Specifika srdečního selhání u jednokomorové cirkulace

V. Chaloupecký

Dětské kardiocentrum 2. LF UK
Fakultní nemocnice v Motole

26. symposium PS Chlopenní a vrozené srdeční vady v dospělosti ČKS
Hradec Králové 6. 3. 2025

Total cavopulmonary connection: A logical alternative to atriopulmonary connection for complex Fontan operations

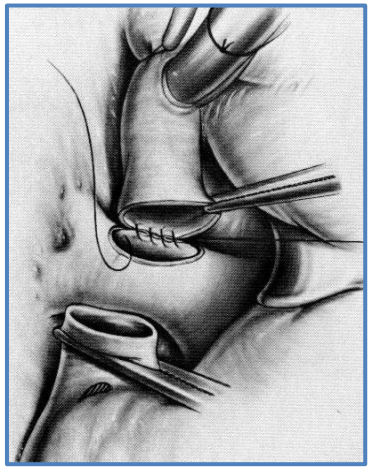
Marc R. de Leval, MD (by invitation), Philip Kilner, MD (by invitation), Marc Gewillig, MD (by invitation), and Catherine Bull, MD (by invitation),

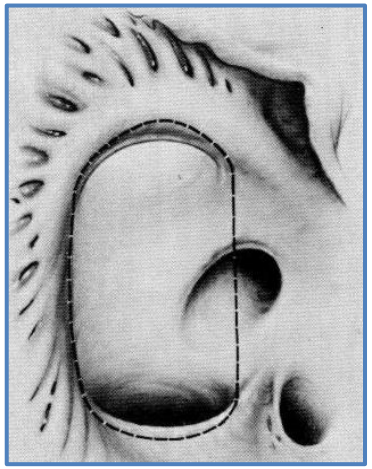
J Thorac Cardiovasc Surg 1988;96:682-95

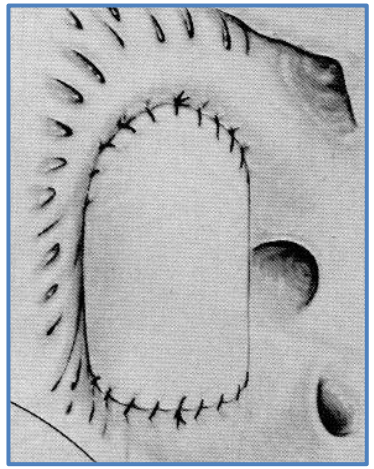
Table I. *Total cavopulmonary connection (March 1987 to March 1988)*

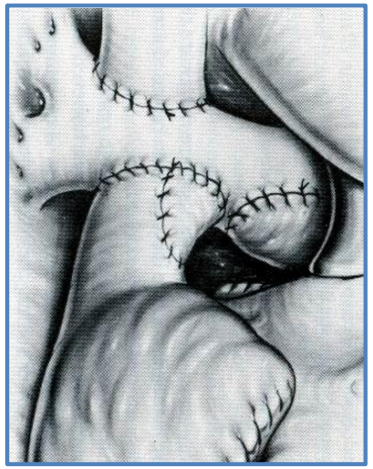
Diagnosis	*Patients*
Double-inlet ventricle	11
Absent right AV connection	2
Pulmonary atresia, IVS	2
DORV, hypoplastic RV	1
VSD, hypoplastic RV	1
TGA, straddling TV	2
AV discordance, hypoplastic RV	1
Total	20

Doba sledování max. 38 let





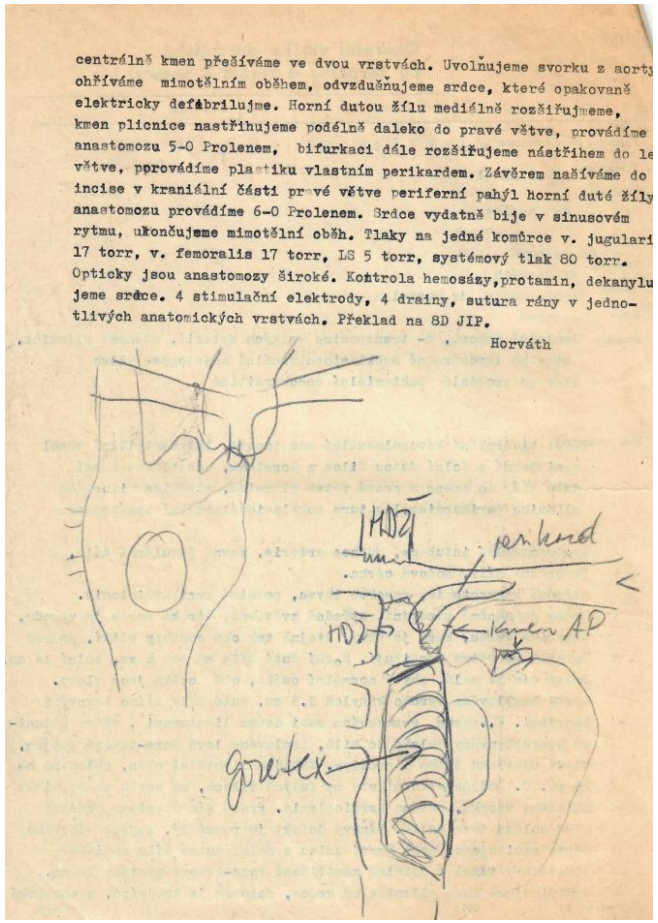




1. Úplné kavopulmonální spojení (TCPC) v Dětském kardiocentru / v ČR

MUDr. Pavel Horváth
+ 1994

KL 5 let, DILV, TCPC 20.11.1991



centrálně kmen přešíváme ve dvou vrstvách. Uvolňujeme svorku z aorty ohříváme mimotělním oběhem, odvzdušňujeme srdce, které opakovaně elektricky defibrilujme. Horní dutou žílu mediálně rozšiřujeme, kmen plicnice nastřihujeme podélně daleko do pravé větve, provádíme anastomozu 5-0 Prolenem, bifurkaci dále rozšiřujeme nástřihem do le větve, pprovádíme plastiku vlastním perikardem. Závěrem našíváme do incise v kraniální části pravé větve periferní pahýl horní duté žíly anastomozu provádíme 6-0 Prolenem. Srdce vydatně bije v sinusovém rytmu, ukončujeme mimotělní oběh. Tlaky na jedné komůrce v. jugulari 17 torr, v. femoralis 17 torr, LS 5 torr, systémový tlak 80 torr. Opticky jsou anastomozy široké. Kontrola hemosázy,protamin, dekanylu jeme srdce. 4 stimulační elektrody, 4 drainy, sutura rány v jednotlivých anatomických vrstvách. Překlad na SD JIP.

Horváth

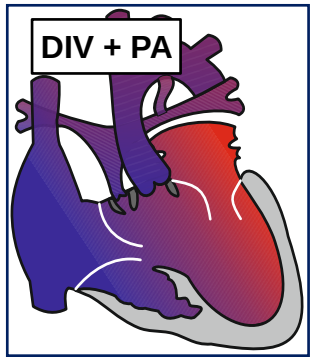
Funkčně jednokomorové srdce – fázové operační řešení

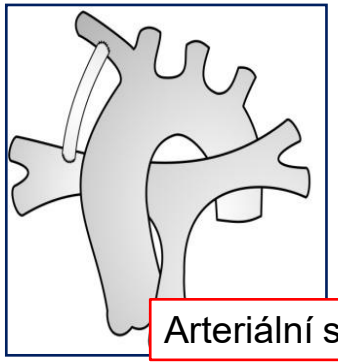
1. fáze
Novorozenec

Plicní průtok

Snížený

Hypoxémie



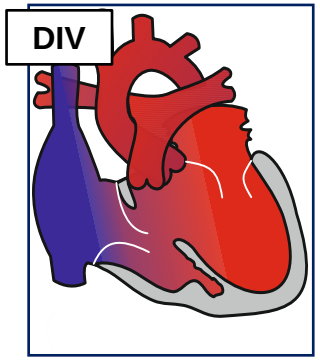


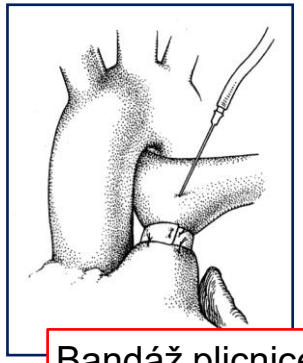
Arteriální spojka

Bidirekční kavopulmonální anastomóza

Zvýšený

Srdeční selhání, plicní hypertenze

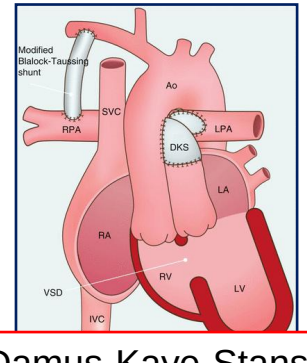




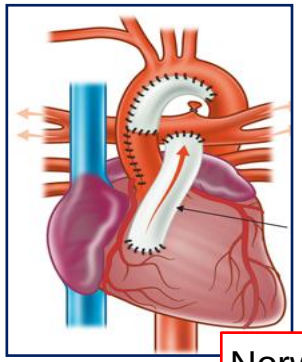
Bandáž plicnice

Extrémně zvýšený / nízký srdeční výdej

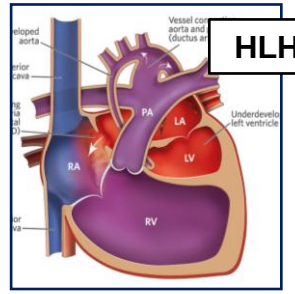
Selhání oběhu, šok



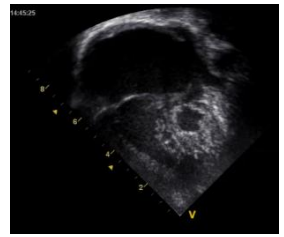
Damus-Kaye-Stansel



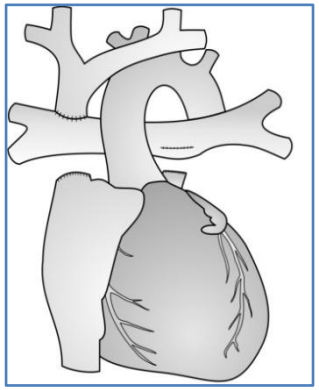
Norwood



HLHS

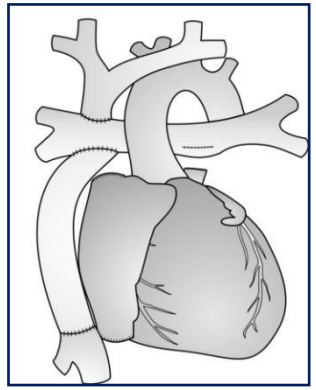


2. fáze
Kojenec
~ 4 měsíce



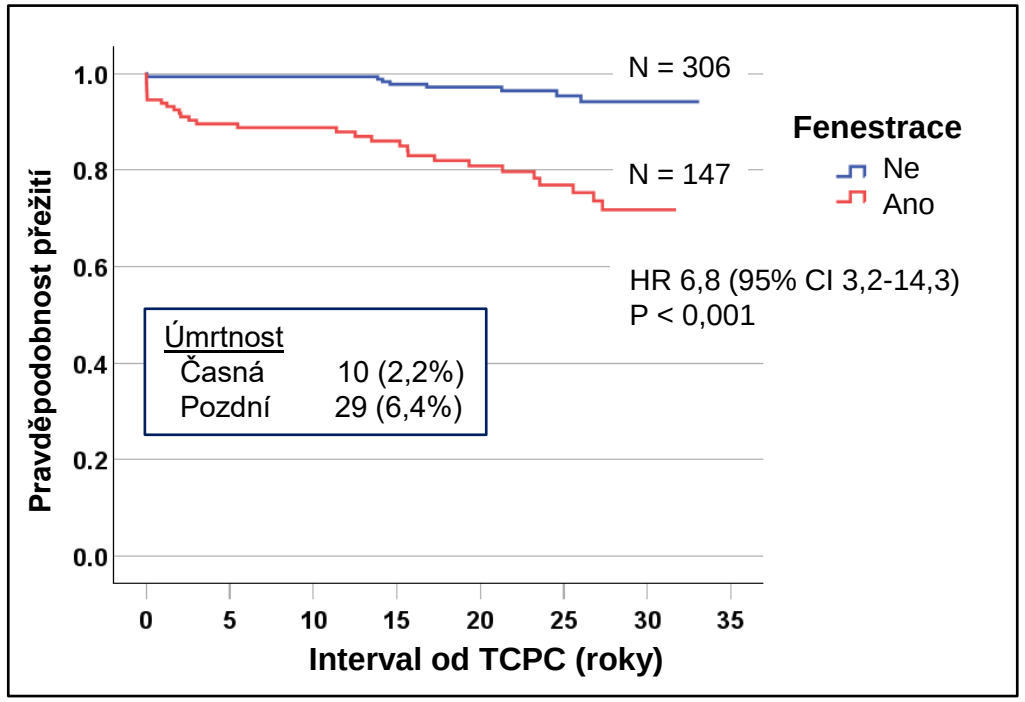
TCPC

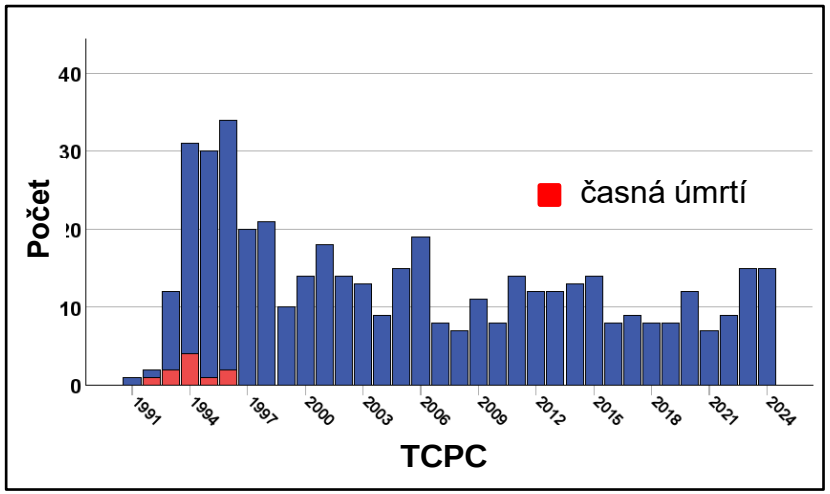
3. fáze
Dítě
~ 4 yrs

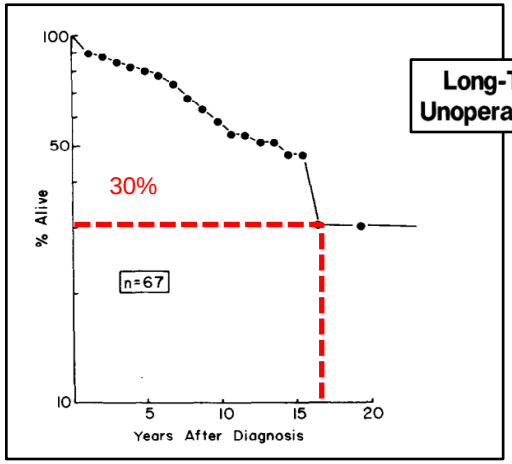


Úplné kavopulmonální spojení v Dětském kardiocentru (1991 - 2024)

N = 453





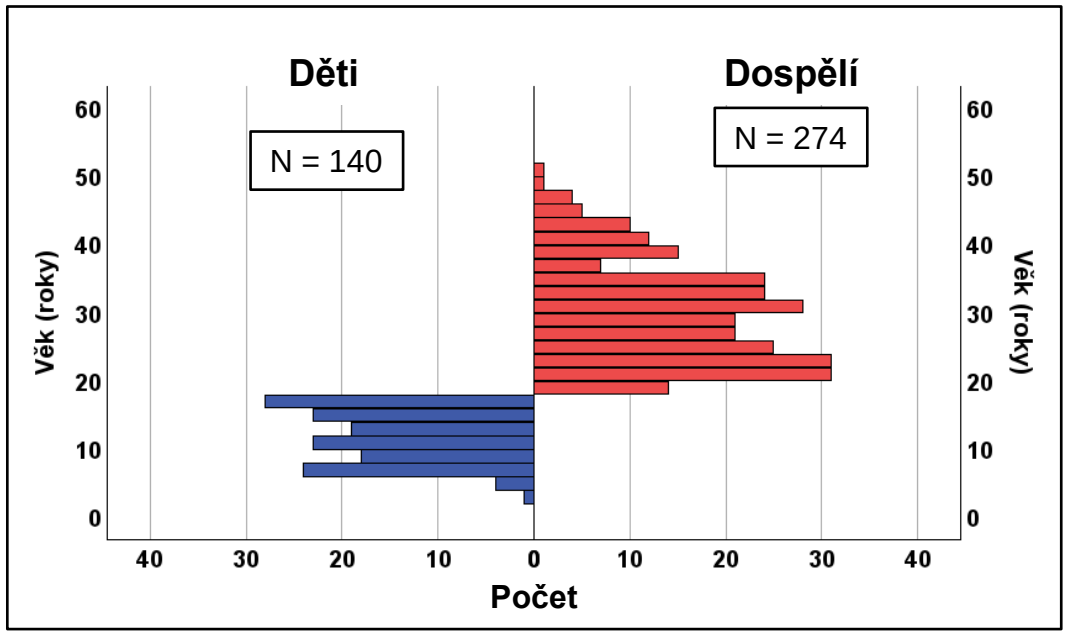


Long-Term Follow-Up in the Unoperated Univentricular Heart

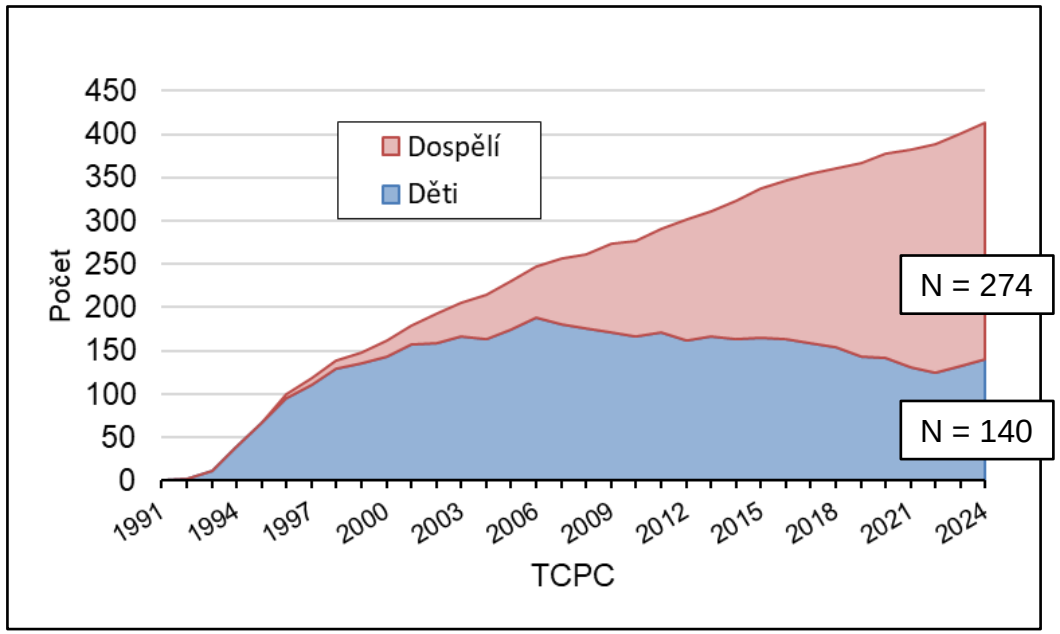
Moodie 1984

Úplné kavopulmonální spojení v DKC (1991 - 2024)

Žijící jedinci k 31. 12. 2024 (N = 414)



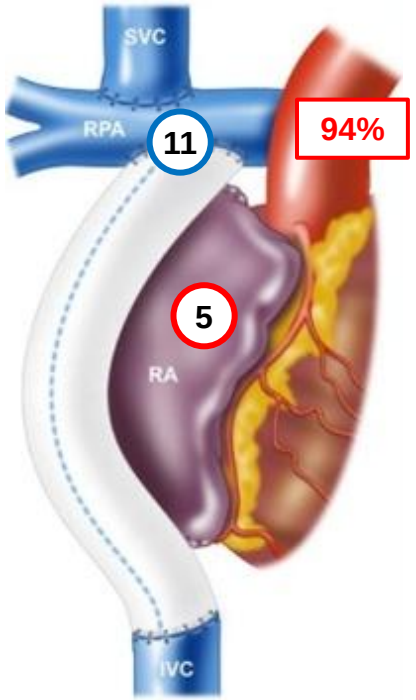
Intervalová kumulativní četnost

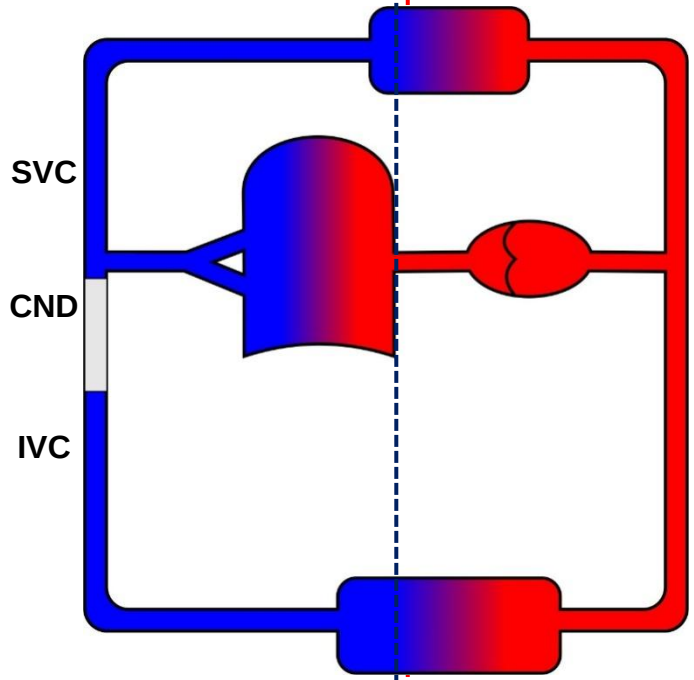


TCPC - patofyziologie krevního oběhu

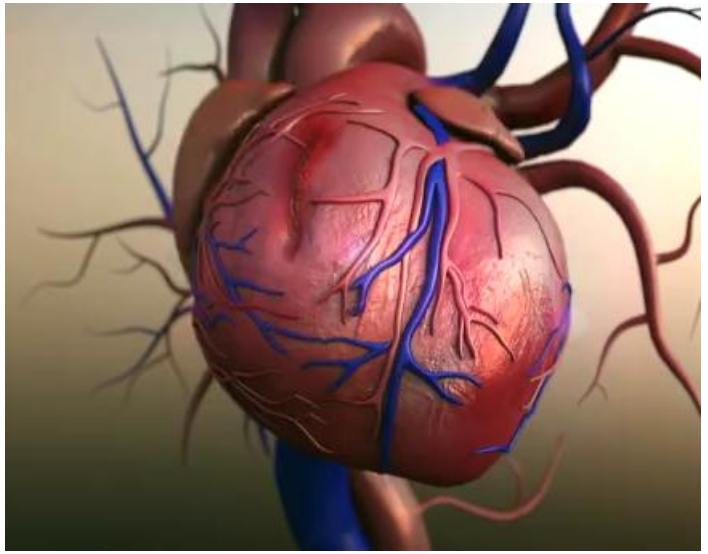
Neoportální systém

Gewillig 2014

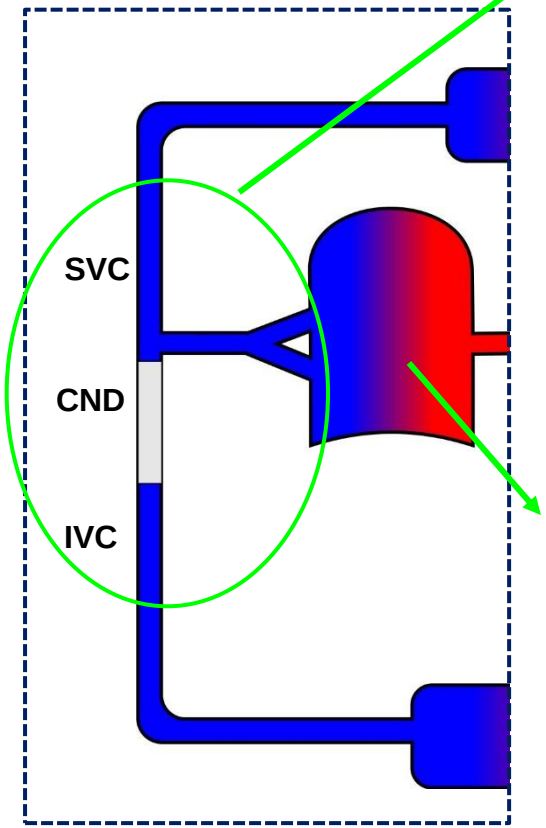




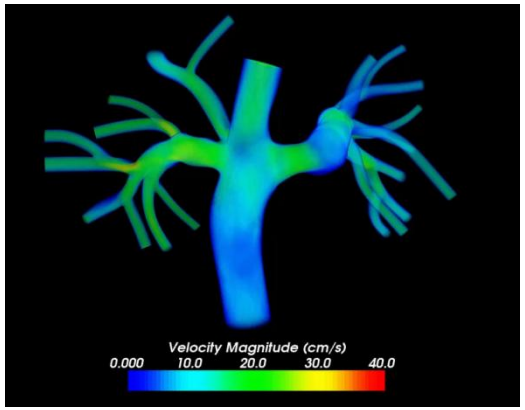
Systémová cirkulace



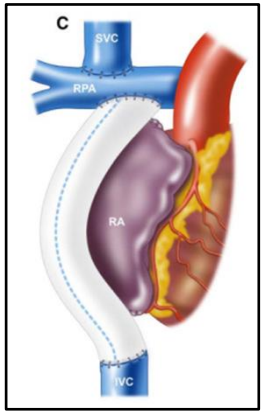
TCPC - Základní komponenty neoportálního systému



