Nutritional Management of Infants with Short Bowel Syndrome

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Executive Summary

Short Bowel Syndrome (SBS) is a clinically significant condition affecting the NICU population. The morbidity and mortality of SBS is most often the result of prolonged dependency on parenteral nutrition. Nutritional management of these patients is complex and studies indicate that clinical outcomes are better among infants with SBS when enteral feedings are initiated sooner rather than later.

Primary Children’s Medical Center (PCMC) in Salt Lake City, UT serves neonatal patients with SBS. Currently there is no guideline in place to manage the complex nutritional needs of these patients. The purpose of this scholarly project was to develop a guideline to manage both parenteral and enteral nutrition in patients with SBS. The objectives for this project included: (1) developing a clinical practice guideline, (2) using the most current evidence and eliciting input from content experts during development of the guideline, (3) incorporation of the guideline into clinical practice, and (4) dissemination of the guideline to a wider audience of professional peers and colleagues.

A clinical practice guideline was developed to serve as a roadmap for neonatal practitioners who manage the complex needs of infants with SBS. Recommendations contained in the guideline were derived from the research literature in addition to input from content experts. The guideline provides management strategies for the three phases of nutritional management. The first phase occurs just after bowel resection when fluid and electrolyte disturbances occur. Recommendations for macronutrients and micronutrients to be included or excluded in the parenteral nutrition are contained in the guideline. Total fluid and caloric goals are also provided. During the second phase, infants with SBS are introduced to enteral feedings. Recommendations are made on when to initiate feeds, the type of feeding to provide, and when to advance or hold enteral feedings. The final phase occurs as the patient is weaned from parenteral nutrition. Recommendations for enteral caloric goals, laboratory monitoring, and expected complications are included. In addition to previously noted recommendations for nutritional management, the guideline also provides information on important laboratory studies to monitor, classes of medications to treat SBS, and symptoms of feeding intolerance.

The guideline has received approval from the content experts. Presentation of the guideline to the Division of Neonatology at PCMC is tentatively scheduled for June 2013. A research study in collaboration with the content experts to evaluate the effectiveness of the guideline has been proposed. Outcomes measures of interest that will be examined include length of hospitalization, time to enteral feeds, growth, length of dependency on parenteral nutrition, and complications. Upon completion of the study, a manuscript will be written for submission to a peer reviewed journal.

The faculty chair of the scholarly project committee is Pamela Phares, PhD, APRN-BC. The program director is Julienne Schiefelbein, DNP, NNP-BC. Committee members also serving as content experts include Daniel Malleske, MD, MS, FAAP, and Cecilia Mulroy, RD, CD, CNSP.
Problem Statement

Short bowel syndrome (SBS) is a form of intestinal failure and a significant contributor to morbidity and mortality in newborns, accounting for approximately 2% of newborn intensive care unit (NICU) admissions (Wales et al., 2004). One of the greatest challenges facing the NICU healthcare team is the nutritional management of patients with SBS. A fine balance exists between parenteral nutrition (PN), enteral feedings, and electrolyte supplementation. Prolonged parenteral nutrition places the neonate at increased risk for complications including catheter associated sepsis, venous thrombosis, and intestinal failure associated liver disease (IFALD). Early initiation of enteral feedings has been shown to minimize these risks (Theyskens & Dams, 2008).

The NICU at Primary Children’s Medical Center in Salt Lake City, Utah serves patients with SBS. Currently there is no clinical guideline or protocol in place for nutritional management of infants with SBS. Several problems exist in managing the nutrition of these patients. These problems include determining: (1) the ideal composition of PN to minimize the risk of IFALD, (2) timing for initiating enteral feedings, (3) which type of feeding should be used, (4) how quickly feedings should be advanced, and (5) which labs need to be monitored and with what frequency. Addressing these clinical problems in the form of an evidence based clinical practice guideline will serve to promote consistency in management of this population across care providers with the ultimate goal of improving patient outcomes.

Clinical Significance

Short bowel syndrome (SBS) is a serious clinical condition affecting 24.5 per 100,000 live births, of which preterm infants are at highest risk (Wales et al., 2004). The incidence of SBS is approximately 100 times greater for preterm infants than for those born at term.
While total incidence of SBS is low, the associated mortality for infants with SBS is high. In a cohort study conducted by Wales et al. (2005), 37.5% of patients who participated in the study died. Infant morbidity is most strongly associated with complications from parenteral nutrition dependency. Other complications of SBS include overgrowth of bacteria, liver disease due to prolonged PN use, diarrhea, lactic acidosis, increased gastric acid secretion, gall stones, kidney stones, sepsis, and nutritional deficiency (Harris, 2007).

