



DGPK Guideline Univentricular Heart

Alfred Hager (DHM, TU München)

Stanislav Ovroutski (DHZB, Berlin)

Robert Cesnjevar (Universitätsklinikum Erlangen)



Conflicts of Interests

Leitlinienkoordinator:		Prof. Dr. med. Jochen Weil		
Leitlinie:		Univentrikuläres Herz		
		PD Dr. med. Alfred Hager	Dr. med. Stanislav Ovroutski	Prof. Dr. med. Robert Cesnjevar
1	Berater- bzw. Gutachtertätigkeit oder bezahlte Mitarbeit in einem wissenschaftlichen Beirat eines Unternehmens der Gesundheitswirtschaft (z.B. Arzneimittelindustrie, Medizinproduktindustrie), eines kommerziell orientierten Auftragsinstituts oder einer Versicherung	Actelion	Ø	Ø
2	Honorare für Vortrags- und Schulungstätigkeiten oder bezahlte Autoren- oder Co-Autorenschaften im Auftrag eines Unternehmens der Gesundheitswirtschaft, eines kommerziell orientierten Auftragsinstituts oder einer Versicherung	Abbott Actelion Encysive Pfizer	Ø	Ø
3	Finanzielle Zuwendungen (Drittmittel) für Forschungsvorhaben oder direkte Finanzierung von Mitarbeitern der Einrichtung von Seiten eines Unternehmens der Gesundheitswirtschaft, eines kommerziell orientierten Auftragsinstituts oder einer Versicherung	Actelion Medtronik Pfizer	Ø	Köhler Chemie Sorin
4	Eigentümerinteresse an Arzneimitteln/Medizinprodukten (z. B. Patent, Urheberrecht, Verkaufslizenz)	Ø	Ø	Ø
5	Besitz von Geschäftsanteilen, Aktien, Fonds mit Beteiligung von Unternehmen der Gesundheitswirtschaft	Siemens	Ø	Ø
6	Persönliche Beziehungen zu einem Vertretungsberechtigten eines Unternehmens Gesundheitswirtschaft	Ø	Ø	Ø
7	Mitglied von in Zusammenhang mit der Leitlinienentwicklung relevanten Fachgesellschaften/Berufsverbänden, Mandatsträger im Rahmen der Leitlinienentwicklung	DGPK, Stellv. Sprecher der AG Belastungsuntersuchungen im Kindesalter (DGPK) ESC / EACPR CPX International	Ø	Ø
8	Politische, akademische (z.B. Zugehörigkeit zu bestimmten „Schulen“), wissenschaftliche oder persönliche Interessen, die mögliche Konflikte begründen könnten	Ø	Ø	Ø
9	Gegenwärtiger Arbeitgeber, relevante frühere Arbeitgeber der letzten 3 Jahre	DHM, TU München	DHZ Berlin	Universitätsklinikum Erlangen



Univentricular Heart

univentricular heart

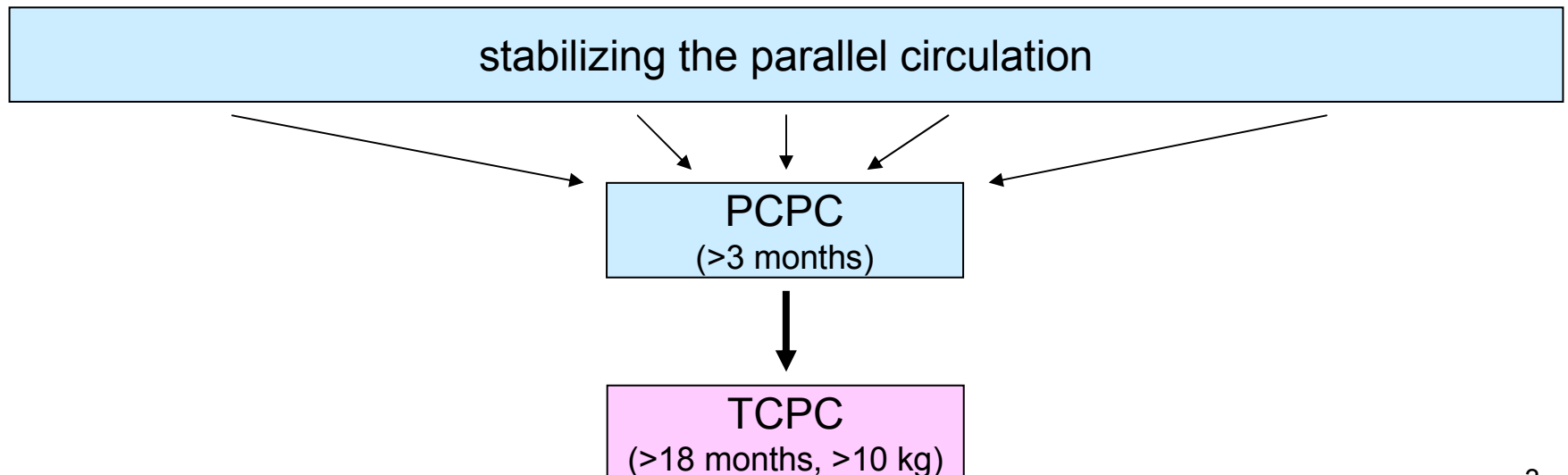
only 1 ventricle with complete inlet and pumping part

