



medically. This article will discuss the advantages and disadvantages of using PN as a treatment in children receiving comprehensive palliative care.

### **The Palliative Care Approach**

True palliative care is not just for children who are dying. It is not about death at all; it is about maximizing the quality of life of children with complex and life-threatening illnesses, from the moment of diagnosis all the way through death. (For more information on this subject, see this previous article describing palliative care: <http://articles.complexchild.com/july2010/00221.html>.)

The World Health Organization defines Palliative Care in the following manner:<sup>2</sup>

- Palliative care for children is the active total care of the child's body, mind and spirit, and also involves giving support to the family.
- It begins when illness is diagnosed, and continues regardless of whether or not a child receives treatment directed at the disease.
- Health providers must evaluate and alleviate a child's physical, psychological, and social distress.
- Effective palliative care requires a broad multidisciplinary approach that includes the family and makes use of available community resources; it can be successfully implemented even if resources are limited.
- It can be provided in tertiary care facilities, in community health centres and even in children's homes.

The most important goal of palliative care is to provide the child with the greatest quality of life achievable, including aggressive treatment of pain.

### **Parenteral Nutrition for Gastrointestinal Pain**

Where does parenteral nutrition fit into this definition of palliative care? It is a known fact that many children with life-limiting conditions or other complex medical issues experience severe gastrointestinal pain related to feeding. In many children, the pain associated with feeds may have a profound effect on their quality of life, causing them to vomit frequently, and in some cases, preventing them from engaging in any other activities due to nausea, pain or vomiting.

These children may or may not have anatomic reasons for this pain. In some children, the pain may be related to poor motility, possibly secondary to narcotic usage. Many children with metabolic disorders and other severe neurological impairments experience chronic intestinal pseudo-obstruction, in which any portion of the gut may have weak, uncoordinated, or absent contractions, causing it to act as if it is physically obstructed. In other children, the problem may be more related to visceral hyperalgesia or hypersensitivity to pain, in which the body perceives anything in the gut as a toxin and responds with pain and vomiting. Some children may also have anatomic malformations,

inflammatory conditions, or other secondary disorders that create additional visceral pain in the gut.

While chronic gut pain is not uncommon and may occur in a wide variety of disorders, it can be unbelievably severe and unrelenting in some children with multi-system disorders. In this case, the child's quality of life is greatly impacted, and adequate nutrition cannot be achieved with oral or tube feeds. It is appropriate to consider using parenteral nutrition in these children as part of an overall palliative care plan.

### **Candidates for PN as a Palliative Care Treatment**

Parenteral nutrition is not without risks, including liver failure, severe infections, and other potentially fatal conditions. In addition, it requires careful preparation, monitoring, and oversight, which may not be possible for some families. As such, it should only be used in children with no other viable option for feeding, or children with severe chronic gastrointestinal pain.

In general, there are two groups of children who should be considered for palliative PN. These include:

- Children with severe life-threatening multi-system disorders with documented severe gastrointestinal dysfunction who have failed tube feeds.
- Children with severe life-threatening multi-system disorders with severe neuropathic or visceral pain that occurs with feeding and continues despite aggressive pain management.

The first group of children typically includes children with chronic intestinal pseudo-obstruction, extremely poor motility of the stomach and small intestine, severe issues with malabsorption, or anatomical/post-surgical malformations making the gut non-functional. In these children, feeding trials have failed, due to feeds moving backwards in the GI tract, not moving at all, not being absorbed, or severe vomiting and/or diarrhea. The second group of children typically includes children who experience incredibly intense pain with feeds, even while on narcotic pain medication. In most cases, children who are candidates for palliative PN experience both significant GI dysfunction and severe pain.

Before beginning PN, it is important to try all other possible alternatives. These include:

- Ruling out treatable other conditions, such as a physical obstruction, eosinophilic disease, inflammatory bowel disease, bacterial overgrowth, cancer, etc.
- Trialing continuous elemental formula feeds into the small intestine, either by NJ tube, GJ tube or a surgically placed J tube. Feeds should be considered failed if the child is unable to take enough to remain hydrated, or experiences extreme losses (vomiting, diarrhea, gastric drainage, ostomy output) that prevent hydration.

