Functional Outcomes after Neonatal Cardiac Surgery

21st Annual SEECMO Conference
Complex Pediatric Therapies Follow-Up Program
Gwen Alton RN, MN
June 5, 2011
Objectives:

• Describe neurodevelopmental outcomes
• Describe the cohort of children
• Introduce the Adaptive Behavioral Assessment System, 2nd ed (ABAS-II)
• Review the functional behavior of children assessed at 2 and 4 years of age
• Describe Heart Transplantation Outcomes
Neurodevelopmental Outcomes

- After Arterial Switch Operation
  - Survival, 99%
  - Disability – rare
  - Pre-operative insult may lead to stroke
  - Concerns
  - Some older children have language/communication disorder

Neurodevelopmental Outcomes

- After TAPVC Repair
  - Survival, 97%
  - Disability – rare
  - Concerns
    - Pulmonary illness
    - > expected Aboriginal children (40%)

Alton et al JTCVS (2007) 133 (4), 905-911
Neurodevelopmental Outcomes

• After palliative surgery for HLHS
  a) Norwood MBTS
    • Survival, 48%
    • Disability – rare
    • Concern – low motor skills
  b) Norwood RVPA
    • Survival, 81% and motor skills much improved
    • Concern – scattered academic abilities → poor academic achievement

Atallah et al, Circulation 2008, 118: 1410-18
Neurodevelopmental Outcomes

- After Heart Transplant at < 6 years
  - Survival, 88%
  - Disability, Cerebral Palsy 13%
  - Concerns
    - the children with congenital heart disease do not do as well as those with cardiomyopathy/myocarditis
    - Lower age at transplant do not do as well

METHODS

• Prospective, cross-sectional cohort study
• Children ≤ 6 weeks of age with complex cardiac surgery
• Assessed for neurodevelopmental outcomes at 18-24 months and 4-5 years
• Parents completed the Adaptive Behavioral Assessment System II at 2 and 4 years
DEFINITION: Adaptive/Functional

- Level to which individuals meet standards of personal self-determination and social conscientiousness that is expected for their age, development and culture.
- Includes real life skills such as grooming, dressing, safety, cleaning, making friends, social skills
- Used in evaluation of children to determine strengths and weaknesses to help improve their success in school and life
Literature Review – Functional Outcomes

• Pediatrics, 2001;108(6), Limperopoulos C., et al.
  – N=131 congenital heart defects, no HLHS
  – 40% had difficulties in ADL (VABS), 7% severe disability in self-care on WeeFIM at 18-24 months of age. 21% functioning within expected age range.

• Pediatric, 2008; 153(1), Majnemer A., et al.
  – 22% below cut off (<75) for self-care (WeeFIM), 11% in ADL, 15% adaptive behavior (VABS) below cutoff in same group at school entry.
  – Developmental and functional assessments correlated
Background

  
  Compared functional outcomes for children with HLHS and TGA at 2 years
  
  4X number of children delayed on GAC on the ABAS II then “normal” population
  
  self-care was lower in children with HLHS than with TGA
Cohort Descriptives

- January 2000 – April 2007, n= 306
- Deaths = 52 (17%) ECMO 18 (6%) Chrom Abn 9 (3%)
- Chrom Abn = 18 alive at 4 years – excluded
- LOST = 4
- 2 year assessments = 52; 37 with ABAS results
- 4 year assessments = 37; 6 with ABAS results (no 2yr)
- ABAS at 2 and 4 years = 143
DEFINITION: SV and non SV groups

• Single Ventricle(SV), n=44: HLHS, HRV, other SV physiology (unbalanced AVSD, TA)

• Non SV, n=99: TGA, Truncus, TAPVC, IAA, PA/VSD, TOF
# Acute care descriptives, n= 143

