ChiPPS Pediatric Palliative Care Newsletter
Issue #13; November, 2008

Edited by Charles A. Corr, PhD, CT, Christy Torkildson, RN, PHN, MSN, and Mary Kay Tyler, RN, MSN, CNP

Issue Topic: Pain and Pain Management in Pediatric Palliative Care

Welcome to the thirteenth issue of the ChiPPS electronic newsletter. This issue of our e-newsletter (and the next issue that will follow) contains a collection of articles contributed by professional colleagues and family members that focus on pain and pain management in pediatric palliative care.

This newsletter is produced by ChiPPS (the Children’s Project on Palliative/Hospice Services), a program of the National Hospice and Palliative Care Organization and, in particular, by NHPCO’s Communications Work Group, co-chaired by Christy Torkildson and Mary Kay Tyler. Comments about the activities of ChiPPS, its Communications Work Group, or this issue of the newsletter are welcomed. We also encourage readers to suggest topics, contributors, and specific ideas for future issues of this newsletter. Please contact Christy at ctorkildson@georgemark.org or Mary Kay at mktyle@hospicewr.org.
It was ten years ago, at a meeting in Dallas that NHPCO and the ChiPPS workgroup began its work together. We celebrate those individuals that have been a part of ChiPPS.

Dr. Friedrichsdorf provides a brief yet comprehensive review of pain and pain management for children and includes several excellent resources that can be used for review and for staff education.

Dr. Faulkner reviews some of the most difficult and challenging pain and symptom management issues for pediatric patients providing an excellent review for all clinicians.

Ms. Fitzsimons, with the gracious permission of Miles Alpert Levin’s mother, Nancy Alpert Levin, again provides the raw emotional reminder that many of our hardest and most important lessons are learned from our patients and families.

Ms. Welsh not only illustrates the role of the child life specialist, often the missing link in pediatric palliative care, but also shares with us important lessons learned from her patients.

Dr. Toce provides a helpful summary and comments on a recent article from Pediatrics that addresses central topics in communicating with children and families.

ChiPPS customarily shares items that may be of interest to our readers.

Please note that the opinions expressed by the contributors to this issue are their own and do not necessarily reflect the views of the editors of this newsletter, ChiPPS and its Communication Work Group, or NHPCO. We invite readers with differing points of view to submit comments or suggestions for possible publication in a future issue.
Tenth Anniversary of the Children’s Project on Palliative/Hospice Services

We are especially pleased in this issue of the newsletter to recognize the 10-year anniversary of ChiPPS! A summary of some of the many ChiPPS activities over the past ten years is attached to this issue.

We owe special thanks to the original leaders of ChiPPS for their work, dedication, and efforts to provide constructive resources for all who are involved with pediatric palliative care, providers and family members alike.

Participants in the original ChiPPS Conclave in Dallas in 1998 included:

**Steering Committee Members:**
- Betty Davies, PhD, RN
- Kate Faulkner, MD, FAAP
- Ann Goldman, MB, FRCP
- Marcia Levetown, MD
- Stephen Liben, MD, FRCP, FAAP

**Meeting Facilitator:**
- Marcia Lattanzi-Licht, MA, RN, LPC

**Participants:**
- David Adams, MSW
- Ann Armstrong-Dailey
- Myra Bluebond-Langner, PhD
- Paul Brenner, MDiv
- Ira Byock, MD
- John Collins, MB, BS, FRCP
- Stephen Connor, PhD
- Charles Corr, PhD
- Tomasz Dangel, MD
- Brenda Eng, RN, MN
- Gerri Frager, MD, RN
- Gerri Haynes, RN
- Doris Howell, MD
- Anne Hunt, RGN, RSCN, MPhil
- Jeanne G. Lewandowski, MD
- Belinda Mitchell, MS, RN, FAAN
- Stacy Orloff, MSW, LCSW
- Danai Papadatou, PhD
- Maureen Pomeitio, MN, RN
- J. Donald Schumacher, PsyD
- Julie Simpson Sligh, RN
- Barbara M. Sourkes, PhD
- Cyndy Simpson Byrne Spitz, OT
- David Steinhorn, MD
- Michael Stevens, MB, BS, FRACP
- Liz Sumner, RN
- Suzanne Toce, MD
- Lisabeth Quesada Tristan, MD
- Christine Wandzura
- J. William Worden, PhD, ABPP

**Current members of the ChiPPS Advisory Council:**

**Co-Chairpersons:**
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- Suzanne Toce, MD
- Christy Torkildson, RN, PHN, MSN
- Mary Kay Tyler, RN, MSN, CNP

Thank you for all your efforts to advance pediatric palliative care!
This month, NHPCO’s Children’s Project on Palliative/Hospice Services (ChiPPS) is celebrating its 10-year anniversary. Over the past decade, this dedicated group of professionals has been committed to making the best-known practices in pediatric palliative care more widely available to care providers and increasing the availability of state-of-the-art services to families.

The work of ChiPPS began with a conclave meeting in November of 1998. This two-day meeting in Dallas, Texas involved 30 leaders from the field who worked to identify and reach consensus about the critical issues facing pediatric palliative care and develop strategies necessary to address those issues.

Since 1998, the accomplishments of ChiPPS have played a vital role in shaping the entire national pediatric palliative care and hospice field. Highlights of the group’s work include:

- Publishing three educational resources for healthcare providers: *Compendium of Pediatric Palliative Care; Pediatric Palliative Care Educational Curriculum;* and *Caring for Kids: How to Develop a Home-Based Support Program;*
- Helping NHPCO in the sponsorship of the first National Conference on Pediatric Palliative and Hospice Care in 2004;
- Coordinating the ChiPPS quarterly e-newsletter which features topical articles by family members as well as professionals;
- Serving as advisor to NHPCO’s consumer engagement initiative, Caring Connections, during development of its educational brochures for families of seriously ill children;
- Developing a national pediatric listserve for NHPCO’s National Council of Hospice and Palliative Professionals;
- Creating the first national standards of care for pediatric palliative care and hospice; and
- Establishing a ‘Pediatric Intensive’ for NHPCO’s 2008 Clinical Team Conference.

NHPCO offers heartfelt thanks to the many members of ChiPPS for their exceptional work to improve the lives of seriously ill children and families across the country.

The NHPCO Web site includes a section about ChiPPS, along with links to the resources noted here—and more. Visit: nhpco.org/pediatrics.
PAIN MANAGEMENT IN PEDIATRIC PALLIATIVE CARE

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Abstract
The majority of the more than 14,000 children dying from life-limiting diseases in the USA each year suffer from pain during their last weeks of life. Data suggest that applying the World Health Organization principles of pain management results in good pain relief for the majority of children with advanced cancer; however less has been reported on the effectiveness of the WHO approach for non-malignant pediatric life-limiting conditions. The management of children with intractable pain remains challenging and requires an interdisciplinary approach. State of the art pain management in the 21st century requires that pharmacological management must be combined with integrative, non-pharmacological therapies to manage a child's pain and suffering effectively.

Introduction
Only a few generations ago it was common in many families that infants and children died during their first years of life. Nowadays the death of a child is a rare and catastrophic incidence in industrialized countries and modern societies are often not prepared to deal with it. Palliative care for children and young people with life-limiting conditions is an active and total approach to care, embracing physical, emotional, social, and spiritual elements. It focuses on enhancement of quality of life for the child and support for the family, and includes management of distressing symptoms, provision of respite, and care through disease, death, and bereavement. Among the many domains of pediatric palliative care, the management of distressing symptoms, especially pain, is one of the most important – but can only be seen in the global picture of a holistic, multidisciplinary approach to the child, siblings, and parents, and cannot be limited to the application of drugs during the last days of life.

1. Broad-band analgesia
Robert Twycross [2] introduced the term “broad-band analgesia” to manage pain in palliative care. In the management of intractable pain in children it may be necessary to combine non-opioids, opioids, integrative therapies, adjuvant analgesia, and anesthetic or neurosurgical interventions. (Figure 1)
2. Pain Assessment
The majority of children dying from a life-limiting disease (both cancer and non-malignant diseases) experience pain during their last week of life. [3, 4, 5, 6] Regular pain assessment followed by appropriate analgesia is necessary to adequately relieve the suffering of these children. Using one-dimensional self-report measures (e.g., visual analogue scales with the anchor points 0=no pain, 10=worst possible pain or faces scale, Figure 2) [7, 8, 9, 10] provides easy pain assessment of alert and responsive children communicating with the caregiver or provider. For infants and children younger than 4 years of age several pain assessment tools have been validated, requiring independent observers recording the physical behaviors, as well as the frequency of their occurrence. [11] Behavioral observation measures to assess pain in cognitively impaired children are increasingly used. [12, 13]

As children may suffer from different pains, such as nociceptive, neuropathic, visceral, or spiritual, to name a few, a single pain rating may not be sufficient to assess the whole dimension of pain. A provider may have to become creative and more detailed-focused to evaluate the different pain aspects of the child. An example could be: “How would you rate your constant achy pain, and how would you rate the occasional shooting pain.” “Do you have pain anywhere else, in your heart or soul?”

