Failing right ventricle

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Prenatal Physiology

Right ventricle
- dominant ventricle
- 59% combined cardiac output

Right ventricular impairment
- ↑ flow foramen ovale
- ↑ performance of the LV

% biventricular cardiac output

Rudolph 2001, Mielke, 2001
Severe fetal right ventricular failure:

- **systolic impairment** (contractility ↓)
- PA-Flow ↓

- **diastolic impairment**
  - Tricuspid regurgitation
  - Doppler venous duct ± Umbilical arteries

- ascites/pericardial effusion/hydrops
Conditions with failing RV

- Myocardial Disease

- Volume / Pressure Overload

- Structural heart disease
Case I  27+4

Cardiomegaly, RV dilatation, myocardial thinning
Case I

PA + arterial duct: antegrade flow
Pulmonary regurgitation

normal pulsatility venous duct

27+4
Suspected: Uhl’s Anomaly

Definition: hypoplastic myocardium of the RV free wall

Histology: apposition of pericardial & endocardial layers absence of musculature

Fetal Echocardiography:
- RV Dilatation
- thinning of the anterior wall of the RV
- RV hypocontractility
- ± abnormal tricuspid valve

Uhl, 1952; Uhl, 1996; Gerlis, 1993; Vaujois, 2014
Case I at 33+6

Functional pulmonary atresia
Duct: retrograde Flow
pericardial effusion

ductus venosus:
neg. atrial contractions

33+6
Prenatal diagnosis: Uhl’s Anomaly

CS, ventilated with NO and O2

RV + LV hypocontractil
Postnatal follow up

Ductal dependend pulmonary circulation
PGE2, adrenaline, NO, O₂

48 hrs later
anterograde pulmonary perfusion
Follow up

3 weeks

MRI no fatty infiltrations, dilated RV, thinning of RV myocardium

6 years
Uhl’s Anomaly - outcome

intrauterine progression:
- failing RV with hydrops
- Fetal or neonatal death

postnatal
- Various presentation
- Fontan/1 ½ ventricle/biventricular
- arrhythmia
- partial forms in adults

Case II  36+6
Dilated RV, prominent coarse trabeculations at the RV apex
RV non-compaction

Dilated RV

Prominent coarse trabeculations

Endocardial fibroelastosis

normal non compaction

Ursell, 2013
RV non-compaction

**Definition:** abnormal excessive trabeculation

**Clinical:**
- Cardiac dysfunction/heart failure/hydrops
- Arrhythmias
- Thrombi
- Endocardial fibrosis

**Isolated/associated with CHD**

Ursell, 2013
Postnatal follow up – day 1
Postnatal follow up – 6/12
Not the same with biventricular non-compaction
Case III
severe RV dysfunction  22+1

+ functional pulmonary atresia
+ reverse flow in the duct
+ pericardial effusion
+ EFE RV

Ductus venosus: reverse a Pulsatile umbilical vein
Case III
severe RV dysfunction 28+2

Ventricular tachycardia – 28+2. SSW
Ventricular rate 218/min, atrial rate 143/min
Fetal hydrops - Cesarian
Postnatal course

- CS 28+ wks, 1300 g
- Severe biventricular impairment

FS 12%
AV-Block 2:1 with VES
Nurses: funny ECG

Long QT-Syndrome Typ 2
AV Block II.

RV Cardiomyopathy
Pulmonary Stenosis

β-Blocker

TdP
Pace-Maker

(1300 g)
Ventricular aneurysm/diverticle

- Rare, RV<<LV
- Outpouching of the RV
- dependent on size
- conservative Thx

Oloron, Ped Cardiol 2011
Pressure and Volume Load
Constriction of the fetal ductus arteriosus

Arterial Duct
39% of the biventricular cardiac output via duct

Ductal occlusion
increased RV afterload
Increased FO flow
Increased pulmonary flow

Rudolph 2001, Hofstadler 1996
Hashima 2012, Zielinsky 2013
Twin 1, 27+2 GA, Indometacin-Thx
Definition of constriction of arterial duct

- Turbulent flow in the duct
- Systolic velocity > 1.4 m/s
- Diastolic velocity > 0.3 m/s
- PI < 2.2 m/s
- Absent flow = total occlusion

