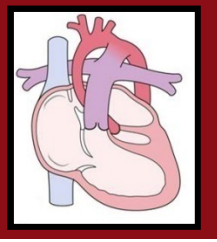
Leitlinien

Hypoplastisches Linksherzsyndrom

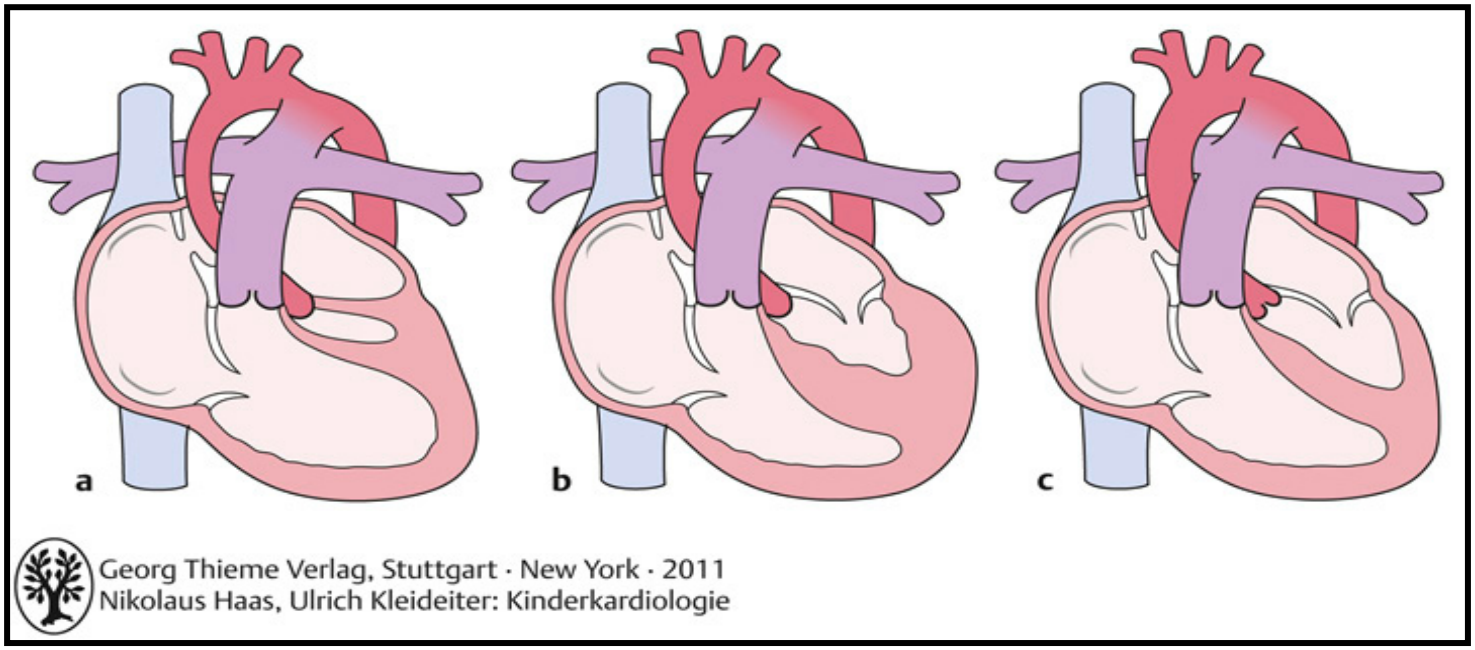


1.Title **Hypoplastic left heart syndrome (HLHS)**

N.A. Haas, Bad Oeynhausen
Ch. Jux, Giessen
J. Photiadis, Berlin
H.-H. Kramer, Kiel

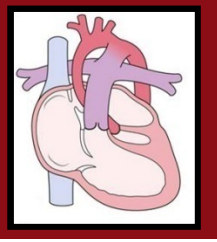
Leitlinien

Hypoplastisches Linksherzsyndrom



Typical forms:

Mitral atresia/Aortic atresia	(MA/AoA)
Mitral stenosis/Aortic stenosis	(MS/AoA)
Mitral stenosis/Aortic stenosis	(MS/AoS)
Mitral atresia/Aortic stenosis (plus VSD)	(MA/AoS)



Additional forms:

- Double outlet right ventricle (DORV) with small LV
- Imbalanced AVSD with small LV
- ccTGA with large VSD and small LV
- Long tubular SubAoS, AS and hypoplastic arch with VSD
- etc.