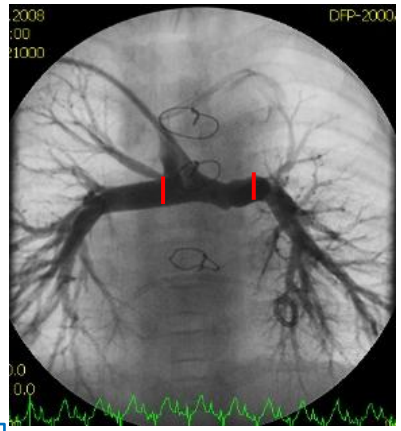
TCPC cesta >>> nepulzatilní tok pomalých rychlostí



Ztráty energie
turbulence
víry
třecí síly
stagnace proudu
kolize proudů
viskozita krve



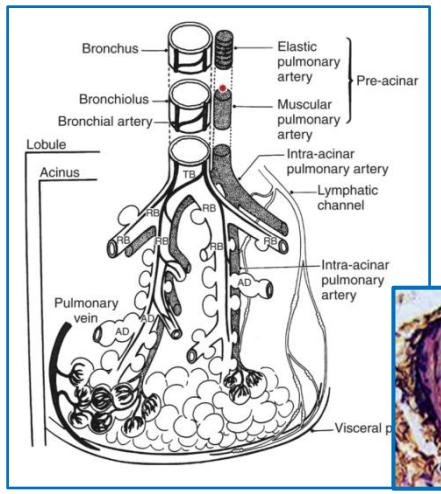
Dostatečná kapacita plicnic

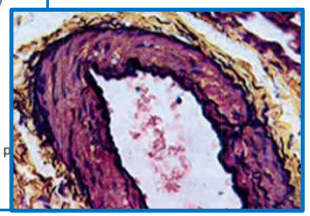


Cévní protéza ≥ 18 mm

- McGoon > 2.0
- Nakata > 250 mm2/m2

Plícní tkáň





Nízký tlak a odpor v plicním řeščišti

- střední tlak v plicnici < 19 mm Hg
- plicní cévní rezistence < 2 wj.m^2

Plicní cévní choroba 1. stupně

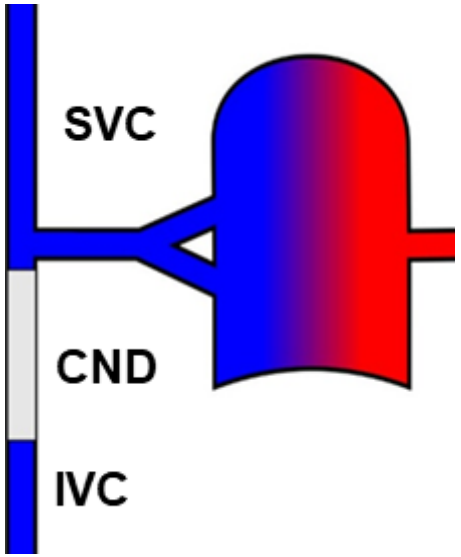
⇩

kontraindikace Fontanovy cirkulace

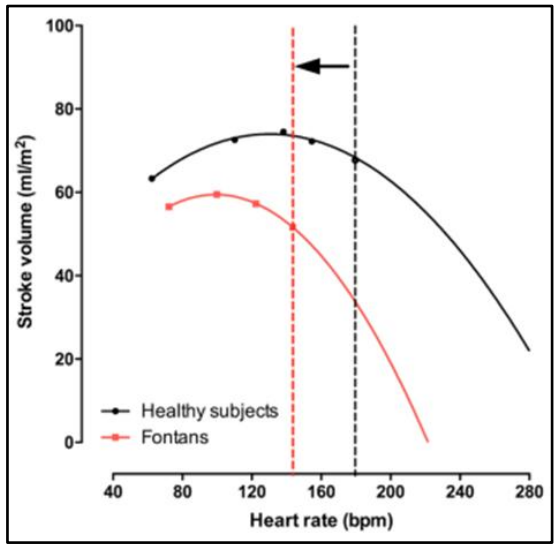
TCPC - limitace srdečního výdeje

Dlouhodobá prognóza ? ⟹

Neoportální systém



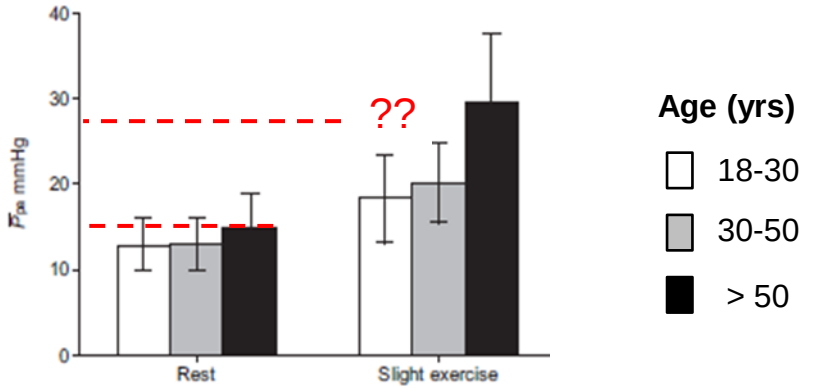
"Bottleneck"



Van De Bruaene, Gewillig 2022

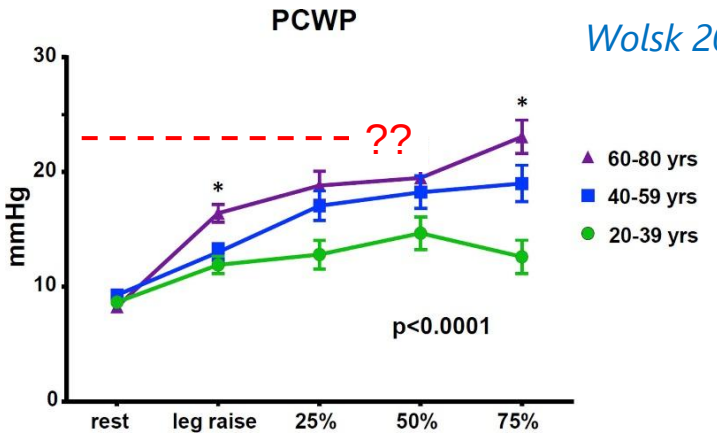
Pulmonary arterial pressure during rest and exercise in healthy subjects: a systematic review

Kovacs 2009



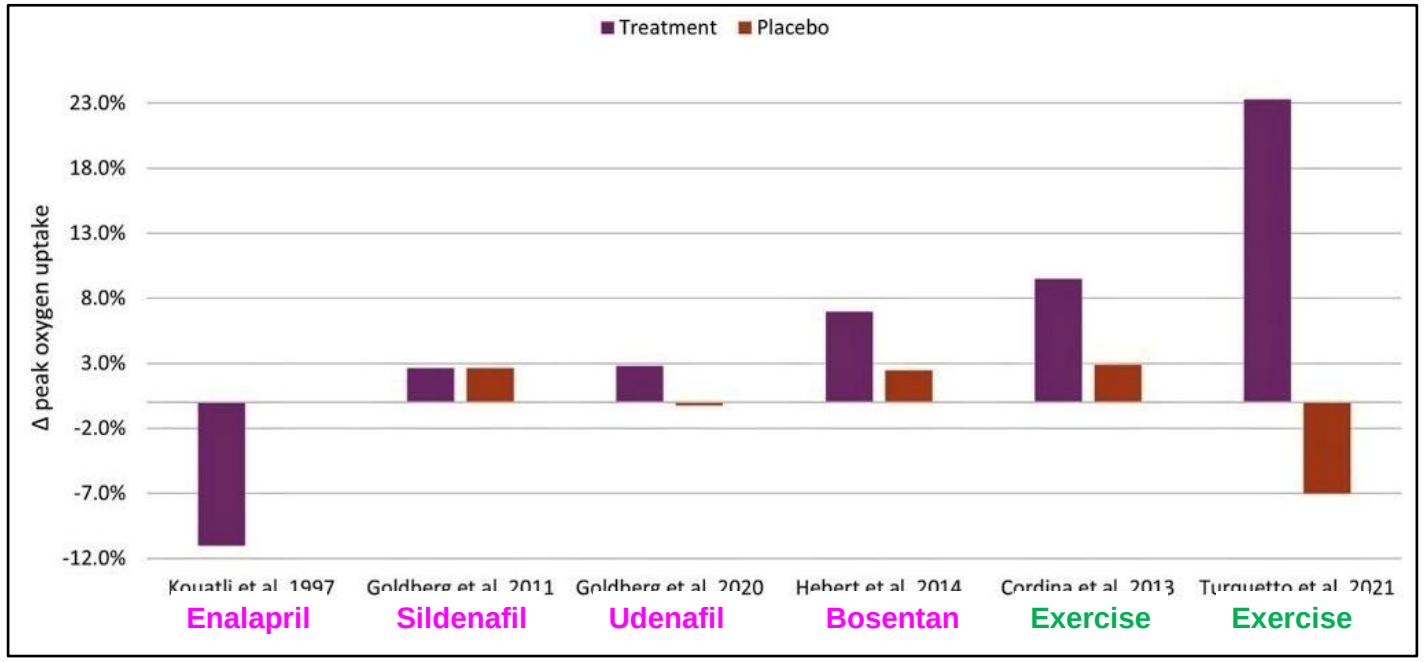
The Influence of Age on Hemodynamic Parameters During Rest and Exercise in Healthy Individuals

Wolsk 2017



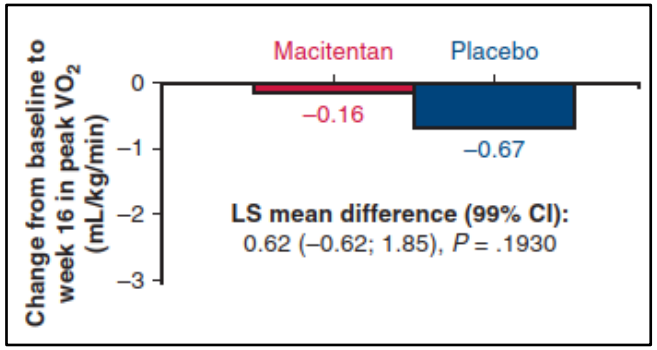
Fontanova cirkulace, vazodilatancia, fyzická výkonnost a cílený trénink

Exercise Intolerance, Benefits, and Prescription for People Living With a Fontan Circulation: The Fontan Fitness Intervention Trial (F-FIT)—Rationale and Design



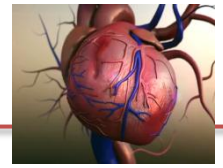
Tran 2021

Efficacy and safety of macitentan in Fontan-palliated patients: 52-week randomized, placebo-controlled RUBATO Phase 3 trial and open-label extension



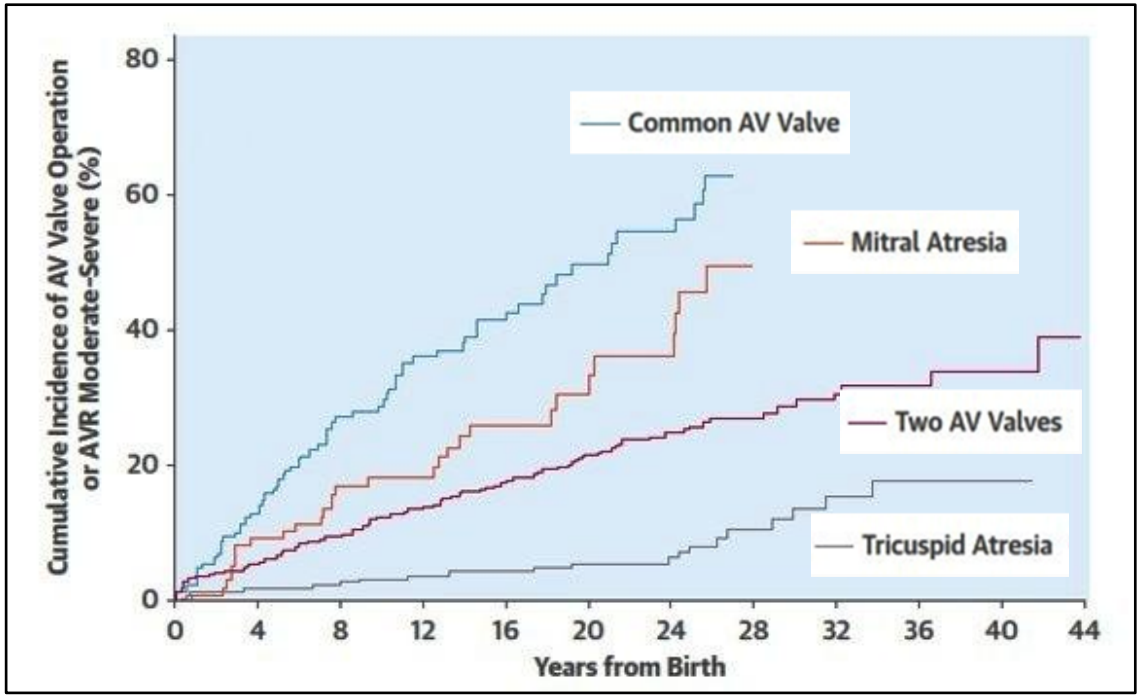
Clift 2025

TCPC - srdeční funkce a systémová cirkulace



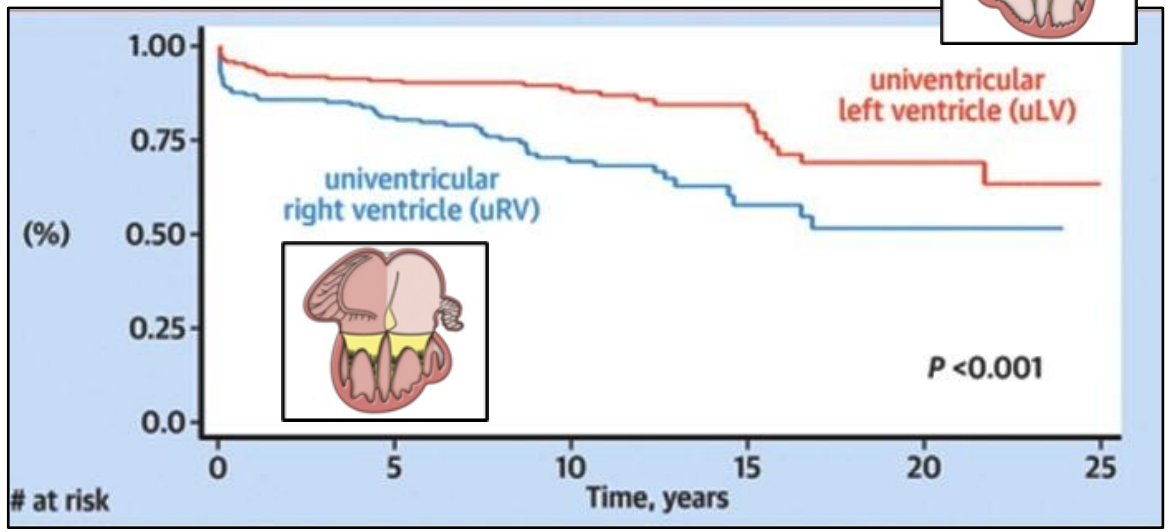
Základní morfologie a funkce vrozené srdeční vady

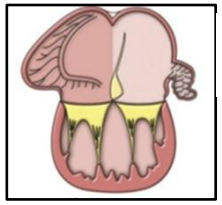
Srdeční chlopně



Upraveno podle King 2019

Dominance srdeční komory



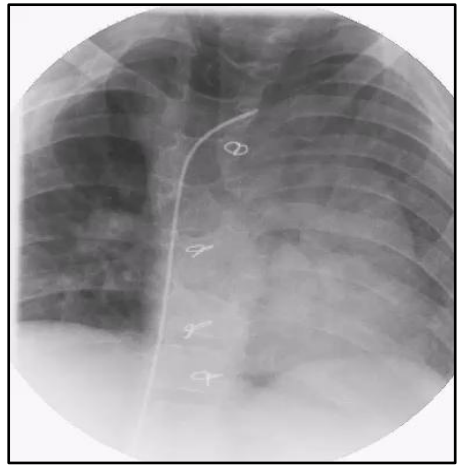


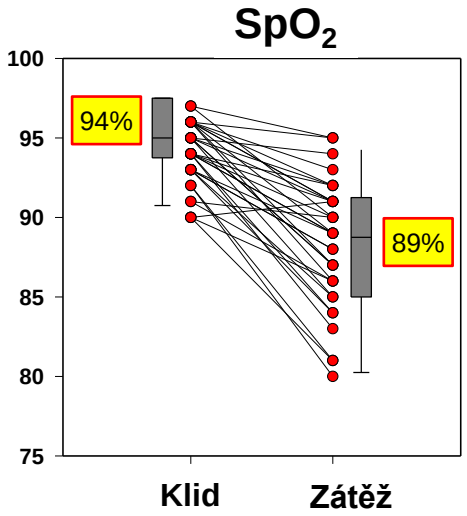
Indetermined ventricle ??

Upraveno podle Dib 2024

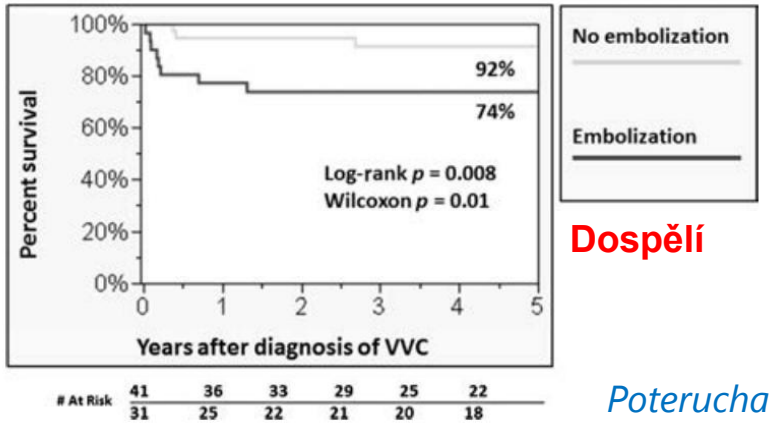
TCPC - kolaterály

Veno - venózní





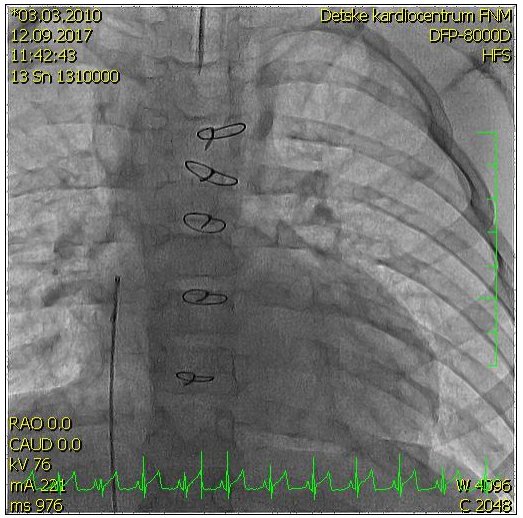
Embolization of Veno-venous Collaterals after the Fontan Operation Is Associated with Decreased Survival

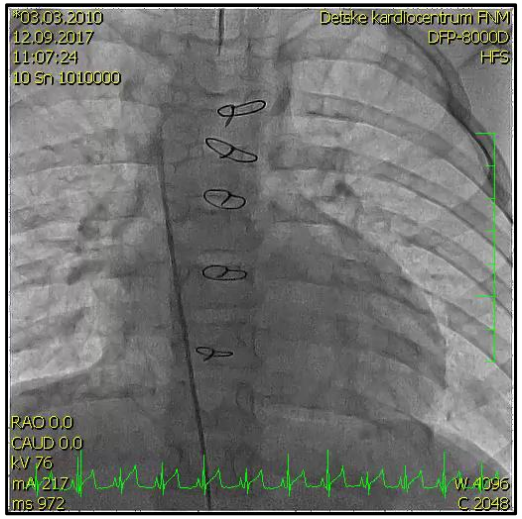


Dospělí

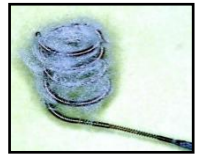
Poterucha 2015

Bronchiální / aortopulmonální





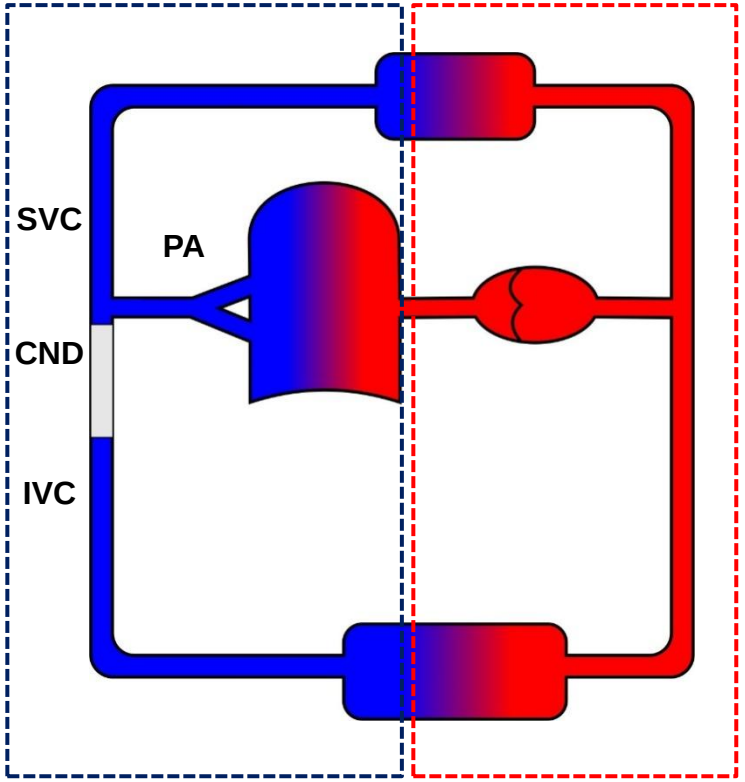
- zvyšují tlak v TCPC >> venózní městnání
- objemová zátěž >> srdeční selhání



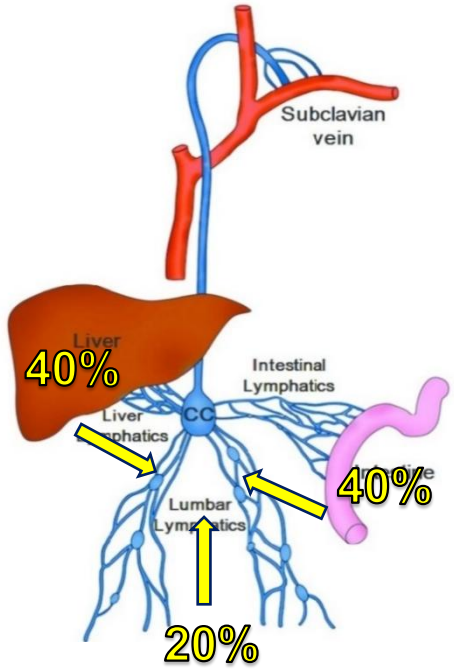
Patofyziologie Fontanovy cirkulace

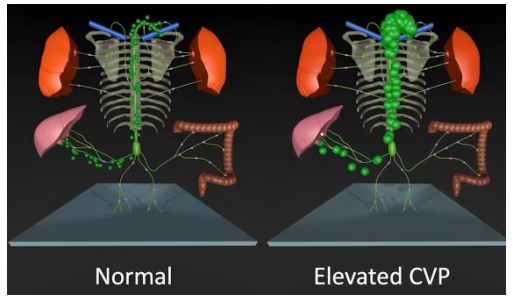
Neoportální systém

Systémová cirkulace

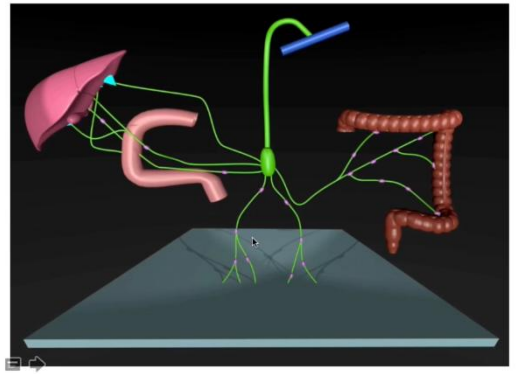


Lymfatický systém





Exudativní enteropatie

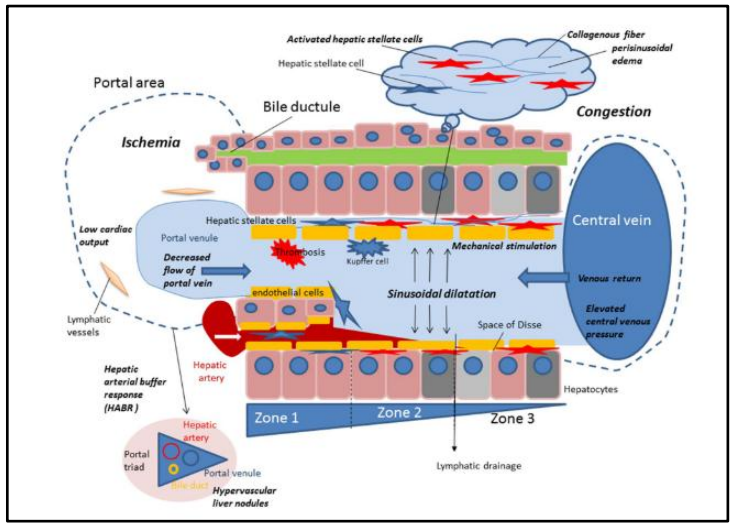


Dr. Yoav Dori: Children´s Hospital of Philadelphia

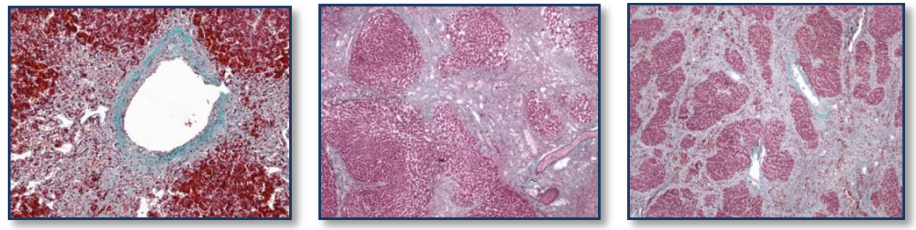
Fontan Associated Liver Disease (FALD)

Patofyziologie

- zvýšený žilní tlak
- hraniční srdeční výdej
- hypoxémie (v klidu, při zátěži)
- před / peri operační poškození jater
- faktor času
- ???



