



Perinatal and Pediatric Palliative Care: Supporting Children and Families Facing Serious Illness

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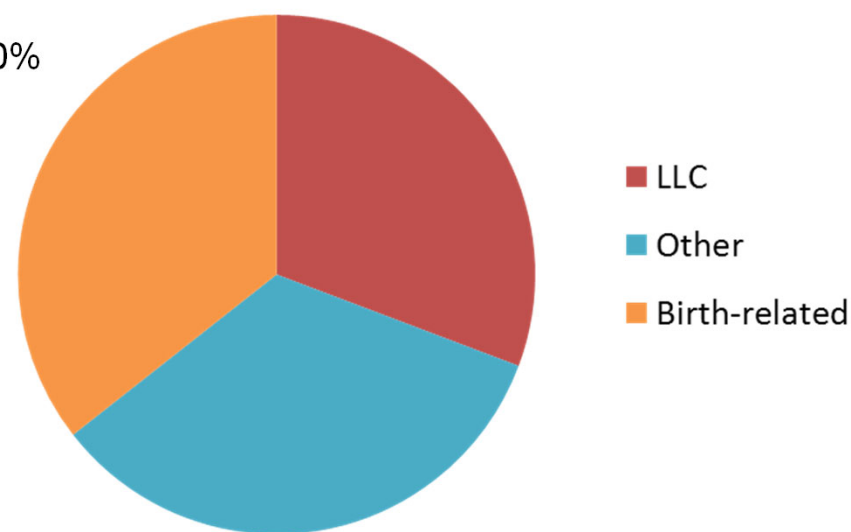
Objective

- Describe how perinatal and pediatric palliative care teams help families of children with serious illness by alleviating suffering, supporting quality of life, improving communication, and supporting decision-making.



Causes of death in children 0-19 years of age

- Over 50,000 childhood deaths annually in USA
- Over 500,000 children living with serious illness
- **2002 data - 51,956 total deaths**
 - Life-limiting conditions – 15,963 – low likelihood for a cure
 - Cancer – about 25%
 - Neurodegenerative/neuromuscular – about 20%
 - Congenital or genetic – about 15%
 - Cardiovascular – about 25%
 - Metabolic or other – 15%
 - Other – 17,506
 - Unintentional injury – 10,952
 - Homicide – 2,604
 - SIDS – 2,050
 - Suicide – 1,900
 - Birth-related conditions – 18,487
 - Complication of delivery or prematurity
- Compare to 1900s – top causes of death were infectious and 2/10 children died before age 5



Source: Friedrischsdorf, personal communication

Changing Face of Death

Before 1900s

- Intergenerational family units
- Limited effective medical interventions
- Common to experience births and deaths in the home

20th Century

- Hospitals and medical technology advance
- Resuscitation (CPR) developed in 1960
- People live longer but with increased morbidity

2000s

- Emphasis on youth and health
- Death seen as a medical failure
- Death occurring in medical facilities > home

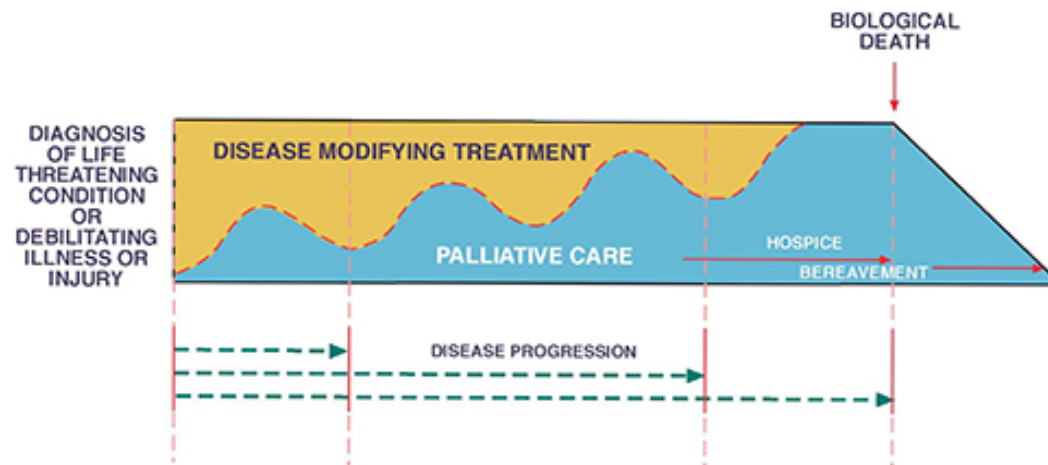
Children with “serious illness”