Delivering optimal nutrition to neonates with SBS is a complex issue requiring careful management of parental nutrition, enteral feedings, and supplementation of electrolytes and vitamins. There is currently no formalized protocol to guide clinicians in the management of nutrition for infants with SBS at Primary Children’s Medical Center. Due to the complex nature of this problem, nutritional management of these infants would most effectively be addressed using a clinical practice guideline.

The purpose of this project is to develop a clinical practice guideline to address the management of nutrition in neonates with SBS for implementation at Primary Children’s Medical Center. It will be a guide for practitioners to standardize care based upon the most current evidence. Recommendations will be made on composition of parenteral nutrition, optimal timing for enteral feedings, when to advance (or hold) feedings, and the appropriate clinical monitoring of infants’ nutritional status during this period. The goal of this project is to promote consistent evidence-based nutritional management thereby improving clinical outcomes for infants with SBS.
Objectives for the DNP Scholarly Project

- Develop a clinical practice guideline to promote nutritional management of neonates with short bowel syndrome, the intent of which is to standardize the nutritional management of neonates with SBS across healthcare providers.
- The proposed guideline will reflect the most current evidence and expert opinion in the field of neonatal nutrition for children with SBS.
- Present the guideline to the Division of Neonatology at Primary Children’s Medical Center for eventual approval to incorporate into the Division's guidelines and protocols for practice.
- Prepare a manuscript for submission to an appropriate peer reviewed journal for dissemination to a broader audience of neonatal practitioners.

Literature Search Strategy

PubMed, Medline, and CINAHL were used for the literature search. The initial search included the terms neonates and short bowel syndrome, and nutritional management. In a second search, additional search terms including incidence, electrolytes, elemental formula, and parenteral nutrition were added. Bibliographies of relevant articles were used to find additional articles.

Review of Literature

Definition

Short bowel syndrome (SBS) is best described as a malabsorptive condition most often due to resection of a large amount of intestine (Wales & Christison-Lagay, 2010). There is no consistent definition of SBS in the literature and substantial variation exists among institutions (Harris, 2007). Wales et. al (2004) defined SBS as post resection bowel length less than 25% of
expected length for gestational age or dependence upon total parenteral nutrition (TPN) for
greater than 42 days post-surgical resection. Understanding the pathophysiology of SBS provides
the clinician with important clues on how to proceed with the nutritional management of these
patients.

**Etiology and Epidemiology**

During the neonatal period, SBS may result from both congenital and acquired diseases.
Prior to 2010, very little epidemiologic data existed to define the extent of the problem. Wales
and Christison-Lagay (2010) conducted a retrospective cohort study in Canada to better describe
the etiology and epidemiology of SBS. Several important findings emerged from their study
including identification of the most common diseases associated with SBS, the projected
incidence of SBS in the neonatal population, and associated mortality rates. Congenital
conditions linked to SBS included intestinal atresia, volvulus, abdominal wall defects, and
Hirschprung’s disease. The leading cause of SBS during the neonatal period was identified as
necrotizing enterocolitis (NEC), accounting for 30% of cases in their study. The incidence of
SBS was 24.5 per 100,000 live births with a mortality rate of 37.5%.

**Pathophysiology**

The pathophysiology and clinical symptoms of SBS depend upon the portion of bowel
that is resected, gestational age, functionality of the remaining bowel, presence of the colon, and
underlying disease processes (Wales & Christison-Lagay, 2010). Short bowel syndrome is
divided into three distinct subtypes based upon the portion of bowel removed and the remaining
bowel anatomy. The first subtype involves resection of the small intestine with preservation of a
portion of the ilium, anastomosis of the remaining small bowel, and an intact colon. The second
subtype includes resection of small bowel and a portion of the colon with enterocolonic
anastomosis. This second subset is the most common form of SBS in infants with NEC. The third subset, and most difficult to manage, is small bowel resection with a high output jejunostomy.