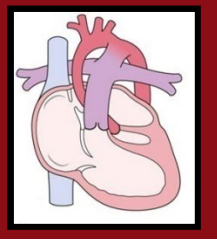
-> hypoplastic left heart complex

- hypoplastic Arch, smallisch LV, dominant RV

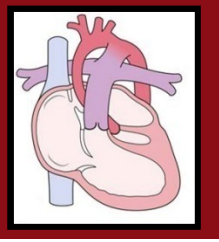
-> univentricular palliation, RV systemic ventricle

Leitlinien

Hypoplastisches Linksherzsyndrom



Epidemiology:	1-2% of all cardiac defects 2/3 male major cause for hart failure in neonates major cause for neonatal death
Pathogenesis:	hypoplasia of LV inflow and outflow secondary ventricular maldevelopment
Associated defects:	VSD anomalous pulmonary vein connections myocardial sinusoids/fistula abnormal coronary arteries tricuspid valve abnormalities abnormalities of the systemic veins

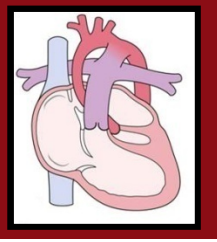


Extracardiac abnormalities:

- between 5 and 18 %
- GI tract (esophageal atresia, duodenal atresia, intestinal malrotation, diaphargmaitic hernia)
- CNS (holoprosencephalopathy, agenesia of corpus callosum, microcepahlus –> neurolog. impairment ?

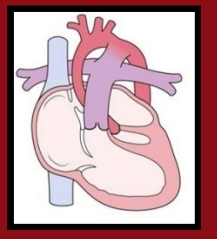
Genetic syndromes:

- Turner-syndrome, Trisomy 13,18, 21, Noonan-syndrome Smith-Lemli-Opitz-syndrom, Holt-Oram-syndrom, Jacobsen syndrome, Ellis-van-Creveld-syndrome CHARGE-Assoziation.



Postnatal decompensation (if untreated)

- constriction of the PDA	- impairment of systemic perfusion - impairment of cerebral perfusion - impairment of cardiac perfusion
- decrease of PVR	- preferntial flow to the lungs - increase of QP/QS - decrease of systemic perfusion - decrease of systemic CO
- compensation	- increase of sympathic activity - increase of SVR - tricuspid insufficiency
- result	- severe acidosis - shock -> organ failure -> death