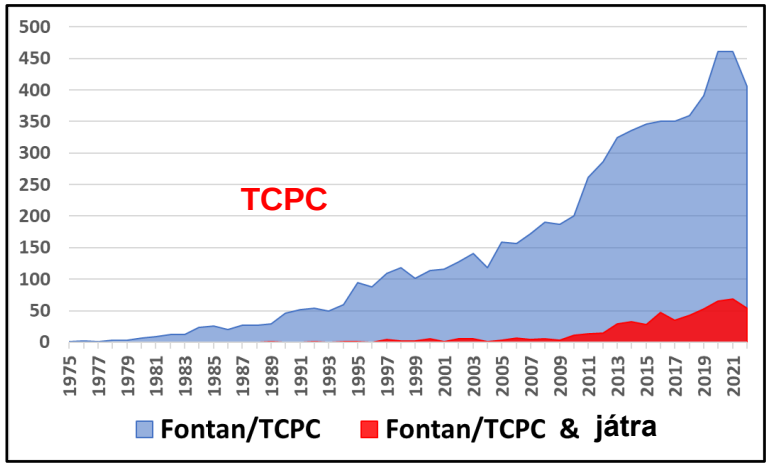
Sinusoidální fibróza | Portální fibróza | Cirrhóza



U dospělých

- obezita
- alkohol

Pubmed: počty publikací



Epidemiology of Fontan-associated liver disease in Japan: Results from a nationwide survey in 2021

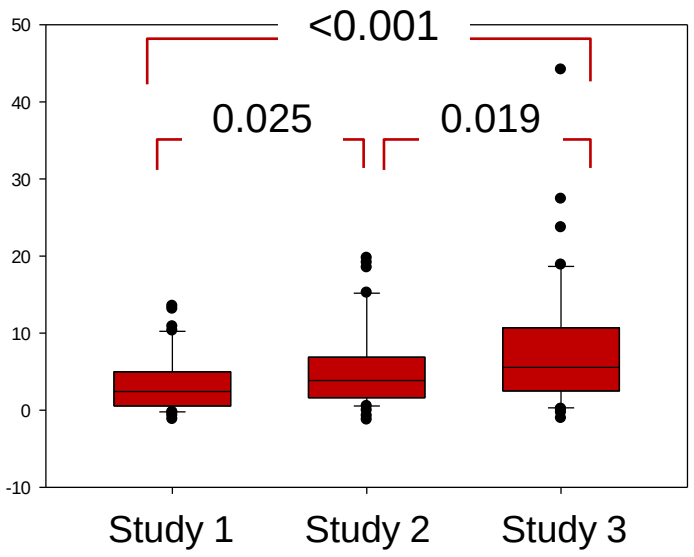
FALD: 3 120 / 7 810 **(40%) pacientů**
Interval od operace: medián 14 let

Ohfuji 2024

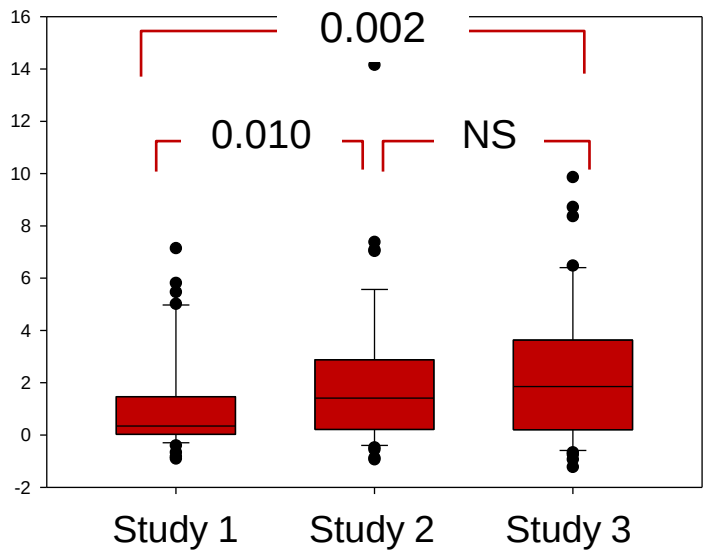
Children´s Heart Centre: TCPC longitudinal study

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

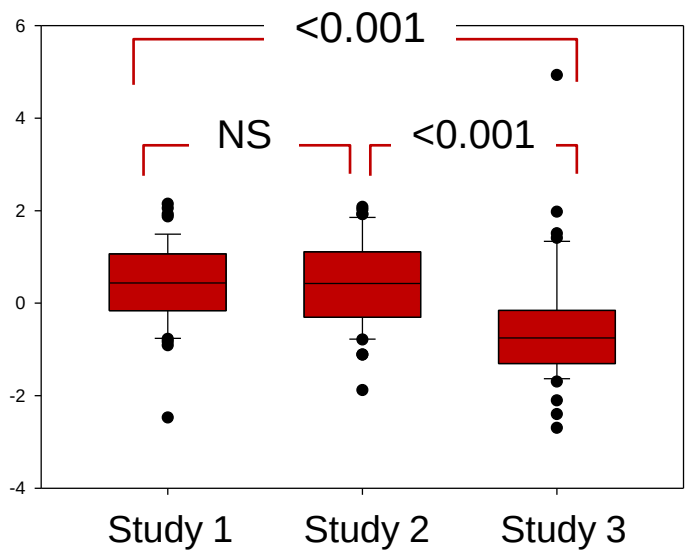
GGT (Z-score)



Bilirubin (Z-score)



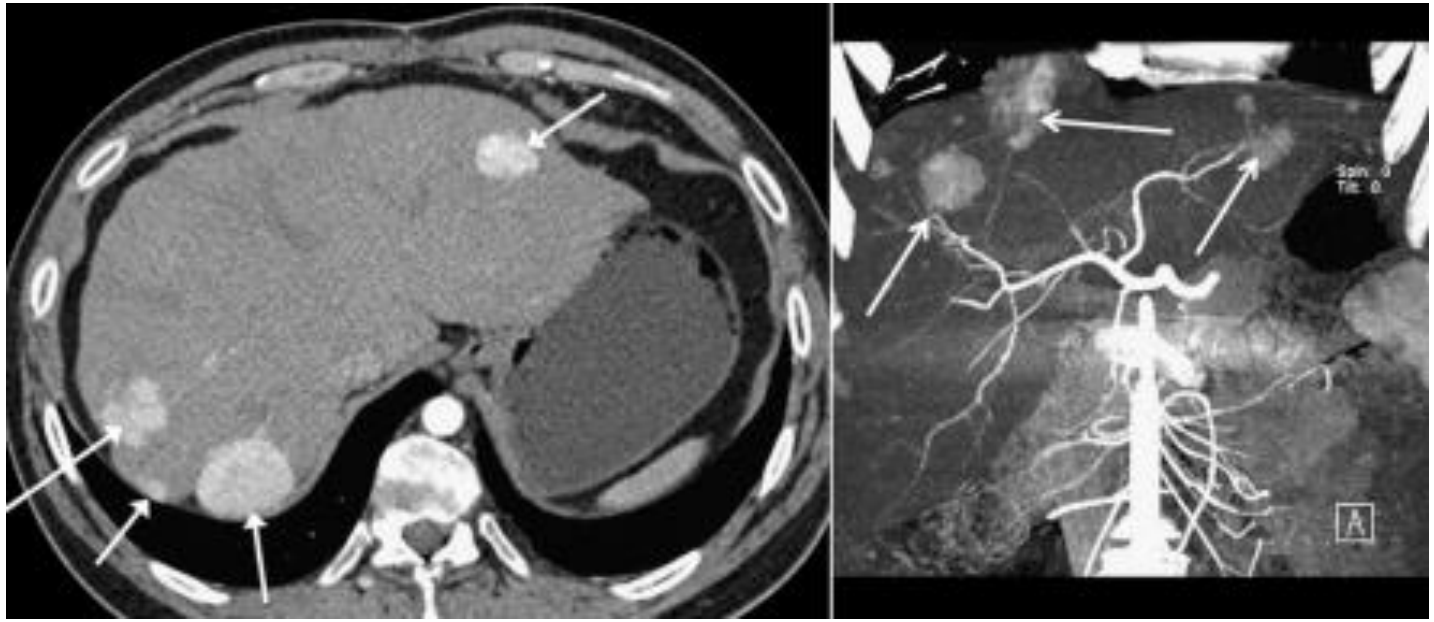
Prealbumin (Z-score)



D. Jičínská

Advanced stage of FALD
Hepatico-cardiac interactions

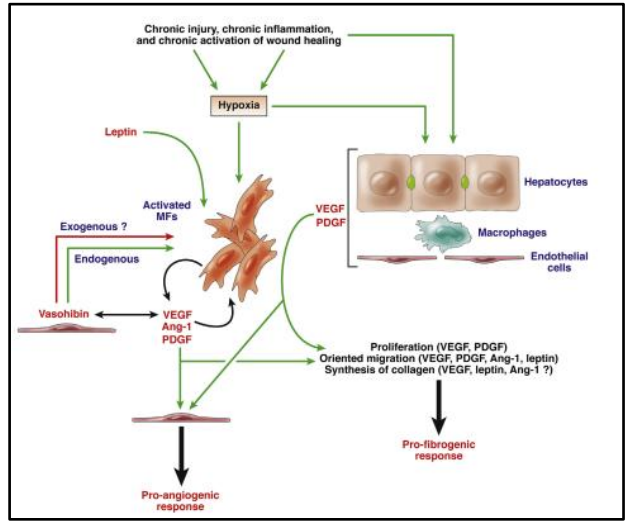
Liver nodules



Prevalence 35% - 48%

Compensatory arterialization of liver parenchyma

Angiogenesis and Fibrogenesis in Chronic Liver Diseases



Bocca 2015

Hepatocellular carcinoma

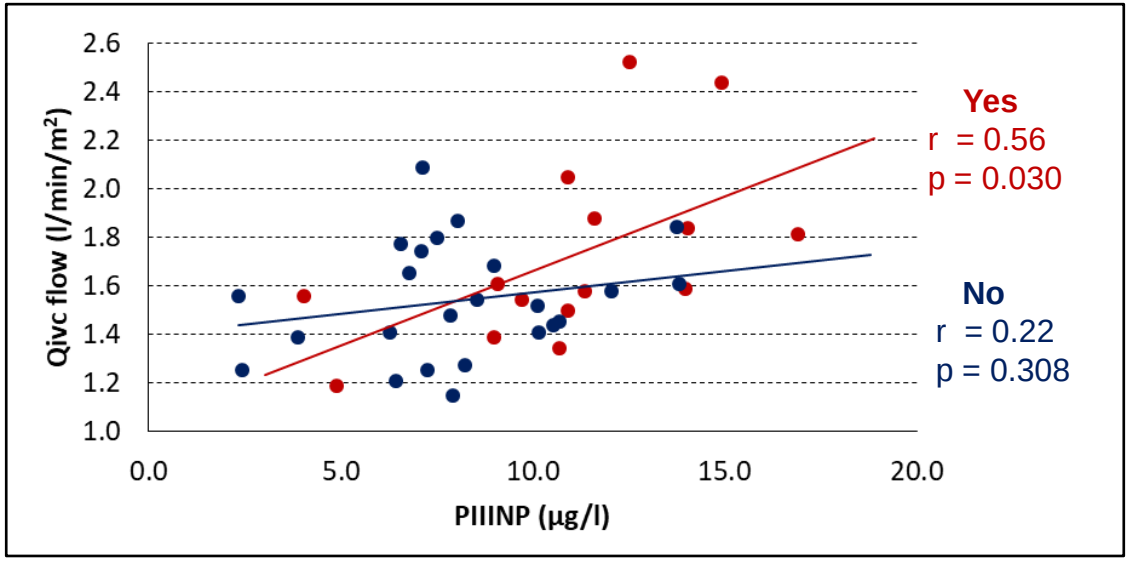
Cumulative incidence 10, 20, and **30 years** postoperatively
0.8, 2.9, and **13.3%**

Téllez 2023

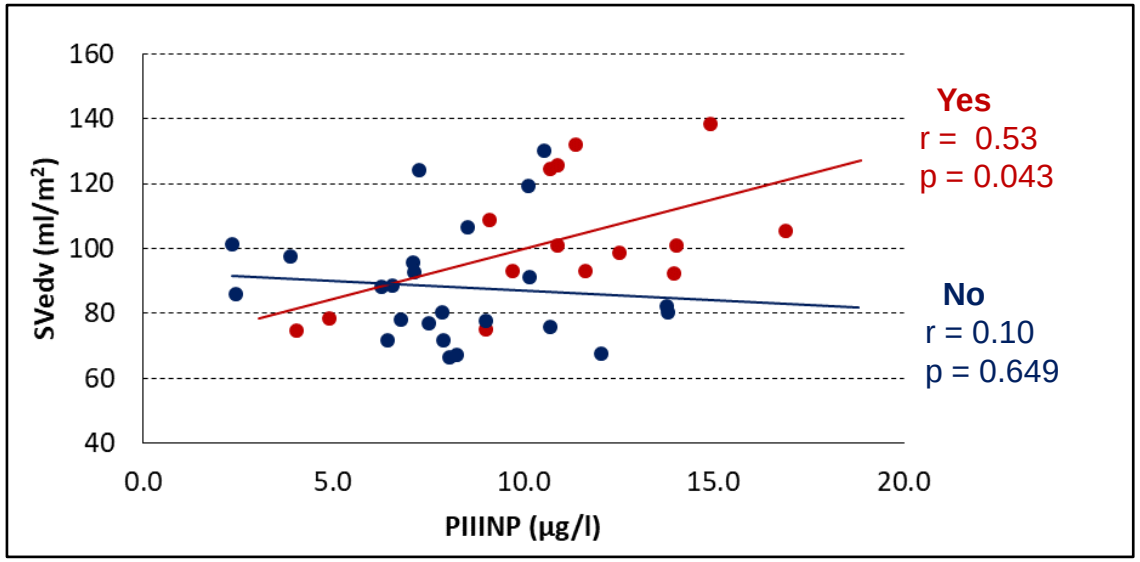
Impact of advanced liver fibrosis on TCPC hemodynamics

Liver nodules: in 15 / 39 (38%) pts
Age at study: 26 (IQR 23 - 28) years
Interval from TCPC: 21 (IQR 20 - 23) years

Inferior vena cava flow



End-diastolic ventricular volume



Chaloupecký 2022

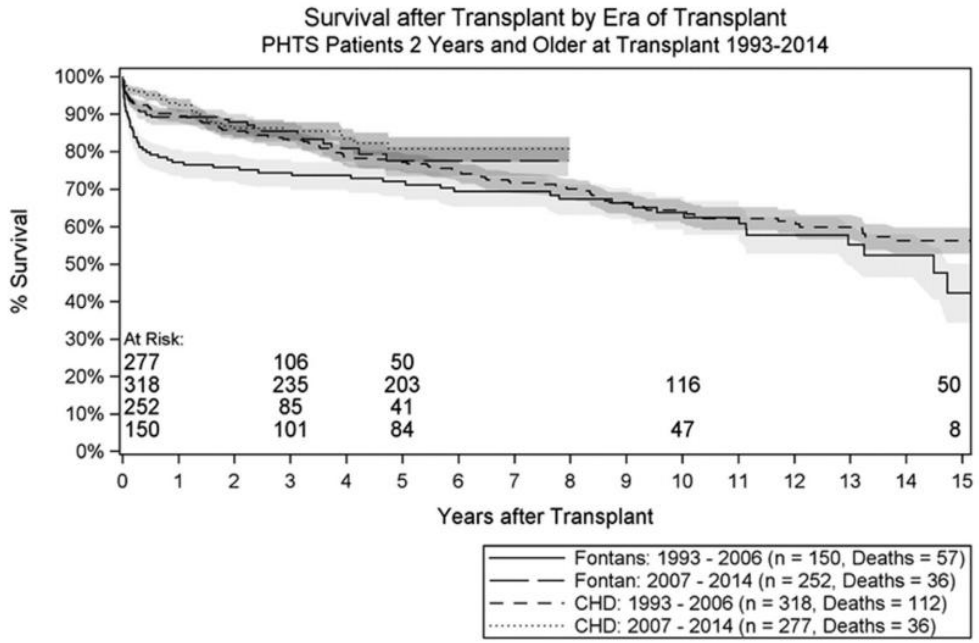
Možnosti léčby těžkého srdečního selhání Fontanovské cirkulace

Dlouhodobá podpora systémové cirkulace

Transplantace srdce

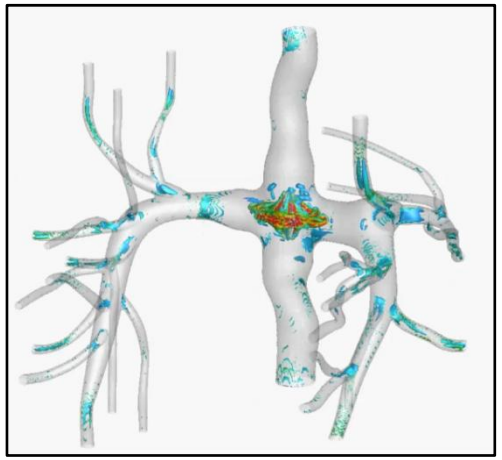
Fontan Patient Survival After Pediatric Heart Transplantation Has Improved in the Current Era

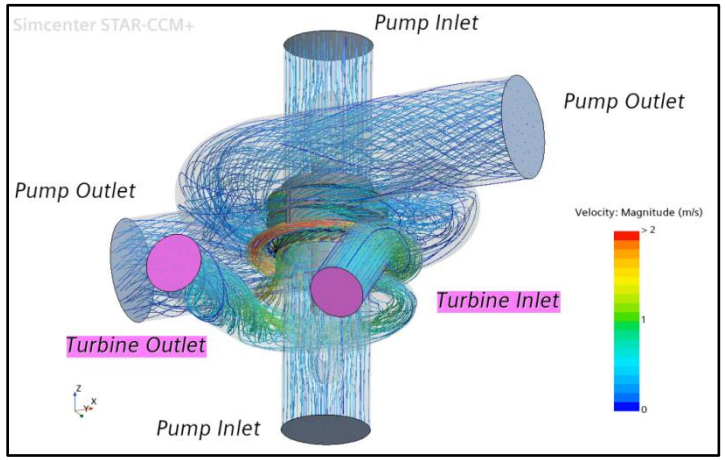
Kathleen E. Simpson, MD, Elizabeth Pruitt, MSPH, James K. Kirklin, MD, David C. Naftel, PhD, Rakesh K. Singh, MD, MS, R. Erik Edens, MD, PhD, Aliessa P. Barnes, MD, and Charles E. Canter, MD

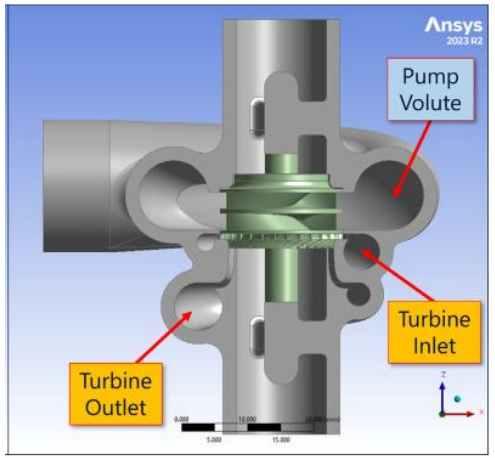


Možnosti podpory neoportálního systému

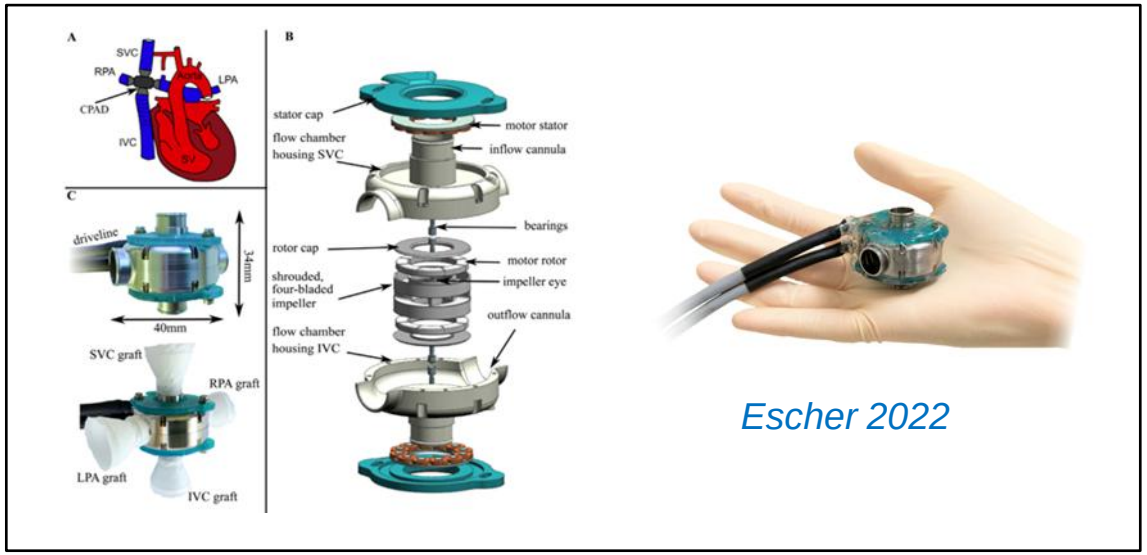
Cíl: snížit alespoň částečně tlak v dolní duté žíle >> zpomalit / zastavit vývoj FALD







Rodefeld 2024



Escher 2022

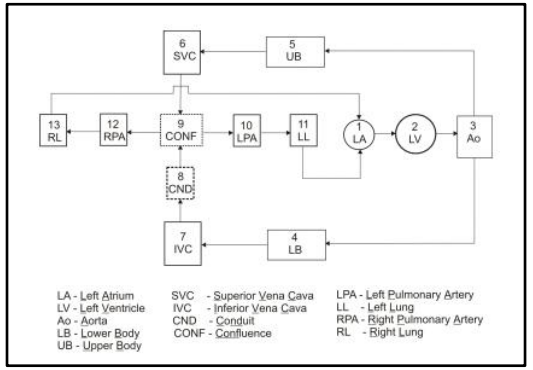
Možnosti podpory neoportálního systému

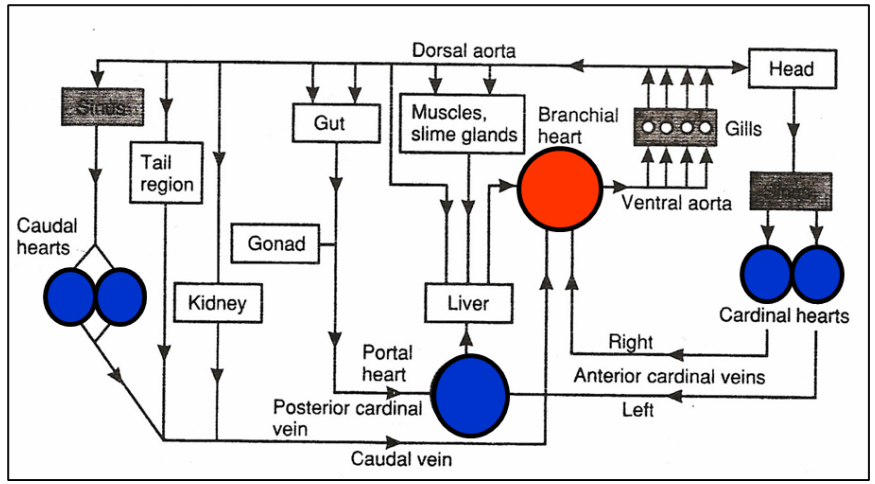
Hagfish (sliznatka)

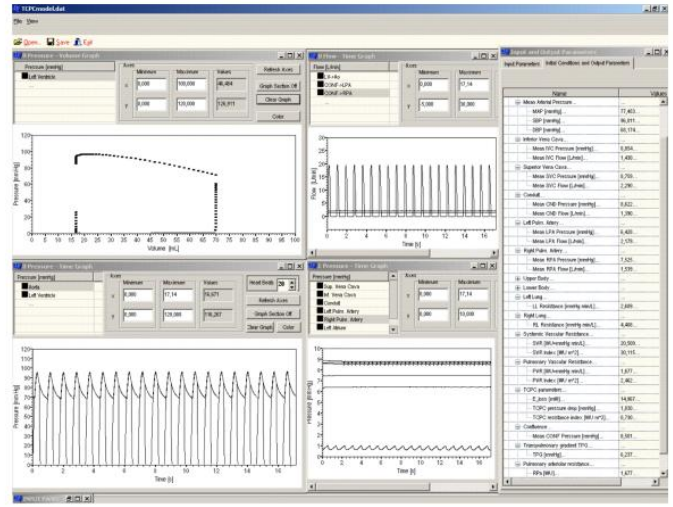
Numerical simulation of TCPC geometry effects on hemodynamics of single ventricle circulation

