Palliative Care Skills for Pediatric Neurology

Palliative care aims to improve quality of life, is appropriate at any stage of illness, and can be provided alongside treatment intended to cure.

By Adrienne M. Schuler, BS, BA; Jennifer Rubin, MD; and Tracy S. Gertler, MD, PhD

CASE. Baby P’s Family Is Concerned

Clinical Presentation

Baby P is 9 months old and was born full-term. She has been admitted to the hospital for failure to gain weight despite multiple adjustments to formula type and adequate recorded daily intake. Neurology was consulted because Baby P’s parents were worried about developmental regression. They report that Baby P had been able to sit independently at 6 months old but now needs a baby seat with straps. They say, “We keep bringing her in and changing formulas, but nothing is helping.”

Physical and Neurologic Examination

Baby P’s physical examination was notable for hirsutism, weight below the 3rd percentile for her age, and height and head at the 10th percentile. Her mental status was age appropriate with early eye contact, tracking of light and sound, a social smile, and spontaneous vocalization. Cranial nerve examination had normal findings. The motor exam was most notable for axial hypertonia with limited passive hip flexion and hyperreflexia at the biceps (3+) and patellae (4+) with sustained clonus. Tactile stimulation was grossly intact, and Baby P reached for toys without dysmetria.

Empathic continuers (Table 1) are potential responses to these indirect cues with statements that address the patient or family’s emotions, validate their feelings, and set the stage for additional conversations. Empathic continuers strengthen the relationship between patient, family, and clinician and create space for patients and families to share information they otherwise may not have chosen to share. Use of empathic continuers helps clinicians better understand the patient and family’s experience; allows them to address emotional, spiritual, and physical concerns; and ultimately leads to decreased anxiety and depression, as well as greater adherence to therapy.

Communication Skills

Communicating with patients and families is for information gathering, information giving, and relationship building. Throughout conversations there are opportunities to respond to cues or clues about families’ concerns with active listening and empathy. For example, rather than voice a specific concern, families may state, “We’re trying everything, and it hasn’t worked.” These indirect cues are often missed by healthcare providers.
Delivering Serious News

When sharing serious medical information with families, the Ask-Tell-Ask cognitive road map (Table 2) is helpful to gauge a family’s baseline knowledge, disclose information in a clear and concise manner, and reassess for understanding. Clinicians should resist the temptation to fill any silence or pauses with excessive medical details and jargon, and instead respond to a family’s emotions with empathic continuers (Table 1).

Managing With Hope and Worry

Over the course of a life-limiting disease, such as neurodegenerative or neuro-oncologic disorders, patients and families may swing between expressions of hope and realism. As in this case, these experiences may be further complicated by the patient’s fluctuating clinical status. Parents may wish to enroll their child in clinical trials, hoping for discovery of a novel, curative treatment, while simultaneously wanting to bring their child home and avoid additional hospitalizations. Adolescents may express a desire to be healthy once again and to grow up to achieve certain goals, but they may also acknowledge their fears of what will happen to their family when they are gone.

Diagnostic Testing

Baby P’s biochemical and metabolic blood and urine test results were notable only for a blood lactate of 3.8 mmol/L. Her brain and spine MRI demonstrated a longitudinally extensive T2/fluid-attenuated inversion recovery (FLAIR) hyperintense signal in the posterior spinal cord columns. She also had subtle diffusion restriction in her basal ganglia, pons, and deep gray cerebellar nuclei bilaterally. Taken together, these results were highly suggestive of a genetic/metabolic etiology. Whole exome sequencing revealed biparental inheritance of compound heterozygous pathogenic variants in cytochrome c oxidase assembly factor (SURF1). This nuclear gene encodes a mitochondrial chaperone protein essential for formation of the cytochrome c oxidase (complex IV) holoenzyme of the electron transport chain. A diagnosis of SURF1-associated Leigh syndrome was discussed with the Baby P’s family.

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CASE (continued). Baby P’s Doctor Receives Confirmatory Test Results

A gastrostomy tube was placed, and overnight continuous feedings helped Baby P gain weight, but she experienced daily emesis, abdominal bloating, and discomfort. Physical and occupational therapy several times per week facilitated more frequent rolling and assisted sitting. Over the next 6 months, however, Baby P’s development regressed again, and she required more truncal support. In the setting of a viral respiratory infection, she developed saccadic eye movements, cerebellar titubation, and difficulty grabbing toys because of past pointing. Although Baby P briefly improved, weeks later she experienced respiratory distress leading to cardiopulmonary arrest that was treated with epinephrine and endotracheal intubation.