3. Integrative Pain Management
State of the art pain management in the 21st century demands that pharmacological management is no longer the sole approach to the management of a child’s pain and suffering. [14] Integrative therapies, used on their own or together with pharmacology, include cognitive behavioral techniques (such as guided imagery, hypnosis, abdominal breathing, distraction) and physical methods (such as cuddle/hug, massage, Transcutaneous Electrical Nerve Stimulation [TENS], comfort positioning, heat, cold, aromatherapy). Children cope better with pain and other distressing symptoms when they understand what is happening and when they are encouraged fully in the process to attain relief from their pain. [15] Comprehensive pain control at the end of life requires tailoring to the needs of the individual child and integrating methods of pain management.
4. Pharmacological Pain Management

4.1. WHO-Principles

Data suggest that applying the World Health Organization (WHO) principles of pain management [16] results in good pain relief for the majority of children with advanced cancer; however less has been reported on the effectiveness of the WHO approach for non-malignant pediatric life-limiting conditions. In our experience, those four principles usually prove to be equally effective in managing children with non-malignant conditions:

4.1.1. “By the Analgesic Ladder”
The choice of analgesic drugs should be based on the WHO analgesic ladder (Figures 3 and 4). An assessment of pain severity dictates the choice of analgesic. Severe pain requires strong pain medication, i.e., opioids. A child with severe pain should not slowly step up the ladder commencing with acetaminophen, then later adding codeine before eventually changing to morphine. In this scenario WHO step III (strong opioids) should have been commenced immediately. There is considerable discussion in the field regarding the usefulness of a step 2, especially as codeine often proves to be a rather unfavorable choice (see below). Some authorities argue to discontinue step 2, using lower doses of “strong” opioids of step 3 instead.

4.1.2. “By the Clock”
Regular scheduling ensures a steady blood level, reducing the peaks and troughs of PRN (“as needed”) dosing. Commonly used opioid drug regimes include immediate release oral morphine every 4 hours or controlled-release morphine twice daily plus (for both strategies) 1/10-1/6 of the 24-hours morphine requirement as an hourly fast-release breakthrough pain medication as needed. (Table 1)

4.1.3. “By the appropriate route”
The least invasive route of administration, chosen by the child, has to be selected, making painful intramuscular injections of pain medication unnecessary and obsolete. Novel routes usually make use of high liphophilicity of certain opioids to cross skin or mucosa.

The oral route (or via nasogastric-tube/PEG-tube) is convenient, non-invasive, and usually preferred by the children and their care providers.

The sublingual application of opioids (morphine, fentanyl, oxycodone, hydromorphone, and methadone) appears safe and well liked by children and caregivers. In fact, this is our preferred route of pediatric opioid application, if oral administration is not feasible and there is no intravenous access (usually in children with nonmalignant conditions). The data for sublingual opioids are somewhat confusing, for morphine suggesting a bioavailability between 9% - 61%. [18, 19] Although morphine has hydrophilic properties, hence is not ideal for the sublingual route, the bioavailability of sublingual and oral administered morphine is interestingly not statistically different. [20, 21]

Oxycodone has a sublingual bioavailability of less than 20% and hydromorphone of 25%. [18] Methadone shows a good bioavailability via sublingual administration [22] and very rapid onset of relief of breakthrough pain in seven adult patients in a dose 2-8 mg. [23]
Case reports suggest good analgesia with sublingual liquid Fentanyl [24], and a commercial sublingual fentanyl application (Fentora™) is now available, demonstrating a bioavailability of 65%.

**Intranasal application** of opioids is pain free and safe. [17] Fentanyl can be diluted in normal saline solution (0.9%) and may be applied as a nasal spray or in drops. The pharmacokinetic profile of intranasal fentanyl seems to be similar to intravenous fentanyl. [25] Intranasal fentanyl does not irritate the nasal mucous membrane and has only minimal ciliotoxic properties. [26, 27] Reported intranasal fentanyl doses in children (1-1.5 mcg/kg) are equal to or only slightly above suggested intravenous doses [26, 28] (Table 1).

**Oral transmucosal fentanyl:** The fentanyl lozenge is a solid drug matrix with berry flavor providing oral transmucosal fentanyl citrate (OTFC). Due to fentanyl’s high lipophilicity, absorption across the oral mucosa directly into the systemic blood is rapid. OTFC has been used for children 3 years of age and above. Recent studies in opioid-naïve children showed typical opioid side effects of OTFC including respiratory depression. Some pediatric trials reported nausea and vomiting commonly, others rarely or not at all. Due to these adverse effects, the indications for OTFC have been changed. Currently, OTFC is indicated exclusively for the treatment of breakthrough pain in cancer patients and is no longer used for sedation or pre-medication. If used for this purpose, certain guidelines should be followed. [29] An FDA approval for late 2008 is expected for an oral adhesive disc technology (BEMA™ Fentanyl): A small, dissolvable, polymer film for application to the buccal membranes, with the smallest fentanyl dose of 200 mcg. Studies on adult subjects have shown a bioavailability of 70% (50% absorbed through mucosa), similar to sublingual fentanyl (Fentora™: 65% [48%]), higher than OTFC (Actiq™: 47% [22%]). [30]

**Transdermal fentanyl** patches are contraindicated for acute pain management due to a long onset time (it may take more than 60 hours to reach peak concentrations in children) [31, 32], inability to rapidly titrate drug delivery, and long elimination half-life (up to 24 hours). Patches can be applied on intact, healthy skin every (48-) 72 hours. They cannot be used for opioid naïve children – patients need to be on the equivalent of 30-60 mg oral morphine/24 hours to safely rotate to a fentanyl patch. The smallest patch delivers 12.5 mcg/hour. Sufficient immediate release breakthrough (rescue) opioid needs to be provided.

The Duragesic™ patch contains a selective semipermeable membrane with a fentanyl reservoir, hence it cannot be divided or cut as this would result in “dose dumping” with potential overdosing. However, a generic fentanyl patch (Mylan Pharmaceuticals) contains fentanyl in a different matrix system. Although the company clearly states “Do not cut or damage fentanyl transdermal system,” the matrix formulation makes dividing the patch theoretically possible, and pediatric experience suggests that cutting this generic matrix may be feasible. Transdermal fentanyl has its role in chronic, stable pain.

**Rectal application** is often unpopular and may deliver a wide variability in therapeutic blood levels through variable absorption, however experience shows good analgesia can be achieved in children when suppositories (or liquid opioids via a small catheter rectally) are administered.

**The intravenous administration** of opioid may be feasible, especially when there is a central line in place. Patient-controlled-analgesia/nurse-controlled-analgesia (PCA/NCA) pumps (e.g., morphine, fentanyl, hydromorphone, methadone) with a continuous background and an as-needed bolus often provide excellent pain management. Alternatively the opioid analgesics can be applied subcutaneously in the same dose as i.v. Many children and their parents we cared for were comfortable in terminal care with a s.c or i.v. PCA/NCA pump providing opioids for the management of pain and dyspnea in the home settings.
4.1.4. “With the Child”

The analgesic treatment should be individualized according to the child’s pain and response to treatment, frequently reassessed, and modified as required. Some children may require extremely high doses of opioids (sometimes more than 100 times the starting dose) to control severe pain. Adjuvant drugs (e.g., amitriptyline, gabapentin, low-dose ketamine, benzodiazepines, bisphosphonates) may be appropriate in the pain management of the individual child.

4.2. Non-Opioids

The most frequently used non-opioids are acetaminophen and ibuprofen (alternative via intravenous administration: ketorolac).

**Acetaminophen** (10-15 mg/kg PO/PR Q4-6h; dose limit: <2 years: 60mg/kg/day, >2 years: 90mg/kg/day) is generally well tolerated by children and lacks gastrointestinal and hematological side-effects. Significant hepatotoxicity [33] is rare, but careful attention to dosing is paramount.

**Ibuprofen** (10mg/kg PO TDS-QID; dose limit 2400mg/day) has the least gastrointestinal side effects among the NSAIDs. It should be used with caution with hepatic or renal impairment, history of GI bleeding or ulcers, and it may inhibit platelet aggregation.