The location of the intestinal resection affects nutrient, vitamin, mineral, and fluid absorption (Harris, 2007). Digestion and absorption of carbohydrates, fats, and nitrogen occurs in the jejunum. In addition, the proximal jejunum is the site of secretin and cholecystokinin production, two hormones responsible for stimulation of pancreatic and bilious secretions. Complications following resection of the jejunum initially include malabsorption of macronutrients, but it is better tolerated than ileal resection. The ileum is the sight of the majority of electrolyte and fluid absorption. In addition, the distal ileum is responsible for absorption of vitamins B₁₂, A, D, E and K, and bile salts. Resection of the ileum may result in diarrhea, steatorrhea, and vitamin deficiencies. Other complications may arise if the ileocecal valve is lost. The ileocecal valve is responsible for two important functions. First, it acts as a barrier that prevents the movement of bacteria from the colon into the small intestine. Second, it regulates the transit time of fluid in the small intestine. With the resection of the ileocecal valve, the infant is at higher risk for bacterial overgrowth and more rapid transit time of intestinal contents, resulting in malabsorption. The colon is responsible for water absorption, some carbohydrate absorption, and synthesis of vitamin K. Resection of the colon may result in dehydration, hyponatremia, hypokalemia, and hypomagnesemia.

**Nutritional Management**

Infants with SBS present with unique nutritional challenges, both in the acute phase following bowel resection and long term as enteral feedings are resumed. The main goals of nutritional management in infants with SBS include providing enough nutrition for proportional
growth, maintaining fluid and electrolyte balance, and supporting bowel adaptation (Wessel & Kocoshis, 2007).

According to Goulet et al. (2012) there are three phases of nutritional management in patients with SBS. First is the acute phase which takes place immediately following bowel resection. Complications during this phase include electrolyte imbalance, impaired gut and intestinal motility, diarrhea, and dehydration. The nutritional goal during this phase is to normalize fluid and electrolytes imbalance through the use of PN and IV replacement fluids. In addition, the GI tract should be used as soon as possible to begin intestinal adaptation.

The second phase is the intermediate phase. This takes place after the initial stabilization and may last several weeks to several months. The main goal during this phase is to provide optimal enteral and parenteral nutrition to maintain the nutritional status of the patient. For term infants, the optimal caloric intake is 100-120 kcal/kg/day and for preterm infants, the range is 110-160 kcal/kg/day (Pierro & Eaton, 2008). Caloric requirements for PN nutrition is lower, ranging from 80-100 kcal/kg/day in term infants. Complications during the intermediate phase may include liver failure, catheter associated sepsis, and small intestine associated bacterial overgrowth (Goulet et al., 2012).

The third phase is the late phase, which involves weaning off of PN once intestinal adaptation is complete. During this phase, the main goal is to gradually wean off PN while maintaining normal growth and optimal nutrition with enteral intake. Close monitoring of macronutrients, micronutrients, electrolytes, and trace elements is important during this phase (Goulet et al., 2012).

**Parenteral nutrition.** The initial post-resection period presents unique nutritional challenges. Parenteral nutrition (PN) provides the nutrients necessary to sustain the infant and is
essential for survival. However, it is a double-edged sword since long term PN use is associated with liver failure in up to 60% of infants with intestinal failure (Kelly, 2006).

Fluid and electrolyte management are critical for patients with SBS. Electrolyte replacement fluids should be used in addition to PN in order to supplement losses from gastric tubes, secretory gastrointestinal output, or ostomy output (Wessel & Kocoshis, 2007). The fluid and electrolytes lost should be measured regularly and replaced. Urine sodium levels more accurately predict total body sodium than serum levels except in infants receiving loop diuretics or those with hyperaldosteronism due to liver failure (Wessel & Kocoshis, 2007).

Glucose and protein are important macronutrient components of TPN, each associated with liver damage when used in excess. In order for the body to use IV lipids as a calorie source, glucose intake must be below basal energy requirements. Jones et al. (1993) found that when the glucose infusion exceeds 18 mg/kg/min, fat oxidation stops and fat synthesis begins, increasing triglyceride and carbon dioxide levels in the blood. Hwang and Shulman (2002) recommend limiting glucose infusion rates to 8 to 12 mg/kg/min in order to decrease the risk for fatty liver disease (FLD). Hyperglycemia has also been found to increase morbidity and mortality in the neonatal population, suggesting adherence to strict glucose management is important for optimal outcomes (Hall, Peters, Eaton, & Pierro, 2004).