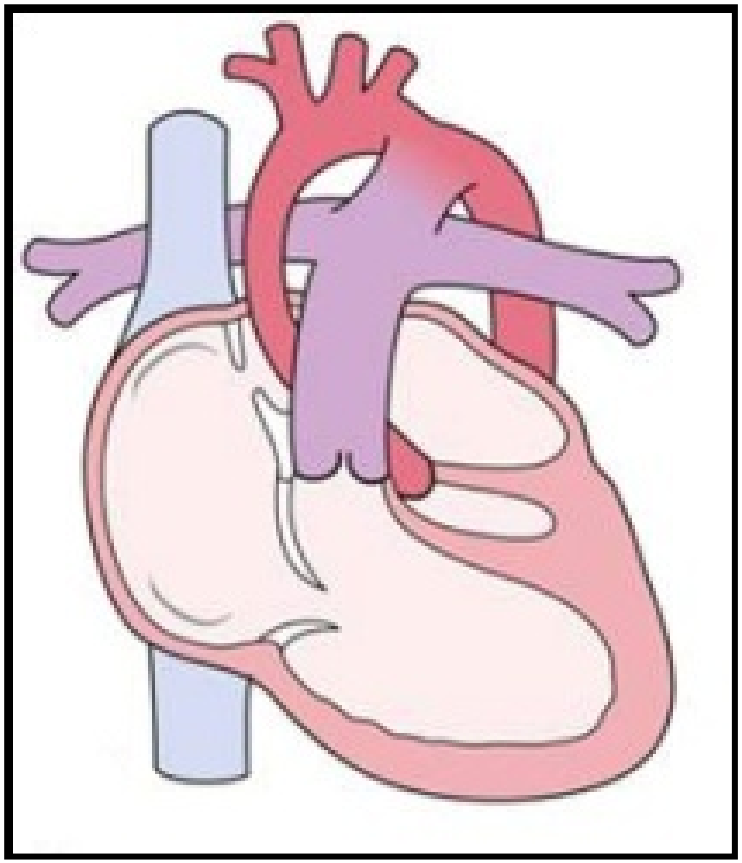


Diagnostics

- Prenatal:
  - basics for adequate postnatal care
  - delivery in a center with affiliated cardiac center experienced in HLHS
- postnatal:
  - complete diagnosis,
  - ? Secondary organ impairment ?
  - ? Additional problems ?
  - ? Genetic disorders ?
- stabilization
  - transfer to a center experienced in HLHS

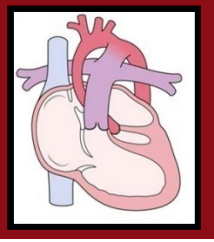
Diagnostics

- blood pressure
- pulse oxymetry
- ECG
- Chest-X-ray
- ECHO
- catheter
- MRI/CT
- pathology



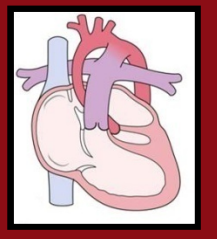
Leitlinien

Hypoplastisches Linksherzsyndrom



Primary diagnostics in HLHS

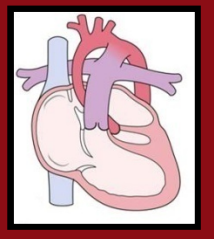
-Blood pressure	- all 4 extremities/limbs
-Pulse oxymetry	- all 4 extremities/limbs
-ECG	- not diagnostic - baseline
-Chest-X-rax	- not diagnostic – baseline pre-op
-catheter	- if pulmonary veins unclear for BAS or hybrid therapy
-MRT, CT	- not necessary
-pathology	- acidosis, organ failure infection, coagulation



Primary diagnostics in HLHS

ECHO: - diagnostic - anatomy and function

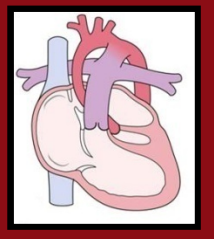
- LV: - size and function
- Aorta - perfusion, size
- arch - anatomy, A.lusoria ?
- ASD - large enough ?
- PDA - stenosis, perfusion
- RV - function, TR ?
- veins - pulmonary anatomy, stenoses ?
- systemic anatomy



Therapeutic options in HLHS

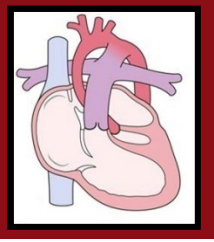
After stabilization of the newborn:

- „Compassionate care“
- Norwood-operation
- Hybrid-therapy
- cardiac transplantation



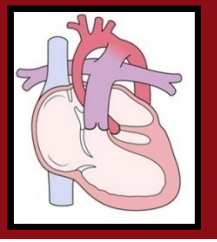
Treatment of the stable newborn with HLHS

- Prostaglandin E-Infusion	- start with 10 – 20 ng/kg/min
	- decrease to 5-10 ng/kg/min if possible
	- check ECHO, caveat: apnoea
- Treat acidosis agressively	- regular check of BGA
- Spontaneous ventilation	- extubate if possible
	- avoid intubation
	- FiO_2 0,21, avoid additional oxygen
	- accept $SatO_2$ of 75-80%
- Afterload reduction	- Na-nitroprusside, phentolamine
	- aim for low normal blood pressure
-Optimize hemoglobin	- transfusion if necessary (Hb 14)