DILV, TA, HLH

„functionally“ univentricular heart

biventricular repair seems impossible

hypoplastic ventricle, straddling av-valves, severe neonatal av-regurge, coronary fistulae in CAVSD, PA-IVS, AS, MS, MA, DORV, VSD, EBS, ...





Symptoms

Varying on anatomical defect

- $\dot{Q}_p < \dot{Q}_s$ severe cyanosis, shock
- $\dot{Q}_p \approx \dot{Q}_s$ mild cyanosis, rather stable
- $\dot{Q}_p \gg \dot{Q}_s$ heart failure, shock

Lung or systemic perfusion might depend on arterial duct !



Diagnostic Tools

- Family history
- Medical history
- Physical examination
- ECG
- Blood gases, pulse oxymetry
- Chest X-ray
- **Echocardiography**
- Catheterization & angiography
- Cardiac MR / CT



Echocardiography

- anatomical diagnosis according to the segmental analysis
- abdominal situs
- myocardial function
- av-valve function
- obstructive outflow tract (AS, CoA)



Stabilizing the Parallel Circulation

Aims

- balanced or only slightly enhanced pulmonary blood flow ($\dot{Q}_p \approx \dot{Q}_s$)
- unobstructive outflow into the aorta and in the aortic arch
- free pulmonary and systemic venous return (in case of an hypoplastic av valve: free flow across an atrial septal defect)



Stabilizing the Parallel Circulation

Acute treatment

- Prostaglandine
- Intensive care

to improve systemic or pulmonary blood flow



Stabilizing the Parallel Circulation

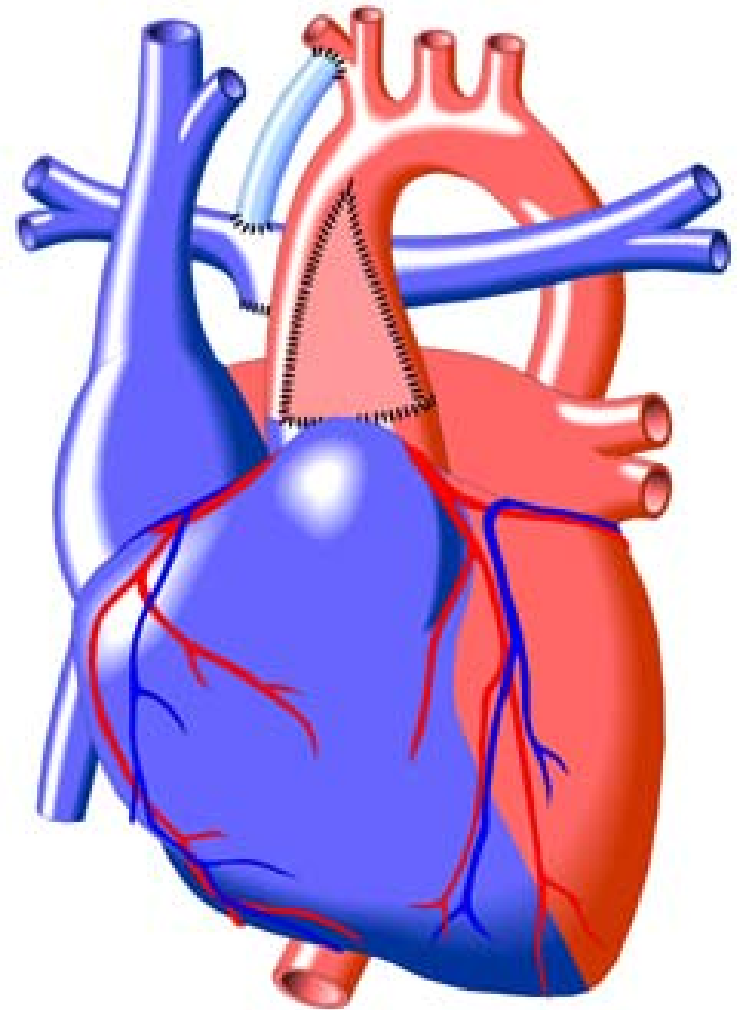
Catheter interventions

- balloon atrioseptostomy
- stent in arterial duct (experimental)

