Family Readiness for Emergencies

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July 1st, 2017
Disclosure

• Conflicts of Interest: None
• Commercial Sponsorship: None
• Disclosures:
  – Roche (Switz) and Genetech (US) Pharmaceuticals, advisory panel research design and outcomes for intervention trials in children with SMA
  – Consultant, Biogen Pharmaceuticals. SMA Advisory Panel (RJG)
  – Site investigator, Cytokinetecs trial in SMA (RJG)
  – Site investigator (upcoming), Avexis trial in SMA (RJG)
  – Site investigator (prior), Isis/Ionis trial in SMA (RJG)
  – Consultant, Audentes Pharmaceuticals, Research design and outcomes for intervention trial in children with XL-Myotubular Myopathy (RJG)
• Clinical Director, C.A.P.E. and Home Ventilation Program (RJG)
• Member of Cure Spinal Muscular Atrophy, Medical Advisory Council (RJG)
Recognition – Thanks in Advance

- Cure SMA, National and our respective chapters, South Florida and New England
- The C.A.P.E. and Home Ventilation Program
Objectives

- Our old world is a new world to most*
- Prepare for the emergencies that WILL happen
- Consider if children and families with SMA different than other populations
- Explore practical considerations when navigating the acute care hospital
- Share experiences and solutions for optimizing care in the emergency and hospital setting (e.g. “go-bags”, generators, hand-held suctions,…medical history binders, and drug information.)
Different Needs → Different Care Paradigm

• Differentiating children and families with neuromuscular disorders and special health care needs
  – Professional Attitudes
  – Reconfigured Parental Roles
  – Idiosyncratic Clinical Trajectory
  – Technologic Advancement / Successes


Professional Attitudes - Children with Disabilities


Professional Attitudes – Care/Support in Neuromuscular Disorders


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I may not be an M.D.

but I am an M.O.M.

(Medically Oriented Mother)
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Vulnerable Population

- Children with technology dependence – 373 Relative Risk of unplanned ICU admission (Dosa et al, Peds 2001)
  - 32% of unscheduled ICU admissions for CSHCN are preventable

<p>| TABLE 2. Risk of Critical Illness in Population Groups |
|-----------------------------------------|-----------------|-----------------|</p>
<table>
<thead>
<tr>
<th>Group</th>
<th>Number of Patients Admitted/Year</th>
<th>Population Size*</th>
<th>RR (95% CI) (Compared With Previously Healthy)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Previously healthy</td>
<td>136</td>
<td>387 040</td>
<td>1</td>
</tr>
<tr>
<td>Chronic conditions</td>
<td>97</td>
<td>84 960</td>
<td>3.3 (2.5–4.2)</td>
</tr>
<tr>
<td>Technology-assisted</td>
<td>29</td>
<td>222</td>
<td>373 (330–422)</td>
</tr>
<tr>
<td>Not technology-assisted</td>
<td>68</td>
<td>84 738</td>
<td>2.3 (1.7–3.0)</td>
</tr>
</tbody>
</table>

* Total pediatric population of the region is 472 000.5 Of these, 18% are assumed to have a chronic condition, and 82% were considered to have been previously healthy.1 The technology-assisted subgroup is assumed include 4.7 per 10 000 children.45

- National KID 1997-2006 increasing proportion of ICU patients, higher mortality, 2x hospital charges, more invasive devices (Odetola et al, 2010)
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EMS for Children, Nat’l Taskforce CSHCN 1997

- All hospitals should have protocols for children with special health care needs (CSHCN)
- Multidisciplinary care
- Contact the primary care immediately upon arrival
- Develop future emergency plans prior to discharge

Annals of Emergency Medicine, 1997; 30(3):274-280

... 15+ years in the future

Do it Yourself...DIY EMS?

• Family-Centered Care - a mixed blessing

• Your pediatrician’s office

• Passports / Emergency Information Form / POLST

• Local and Regional facilities

• Pediatric Transport Teams

• Contingency Plans when away from home

• Advanced Directives, Healthcare Proxies
We made it, a sigh of relief or take a deep breath?

• ED and ICU Care Plans
• Direct Admissions

• Reach-out (Primary Care, Specialists, your advocate...local or elsewhere)

• Triage Process
  – Transfer hospitals
  – Ward
  – Intermediate or Intensive Care Unit
  – Home
Not a red herring…but swimming upstream
The ICU is not a vacation

• 1. Know my child’s baseline
• 2. Integrate and bridge multiple services
• 3. Disconnect between role of parent at home vs. parent in the PICU
• 4. PICU care does not equate with respite
• 5. High stakes learning environment
• 6. Heterogeneity within group
• 7. Lack of fit within the acute care model

It may feel like Hotel California ♫

- Respiratory issues as Primary or Secondary
  - Bring your equipment and protocols

- Calibrating providers to SMA
  - Feeding
  - Technology supports
  - Airway

- Calibrating providers to you

- UP To DATE on SMA?

- Again, reach out
Peri-Anesthesia and Procedural Sedation

• No such thing as “Conscious Sedation”
• Consider different options for ventilator support
• Regional anesthetics
• When do you extubate?
• Oxygen is not the enemy but heed the warning
• Aggressive airway clearance and NIV
• Pain control
• Nutrition and bowel regimens
Culture of Cure – Other Practical Implications

• Parent Presence

• Resuscitation and the OR

• Assessment of distress

• Transitions in Care / Adult Hospitals

• Extending critical care services beyond the boundaries of the ICU
Revisiting our Objectives

• Anticipating and preparing for the undesired event

• Individuals with SMA and their families as part of the pediatric population

• Explored practical considerations when navigating the acute care hospital

• Let’s share experiences and solutions for optimizing care in the emergency and hospital setting
SMA Conference Survey

Please complete your conference survey at this link: https://www.surveymonkey.com/r/2017SMAAnnualConference
Or fill out the paper survey in your conference folder.

• All participants who complete a survey by 10:30 am on Sunday July 2nd, will have their name entered into a raffle for a brand new iPad!
• The winners will be drawn and announced on Sunday, July 2nd at the Closing General Session/It’s a Wonderful Life.