**Ketorolac** has the advantage of IV administration, but should be rotated to oral ibuprofen, as soon as tolerated (< 2 years= 0.25mg/kg TID; > 2 years: 0.5 mg/kg q6h; max. 30 mg/dose; recommended dosing no longer than five days).

4.3 “Weak” Opioids

Codeine and tramadol are frequently used for mild-moderate pain and are so called “weak opioids” due to their ceiling effect (increasing above recommended dosing does increase adverse effects, but does not increase analgesia).

**Codeine** cannot be recommended in pediatric analgesia: Not only has codeine a variable bioavailability (15-80 %), but also produces its analgesic effect only through its metabolite morphine. [This pathway depends on the activity of the enzyme cytochrome P450 2D6 (CYP 2D6). Slow metabolizers (in white Caucasians 10%, in Chinese 30%) therefore do not achieve analgesia by codeine. On the other hand, around 5% of the general population have multiple copies of CYP 2D6 and are ultra rapid metabolizers [34], and therefore metabolize unusually high doses of morphine.] Individuals may vary widely in the rate at which they metabolize codeine. The author recently cared for a 10-year old girl who after receiving an appropriate dose of 1mg/kg codeine for post surgical pain displayed significant respiratory depression (rate 6/minute) and required several doses of the opioid-antagonist naloxone (with immediate response).
A better pediatric choice of a “weak” opioid is tramadol (Ultram™), which has been used in pediatrics since the 1970’s in Europe. It been trialed in neonates and children (mainly postoperative) and shown to be safe and effective. [35, 36] The analgesic strength of tramadol (a weak mu-receptor agonist - even weaker for delta and kappa) is augmented by an additional effect in inhibiting monoamine neurotransmitter (norepinephrine/serotonin) reuptake and it has a potency intermediate between codeine and morphine. [37] Although tramadol is metabolized by CYP 2D6 (and to a lesser degree by CYP 3A4) into the more potent O-desmethyltramadol, tramadol itself is a potent analgesic. For slow CYP 2D6 metabolizers the parent compound (tramadol) remains active, hence those individuals experience no decrease or only a slightly diminished effect on their analgesia. [38] Common adverse effects include nausea, vomiting, dizziness, constipation, and sedation. A rare, but severe side effect is the serotonergic syndrome. Tramadol appears not to increase the risk of ideopathic seizures; but patients with seizure tendency or medications that lower seizure threshold (tricyclic antidepressents, SSRI, MAOI, antipsychotics) may be at increased risk. Tramadol appears fairly safe regarding respiratory depression with overdose: No symptoms noted in children < 6 years who ingested 10/mg/kg or less, and in 87 adult patients with overdose only two demonstrated respiratory depression. In the USA tramadol is available in tablets only, however it’s easy to compound in a stable liquid, so both our inpatient and many outpatient pharmacies in our region now compound the liquid tramadol.

“Weak” and “strong” opioids should not be combined due to an unfavorable side effect profile.

4.4. “Strong” Opioids
The most frequently used opioid in pediatrics for moderate to severe pain remains morphine. Opioid-associated side effects (e.g., constipation, pruritus, nausea) have to be expected and treated accordingly. For recommended starting doses see Table 1. Morphine undergoes a strong first-pass metabolism (hence oral:intravenous conversion of 3:1), and is metabolized by liver glucuronyl transferase into Morphine-6 glucuronide (M6G) and Morphine-3 glucuronide (M3G). [M6G is a much stronger analgesic (x40-100) and displays adverse effects including nausea, vomiting, sedation, and respiratory depression. M3G is not an analgesic and rather a mu-antidote with unique adverse effects, especially hyperexcitability/neurotoxicity]. The ratio of M6G/M3G thereby defines in parts its analgesia to adverse effect profile in individuals. Both metabolites need to be excreted by the kidney, and children in kidney failure have a higher risk of unwanted side effects. Fentanyl or methadone, both not excreted renally, are likely to be a better choice in this scenario.

Opioids can be categorized into separate families: Phenanthrene derivatives (Morphine, Hydromorphone, Oxycodone, Hydrocodone), phenylpiperidine derivatives (Fentanyl, Meperidine), and diphenylheptane derivatives (Methadone, Propoxyphene). An opioid rotation may be necessary, if dose limiting opioid toxicity occurs. [39] This is necessary in about 10 per cent of the children provided with opioids by the “Pain & Palliative Care Team” at the Children’s Hospitals.
and Clinics of Minnesota in Minneapolis/St. Paul. An observation is that a switch from one opioid to another is often accompanied by change in the balance between analgesia and side effects. [40] A favorable change in opioid analgesia to side-effect profile will be experienced if there is less cross-tolerance at the opioid receptors mediating analgesia than at those mediating adverse effects. [41] Because of incomplete cross-tolerance, when changing between opioids with short duration of action, start new opioid at 50% of equianalgesic dose and titrate to effect. Even when a child may become unconscious during the last days of life due to the underlying disease (and not as an opioid toxicity), ceasing regular opioid analgesic drugs may provoke unpleasant withdrawal.

**Oxycodone** is a selective mu-opioid receptor agonist, although some animal studies suggest a kappa receptor agonist activity. [42] The oral potency of oral Oxycodone to Morphine is between 1:1 to 2:1. [43] One advantage of oxycodone over morphine is the slightly longer half-life, frequently allowing a Q6h dosing (as oppose to Q4h in morphine). Renal and hepatic impairment increases the oxycodone serum level. [44]

**Hydromorphone** is another selective mu-opioid receptor agonist. Unlike morphine metabolism, there is no hydromorphone-6-glucuronide (H6G), but similar to the morphine metabolism there is hydromorphone-3-glucuronide (H3G). Opioid hyperexcitability has been reported in patients with renal failure taking hydromorphone. [45, 46]

**Fentanyl** is a popular opioid for analgesia prior to painful procedures due to its rapid onset (about 1 minute) and its brief duration of action (30-45 minutes). It is also used in the pain management of children with cancer, for intra- and postoperative analgesia, in pediatric palliative care, and in sedation analgesia for ventilated children on the intensive care unit. Fentanyl provides a good alternative to morphine when dose-limiting side effects of the latter mandate a rotation of opioid drug. [47, 48, 49]

**Methadone** is an excellent opioid choice in pediatric palliative care and remains underutilized. It is a mu (delta, kappa)- opioid receptor agonist, a NMDA-receptor antagonist, and presynaptic blocker of serotonin and norepinephrine re-uptake.

Advantages include Methadone’s long half-life (allowing BID or TID dosing), high effectiveness in chronic pain relief as well as in the management of neuropathic pain, NMDA receptor antagonist mechanism (helps preventing tolerance), lower incidence of constipation, absent active metabolites, safe usage in renal failure and in stable liver disease, and its inexpensiveness. There are disadvantages though, including wide dosing variation, long half-life (may lead to accumulation; making quick titration difficult), and more complex equianalgesic conversion, which requires a much longer and closer patient observation than other opioids.

We are using an equianalgesic conversion chart (Table 2) when switching to oral (or sublingual) methadone in our pediatric patients. When switching from the oral to intravenous route of administration (either TID, or continuous infusion via a PCA-pump with additional boluses) we use 50-80% of the oral daily methadone dose.
4.5 Combination Analgesia
Fixed combination analgesia, usually acetaminophen plus an opioid should not be used in pediatric analgesia. Examples include Acetaminophen/Hydrocodone (e.g., Vicodine™), Acetaminophen/Oxycodone (e.g., Percocet™, Roxicet™) or Acetaminophen/Codeine (e.g., Tylenol No3™). The fixed ratio of acetaminophen to the opioid leaves dangerous choices: Either using suboptimal opioid doses or, when using adequate opioid doses, administering a liver-toxic dose of acetaminophen. Also it is unclear, if a child takes a scheduled combination formulation, what to choose for a rescue (breakthrough) dose – can we be certain that caregivers will not administer additional doses of the drug if their child remains in pain (and thereby grossly increasing the risk of an acetaminophen overdose)? Using the above combination analgesia inhibits titration to effect, due to acetaminophen toxicity. State of the art pediatric analgesia therefore requires the individual titration of stand-alone acetaminophen with a single opioid, the latter titrated to effect.

5. Obstacles
Many myths still remain and may be responsible for the inadequate pain management of many children in palliative care. Especially infants and very young children, as well as severely impaired children and teens, often do not receive sufficient analgesia, because their discomfort is different from that of adults. It is fallacious to believe, that children’s nervous systems are immature and therefore unable to experience, perceive, and remember pain. All available data suggest that those theories are wrong. [50] The application of an opioid to treat pain or dyspnea does not hasten a child’s death, if titrated by effect. The correct provision of opioids for symptom management not only improves the quality of life of a dying child significantly, but often prolongs the end-of-life period due to the improved quality of life.