Surgery

- Shunt
 - aortopulmonary shunt
 - modified Blalock-Taussig shunt
- Closure of the pulmonary valve and shunt
- Damus-Kaye-Stansel procedure
(end-to-side anastomosis PA→Ao, modified BTS, atrioseptectomy)

Shunt -> antiplatelets (ASS, clopidogrel)





PCPC Pre-OP Evaluation

- Medical history
- Physical examination
- ECG
- Blood gases, pulse oxymetry
- Chest X-ray
- **Echocardiography**
- **Catheterization & angiography**
- Cardiac MR / CT



PCPC Pre-OP Evaluation

Echocardiography

- ventricular function
- av valve function
- aortic outflow



PCPC Pre-OP Evaluation

Diagnostic catheterization

- pulmonary vascular resistance
- transpulmonary gradient
- vessel diameter
 - Nakata index > 300 mm²/m²
 - McGoon index > 2
- ventricular function
- aortic outflow to

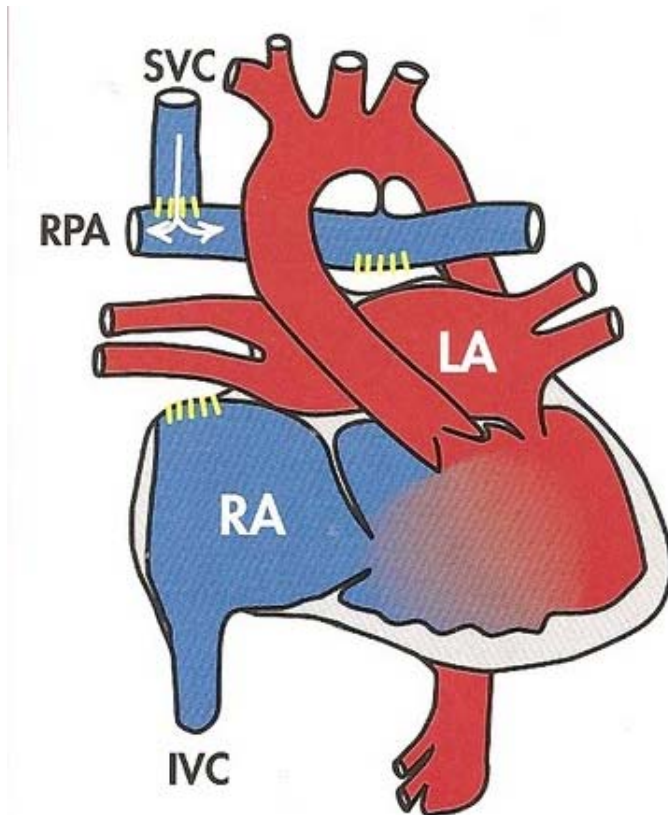
$$Nakata.index = \frac{RPA.area(mm^2) + LPA.area(mm^2)}{BSA(m^2)}$$

$$McGoon.index = \frac{RPA.diameter(mm) + LPA.diameter(mm)}{diaphragmatic.aortic.diameter(mm)}$$