Světlana Převorovská, František Maršík, Václav Chaloupecký

Institute of Thermomechanics CAS **2008**



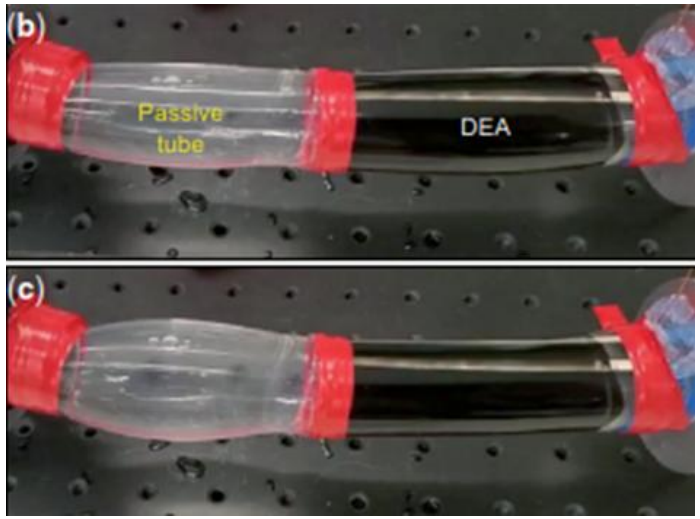


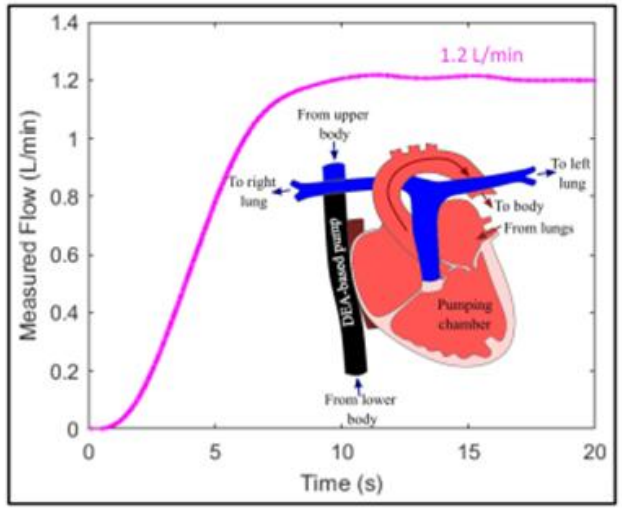


Možnosti podpory neoportálního systému

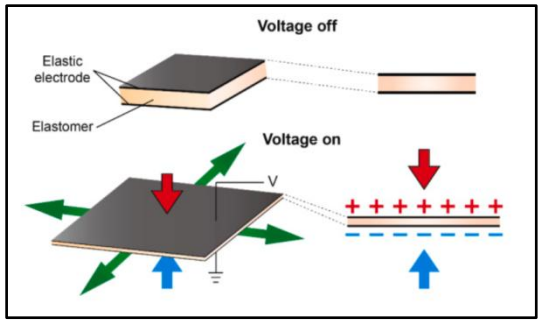
Dielectric Elastomer Actuators (DEAs)

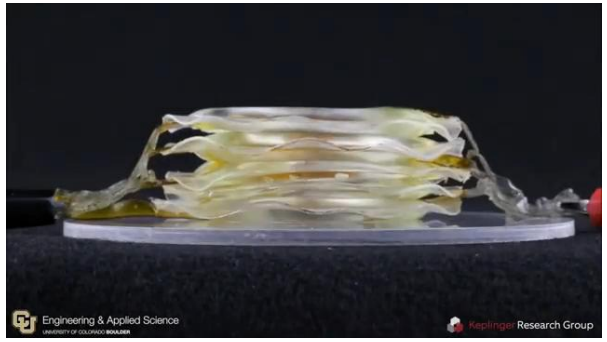
Dielectric elastomer actuator-based valveless pump as Fontan failure assist device: introduction and preliminary study

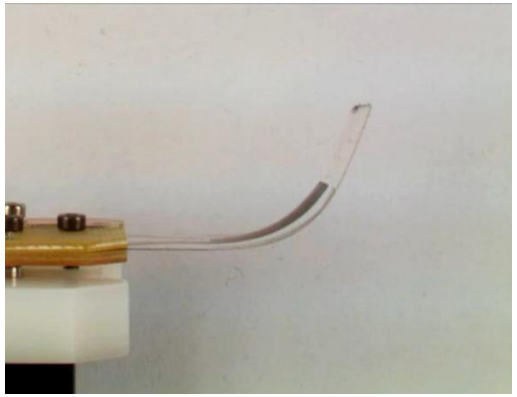




Benouhiba 2024

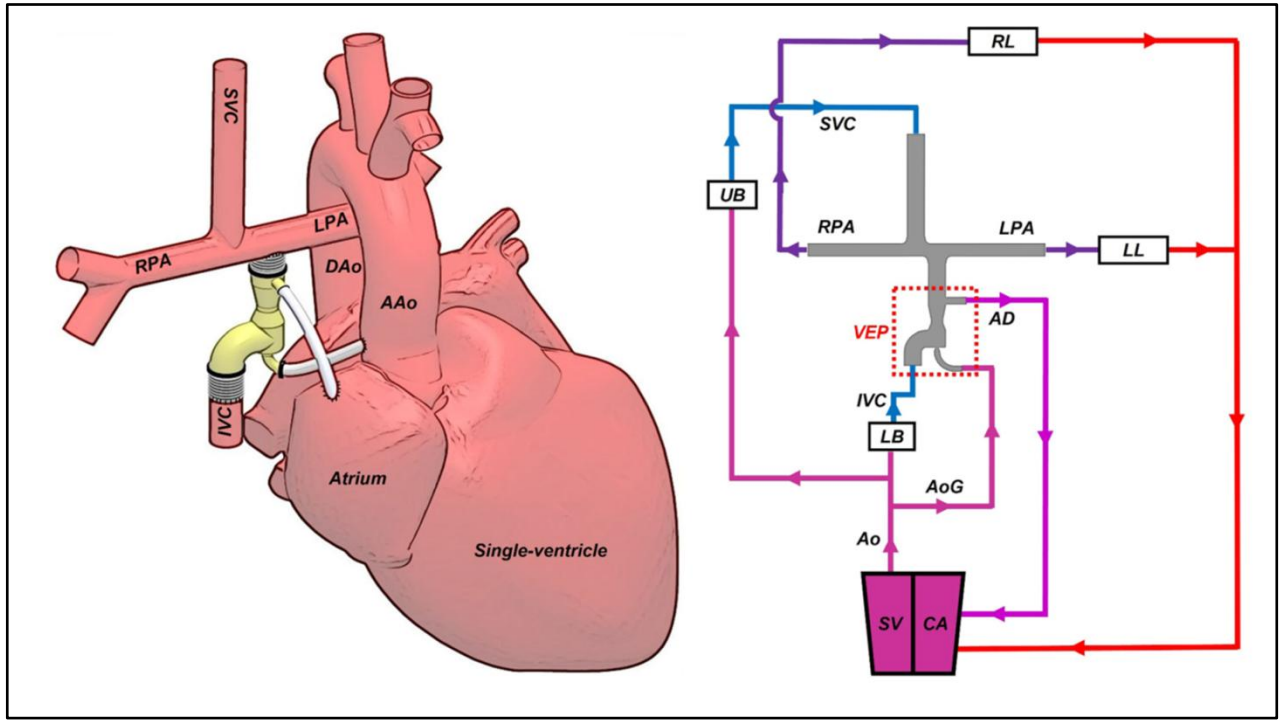




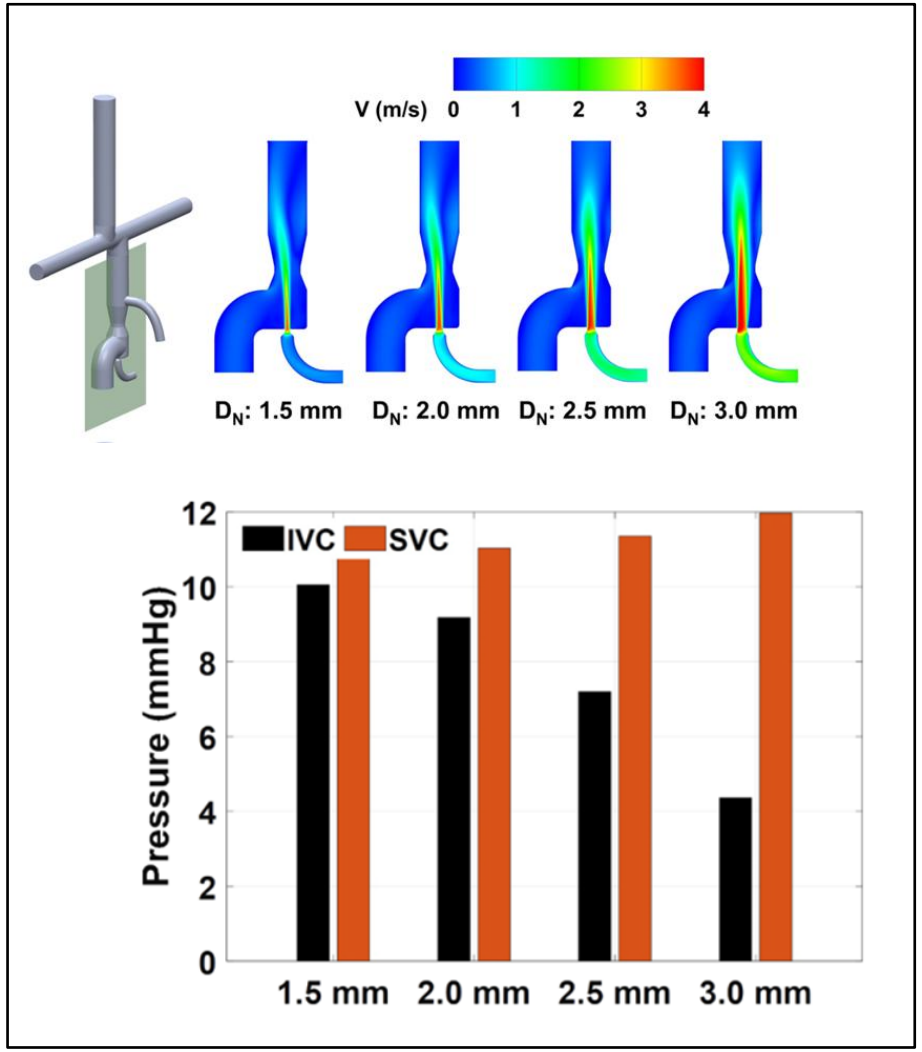


Možnosti podpory neoportálního systému

In Silico Evaluation of a Self-powered Venous Ejector Pump for Fontan Patients



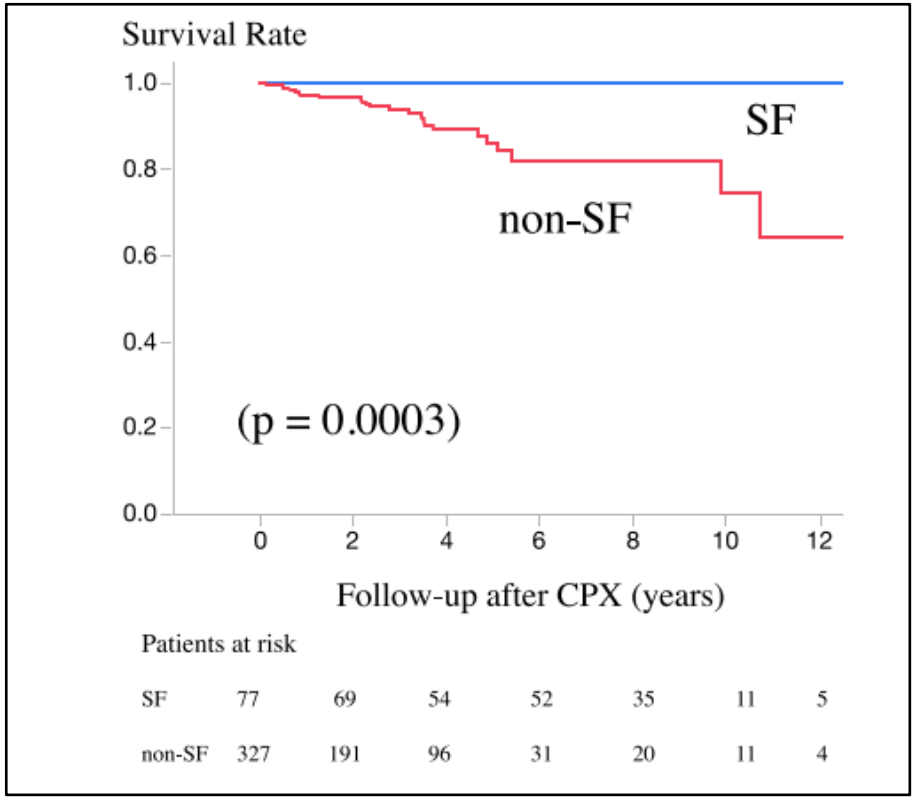
Rasooli 2023



"Super Fontan"

Prevalence and clinical correlates and characteristics of "Super Fontan"

Exercise capacity ≥ 80% predicted normal value



77 (19%)

327 (81%)

Ohuchi 2023

Multidisciplinární celoživotní sledování jedinců s Fontanovou cirkulací

Specializovaná centra /ambulance

kardiolog
kardiochirurg
rentgenolog
hepatolog
hematolog

Doporučení pro jedince > 14 let věku

Vyšetření	Interval (roky)
ECHO Holter Zátěž	2
Sono jater Elastografie	2
CMRI (funkce, průtoky) MRI jater	4

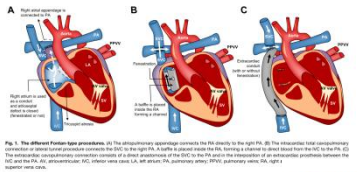
Laboratorní vyšetření		Interval (roky)
Biochemie	Iontogram + P, Ca metabolismus AST, ALT, GGT, ALP, BIL Urea, kreatinin, k. močová, cystatin C Celk. bílkovina, albumin, prealbumin NTproBNP ELF test Alfa-fetoprotein (při patol. sono jater)	2
Hematologie	KO+diff+trombo	
	APTT, QUICK, INR, TT Fibrinogen Antitrombin Faktor V, VII, VIII Protein C, S	
Trombofilní markery	Při vstupu dítěte do programu jednokomorové cirkulace	

Upraveno podle Rychik 2022, Téllez 2023


Keep calm
and
believe in science

EASL-ERN position paper on liver involvement in patients with Fontan-type circulation

Luis Téllez[1,2], Audrey Payancé[3,4], Eric Tjwa[5], María Jesús del Cerro[6,7], Lars Idorn[8], Stanislav Ovroutski[9], Ruth De Bruyne[10,†], Henkjan J. Verkade[11,†], Fabrizio De Rita[12], Charlotte de Lange[13], Annalisa Angelini[14], Valérie Paradis[15,16], Pierre Emmanuel Rautou[17,18,†], Juan Carlos García-Pagán[19,20,*]



European Association for The Study of the Liver (EASL) and the European Reference Network
on Rare Liver Diseases (ERN RARE-LIVER)

Clinical evaluation and staging of Fontan-associated liver disease

Statements

- FALD is typified by gross architectural distortion, massive sinusoidal dilation, and perisinusoidal fibrosis in the absence of significant parenchymal inflammation (**High LoE**).
- In patients with FALD, liver fibrosis is nearly universal and distinctively involves both centrilobular and portal areas (**High LoE**).
- The patchy distribution of fibrosis in FALD may lead to underestimation of its stage (**Moderate LoE**).

What are the typical laboratory abnormalities of FALD? Are serum biomarkers useful for the diagnosis and staging of FALD?

Statements

- A mild increase in serum gamma-glutamyltransferase is the most common and early laboratory liver abnormality in Fontan patients. Slight elevations of serum bilirubin may also be present (**High LoE**).
- Most scores (*i.e.*, model for end-stage liver disease [MELD], MELD-Na, and Child-Pugh classification) may not be accurate to stage liver disease severity in this population (**Moderate LoE**).

Recommendations

- Serological and combined serum-clinical tests used in other liver aetiologies (aspartate aminotransferase-to-platelet ratio index [APRI], ELF, FIB-4, Forns index, and FibroTest®) have a modest discriminatory power in identifying patients with severe fibrosis and cannot be routinely recommended for staging the liver disease (**Low LoE, strong recommendation**).

Statement

- Nodular hepatocellular lesions of different sizes are common in FALD and are mostly benign (**Low LoE**).

Recommendations

- Compared with the age-matched population of HCC in Fontan patients is increased, and programme is recommended (**Low recommendation**).
- HCC surveillance should be initiated 10 year type surgery and strongly considered earlier tan circulatory failure (**Low LoE, strong reco**
- Surveillance should be performed by personnel using abdominal ultrasound exam months. This should be complemented by contrast-enhanced imaging every 1-2 ye **LoE, strong recommendation**).

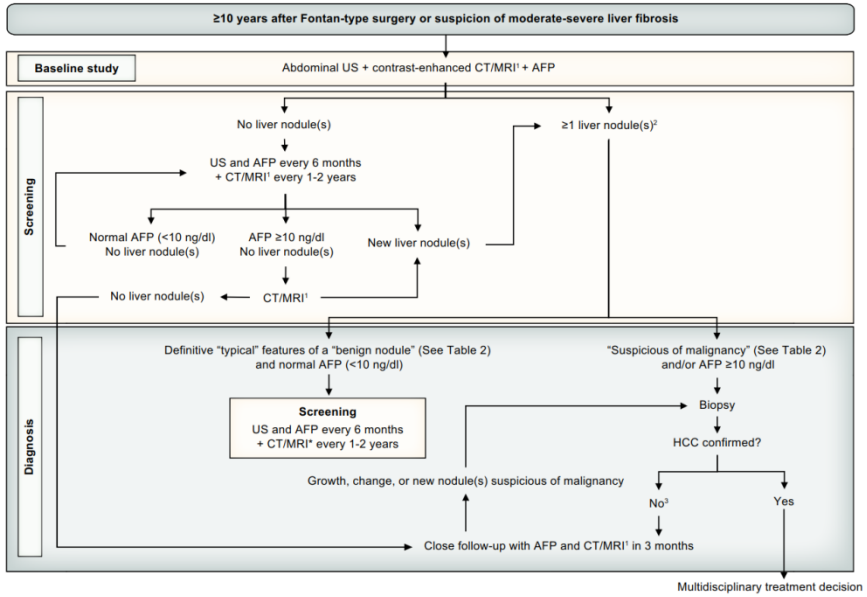


Fig. 3. Flow diagram for evaluation of liver nodules and HCC surveillance and diagnosis in FALD. (1) Multiphasic contrast-enhaced CT/multiphasic contrast-enhaced MRI, or (preferable) gadoxetic-enhaced MRI; (2) The optimal management for nodules <1 cm showing typical HCC patterns has yet to be clarified, and a

Dielectric elastomer actuator-based valveless pump as Fontan failure assist device: introduction and preliminary study

Amine Benouhiba [a,*], Armando Walter [a], Silje Ekroll Jahren [a,b], Thomas Martinez[a], Francesco Clavica [a,b], Paul Philipp Heinisch [c,d], Dominik Obrist [b], Yoan Civet [a] and Yves Perriard [a]

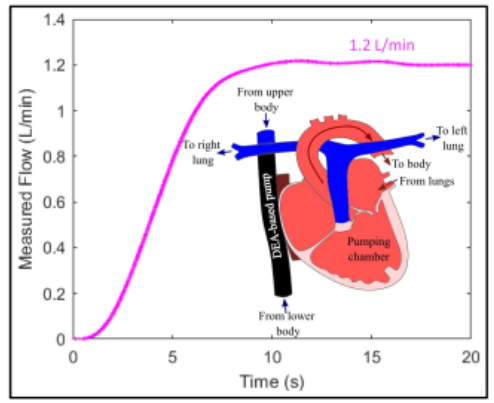
[a] Integrated Actuators Laboratory (LAI), École polytechnique fédérale de Lausanne (EPFL), Neuchâtel, Switzerland
[b] ARTORG Center for Biomedical Engineering Research, University of Bern, Bern, Switzerland
[c] Division of Congenital and Pediatric Heart Surgery, University Hospital of Munich, Ludwig-Maximilians-University, Munich, Germany
[d] Department of Congenital and Pediatric Heart Surgery, German Heart Center Munich, Technical University of Munich, Munich, Germany

* Corresponding author. Integrated Actuators Laboratory (LAI), École polytechnique fédérale de Lausanne (EPFL), Rue de la Maladière 71 B, 2000 Neuchâtel, Switzerland. Tel: +41-21-693-80-89; e-mail: amine.benouhiba@epfl.ch (A. Benouhiba).

Novel dielectric elastomer actuator pump as a substitute for the Fontan conduit

Summary

This *in-vitro* study explores the viability of a bladeless impedance-driven cavopulmonary assist device utilizing dielectric elastomer actuators (DEA) as a potential solution for Fontan failure. The DEA-based pump undergoes testing within a closed-loop setup, demonstrating its capacity to produce substantial flow rates across various pressure levels.



Legend: Novel DEA-based pump as a substitute for the Fontan conduit and *in-vitro* achieved flow rate.

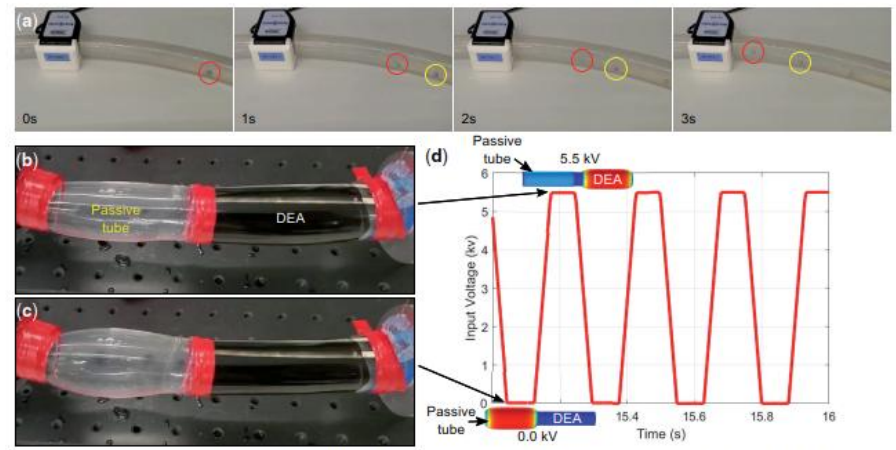


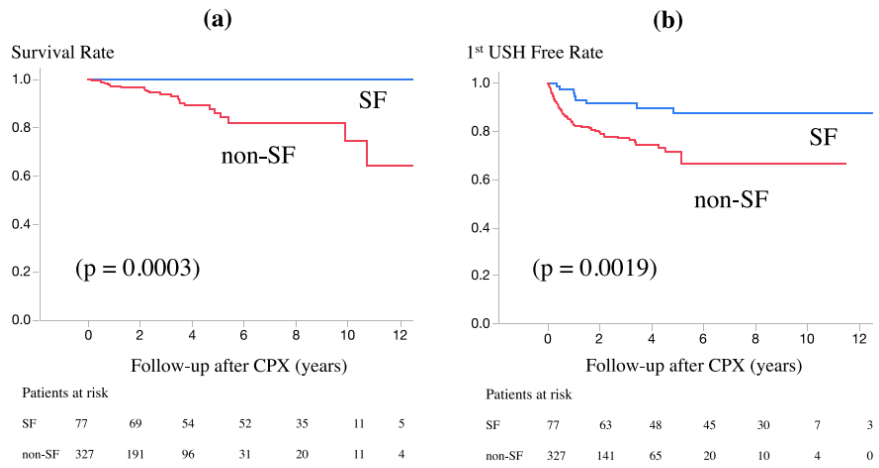
Figure 4: The working behaviour of the DEA pump observed at the resonant frequency of 4 Hz and an internal pressure of 15 mmHg. (**a**) A time-lapse representation of particles' movement within the fluid during the activation process, (**b**) DEA pump at 5.5 kV, (**c**) DEA pump at 0 kV and (**d**) the applied voltage signal. DEA: dielectric elastomer actuator.

Prevalence and clinical correlates and characteristics of "Super Fontan"

Hideo Ohuchi, MD, PhD [a,b], Aki Mori, MD [a,b], Kenichi Kurosaki, MD, PhD [a], Isao Shiraishi, MD, PhD [a], and Michikazu Nakai, PhD [c] *Osaka, Japan*

have been conducted and the beneficial effect of improving exercise capacity has been demonstrated.[2] On the other hand, Fontan patients with normal exercise capacity (≥80% predicted normal value) have been defined as "Super-Fontan (SF)"[3]; they are an excellent clinical phenotype that should be the optimal therapeutic target in our practice.

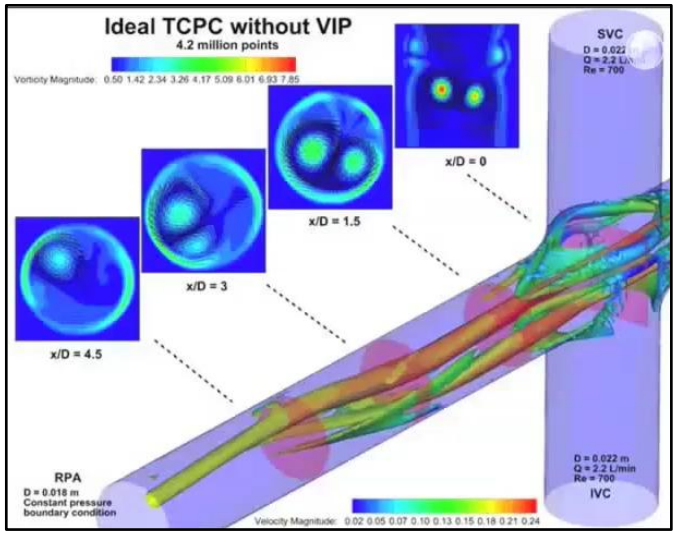
Figure 2



Comparison of survival curves **A,** and 1st unscheduled hospitalization (USH) free curves **B,** between SF and non-SF patients.

