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Providing Neuro-Palliative Care for Children Who Have Severe Neurodevelopmental Impairments



Julie Hauer, M.D.

Palliative care is a model of interdisciplinary care for patients with serious, complex illnesses. It addresses suffering and provides assistance for people faced with significant medical decisions. Palliative care for children aims to improve the quality of life for patients and their families. It seeks to prevent and relieve suffering by promptly identifying and treating pain and other problems, whether they are physical, psychosocial or spiritual (Liben, 2007; Himelstein, 2004). Interdisciplinary palliative care teams often include a palliative care physician, an advanced practice nurse, a chaplain and a social worker.

Children who have severe neurodevelopmental impairments — whether from neurologic devastation, genetic disorders, congenital anomalies or metabolic disorders — often have chronic and complex medical needs as well. Pediatric palliative care teams commonly see children with such conditions. The teams at Children's Hospital Boston/Dana Farber Cancer Institute and at Seattle Children's Hospital identified that 47 to 59 percent of consultations fell into those medical categories. The program at Akron Children's Hospital stated that "the largest number of referrals for palliative care is for neurologic, neurodegenerative and genetic disorders."

The families and clinicians who care for such children often encounter challenges related to the child's uncertain future and the need to manage unique symptoms. Concern about a child's functional outcome and health-status trajectory can cause stress for the family. Symptoms that often need management include chronic irritability, gastrointestinal distress and recurrent respiratory distress.

This article will address how the model of palliative care can guide families through such uncertainty while integrating symptom management into the child's medical care.

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Addressing Uncertainty

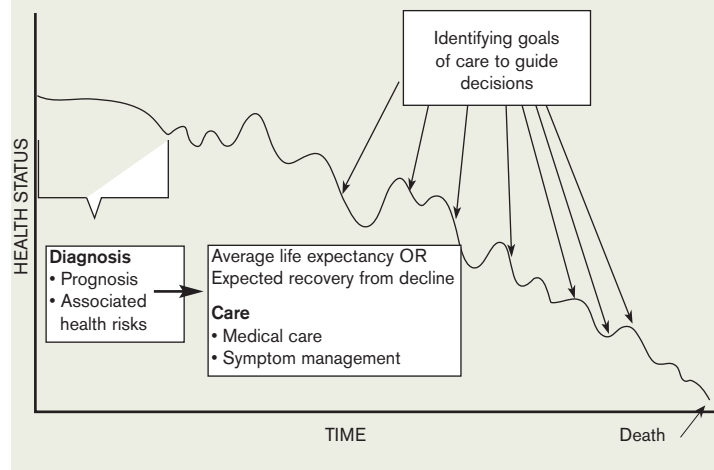
Rachel is a 3¹/₂-year-old who has refractory epilepsy. She has not improved despite multiple medication trials, resection of a dysplastic left parietal area, the ketogenic diet, intravenous immunoglobulin (IVIG) therapy and a left hemispherectomy. Rachel lost previously acquired motor and communication skills six months ago and remains unable to ambulate or communicate. She has had six hospital admissions in the past five months and has experienced recurrent bouts of aspiration pneumonia despite anti-reflux surgery and placement of a gastrostomy feeding tube. Rachel's doctor recommends a vagus nerve stimulator.

Figure 1 shows a hypothetical disease trajectory that represents children with severe neurodevelopmental impairments. The figure demonstrates that identifying a diagnosis — whether a specific medical diagnosis or one defined by the severity of the neurodevelopmental impairment — allows us to anticipate a child's clinical course and associated health problems. Health issues can be specific to a diagnosis, such as identifying a cardiac defect or central apnea in a child with a genetic disorder. On the other hand, issues that affect health can be specific to the severity of a child's motor impairment and associated tone abnormality, regardless of the cause of the impairment. Examples include oral motor dysfunction and recurrent pulmonary aspiration.