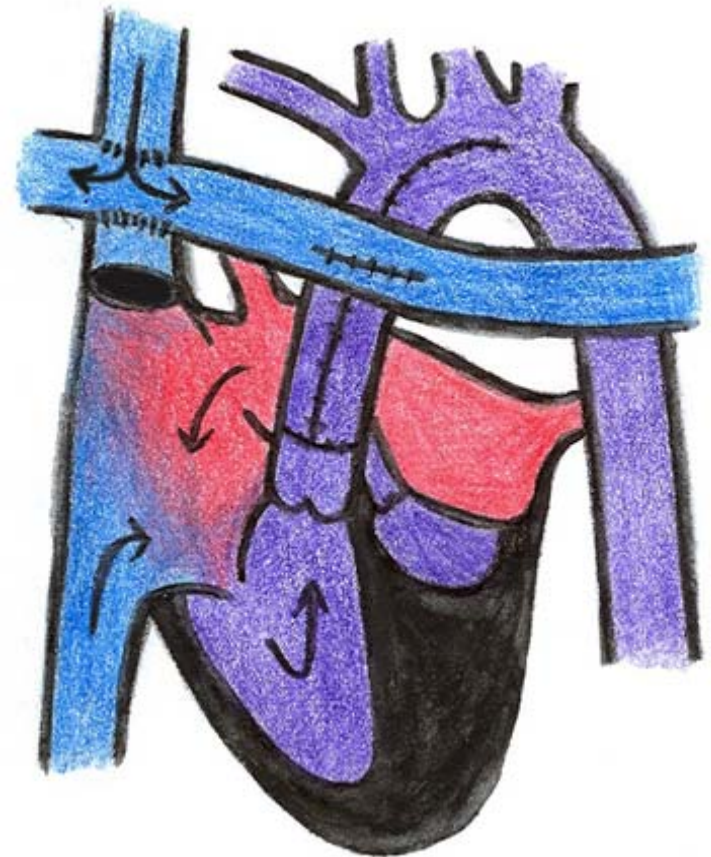
Therapeutic catheterization

- coarctation
- pulmonary artery stenosis
- embolization of major ap collateral arteries (MAPCA)

**bidirectional superior cavopulmonary shunt
(bidirectional Glenn)**



modified hemi-Fontan





PCPC post-OP Management

thromboembolic prophylaxis ?

If severely cyanotic

- dilatation, stent implantation (?) at the PCPC or pulmonary arteries
- embolization of MAPCA
- embolization of veno-venous shunts



TCPC Pre-OP Evaluation

- Medical history
- Physical examination
- ECG
- Blood gases, pulse oxymetry
- Chest X-ray
- **Echocardiography**
- **Catheterization & angiography**
- Cardiac MR / CT



TCPC Pre-OP Evaluation

Echocardiography

- ventricular function
- av valve function
- aortic outflow



TCPC Pre-OP Evaluation

Diagnostic catheterization

- pulmonary vascular resistance
- transpulmonary gradient
- vessel diameter (Nakata index, McGoon index)
- ventricular function
- aortic outflow tract and aortic arch

Therapeutic catheterization

- coarctation
- stenosis at PCPC, pulmonary artery stenosis
- embolization of major ap collateral arteries (MAPCA)
- embolization of veno-venous shunts



TCPC „The last commandments“

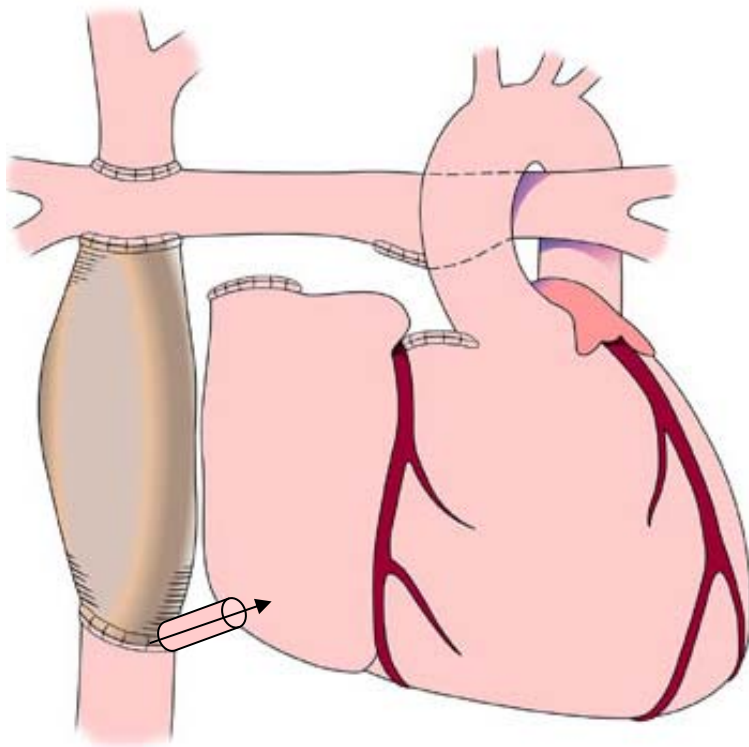
- ~~1. age > 4 years~~ 18-24 (-36) months, >10 kg
2. no tortuous pulmonary arteries (after surgery) ✓ surgery
3. normal venous return ✓ surgery
4. normal ventricular function (EF > 60%)
5. adequate size of pulmonary arteries
(PA diameter / aortic diameter >0,75)
6. No av valve regurge ✓ surgery
7. low mean pulmonary artery pressure (<15 mmHg)
8. low pulmonary vascular resistance (<4 Wood units)
- ~~9. sinus rhythm~~
- ~~10. normal sized right atrium~~