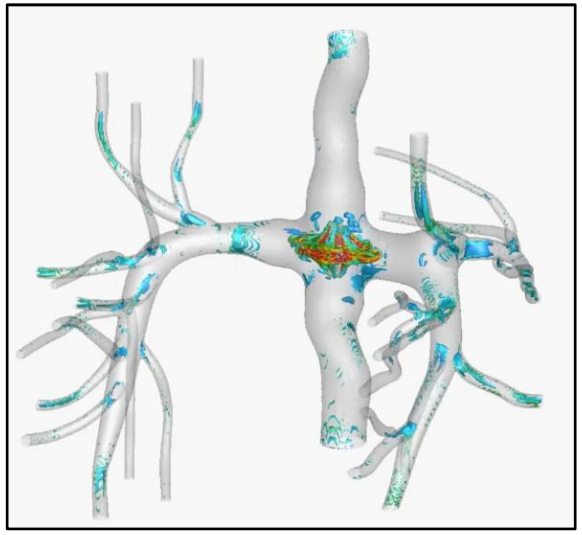
Subpulmonary pump

TCPC
Large Edy Simulation



https://www.youtube.com/watch?v=0FE3sEDWcCg

TCPC pump
Computational surgery



https://www.youtube.com/watch?v=G3ie15dcZPk

NASA prototype
bi-conical heart pump

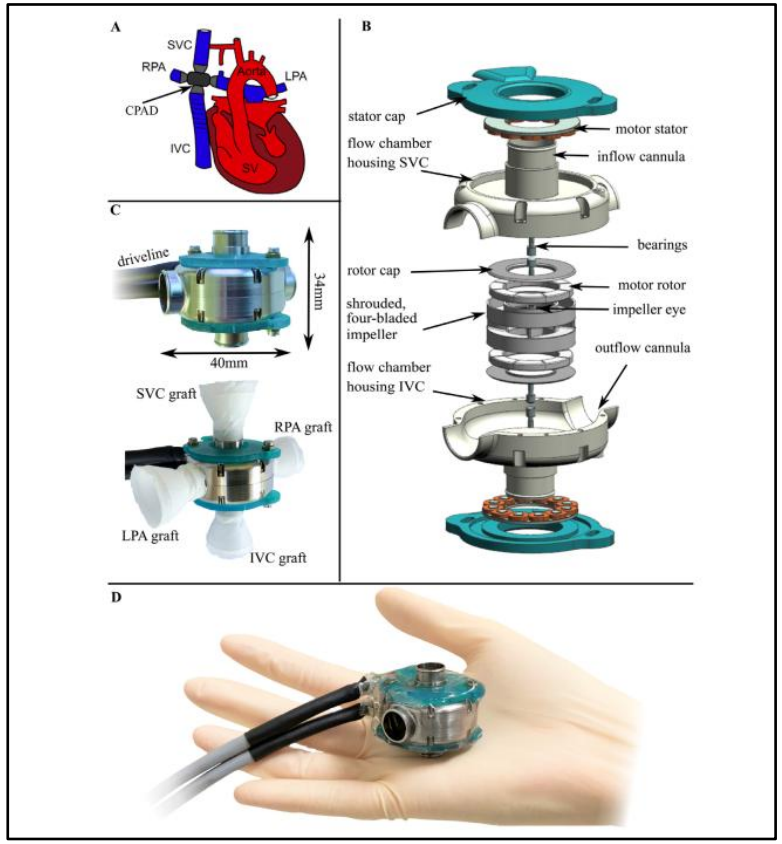
https://www.nasa.gov/feature/nasa-glenn-technology-pumps-hope-into-broken-hearts

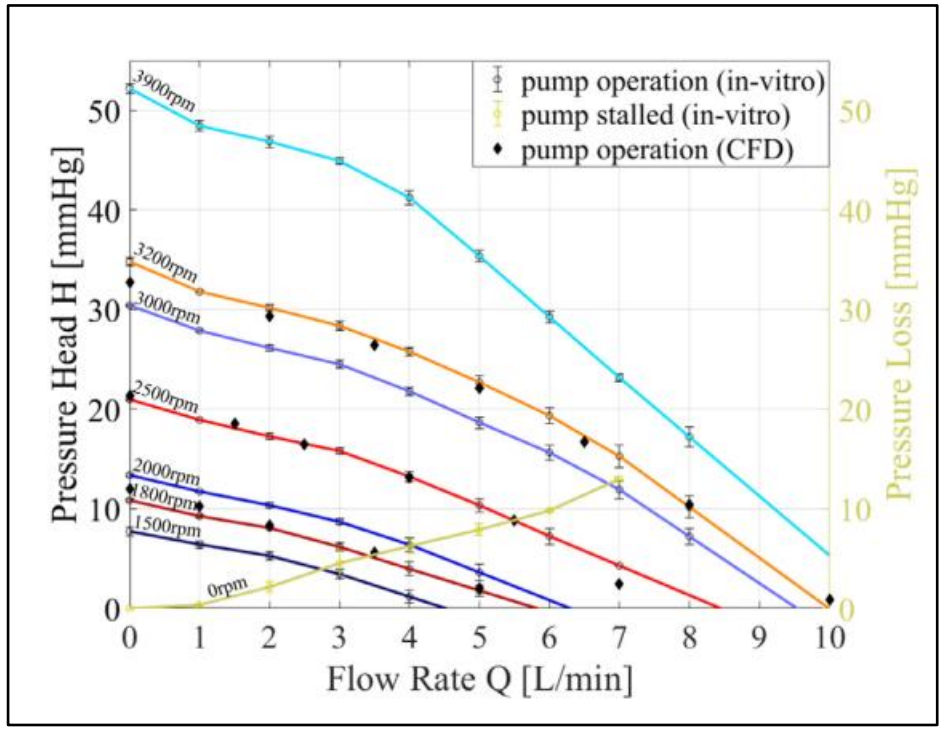
Steven Frankel

A Cavopulmonary Assist Device for Long-Term Therapy of Fontan Patients

Andreas Escher, MSc,[,†] Carsten Strauch, MSc,[‡] Emanuel J. Hubmann, MSc,[§] Michael Hübler, MD,[¶] Dominik Bortis, PhD,[§] Bente Thamsen, PhD,[*] Marc Mueller, PhD,[**] Ulrich Kertzscher, PhD,[*] Paul U. Thamsen, PhD,[‡] Johann W. Kolar, PhD,[§] Daniel Zimpfer, MD,[†] and Marcus Granegger, PhD[*,†]*

Semin Thorac Cardiovasc Surg 2022



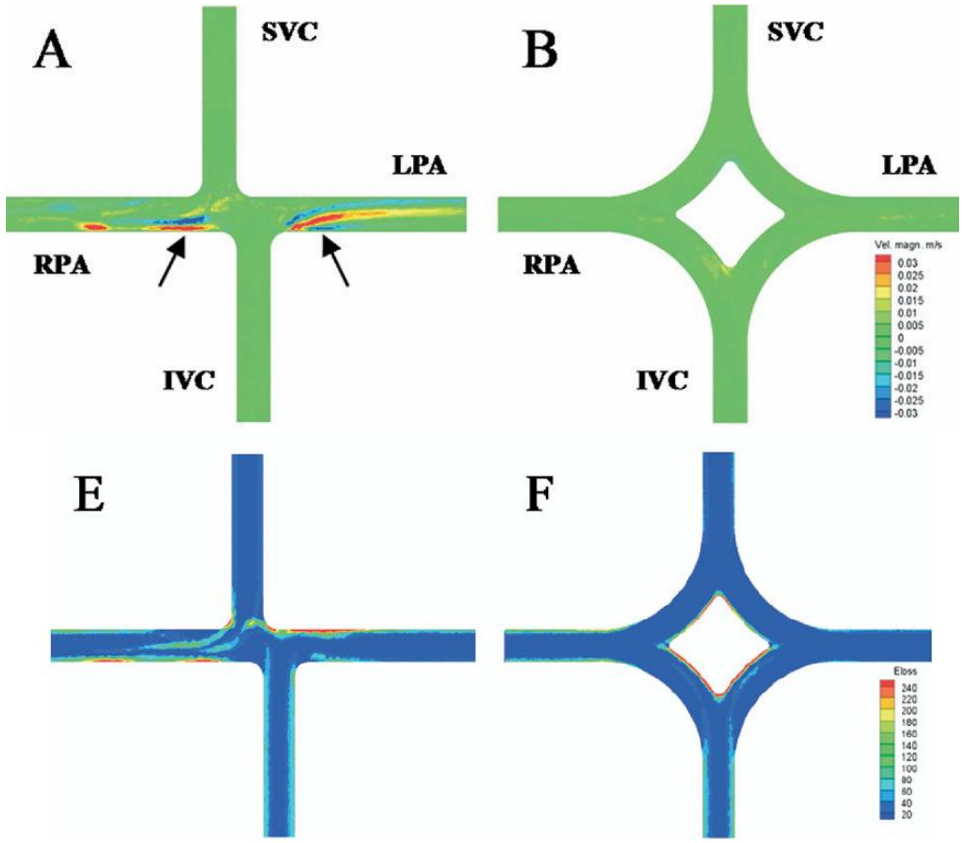


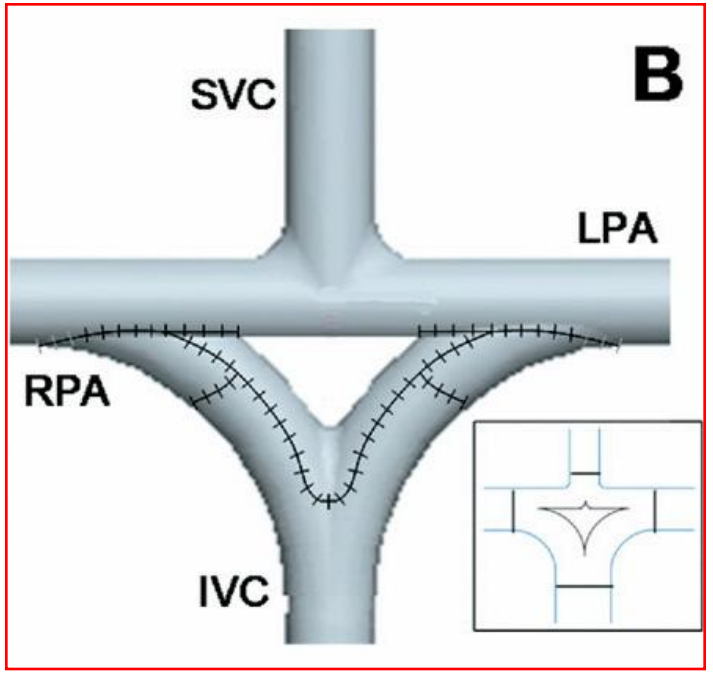
Introduction of a New Optimized Total Cavopulmonary Connection

Dennis D. Soerensen, MS, Kerem Pekkan, PhD, Diane de Zélicourt, MS,
Shiva Sharma, MD, Kirk Kanter, MD, Mark Fogel, MD, and Ajit P. Yoganathan, PhD

Ann Thorac Surg 2007;83:2182–90

bifurcated Y - graft "Optiflo"

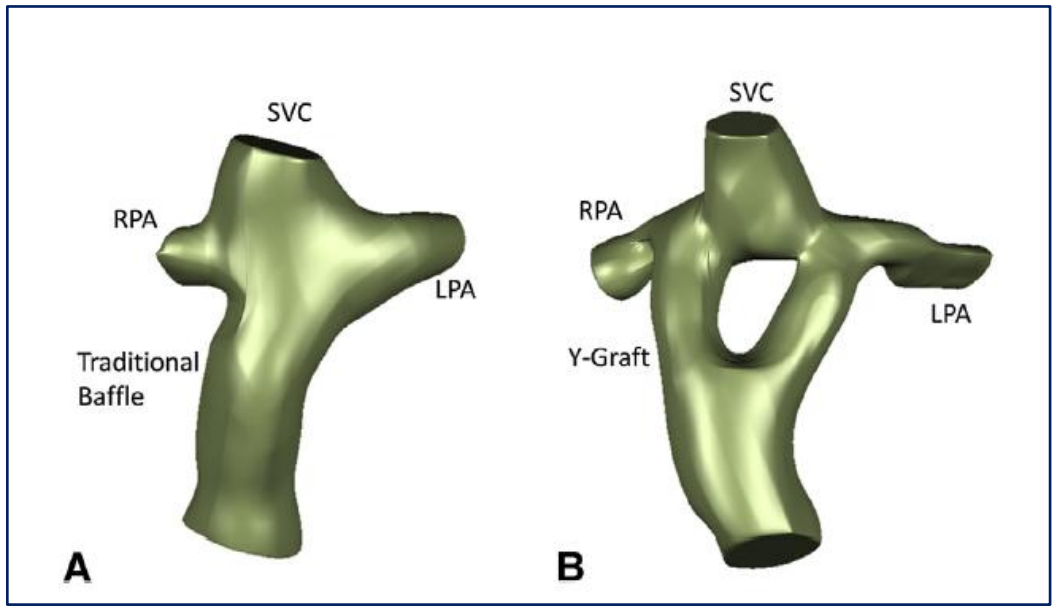


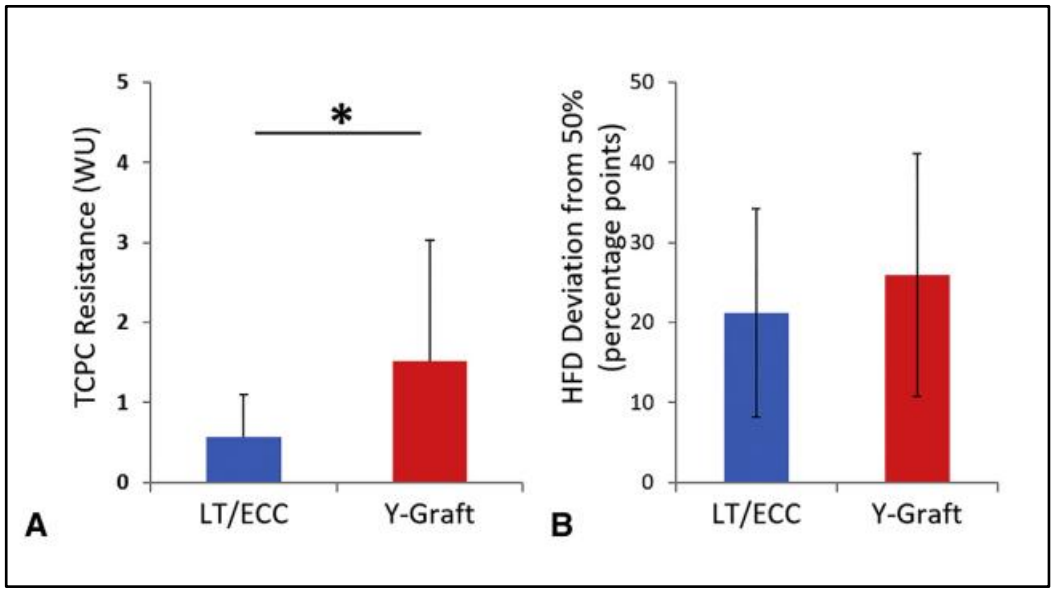


A pulsatile hemodynamic evaluation of the commercially available bifurcated Y-graft Fontan modification and comparison with the lateral tunnel and extracardiac conduits

Phillip M. Trusty, BS,[a] Maria Restrepo, PhD,[a] Kirk R. Kanter, MD,[b] Ajit P. Yoganathan, PhD,[a] Mark A. Fogel, MD,[c] and Timothy C. Slesnick, MD[d]

J Thorac Cardiovasc Surg 2016





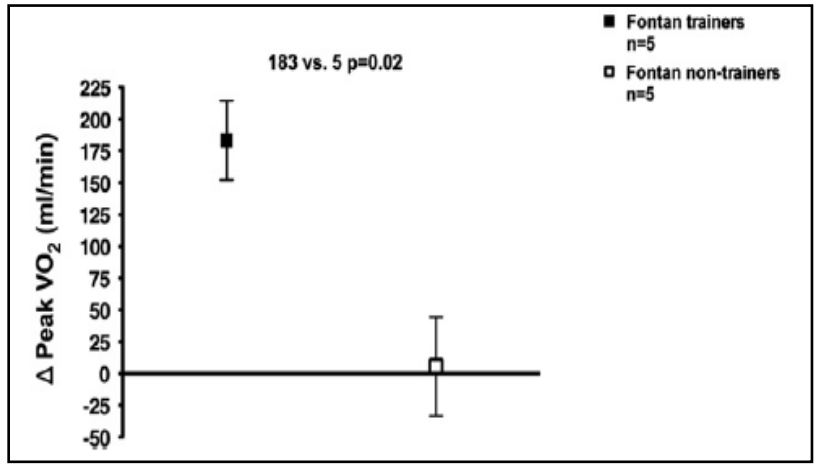
30 pts

30 pts

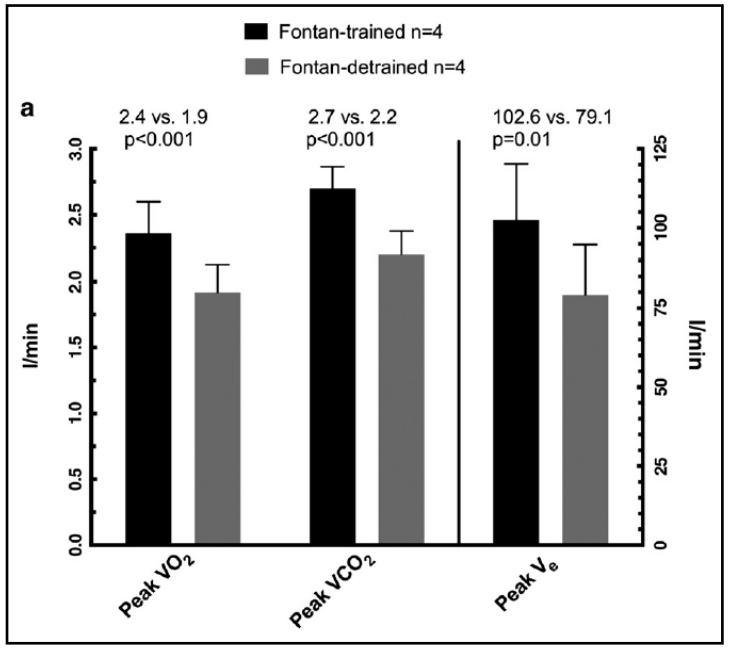
Vliv tréninku na zdatnost

Cordina RL et al., Resistance training improves cardiac output, exercise capacity and tolerance to positive airway pressure in Fontan physiology. Int J Cardiol. 2013.

po 20 týdnech tréninku



1 rok po ukončení tréninku



Efficacy and safety of macitentan in Fontan-palliated patients: 52-week randomized, placebo-controlled RUBATO Phase 3 trial and open-label extension

Paul Clift, MD,[a] Felix Berger, MD,[b] Lars Sondergaard, MD, MDSc,[c] Petra Antonova, MD,[d] Patrick Disney, MD,[e] Jeremy Nicolarsen, MD,[f] Jean-Benoît Thambo, MD,[g,h] Lidia Tomkiewicz Pajak, MD,[i] Jou-kou Wang, MD,[j] Annette Schophuus Jensen, MD, PhD,[k] Michela Efficace, MSc,[l] Michael Friberg, PhD,[m] Diana Haberle, PharmD, PhD,[m] Verena Walter, Diplom-Mathematikerin (FH),[n] and Yves d'Udekem, MD[o]

Conclusions: The primary end point of RUBATO-DB was not met; macitentan did not improve exercise capacity versus placebo in patients with Fontan palliation. Macitentan was generally well tolerated over long-term treatment. (J Thorac Cardiovasc Surg 2025;169:385-94)

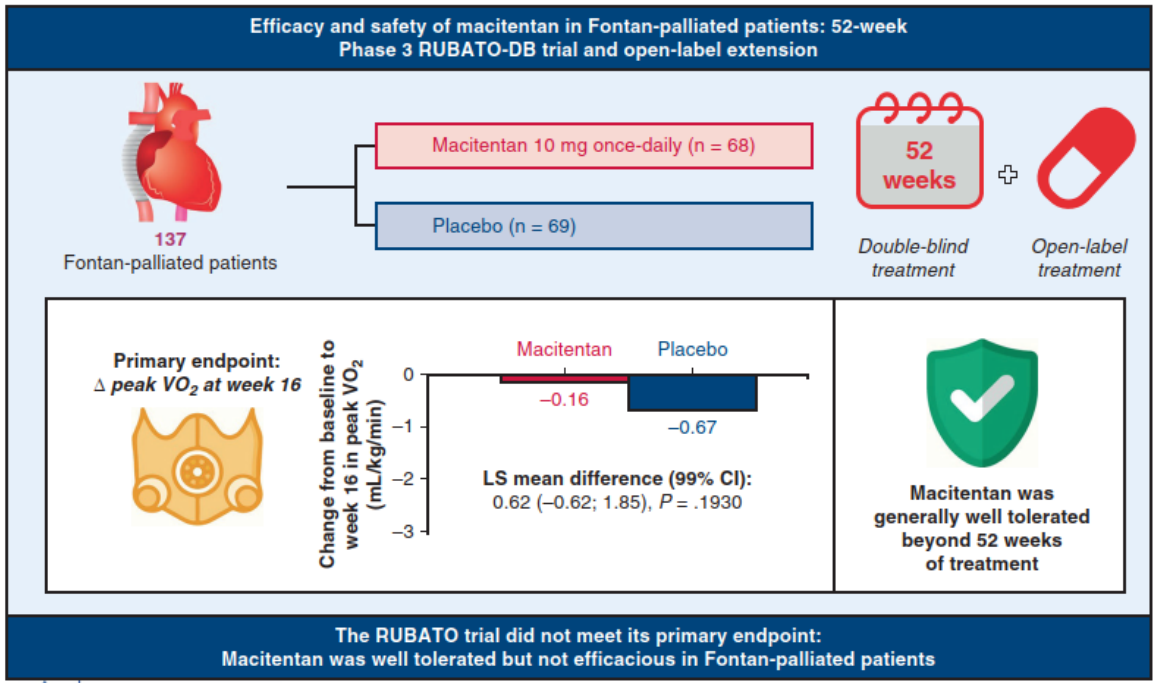
ABSTRACT

Objectives: The efficacy and safety of macitentan, an endothelin receptor antagonist, were assessed in a 52-week, prospective, multicenter, double-blind, randomized, placebo-controlled, parallel-group study assessing the efficacy and safety of macitentan in Fontan-palliated adult and adolescent patients (RUBATO-DB) and an open-label extension trial (RUBATO-OL).

Methods: Patients aged 12 years and older with New York Heart Association functional class II or III underwent total cavopulmonary connection more than 1 year before screening and showed no signs of Fontan failure/clinical deterioration. In RUBATO-DB, the primary efficacy end point was change in peak oxygen consumption from baseline to week 16; secondary end points were change from baseline over 52 weeks in peak oxygen consumption and change in mean count/minute of daily physical activity via accelerometer from baseline to week 16. Safety was assessed throughout both studies.

Results: In RUBATO-DB, 137 patients were randomized to macitentan 10 mg (n = 68) or placebo (n = 69); 92.7% completed 52-week double-blind treatment. At week 16, mean ± SD change in peak oxygen consumption was −0.16 ± 2.86 versus −0.67 ± 2.66 mL/kg/minute with macitentan versus placebo (median unbiased treatment difference estimate, 0.62 mL/kg/minute [99% repeated CI, −0.62 to 1.85]; $P = .19$). No treatment effect was observed in either of the secondary end points. During RUBATO-DB, most common adverse events with macitentan were headache, nasopharyngitis, and pyrexia. Across RUBATO-DB and RUBATO-OL, most common adverse events were COVID-19, headache, and fatigue. RUBATO-OL was prematurely discontinued because RUBATO-DB did not meet its primary or secondary end point.

