Bi-Ventricular Repair for the Borderline Left Ventricle

Innovations and Anesthesia Challenges

Annette Schure, MD, DEAA, FAAP
Boston Children’s Hospital
Harvard Medical School
No conflicts of interest
No disclosures
Objectives

Bi-ventricular Repair for the Borderline Left Heart

- Indications and surgical challenges
- Staging and LV recruitment
- Outcome variables
- Considerations for the anesthetic management
First Joint Meeting of the Congenital Heart Surgeons’ Society and the European Congenital Heart Surgeons Association
Montreal, Canada, Oct. 2004

~ 100 experienced surgeons
Audience Response Survey 2004

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A11) A 26-year-old woman presents to you, 14 weeks pregnant, with a fetal diagnosis of HLHS. She is a single mother who has 4 other children, all under the age of 6. She has no education beyond high school and has limited resources. The baby’s father is no longer involved in her life. She is willing to do whatever you recommend. You would:

1. Advise her to deliver the baby and have a Norwood procedure
2. Advise her to deliver the baby and plan for a heart transplantation
3. Advise her to have a therapeutic abortion
4. Advise her to deliver the baby, hold it, but do nothing to treat it

<table>
<thead>
<tr>
<th>Option</th>
<th>Votes</th>
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<tbody>
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<td>4.</td>
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Life is more complex

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<td>coin</td>
<td>fossil</td>
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![Apples of various varieties](image-url)
Wide spectrum of LV hypoplasia

- Mild hypoplasia; near-normal LV
- Hypoplasia +/- hypertrophied LV
- Rudimentary LV

2V repair
Borderline LV
1V repair
Good size LV? Aortic/Mitral valve?

- **Yes**: Biventricular Repair (Valvuloplasty, Ross, Yasui Arch/Coarctation repair, etc.)
- **No**: Single Ventricle Pathway
  - Stage I => Glenn => Fontan

Definition? “Almost apex forming, not too bad…”
“Age-old” Question:

“Is a high risk bi-ventricular repair better than a low risk uni-ventricular repair?”

Dr. Ed Bove in a discussion at the 87th Annual Meeting of the American Association for Thoracic Surgery
Washington, DC, May 5-9, 2007
IS A HIGH-RISK BIVENTRICULAR REPAIR ALWAYS PREFERABLE TO CONVERSION TO A SINGLE VENTRICLE REPAIR?

Ralph E. Delius, MD
Marc A. Rademecker, MD
Marc R. de Leval, MD
Martin J. Elliott, MD
Jaroslav Stark, MD

87th Annual Meeting of the American Association for Thoracic Surgery,
Washington, DC, May 5-9, 2007

2-V or not 2-V: That is the question... plus some musings on thinking out of the box

Ralph E. Delius, MD

Borderline left ventricle: Trying to see the forest for the trees

Emile Bacha, MD
20 years of discussion - and no answer in sight?

Lessons learned!
# 1: A Fontan is not a perfect solution

Despite:

• **Expanded indications:**
  – Tricuspid atresia => all “functional” SVs

• **New techniques:**
  – Atriopulmonary => lateral tunnel => extracardiac

• **Improved inter-stage survival:**
  – 2002: ~68%  
    2017: ~85%

• **Improved short and long term mortality after Fontan**
Long-term survival after the Fontan operation: Twenty years of experience at a single center

Tacy E. Downing, MD,a Kiona Y. Allen, MD,a Andrew C. Glatz, MD, MSCE,a Lindsay S. Rogers, MD,c Chitra Ravishankar, MD,a Jack Rychik, MD,a Jennifer A. Faerber, PhD,b Stephanie Fuller, MD,c Lisa M. Montenegro, MD,d James M. Steven, MD,d Thomas L. Spray, MD,e Susan C. Nicolson, MD,d J. William Gaynor, MD,f and David J. Goldberg, MDa

TABLE E1. Comparison of population characteristics and survival outcomes in recent Fontan follow-up studies