Treatment of the newborn with HLHS in shock

- Prostaglandin E-Infusion	- start with 20 ng/kg/min
	- increase if PDA restriction
	- check ECHO
-Treat acidosis agressively	- regular check of BGA
- ventilation	- aim for $paCO_2$ of 40-45
	- if Sats are high - $paCO_2$ of 50
	- aim for paO_2 of 40 ($SatO_2$ of 75-80%)
- optimize blood pressure	- volume bolus
- optimize cardiac function	- milrinone, rarely catecholamines
- Afterload reduction	- careful
- optimize hemoglobin	- transfusion if necessary (Hb 14)



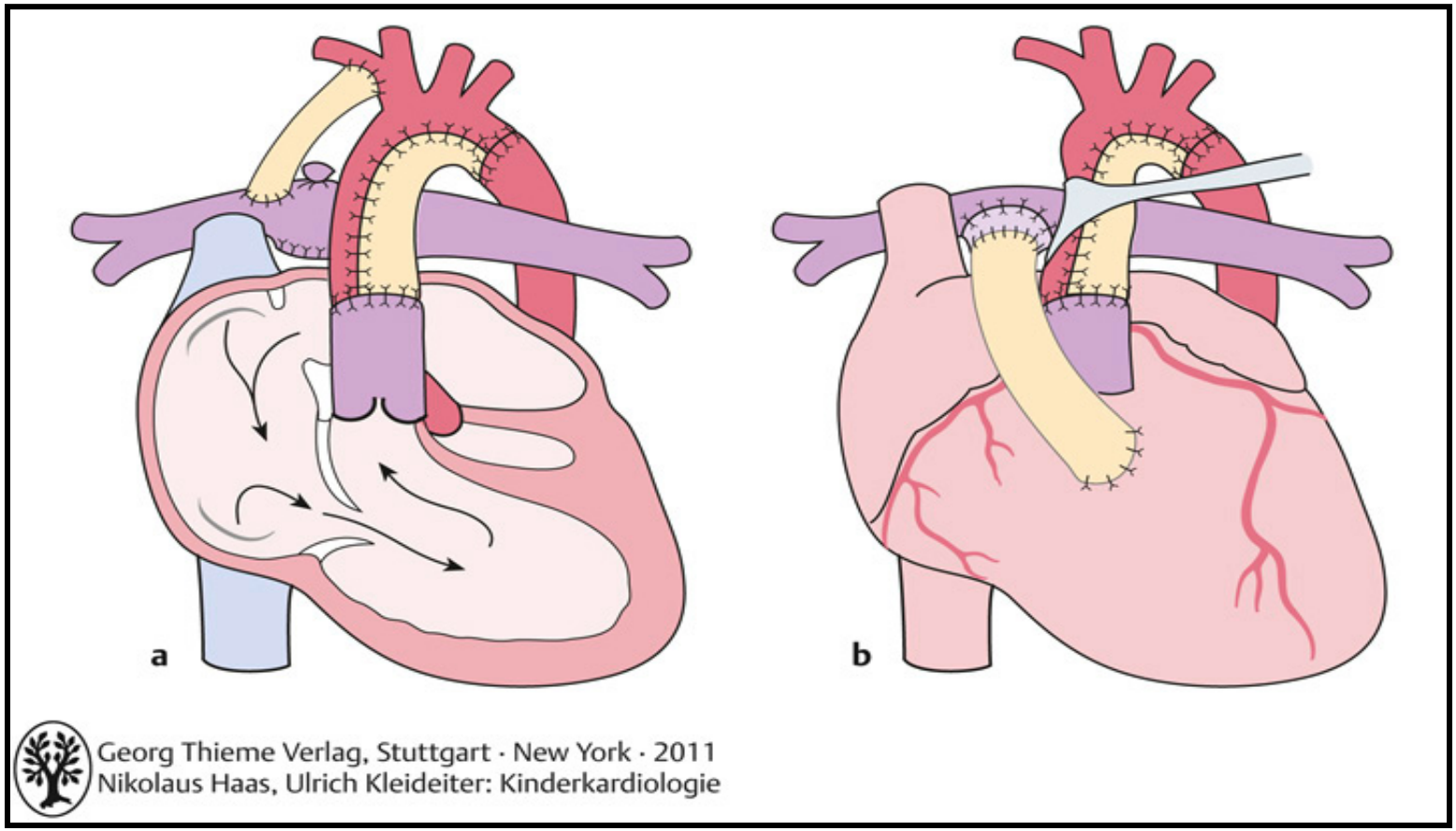
Surgical treatment options