*Tulzer 1991, Zielinsky 2013*
Sonographic features in isolated DA closure

<table>
<thead>
<tr>
<th>Finding</th>
<th>Number (n)</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right atrium</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dilation</td>
<td>8</td>
<td>75</td>
</tr>
<tr>
<td>Tricuspid valve regurgitation (moderate–severe)</td>
<td>12</td>
<td>100</td>
</tr>
<tr>
<td>Right ventricle</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hypertrophy</td>
<td>12</td>
<td>100</td>
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<tr>
<td>Bipartite</td>
<td>8</td>
<td>75</td>
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<tr>
<td>Pulmonary valve</td>
<td></td>
<td></td>
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<tr>
<td>Stenosis</td>
<td>2</td>
<td>17</td>
</tr>
<tr>
<td>Regurgitation (moderate–severe)</td>
<td>11</td>
<td>92</td>
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<tr>
<td>Dysplastic</td>
<td>3</td>
<td>25</td>
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<tr>
<td>Pulmonary artery</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dilatation trunk and branch pulmonary arteries</td>
<td>5</td>
<td>42</td>
</tr>
<tr>
<td>Foetal suprasystemic pulmonary pressure (measurement TR)</td>
<td>2</td>
<td>17</td>
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<tr>
<td>Functional pulmonary atresia</td>
<td>1</td>
<td>8</td>
</tr>
<tr>
<td>Other</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pericardial effusion</td>
<td>1</td>
<td>8</td>
</tr>
<tr>
<td>Aneurysm ductus with thrombus</td>
<td>1</td>
<td>8</td>
</tr>
<tr>
<td>Hydrops foetalis</td>
<td>1</td>
<td>8</td>
</tr>
<tr>
<td>Microcystic lung changes</td>
<td>1</td>
<td>8</td>
</tr>
</tbody>
</table>

- Severe RV Hypertrophy
- Tricuspid valve regurgitation
- Pulmonary valve regurgitation

Gewillig, 2009
## Outcome in isolated DA closure

### Table 3 Management and outcomes

<table>
<thead>
<tr>
<th></th>
<th>Number (n)</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Foetal</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Premature induction of delivery</td>
<td>5</td>
<td>62</td>
</tr>
<tr>
<td>Neonatal treatment</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Observation and oxygen administration</td>
<td>7</td>
<td>58</td>
</tr>
<tr>
<td>Ventilation with administration of pulmonary vasodilators</td>
<td>5</td>
<td>42</td>
</tr>
<tr>
<td>ECMO</td>
<td>1</td>
<td>8</td>
</tr>
<tr>
<td>Surgery</td>
<td></td>
<td></td>
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<tr>
<td>Resection thrombosed aneurismal ductus—occlusion LPA</td>
<td>1</td>
<td>8</td>
</tr>
<tr>
<td>Outcome/follow-up</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neonatal death</td>
<td>3</td>
<td>25</td>
</tr>
<tr>
<td>Cardiovascular</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Balloon angioplasty of PS (7 months) and closure ASD (9 years)</td>
<td>1</td>
<td>8</td>
</tr>
<tr>
<td>Homograft reconstruction pulmonary artery</td>
<td>1</td>
<td>8</td>
</tr>
<tr>
<td>Cardiomyopathy (non-compaction)</td>
<td>1</td>
<td>8</td>
</tr>
<tr>
<td>Psychomotor delay, mild</td>
<td>2</td>
<td>17</td>
</tr>
</tbody>
</table>

ECMO, extracorporeal membrane oxygenation; LPA, left pulmonary artery; PS, pulmonary valve stenosis; ASD, atrial septal defect. See text for details.

- Fetal hydrops
- IUFD
- Premature delivery
- Relevant early and late morbidity
- Pulmonary hypertension

Gewillig, 2009
Twin-Twin Transfusion Syndrome - TTTS

- imbalance of placental anastomoses in monochorionic-diamnotic twin pregnancies

- volume shift from donor to recipient

- Recipient: increased pre- & afterload
  - Myocardial hypertrophy
  - Diastolic dysfunction
  - Cardiac failure
# Preoperative cardiac function

