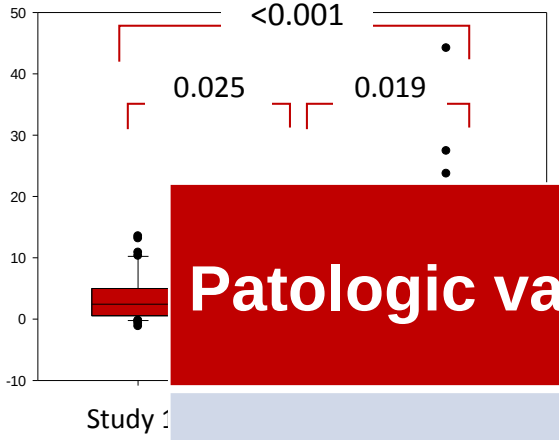
FORNS



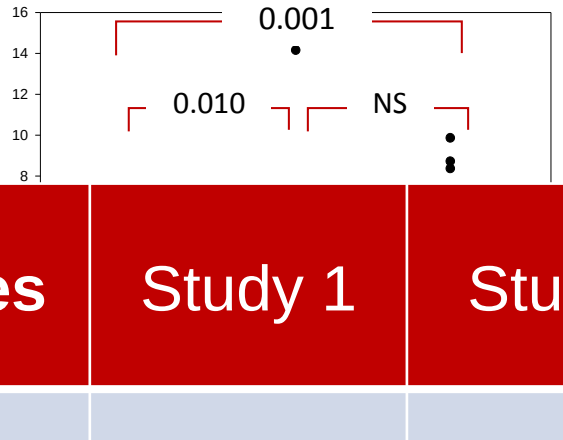
FORNS index = 7,811 – 3,131.ln(PLT) + 0,781.ln(GGT) + 3,467.ln(age) – 0,014.(cholesterol)

Liver test

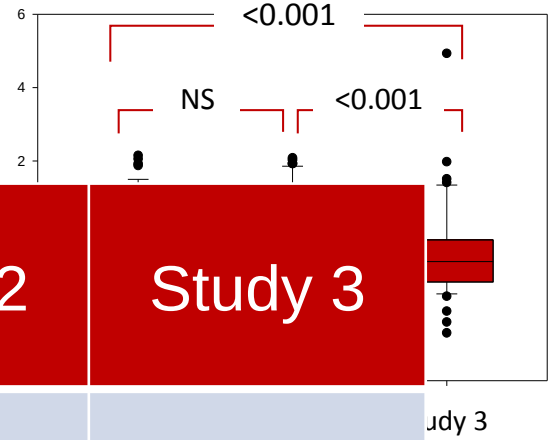
GGT (Z-score)



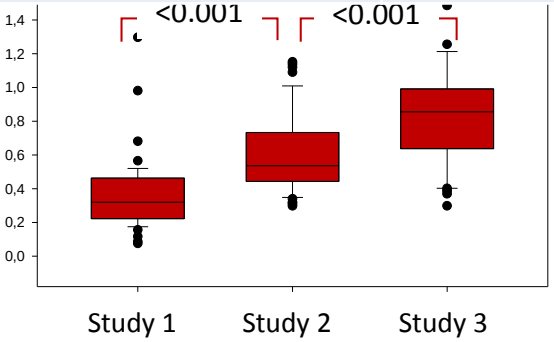
Bilirubin (Z-score)

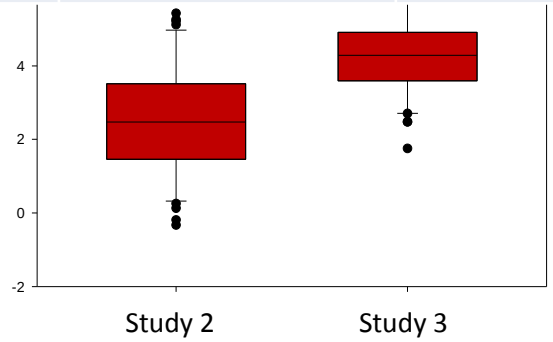


Prealbumin (Z-score)