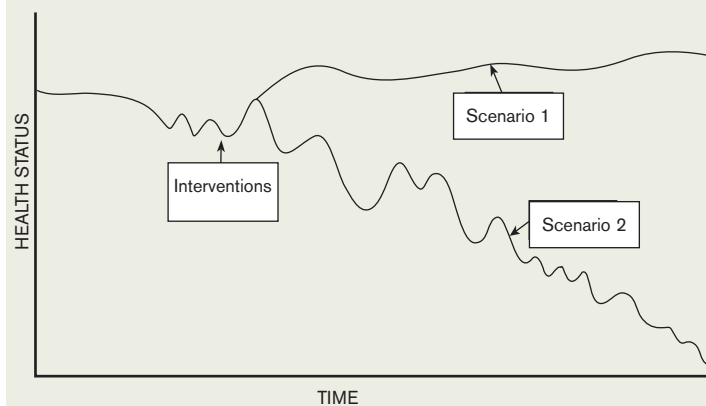
Such knowledge guides us toward identifying chronic and acute medical treatments to minimize health effects while maximizing functional outcomes. By monitoring for changes in health status and noting any decreased benefit from treatments, we can identify children who have life-limiting conditions and are at risk of experiencing life-threatening events.

Figure 2 further highlights the uncertainty of knowing what level of recovery will result from the interventions used. In addition, information concerning average life expectancy is of limited use for an individual child. Instead, the greatest benefit comes from allowing each child's clinical and life story to

■ Figure 1: Integrating Medical and Palliative Care



■ Figure 2: Hope for the Best, Prepare for the Worst



*Examples of interventions might include: gastrostomy feeding tube, treatment of gastroesophageal reflux, antireflux surgery, treatment of acute respiratory exacerbations, and medication changes for intractable epilepsy.

■ **Table 1: Communication Tools Used In Palliative Care**

OBJECTIVES	SUGGESTED LANGUAGE
Determine goals of care as a guide to making decisions	<ul style="list-style-type: none"> • We'll always review how the interventions available meet the goals you've identified. • Goals commonly include comfort and quality of life, might shift to providing care at home when less benefit is seen from care in the hospital, and can be specific activities, such as maintaining current health status for a family trip.
Focus on parental expertise	<ul style="list-style-type: none"> • Though I will honestly reflect on changes in your daughter's functional and health status, you are the expert in how such changes affect her quality of life. • Though we bring expertise in evaluating and managing health problems in children, you are the expert in how such problems look in your daughter.
Anticipate issues	<ul style="list-style-type: none"> • We know that worries of parents at such times often include: Is my child suffering? Will my child get better? Is there more we can do?
Reflect on and acknowledge the limits of interventions	<ul style="list-style-type: none"> • Your daughter continues to receive exceptional care at home. My observation is that we are seeing fewer benefits from the treatment options. What have you observed?
Hope for the best; prepare for the worst	<ul style="list-style-type: none"> • We know families often hold simultaneously their hope for the best possible outcome with their realistic understanding of the severity of the problem.
Offer continuity with a safety net	<ul style="list-style-type: none"> • We might not know when your daughter will have more problems, but we can ensure that you have a team available to help guide and support you on this journey.
Identify decision-making as a dynamic process	<ul style="list-style-type: none"> • What might seem like the "right" decision today could change. Your decisions are never final and can change as the clinical course changes.

unfold. Palliative care allows time to be our guide while we “hope for the best and prepare for the worst.”

Rachel's mother is concerned that more interventions will provide no long-term benefit while resulting in more admissions and discomfort for her daughter. She believes she has “lost” part of Rachel. She still hopes for improvement in Rachel's functional and health status, but she wishes to avoid further invasive procedures. During several meetings, the medical team, palliative care team, and Rachel's mother discuss medical options along with the family's goals, hopes and fears.

Table 1 outlines some of the communication tools used in palliative care. Defining goals of care is a critical part of ensuring that interventions meet identified goals rather than treat isolated medical problems. Palliative care recognizes that it is distressing for parents and physicians to encounter the limits of medicine. At such times, people often try to do **more** out of a sense that they need to do **something**.

Palliative care minimizes emotional, existential and physical distress at such times. It brings expertise in discussing the limits of medical care, focuses on goals of care to guide decisions, and identifies other care options, such as symptom management. It recognizes that there is always care to provide as goals shift from treatment and cure to quality of life and comfort. With palliative care, we never accept that “there is nothing more we can do,” even as we reach the limitations of medical care.