<table>
<thead>
<tr>
<th>Variable</th>
<th>Total n=143</th>
<th>SV n=44</th>
<th>Non SV n=99</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender – Male</td>
<td>94(66%)</td>
<td>27(61%)</td>
<td>67(68%)</td>
<td>.463*</td>
</tr>
<tr>
<td>Antenatal Diagnosis</td>
<td>43(30%)</td>
<td>28(64%)</td>
<td>15(15%)</td>
<td>.000*</td>
</tr>
<tr>
<td>Birth Gestation</td>
<td>38.9(1.8)</td>
<td>39(1.5)</td>
<td>38.8(1.9)</td>
<td>.339</td>
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<tr>
<td>Pre-op inotrope score</td>
<td>6.4(10.5)</td>
<td>5.5(10.2)</td>
<td>6.7(10.7)</td>
<td>.527</td>
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<tr>
<td>Pre-op serum lactate</td>
<td>4.1(4.2)</td>
<td>3.5(3.1)</td>
<td>4.4(4.6)</td>
<td>.181</td>
</tr>
<tr>
<td>Age at Treatment (days)</td>
<td>12.7(9.9)</td>
<td>10.8(8.2)</td>
<td>13.5(10.4)</td>
<td>.123</td>
</tr>
</tbody>
</table>

* Fisher’s Exact – 2 sided
### Acute care descriptives, n= 143 mean(SD)

<table>
<thead>
<tr>
<th>Variable</th>
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</thead>
<tbody>
<tr>
<td>CPB, minutes</td>
<td>119 (54)</td>
<td>117(39)</td>
<td>120(60)</td>
<td>.794</td>
</tr>
<tr>
<td>DHCA (n=96), minutes</td>
<td>21(16)</td>
<td>23(13)</td>
<td>19(17)</td>
<td>.197</td>
</tr>
<tr>
<td>PO day 1 highest Inotrope Score</td>
<td>15(12)</td>
<td>17(13)</td>
<td>14(11)</td>
<td>.111</td>
</tr>
<tr>
<td>PO day 1 highest lactate</td>
<td>5.8(2.9)</td>
<td>6.9(3.4)</td>
<td>5.3(2.5)</td>
<td>.006</td>
</tr>
</tbody>
</table>
# Acute care descriptives, n= 143

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</thead>
<tbody>
<tr>
<td>PO day 2-5 highest inotrope score</td>
<td>12 (10)</td>
<td>15(10)</td>
<td>12(9)</td>
<td>.099</td>
</tr>
<tr>
<td>PO day 2-5 highest lactate</td>
<td>2.8(1.7)</td>
<td>2.9(1.5)</td>
<td>2.8(1.8)</td>
<td>.796</td>
</tr>
<tr>
<td>PO day 6+ highest inotrope score</td>
<td>4(7)</td>
<td>7(8)</td>
<td>3(7)</td>
<td>.007</td>
</tr>
<tr>
<td>PO day 6+ highest lactate</td>
<td>1.7(1.4)</td>
<td>2.1(1.9)</td>
<td>1.4(1.0)</td>
<td>.060</td>
</tr>
</tbody>
</table>
## Acute care descriptives, n= 143

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</tr>
</thead>
<tbody>
<tr>
<td>PO days of ventilation</td>
<td>12(15)</td>
<td>14(13)</td>
<td>11(16)</td>
<td>.195</td>
</tr>
<tr>
<td>PO ICU days</td>
<td>19(18)</td>
<td>25(19)</td>
<td>16(17)</td>
<td>.005</td>
</tr>
<tr>
<td>Dialysis, anytime in ICU</td>
<td>10(7%)</td>
<td>3(7%)</td>
<td>7(7%)</td>
<td>1.000*</td>
</tr>
<tr>
<td>CPR, anytime in ICU</td>
<td>12(8.4%)</td>
<td>2(4.5%)</td>
<td>10(10.1%)</td>
<td>.344*</td>
</tr>
<tr>
<td>ECMO, post-op</td>
<td>6(4.2%)</td>
<td>2(4.5%)</td>
<td>4(4%)</td>
<td>1.000*</td>
</tr>
</tbody>
</table>

* Fisher’s Exact – 2-sided
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<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at 18-24 month assessment</td>
<td>21.7(4.7)</td>
<td>22(5.2)</td>
<td>21.6(4.4)</td>
<td>.607</td>
</tr>
<tr>
<td>Age at 4-5 year assessment</td>
<td>53.7(5.4)</td>
<td>53.5(5.1)</td>
<td>53.9(5.6)</td>
<td>.701</td>
</tr>
<tr>
<td>Mother’s educ at 4yr</td>
<td>13.3(2.4)</td>
<td>13.1(2.2)</td>
<td>13.4(2.5)</td>
<td>.575</td>
</tr>
<tr>
<td>Ht&lt;5th% at 4 years</td>
<td>14(10%)</td>
<td>9(20%)</td>
<td>5(5.1%)</td>
<td>.011*</td>
</tr>
<tr>
<td>Wt &lt;5th% at 4 years</td>
<td>13(9%)</td>
<td>7(16%)</td>
<td>7(7.1%)</td>
<td>.101*</td>
</tr>
<tr>
<td>HC below-2SD at 4yr</td>
<td>8(5.6%)</td>
<td>4(9.1%)</td>
<td>4(4%)</td>
<td>.251*</td>
</tr>
<tr>
<td>Tube feedings at 4yr</td>
<td>8(5.6%)</td>
<td>6(14%)</td>
<td>3(3%)</td>
<td>.016*</td>
</tr>
</tbody>
</table>
## 4-year outcomes for survivors, n(%) Table

<table>
<thead>
<tr>
<th>Variable</th>
<th>Total N=143</th>
<th>SV n=44</th>
<th>Non SV n=99</th>
<th>Fisher’s Exact</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cognitive delay @ 4yr</td>
<td>5(3.5%)</td>
<td>1(2.2%)</td>
<td>4(4.1%)</td>
<td>1.000</td>
</tr>
<tr>
<td>Cerebral Palsy at 4yr</td>
<td>6(4.2%)</td>
<td>3(6.7%)</td>
<td>3(3.1%)</td>
<td>.372</td>
</tr>
<tr>
<td>Hearing Loss at 4 yr</td>
<td>9(6.3%)</td>
<td>8(18%)</td>
<td>1(1.0%)</td>
<td>.000</td>
</tr>
<tr>
<td></td>
<td></td>
<td>8 amplified</td>
<td>1 amplified</td>
<td></td>
</tr>
</tbody>
</table>

No vision loss
# ABAS II Profile of Function

<table>
<thead>
<tr>
<th>COMPOSITE DOMAINS 100(15)</th>
<th>10 SKILL AREAS 10(3)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Motor</td>
</tr>
<tr>
<td>CONCEPTUAL</td>
<td>Communication</td>
</tr>
<tr>
<td></td>
<td>Functional Pre/Academics</td>
</tr>
<tr>
<td></td>
<td>Self-Direction</td>
</tr>
<tr>
<td>SOCIAL</td>
<td>Leisure</td>
</tr>
<tr>
<td></td>
<td>Social</td>
</tr>
<tr>
<td>PRACTICAL</td>
<td>Community Use</td>
</tr>
<tr>
<td></td>
<td>Home Living</td>
</tr>
<tr>
<td></td>
<td>Health and Safety</td>
</tr>
<tr>
<td></td>
<td>Self-Care</td>
</tr>
</tbody>
</table>

General Adaptive Composite Score (GAC) includes all 10 skill areas
2- & 4-y Domain differences, SV, m(SD)

n = 44, mean 100(15)
2-\&4-y Skill area differences; SV,m(SD)

n=45, mean 10(3)

Com  CU  FA*  HL  H&S  LS  SC*  SD  Soc  MO

p= .004  .025  .042  .002

2 year  4 year
2-4-y Domain differences, nonSV,m(SD)

n= 99, mean 100(15)

- GAC: p= .035
- CON: .415
- Social: .091
- Practical: .000

2 Year
4 year
2-\&4-y Skill area differences; nonSV, m(SD)

n= 99, mean 10 (3)
General Adaptive Composite Delay

- SV – 10 (23%) - GAC<70

- Non SV – 11 (11%) – GAC <70
Pediatric Nursing, March-April 2008 34(2). Ling Fan

- Self-care behaviors of school-age (6-12y) children with heart defects in China (ASD, VSD, TOF) – **Orem’s Self-Care Theory**
- self-care activities decreased in children with more severe heart function and more health symptoms
- moderate levels of self-care possible explanations:
  - health impact on growth and development – critical period
  - health education – long-term, follow-up programs suggested
  - parent education – predictor of self-care practices
  - family support (parental fostered dependency)
“Children with executive dysfunction have fewer problems with what they know but more difficulty with what they can do (Denckla, 1996), especially under demands for independence.”

Bellinger – Theory of Mind

*Bellinger, D. (2008) Are children with congenital cardiac malformations at increased risk of deficits in social cognition? Cardiol Young (Boston Trial group – TGA at 8 years)
  - Theory of mind – (ability to read people and interpret actions) concerns
    - children appear to have difficulty identifying and describing their own internal states
    - studies now addressing burden of these deficits on social function and disability during adolescence
Calderon - Executive Function


reported on executive function and theory of mind in school-aged children after neonatal surgery for TGA general IQ within normal limits – more subtle cognitive deficits, cognitive and behavioral inhibition impaired – minimum of 2 years’ delay (longer reaction times and difficult to self-regulate behavior)
• MRI’s in the peri-operative period
• White matter injury was observed in 13 (32%) of children with CHD and in no controls
• Need to explore this further with more advanced MRI techniques and cerebral blood flow patterns in fetus with CHD
Predictors of 4 year outcomes - ABASII

- Mother’s education – Social and Conceptual Domains
- Pre-op highest plasma lactate
  - (Soreh Taghaddos, statistician Master student)
- Oakland & Harrison (2008) – found differences in children’s GAC scores from parents with < high school or a HS diploma compared to parents with some college
Educational & Rehab Services

- Cardio in the Young, 2008: 18. Majnemer A. et al
  - patterns of use of educational & rehab services at school age – 60 families (mean age 8 years)
  - Same cohort as 2001, no HLHS
- Concerns identified by parents: classroom work, handwriting, mathematics, behavior and communication difficulties.
  - 22% received educational supports (supplemental tutoring)
  - 17% of children not receiving support, parents felt their child needed services to optimize performance
# Rehabilitation services utilized

<table>
<thead>
<tr>
<th>Services</th>
<th>SV n=44</th>
<th>Non SV n=99</th>
</tr>
</thead>
<tbody>
<tr>
<td>Early Intervention</td>
<td>20(45%)</td>
<td>31(31%)</td>
</tr>
<tr>
<td>Daycare</td>
<td>9(20%)</td>
<td>22(22%)</td>
</tr>
<tr>
<td>Playschool</td>
<td></td>
<td></td>
</tr>
<tr>
<td>No aid</td>
<td></td>
<td></td>
</tr>
<tr>
<td>No services</td>
<td>15(35%)</td>
<td>46(47%)</td>
</tr>
</tbody>
</table>
Publication – Heart Transplantation

Heart Transplantation Outcomes

- n = 33 children under 6 years of age – 1999 – 2006
  - Congenital Heart Disease (CHD) = 18
  - Cardiomyopathy-myocarditis = 15
- ECMO – 6 (4 deaths-CHD)
- Mortality (18-24 month assessment) – 12%
Heart Transplantation Outcomes

• General Adaptive Score (ABASII)
  74.3(19.9)
  – 14(48%) delayed on the GAC

• Mental delay (scores <70) 10(34%)
Summary

• Functional Pre-academics, and self-care significantly decrease at 4 years of age
• Active participation in promoting a child’s independence has several benefits: improve motor skills, processing skills, cognitive skills and promote mastery of tasks that are both meaningful and important socially
• Understand the sequence of learning new skills in order to introduce new tasks or challenges at appropriate developmental times in the child’s life
Promote Early Intervention Programs (0-3yrs) and Early Educational Support (3-6yrs)
Facilitate parenting education – foster a healthy living environment for these children and their families
Thank you

• All the families and children who have participated – Complex Pediatric Therapies Follow-Up Program
• Other sites across Western Canada, Hospitals and Developmental Clinics
• NICU, PICU, surgical staff – Stollery Children’s Hospital
• 4C staff & therapists – Stollery Children’s Hospital
• Glenrose Rehabilitation Hospital
  – therapist and support staff
• Co-chairs – Dr. Charlene Robertson,
  Dr. Ari Joffe, Dr. Reg Sauvé
Parent Stress

- Majnemer et al (2006) using the Parenting Stress Index reported on the stress level of parents with children 5 years post cardiac surgery
  - 25% high stress levels
  - 17% defensive
  - 15% high stress and defensive

- Heart Transplant children: 30% of parents diagnosed with PTSD