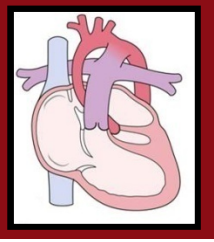
Principle:
- at the end of the first week of life
- stenosis free flow to the heart
  ->pulmonary veins, atrial septum
- stenosis free flow to the body
  ->aortic arch
- adequate arch and coronary perfusion
- balanced pulmonary perfusion

2 surgical options
- classical Norwood operation
- Norwood-Sano operation

Leitlinien

Hypoplastisches Linksherzsyndrom



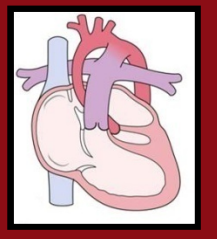


Surgical treatment options

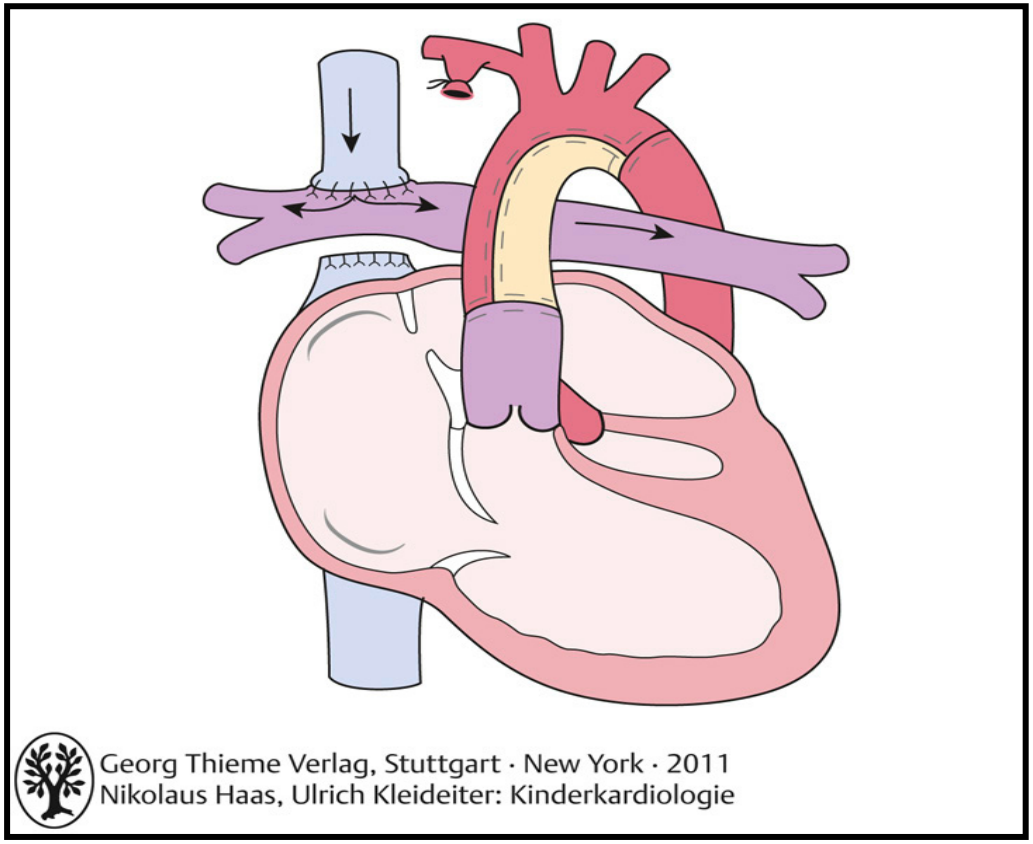
Norwood operation: atrioseptectomy, arch augmentation
modified Blalock-Taussig shunt