<table>
<thead>
<tr>
<th>Study</th>
<th>Cohort years</th>
<th>n</th>
<th>Median follow-up, y</th>
<th>Follow-up rate</th>
<th>Fontan type</th>
<th>Systemic RV</th>
<th>HLHS</th>
<th>Survival</th>
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<td>CHOP cohort</td>
<td>1992-2009</td>
<td>773</td>
<td>90%</td>
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<td>AP</td>
<td>53% LT</td>
<td>47% EC</td>
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<tr>
<td>Pandi et al.12</td>
<td>2015</td>
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<td>Nakano et al.10</td>
<td>2015</td>
<td>500</td>
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<td>AP</td>
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<tr>
<td>d’Udekem et al.11</td>
<td>2014</td>
<td>1006</td>
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<td>AP</td>
<td>27% LT</td>
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<td>Khairy et al.14</td>
<td>2008</td>
<td>261</td>
<td>12</td>
<td>&lt;50%</td>
<td>62%</td>
<td>AP</td>
<td>37% LT</td>
<td>1% EC</td>
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<td>Hirsch et al.15</td>
<td>2008</td>
<td>636</td>
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<td>NR</td>
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<td>AP</td>
<td>92% LT</td>
<td>8% EC</td>
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</table>

RV: Right ventricle; HLHS, hypoplastic left heart syndrome; CHOP, Children’s Hospital of Philadelphia; AP, anterosuperior; LT, lateral tunnel; EC, extracardiac conduit; NR, not reported.
Survival Prospects and Circumstances of Death in Contemporary Adult Congenital Heart Disease Patients Under Follow-Up at a Large Tertiary Centre

Gerhard-Paul Diller, MD, MSc, PhD, MBA; Aleksander Kempny, MD; Rafael Alonso-Gonzalez, MD, MSc; Lorna Swan, MD, FRCP; Anselm Uebing, MD, PhD; Wei Li, MD, PhD; Sonya Babu-Narayan MB, BS, BSc, MRCP, PhD; Stephen J. Wort, PhD; Konstantinos Dimopoulos, MD, MSc, PhD; Michael A. Gatzoulis, MD, PhD

<table>
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<tr>
<th>Patient's age (years)</th>
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<th>30</th>
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<td>Fontan</td>
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<td>78</td>
<td>82</td>
<td>86</td>
<td>91</td>
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</tr>
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</table>
Forget about Survival! What about Quality of Life?
Quality of Life?

- **Arrhythmias:**
  - SVTs 50%, VT 3%, sudden death

- **Decreased Exercise Tolerance**
  - NYHA class II 10 y post op => NYHA III 20 y (~ 50%)

- **“Man made” form of Chronic Heart Failure**
  - Impaired systolic and diastolic function, increased PVR, hypoxemia

Yves d’Udekem et al. Circulation. 2014;130:S32-S38
Multiple Long Term Problems

• **Thromboembolism**
  – Risk for cerebrovascular accidents

• **Hepatic and Renal Dysfunction**

• **Protein-losing Enteropathy**
  – Prevalence: 3.7-24%, 13% @10y

• **Plastic bronchitis**
  – Rare, but potentially fatal
Bridge to Transplant?

High waiting list mortality
Significant wait time: 54d (3-452)

Only 50% 5y survival

High Risk Candidates for Fontan

• Atrioventricular valve regurgitation
• Pulmonary artery or vein stenosis
• Ventricular dysfunction
• Genetic syndromes: Trisomy 21
• Heterotaxy

=> Poor outcomes with Fontan circulation: Better off with BiVi
# 2: A Biventricular is not always better

**IS A HIGH-RISK BIVENTRICULAR REPAIR ALWAYS PREFERABLE TO CONVERSION TO A SINGLE VENTRICLE REPAIR?**


- 50 patients: complex intracardiac anatomy, good size ventricles
- 34 for BiVi repair, 16 for SVP, f/u over 8-10y
- => higher mortality & morbidity in BiVi group
  