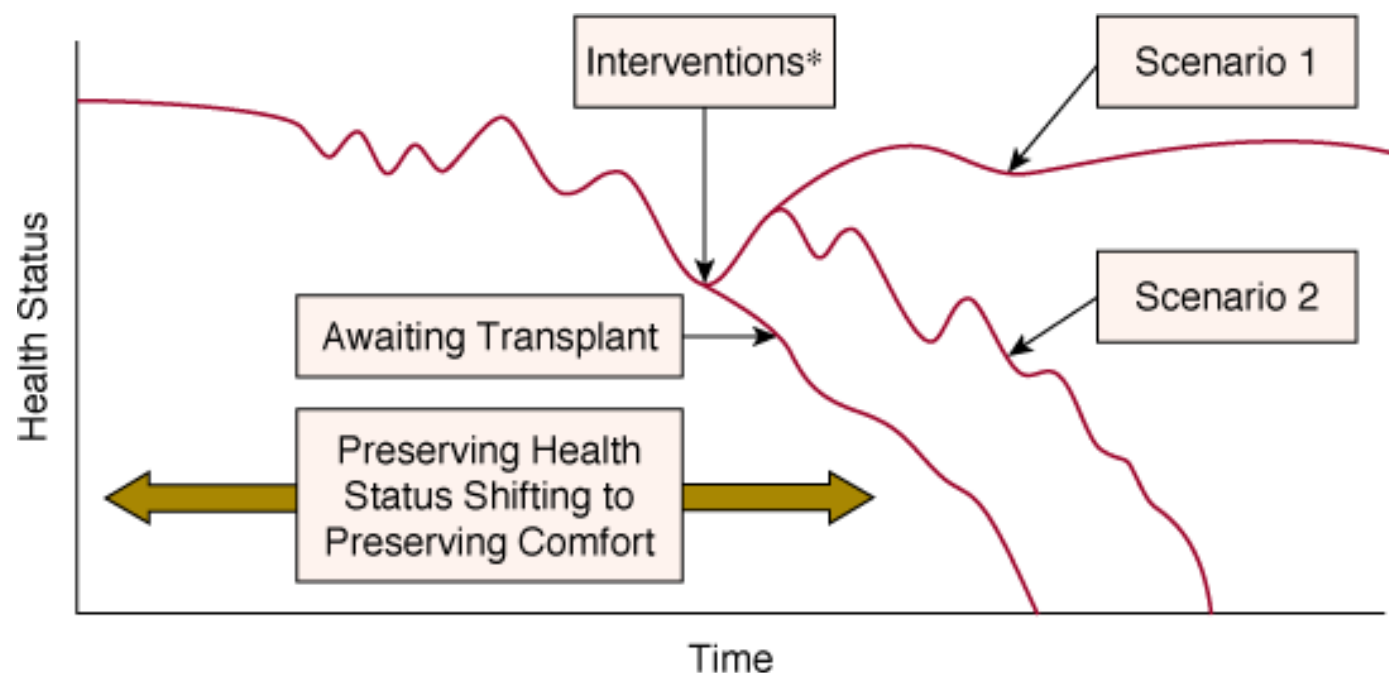


- **Life-Limiting Condition (LLC)** –there is no reasonable hope for cure, and will likely ultimately be fatal (e.g. muscular dystrophy)
- **Life-Threatening Condition (LTC)** – curative treatment may be feasible but may fail (e.g. cancer)
- **Chronic Complex Conditions (CCC)** - Any medical condition expected to last at least 12 months (unless death intervenes) and/or any condition that involves several organ systems of one organ system enough to require specialized pediatric care, often at a tertiary medical center (congenital heart disease, cerebral palsy, trisomy)
- Many of these children will benefit from palliative care to help with symptom management, decision-making, support

Perinatal and Pediatric Palliative Care

- Support quality of life and attempt to relieve suffering for children and families with serious illnesses, regardless of the stage of illness
- Make each day the best possible and provide the best possible care
- Support can and should be initiated early and integrated with curative treatment
- In a large multi-center cohort trial, over 2/3 of palliative care referrals were alive one year later





* Examples of interventions: chemotherapy, radiation therapy, organ transplantation, gastrostomy feeding tube, antireflux surgery, treatment of acute respiratory exacerbations from chronic pulmonary aspiration, medication changes for intractable epilepsy.



Palliative Care...