Protein supplementation is important for growth and wound healing. Some experts recommend supplementing with 3.5 mg/kg/day while others feel that excessive protein is another factor found to contribute to liver damage and as such, prefer to restrict supplementation. Senger et al. (1986) found an association between elevated protein supplementation and cholestasis. According to Wessel and Kocoshis (2007) studies that provide clear guidance for appropriate protein supplementation are lacking.
Lipid infusions are an important source of calories but are associated with long term complications of PN. Clayton et. al (1993) found that phytosterols in lipid emulsions were associated with cholestatic liver disease in infants who were maintained on long-term PN. Other researchers found that changes in lipid administration were followed by hepatic complications (Colomb et al., 2000). It was found that as lipid doses were decreased, a subsequent decrease in bilirubin levels occurred. The authors concluded that lipids increased risk for cholestasis in infants requiring prolonged PN therapy. A retrospective case-control study conducted by Angsten et al. (2012) investigated the outcomes of infants receiving parenteral fish oil versus traditional soybean based lipid emulsions. The authors found that all of the infants receiving the fish-oil based lipids showed normalization of bilirubin levels while only 10% of the control population had a reversal of bilirubin level. A major limitation of this study was the small sample size (n=20). Fish oil based lipids have not been approved by the FDA for use in newborns and controlled trials are needed to establish safety for their use. Despite a paucity of data, the results of these recent studies are promising.

Historically, copper and manganese are removed from TPN for patients experiencing cholestasis (Wessel & Kocoshis, 2007). Copper accumulation in the liver has been described in adult populations receiving long-term PN. Researchers in one study found no association between length of TPN therapy and elevated levels of copper in the liver (Blaszyk, Wild, Oliveira, Kelly, & Burgart, 2005). According to Wessel and Kocoshis (2007), more studies are needed to determine if the practice of removing copper from TPN is warranted, given resulting complications such as pancytopenia, anemia, and neutropenia.

Manganese neurotoxicity was reported in one infant on long term PN (Suita, Masumoto, Yamanouchi, Nagano, & Nakamura, 1999). According to the authors, manganese exists as a
contaminant in TPN at levels high enough to provide the nutrient without additional supplementation. Based upon these findings, Wessel and Kocoshis (2007) recommend removing supplemental manganese from TPN.

Zinc supplementation is another important consideration. In one study, zinc content in ileostomy fluid ranged between 12 to 17 mg/dL (Shulman, 1989). While zinc levels were low in the post-operative period, the use of zinc supplementation for two weeks in the TPN resulted in normal serum levels. Serum zinc levels may not accurately reflect total body zinc stores, so some clinicians recommend using a lower than expected alkaline phosphatase level as an indicator of deficient zinc stores (Wessel & Kocoshis, 2007).

**Enteral nutrition.** In order to decrease risk of complications from long term PN and initiate intestinal adaptation, enteral feedings should begin as soon as possible. Timing for enteral feeding initiation has not been well established in the literature. Bohnhorst et al. (2003) found that infants recovering from NEC who were fed at a median of four days following resolution of NEC by radiological confirmation experienced shorter hospitalization, fewer cases of sepsis, and shorter time to achieve full feedings versus infants fed at a median of 10 days. In a prospective multicenter study infants fed within a mean of 12 hours following abdominal surgery tolerated those feedings well (Ekingen, Ceran, Guvenc, Tuzlaci, & Kahraman, 2005). In addition, the treatment group tolerated full enteral feedings sooner and had fewer hospitalization days when compared to the control group, which did not receive enteral feeds until resolution of post-surgical ileus. The limitation of this study is that only infants weighing between 1500-4300 grams with congenital bowel obstructions were included in the sample.

The type of feeding an infant receives plays an important role in intestinal adaption and may decrease the days of PN dependency. Andorsky et. al (2001) found that infants who were
fed breast milk had mean duration of PN dependency of 290 days versus 720 days in formula fed infants. Enteral feedings of amino acid based, or elemental, formulas were also associated with shorter duration of PN dependency. Based on these findings, Olieman et al. (2010) recommended using breast milk first. When breast milk is not available, an appropriate infant formula must be selected. There are no clear recommendations as to the type of infant formula to use. One group of researchers conducted a randomized controlled trial comparing different infant formulas (Ksiazyk, Piena, Kierkus, & Lyszkowska, 2002). They found no difference in intestinal absorption, weight gain, energy, or nitrogen balance between infants receiving hydrolyzed formula versus non-hydrolyzed formula. If commercial term and preterm formulas are not tolerated, elemental formulas should be considered (Bines, Francis, & Hill, 1998).