Norwood-Sano-operation: atrioseptectomy, arch augmentation
5-6 mm Goretex conduit RV-> PA
no diastolic run-off
RV-scar, PA-scars

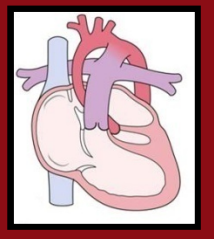
Results: 30 day mortality about 20%



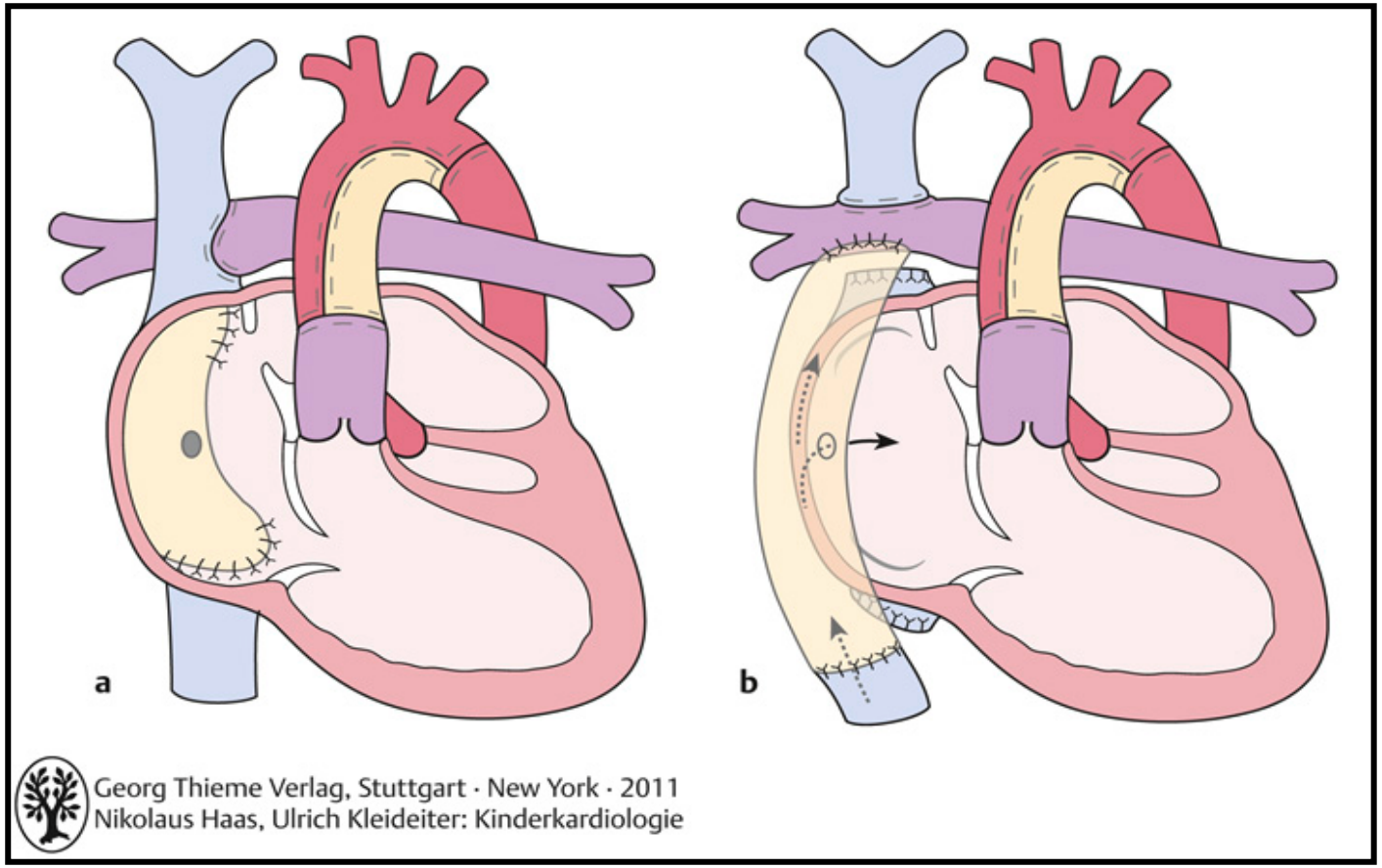
2. step: Glenn (superior) cavopulmonary anastomosis