  7y survival: BiVi 68% vs SVP 93%

It’s all about choosing the right candidates
Rhodes Score:

Threshold score:
- Indexed aortic root diameter ≤ 3.5cm/m²
- Indexed MV area: ≤ 4.75cm²/m²
- LAR ≤ 0.8 (ratio long axis dimension LV to long axis dimension heart)
- Indexed LV mass ≤ 35g/m²

Score ≥ 2: poor BiVi outcome

Echo: 5% measurement variability => change in predicted outcome in 10-15%

Score = 14.0 (BSA) + 0.943 (ROOT₁) + 4.78 (LAR) + 0.157 (MVA₁) − 12.03

≤ - 0.35 : poor outcome for BiVi
Hypoplastic Left Heart Complex

1. MV and AV hypoplasia (annulus z-score ≤ -2)
   Without evident intrinsic stenosis
2. LV hypoplasia (LV volume < 20ml/m² Simpson)
3. Ascending aorta, proximal and distal arch hypoplasia (z ≤ -2)
   With or without coarctation
4. Antegrade flow through LH and ascending aorta
5. Ductus-dependent systemic blood flow

Tchervenkov CI, Tahta SA, Jutras LC, Beland MJ.
• 326 neonates with critical LVOTO, 26 institutions, 1994-2001
• Follow up over 5 years
• Initial procedure indicated intended repair:
  - Univentricular Repair: 223 (Norwood)
  - BiVi Repair: 139 (BAV, AVplasty, Ross Konno, Yasui)
=> No statistical difference in actual survival
Predictors for death for UVR:
- Degree of TR
- Z-score of MV annulus
- Presence of large VSD
- Length of apex forming ventricle

Predictors for death for BVR:
- Minimum diameter of LVOT
- Presence of LV dysfunction
- Grade of endocardial fibroelastosis
- Diameter of the mid-aortic arch

Borderline patients: => often BVR
“two ventricles are better than one”
Wrong choice: worse survival effects for BVR
Predictors for death for UVR:
- Degree of TR
- Z-score of MV annulus
- Presence of large VSD
- Length of apex forming ventricle

Survival benefit = 30.55 (inverse of age at study entry [d] + 1)
  - 6.20 (aortic root z-score)
  + 12.14 (echocardiographic grade of EFE)
  + 23.33 (logarithm of ascending aortic [mm])
  - 28.30 (presence of moderate or severe tricuspid regurgitation)
  - 0.70 (LV long-axis length z-score) − 86.47

Borderline patients: => often BVR
“two ventricles are better than one”
Wrong choice: worse survival effects for BVR

Predicted 5 year survival difference between UVR and BVR:
• + value favors UVR
• - value favors BVR
• Number = magnitude of difference
Biventricular strategies for neonatal critical aortic stenosis: High mortality associated with early reintervention

Edward J. Hickey, MD, Christopher A. Caldarone, MD, Eugene H. Blackstone, MD, William G. Williams, MD, Tom Yeh, Jr, MD, Christian Pizarro, MD, Gary Lofland, MD, Christo I. Tchervenkov, MD, Frank Pigula, MD, Brian W. McCrindle, MD, and the Congenital Heart Surgeons’ Society

The Journal of Thoracic and Cardiovascular Surgery • August 2012

Feasibility and Safety of Biventricular Repair in Neonates with Hypoplastic Left Heart Complex


• Good early and late survival: 80-90%
• High rate of reinterventions: 30-50%, often multiple
  => Poor outcome if reintervention within 30 days: 60% mortality
  => Close follow up necessary: AV, MV, subaortic stenosis?
Follow up study on 139 patients with BVR
- 64 for reintervention – 27 for 2\textsuperscript{nd} reintervention – 8 for 3\textsuperscript{rd}.