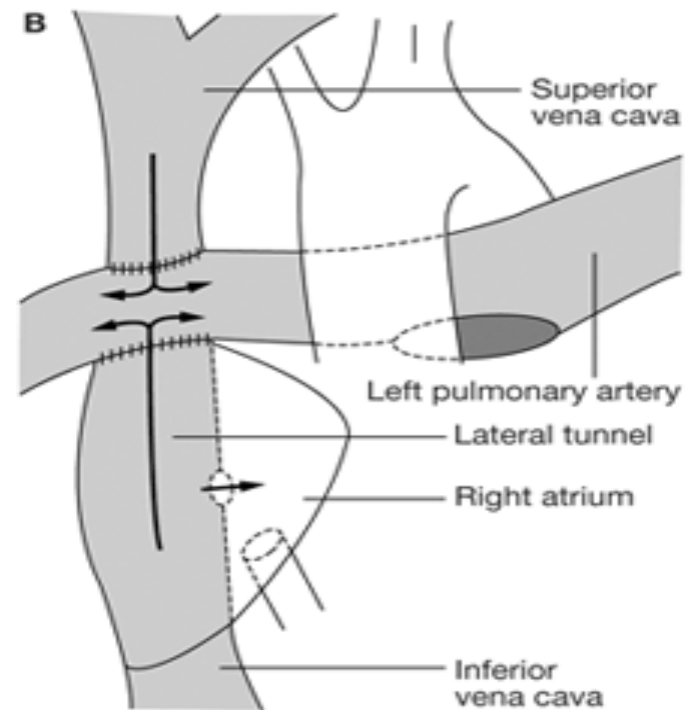
TCPC „The last commandments“

- ~~1. age > 4 years~~ **18-24 (-36) months, >10 kg**
2. no tortuous pulmonary arteries (after surgery) **✓ surgery**
3. normal venous return
4. normal ventricular function
5. adequate systemic flow (PA diameter) ideally
normal ventricular function
normal av valve function
unrestricted flow into the aorta
6. No av valve disease
7. low mean pulmonary artery pressure (ideally Mc-Goon-Ratio >2; Nakata-Index >300 mm²/m²)
mean pulmonary artery pressure <15 mmHg
8. low pulmonary vascular resistance
transpulmonary gradient <5 mmHg
Rp <4 WE×m² (ideally <2-3 WE×m²)
- ~~9. sinus rhythm~~
- ~~10. normal sized right atrium~~

Extracardial Conduit (EC-TCPC)



Intracardial Tunnel (LT-TCPC)





Fenestration ?

Fenestration Improves Clinical Outcome of the Fontan Procedure: A Prospective, Randomized Study

Matthew S. Lemler, William A. Scott, Steven R. Leonard, Daniel Stromberg and Claudio Ramaciotti

Circulation 2002;105:207-212

DOI: 10.1161/hc0202.102237

Randomized clinical trial

decreasing

- pleural drainage
- hospital length of stay
- need for additional postoperative procedures

catheter closure 6-12 months after surgery

Nonfenestrated Extracardiac Total Cavopulmonary Connection in 132 Consecutive Patients

Christian Schreiber, Jürgen Hörer, Manfred Vogt, Julie Cleuziou, Zsolt Prodan and Rüdiger Lange

Ann Thorac Surg 2007;84:894-899

DOI: 10.1016/j.athoracsur.2007.04.034

Institutional report

fenestration not necessary



TCPC Follow-up

- medical history, physical examination
- ECG, ambulatory ECG, **electrophysiologic study**
- blood gases, pulse oxymetry,
- blood tests: hemoglobin, coagulation, GGT, protein/albumin, (NT-pro)BNP (every year)
- cardiopulmonary exercise testing (if >10 years old, every 3 years)
- chest X-ray
- echocardiography
- **catheterization & angiography**
- **cardiac MR, (cardiac CT)**

if signs of Fontan failure



TCPC Follow-up

- medical history, physical examination
- ECG, ambulatory ECG, **electrophysiologic study**
- blood gases, pulse oxymetry,
- blood tests: hemoglobin, coagulation, GGT, protein/albumin, (NT-pro)BNP (every year)
- cardiopulmonary exercise testing (if >10 years old)
- chest X-ray
- echocardiography
- **catheterization & angiography**
- **cardiac MR, (cardiac CT)**

throughout life !