6. Conclusions
Children in severe pain quite often need strong pain medication, i.e., morphine or other strong opioids. A dose limiting side effect may require an opioid rotation. Pediatric evidence and experience also supports novel routes of opioid application: transmucosal, transdermal, and intranasal opioid applications are well tolerated by children, effective, and safe. But neither transdermal nor transmucosal opioids must be used in opioid-naïve children and transdermal opioids are contraindicated in acute pediatric pain management. Providing a good pain management for a dying child usually requires a holistic, multidisciplinary approach and the knowledge to apply appropriate analgesic drugs in combination with integrative non-drug therapies.

Managing intractable pain in children at the end-of-life will usually require the integration of pharmacology (non-opioids plus opioids - following the WHO-principles) with non-
pharmacological, integrative therapies. (Figure 1). Not uncommonly children may require the addition of adjuvant analgesia or invasive approaches. Only if all six circles of Figure 1 have been exhausted, and not earlier, would it be necessary to consider sedation to unconsciousness, hence making the latter a very rarely needed intervention (estimated less than once per year in large pediatric palliative care programs).

Excellent pharmacologic and integrative, non pharmacologic management of pain at end of life is paramount and consistent with our goals of palliative care for the child and family, and will enhance their quality of life for the time remaining to them.

### References:

1. Association for Children with Life-threatening or Terminal Conditions and their Families and The Royal College of Paediatrics and Child Health: A Guide to the Development of Children's Palliative Care Services. 2nd Ed, ACT, Bristol (UK), 2003
3. Dangel T: Domowa opieka paliatywna nad dziecmi w Polsce. Department of Palliative Care – Institute for Mother and Child, Warsaw, Poland, 2001
16. World Health Organization: Cancer Pain Relief and Palliative Care in Children, 1999
43. Kalso E: How different is oxycodone from morphine? *Pain* 2007; 132(3):227-8
Table 1 Opioid analgesics: usual starting doses [11, 17]

<table>
<thead>
<tr>
<th>Drug (Route of administration)</th>
<th>Equianalgesic dose (parenteral)</th>
<th>Starting dose IV</th>
<th>IV:PO ratio</th>
<th>Starting dose PO (transdermal)</th>
<th>Starting dose controlled release</th>
</tr>
</thead>
<tbody>
<tr>
<td>Morphine (PO, SL, IV, SC, PR)</td>
<td>10 mg</td>
<td>Bolus dose: 50-100 mcg/kg every 2-4 h Continuous Infusion: 10-30 mcg/kg/h</td>
<td>1:3</td>
<td>0.15-0.3 mg/kg every 4 h</td>
<td>0.45-0.9 mg every 12 hours</td>
</tr>
<tr>
<td>Fentanyl (IV, SC, SL, transdermal, buccal)</td>
<td>100-250 mcg</td>
<td>Bolus dose: 1-3 mcg/kg (slowly over 3-5 minutes - fast bolus may cause thorax rigidity) Continuous Infusion: 1-2 mcg/kg/h</td>
<td>1:1 (IV to Transdermal)</td>
<td>12 mcg/h patch (must be on the equivalent of at least 30 mg oral morphine/24 hours, before switched to patch)</td>
<td>n/a</td>
</tr>
<tr>
<td>Hydromorphone (PO, SL, IV, SC, PR)</td>
<td>1.5 mg</td>
<td>Bolus dose: 15-20 mcg/kg every 4 h Continuous Infusion: 5 mcg/kg/h</td>
<td>1:5</td>
<td>60 mcg/kg every 3-4 h</td>
<td>180 mcg/kg every 12 h – currently not available in USA</td>
</tr>
<tr>
<td>Oxycodone (PO, SL, PR)</td>
<td>5-10 mg</td>
<td>n/a</td>
<td>n/a</td>
<td>0.1-0.2 mg/kg every 4-6 h</td>
<td>0.3-0.9 mg/kg every 12 h</td>
</tr>
<tr>
<td>Codeine (not recommended)</td>
<td>120 mg</td>
<td>n/a</td>
<td>n/a</td>
<td>0.5-1 mg/kg every 3-4 h</td>
<td>2-4 mg/kg every 12 hours</td>
</tr>
<tr>
<td>Tramadol (PO, PR)</td>
<td>100 mg</td>
<td>IV not available in USA [Bolus dose: 1 mg/kg every 3-4 h Continuous Infusion: 0.25 mcg/kg/h]</td>
<td>1:1</td>
<td>1-2 mg/kg every 3-4 h, max. of 8 mg/kg/day (&gt; 50kg; max. of 400 mg/day)</td>
<td>n/a</td>
</tr>
</tbody>
</table>

Calculated rescue (breakthrough) dose: 10-16 % of 24-hour opioid dose to be given every 1-2 hours as needed

IV = intravenous, PO = by mouth, SL = sublingual, SC = subcutaneous, PR = rectal, n/a = not applicable
### Table 2: Equianalgesic Methadone Chart

<table>
<thead>
<tr>
<th>Total Daily Oral Morphine Dose</th>
<th>Estimated Daily Oral Methadone Requirement</th>
<th>Source 1</th>
<th>Source 2</th>
<th>Source 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 100 mg</td>
<td>3:1</td>
<td>20% - 30%</td>
<td>33 %</td>
<td></td>
</tr>
<tr>
<td>101mg - 300mg</td>
<td>5:1</td>
<td>10% - 20%</td>
<td>20 %</td>
<td></td>
</tr>
<tr>
<td>301mg - 600mg</td>
<td>10:1</td>
<td>8% - 12%</td>
<td>10 %</td>
<td></td>
</tr>
<tr>
<td>601mg - 800mg</td>
<td>12:1</td>
<td>5% - 10%</td>
<td>8 %</td>
<td></td>
</tr>
<tr>
<td>801mg - 1000mg</td>
<td>15:1</td>
<td>5% - 10%</td>
<td>7 %</td>
<td></td>
</tr>
<tr>
<td>&gt; 1000mg</td>
<td>20:1</td>
<td>&lt; 5 %</td>
<td>5 %</td>
<td></td>
</tr>
</tbody>
</table>
Figure 1 Managing Children in Intractable Pain: Broad-Band Analgesia
(Blue circles: Standard approach; Yellow circles: Advanced management in selected cases)

- **Non-Opioids**
  - Acetaminophen
  - NSAIDs

- **Opioids**
  - Tramadol ("weak")
  - Morphine ("strong")

- **Adjuvants**
  - Anticonvulsants
  - Antidepressants
  - Antiemetics
  - Benzodiazepines
  - Bisphosphonates
  - Corticosteroids
  - Muscle relaxants
  - NMDA-receptor-channel blockers
  - Radiopharmaceuticals

- **WHO-Principles**
  - "By the clock"
  - "By the child"
  - "By the appropriate route"
  - "By the WHO ladder"

- **Integrative Therapies**
  - Massage
  - Heat/cold
  - Breathing
  - Biofeedback
  - Hypnosis

- **Invasive Approaches**
  - Regional anesthesia
  - Epidural or intrathecal
  - Neurolytic blocks
  - [Intraventricular opioids?]
  - [Percutaneous cervical cordotomy?]
Figure 2: Faces Pain Scale Revised (FPS-R) [7]

Please check the web site: www.painsourcebook.ca for correct administration and translations into many languages of these instructions.
Figure 3: The WHO three-step analgesic ladder [16]
Figure 4: The WHO three-step analgesic ladder with pediatric first-line drug suggestions

- Pain persisting or increasing
  - Opioid for mild to moderate pain
    - Tramadol
      - (Codeine not recommended)
    - Non-opioid
    - ± Adjuvant
  - Pain persisting or increasing
    - Opioid for moderate pain
      - Morphine
        - (Fentanyl, Hydromorphone, Oxydodec, Methadone)
      - ± Adjuvant
  - Pain persisting or increasing
    - Acetaminophen and/or ibuprofen
      - ± Adjuvant (IV: Ketalolac)
NERVE PAIN, SLEEP DISTURBANCES, AND THEIR TREATMENT

Kate Faulkner, MD
kathleenfaulkner@gmail.com

Neuropathic Pain

Neuropathic pain arises from injury, disease, or altered excitability of portions of the peripheral, central, or autonomic nervous system. It is one of the few types of pain that is not protective, since the painful sensation persists independent of ongoing tissue injury or inflammation. Common features of neuropathic pain conditions include sensory disturbances such as allodynia (feeling pain in response to non-painful stimuli, like stroking), cold hypersensitivity, paresthesias (tingling), and sensory deficits. There are sometimes motor findings, such as spasm, tremor, weakness, and atrophy. Possible autonomic abnormalities include cyanosis, redness, mottling, edema, and increased sweating. Neuropathic pain is frequently described by verbal children as having shock-like characteristics of burning, stabbing, “zinging,” or a lightening strike.

