

Family Readiness for Emergencies

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Make today a breakthrough.

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, Cytokinetics trial in SMA (RJG)
 - Site investigator (upcoming), Avexis trial in SMA (RJG)
 - Site investigator (prior), Isis/Ionis trial in SMA (RJG)
 - Consultant, Audentes Pharmacueticals, 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 suction, ...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

Graham RJ and Robinson WM. Integrating palliative care into chronic care for children with severe neurodevelopmental disabilities. *J Dev Behav Pediatr* 2005;26(5):361-365.

Klick JC and Ballantine A. Providing care in chronic disease: The ever-changing balance of integrating palliative and restorative medicine. *Pediatr Clin N Am* 2007;54(5):799-812.

Professional Attitudes - Children with Disabilities

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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

TABLE 2. Risk of Critical Illness in Population Groups

Group	Number of Patients Admitted/Year	Population Size ^a	RR (95% CI) (Compared With Previously Healthy)
Previously healthy	136	387 040	1
Chronic conditions	97	84 960	3.3 (2.5–4.2)
Technology-assisted	29	222	373 (330–422)
Not technology-assisted	68	84 738	2.3 (1.7–3.0)

^a 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.^{4,5}

- 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

Carlos Lerner et. al. Medical Transport of Children with Complex Chronic Conditions. *Emerg Med Intl.* 2012 “*little published evidence*”



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

Graham RJ, Pemstein DM, Curley MA. Experiencing the pediatric intensive care unit: perspective from parents of children with severe antecedent disabilities. *Crit Care Med* 2009;37:2064-70.

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

- Dingeman RS, Mitchell EA, Meyer EC, Curley MA. Parent presence during complex invasive procedures and cardiopulmonary resuscitation: a systematic review of the literature. *Pediatrics* 2007;120:842-54.

- Resuscitation and the OR

- Truog RD, Waisel DB, Burns JP. DNR in the OR: a goal-directed approach. *Anesthesiology*. Jan 1999;90(1):289-295.

- Assessment of distress

- Regnard C et al. Understanding distress in people with severe communication difficulties: developing and assessing the Disability Distress Assessment Tool (DisDAT). *J Intellect Disabil Res* 2007;51(4):277-292.

- 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.**