How to provide enteral feedings and when to advance feedings are important clinical questions to address in infants with SBS. Studies have been inconclusive as to whether continuous versus intermittent feeds are more beneficial. Parker, Stroop, and Greene (1981) concluded that infants on continuous feedings had improved enteral balance of major nutrients compared to those who received bolus feedings. In contrast to their findings, Silvestre et. al (1996) found no difference in growth or macronutrient retention rates between the continuous and bolus feeding groups. Wessel and Kocoshis (2007) recommended initiating oral feedings as soon as developmentally appropriate and to involve speech and occupational therapists to enhance success of feedings.

Tolerance of enteral feedings has historically been monitored by stool output. In general, infants experiencing 40 to 50 mL/kg/day of fluid loss in the form of stools will begin to have electrolyte imbalances (Wessel & Kocoshis, 2007). Researchers in another study took a different approach to determine feeding tolerance by looking at hydration, electrolyte status, and acid-base
status (Alkalay et al., 1995). Infants were provided high volume feedings and tolerated ileostomy outputs ranging from 113 mL/kg/day up to 228 mL/kg/day. Findings from this study should be interpreted cautiously due to its small sample size (n=3). Until more evidence is gathered to the contrary, stool output should continue to be used as a guide for feeding tolerance.

Project Implementation

The primary purpose of the project was to develop a clinical practice guideline to be used by clinicians in order to provide consistent, evidence-based care. The project objectives were designed to achieve this overall goal. The following section discusses the implementation of the project objectives.

Objective 1: Development of the Evidence-based Clinical Guideline

The first objective was to develop a clinical practice guideline based upon the most current research literature. An extensive review of literature was conducted in September 2012. An initial review of literature document was submitted to the project chair for review in October 2012. Revisions were made and the final proposal document was submitted in December 2012. In January 2013 the proposal document was forwarded to the content experts. Based upon their feedback, an additional review of literature was conducted during January and February 2013.

There were challenges encountered when searching the literature. First, in the newborn population there are limited published studies addressing SBS. Finding high level evidence became a challenge with most nutritional management strategies based upon expert opinion. Bibliographies from review articles were utilized to find additional primary research articles. Content experts were also utilized to assist in identifying research articles to support the recommendations in the guideline.

In February 2013, the first draft of the clinical pathway was developed. The algorithm was scrutinized in meetings with the content experts. The input of the content experts and the
most current research literature were utilized to fine tune the guideline. It was then forwarded to
the manager of the Neonatal Nurse Practitioner group at PCMC for further evaluation and
suggestions for improvement. After several sessions, this objective was successfully completed
with the final guideline being finished in March, 2013.

The clinical algorithm was created as a quick reference to assist neonatologists and
neonatal nurse practitioners manage both the parenteral and enteral nutrition of neonates with
short bowel syndrome. In addition to the nutrition recommendations, the guideline provides
clinicians with additional considerations to help guide the management of these patients. There is
a list of laboratory studies and commonly used medications. There is also a list of signs of
feedings intolerance and a list of potential complications that may be encountered by this
population. A copy of the completed guideline is found in the Appendix A.

Objective 2: Utilize Content Experts to Develop the Guideline

Content experts were vital to the development of the project. NNP program faculty and
the project chair were consulted to develop a list of potential content experts. Cecilia Mulroy,
RD, CD, CNSP is the lead dietician in the NICU at PCMC. She was approached to assist with
the project given her extensive background working with patients with SBS. Daniel Malleske,
MD, MS, FAAP is one of the neonatologists working at PCMC with clinical experience working
with this population and an interest in taking the project to the next level by conducting a
research study to evaluate the effectiveness of the guideline. In November 2012, agreements
were signed by both content experts to take part in the project. Their input has been invaluable
during development of the guideline. Both individuals have offered unique clinical perspective
and were able to help identify the most important components to include in the guideline. Ms.
Mulroy and Dr. Malleske provided extensive feedback, editing, and suggestions for improving
the guideline, as well as approving the final version of the document to be presented to the
Division of Neonatology in June 2013.
Objective 3: Eventual Adoption of the Proposed Guideline

The third objective was to present the guideline to the Division of Neonatology at PCMC for approval to incorporate it into the division's guidelines and protocols for practice. Although the initial target date for achieving this objective was set for February 2013, the content experts have recommended delaying presentation until June of this year to allow for refinement of the guideline. A preliminary Power Point presentation was completed in mid-April, but this is currently being revised in collaboration with the content experts to build a compelling case for the guideline’s adoption. The author’s time-frame for achieving this goal may have been too unrealistic given the lengthy institutional process at PCMC for adopting new evidence-based clinical practice guidelines.