In palliative care and hospice, solid tumor invasion or compression of nerves and/or the spinal cord are common causes of neuropathic pain. Phantom limb pain is also a type of neuropathic pain that occurs less commonly now that pre-emptive analgesia is used before amputation. Neuropathic pain occurs in some of the congenital neuro-degenerative disorders, such as the Charcot-Marie-Tooth disorder.

An interdisciplinary approach works bests with neuropathic pain, as it is rarely possible to achieve resolution of the pain with any one therapy. Depending on the etiology of the pain, different types of approaches may be emphasized. Physical therapy and rehabilitation play a main role, and many children find cognitive-behavioral treatments helpful in decreasing pain, improving strength, and promoting functional improvement. Neurosurgical interventions, or nerve blocks, may be indicated for some problems. Acupuncture and hypnosis have been successfully used to treat chronic pediatric pain.

Medical management of neuropathic pain relies on two classes of drug therapy. In the first are membrane stabilizing agents, such as certain antiepileptic drugs (carbamazepine, phenytoin, valproate), steroids, and antiarrhythmics (lidocaine, mexiletine). Drugs that enhance dorsal horn inhibition, including antidepressants (amitriptyline, nortriptyline, paroxetine), GABA-agonists (baclofen), and other antiepileptic drugs (clonazepam, gabapentin) comprise the second class. The role of opioids in neuropathic pain remains controversial. Traditional teaching is that this type of pain does not respond as well to opioid therapy as does nociceptive pain, but opioids are clearly effective in the treatment of some adults and children with neuropathic pain syndromes. What remains unknown are the long-term side-effects and consequences of chronic opioid therapy.

Nortriptyline and gabapentin dose escalation schedules for neuropathic pain are as follows (Adapted with permission from Berde CB, Lebel AA, Olsson G. Neuropathic pain in children. In NL Schechter & CB Berde, eds. Pain in Infants, Children and Adolescents (2nd ed.). Philadelphia: Lippincott Williams & Wilkins, 2003, p.626.).

Nortriptyline dose escalation schedule:

A. For ambulatory patients

<table>
<thead>
<tr>
<th></th>
<th>&lt;50 kg</th>
<th>&gt;50 kg</th>
</tr>
</thead>
<tbody>
<tr>
<td>Day 1-4</td>
<td>0.2 mg/kg qhs</td>
<td>10 mg qhs</td>
</tr>
<tr>
<td>Day 5-8</td>
<td>0.4 mg/kg qhs</td>
<td>20 mg qhs</td>
</tr>
</tbody>
</table>
Increase as tolerated every 4-6 days until:
1. good analgesia is achieved
2. limiting side effects occur, or
3. dosing reaches 1 mg/kg/day (<50 kg) or 50 mg (>50 kg).

B. For inpatients or others with severe and uncontrolled pain, begin with the above doses but titrate upwards every 1-2 days.

Gabapentin dose escalation schedule:
A. For ambulatory patients

<table>
<thead>
<tr>
<th>Day</th>
<th>&lt;50 kg</th>
<th>&gt;50 kg</th>
</tr>
</thead>
<tbody>
<tr>
<td>Day 1</td>
<td>2 mg/kg qhs</td>
<td>100 mg qhs</td>
</tr>
<tr>
<td>Day 2</td>
<td>2 mg/kg tid</td>
<td>100 mg tid</td>
</tr>
<tr>
<td>Day 3</td>
<td>2 mg/kg am and midday,</td>
<td>4 mg/kg qhs</td>
</tr>
<tr>
<td>Day 4</td>
<td>4 mg/kg qhs</td>
<td>200 mg q hs</td>
</tr>
</tbody>
</table>

Continue to increase by 2 mg/kg (<50 kg) or 100 mg (>50 kg) each day, alternating the timing of the increased dose so that at least half the daily dose is at nighttime. Dose escalation should continue until:
1. good analgesia is achieved
2. side effects occur, or
3. dosing reaches 60 mg/kg.

B. For inpatients or others with severe and uncontrolled pain, a similar scheme is used with but triple the dose is given at each increment.

Neuroirritability

The incidence of pain in children with metabolic diseases and other types of neurodegenerative disorders remains unknown. Clinicians recognize that many of these children may present with or develop a syndrome of long-standing severe irritability or persistent crying and screaming. These symptoms may be particularly prominent in children with leukodystrophies and mitochondrial disorders. Identifiable etiologies for the irritability include pain from muscle spasm, joint involvement, and spasticity. Sources of neuropathic pain in this population include visceral nerve involvement with associated gastrointestinal dysmotility, and peripheral demyelination. However, often a treatable etiology remains unrecognized. A trial of gabapentin or anticonvulsant therapy is probably indicated in these situations, since there have been anecdotal reports of improvement.

Sleep Disturbances

More than 20% of a general pediatric practice concerns issues relating to sleep, so it is no surprise that sleep disorders come up frequently in palliative care. Sleep-related disorders have a profound impact on daily living for both children and their families. Lack of restful sleep can lead to daytime drowsiness and inattention, headaches, depression, and school or work problems for children and parents alike.

An understanding of normal developmental changes in children’s sleep patterns is helpful for practitioners and families. Newborns start out sleeping 16-20 hours/day, with one-two hour awake periods alternating with one-four hour sleep periods, around the clock. Between six weeks to three months, night differentiation develops, and by nine months about three-quarters of infants sleep through the night. Total sleep needs decline to 11-12 hours/day by school entry, and most children give up naps by age five. Most children sleep about 10 hours/day during the middle childhood years, and most adolescents should sleep nine hours. Common sleep disturbances in well and sick children alike include: increased latency (the time it takes to fall asleep), parasomnias (sleepwalking, night terrors, nightmares, and rhythmic movement disorders), and night awakenings in which children need parental intervention to re-establish sleep.
Anatomic prerequisites exist for the development of a normal circadian cycle. States of wakefulness are thought to be regulated by diencephalic and brainstem nuclei, whereas the establishment of circadian rhythms require the development of the suprachiasmatic nucleus of the hypothalamus and its connections. Therefore, children with midline brain maldevelopment are at high risk for sleep disorders. Some portions of the cerebral hemispheres also contribute to sleep-wake cycles, because children with hydronencephaly, lacking these but having an intact brainstem and cerebellum, also have profound sleep disturbances.

Sleep-related breathing disorders may also be associated with anatomic abnormalities. Craniofacial deformities, common in children with trisomy defects and myelomeningocele, can lead to nighttime obstructive apnea. The hypercapnic ventilatory and arousal response is also frequently blunted. Central and obstructive apneas are also common complications for children with myopathies and neuromuscular disease. In addition, such children face the real risk of ventilatory muscle fatigue, particularly in the latter hours of the night. This hypoventilation leads to arousal, daytime sleepiness, and headaches. Yet another anatomical source of sleep disorders is seizures. Sleep-related epilepsy accounts for 30% of seizure disorders in children. Frequent nocturnal seizures can fragment sleep and negatively affect daytime performance.

Establishment of circadian rhythm also benefits from children’s perceptions of environmental cues, known as “zeitgebers.” Disturbed sleep can arise from blindness or poor vision, therefore, because of children’s inability to distinguish light-darkness cycles. In general, children who are moderately or profoundly mentally challenged have difficulty in interpreting the social cues families use to promote healthy sleep cycles. Infants who are exposed to constant light, as in some neonatal intensive care units, can also suffer from lack of circadian rhythmicity.

A variety of other medical conditions frequently associated with neurological disorders may also hamper restful sleep. These include reflux, colic, hypoxia and pulmonary edema associated with cardiac disease, pain, muscle spasm, headaches, and movement disorders. In addition, many of the therapeutic drugs used in pediatric palliative care can disrupt normal sleep patterns. These include opioids, antiepileptic drugs, stimulating agents, and antiasthmatic medication. Hospitalization and episodic illness can also interfere with consistent sleep, because of disruption of normal routines.

Psychologic stressors that hinder sleep onset are common in palliative care. Children may have worries about their situation, fears of the dark compounded by fears of death, extreme separation anxiety, or a variety of other legitimate concerns that manifest by poor sleep patterns. They may also have negative associations with their bed if it is linked with stressors such as pain or procedures.