15 neonates required reintervention within 30 days
=> highest 5y mortality ~ 60%

\textbf{Risk factors:} extensive EFE, LV dysfunction, AV cusp thickness, monocusp valves, subvalvar obstruction

\textbf{Importance of initial selection}
• Hypoplastic Left Heart Complex:
  – Subset of HLHS with milder form of LV hypoplasia and no valvar stenosis

• Retrospective analysis, 30 neonates with HLHC and primary biventricular repair at 1 hospital between 12/01 and 7/13 (12.5 y)

• During same period: 58 patients for UV repair
Primary Outcomes and Follow up

Primary:
- 97% survival, 1 death
- 3 x ECMO, 2 x Ischemic stroke
- 17 pts = 56% Reops/interventions

Follow up @ 5y:
- ≈ 30% reoperation
- ≈ 40% cath procedures, most within first 6 mo
  only 43% without intervention
# 3: It’s all about Genetics – and little do we know

- **Primary Mutations** affecting endocardial cushions, arch and vessels
- **Abnormal flow dynamics & shear stresses**
- **Further mutations** affecting LV growth
- **Myogenic switch**: cells mitosis stops, only hypertrophy, timing?
- **Remodeling**

Good size LV? Aortic/Mitral valve?

Yes
Rhodes Score
HLHS Score
CHSS Score
Biventricular Repair
(Valvuloplasty, Ross, Yasui Arch/Coarctation repair, etc.)

No

Single Ventricle Pathway
Stage I => Glenn => Fontan
What if, we could....

Promote LV growth and remodeling
New Strategy: LV rehabilitation and Staging

=> Series of Surgical Techniques

Goal: Promotion of LV Growth

- Relief of Inflow and outflow obstructions
- Improved flow through the ventricles
- Resection of Endocardial Fibroelastosis (EFE)
LV rehabilitation and Staging

Typical Candidates:

- Real borderline left heart: z-scores -5 to -0.5
  - Aortic and mitral valve stenosis, coarctation, small LV volume, EFE
  - Intact ventricular septum

- Unbalanced right-dominant AV canal: AV valve index

Contraindications:

Mitral or aortic atresia with intact ventricular septum
Surgical techniques and procedures

- Fetal transcatheter aortic balloon valvuloplasty
- EFE resection
- Mitral and Aortic valvuloplasty
- AV valve reconstruction for unbalanced AV canals
- Restriction of atrial septal defect
- Transcatheter Interventions: AV, MV, ASD: dilations, stenting
- Addition of accessory pulmonary blood flow
LV rehabilitation and Staging

**Primary LV recruitment**  \((z\text{-scores} > -3, \ AV\ valve\ index > 0.19)\)

- 2-V physiology at birth but increasing LA HTN, pHTN
  => Aortic and mitral valvuloplasty, coarctation repair, EFE resection

**Staged LV recruitment**  \((z\text{-scores} < -3, \ AV\ valve\ index < 0.19)\)

- Traditional SVP or high risk candidates for BiVi repair
  => Initial single ventricle approach:  Stage I => Glenn
  => Simultaneous LV rehabilitation
  => Conversion to BiVi circulation at later stage
Diagnostic Imaging

Cardiac MRI:

- Quantification of LV blood flow
  - ASD and left-to-right atrial shunting
- Detection of EFE

3D Echo:

- Aortic and mitral valve pathology

Fetal Aortic Valvuloplasty

- 20-24 wks, multidisciplinary team
- Fetal Candidates: Aortic stenosis
  - Progression to HLHS?
    - LV systolic dysfunction
    - Retrograde flow in transverse arch
    - Monophasic mitral inflow
    - Left-to-right flow across foramen
  - LV salvageable?
    - LV long axis z-score > 0
    - LV short axis z-score > 0
    - Aortic annulus z-score > -3.5
    - Mitral valve annulus z-score > -2
    - Aortic stenosis: max gradient > 20mmHg
- Mother suitable candidate?