Symptom Management

Palliative care also addresses physical needs by integrating symptom management into the medical care plan. It supports medical care that both minimizes health effects of associated problems and maximizes quality of life and comfort. Palliative care clinicians provide expertise in managing distressing symptoms that are unique to children with severe neurodevelopmental impairment: chronic irritability and agitation, chronic gastrointestinal distress, and dyspnea during acute respiratory illnesses.

Chronic Irritability, Agitation and Associated Gastrointestinal Symptoms

Children with neurological impairments experience pain more frequently than the general pediatric population does. In one study,

caregivers reported that 44 percent of children with severe cognitive impairments experienced pain each week during a four-week interval. Pain frequency was highest in the most impaired group of children. Parents identified the gastrointestinal tract as the most frequent source of pain (Breau, 2004).

Another study found that children with gastrostomy tubes, and those who take medications for gastroesophageal reflux or gastrointestinal motility, have significantly higher rates of pain (Houlihan, 2004). The association between pain and the gastrointestinal tract led to speculation by this author that visceral hyperalgesia is a source of chronic irritability and agitation in neurologically impaired children. In a retrospective series of nine neurologically impaired children who experienced significant unexplained pain and irritability, marked symptom improvement was identified when the children received gabapentin therapy titrated to standard doses (Hauer, 2007). For such children with persistent irritability, we suggest using medications, such as gabapentin, that have demonstrated benefits for neuropathic pain syndromes.

Dyspnea

Recurrent respiratory illness leading to respiratory failure is the most common cause of mortality in children with severe cerebral palsy. Aspiration is a frequent factor, identified in 31 to 68 percent of such children. This author suggests that aspiration pneumonia in neurologically impaired children is best understood as an acute exacerbation resulting from chronic contributing issues. Those issues include an ineffective cough, an impaired ability to manage routine oral secretions, hypoventilation, chronic aspiration resulting in inflammation and the introduction of oral bacteria, and development of mucous plugs with ventilation/perfusion (V/Q) mismatch.

Table 2 outlines chronic and acute home-care strategies, based on experience and evidence, for children with severe neurodevelopmental impairments and associated aspiration of oral secretions.

■ **Table 2: Home Management – Medical Treatment and Comfort Strategies**

CHRONIC INTERVENTIONS

Suctioning	As needed for comfort
Oxygen	Assessed by appearance of patient or by oximeter
Albuterol nebulizer	Every 3-4 hours for coughing, wheezing, congestion
Ipratropium (Atrovent) nebulizer	Every 3-4 hours for coughing, wheezing, congestion
Saline or Mucomyst nebulizer	As needed for coughing, wheezing, congestion
Chest physiotherapy or vest	2 times/day, increase to 3-4 times/day with increased symptoms*
Nebulized Budesonide (Pulmicort)	2 times/day, increase to 3-4 times/day with increased symptoms*
Salmeterol (Serevent)	If family history of allergies or improvement from daily Albuterol
Proton pump inhibitor	If clinical or diagnostic evidence of gastroesophageal reflux

ACUTE INTERVENTIONS FOR RESPIRATORY EXACERBATIONS FROM CHRONIC ASPIRATION

Clindamycin, Augmentin or Levofloxacin/Moxifloxacin [†]	10 – 14 days
Systemic steroids (Prednisone) [‡]	5 days

ADDITIONAL INTERVENTIONS FOR SYMPTOM MANAGEMENT AND END-OF-LIFE CARE

Fan on face	Relieves sensation of breathlessness
Morphine sulfate	Use for discomfort or respiratory distress; starting dose 0.1 mg/kg/dose PO/SL/Gtube; may increase by 30% until comfortable
Glycopyrrolate (Robinul) or Scopolamine	Might contribute to mucous plugging; decreases oral and respiratory secretions in end-of-life care

* Symptoms include increased coughing, secretions, congestion, respiratory rate and breathing effort.

[†] Use in children with aspiration when symptoms persist despite an increase in chronic interventions.

[‡] Include with third or fourth exacerbation, sooner if symptoms return within two months of antibiotic course.

For example, the table identifies antibiotics that provide coverage of anaerobic bacteria along with other oral bacteria when treating people who chronically aspirate oral secretions.