It should be clear that a complete history is always necessary, and a diagnostic workup sometimes necessary, to hope to successfully address sleep disorders in this group of children. The obstructive apnea of a child with trisomy may best be addressed via adenoidectomy; seizures with antiepileptic medication; headaches due to brain tumor growth with steroids; muscle fatigue with nighttime ventilatory assistance, and so forth.

A number of studies have documented the effectiveness of melatonin to reduce sleep latency in many children with developmental disorders. Administered in doses ranging from 2 mg. to 10 mg. two hours before bedtime, approximately three-quarters of children are able to fall asleep faster, and may stay asleep longer.

The use of hypnotics in children is less satisfactory. Two benzodiazepines (flurazepam and delorazepam), one antihistamine (niaprazine), and one phenothiazine (trimeprazine), have been shown to be effective in the short-term treatment of insomnia in children, but none are officially approved.
Tachyphylaxis precludes the long-term use of these medications. If short-term therapy is indicated, hospices have tended to use drugs that might already be in the home for other reasons. Therefore, diphenhydramine 1 mg./kg./dose p.o. at bedtime, with a repeat in an hour if necessary, or lorazepam 0.05 mg./kg./dose p.o. q. 4-8 h. are commonly used, as is chloral hydrate 5-15 mg./kg./dose p.o. q. 6-8 h.  Several complementary and alternative treatments show promise for insomnia, including herbal therapy with valerian, and aromatherapy with bitter orange essential oil. Anecdotal evidence from a palliative in-patient use promotes the use of lavender oils in a warm bath, and music therapy in selected cases.

The palliative care team would do well to teach and reinforce basic principles of sleep hygiene for children, since even ill children are likely to benefit. These include keeping a child busy and active during the day, and limiting naps after mid-afternoon. Children sleep better after exercise and after spending time outside each day. Even very sick children might benefit from having different places to spend the days and the nights. Children should not be hungry at bedtime, but are soothed more with a light snack than a full meal. A set bedtime should be established, along with a set bedtime routine. The hour before bedtime should be quiet time for shared pleasures, with television, computer games, and other stimulating activities restricted. A cool, quiet, comfortable bedroom will promote sleep. Children are often comforted by a dim nightlight, and a familiar transitional object such as a stuffed animal. Finally, restful sleep is promoted by arising at about the same time each day, both on school nights and weekends. Simple measures such as these may go a long way to increasing restful sleep for the whole family, particularly if the palliative care team keeps nighttime medications and other interventions to a bare minimum.
“RIDING THE PAIN ROLLER COASTER”: A MOM AND HER DYING SON’S REFLECTIONS ON HIS PAIN AND SYMPTOM MANAGEMENT

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With Excerpts from Miles Levin’s Blog, Levinstory on Carepages, Inc.
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“Pain is inevitable. Suffering is optional.”
Anonymous

Background

Pain and symptom management can mean different things to different people. Webster’s Dictionary defines pain as “physical or mental suffering caused by injury, disease, anxiety, grief, etc.” For pediatric patients, living and dying with their terminal illnesses is likely to involve all of these types of pain. One obvious goal in caring for these children and their families under palliative and hospice care is to help alleviate (or at least manage) their pain and suffering, regardless of the source (i.e., physical, mental, and/or emotional). However, to do this well, one must understand this pain from their point of view.

Pediatric patients and their families have always been our best teachers for gaining the insights we need to learn on how to better care for these patients and their families.

Miles Levin was diagnosed with Alveolar Rhabdomyosarcoma at 16. He succumbed to the disease two years later at age 18 on August 19, 2007, but not without leaving an incredible legacy behind. Throughout his two-year fight with cancer, he shared from the depths of his heart and soul what this journey was like for him on his CarePages.com space. His writings are raw, yet rich, with the wisdom Miles was gaining daily as he approached the end of his life. On days when Miles was unable to write, his Mom, Nancy, stepped in. Some of the excerpts from this insightful journey are included below. These specifically have been selected from all that Miles and Nancy wrote to shed some light on what his pain and suffering was like, and what his Mom experienced as she watched his daily struggles with the many types of pain that the cancer caused her son. These excerpts are not shared as a criticism in any way of the care that Miles received; but instead, as an illustration of how very present his pain and suffering was to him at times, and presumably, for many other pediatric patients also.

Some of the pain and symptom management themes which surface in these writings worth reflecting on include:

• Pain is so much broader than just the physical
• Pain management cannot be “cookie cutter” for peds patients as every child, every illness, every moment, every day brings new pain-related challenges and issues to work around to try and control; what’s important is that it is controlled and managed in the eyes of the child and their families
• Sometimes mental or emotional pain is caused (unnecessarily) by how some tests/procedures are explained or portrayed by the healthcare team (suggesting a review of these patient communication tools for pediatric patients and their families may be needed)
Identifying outside resources for pediatric patients like blogs, websites, online chat groups, etc. (which can be accessed while in the hospital/clinics), may provide an outlet, a release, which can’t be addressed by traditional pain management techniques.

Sometimes the actions in caring for these patients in pain speak louder than the words; the patients and their families are watching and taking cues from the healthcare team’s behavior; sending the right message in word AND action is important.

**Miles’ and Nancy’s Excerpts**

Note: Where the author is not noted, it’s Miles. Nancy, his Mom’s writings, are specifically identified as such.

“*My hair is starting to fall out now. I’m finding it very distressing.*” (7/5/05)

“We’re about to head out for the hospital to start another chemo round. I’m dreading these next few days because I’ve just started feeling really strong in the last couple of days and now the disintegration process starts all over again.” (8/2/05)

“The last round took a turn for the worse after day 2. We used to think there was a pattern of delay, peak and gradual subsiding of nausea. It seems now that there is no pattern.” (8/10/05)

“We got to the chapter on what to expect on the simulation process. Images of some strange apparatus resembling a cross between a garden hose and a medieval torture device told me this would not be fun. Nature has designed certain outlets of the body to be one-way streets, something these tubes were quite ready to defy. The video assured me that to any and all openings remotely near the pelvis, something bad was going to happen to them. I was horrified. Furthermore, it wasn’t quick either, the simulations took 1-2 hours. The video ended, leaving me in a dread that the inappropriately happy and upbeat credits music tried and failed—to assuage. Our nurse came in and asked if we had any questions. Did I ever. Question after question I stalled her, wildly devising an escape plan. I realized this was something that needed to be done and walked slowly into the hall like I was walking the plank of a pirate ship…” (8/22/05)

“My motto about the cancer experience is quickly becoming, “it’s always something.” Once you get over the nausea, your counts start to drop, then due to low counts you get sick. Once you are finally starting to feel better from this illness, you have chemo again.” (9/14/05)

“Miles has given me permission to do his update. He is emerging from the last round of in-patient chemo and getting ready for radiation…We had an experience in the hospital that in my view was powerful. Miles had completed the required six hour hydration period prior to receiving cytoxan. This process is necessary because the chemo drugs can burn the bladder. Yes. The nurse then hung the bag of chemo on the iv pole and a few drops of the chemo drug accidentally spilled on the floor. She immediately stepped back and threw several towels (which were later discarded) on the few drips. She then called someone from “Environmental Services” who arrived promptly, wearing large rubber gloves, and carrying a mop with strong detergent. What was striking about this picture was that no one flinched when the entire contents of the bag of this toxic substance was dripped into Mile’s body. One day, years from now, we will look back at these days of cancer treatment and view them as barbaric. Now that I’ve been up close and personal, I already do.” (Posted by Nancy, Miles’ Mom, 9/27/05)

“Travel will be challenging as Miles is not feeling well. As he referenced in his last update, he had outpatient chemo…Perhaps the level of toxicity reached a certain threshold, I don’t know, but Miles has not been able to tolerate it. In addition to vomiting, he has developed muscular pain, particularly in his hip. He cannot walk. Contrary to his usual stance, he has accepted some Tylenol. We are scouring the neighborhood for crutches. These symptoms can abate in a few days, or persist for many weeks. He has
more symptoms than I’m describing here, but I think you get the picture…” (Posted by Nancy, Miles’ Mom, 11/13/05)

“The bad news is that there is bad news. The cancer is pretty widespread now throughout my body and it’s growing rapidly. Time is of the essence.” (3/13/07)

“This is a very hard pill to swallow for all who care about me. Hear this, though; firstly, I won’t tell you that if there’s a will, there’s a way but I will tell you that if there’s a way, we’re going to do it. And secondly, it looks now more than ever that I may very well be vanquished in body, but know that I will not be vanquished in spirits.” (3/14/07)