Patologic values	Study 1	Study 2	Study 3
bilirubin	27%	32%	45%
GGT	54%	66%	74%



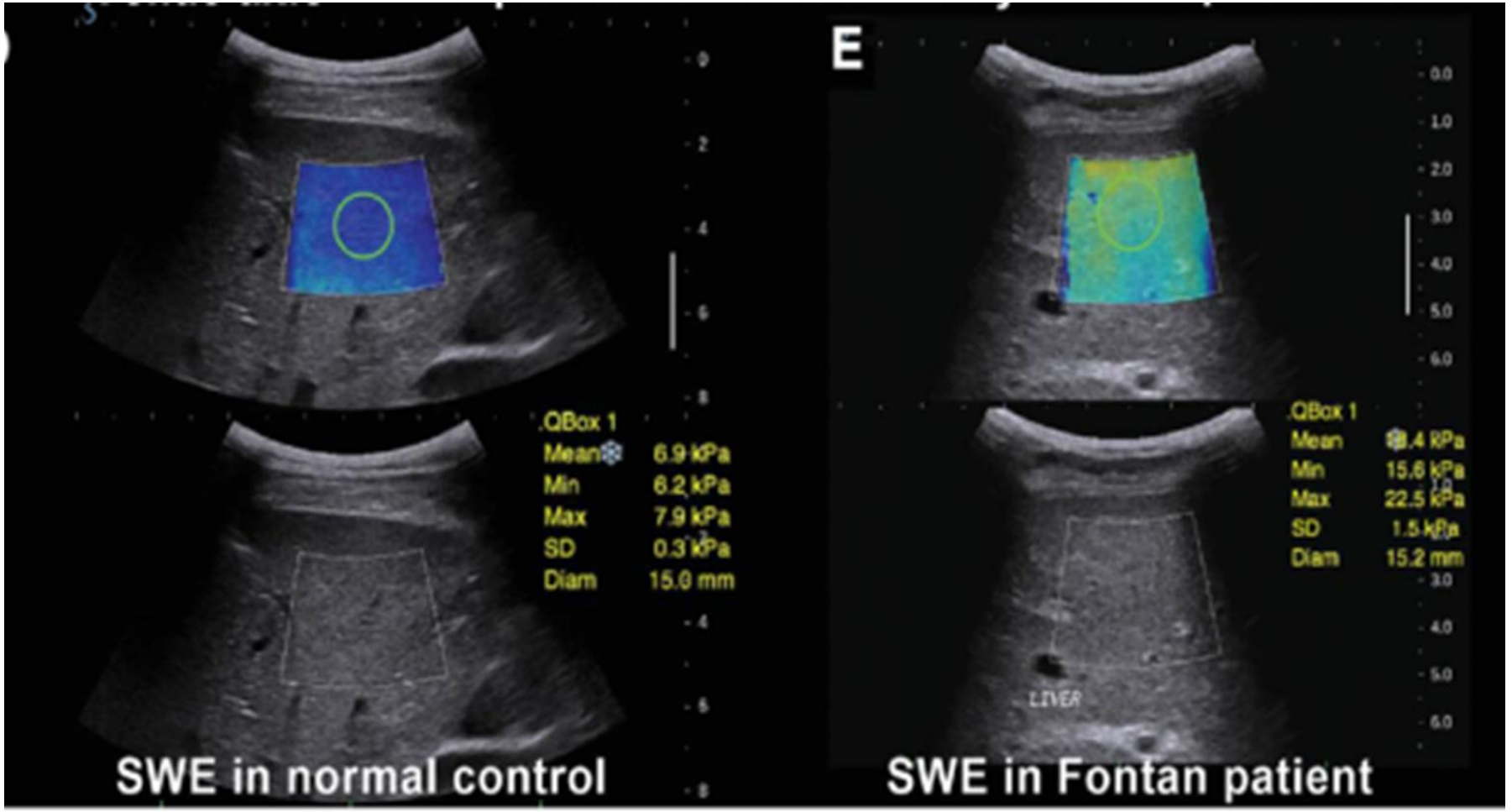


FIB-4 = age*AST/(PLT*(ALT)^0.5)

FORNS index = 7,811 – 3,131.ln(PLT) + 0,781.ln(GGT) + 3,467.ln(age) – 0,014.(cholesterol)

