Antenatal & Neonatal Cardiac Diagnosis: When active management is not possible

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Congenital cardiac lesions

Cardiac lesions which are possible to diagnose antenatally

- AVSD, VSD
- Aortic stenosis
- Pulmonary stenosis
- Coarctation of aorta/Hypoplastic aortic arch
- Cardiac masses
- Transposition of great arteries (TGA)
- Tetralogy of Fallot; Pulmonary atresia/VSD
- Truncus arteriosus
- Unbalanced ventricles – HLHS; Tricuspid atresia; complex combination

Cardiac lesions which cannot be diagnosed till after birth

- ASD
- PDA
- Anomalous pulmonay veins
- Milder forms of aortic stenosis, pulmonary stenosis, coarctation of aorta
Congenital structural cardiac lesion

- Diagnosis
- Management
- Options
Management

Multidisciplinary team

- Paediatric cardiologist (and cardiac surgeon)
- Obstetrician
- Radiologist
- Paediatrician/Neonatologist
- Cardiac liaison sister / midwife
- Palliative care

Aim

- Provide diagnosis/information for parents
- Options explained
- Planned delivery
- Early transfer to tertiary cardiac centre for further management
- Avoid postnatal cardiovascular collapse
Management

• Counselling / information
• Further tests (rule out chromosomal association, extracardiac anomaly)
• Other teams involved – fetal medicine, genetics, palliative care team, etc
• Follow-up
• Plan for delivery/management
Management

Diagnosis

Counselling
(Further investigations, treatment, options explained, cardiac liaison sister)

Follow-up review
(evolution of lesion)

Planned delivery
(issues with prematurity, size)

Postnatal review and management

IUD / Termination

Active Management

Compassionate Care
Management of delivery

Ideal management of delivery

– Those fetuses who are duct or “mixing” dependent should be delivered at (or close to) the site of cardiac surgery

– Rarely necessary to alter timing or mode of delivery
Management of CHD

• Prognosis worsened if associated
  – non-cardiac issues
  – chromosomal association
  – Prematurity/small for gestational age

• **Most of the spectrum** of antenatally diagnosed cardiac lesions
  – May progress in severity or get better with gestation of pregnancy
  – Some will need only medical intervention (medications/watchful wait)
  – Some major structural cardiac lesions will need “corrective” surgery in the first year of life (single or multiple staged surgery)
  – For some cardiac lesions – despite surgery, the heart is not entirely normal (only near normal) with limitations on exercise ability
  – Most have good long term survival

• **Small percentage** of antenatally diagnosed cardiac lesions
  – May not be amenable to “corrective” surgery, hence only “palliative surgery” – to compensate for structural deficiencies
  – Some patients will eventually deteriorate with time and have limited long-term survival
  – Variable quality of life
Management

Diagnosis

Counselling
(Further investigations, treatment, options explained, cardiac liaison sister)

Follow-up review
(evolution of lesion)

Planned delivery
(issues with prematurity, size)

Postnatal review and management

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Active Management

Compassionate Care
Postnatal management options for complex cardiac lesions not suitable for corrective surgery

- Active management with palliative surgery (non-corrective)
- Compassionate care
Unbalanced Ventrices - Palliative surgery (HLHS; Tricuspid atresia; Unbalanced AVSD)

- Usually prostaglandin dependent
- 3 stages in the first 4-6 years of life
  - Stage 1: usually 7-10 days of life (50-70% survival)
  - Stage 2: 4-6 months of life
  - Stage 3: 4-6 years of life (30% survival in long-term)
- Survival: usually 20-30 years
- Quality of life
  - variable with gradual deterioration over time
  - Long-term medications
  - Life-long follow-up
  - May need more interventions
  - Transplant may be an option, although not realistic (donor heart availability; other organ dysfunction)
Stage 1

• Usually done at around 7-10 days of life

• Options:
  – Norwood/Norwood-Sano procedure (for HLHS)
  – BT shunt or stent of PDA (for Tricuspid atresia; pulmonary atresia with small RV)
  – Sometimes delayed to 4-6 weeks – Pulmonary artery band (to limit amount of blood flow to the lungs)
Norwood/Norwood-Sano Procedure

- For hypoplastic left ventricle
- To enlarge body vessel and ensure good flow to rest of body
- Limit amount blood flow to lungs
BT shunt or PDA stenting

Tricuspid or pulmonary atresia
• The systemic/body blood supply is adequate
• BUT limited blood supply to lungs

Aim to maintain continuity of blood supply to lungs
• Prostaglandin (short term use)
• BT shunt (surgical procedure)
• PDA stenting (catheter intervention)
Pulmonary artery banding

For unbalanced ventricles with
• Good size aorta
• Too much blood flow to the lungs

Options:
Conventional band (using dacron tape - fixed)
Flowatch PA band (adjustable via external magnetic control)
Compassionate care