• 127 fetal interventions over 15 years: early and late era
• Improved success rate: 83%
• Recently BiVi outcome more likely: 59% vs 26%
  - High probability for BiVi: LV pressure > 47 mmHg + Asc. Ao z-score > 0.57
  - Low probability: low LV pressure, mitral z-score < 0.1, MV inflow time z-score < -2
• BiVi patients: ~ 46% re-interventions (AV, MV etc)

Neurodevelopmental Outcome in Children after Fetal Cardiac Intervention for Aortic Stenosis with Evolving Hypoplastic Left Heart Syndrome

• 69 patients after FAV: 33 BiVi, 36 SVP, median age 5y (1.3-13y)
• Assessment of neurodevelopment (questionnaire, testing etc)
• Results: Scores lower than general population, BiVi 97 / SVP 89

Fetal intervention and/or Biventricular circulation did not improve outcomes

Predictor for poor outcome:
=> Total duration of hospital stay in first year of life

Laraja K et al: J Pediatr 2017;184:130-6 e4
Proof of concept

Primary left ventricular rehabilitation is effective in maintaining two-ventricle physiology in the borderline left heart

- 9 patients
- Borderline LH + EFE
- CHSS score 6.1 => favoring UVR
- Initial procedures:
  - Prenatal and postnatal AV BD, Coarctation
- Primary LV rehabilitation
  - @ 5.6 mo (19d-3y), CHF and LA HTN
- Median F/u 25mo (6mo - 10y)
  - 1 death (car accident)
  - 2 pt for reinterventions

<table>
<thead>
<tr>
<th>Preoperative interventions</th>
<th>Age at LV rehabilitation (mo)</th>
<th>Details of LV rehabilitation</th>
<th>Reintervention</th>
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<tbody>
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<td>1  Fetal balloon dilation</td>
<td>4</td>
<td>Mitral valvuloplasty, EFE resection</td>
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<tr>
<td>2  Postnatal balloon dilation of aortic valve, coarctation repair through thoracotomy</td>
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<tr>
<td>3  Left thoracotomy for coarctation repair, postnatal balloon dilation of aortic and mitral valves</td>
<td>7</td>
<td>Mitral valvuloplasty, EFE resection</td>
<td>2 reoperations for mitral replacement</td>
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<td>4  Postnatal balloon dilation of aortic valve</td>
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<tr>
<td>5  Left thoracotomy for coarctation repair</td>
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<td>Mitral valvuloplasty, EFE resection, resection of subaortic obstruction</td>
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<td>6  Fetal and postnatal balloon dilations of aortic valve</td>
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<td>7  Fetal and postnatal balloon dilations of aortic valve</td>
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<td>8  Fetal and postnatal balloon dilations of aortic valve</td>
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</table>

*LV, Left ventricle; EFE, endocardial fibroelastosis; ASD, atrial septal defect.*
Staged Left Ventricular Recruitment After Single-Ventricle Palliation in Patients With Borderline Left Heart Hypoplasia

- Retrospective analysis, Staged LV rehab vs SVP, 1995 - 2010
- 34 patients in each group
- Atypical case control: SVP 1995-2010, LV recruitment after 2001
- Primary Outcomes:
  - Change in size of LH structure over time (BSA, MRI & Echo)
  - Clinical Outcomes (mortality, biventricular conversion)

LV rehab techniques mainly during planned staged surgeries

11 additional procedures in 9 patients
Median f/u of 5.2y (1-9y)

Median f/u of 9 y (1-16y)
LV recruitment

- A: LVTIV Z-Score
- B: LV Long Z-Score
- C: MVP Leaf Z-Score
- D: AIV Z-Score

Graphs show comparisons prior to Stage I, prior to BDG, and prior to Fontan/Biventricular Repair.