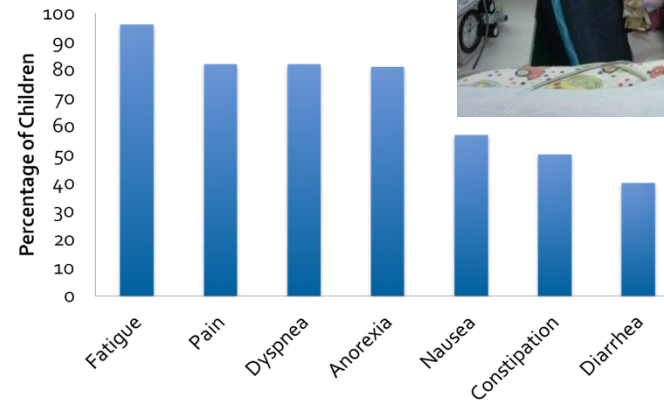
- is NOT closing the door on curative treatment
- is NOT pushing for less aggressive therapy
- is NOT only hospice and end of life care
- is NOT taking away hope
- is NOT giving up

Tasks of Palliative Care

- Communication
 - Identifying problems and challenges
 - Understanding illness
 - Developing prognostic awareness ***
 - Exploring hopes, concerns
 - Setting goals
 - Advanced care planning
 - Making decisions
- Care coordination
 - Collaborating with other providers
 - Facilitating logistics of medical & social needs
 - Partnering with community programs
 - Identifying community resources
 - Transition to home
- Symptom management
 - Daily issues
 - Anticipated symptoms
 - Emergency issues
- Bereavement Care for family
 - Assessing for anticipatory grief
 - Tools for healthy grieving
 - Follow up

“Words are, in my not-so-humble opinion, our most inexhaustible source of magic; capable of both inflicting injury, and remedying it.”

**- Albus Dumbledore,
Harry Potter and the Deathly Hallows**



Wolfe, J., et al. *N Engl J Med.* 2000; 342:326-33.



Quad-City Times

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HOT TOPICS: GO & VIEW | SHUTDOWN'S EFFECTS | IOWA REDISTRICTING | ASK THE TIMES: SIRENS | NOISY SEMIS | M

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QCTIMES.COM/MAGGIE

Maggie faced death, chose to live life her way

Story Discussion Video Image (2) Audio Font Size: - +

Kay Luna | Posted: Monday, April 5, 2010 4:47 am | Loading...



Maggie's suffering is over.

Her mother imagines the 12-year-old girl running, dancing and flying with the angels.

Maggie always did want to fly, Diane Wiederholt somberly says while sitting in what was her youngest daughter's bedroom at their rural Walcott, Iowa, home.

The thought reminds her of when Margaret "Maggie" Rose Wiederholt was wee-little. She used to jump off the family's picnic table, expecting to soar with the birds.

Instead, her fragile body would crash to the ground.

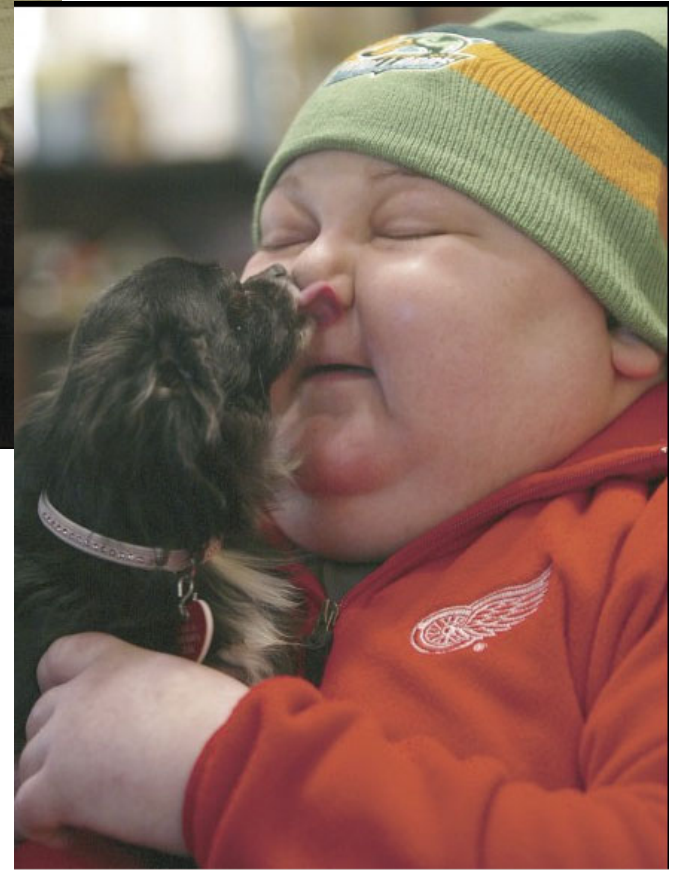
- Maggie
- 12 year-old girl with an autoimmune disease causing multiorgan failure
- Treatment no longer clearly providing benefit, but causes high burden
- Parents want her to be able to give input into treatment plan given the poor prognosis and uncertainty ahead...



HOME // GALLERY // MAGGIE DECIDES TO END THE FIGHT



How not to have a code status discussion...