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### TABLE 1. NURSE PROTOCOL FOR RESPONDING TO EMOTIVE STATEMENTS

<table>
<thead>
<tr>
<th>Steps</th>
<th>Definition</th>
<th>Examples</th>
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<tr>
<td>N</td>
<td>Naming emotions</td>
<td>Use verbal and nonverbal communication (eg, sighs, tears, body language) to label difficult emotions</td>
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<tr>
<td>U</td>
<td>Understanding</td>
<td>Verbally recognize the emotional and psychologic gravity of a situation</td>
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<td>R</td>
<td>Respecting roles</td>
<td>Acknowledge the family’s role in caring for the child</td>
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<td>S</td>
<td>Supporting needs</td>
<td>Inform the family they are not alone should they need emotional support or medical information</td>
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<tr>
<td>E</td>
<td>Exploring</td>
<td>Open the door to conversations regarding emotions</td>
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### TABLE 2. ASK-TELL-ASK METHOD TO DELIVER DIFFICULT NEWS

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<th>Ask</th>
<th>Tell</th>
<th>Ask</th>
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<td>Evaluate understanding of treatment course and prior discussions</td>
<td>“To make sure we start from the same place, what is your understanding of why we performed genetic testing?”</td>
<td>“That was a lot of information. What questions do you have for me?”</td>
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<tr>
<td>Disclose information</td>
<td>“I have some tough news to share. The genetic test confirmed that your daughter has a serious condition called Leigh syndrome.”</td>
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Although it may be difficult to balance these seemingly contradictory emotions, emphasizing only one or the other aspect can be detrimental to the patient and family. Sole focus on hope can deprive patients of conversations about fear and pain, whereas a focus only on realism can cause patients to feel abandoned in their hope. Acknowledging this duality can be helpful in identifying and naming emotions.\(^4\)\(^5\)

Statements that include both hope and worry can be an impactful way of acknowledging and holding space for hope, while still introducing potentially worrisome scenarios. Avoid the temptation to use the word “but” to connect hope and worry statements because it may unintentionally move the focus to what comes after the “but” and negate what is before. For example, one might say, “I too hope her breathing improves and we can safely get her off the respirator. I worry her muscles are too weak to support breathing on their own.”

### Requesting a Palliative Care Consult

#### CASE (continued). Baby P’s Team Consults Specialty Palliative Care

Over the next several weeks in the hospital, Baby P had transient improvements in her respiratory capability, tolerance of gastrostomy feeds, and when treatments for pulmonary clearance were escalated, mental status. She was, however, unable to maintain adequate oxygen saturation with noninvasive ventilation. Profound autonomic dysregulation resulted in multiple code events with hypotensive bradycardia. Baby P became increasingly irritable with routine care and less interactive with family, therapists, and other healthcare workers. Repeat brain imaging was obtained to exclude intracranial infections or hydrocephalus and showed clear progression of her metabolic disease with increasingly large areas of restricted diffusion and cortical atrophy.

Care conferences began to focus on clarifying the goals of care. Baby P’s parents felt strongly that a life consisting of recurrent intubation, sedation, and withholding of nutrition was not consistent with their values. They also worried that their religious community and extended family members had different perspectives and wondered if withdrawal of care was ethical. Ultimately, given the circumstances, Baby P’s parents were united in their wish to allow her one more opportunity to feel sunshine on her skin, sense the motion of her stroller as it rolled down the street, and hold her favorite toys from home. A decision to decline tracheostomy and pursue compassionate extubation with discharge home was agreed upon. Baby P was sent home with palliative care nursing available to manage symptoms, including opioid administration for respiratory discomfort and air hunger. She passed away at home 28 hours after hospital discharge.

Although child neurologists should have the practical skills to break bad news and respond to complex emotions, they should also know when to consult specialty palliative care teams for chronic and serious illnesses. Specialty palliative care can provide expertise in navigating complex medical decision making and can help simplify communication when there are multiple medical teams. Palliative care specialists can aid in decisions regarding life-prolonging treatments and withdrawal of care. Specialty palliative care can also provide valuable input in symptoms including managing secretions, pain relief, and balancing air hunger and respiratory depression.