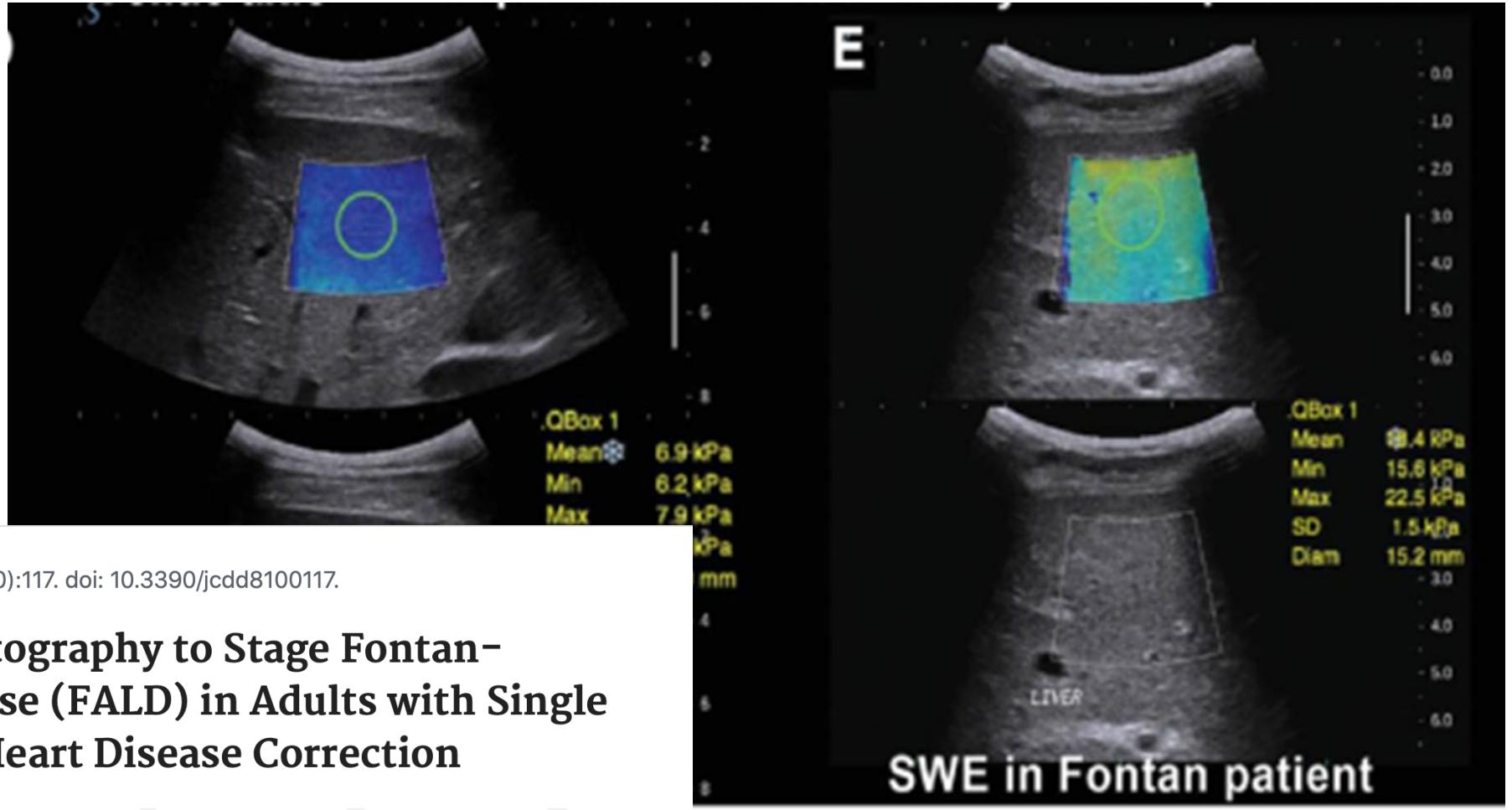
Elastography

abnormal elastography (>7.2 kPa) in all subjects



Elastography

abnormal elastography (>7.2 kPa) in all subjects



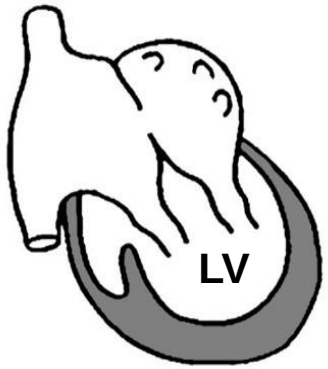
J Cardiovasc Dev Dis. 2021 Sep 23;8(10):117. doi: 10.3390/jcdd8100117.