Questions to guide informed decision making

From Atul Gawande's book *Being Mortal*
and NY Times article, *The Best Possible Day*



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- What is your understanding of your health or condition?
 - How have things changed over the past year?
 - What have the doctors told you about what to expect in the future?
 - What are you hoping this treatment will do for you?
- What are your goals if your condition worsens?
 - If the treatment doesn't help like we hope it will, what else is important to you?
 - How does being comfortable or being at home compare in priority to living longer?
- What are your fears?
 - What are your concerns about the future?
 - Is there anything that keeps you up at night worrying?
- What are trade offs are you willing to make or not to make?
 - What are you willing to live with?
 - What are you willing to live without?
- Then, communicate clearly and compassionately about achievable outcomes
 - “I hope... but I worry...” or “I wish... I worry... I wonder...”
 - Give best estimate of possible outcome:
 - Time ranges: days to weeks, weeks to months
 - Range of functional outcomes with concrete examples

How can we guide families through...

What parents want...

- Honest information so they can make good decisions, even if it is difficult news
- Ongoing relationships with trusted providers
- To be able to continue to provide love, hope, comfort, and care – to be seen as “a good parent” and to have “done everything”
- A sense of security and privacy for the family
- To exercise responsibility for what happens to the child to an appropriate degree, but not to feel responsible for causing their death

One tool to consider...

Time-limited trial

- When the benefit of an intervention is unclear, a time-limited trial can be implemented to determine benefit vs. burden
- Establish clear parameters and timelines in advance
- Appropriate only when there is a **reasonable chance** that therapy will have a net benefit to the patient and help achieve goals
- Consider making a specific recommendation when you feel the goals of care align with a specific treatment plan

What about the patient?

Children and adolescents

- Often rate symptom burden and quality of life much differently from parents or healthcare providers
- May or may not want to be involved in various parts of the healthcare discussion
- Can suffer from isolation, guilt, and uncertainty if they feel the topic is taboo and cannot talk about it
- Benefit from controlling at least certain aspects of their care, having opportunity to ask questions, and give final gifts



Dyspnea and Secretions

- Dyspnea “...subjective experience of breathing discomfort that consists of qualitatively distinct sensations” often described as “air hunger”
 - Mismatch between neural stimulus to breathe & mechanical output
 - Imbalance of metabolic demand vs. respiratory ability
 - Combination of physiological, psychological, social, environmental factors
 - Etiologies: Hypoventilation, shunt, diffusion impairment, VQ mismatch, airway obstruction, impaired chest expansion, neuromuscular disease, depressed respiratory drive, increased demand

American Thoracic Society. *Am J Resp Crit Care Med*. 1999; 159(1):321-40.

- Secretions – especially problematic for many with neurologic conditions
 - Etiologies: hyporesponsive gag, dysfunctional swallow, too weak to cough
 - Can lead to episodes of choking, cough, or to silent aspiration
 - May have gurgling noise from build up of secretions in posterior oropharynx

Wildiers, H., et al. *J Pain Symptom Manage*. 2009; 38:124-33.

Wildiers, H, & Menten, J. *J Pain Symptom Manage*. 2002; 23(4):310-7.

Management of Dyspnea near End of Life

Non-pharmacologic treatments

- Elimination of respiratory irritants
- Elevation of head of bed
- Positioning
- Cool cloth to face/neck
- Fan
- Nasal strips
- Relaxation techniques
- Essential oils
- Suctioning/pulmonary toilet
- Cough assist

Ullrich, CK & Mayer, OH. *Pediatr Clin N Am.* 2007; 54: 735-56.

Abernethy, AP, et al. *Lancet.* 2010; 376:784-93.

Jennings, AL, et al. *Cochrane Database of Scientific Reviews.* 2001(3).

Jennings, AL, et al. *Thorax.* 2002; 57(11):939-44.



Medical treatments

- Supplemental O₂ if hypoxic
- Non-invasive ventilation
- Anxiolytics if anxious
- Steroids or cough suppressants if indicated
- Opioids
 - Reduce central perception of dyspnea
 - Can reduce oxygen consumption and work of ventilation
 - Systemic can be dosed 50-75% lower than for pain relief initially and titrated as needed
 - Nebulized – no proven benefit
 - Survival time after compassionate extubation was no different after giving appropriate dose of morphine

Mahler, DA, et al. *Chest.* 2010; 137:674-91.