“When told I’d relapsed, I thought to myself “Two years ago the waves swept me overboard. Why did I keep kicking my feet even as the currents carried me away?” Here I fought as valiantly as I possibly could; I continued to undergo chemotherapy even after the conventional protocol ended, stopping only when my body physically could tolerate no more. We did everything. What do I make of a world in which that’s not good enough?” (3/16/07)

“Why didn’t I call it a day two years ago and save myself endless vomiting and suffering? Because the only way to know if she will go to Prom with you is to ask her out. Our power is not infinite, and there are times when we fall short despite all our efforts, but the only way to find out is to find out. I have no regrets.” (3/16/07)

“It’s not much pleasure being in my body anymore, and I could stop treatment and let it all be over any time I wish, but these affirmations remind me that my remaining life is now about something bigger than that. I must keep on.” (3/26/07)

“I’m in rough seas now, with a cancer weight clasped around my ankle, pulling me down beneath the waves. I’ve come close to drowning a couple of times, but lately it looks like all this furious kicking might just being me back to the surface, at least for a while.” (4/6/07)

“The idea of jutting off to Baltimore, Nashville or even another hospital chasing a drug while Miles is in excruciating pain with the amount of required pain meds on the rise became obviously foolish…The boy needs relief. Plan is to zap it (i.e., radiate) the tumor in the rib, the primary culprit, to provide some palliative treatment. Chemo is also on the menu, but the recipe is still in the works. What isn’t uncertain is the cook.” (Posted by Nancy, Miles’ Mom, 4/8/07)

“When I need pain relief, I take more morphine. When I need emotional relief, I read the message boards.” (4/10/07)

“As my Doctor presented the option to discontinue treatment, I thought of you. I pledged that if it came to it, I would go down in magnificent flames for the principle glory of the thing. This treatment may result in nothing more than causing me additional unpleasantness for the last two or three weeks of my life, but my strife continues. I want to show everyone how it’s done. You don’t stop fighting until it’s over. Meanwhile, we must be realistic in our understanding that while this isn’t over yet, it looks to be soon. Thus my mantra has become…Keep fighting, but stop struggling.” (4/11/07)

“…The only thing more brutal than the cancer treatment is what cancer can and does do to the body and eventually the spirit. It erodes the person we know and replaces the personality with a shadow of himself. You can only imagine how excruciating it is to watch Miles waste away, all the while hoping and praying that your visions, your prayers, plus the chemo will reverse the course. As wonderful as you remember him, as wonderful as he is in his essence, he is going through trial by fire, the kind that makes most of us shudder and crumble.” (Posted by Nancy, Miles’ Mom, 4/13/07)
“Someone is really testing my resolve now. Last evening, I started having pain in both my shoulders, known born tumor sites. We started giving me morphine, but the pain continued to worsen anyway. Within an hour, it had spiraled out of control. My chest hurt so much that I didn’t feel I could take in a breath. More and more morphine only succeeded in making me sick and throwing up. By 11 o’clock we were in the emergency room and I was screaming like a pregnant lady. The IV dose of morphine barely did a thing. Eventually, around 2 am, I was given a narcotic called dilaudid, which got the pain under control. We got back to our room at 4 am and had to sleep upright because it hurt too much to lie flat.” (4/19/07)

“I made it. This past week of chemo was one of the longest, hardest, most miserable weeks of my life. I’ve come to know my body and it’s telling me it’s almost done with the poisonings. I’m not sure how many more rounds of chemo I can crank out…Yea, chemo is worse that I can convey. The feeling can’t really be put into any sort of terms that someone who hasn’t experienced it could understand…I would describe it as a distinct feeling of having been poisoned and every cell in your body saying get this stuff out of me!!! It’s a toxic feeling more than anything.” (5/7/07)

“Miles is dying in the same graceful, courageous, and honest way he has confronted every step of his illness. The hospice nurse told me that she has never seen a patient, of any age, with such openness and curiosity about the process, and she has been touched. He still looks beautiful, with his sculpted features, his long beautiful fingers, and his peaceful countenance. There is less and less of Miles, and more and more of just pure love.” (8/14/07, Posted by Nancy, Miles’ Mom)

“Some days are “better” than others; some hours are “better” than others---in terms of his comfort level. In terms of which world he is in, and in terms of his anguish and restlessness. He has made it clear; he wants out of this body. He’s done.” (8/14/07 Posted by Nancy, Miles’ Mom)

“…Miles’ earthly body has left us, early this morning. This is the day we have been dreading since June of ‘05…” (8/19/07, Posted by Nancy, Miles’ Mom)

“…There is relief that he is no longer in pain. With the exception of a handful of days, he was in varying degrees of pain for most of the last two years; he is free of that now.” (8/21/07, Posted by Nancy, Miles’ Mom)

“Suffering is the sandpaper of our life.
It does its work of shaping us.
Suffering is part of our training program for becoming wise.”

-By Ram Dass

Miles and Nancy and many other pediatric patients have been shaped by their experiences and we thank them for allowing us to enter their journeys and learn from them. May we become wiser from their stories so that we can better help with the pain and suffering of the pediatric patients and their families that we have yet to meet.

-###-
CHILD LIFE INTERVENTIONS WITH PAIN AT THE END OF LIFE

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Child life specialists work everyday to help children cope with stressful or painful situations in medical care. We blow bubbles, model deep breaths, help imagine a beautiful, peaceful place, and hold hands. Helping children and adolescents develop and access coping tools is, in fact, a key component of our profession. This particular job function is used often when working with children and teens who live with chronic or terminal medical conditions. I would like to share with you a story from my child life practice that illustrates some of the physiological and psychological distress I have encountered and how these children and teens have coped. Most importantly, I would like to share how much they have taught me.

I work with children, teens, and young adults who are HIV positive in an outpatient clinic. However, I frequently follow children, teens, or young adults as they are admitted to the inpatient unit in order to provide continuity of care. For this population of children, teens, or young adults, there is significance in being a long term presence in their lives. My role with children admitted to an inpatient unit is frequently focused on physical and psychosocial pain management. Many children, teens, and young adults with chronic illnesses experience total pain, where their pain has more than a physical manifestation. This type of pain can be seen in many patients who are approaching the end of their lives. Pain like this cannot be addressed solely through the use of pharmacological interventions, though interventions often include the use of medicine for physical pain management, as well as anxiety management. Additionally, pastoral care, social work, nursing, and physician care focused on psychosocial needs, and, of course, child life services can beautifully accompany this treatment.

My story is about a young man. Adam celebrated his eighteenth birthday in the hospital. He had spent the last few months in and out of the hospital due to his increasingly complex medical needs. He had multiple complications of his HIV and was very sick. Adam had previously spent long periods of time in the hospital. These long term hospitalizations caused a lot of distress for a young man attempting to create a life for himself outside of the hospital.

When he was inpatient (and during clinic visits) he expressed significant fear about being in pain. He would often say that he feared pain more than anything, even death. Due to his complex medical needs, he was often in a lot of pain, and as a result he was frequently without further pharmacologic pain management options. He and I worked together to find other methods that could work in conjunction with the pharmacologic pain medication he already used. Adam would often want someone to sit, hold his hand, and talk about the mundane details of life. Really, Adam needed something other than his pain on which to focus. He was a great conversationalist, asking questions about food, family, and pets. I was surprised time after time that even in his discomfort, Adam wanted to reach out to others and hear about their lives.

Adam also responded well to informal guided imagery, often imagining accomplishments beyond his physical capability. These imaginations served a number of purposes. He could create scenarios where he was not sick, had no physical limitations, and could accomplish anything. He would often tell me the bare bones of a situation and ask me to write him a “guided imagery” for him to practice. These scenarios also allowed him a great deal of control for at least one part of his life. Some of the most complex psychosocial problems with Adam stemmed from a lack of control of his body, his need for medicine, and
his family life. He also was able to be as involved or as passive as he felt like being at that moment. He could just lay there and listen, or start to create a new place to go, contribute his ideas, and change the ending. These imaginings were a place where he could express his strength of character. In the interactions I had with Adam and his family, there were many situations where pain was a factor. Sometimes Adam and I would do deep breaths to help him cope with an I.V. start. Sometimes Adam and I would imagine he was an NBA star, with lots of money, friends, and family always around him. Sometimes Adam would talk and cry and try to figure out why life was so unfair, and I would listen.