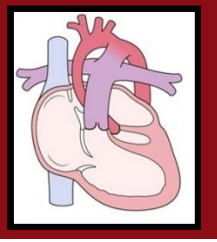
Age 4-6 months, if necessary tricuspid valve repair



3. step: total cavopulmonary anastomosis



Age 2-3 years, extra- intracardiac tunnel, fenestration ?

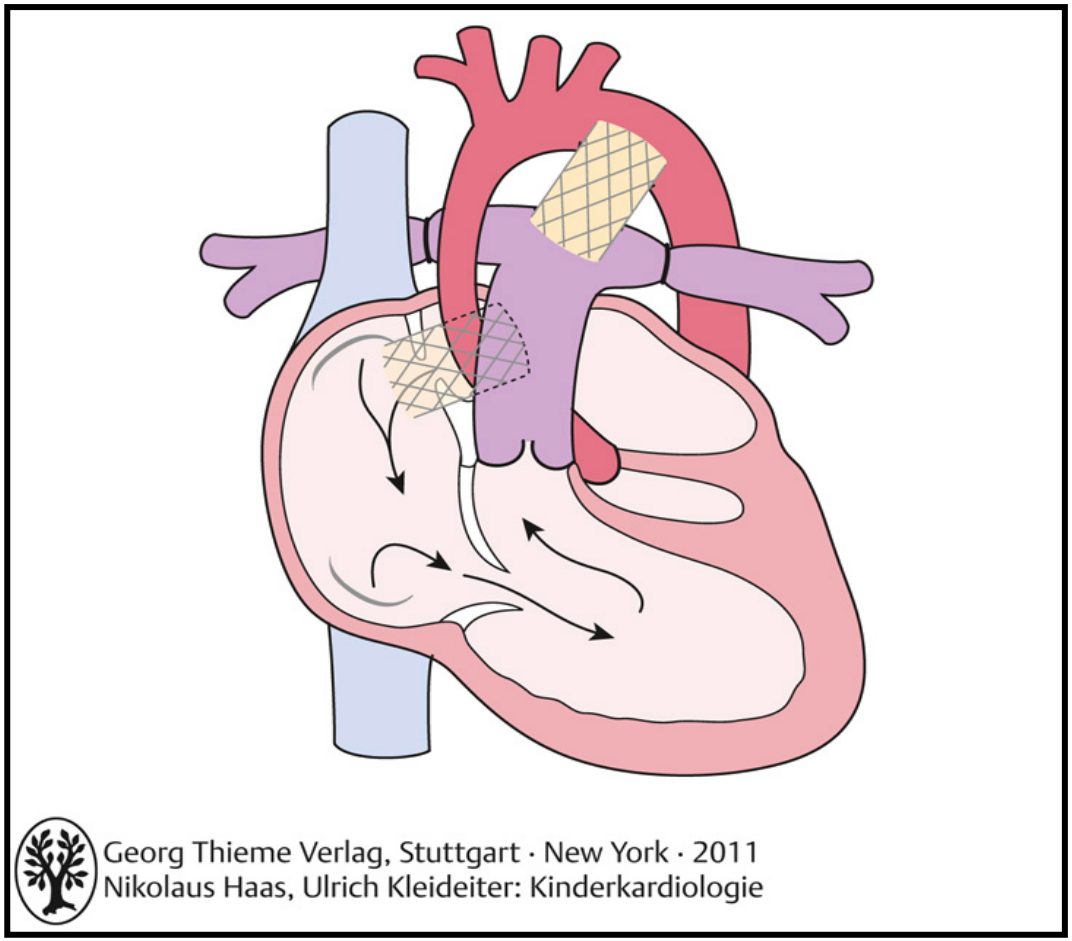


Hybrid therapy – palliation of HLHS

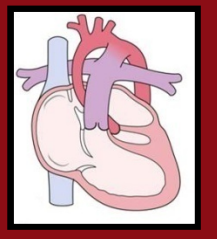
-Treatment option	- alternative - in complications (NEC, etc.)
- technique:	- bilateral banding - PDA stenting – systemic perfusion - ASD (BAS or Stent)
-Step II	- „comprehensive stage II“ - Norwood I and II

Leitlinien

Hypoplastisches Linksherzsyndrom



Georg Thieme Verlag, Stuttgart · New York · 2011
Nikolaus Haas, Ulrich Kleideiter: Kinderkardiologie

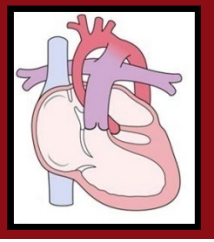


Hybrid therapy – palliation of HLHS

- advantage
  - no cardiopulmonary bypass
  - no SIRS etc. in newborn age
  - usable in complications (NEC, etc.)
  - bridge to transplant
  - borderline LV -> growth ?
    -> biventricular ?

- disadvantage
  - bilateral banding – scars at PA
  - coarctation – brain perfusion ?
  - high rate of reinterventions

- results
  - no benefit regarding morbidity/mortality
  - no benefit regarding neurologic outcome



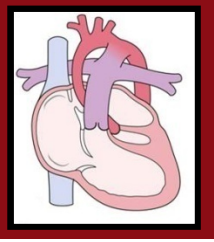
Cardiac transplantation in HLHS

- no veritable treatment option in newborns

- shortage of donor organs

- in failing hearts at any stage of palliation possible

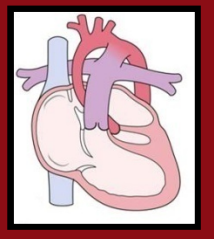
- mortality comparable to Norwood- operation



Compassionate care in HLHS