Management of Secretions near End of Life



Non-pharmacologic treatments

- Positioning – turn on side so can pool and exit orally
- Suctioning if benefit greater than burden (minimize invasiveness)
- Decision-making about route and type of artificial nutrition and hydration
- Educate family and providers about the normal process the body is going through

Medical treatments

- Glycopyrrolate (Robinul), scopolamine (patch), atropine (sublingual use of ophthalmic drops), hyoscyamine
- Consider anti-reflux medications if felt to be a large component, but not a cure a
- Botox injections or removal of salivary glands
- Tracheostomy or laryngotracheal separation

Perinatal and Pediatric Palliative Care in Iowa



- Pediatric Pain and Palliative Care Program at the UI Stead Family Children's Hospital - formed in 2009
- SHINE Perinatal & Pediatric Palliative Care Program at Blank Children's Hospital, UnityPoint Health, Des Moines
- ChildServe offers palliative care
- Many hospices across the state are willing to take pediatric patients



Tools for health care professionals

- Conversation guides for families
 - <https://theconversationproject.org/wp-content/uploads/2017/02/ConversationProject-StarterKit-Pediatric-English.pdf>
- Advance care planning
 - Emergency information form
 - Advance directives
 - My Wishes (children)
 - Voicing My Choices (teens)
 - Honoring Your Wishes (adults)
- IPOST
- EMR Order sets
 - Newborn palliative
 - Comfort Care/EOL
 - Compassionate extubation
- Hospice
 - Concurrent care

Emergency Information Form for Children With Special Needs

American College of Emergency Physicians | American Academy of Pediatrics

Form completed By: _____ Reviewed: _____ Initials: _____

Name: _____ Birth date: _____ Nickname: _____

Home Address: _____ Home/Work Phone: _____

Parent/Guardian: _____ Emergency Contact Names & Relationship: _____

Signature/Consent: _____

Primary Language: _____ Phone Number(s): _____

Physicians:

Primary care physician: _____ Emergency Phone: _____

Current Specialty physician: _____ Fax: _____

Specialty: _____ Emergency Phone: _____

Current Specialty physician: _____ Fax: _____

Anticipated Primary ED: _____ Pharmacy: _____

Anticipated Tertiary Care Center: _____

Diagnoses/Past Procedures/Physical Exam:

1. _____ Baseline physical findings: _____

2. _____

3. _____ Baseline vital signs: _____

4. _____

Synopsis: _____ Baseline neurological status: _____

Immunizations (mmv):

DTaP: _____ Tdap: _____

OPV: _____ Hib: _____

MMR: _____ MMR2: _____

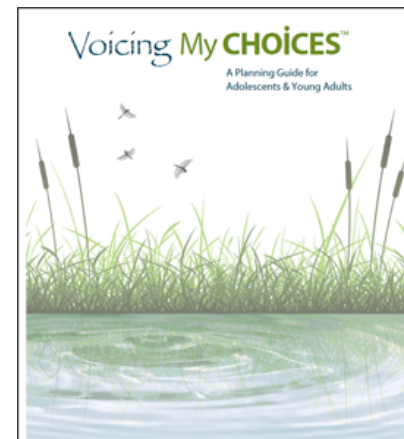
Antibiotic prophylaxis: _____ Indication: _____ Medication and dose: _____

Common Presenting Problems/Findings With Spec:

Problems: _____ Suggested diagnosis: No

Comments on child, family, or other specific medical issues: _____

Physician/Provider Signature: _____



Pediatric Starter Kit

Having the conversation with your seriously ill child

IPOST

Iowa Physician Orders for Scope of Treatment



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- **Recipients:** Intended for individuals who are frail and elderly **or who have a chronic, critical medical condition or a terminal illness**
- Transfers advance care planning into medical orders that are applicable across care settings
 - In the home or daycare (if EMS is called for help)
 - In a long term care facility or rehab facility
 - In the hospital, clinic, or dialysis setting
- Reflects a discussion of prognosis and goals and can be revoked or changed over time
- If you see that a patient has one, and it is not being followed, speak up!

IPOST



Iowa Physician Orders for Scope of Treatment

HIPAA PERMITS DISCLOSURE OF IPOST TO OTHER HEALTH CARE PROVIDERS AS NECESSARY

**Iowa Physician Orders
for Scope of Treatment (IPOST)**

First follow these orders, THEN contact physician or nurse practitioner. This is a Physician order sheet based on the person's current medical condition and wishes. Any section not completed implies full treatment for that section. Everyone shall be treated with dignity and respect.