Statistical significance:
- Prior to Stage I: P=0.001
- Prior to BDG: P=0.002
- Prior to Fontan/Biventricular Repair: P=0.001

Ejection Fraction:
- Prior to Stage I: P=0.03
- Prior to BDG: P=0.001
- Prior to Fontan/Biventricular Repair:
Important Concept: 
ASD restriction to promote LV growth
But it comes at a prize...

- More surgical procedures:
  - 3 for SV repair
  - 4 for LV rehab before conversion, 11 add. procedures (9 pts)
- Longer hospitalization (cumulative days in hospital):
  - Single Ventricle Group: 54.5 days (8-348)
  - LV recruitment Group: 94 days (27-518)
- LA hypertension:
  - Need for BD +/- stenting of ASD
LA Hypertension

Restrictive ASD to promote LV growth (n=19)

=> respiratory insufficiency, need for re-intubation (n=3)
=> persistent inotropic support (n=2)
=> unplanned Cath and intervention: mean Lap 19 (n=6)
=> asymptomatic LA hypertension on cath (n=7)

• Mean ventilatory time: 9.4 days / 17.5 ± 22 days in CICU
• Repeat restriction at subsequent surgery (n=4)
After Biventricular Conversion

• 12 patients, median follow up: 2.9 years (1-6y)
• No mortality
• Reoperation in 4 patients:
  – MV repair x 1, MV replacement x 1, AV repair x 1
  – VAD => cardiac transplant

• Limited study:

=> Long term outcomes and complications?
=> Increased LV dimension: dilation or growth?
Complex BiVi Repair Program

Institutional Approach:

– Staged LV recruitment for all borderline LV
– LH rehabilitation mainly during stage II
– ASD restricted to 4mm at the time of BDG
– Asymptomatic LA HTN will be tolerated
– ASD enlargement only for symptomatic patients

• Multidisciplinary group and regular conferences
Hemodynamic parameters predict adverse outcomes following biventricular conversion with single-ventricle palliation takedown


- Median age at conversion 2.7y
- Median weight 11.9kg
- Median f/u 2.7y (0.2mo-12y)
- 22% adverse outcome
dead (7), transplant (3) takedown (1)
- “Survival” 80% at 3y and 73% at 5y
A few numbers

- 25% add. procedure after Glenn
- Time to BiVi conversion 1.3y (IQR 0.6-2.3y)
- Hospital length of stay 25 days (IQR 12-38d)
- Surgical reintervention: 49%
Predictors for poor outcome

- Borderline HLHS
  - bHLHS 30% vs uAVC 6%
- LVEDP > 13mmHg
- RVP > \( \frac{3}{4} \) systemic

Diastolic dysfunction despite LV growth
Size doesn't always matter ....

• New focus: => **Diastolic Function** => Aggressive treatment in early stages => Decision making and family counseling

  Poor compliance:
  ─ definitive SVP if no contraindications
  ─ Ongoing observation – delayed conversion
  ─ Re-evaluation for treatable causes

• New techniques: Mitochondrial Transfer?
Anesthetic Considerations

• LA and pulmonary hypertension
• Diastolic LV dysfunction and RV dysfunction
• Chronic pulmonary edema
• Arrhythmias
• Multiple previous surgeries and interventions
  – Poor vascular access, tolerance to sedatives
• Prolonged hospitalizations
  – Developmental delay, PTSD, behavioral problems
And almost everywhere.....
Here is just one example........

• 7 yo, born with hypoplastic LV, MS, AS
  (z-scores: LV ED – 1.77, AV – 2.99 MV -2.02)
  s/p Ross-Konno, MVR x 3, PVR, EFE resections
• 10 cath (incl. fetal intervention) & 5 cardiac surgeries
• 16 anesthetics
• Total days in hospital: 211 (143 within first year of life)
• Last cath: LAP 18, PVR 6, RVP ½ syst., CI 2.8

=> Presenting for PICC placement in IR (milrinone tx –waiting for OHT)
Preoperative Evaluation

• Type/Morphology:
  – HLHS (primary or staged): => diastolic LV function? Unbalanced AV canal: => AV valve status?