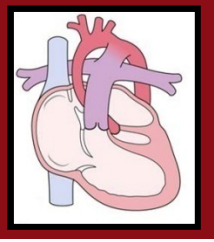
- has to be adressed in patient consultation

- is veritable treatment option in newborns
  - high late morbidity and mortality
  - impaired life expectancy
  - neurologic long term follow-up



Follow-up after HLHS palliation

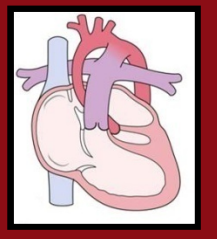
f/u after stage I:	- close follow-up
	- ECHO: coarctation, heart failure
	ASD, TR
	- saturation: 75-80%
	shunt stenosis, QP/QS
f/u after hybrid stage I:	- flow PA bands, flow PDA -> aorta
	- coarctation, retrograde flow, ASD ?
Medication:	- ASS
Nutrition:	- often feeding problems



Follow-up after HLHS palliation

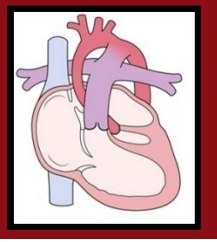
Interstage mortality :
- up to 15%
- reason unclear
- close follow-up ->

Home monitoring programs:
daily weight, saturations
weekly telephone calls
high alert to infections
early hospital admission
close contact to pediatricians



Late follow-up after HLHS palliation

somatic	- impaired exercise capacity - reduced life expectancy - RV systemic ventricle, TR
social	- no normal professional development - insurance ?
neurology	- very high rate of neurologic impairment - decerased mean IQ - early supportive measures necessary - modern surgical techniques ?
transplant	- immunosuppression - transplantation service



Prevention of HLHS

- Not possible

- Fetal ECHO should be used
  - decision making
  - optimal postnatal care

- Discrete increased familiar risk ?- genetic counselling

- Intrauterine treatment
  - not successful/high risk