Conclusions: The primary end point of RUBATO-DB was not met; macitentan did not improve exercise capacity versus placebo in patients with Fontan palliation. Macitentan was generally well tolerated over long-term treatment. (J Thorac Cardiovasc Surg 2025;169:385-94)

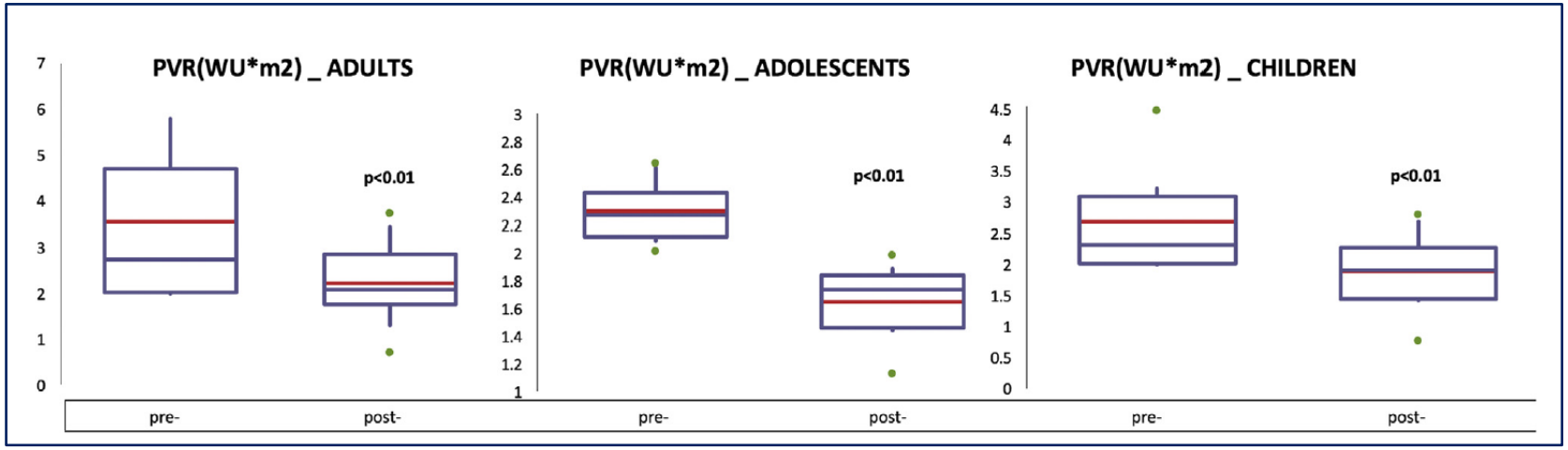


Endothelin inhibitors lower pulmonary vascular resistance and improve functional capacity in patients with Fontan circulation

Gabriella Agnoletti, PhD,[a] Simona Gala, MD,[a] Francesca Ferroni, MD,[a] Roberto Bordese, MD,[a] Lorenzo Appendini, MD,[b] Carlo Pace Napoleone, MD,[c] and Laura Bergamasco, PhD[b]

Agnoletti 2017

Patients with increased Rp > 2 U * m^2



Physical exercise training in patients with a Fontan circulation: A systematic review

Linda E. Scheffers[1], Linda E.M. vd Berg[1,2], Gamida Ismailova[2], Karolijn Dulfer[3], Johanna J.M. Takkenberg[4], and Wim A. Helbing[1]*

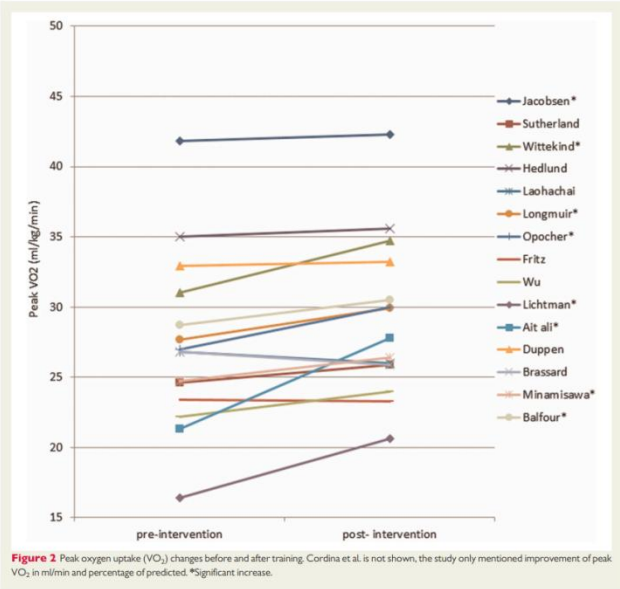


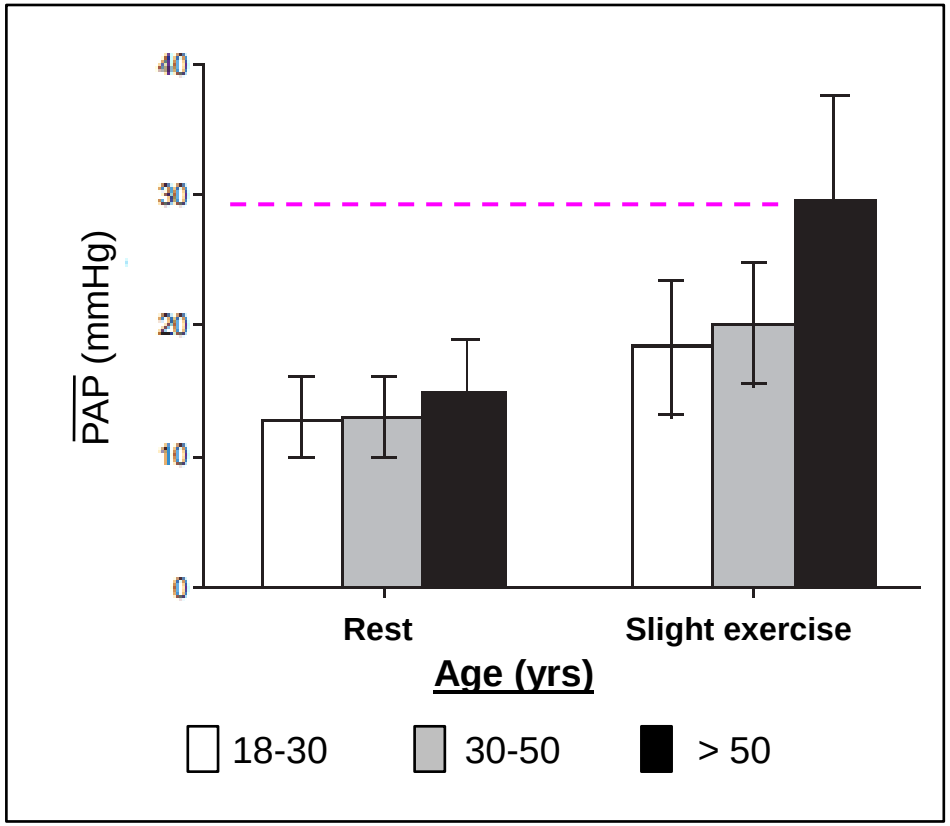
Figure 2 Peak oxygen uptake (VO_2) changes before and after training. Cordina et al. is not shown, the study only mentioned improvement of peak VO_2 in ml/min and percentage of predicted. *Significant increase.

Pulmonary arterial pressure during rest and exercise in healthy subjects: a systematic review

G. Kovacs*, A. Berghold#, S. Scheidl* and H. Olschewski*

Eur Resp J 2009

Jak se bude dařit jedincům s Fontanovou cirkulací v 50ti letech ??

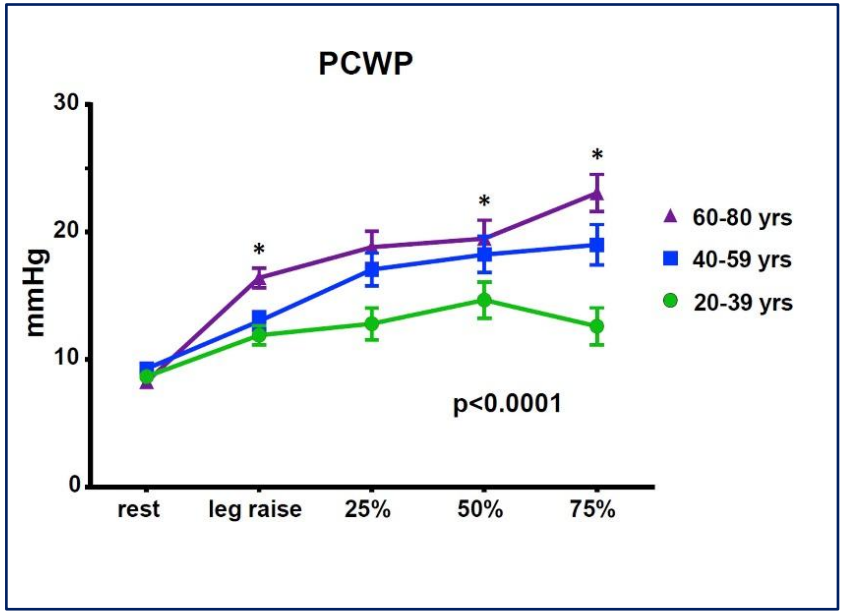


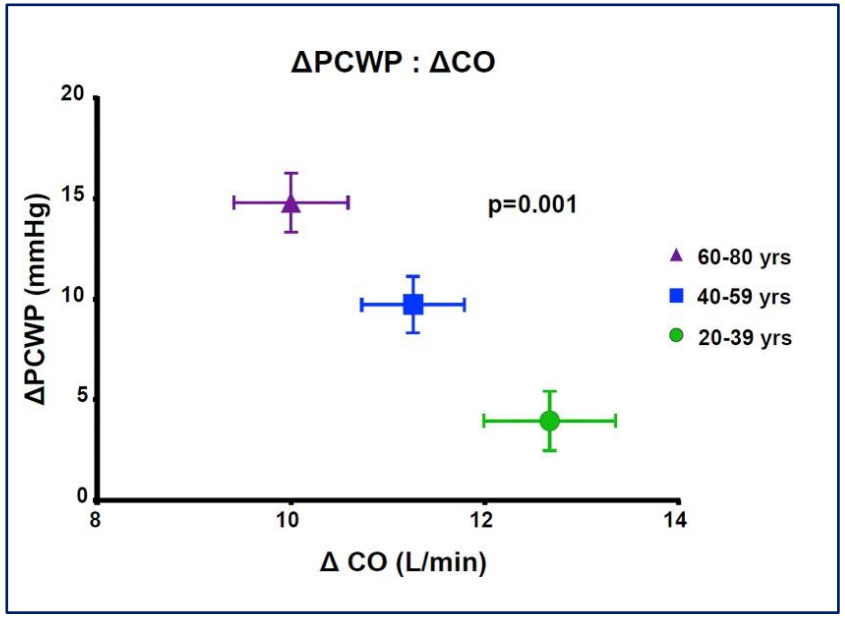
The Influence of Age on Hemodynamic Parameters During Rest and Exercise in Healthy Individuals

Emil Wolsk, MD, PhD,[a] Rine Bakkestrøm, MD,[b] Jacob H. Thomsen, MD,[a] Louise Balling, MD,[a] Mads J. Andersen, MD, PhD,[a] Jordi S. Dahl, MD, PhD,[b] Christian Hassager, MD, DMSc,[a] Jakob E. Møller, MD, DMSc,[b] Finn Gustafsson, MD, DMSc[a]

JACC Heart Fail 2017;5: 337-346

Jak se bude dařit jedincům s Fontanovou cirkulaci v 50ti letech ??



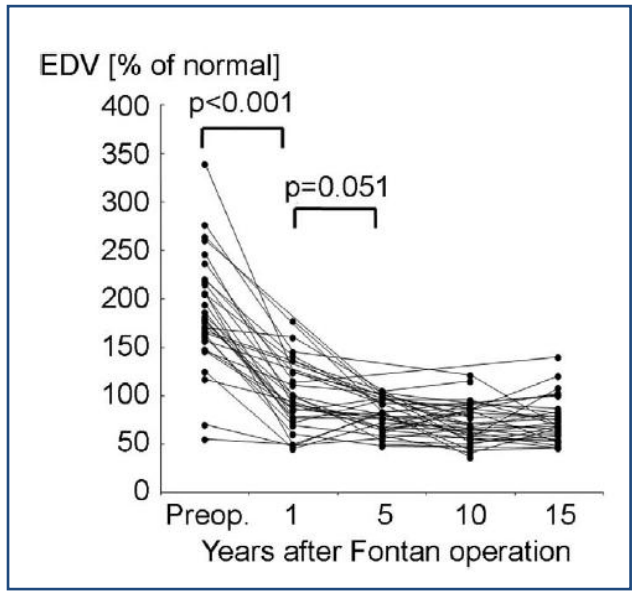


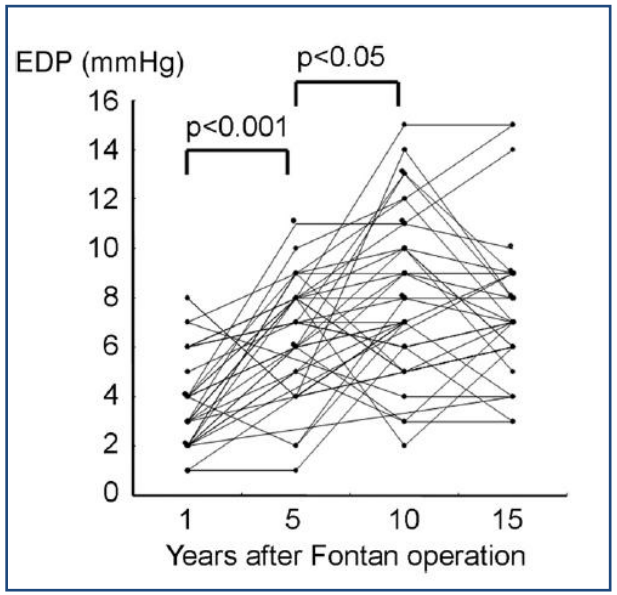
62 zdravých jedinců

Ventricular Performance in Long-Term Survivors After Fontan Operation

Yuki Nakamura, MD, Toshikatsu Yagihara, MD, Koji Kagisaki, MD, Ikuo Hagino, MD, and Junjiro Kobayashi, MD

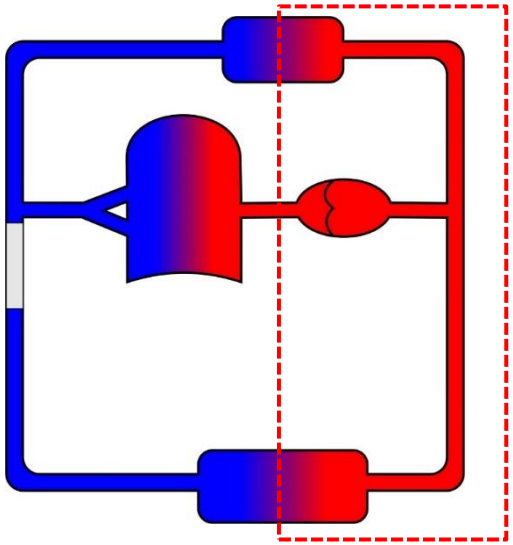
Ann Thorac Surg 2011;91:172

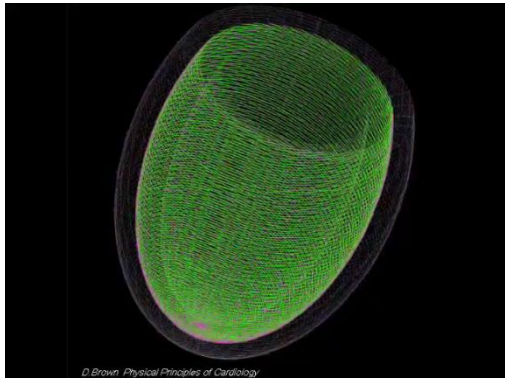




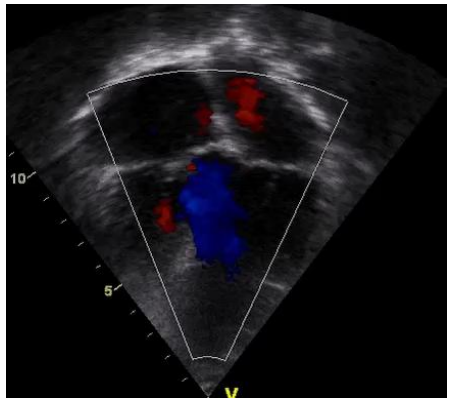
Patofyziologie Fontanovy cirkulace

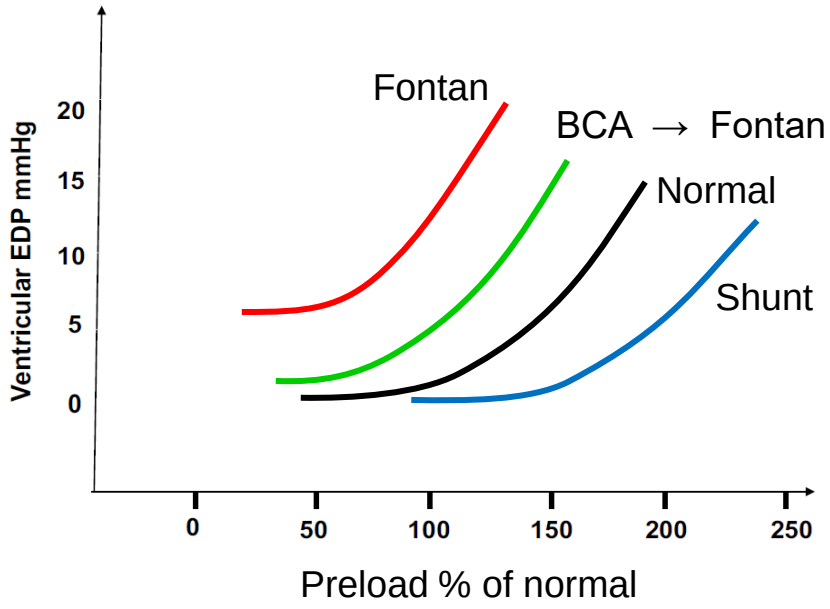
Systémová cirkulace

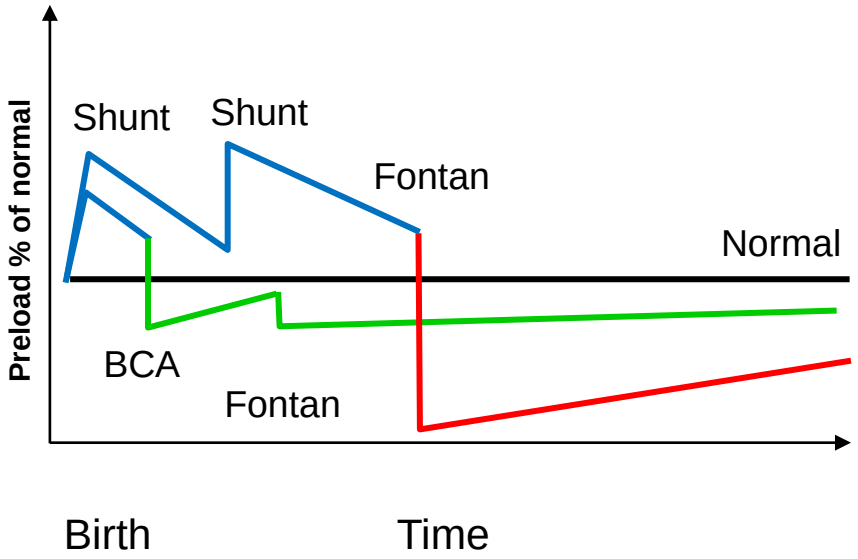




Srdeční funkce







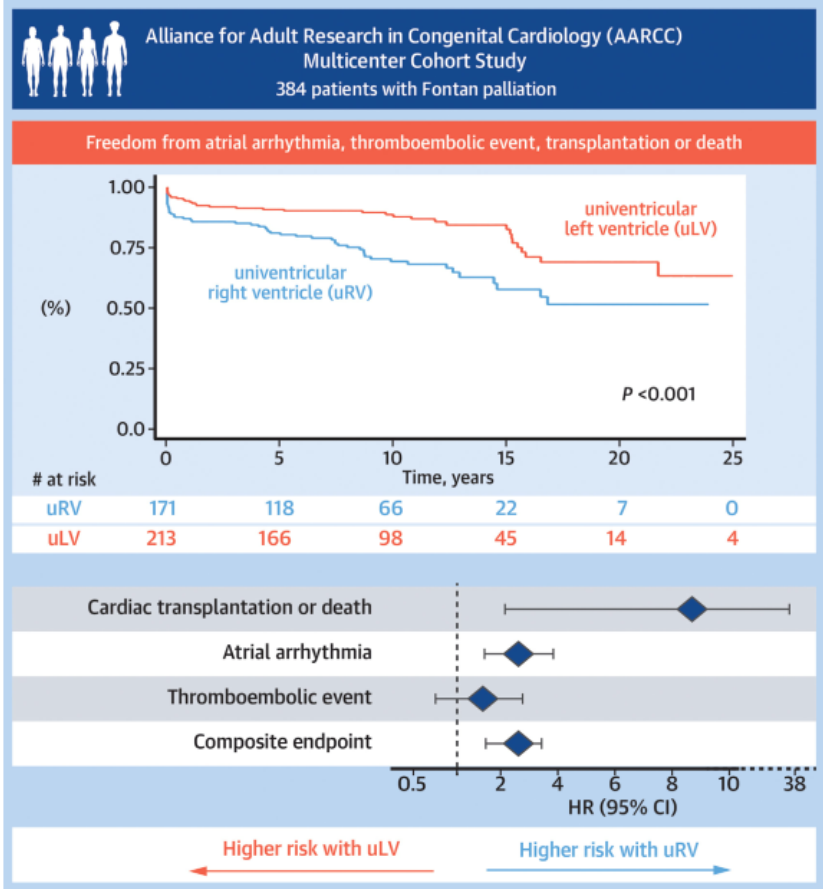
Upraveno podle: Gewilliga 2014

Cardiovascular Outcomes in Fontan Patients With Right vs Left Univentricular Morphology

A Multicenter Study

Nabil Dib, MD,[a] Marie-A. Chaix, MD, PhD,[a] Michelle Samuel, MPH, PhD,[a] Sewanou Hermann Honfo, PhD,[a] Robert M. Hamilton, MD,[b] Jamil Aboulhosn, MD,[c] Craig S. Broberg, MD,[d] Scott Cohen, MD,[e] Stephen Cook, MD,[f] Annie Dore, MD,[a] Susan M. Jameson, LPD, PA-C,[g] Anne Fournier, MD,[h] Reda Ibrahim, MD,[a] Joseph Kay, MD,[i] François-Pierre Mongeon, MD, SM,[a] Alexander R. Opotowsky, MD, MMSc,[j] Ali Zaidi, MD,[k] Nancy Poirier, MD,[a] Paul Khairy, MD, PhD[a]

CENTRAL ILLUSTRATION Cardiovascular Outcomes in Fontan Patients With Right vs Left Univentricular Morphology



Dib N, et al. JACC Adv. 2024;3(4):100871.

ABSTRACT

BACKGROUND There is a paucity of data on long-term outcomes after Fontan palliation in patients with a dominant morphological univentricular right (uRV) vs left (uLV) ventricle.

OBJECTIVES The purpose of this study was to compare the incidence of atrial arrhythmias, thromboembolic events, cardiac transplantation, and death following Fontan palliation in patients with uRV vs uLV.

METHODS The Alliance for Adult Research in Congenital Cardiology conducted a multicenter retrospective cohort study on patients with total cavopulmonary connection Fontan palliation across 12 centers in North America. All components of the composite outcome, that is, atrial arrhythmias, thromboembolic events, cardiac transplantation, and death, were reviewed and classified by a blinded adjudicating committee. Time-to-event analyses were performed that accounted for competing risks.

RESULTS A total of 384 patients were followed for 10.5 ± 5.9 years. The composite outcome occurred in 3.7 vs 1.7 cases per 100 person-years for uRV (N = 171) vs uLV (N = 213), respectively ($P < 0.001$). In multivariable analyses, uRV conferred a >2-fold higher risk of the composite outcome (HR: 2.17, 95% CI: 1.45-3.45, $P < 0.001$). In secondary analyses of components of the primary outcome, uRV was significantly associated with a greater risk of cardiac transplantation or death (HR: 9.09, 95% CI: 2.17-38.46, $P < 0.001$) and atrial arrhythmias (HR: 2.17, 95% CI: 1.20-4.00, $P = 0.010$) but not thromboembolic events (HR: 1.64, 95% CI: 0.86-3.16, $P = 0.131$).

CONCLUSIONS Fontan patients with uRV vs uLV morphology have a higher incidence of adverse cardiovascular events, including atrial arrhythmia, cardiac transplantation, and all-cause mortality. (JACC Adv 2024;3:100871)

Late Fontan Circulatory Failure. What Drives Systemic Venous Congestion and Low Cardiac Output in Adult Fontan Patients?

Alexander Van De Bruaene[1], Guido Claessen[1], Thomas Salaets[2] and Marc Gewillig[2]*

[1] *Division of Cardiology, Department of Cardiovascular Sciences, University Hospitals Leuven, KU Leuven, Leuven, Belgium,*
[2] *Division of Pediatric Cardiology, Department of Cardiovascular Sciences, University Hospitals Leuven, KU Leuven, Leuven, Belgium*

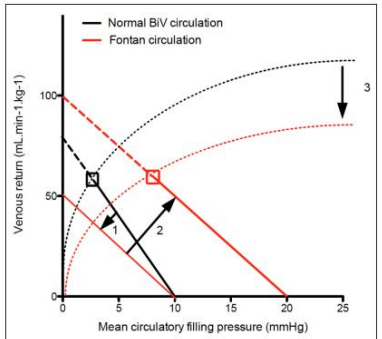


FIGURE 1 | Venous return curve. In Fontan patients the venous return curve shows on downward rotation due the pulmonary vasculature (and hence PVR) being incorporated in venous return (1). Volume load brings back venous return at the level of biventricular circulation at the expense of an increase in mean circulatory filling pressure (2). Finally since left atrial pressure is the backpressure for venous return, increased left atrial pressure will result in decrease cardiac output. The dashed lines indicate the Frank Starling curve, indicating lower contractility (3).

reason that factors affecting mean filling pressure (blood volume

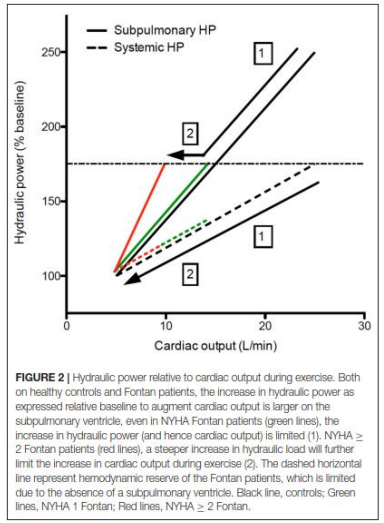


FIGURE 2 | Hydraulic power relative to cardiac output during exercise. Both on healthy controls and Fontan patients, the increase in hydraulic power as expressed relative baseline to augment cardiac output is larger on the subpulmonary ventricle, even in NYHA Fontan patients (green lines), the increase in hydraulic power (and hence cardiac output) is limited (1). NYHA ≥ 2 Fontan patients (red lines), a steeper increase in hydraulic load will further limit the increase in cardiac output during exercise (2). The dashed horizontal line represent hemodynamic reserve of the Fontan patients, which is limited due to the absence of a subpulmonary ventricle. Black line, controls; Green lines, NYHA 1 Fontan; Red lines, NYHA ≥ 2 Fontan.