Rachel's care team supports her mother's decision to focus on active home-treatment strategies for respiratory exacerbations. A medical and symptom-management protocol is outlined, which includes morphine sulfate for respiratory distress, ativan for agitation and robinul for secretions. The first time Rachel's caregivers use the plan, they report that Rachel appears more comfortable and interactive than she has in months.

Often missing in care plans is management of dyspnea. Table 2 includes morphine sulfate for consideration as goals shift from medical treatment in the early phase to treatment and comfort as a decline in health status is observed. Evidence from adults indicates the effectiveness of morphine sulfate for dyspnea starting at 30 percent of the dose used for pain (Abernethy, 2003; Del Fabbro, 2006). Further evidence indicates the safety of morphine sulfate for managing dyspnea without development of respiratory depression (Clemens, 2007).

For such reasons, we likely underutilize morphine sulfate when managing respiratory distress, instead reserving its use for comfort at the end of life. There is a need to study the integration of morphine sulfate into the care plans of patients who have chronic aspiration and recurrent, distressing respiratory exacerbations.

Conclusion

Palliative care practitioners provide expertise that helps families and medical teams provide care that manages symptoms and ensures the best quality of life possible. Palliative care teams provide guideposts along a journey that often includes medical and developmental uncertainty. The interdisciplinary approach can help manage pain and other distressing symptoms while supporting spiritual and emotional needs. Further study is needed to determine how best to meet the unique needs of this patient population.

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Breath-holding Spells in Infants and Toddlers



Galen Breningstall, M.D.

In infants and toddlers, breath-holding spells are frequently mistaken for epileptic seizures. One type of breath-holding spell is described by Stephenson:

An unsteady toddler on his own trips and falls. His mother hears the bump but no succeeding cry and hurries to him. She finds her child lying deathly still with eyes fixed upwards, lips dusky. As she lifts him, he abruptly stiffens into rigid extension with jaw clenched and hands fisted, gives a few jerks, and after what seems an age (but in fact is less than half a minute) relaxes limply with an absent far-away look. Then he opens his eyes, at once recognizes his mother, cries a little, and drifts off to sleep, his face distinctly pale.

(Stephenson, 1978)

A more common type of breath-holding spell occurs in infants and toddlers who are upset, rather than injured. The children cry vigorously, turning red and then purple. Next they abruptly cease crying, sometimes stiffening their extremities and arching the back. They might flail an arm repetitively, then gasp. At that point, children lose consciousness and lie deathly still. Arousal follows, and crying often resumes.

The 17th-century French philosopher, Jean-Jacques Rousseau, described a breath-holding spell that terminated, as such spells often do, before syncope (loss of consciousness from lack of oxygen):

I shall never forget how I once observed one of these obnoxious screamers whom his nurse had slapped. He suddenly became quiet; I thought he was intimidated. I said to myself: One day this will be a servile mind which will be influenced only by sternness. But I was wrong; the unhappy child was suffocating from anger and had lost his breath; I saw him turn purple. A moment later a piercing scream erupted. All the signs of indignation and desperation of this age could be heard in his screaming: I was afraid he would succumb to this excitement. . . . I am sure a fire brand accidentally falling on the child's hand could not have excited him more than the rather light slap which had been delivered with the apparent intent of chastising him.

(Rousseau)

Who Experiences Breath-holding Spells?

Breath-holding spells typically occur in children 6 months to 4 years of age. The median age of onset is 6 to 12 months, and the median age of peak occurrence is 12 months to 18 months.

A majority of children have one to six spells per week, and 15 percent have more than one spell per day. In contrast, 25 percent have one spell or fewer per month. Of children who experience breath-holding spells, 54 to 62 percent are cyanotic, 19 to 22 percent are pallid, and 19 to 24 percent are mixed or unclassifiable. Children may have both types of spells.

Because breath-holding spells frequently occur in upset children (second scenario above), there is a misconception that breath-holding spells are symptomatic of psychosocial problems. Studies of the temperament of breath-holders, however, indicate that such children have tantrums no more frequently than other children do and that the parents of breath-holders have a quite normal parenting style. Breath-holders are not particularly difficult children.