if signs of Fontan failure



The Failing Fontan

Symptoms and Syndromes

- „congestive right heart failure“
- right-left shunts, cyanosis
- arrhythmia (SND, atrial re-entry tachycardia)
- thrombosis, thromboembolic events
- hepatic dysfunction and fibrosis
- protein losing enteropathy
- plastic bronchitis



The Failing Fontan

Underlying pathophysiology

- stenosis at TCPC or pulmonary arteries ?
- increased pulmonary vascular resistance ?
- intracardiac stenosis, coarctation, valve regurge ?
- myocardial heart failure ?
- diaphragmatic palsy, scoliosis ?



The Failing Fontan

Underlying pathophysiology

- stenosis at T...
- increased
- intra
- valv
- myoc
- diaphrag

Detailed diagnostic work-up

Catheterization & angiography

TTE/TEE

cardiac MR (cardiac CT)

Holter ECG / EP study



The Failing Fontan

Treat underlying pathophysiology !!

- remove stenosis at TCPC or pulmonary arteries
- pulmonary vasodilators
- dilate/repair intracardiac stenosis, coarctation, repair valve regurge
- treat myocardial heart failure
- diaphragm plication, scoliosis



The Failing Fontan

Then treat the symptoms

- „congestive right heart failure“ diuretics
- right-left shunts, cyanosis shunt embolization
- arrhythmia (SND, atrial re-entry tachycardia) ablation
- thrombosis, thromboembolic events lysis, anticoagulation
- hepatic dysfunction and fibrosis ?
- protein losing enteropathy albumin substitution, budenosid
- plastic bronchitis bronchoscopic removal of casts, endotracheal lysis



The Failing Fontan

If nothing helps

- fenestration
- Fontan take-down
- heart transplantation





Thromboembolic Prophylaxis

A Multicenter, Randomized Trial Comparing Heparin/Warfarin and Acetylsalicylic Acid as Primary Thromboprophylaxis for 2 Years After the Fontan Procedure in Children

Paul Monagle, MD, MSc, MBBS,* Andrew Cochrane, MD,* Robin Roberts, MSc,†
Cedric Manlhiot, BSc,‡ Robert Weintraub, MBBS,* Barbara Szechtman, BA,†
Marina Hughes, DPHIL,§ Maureen Andrew, MD,‡ Brian W. McCrindle, MD, MPH,‡
for the Fontan Anticoagulation Study Group

Melbourne, Victoria, Australia; and Toronto and Hamilton, Ontario, Canada

- 111 patients
- randomized to 2 years of open-label
 - ASA 5 mg/kg/day
 - heparin / warfarin (INR 2-3)
- thrombosis rate (TEE 3 and 24 months after surgery, thromboembolic events) was suboptimal in both groups (21% versus 24%)
- low compliance recruitment: failed the recruitment goal of 242 patients
- low compliance follow-up: only 48% had both TEE
- low compliance warfarin therapy: 41% of all INR measurements were <2



Thromboembolic Prophylaxis

No general recommendation!

High risk factors with clear indication for oral anticoagulation

- arrhythmia
- azygos continuity
- previous thrombosis, thromboembolic event

Other risk factors

- bilateral PCPC
- fenestration, veno-venous shunts
- stenosis at TCPC
- small pulmonary arteries
- protein losing enteropathy



Thromboembolic Prophylaxis

No general recommendation!

High risk factors with clear indication for oral anticoagulation

- arrhythmia
- azygos conti
- previous

Other ri

- bilate
- fenest
- stenosis
- small pulmona
- protein losing enteropa

oral direct thrombin inhibitors
oral direct factor Xa inhibitors

not yet tested in children !!!
not yet tested in Fontan patients !!!



Infection Prophylaxis

- endocarditis
 - oral health
 - antibiotic prophylaxis for risk patients
- RSV immunoprophylaxis
- annual flu shot
- pneumococcal vaccination (P23)
- special recommendations for asplenia



Fontan Surveillance

- neuro-developmental surveillance
- life-long congenital-cardiologic surveillance

Counselling

- leisure sport is recommended
- education, profession, job
- pregnancy, contraception



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www.kinderkardiologie.org/dgpkLeitlinien.shtml