- Aggressively managing constipation, both medically or surgically, using a colostomy, ileostomy, or ACE (antegrade continence enema) procedure.
- Trialing motility medication, such as reglan (if not contraindicated), an antibiotic such as erythromycin, or a compassionate use medication such as cisapride.
- Trialing pain medication for visceral/neuropathic pain. These medications may include tricyclic or other types of antidepressants (such as Elavil or Cymbalta), anti-seizure medications such as Neurontin or Lyrica, and other medications, including Periactin. Note that these may take a month or longer to reach full effect.
- Trialing anti-nausea medications, including Zofran, Kytril, Benadryl, etc.

If a child with multi-system disease continues to experience severe GI pain and dysfunction despite attempting the aforementioned alternatives, palliative PN should be considered.

### **Ethical Considerations**

Managing PN in the home requires a family that is willing to put in the time, effort, dedication, and commitment to doing it safely and correctly. If a child does not come from such a family, it is prudent to try to establish private duty nursing care for the child. Without a committed family or consistent nursing care, PN cannot be administered safely and is likely to cause more problems than it solves.

Some doctors, health systems, or national health programs do not believe palliative PN is an option for children with severe multi-system disorders, especially when these disorders are neurological in nature. These individuals or systems believe that long term PN prolongs life in children with severe disease who otherwise would not survive. The primary argument in this situation is that the difficulties in providing PN in the home, the financial cost, as well as the potential burden of side effects such as liver disease and sepsis, overwhelmingly negate the positive impact of PN in these children.

This system of thought is based on somewhat antiquated information. In the past, home PN was difficult to administer due to a lack of portable equipment, home infusion providers, and other factors. Currently, portable infusion pumps, supplies, and expert home infusion providers are widely available, making this no longer an issue. In addition, the expertise in creating an appropriate PN recipe, as well as preventing and treating side effects such as liver disease, has increased dramatically over the past twenty years, meaning PN is much safer than it was in the past. Finally, methods of preventing infection have improved dramatically in recent years, again increasing safety.

The financial cost of PN does remain high. It is not, however, so high that it precludes use for any child who may regain or improve his or her quality of life on it. It also remains difficult to arrange appropriate nursing care or parental support for many of these children. Changes in health insurance benefits may be necessary to overcome this barrier.

## Palliative PN

Over the past few years, children with life-threatening multi-system disorders have begun receiving PN as part of an overall strategy for improving their pain and quality of life. Many children have thrived on PN, with minimal side effects and a vast improvement in pain. Many children have also found that the nutritional benefits of being on PN have dramatically increased their health and energy levels, improving their quality of life and reducing their disease process even more. Unfortunately, few studies have been done on the quality of life of children on PN, and none on children with multi-system disorders who are candidates for palliative PN.<sup>3</sup> These sorts of studies are absolutely essential to help determine which children are good candidates likely to experience significant improvement in their quality of life from PN.

PN is definitely not for all children, however. Some children do experience severe infections, including fatal episodes of sepsis, liver complications, and other serious side effects. Other children may continue to have severe pain even on PN. The risks and benefits must be carefully and individually weighed for each child. The potential for improved quality of life is enormous, but the potential for severe side effects or fatal infection is equally possible. Palliative PN must be reserved for children with no other alternatives who experience a great increase in health and quality of life as a direct result of the PN.

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<sup>1</sup> See <http://www.merckmanuals.com/professional/sec01/ch003/ch003c.html#sec01-ch003-ch003c-210>

<sup>2</sup> <http://www.who.int/cancer/palliative/definition/en/>

<sup>3</sup> See, for example, Emedo MJ, Godfrey EI, Hill SM. A qualitative study of the quality of life of children receiving intravenous nutrition at home. *J Pediatr Gastroenterol Nutr.* 2010;50(4):431-40.