Physiology of Breath-holding Spells

Breath-holding spells can create a transitory shortage in the supply of blood and oxygen to the brain, resulting in anoxic seizures. Studies in adult volunteers have shown that myoclonus (seizure-like movements) accompanies fainting (syncope) in 90 percent of episodes. (The study volunteers induce syncope with a combination of hyperventilation, orthostatic change, and a Valsalva maneuver, which tightens the chest against a closed airway.)

During an anoxic seizure, a lack of oxygen temporarily silences the upper brain centers. Presumably that silencing, in turn, releases the upper brain's control of the lower brain centers, allowing a seizure to occur. During an anoxic seizure, an electroencephalogram (EEG) pattern becomes diffusely slow and then flattens. Epileptic seizures, by contrast, involve an exuberant discharge of electrical activity in the brain.

Cyanotic Breath-holders

Cyanotic breath-holding (the type that follows vigorous crying) involves hyperventilation. Because hyperventilation decreases carbon dioxide in the blood, it also decreases blood flow and oxygen supply to the brain.

Breath-holding maladaptively occurs during expiration rather than following inspiration, thus reducing oxygen capacity in the lungs. The chest tightens because of a Valsalva maneuver, which increases intrathoracic pressure and impedes the return of cerebral venous blood to the heart. In addition, intrinsic pulmonary mechanisms — with ventilation perfusion mismatch or flow through abnormal bronchopulmonary arteriovenous anastomoses (interconnections) — might impair the oxygenation of blood in the lungs. The combined factors cause a transitory, critical reduction in the supply of oxygen to the brain.

Studies of the autonomic responses and sleep characteristics of such breath-holders have demonstrated some differences from those of other children. Anemia might contribute to breath-holding episodes in some children. Iron deficiency, even without anemia, might adversely affect autonomic function.

Pallid Breath-holders

Pallid breath-holders (first scenario, Page 6) frequently have parasympathetic hyperactivity, with excessive vagus nerve slowing of the heart following an unexpected injury — often a mild blow to the head. Indeed, the heart might stop beating for 10 seconds or more. In pallid breath-holders, that cessation seems to be the primary factor causing a transitory critical reduction in the brain's oxygen supply.

Diagnosis and Treatment

Breath-holding spells are invariably provoked, as previously described. It is unlikely for a seizure that follows crying or minor injury to be a manifestation of epilepsy.

Accordingly, a careful history is critical to diagnosing breath-holding spells. Examinations of breath-holders are expected to be entirely normal. In some instances, there is a family history of breath-holding spells or, more likely, of fainting. Laboratory evaluations are generally unrewarding. In some instances, anemia due to iron deficiency is present. In some children who experience breath-holding spells and have anemia, iron supplementation has seemed to reduce their susceptibility to breath-holding spells. An electrocardiogram (EKG) can screen for the unlikely occurrence of prolonged QT syndrome.

The daughter, almost 3, of missionaries was airlifted to the U.S. for evaluation of seizures. About a year earlier, she had fallen 6 feet from a ladder onto a cement floor. Perhaps surprisingly, she did not sustain serious injury. She was unconscious, however, and lay prone — with her back arched and arms extended — for a few seconds. Three weeks later, a similar episode followed a

minor injury. One month before her evaluation, she stumbled, and her mother pulled her to keep her from falling. Another stiffening episode occurred, this one associated with vigorous crying. Ultimately, the diagnosis was breath-holding spells.

Breath-holding spells, even when frequent, are quite unlikely to cause immediate or chronic injury. They are almost always self-limited events, requiring no intervention to terminate. During a breath-holding spell, some parents try counter-stimulation: for example, they blow on the child's face. It is reasonable to remove the child from any risk of falling and to lay the child on his or her side.

Aside from the possible efficacy of iron supplementation in some children, no particular intervention is effective in preventing cyanotic breath-holding spells. Certainly, it is undesirable to alter good parenting in an effort to prevent a child from becoming upset. Neither do pallid breath-holding spells generally require treatment. In unusual instances, some children have seemed to benefit from treatment with atropine or scopolamine. In truly unusual instances, patients with pallid breath-holding spells have been treated with cardiac pacemaker placements.

Generally, however, only the passage of time is necessary. Children simply outgrow their tendency to experience breath-holding spells. Such children might, however, have an increased risk of syncope in adolescence or adulthood.

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