### Addressing “Care for Rare”

#### CASE (continued). Baby P’s Family Considered Clinical Trials

Baby P’s family was informed that clinical research trials for Leigh disease are open yet limited by inavailability of specific therapies beyond vitamin supplementation. External clinicians with expertise in mitochondrial diseases confirmed no therapeutic strategies that could have altered Baby P’s clinical course had been overlooked and that there were no novel therapies that would be of benefit. Her parents felt comforted by the alliance with others with similar challenging circumstances. Baby P’s blood sample was collected 24 hours before discharge under an Institutional Review Board (IRB)-approved protocol for storage of deidentified clinical specimens in a biorepository for rare neurogenetic diseases.

Providing care to a child with a rare disease may be overwhelming to clinicians, who may feel uncertain of whether they exhausted therapeutic options when encountering a disease they have not treated before. It is especially complicated when there are no established standards of care or targeted therapies available. Contacting providers at different institutions who may have specialized in or researched this disease may be helpful in supporting the physician while deciding if enrollment in clinical trials would impact prognosis.

External connections can benefit the family members as well. Joining a support group can help parents and family members feel less isolated in their child’s treatment journey. It allows them to gain coping strategies, share experiences, and find comfort with people who have uncommon yet shared experiences. After the loss of a loved one, parents also may feel comfort in knowing that their loss may help others. When appropriate, discussion of potential ongoing research trials may help loved ones find meaning in their loss.

### Conclusion

Considering the nature of neurologic disease, child neurologists are challenged to support an entire family through
TABLE 3. ADDITIONAL EXAMPLES OF PEDIATRIC-SPECIFIC CONSIDERATIONS FOR PALLIATIVE CARE

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<th>Address children directly</th>
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<td>Clinicians often speak directly to parents or guardians who are the legal decision makers, although children and adolescents can weigh in about decisions about end-of-life care. Speaking directly with the patient at all ages can be beneficial, and adolescents especially value autonomous decision making. In addition, these conversations increase acceptance of decisions regarding withdrawal of care.</td>
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<th>Ensure adequate pain management</th>
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<td>The revised Face, Legs, Activity, Cry, Consolability (FLACC) scale has significant evidence supporting its validity and reliability for assessing pain in children with cognitive impairments and incorporates pain descriptors commonly exhibited in children with cognitive impairment, including verbal outbursts, tremors, increased spasticity, jerking movements, and respiratory changes. Pain medication dosing in pediatrics is age- and weight-dependent, and pain may be underreported considering developmental and/or disease stages. Recommendations should be offered where possible by involved clinicians and revised frequently as needed.</td>
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<th>Consider transition needs</th>
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<td>Adolescents may be approached for transition to adult providers depending on disease prognosis and capacity/capability of the adolescent (ie, need for guardianship). Presence or absence of puberty-associated physical changes may make this period additionally challenging. The recommended age for transitioning from pediatric to adult care is variable based on healthcare systems and should be tailored as much as possible to the individual child.</td>
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<th>Address family unit, including siblings</th>
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<td>Although a chronic and/or life-threatening disease may be challenging for the entire family, siblings who will likely also be at different developmental stages should be offered information congruent with their level of curiosity and understanding. Support from familiar teachers, school personnel, child life services in a hospital, and/or individual therapists should be introduced as needed. Respite may be needed to avoid caregiver burnout for parents of children requiring medically complex care.</td>
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<th>Limit prognostication</th>
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<td>Providing prognoses for children born with rare diseases about which there is little information or diagnosed with a serious disease at different timepoints during ongoing developmental maturation can be especially challenging. Resilience and adaptation to disease-related deficits make prognosticating about the future more difficult, and this uncertainty should be disclosed rather than ignored.</td>
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a complex disease process with unique pediatric-specific considerations (Table 3). Child neurologists must feel equipped to disclose difficult information empathetically and respond holistically to complex emotions. These skills improve communication and provide high-quality care for children with serious and potentially life-limiting conditions. It is our hope that child neurologists can use the tools discussed to navigate these conversations and experiences. Child neurologists should also be comfortable consulting specialty palliative care to help assess goals of care, complex medical decision making, and symptom burden in any child with a serious neurologic condition where prognostic awareness and medical decision making complicate care. These discussions should occur and continue at all stages of the child’s condition beginning at diagnosis.


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Disclosures
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JR and TSG have disclosures at www.practicalneurology.com