Family Readiness for Emergencies

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Make today a breakthrough.

Disclosure

- Conflicts of Interest: None
- Commercial Sponsorship: None
- Disclosures:
  - Site investigator, Cytokinetics trial in SMA (RJG)
  - Site investigator, Avexis trial in SMA (RJG)
  - Site investigator, Roche trial in SMA (RJG)
  - Consultant, Audentes Pharmacueticals, Research design and outcomes for intervention trial in children with XL-Myotubular Myopathy (RJG)
  - Prior:
    - 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, Isis/Ionis trial in SMA (RJG)
- Clinical Director, C.A.P.E. and Home Ventilation Program (RJG)
- Member of Cure Spinal Muscular Atrophy, Board of Directors (probationary) (RJG)

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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

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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.)

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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.

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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*	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)

* 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

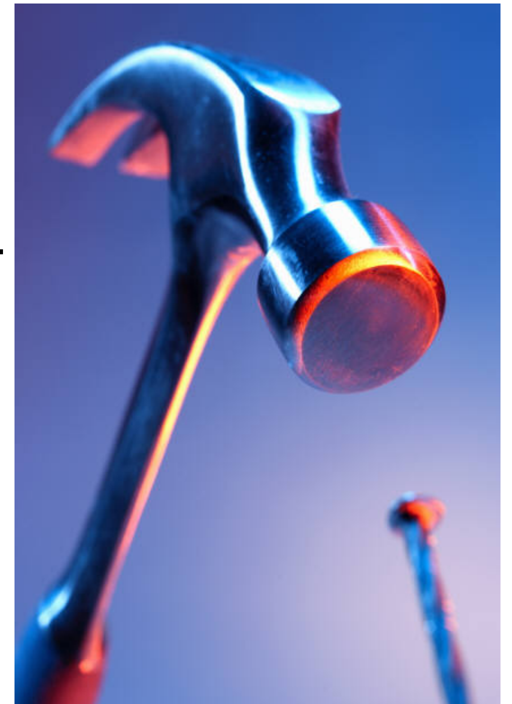
Annals of Emergency Medicine, 1997; 30(3):274-280

… 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



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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

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Not a red herring…but swimming upstream

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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

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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

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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

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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

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