1. Title  
Hypoplastic left heart syndrome (HLHS)

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Typical forms:
Mitral atresia/Aortic atresia  (MA/AoA)
Mitral stenosis/Aortic stenosis  (MS/AoA)
Mitral stenosis/Aortic stenosis (plus VSD)  (MS/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
Epidemiology: 1-2% of all cardiac defects
2/3 male
major cause for heart 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, diaphragmatic hernia)
- CNS (holoprosencephalopathy, agenesia of corpus callosum, microcephalus → neurolog. impairment ?

Genetic syndromes:
Postnatal decompensation (if untreated)

- constriction of the PDA
- impairment of systemic perfusion
- impairment of cerebral perfusion
- impairment of cardiac perfusion

- decrease of PVR
- preferential flow to the lungs
- increase of QP/QS
- decrease of systemic perfusion
- decrease of systemic CO

- compensation
- increase of sympathetic 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
### 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 aggressively - regular check of BGA

- Spontaneous ventilation - extubate if possible
  - avoid intubation
  - FiO₂ 0,21, avoid additional oxygen
  - accept SatO₂ 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 aggressively
  - regular check of BGA
  - 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%)

- ventilation
- optimize blood pressure
  - volume bolus

- optimize cardiac function
  - milrinone, rarely catecholamines
  - careful

- Afterload reduction
- 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
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 $\rightarrow$ growth?
  $\rightarrow$ 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 addressed 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
- decreased 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