Another area of great strength for Adam was his spirituality. Collaborating with Pastoral Care was an essential part of Adam accessing these resources. He would often pray (and ask me and others to join in) before painful or frightening procedures. He also was able to have some level of peace about what became an inevitable death through his many conversations with us about his God. He sought, and found, a greater connection to people through these conversations.

When it became clear that Adam would die, we held a meeting with Adams’s father and members of our clinic and Intensive Care Unit staff. At this point, Adam was not conscious and he had designated his father as surrogate decision maker. We are lucky to have a collaborative interdisciplinary group in our outpatient clinic, and have a long standing relationship with Adam’s father. In that meeting, I was faced with a different type of pain. The pain Adam’s father felt, hearing that medical interventions would most likely be in vain, demanded a different skill set than I used with Adam. Adam’s father needed to have his grief be heard, he needed space to be angry and space to express the excruciating pain of losing a child. For Adam’s father I could be a sounding board, I could use my knowledge of development and bereavement and family systems to provide him with anticipatory guidance. Like many professionals, Child Life Specialists sometimes just need to be the silent witness to a family’s pain. We (and other professionals) need to know that we cannot, and should not, fix everything. There are times when living and practicing in that place can be difficult for us. We have to be able to access support ourselves, in order to remain connected and available to families.

After this discussion Adam’s father decided to remove all artificial methods of keeping Adam alive. It was too painful for him to stay to watch this happen, and so he went home. Our clinic medical director, the nurse practitioner who had taken care of Adam his whole life, a Pediatric Intensive Care (PICU) nurse, Chaplain, and I all stood around Adam’s bed as he was extubated and his medications were stopped. The Chaplain read the 23rd Psalm with us, we each took turns saying out loud our favorite memories of Adam, and eventually went home to our families. Adam was still alive! The next morning Adam’s father returned. Within minutes of his arrival, Adam died. “He waited for me” was the only response his father was able to communicate. Wonderfully, the PICU child life specialist was with him in this moment, and later walked with Adam’s father back out to his truck, clutching the blanket from Adam’s bed.

A few months later, some members of my outpatient clinic and I brought items to Adam’s family’s house. We had finished a scrapbook that Adam had begun. There were pages and pages of photographs of Adam at camp, at clinic, and with his family. We brought hand molds that I made of Adam’s hands in the days before his death. We brought a teddy bear that had lived on Adam’s bed during his last hospitalization. His father was glad to see us, and glad to have these items. Mostly, though, I think he was glad to have the chance to talk about Adam, and to know that Adam was not forgotten. I remember moments of Adam’s life where he was in less pain, when he laughed a lot, and flirted with the girls at camp, and sometimes I get to share those memories with others.

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Readers’ Corner


Abstract: Health care communication is a skill that is critical to safe and effective medical practice; it can and must be taught. Communication skill influences patient disclosure, treatment adherence and outcome, adaptation to illness, and bereavement. This article provides a review of the evidence regarding clinical communication in the pediatric setting, covering the spectrum from outpatient primary care consultation to death notification, and provides practical suggestions to improve communication with patients and families, enabling more effective, efficient, and empathic pediatric health care.

Who is the audience for this information? Originally intended for pediatricians, this article is pertinent for all members of the pediatric palliative care team.

What is special about this article? No new and startling information is presented in this article. It is, however, very well organized and user friendly. I particularly like the suggestions for delivering bad news with skill and empathy, usable strategies in the hospital, and suggestions for communicating in a way that is perceived by families in the way we intend. (The samples of the perceived message in response to the health care provider’s traditional communication are quite humbling.)

Where and how can I apply this information? As communication is a skill that can be taught and learned, these techniques can be applied in any setting: home, hospital, clinic, care facility, emergency room, etc.

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Items of Interest:
In each issue of our ChiPPS e-newsletter, we offer additional items of interest.

1. **Reader's Corner.** In this issue, we inaugurate a new, occasional feature (see page 32). The Reader's Corner will provide brief summaries and bibliographical information about journal articles and other publications that are important and likely to be of widespread interest to individuals who are involved or interested in pediatric palliative care, but that may not be known to all readers of this newsletter. Contributions to the Reader's Corner will include an abstract of the publication, a description of the audience for this information, comments on what is special about the publication, and suggestions as to where and how this information can be applied. We would welcome suggestions for publications to include in our Reader's Corner and/or summaries and comments on such articles following the model in this issue. Please send all such suggestions to Mary Kay Tyler at mktyler@hospicewr.org.

2. **Subjects and Contributors for Future Issues of This Newsletter.** In past issues of this newsletter, we have addressed a wide range of subjects, including bereavement, sibling bereavement, self care, ill and dying teens, perinatal loss, neonatal loss, spirituality, and unsung heroes in the circle of care. In this issue and in the one that will follow, we are examining issues related to pain and symptom management for children with life-threatening illnesses and their families. For future issues, we are considering subjects that focus on overcoming barriers to pediatric palliative care, making memories or legacy building, and how to prepare families to take a child home and to cope with a child who dies at home. If you know of good topics and/or contributors (including yourself) for any of these and/or other future issues of this newsletter, please do not be shy! Step right up and contact any of the following: Christy Torkildson at torkc@sbcglobal.net; Mary Kay Tyler at mktyler@hospicewr.org; or Chuck Corr at charlescorr@mindspring.com. We will work with you!

3. **NHPCO's Audio Web Seminar.** NHPCO is sponsoring an Audio Web Seminar, *Providing Comfort to Seriously Ill Children: Pediatric Pain Management*, on December 11, 2:00 – 3:30 pm ET. This seminar provides a valuable learning opportunity that will help staff develop new insights, strategies and practices to ensure the delivery of quality pediatric hospice and palliative care. NHPCO's Audio Web Seminars are brought to you, live, in your location via phone line and internet access. Participants dial in to listen to presenters and view PowerPoint presentations in real time on their computers. You can participate in interactive features and submit questions electronically. Participation requires only a phone (for audio) and computer with internet connection. To learn more and register, go to www.nhpco.org/conferences and click on the link for Audio Web Seminars.

4. **Online Courses in Collaboration with the National Center for Death Education.** NHPCO is proud to collaborate with the NCDE at Mt. Ida College in Newton, MA to provide online educational programs for hospice and palliative care professionals and volunteers. The next scheduled course, *Pain Management for Children in Hospice and Palliative Care*, runs from November 17 to December 19, 2008 (no class Thanksgiving week) with faculty Susan M. Huff, RN, MSN. For more information about NCDE, visit www.mountida.edu/ncde or contact NCDE at 617-928-4649.

5. **NHPCO and ChiPPS Pediatric Listserv.** NHPCO and ChiPPS have created a special pediatric listserv for NCHPP or eNCHPP members who provide services for children with life-threatening conditions and their families. Read more about the listserv and eligibility requirements to participate - and sign up online.

7. Support Partnering for Children/Wear a Bracelet. Partnering for Children is a national awareness campaign that was launched November 2007. The goal of Partnering for Children is to help get the word out about compassionate, family-centered healthcare for children with life-threatening conditions. The ChiPPS work group and the resources ChiPPS makes available is an important part of this campaign. In the memory of the many children whose wisdom and courage inspire us, inspirational bracelets developed by Children’s Hospice and Palliative Care Coalition are now available through Partnering for Children. These inspirational bracelets which bear poignant messages from children can be ordered directly online at partneringforchildren.org or by calling 800/646-6460. One hundred percent of the net proceeds of these bracelets go directly to improving care and quality of life for children with life-threatening conditions.

For more information on the Partnering for Children, including how to join as a campaign partner, please visit www.partneringforchildren.org.

8. Calendar of Events. As a reminder, there is a calendar of pediatric educational opportunities on the ChiPPS section of the Web site, www.nhpco.org/pediatrics. ChiPPS is a program of the National Hospice and Palliative Care Organization. Learn more at www.nhpco.org/pediatrics. Please e-mail Christy Torkildson at torkc@sbcglobal.net to have your pediatric palliative care educational offering listed.

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Thank you for taking time to read this issue and for any feedback that you can offer us. Providing pediatric palliative and hospice care to children, adolescents, and their family members has made great strides in recent years, even though it is certainly not always easy and still faces many challenges and obstacles. We wish you all the best in your good work. If you are not on our mailing list and received this newsletter from a friend or some other source, please send an email message to CHIPPS2@NHPCO.org requesting to be added to our mailing list. If you are a member of NHPCO, you can go to the Communications Preferences tab in your individual member record online and “opt-in” for communications from ChiPPS. Member Services will be happy to help you adjust your communications preferences; contact them at 800/646-6460.

Visit the ChiPPS Web page at www.nhpco.org/pediatrics for further materials and resources of interest.

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