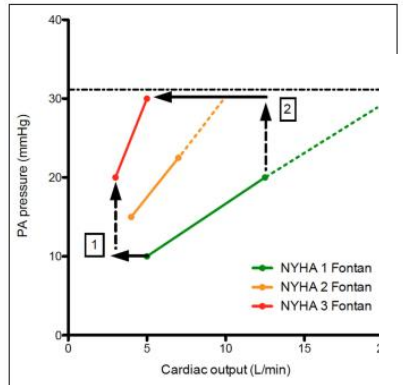


FIGURE 3 | Pressure flow plots indicating the increase in PA pressure vs. cardiac output during exercise. When comparing NYHA 1 Fontan patients wi NYHA 2&3 Fontan patients, note there is a decrease in cardiac output despit increase in resting PA pressure (1). During exercise, the increase in PA pressure is steeper for NYHA 2&3 patients. A hypothetical maximum of PA pressure (systematic venous pressure) results is an exacerbated decrease in cardiac output resulting in significant functional limitation of the Fontan patier (2). The dashed lines represent hemodynamic reserve of the Fontan patient.

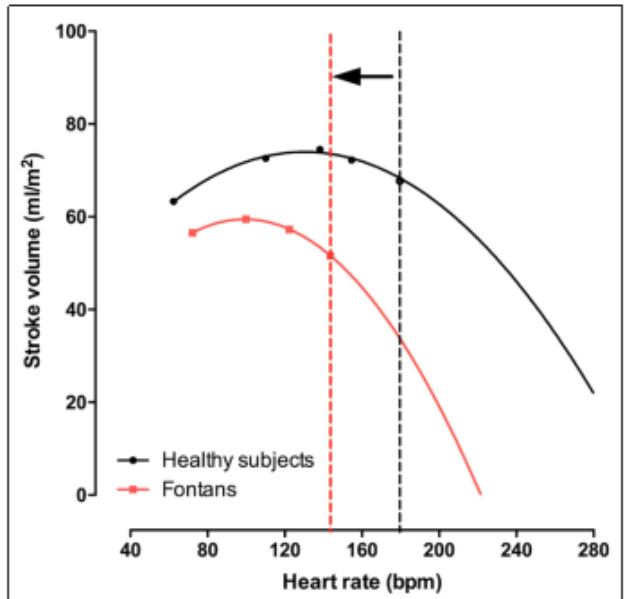


FIGURE 5 | Stroke volume index vs. heart rate during exercise. In Fontan patient heart rate and exercise capacity is limited. In a significant proportion of Fontan patients, heart rate relative to workload is preserved (or even increased). Nevertheless, heart rate reserve is still reduced, which may be a physiologic mechanism preventing a full in stroke volume (and cardiac output). This highlighted in the figure, where the infliction point (vertical dashed line) beyond which a further increase in heart rate would result in falling cardiac output.

The Epidemiology of Persons Living with Fontan in 2020 and Projections for 2030: Development of an Epidemiology Model Providing Multinational Estimates

Leandra Plappert · Susan Edwards · Assunta Senatore · Angela De Martini

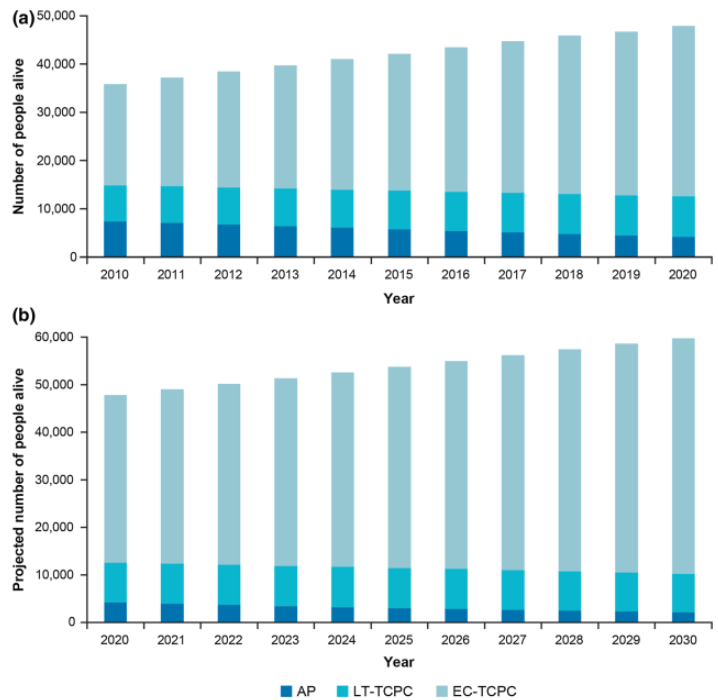


Fig. 2 Estimates of the number of people alive with a Fontan circuit in **A** 2010–2020 and projection through **B** 2020–2030, by procedural subtype. *AP* atriopulmonary connection, *EC-TCPC* extracardiac total cavopulmonary connection, *LT-TCPC* lateral tunnel total cavopulmonary connection

Diastolic Dysfunction With Preserved Ejection Fraction After the Fontan Procedure

Shahryar M. Chowdhury, MD, MSCR; Eric M. Graham, MD; Carolyn L. Taylor, MD; Andrew Savage, MD; Kimberly E. McHugh, MD; Stephanie Gaydos, MD; Arni C. Nutting, MD; Michael R. Zile, MD; Andrew M. Atz, MD

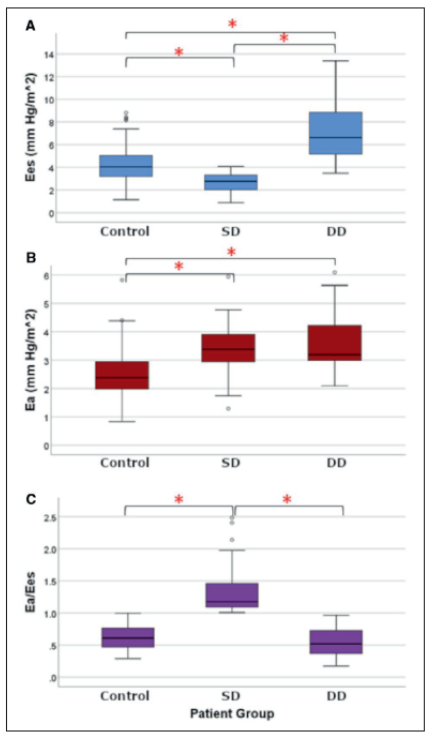


Figure 1. Differences in cardiac mechanics between controls, systolic dysfunction, and diastolic dysfunction. **A**, Differences in Ees (contractility) between controls, SD, and DD. **B**, Differences in Ea (afterload) between controls, SD, and DD. **C**, Differences in Ea/Ees (ventriculo-arterial coupling) between controls, SD, and DD. *over a bracket represents $P<0.01$ between the groups. DD indicates diastolic dysfunction; Ea, arterial elastance; Ees, end-systolic elastance; and SD, systolic dysfunction.

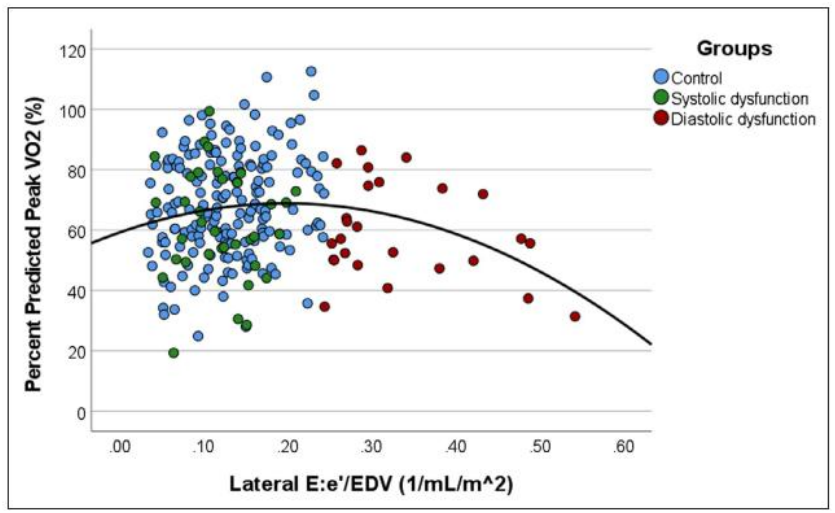
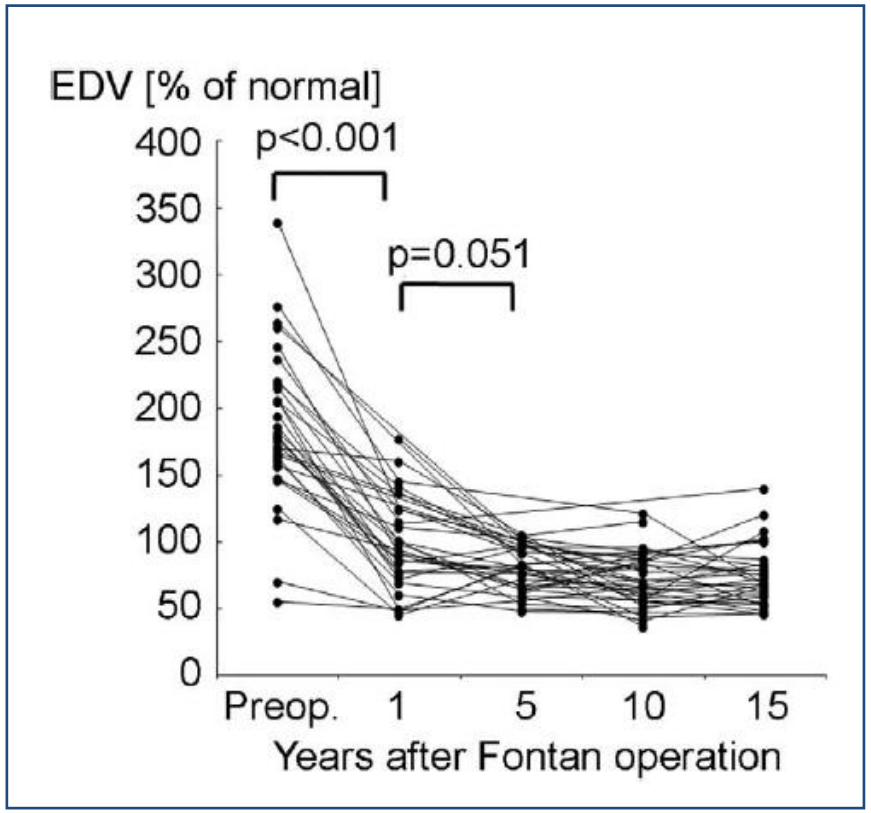


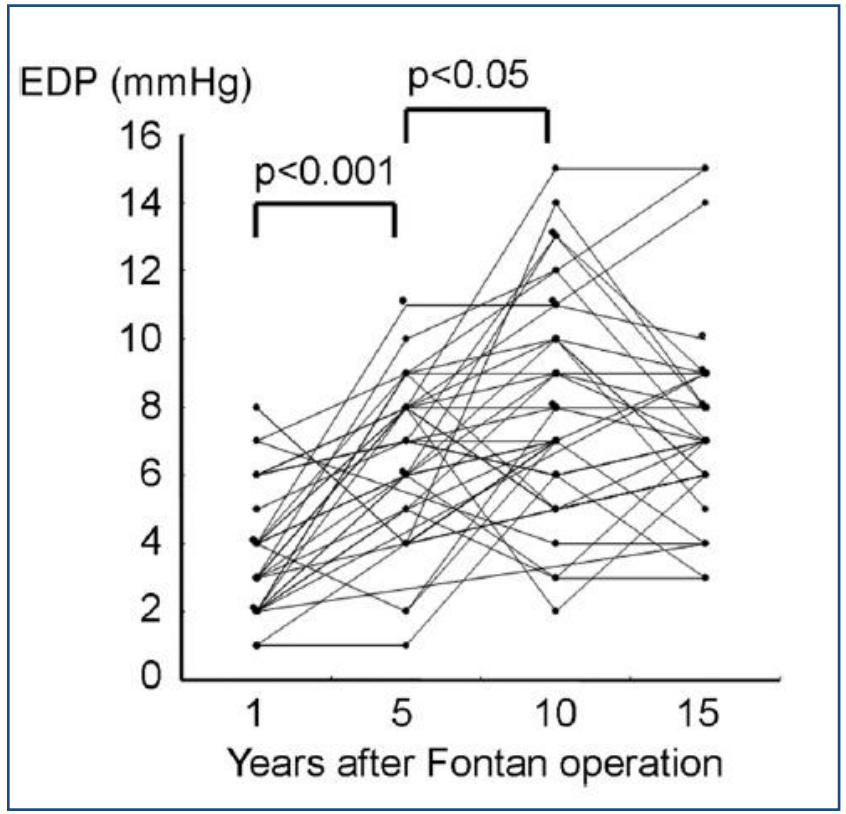
Figure 2. Relationship between lateral E:e'/EDV and percent predicted peak VO_2. Scatterplot of lateral E:e'/EDV and percent predicted peak VO_2. Patient groups are separated by color coding. Quadratic fit line shown ($F=4.76$, $P=0.009$). EDV indicates end-diastolic volume.

Ventricular Performance in Long-Term Survivors After Fontan Operation

Yuki Nakamura, MD, Toshikatsu Yagihara, MD, Koji Kagisaki, MD, Ikuo Hagino, MD, and Junjiro Kobayashi, MD

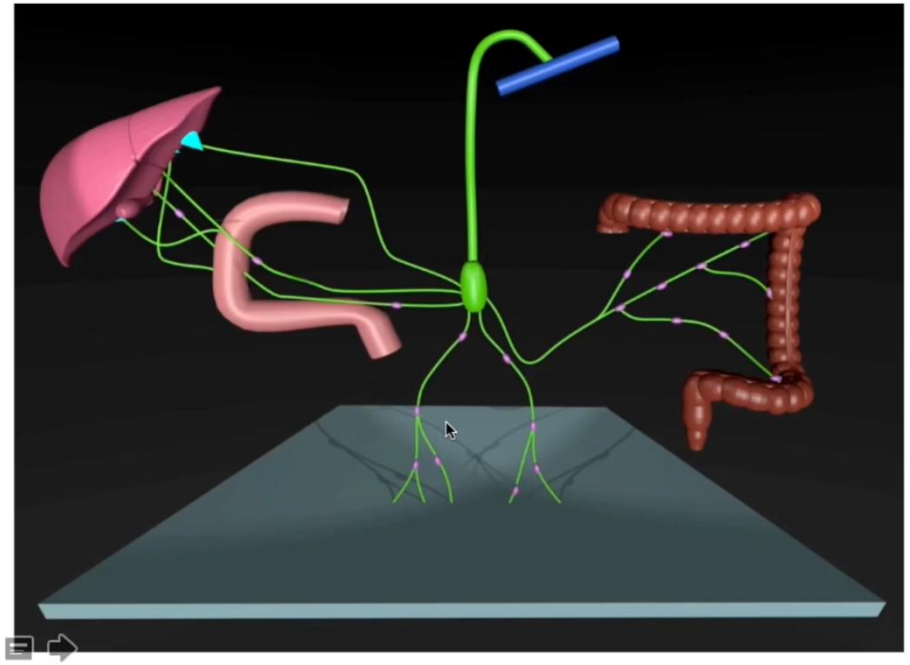
Ann Thorac Surg 2011;91:172

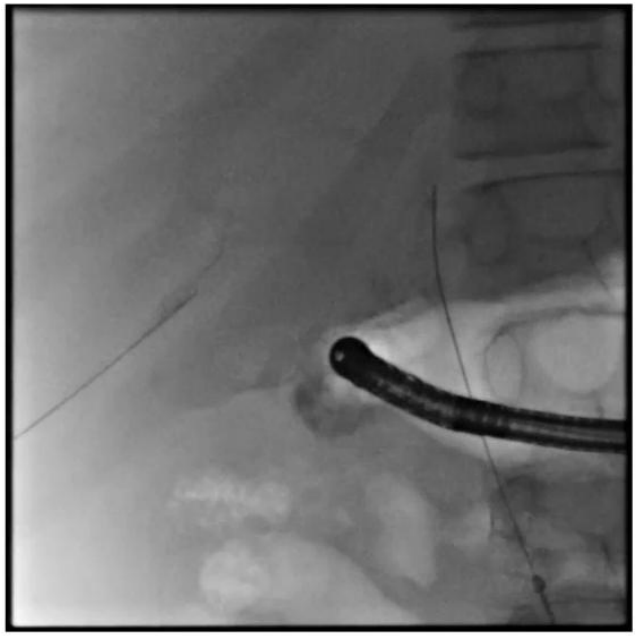


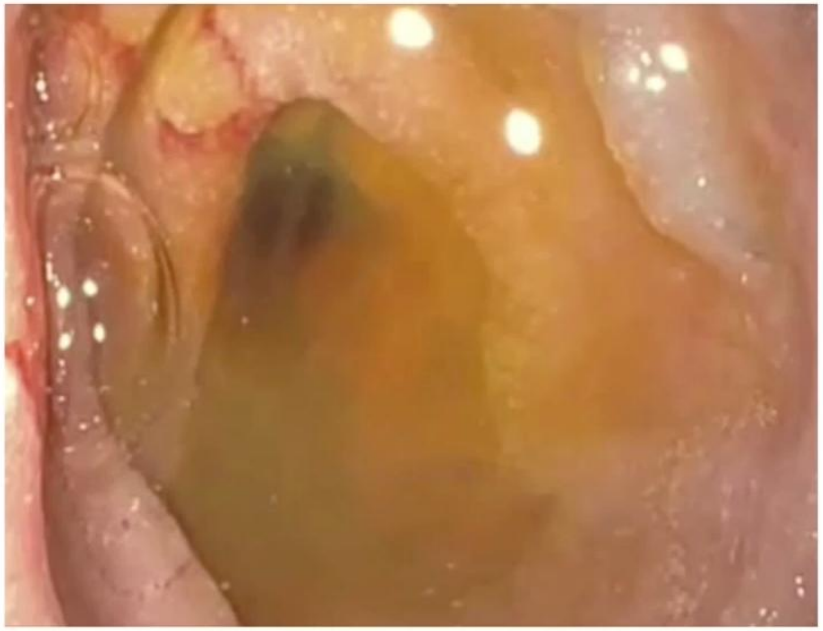


Exudativní enteropatie po TCPC

Lymfatická drenáž

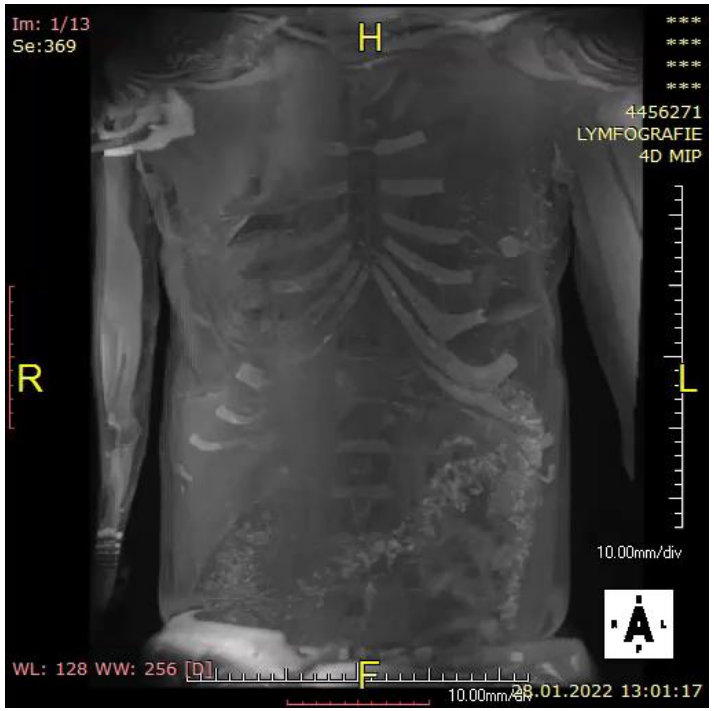


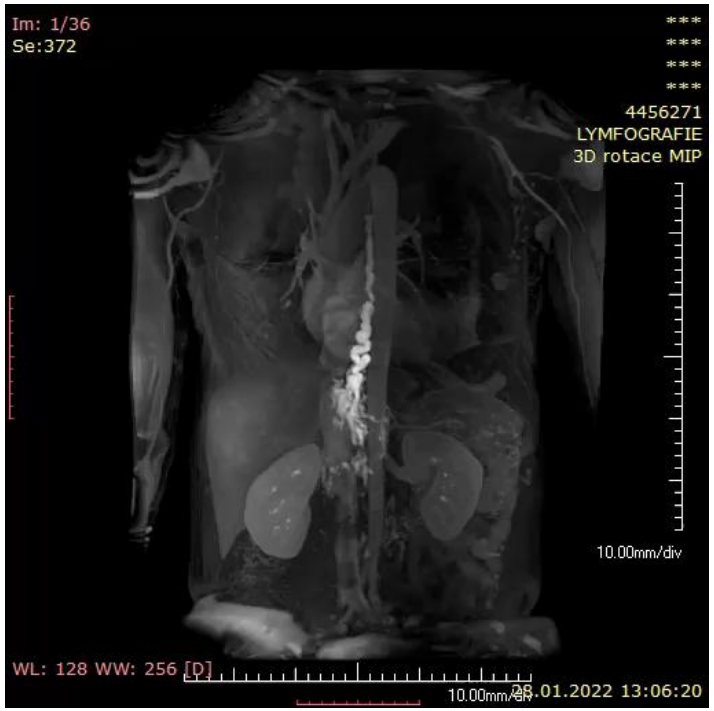




Itkin: CHOP 2017

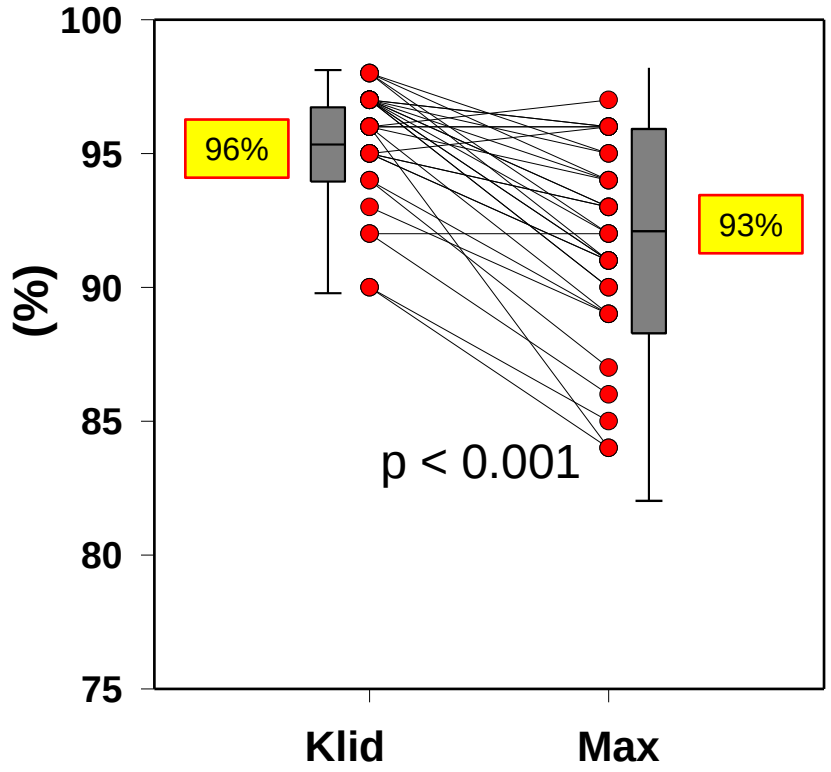
MRI – chylothorax





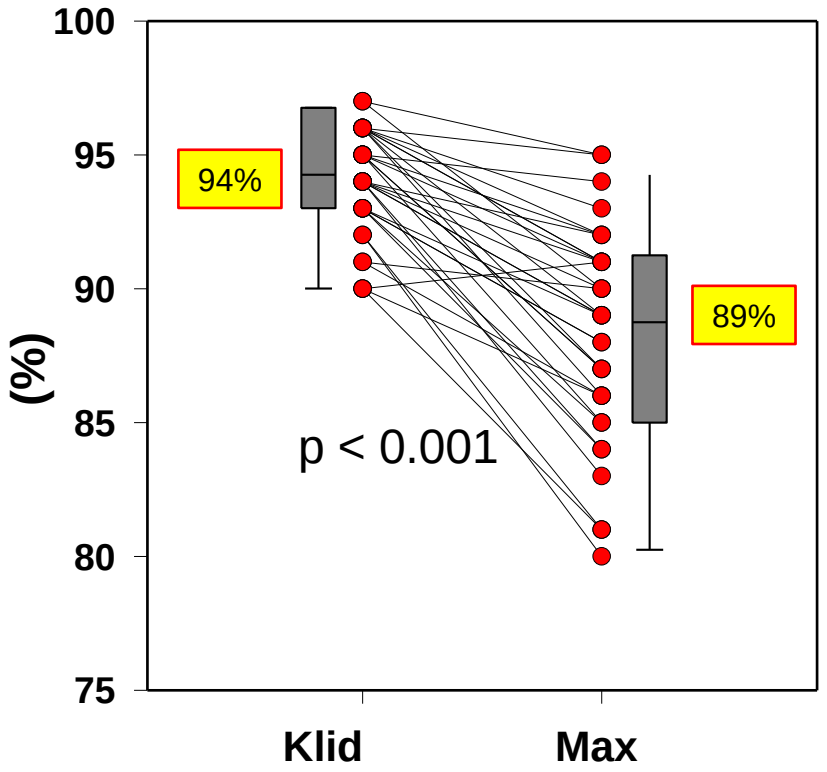
Pulzní oxymetrie po TCPC při zátěži

35 jedinců bez fenestrace



Studie 1 (roky)