• Diagnostic studies:
  – Echo: intracardiac shunts? Size of ASD? Gradients?
  – MRI: size of LH structures, MR, EFE?
  – Cath: HD data, interventions, vasodilator testing

• History
  – Previous surgeries and interventions, time in ICU, on ventilator

• Clinical status:
  – O₂ requirements, tachypnea, weight gain, baseline neuro status, vascular access

• Meds and Labs:
  – Diuretics, anticoagulation, antiarrhythmics, pulmonary vasodilators, ACE inhibitors
  – Anemia, hypokalemia, liver and renal dysfunction, coagulation status
Anesthesia Plan

- **Sedation:** PTSD – poor pulmonary reserve – high tolerance
- **Transport:** fluid shifts, O\textsubscript{2} requirements, monitoring
- **Induction:** “bubble precautions”, early ventilatory support, hypotension and arrhythmias, careful fluid management, early inotropic support
- **Difficult vascular access:** early support: equipment and staff
- **Maintenance:** inotropic support – adequate perfusion pressure, “fluids/diuretics sandwich”, ventilator adjustments
- **Post Op:** adequate observation, poor reserves, fever?
<table>
<thead>
<tr>
<th>Question</th>
<th>Total N</th>
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<th>America N</th>
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</tr>
</thead>
<tbody>
<tr>
<td>A11) A 26-year-old woman presents to you, 14 weeks pregnant, with a</td>
<td>72</td>
<td>39%</td>
<td>51</td>
<td>49%</td>
<td>21</td>
<td>14%</td>
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<tr>
<td>fetal diagnosis of HLHS. She is a single mother who has 4 other</td>
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<td>children, all under the age of 6. She has no education beyond high</td>
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<td>school and has limited resources. The baby’s father is no longer</td>
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<td>involved in her life. She is willing to do whatever you recommend.</td>
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<tr>
<td>You would:</td>
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</tr>
<tr>
<td>1. Advise her to deliver the baby and have a Norwood procedure</td>
<td>28</td>
<td>39%</td>
<td>25</td>
<td>49%</td>
<td>3</td>
<td>14%</td>
</tr>
<tr>
<td>2. Advise her to deliver the baby and plan for a heart transplantation</td>
<td>1</td>
<td>1%</td>
<td>1</td>
<td>2%</td>
<td>0</td>
<td>0%</td>
</tr>
<tr>
<td>3. Advise her to have a therapeutic abortion</td>
<td>41</td>
<td>57%</td>
<td>24</td>
<td>47%</td>
<td>17</td>
<td>81%</td>
</tr>
<tr>
<td>4. Advise her to deliver the baby, hold it, but do nothing to treat it</td>
<td>2</td>
<td>3%</td>
<td>1</td>
<td>2%</td>
<td>1</td>
<td>5%</td>
</tr>
</tbody>
</table>
A9) A 40-year-old woman presents to you, 14 weeks pregnant, with a fetal diagnosis of HLHS. She and her husband have been trying for 8 years to have a baby and have spent thousands of dollars on medical care to get to this point. They want this baby very badly “if the baby could have a good life.” They want your recommendation based on your experience and knowledge. You would:

Stage I: 73%
Termination: 21%
Compassionate care: 6%

A10) Your daughter (or son’s wife) is 14 weeks pregnant and the fetal diagnosis is HLHS. This would be their first child. They really want your advice, based on your experience and knowledge. You would:

Stage I: 26%
Termination: 67%
Compassionate Care: 5%
This was back in 2004
What about today?

5. LV rehab and biventricular repair
Life is complex

“Pushing the envelope”
- Giving Hope
- Facing Challenges
- Developing new therapies
- Experimenting with strategies
- Keeping busy

“Burden”
- Family
- Patients
- Medical system
Thank you!

Annette.Schure@childrens.harvard.edu
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LV Rehabilitation and Staging:


