Dealing with Success → Transition of Survivors of Complex Pediatric Diseases to Adulthood

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Xi’an China

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Objectives

- Define the numbers
- What are the emerging populations?
- Strategies: Survivors of NICU/Prematurity
- Survivors of Diaphragmatic Hernia
- Congenital Cardiac Disease
- Intestinal Failure/Abdominal Surgery
- Recommendations: Role of Primary Pediatrician
The Numbers: Survival Increasing

- World wide ↑ Survival of Prematurity
- 26 weeker: 50% survival 1997 → 75% 2011

From Ancel et al. Survival of < 32 w in France, JAMA Pediatr. 2015;169:230-238
Survival and Numbers of “Premature” Infants Increasing World-wide

Greatest Numbers In Asia

Figure 3: Estimated preterm birth rates by country for the year 2010
But survivors, if less than 27w, have high morbidity

Follow-up of NICU graduates

- General Pediatrician
- Neonatologist
- Developmental Peds/neurology
- Psychology
- OT/PT/Speech Therapy
- Pulmonologist
- GI/Surgery

From Bockli et al. Trends and challenges in United States neonatal intensive care units follow-up clinics J Perinatol. 2014; 34:71-74
Outcome after repair of Diaphragmatic Hernia

Large Hernia with Pulmonary Hypoplasia requiring ECMO
Neurological Outcome following repair of CDH: 25% Impaired

BSID-III outcomes of the entire cohort as compared to population norms [16].

<table>
<thead>
<tr>
<th></th>
<th>BSID-III score</th>
<th>P-value</th>
<th>Proportion &gt; 1 SD and &lt; 2 SD below mean, %</th>
<th>Proportion &gt; 2 SD below mean, %</th>
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<tbody>
<tr>
<td>Cognitive composite score</td>
<td>93.7 ± 14.4 (55–115)</td>
<td>0.001</td>
<td>14</td>
<td>8</td>
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<tr>
<td>Language composite score</td>
<td>85.9 ± 13.8 (56–124)</td>
<td>0.0001</td>
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<td>Motor composite score</td>
<td>89.6 ± 14.6 (46–124)</td>
<td>0.0001</td>
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<td>Receptive language score</td>
<td>7.4 ± 2.4 (1–13)</td>
<td>0.0001</td>
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<td>Expressive language score</td>
<td>8.0 ± 2.6 (2–13)</td>
<td>0.0001</td>
<td>19</td>
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<td>Fine motor score</td>
<td>8.9 ± 2.5 (1–15)</td>
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<tr>
<td>Gross motor score</td>
<td>7.8 ± 3.2 (1–13)</td>
<td>0.0001</td>
<td>12</td>
<td>14</td>
</tr>
</tbody>
</table>

Data presented as mean ± SD (range) or proportions as appropriate.

From Danzer et al. Neurodevelopmental outcome at one year of age in congenital diaphragmatic hernia infants not treated with ECMO J PedSurg. 2015;50:898-903
Problems in CDH Patients?

- Pulmonary: Short term: >50% steroids and bronchodilators in first 2 years only
- Neurodevelopmental: 30% affected
- GI: Reflux common! 50% of patients ~ 8% with Barrett’s esophagitis
- Musculoskeletal: Scoliosis, chest wall deformity ~ 10%
- F.U. Implications: Specialized Clinic + Primary Pediatrician
Follow-up of Congenital Heart Disease: Scope of the Problem

- Congenital Heart Disease: Currently, there are more than one million adult patients with congenital heart disease (un-repaired, repaired, and palliated) in the US
Interventional Therapies For
Congenital & Structural Heart Disease

CHD: Based on Physiology

A. Left-to-right shunting lesions.
B. Right-to-left shunting lesions.
C. Right heart obstructive lesions.
D. Left heart obstructive lesions.
E. Post Fontan operation.
F. Post Tetralogy repair patients.
G. Miscellaneous
Adult Survivors with Congenital Heart Disease > Pediatric Patients

Figure 4. Age range of patients with CHD in two tertiary care centers.
Interventional Therapies For Congenital & Structural Heart Disease

Atrial Septal Defect
Interventional Therapies For Congenital & Structural Heart

Devices

1. Amplatzer Septal Occluder
2. Gore Devices
3. CardioSeal/StarFlex
4. Cardia Intrasept
5. Occlutech-Figulla Flex II
6. Solysafe
7. Patch Sideris
8. Others
Interventional Therapies For Congenital & Structural Heart Disease

The Team

1. Interventional Adult Cardiologist-Expertise in CHD
2. Interventional Pediatric Cardiologist
3. Cardiac Surgeon-Expertise in CHD & Valve disease
4. Echocardiographer-Expertise in CHD
5. Anesthesia-Expertise in CHD/ShD
6. Nurses/Techs/Perfusionists/etc
Closure Protocol

Local anesthesia with ICE
R & L Heart Catheterization
RUPV Angiogram (LAO/Cr)
Balloon Sizing
Closure
ICE/Angiogram (RA/PA)
Interventional Therapies For Congenital & Structural Heart Disease
Congenital Heart Disease:

Interventional Therapies For Congenital & Structural Heart Disease

All patients born with Congenital Heart Defects 1.6 M Worldwide

RVOT (22%) 352,000

Pulmonary Stenosis
Tetralogy of Fallot
Truncus Arteriosus
Transposition of the Great Arteries
Double Outlet Right Ventricle (DORV)
Pulmonary Atresia

Non-conduit Patients

Conduit Pt (30%) 105,600
Follow-up of Congenital Heart Disease into Adulthood

1. Simple adult CHD cases can be followed by local cardiologists.

2. Moderate adult CHD can be followed by local cardiologists with consultation from the regional center

3. Complex adult CHD need to be followed indefinitely by the regional center (serving population of 10-15 M)

Outcome with Intestinal Failure (SBS) in the Neonate: Effect of a Multi-Disciplinary Care Team

Sigalet et al, Elements of Successful Intestinal Rehabilitation J Ped Surg 2011
IF: Improved Support $\rightarrow$ ↑ Survival and patients on long term PN

18% of patients on home PN at 2 years
10% at 3 years: Cost/Quality of Life?
Need Better Therapy to ↑ adaptation!
Solution: Multidisciplinary Longterm follow-up clinics

- Core Pediatrics and Neonatology follow-up
- Surgical/Pulmonary specialists as part of team
- Experienced, consistent caregivers Critical.
Long Term Follow-up: Partnership between Neonatologist and Pediatrician and Associated Specialists

Childhood →

Age: 0 1 2 6 12 24 40+ years

Neuronal development Socialization, Sports
Long term follow-up essential → Yearly Review
Neonatology → Core Pediatrician
Allied Health: Speech, Physio, OT
Pulmonary
Surgery and Cardiology

General Pediatrician → Adult Medicine

Adult
Conclusions

- Long term follow-up **essential**
- Regional centers for follow-up
- Graded transition from Neonatal to Pediatric to Adult Care clinic
- Coordination and multi-disciplinary team involvement optimal for best results
- As technology advances ↑ frequency of survivors with ↑ complex problems