Last Name

First/Middle Name

Date of Birth

A Check one	CARDIOPULMONARY RESUSCITATION (CPR): <u>Person has no pulse AND is not breathing.</u>	
	<input type="checkbox"/> CPR/Attempt Resuscitation <input type="checkbox"/> DNR/Do Not Attempt Resuscitation	
B Check one	MEDICAL INTERVENTIONS: <u>Person has a pulse AND/OR is breathing.</u>	
	<input type="checkbox"/> COMFORT MEASURES ONLY Use medication by any route, positioning, wound care and other measures to relieve pain and suffering. Use oxygen, suction and manual treatment of airway obstruction as needed for comfort. <i>Patient prefers no transfer to hospital for life-sustaining treatment. Transfer if comfort needs cannot be met in current location.</i>	
	<input type="checkbox"/> LIMITED ADDITIONAL INTERVENTIONS Includes care described above. Use medical treatment, cardiac monitor, oral/IV fluids and medications as indicated. Do not use intubation, or mechanical ventilation. May consider less invasive airway support (BiPAP, CPAP). May use vasopressors. <i>Transfer to hospital if indicated, may include critical care.</i>	
	<input type="checkbox"/> FULL TREATMENT Includes care described above. Use intubation, advanced airway interventions, mechanical ventilation and cardioversion as indicated. <i>Transfer to hospital if indicated. Includes critical care.</i>	
Additional Orders: _____		
C Check one	ARTIFICIALLY ADMINISTERED NUTRITION Always offer food by mouth if feasible.	
	<input type="checkbox"/> No artificial nutrition by tube. <input type="checkbox"/> Defined trial period of artificial nutrition by tube. <input type="checkbox"/> Long-term artificial nutrition by tube.	
D	MEDICAL DECISION MAKING	
	Directed by: (listed in order of Iowa Code/Statute for Priority of Surrogates; check only one) <input type="checkbox"/> Patient <input type="checkbox"/> Durable Power of Attorney for Health Care <input type="checkbox"/> Spouse <input type="checkbox"/> Majority of Adult Children <input type="checkbox"/> Parents <input type="checkbox"/> Majority rule for nearest relative <input type="checkbox"/> Other: _____	Rationale for these orders: (check all that apply) <input type="checkbox"/> Advance Directives Year AD completed: _____ <input type="checkbox"/> Patient's known preference <input type="checkbox"/> Limited treatment options <input type="checkbox"/> Poor prognosis <input type="checkbox"/> Other: _____

Physician/ARNP/signature (mandatory) Print Physician/ARNP/ Name Date Phone Number

Patient/Resident or Legal Surrogate for Health Care Signature (mandatory) Date

SEND FORM WITH PERSON WHENEVER TRANSFERRED OR DISCHARGED



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Iowa Physician Orders for Scope of Treatment