## TABLE 3
Preoperative cardiac function in TTTS

<table>
<thead>
<tr>
<th>Variable</th>
<th>Control subjects (n = 172)</th>
<th>Donors (n = 39)</th>
<th>P value, donors</th>
<th>Recipients (n = 39)</th>
<th>P value, recipients</th>
</tr>
</thead>
<tbody>
<tr>
<td>LV-MPI (z score)</td>
<td>-0.02 ± 1.00</td>
<td>-0.24 ± 1.09</td>
<td>.19</td>
<td>2.43 ± 2.47</td>
<td>&lt; .001</td>
</tr>
<tr>
<td>RV-MPI (z score)</td>
<td>-0.01 ± 1.00</td>
<td>-0.24 ± 1.21</td>
<td>.41</td>
<td>2.84 ± 1.89</td>
<td>&lt; .001</td>
</tr>
<tr>
<td>DV abnormal, n (%)</td>
<td>0 (0)</td>
<td>2 (6)</td>
<td>.03</td>
<td>14 (36)</td>
<td>&lt; .001</td>
</tr>
<tr>
<td>UV pulsations, n (%)</td>
<td>2 (1)</td>
<td>1 (3)</td>
<td>.09</td>
<td>12 (31)</td>
<td>&lt; .001</td>
</tr>
<tr>
<td>MI, n (%)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>NA</td>
<td>5 (13)</td>
<td>&lt; .001</td>
</tr>
<tr>
<td>TI, n (%)</td>
<td>1 (1)</td>
<td>1 (3)</td>
<td>.34</td>
<td>16 (41)</td>
<td>&lt; .001</td>
</tr>
<tr>
<td>Mitral fusion, n (%)</td>
<td>1 (1)</td>
<td>0 (0)</td>
<td>1.00</td>
<td>10 (26)</td>
<td>&lt; .001</td>
</tr>
<tr>
<td>Tricuspid fusion, n (%)</td>
<td>4 (2)</td>
<td>0 (0)</td>
<td>1.00</td>
<td>14 (36)</td>
<td>&lt; .001</td>
</tr>
</tbody>
</table>

*DV, ductus venosus; LV, left ventricle; MI, mitral insufficiency; MPI, myocardial performance index; NA, not available; RV, right ventricle; TI, tricuspid insufficiency; TTTS, twin-to-twin transfusion syndrome; UV, umbilical vein.

## Postoperative cardiac function

### TABLE 4
Preoperative and short-term postoperative cardiac function in TTTS

<table>
<thead>
<tr>
<th>Variable</th>
<th>Donors (n = 33)</th>
<th>Recipients (n = 35)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Preoperative</td>
<td>24-48 h postoperative</td>
</tr>
<tr>
<td>LV-MPI (z score)</td>
<td>-0.29 ± 1.15</td>
<td>-0.73 ± 1.89</td>
</tr>
<tr>
<td>RV-MPI (z score)</td>
<td>-0.29 ± 1.24</td>
<td>-0.42 ± 1.46</td>
</tr>
<tr>
<td>DV abnormal, n (%)</td>
<td>2 (6)</td>
<td>8 (24)</td>
</tr>
<tr>
<td>UV pulsations, n (%)</td>
<td>1 (3)</td>
<td>4 (12)</td>
</tr>
<tr>
<td>MI, n (%)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>TI, n (%)</td>
<td>1 (3)</td>
<td>4 (12)</td>
</tr>
<tr>
<td>Mitral fusion, n (%)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Tricuspid fusion, n (%)</td>
<td>0 (0)</td>
<td>1 (3)</td>
</tr>
</tbody>
</table>

*DV, ductus venosus; LV, left ventricle; MI, mitral insufficiency; MPI, myocardial performance index syndrome; UV, umbilical vein.*

*Van Mieghem, Assessment of fetal cardiac function before and after therapy for TTTS. Am*
Conditions with failing RV

❤️ **Myocardial Disease**
- Uhl’s Anomaly
- RV Non-Compaction
- Diverticula/Aneurysms

❤️ **Volume / Pressure Overload**
- TTTS
- Ductal constriction
- Shunts: AV-malformations, ductus venosus agenesis,…

❤️ **Structural heart disease**
- Ebstein’s Anomaly
- Absent pulmonary valve syndrome
Postnatale Adaptation

Fall of pulmonary vascular resistance

Morin, 1992, Soifer, 1989
Perinatal Management of Fetal right heart failure

Optimize ventricular function

RV  pulmonary vascular resistance ↓
keep duct open until LR-Shunt

LV  +/- careful catecholamin thx

Reduction of pulmonary vascular resistance

NO, O₂, Ventilation

Gain time
Predictors of perinatal outcome in fetal CMP

Dysfunction of both ventricles  p 0.01

Holosystolic AV-regurgitation  p 0.002

Abnormal diastolic Function
-DV neg a
-UV Pulsation  p 0.047

N=55

*Predictors of perinatal outcome in fetal CMP*
*Pedra, ... Hornberger, Circulation 2002*
Conclusion

- The course of the failing fetal RV is dependent on the function of the remaining LV to obtain cardiac output

- cut off values to predict perinatal outcome are still missing

- postnatally, fall of RV afterload allows improvement of RV function→ wait and see
Thank you for your attention