Věk 9
Interval 5



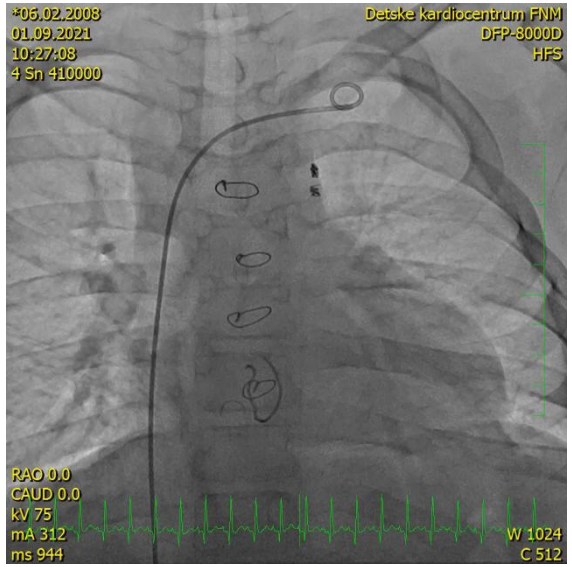
Studie 2 (roky)

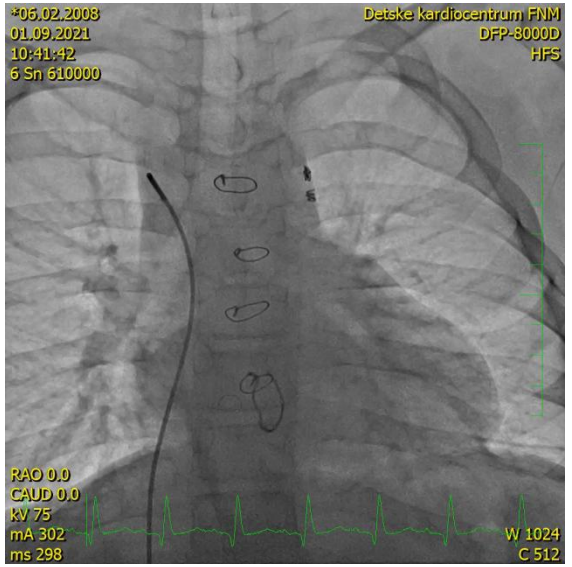
Věk 17
Interval 13

Patofyziologie Fontanovy cirkulace

Veno - venózní kolaterály

- ↙ plicní průtok / tlak v TCPC
- ↙ **arteriální saturaci**

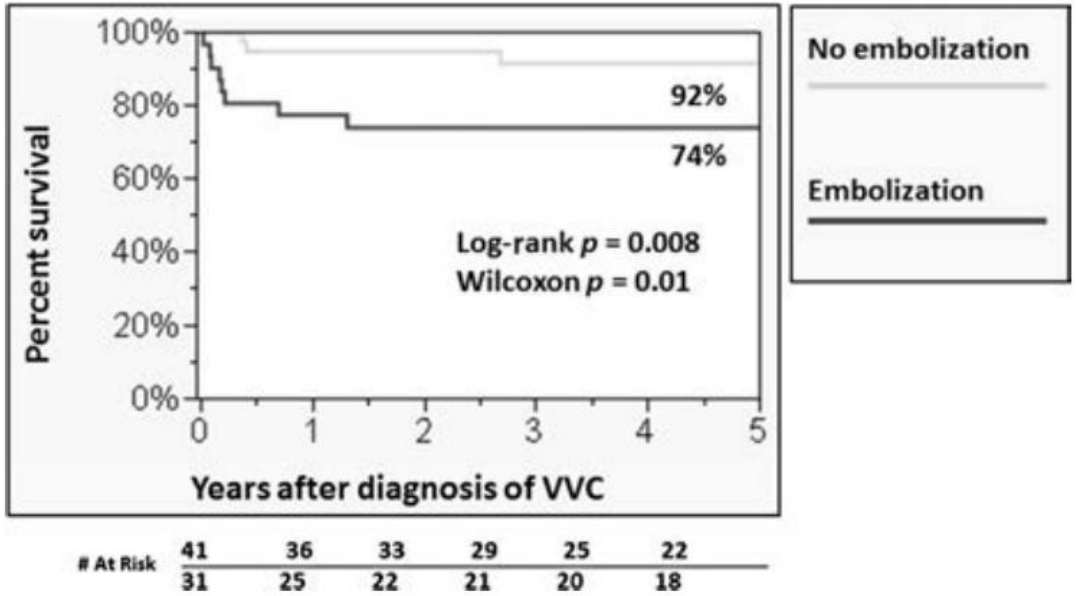




Embolization of Veno-venous Collaterals after the Fontan Operation Is Associated with Decreased Survival

Joseph T. Poterucha, DO,* Jonathan N. Johnson, MD,* Nathan W. Taggart, MD,* Allison K. Cabalka, MD,* Donald J. Hagler, MD,*† David J. Driscoll, MD,*† and Frank Cetta,

Congenit Heart Dis 2015



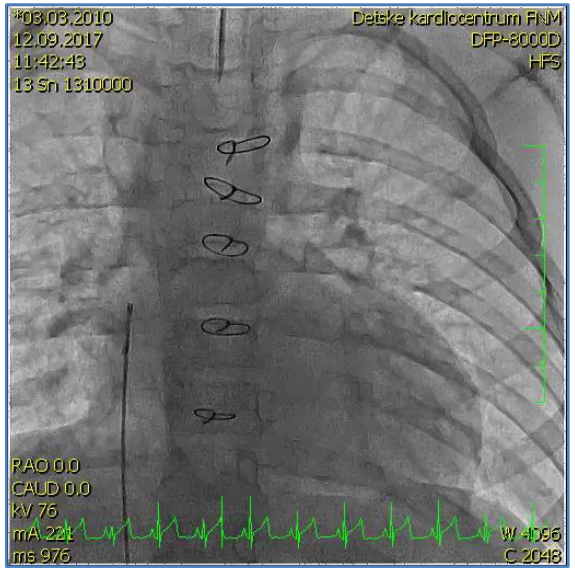
Dospělí

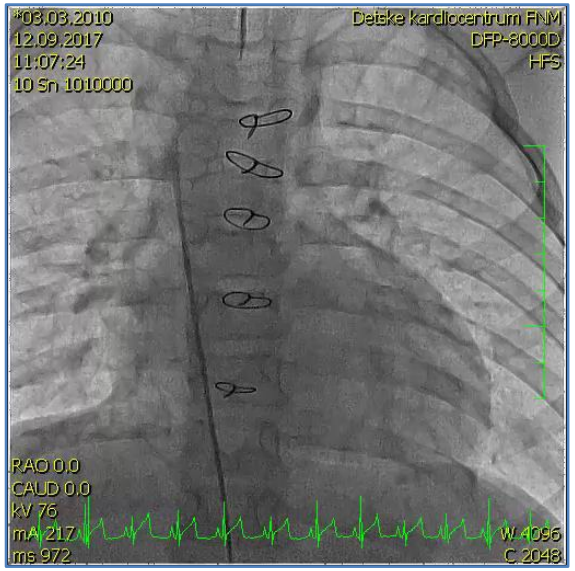
Patofyziologie Fontanovy cirkulace

Aortopulmonální kolaterály

↗ plicní průtok a tlak v TCPC
↗ objemová zátěž komory

} **žilní městnání**

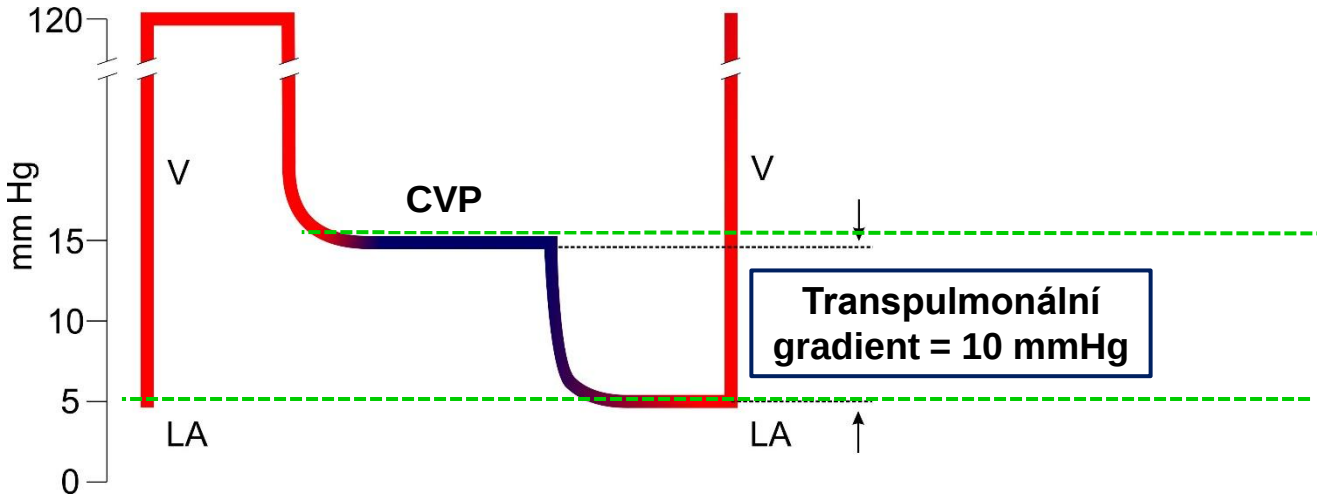


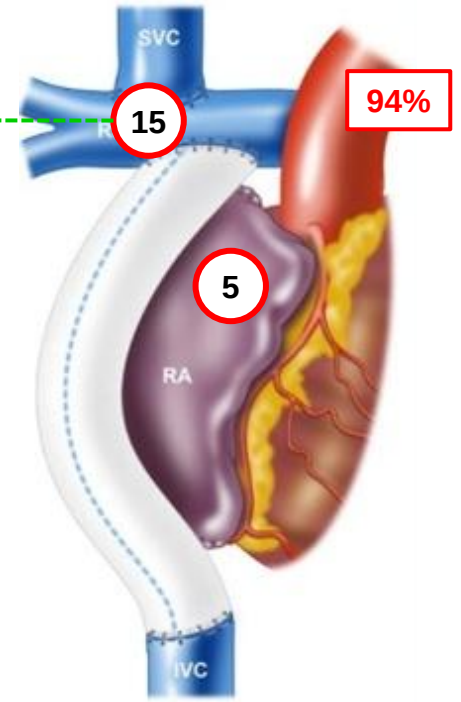


→ Katetrizační uzávěr

TCPC

Nedostatečná kapacita plicnic
Zvýšená plicní cévní rezistence



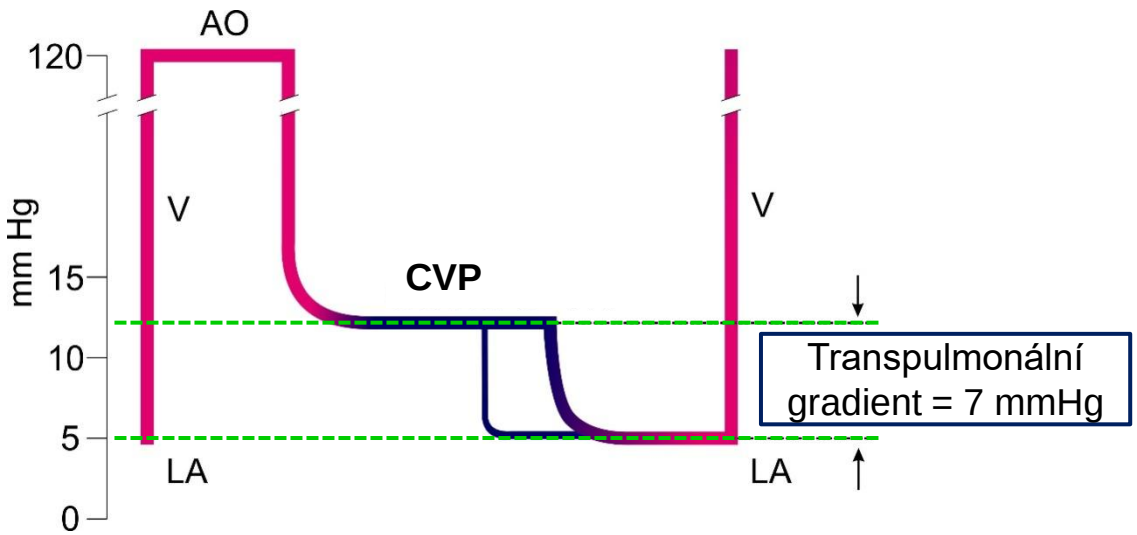


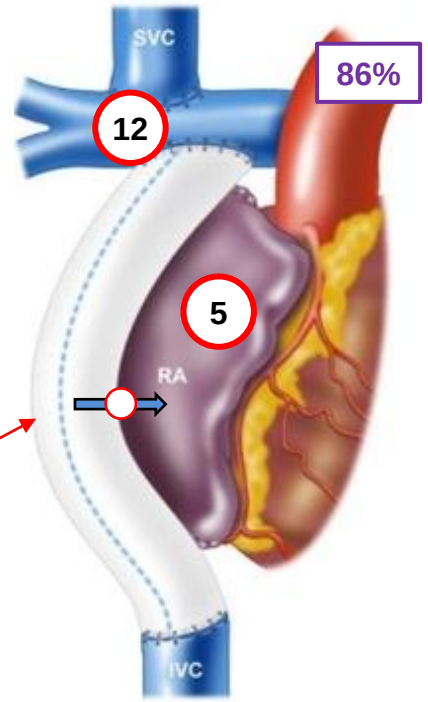
Závažné systémové žilní městnání
- edémy
- fluidothorax
- ascites

Nízký srdeční výdej
- renální selhání
- hepatální selhání

TCPC s fenestrací

Nedostatečná kapacita plicnic
Zvýšená plicní cévní rezistence

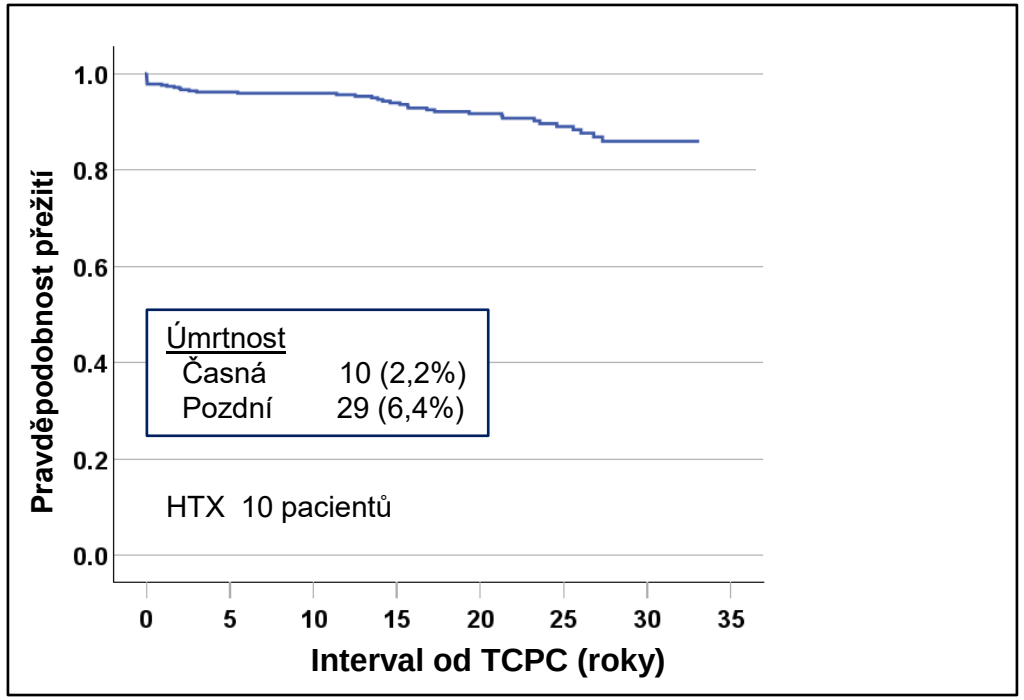


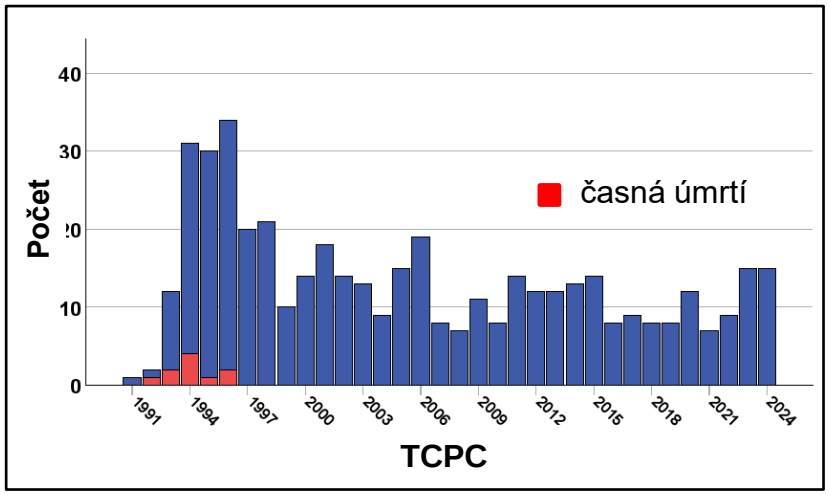


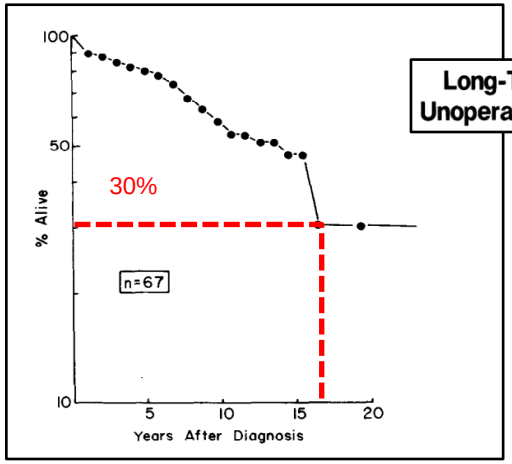
Fenestrace

Úplné kavopulmonální spojení v Dětském kardiocentru (1991 - 2024)

N = 453







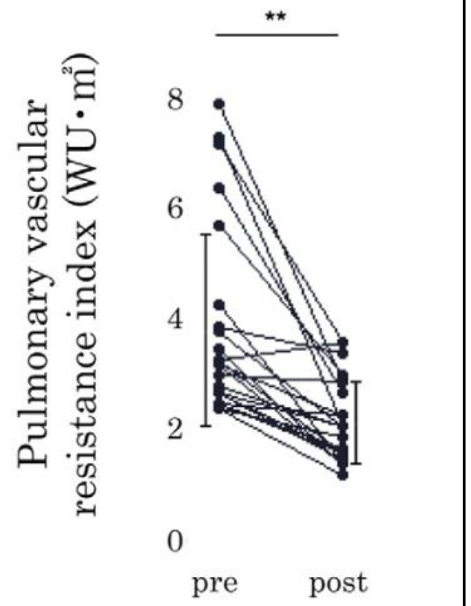
Long-Term Follow-Up in the Unoperated Univentricular Heart

Moodie 1984

Fontanova cirkulace a plicní vazodilatancia

Testy byly provedeny u jedinců se zvýšenou plicní cévní rezistencií > 2,0 WJ*m^2

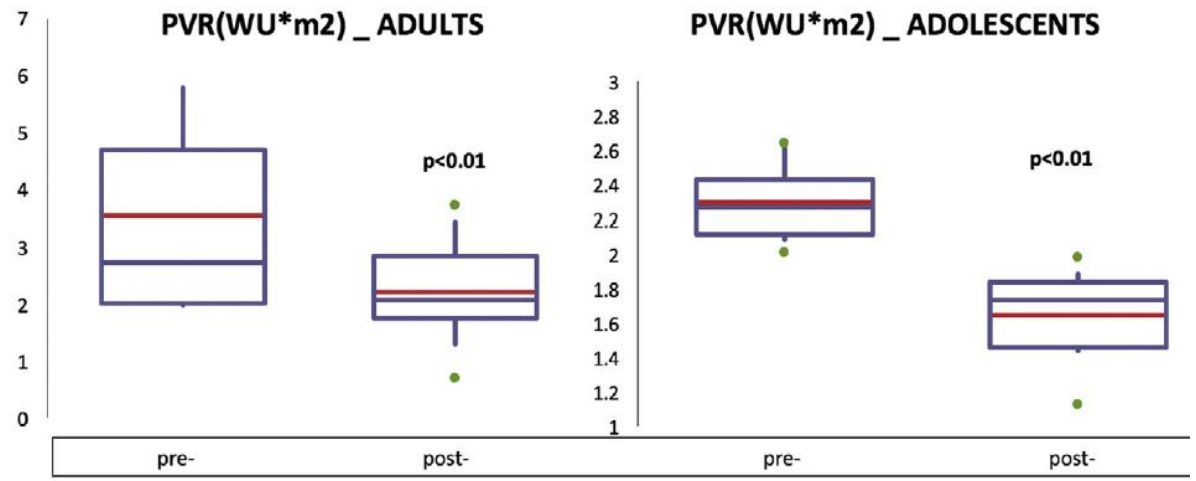
Sildenafil



Mori 2016

Macicentan

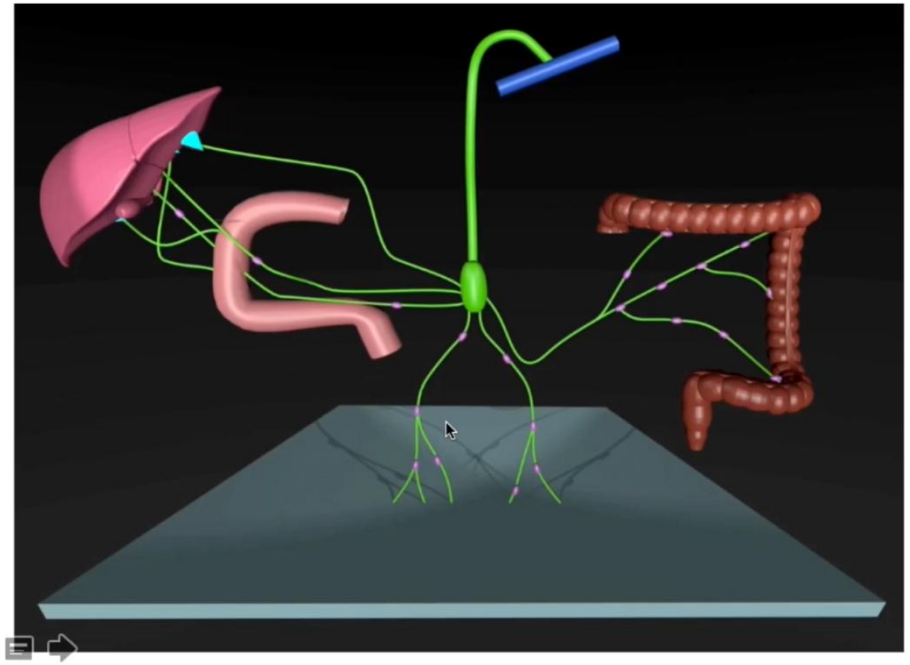
Bosentan

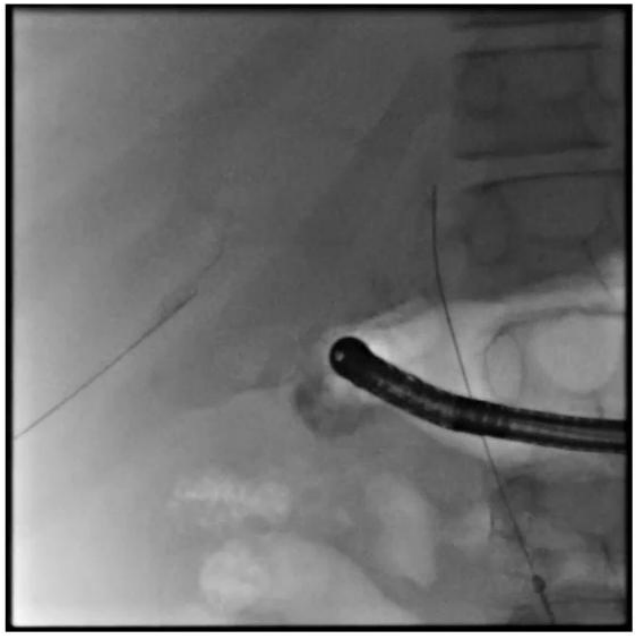


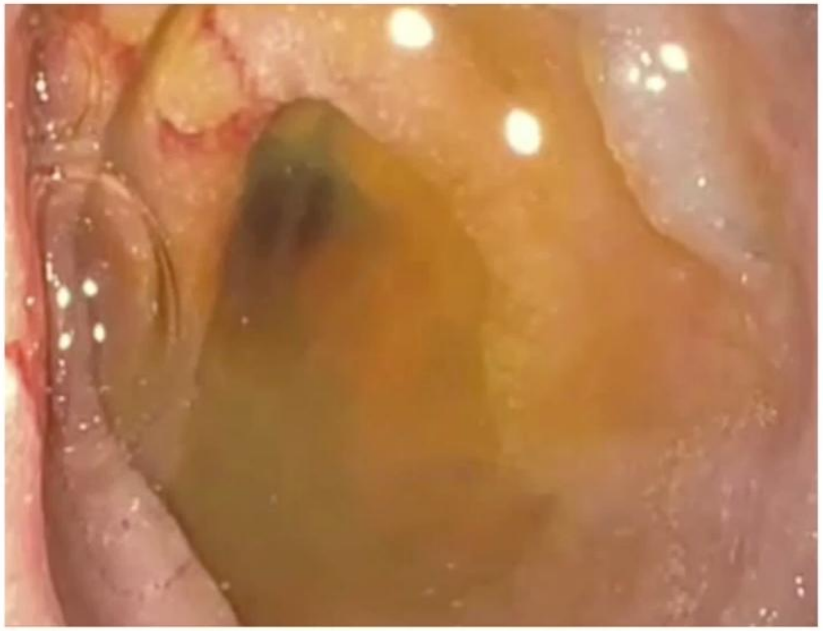
Agnoletti 2017

Exudativní enteropatie po TCPC

Lymfatická drenáž







Itkin: CHOP 2017