UISF Children's Hospital IPOST Metrics

November 1, 2012—May 17, 2018



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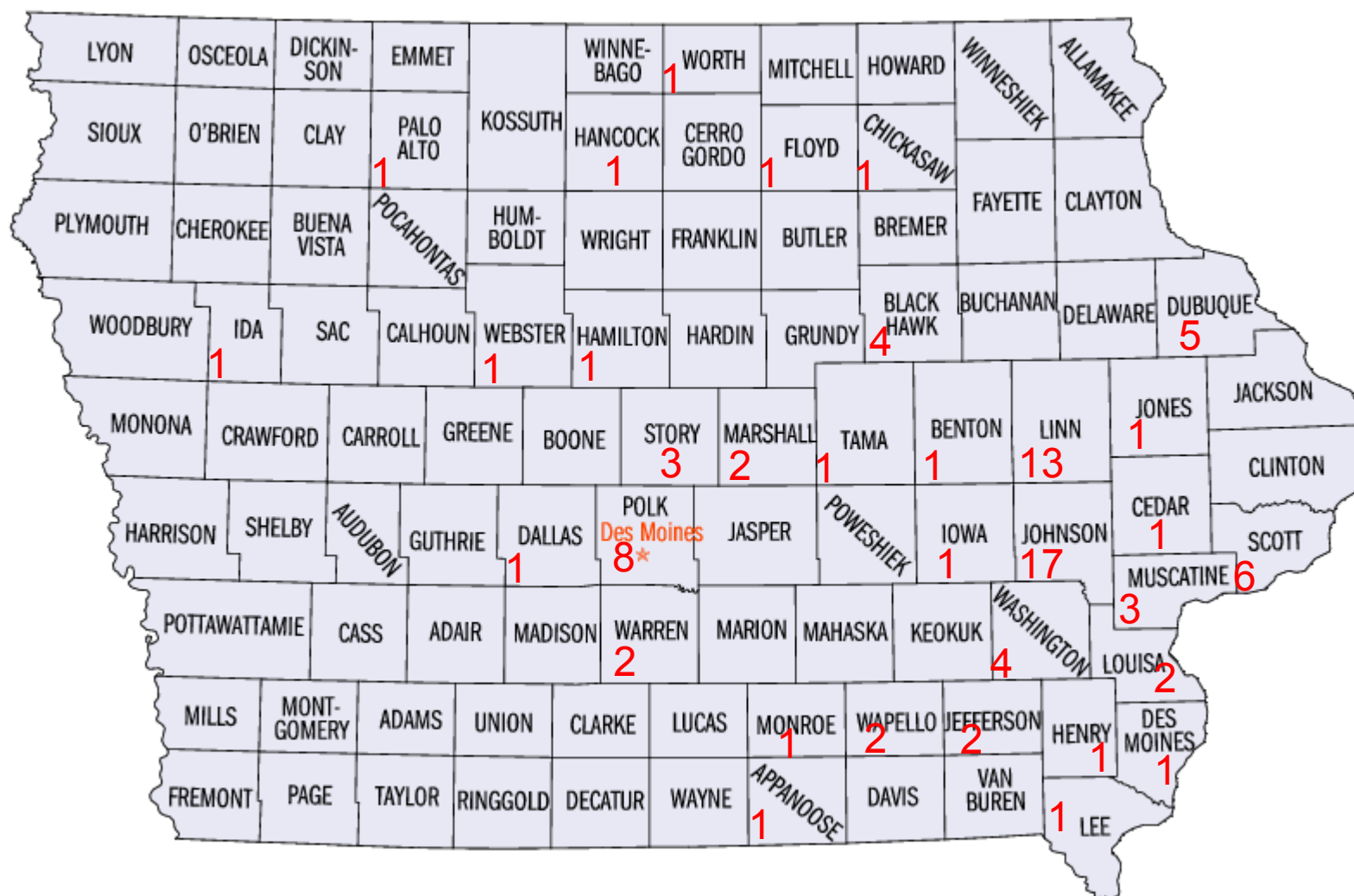
- **IPOST forms completed:** 92
- **Indications**
 - DNR/Comfort: 35
 - DNR/Limited: 31
 - DNR/Full Treatment: 15
 - CPR/Full Treatment: 11
- **Conditions**
 - Hem/Onc: 19
 - Neuro: 52
 - Genetic: 8
 - Cardiac: 5
 - Duchenne MD: 6
 - Cystic Fibrosis: 1
 - Other: 1
- **Patient Outcomes**
 - Living patients: 47
 - Deceased patients: 45
- **Patients receiving hospice support:** 32 of 92
- **Number of Iowa counties where children with IPOST forms reside:** 33
- **IPOST is dynamic**
 - At least 10 families have changed their IPOST preferences

IPOST data slides courtesy of Sheila Frascht, RN, CHPPN, CPLC
Nurse Clinician - Pediatric Pain & Palliative Care
University of Iowa Children's Hospital