• For duct dependent lesions (prostaglandin dependent)
  – If prostaglandin stopped → duct will close (variable duration: hours-days-weeks)
  – Progressive reduction in systemic/pulmonary blood flow
  – Progressive hypoxia/systemic dysfunction

• Non duct dependent lesions (complex anatomy with precarious balance of circulation)
  – May survive for variable duration
  – Progressive deterioration with time (months to years)
  – Surgery may occasionally hasten death
Stage 2: 4-6 months of life

Cavopulmonary anastomosis

• To ensure more definitive blood supply to the lungs

• Patient still remains relatively “blue” (saturations in 80s)
Stage 3: 4-6 years of life

Fontan surgery

- Connecting all venous blood (SVC, IVC, hepatic veins) to pulmonary circulation
- Separating pink from blue blood
- Using larger/stronger ventricle to pump blood to systemic circulation
- Patient now much “pinker” (saturations low-mid 90s)
Audit of Outcome of Unbalanced Cardiac Ventricles
Data

Retrospective review of all patients born with unbalanced ventricles between 2001 and 2011, from
– Antenatal records/ fetal database at Liverpool Women’s Hospital
– Cardiac database at Alder Hey Children’s Hospital.

Fetuses with borderline small Right or Left sides also included. Postnatally, only confirmed unbalanced ventricles were included (ie suitable for less than biventricular repair).

Comparison made between the AN and PN groups.
Outcomes of 212 affected pregnancies

- TOP: 90
- IUD/SB: 12
- Born alive: 110
On a yearly basis the mean TOP rate was 46.9% (± SD 23%); mean livebirth rate was 44%
Postnatal management of liveborn infants with unbalanced ventricles

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<thead>
<tr>
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<th>Active Management</th>
<th>Compassionate care</th>
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<tbody>
<tr>
<td>Antenatal Dx (110)</td>
<td>76</td>
<td>34</td>
</tr>
<tr>
<td>Postnatal Dx (51)</td>
<td>48</td>
<td>3</td>
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Follow-up of patients

• The cohort has been under follow-up for a decade.
  – 52 (68%) of 76 actively managed AN patients are alive.
  – 35 (72%) of 48 actively managed PN patients are alive.

• Long term survival does not differ significantly between AN versus PN -diagnosed patients with unbalanced ventricles
Descriptive Review of Neonatal Compassionate Care for Antenatally and Postnatally Diagnosed Complex Cardiac Diagnosis

R Ramaraj, L Brook, C Jones, G Gladman, D Roberts, J Lim
Alder Hey Hospital, Liverpool Women’s Hospital, Manchester Children’s Hospital
Method

• Between 2009-2014
• Descriptive retrospective review
• Patients with complex congenital cardiac diagnosis (CHD) with decision for compassionate care
• Diagnosed antenatally (AN) or postnatally (PN)
• Patients from Northwest and North Wales
Results

• 24 neonates whose parents elected for compassionate care
• 14 AN ; 10 PN
• 6 patients with lethal trisomies (2 AN, 4 PN) with cardiac defects
• 18 patients with complex CHD
  – 13 HLHS
  – 2 unbalanced AVSD
  – 1 TAPVD with veno-occlusive disease
  – 1 PA/VSD/Aortic obstruction
  – 1 PA/IVS
## Results

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<thead>
<tr>
<th></th>
<th>AN dx</th>
<th>PN dx</th>
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<tbody>
<tr>
<td>Alive</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Dead</td>
<td>12</td>
<td>9</td>
</tr>
<tr>
<td>Total</td>
<td>14</td>
<td>10</td>
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## Results

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<tr>
<th>Place of compassionate care</th>
<th>AN diagnosis</th>
<th>PN diagnosis</th>
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<tbody>
<tr>
<td>Hospice</td>
<td>6 (5 complex cardiac; 1 lethal trisomy)</td>
<td>4 (4 lethal trisomy)</td>
</tr>
<tr>
<td>Home</td>
<td>4 (3 complex cardiac; 1 lethal trisomy)</td>
<td>3 (3 complex cardiac)</td>
</tr>
<tr>
<td>Hospital</td>
<td>2 (2 complex cardiac)</td>
<td>2 (2 complex cardiac)</td>
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Findings

• Preference for hospice/home care for families who have had AN contact with Palliative Care team

• Preference for hospital care for those diagnosed PN
Practice in Northwest/North Wales

AN diagnosis of Complex CHD – with potential for grim prognosis
• Offered options of palliative surgery, compassionate care or termination
• MDT – cardiologist, obstetrician, neonatologist, cardiac liaison nurse, palliative care team
• Palliative care team (Dr Lynda Brook’s team) introduced to parents if they are considering compassionate care postnatally
• PN confirmation of diagnosis
• Palliative care team involved postnatally only if parents decide for compassionate care
Summary

• AN diagnosis and discussion of palliative care allowed families to explore available options – including visiting children’s hospice before baby is born

• More informed choice

• AN involvement of palliative care team beneficial for postnatal planning
Thank you