Role of Transient Elastography to Stage Fontan-Associated Liver Disease (FALD) in Adults with Single Ventricle Congenital Heart Disease Correction

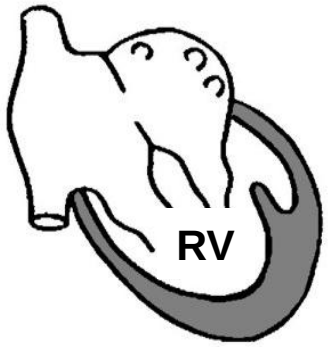
Liliana Chemello [1], Massimo Padalino [2], Chiara Zanon [1], Luisa Benvegnu' [3], Roberta Biffanti [4], Daniela Mancuso [5], Luisa Cavalletto [1]

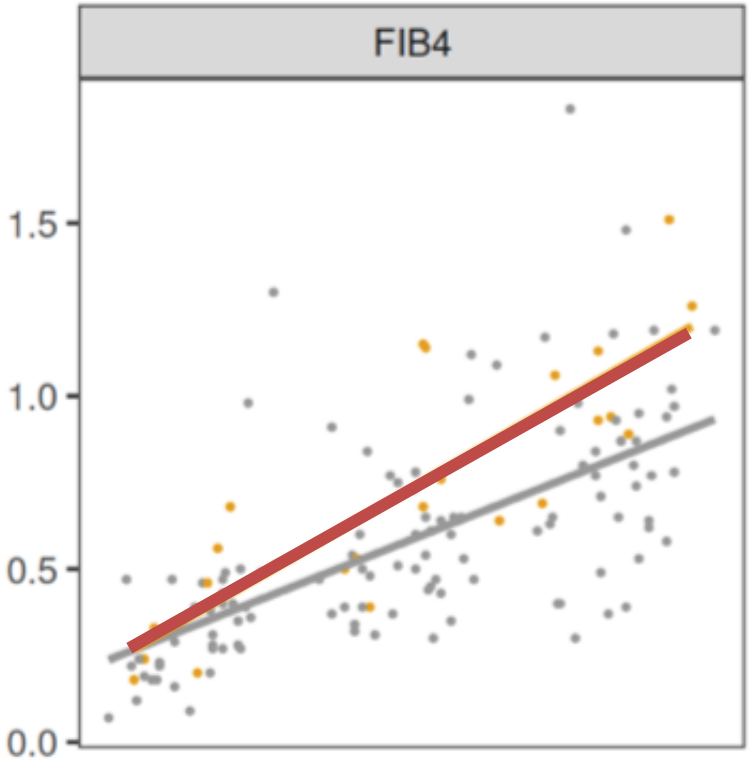
Risk factors – right ventricular morphology

Double inlet left ventricle

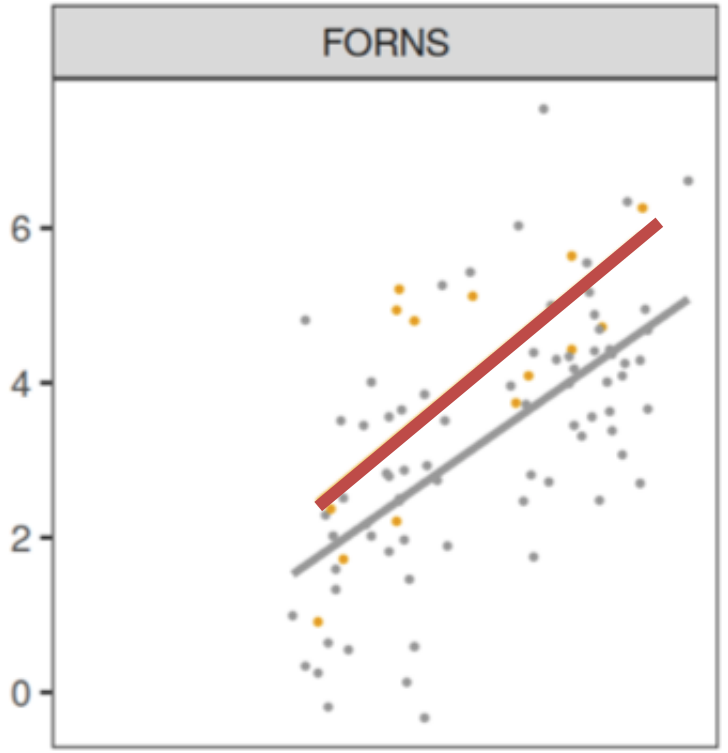


Double inlet right ventricle





Steeper increase in FIB-4



Higher values of Forns index

Quality of life

SF-36 questionare:

Subjective rating of personal health

Conclusion

- FALD is gradually progressing even in patients in a good clinical condition with a good or mildly decreased single ventricle function

- Close cooperation between cardiologist and hepatologist in adult patients after TCPC is inevitable

Thank you for your attention

denisa.jicinska@fnmotol.cz