UI Children's Hospital Patients with IPOSTs



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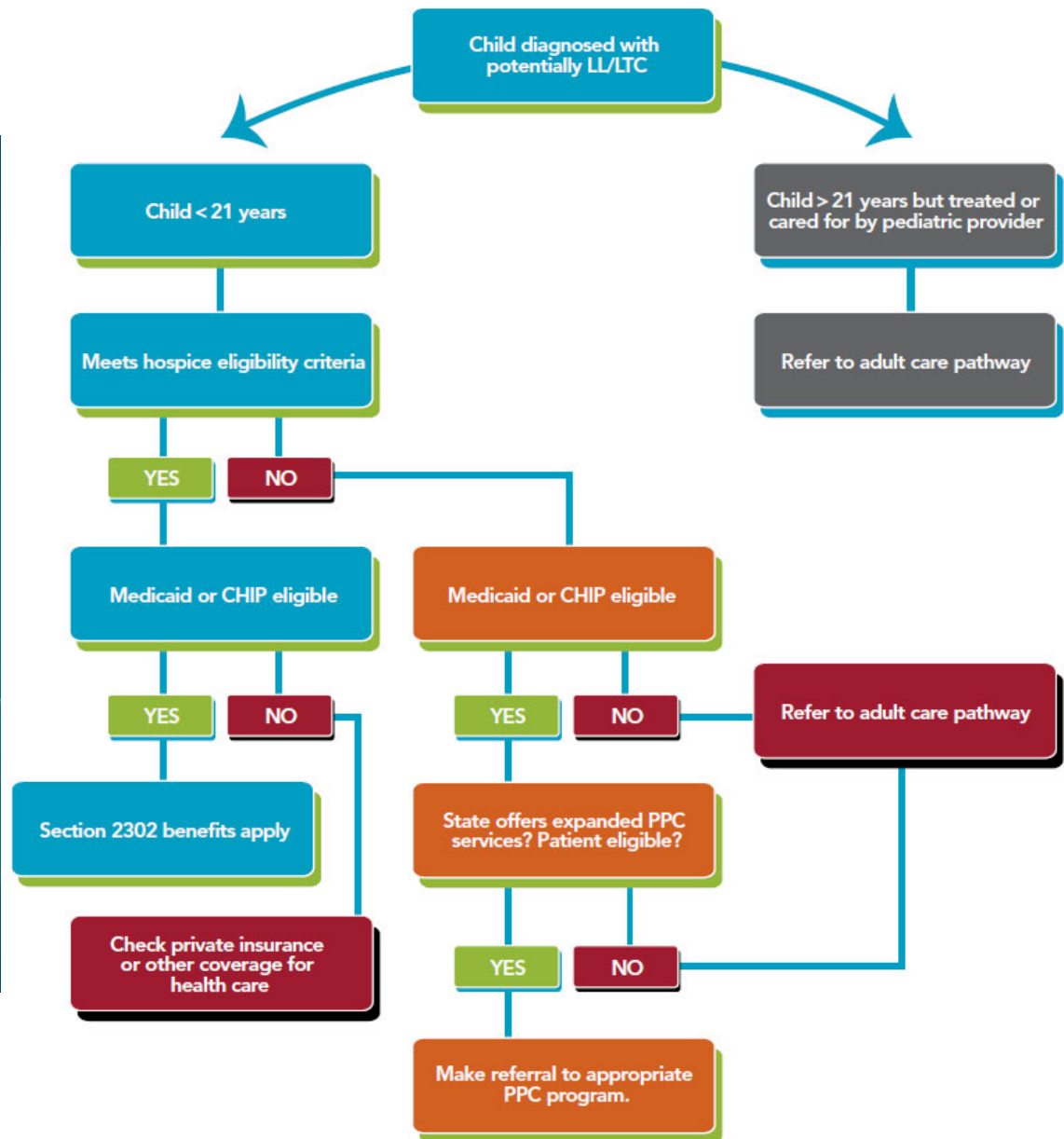
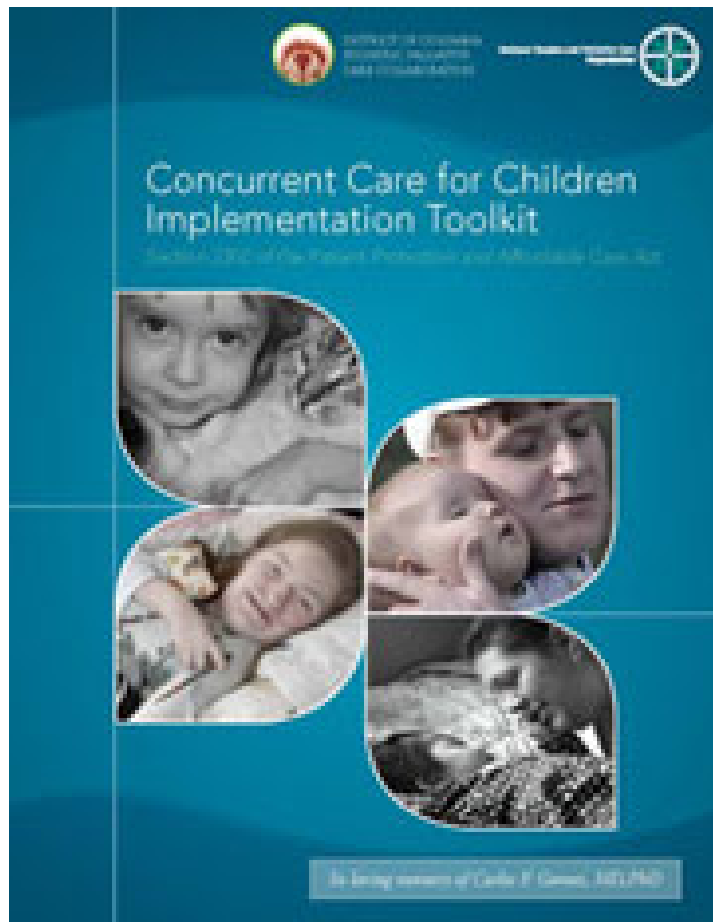
Data Collected from November 2012- May 2018

Concurrent Care for Children – Hospice plus



- Signed into law in 2010 by President Obama as part of the ACA
- Applies to children (< 21 years of age) who:
 - Are covered by Medicaid and Children's Health Insurance Program (CHIP), or EPSDT
 - Have a 6 month or less prognosis and are eligible for hospice services
- State Medicaid pays for curative treatment and hospice care separately (after private insurance does its part)
- Iowa has no specific plan to implement this, but it has been successfully done on a case by case basis

Healthcare Professional and Hospice Decision Process
Section 2302 and Pediatric Palliative Care



Other Resources

- November 15 Peds Pall conf
- Getpalliativecare.org
- Vitaltalk.com
- Courageousparentnetwork.com