Objective 4: Dissemination of the Guideline

The fourth and final objective for the project was to disseminate the guideline to a wider audience of neonatal practitioners. The original plan was to prepare a manuscript for submission to a peer reviewed journal. This objective has been modified based upon the new direction the project has taken namely, the recommendation from Dr. Malleske to first perform a comparative study to test the new clinical practice guideline before publishing a paper. Following the conclusion of the proposed study, a manuscript will be written and submitted to an appropriate peer reviewed journal.

The proposed clinical practice guideline was presented to nursing colleagues during a poster defense of the project which took place on April 18, 2013. The poster presentation was open to all students, faculty and employees of the University of Utah Health Sciences Center, during which questions were answered by the project’s author. A copy of the poster is located in Appendix B.

Further dissemination of the clinical practice guideline will take place in June 2013 when it is presented to the Division of Neonatology during fellow’s rounds. This meeting is open to pediatric residents, neonatal fellows, neonatologists, nurse practitioners, nurses, and students. The presentation provides a brief background discussing the clinical significance of SBS, the
pathophysiology, and the underlying clinical rationale for its use based on current evidence. The majority of the presentation focuses on the guideline and the strength of the evidence supporting the recommendations.

**Limitations**

There were a number of limitations encountered during the development of the project. First among these was an unrealistic time frame in which to achieve all the objectives given the complexity of the issue being addressed. Because of the paucity of literature and strength of the evidence which relied mostly on expert opinion, collating the evidence and expertise of content experts to support construction of the guideline was time-consuming. The guideline was also re-submitted several times for feedback and revisions, also delaying achievement of goals.

Another limitation encountered was the lengthy process by which clinical practice guidelines are evaluated, approved, and adopted by PCMC. Numerous steps are required in the approval process. Meetings are held monthly and additions to the agendas of these meetings require permission, which delayed the window of opportunity to present the guideline to the Division of Neonatology within the project’s time-frame. This in turn delayed goal achievement.

Despite these limitations, the content experts with whom I worked during this project are committed to seeing the implementation of the guideline. Work on the project will continue beyond graduation. The next step will be presenting the guideline in June 2013 to the Division. Recommendations from the neonatologists will be incorporated into the guideline with a goal of implementing the guideline in clinical practice in late summer or early fall of 2013.

**Opportunities**

Although limitations arose, unintended opportunities presented themselves during the implementation of the project. Dr. Malleske and Ms. Mulroy were very enthusiastic about this
project and expressed a desire to extend work on it beyond the time restriction for the scholarly project. In addition to development and approval of the guideline for clinical use at PCMC, a research study is planned in order to evaluate and compare patient outcomes using the guideline. Proposed outcome criteria include length of hospitalization, time to full enteral feedings, growth, length of PN dependency, and complications or adverse outcomes. Work on the proposed study is currently underway.

**Future Recommendations**

Numerous opportunities exist for further evaluation of the proposed guideline along with additional research opportunities.

- The guideline will need to be evaluated clinically to determine its effect on outcomes for neonates with SBS. Outcome measures of interest include length of PN dependency, time to achieve full enteral feedings, length of hospitalization, and incidence of complications.

- If the guideline successfully improves patient outcomes, findings will need to be distributed to a wider audience of neonatal practitioners which may involve future publications or presentations at national conferences.

- Studies are needed to evaluate the optimal enteral formula. While studies show that breast milk is the best enteral feeding choice, alternative choices have not been adequately studied. Further research in this area needs to take place.

- Studies need to be conducted to examine alternative lipid sources in PN. Lipid infusion is a known cause of liver failure in infants with SBS. Promising results using fish-based oils have been demonstrated in studies; however, the strength of the evidence is questionable.

- Pharmacologic interventions were beyond the scope of this project. Very general recommendations on classes of medications were made in the guideline. A systematic
review of the current evidence needs to be conducted to determine what is known about the safety and efficacy of various pharmacologic agents in the management of SBS.

- The guideline developed for this project is intended for use with hospitalized neonates initially diagnosed with and treated for SBS. Guidelines for home management of these children need to be developed. Strategies such as cycling TPN and introducing solid foods at home or later in the hospitalization course are not included in this guideline and will therefore need to be incorporated at a later time.

Conclusion

Short bowel syndrome is a serious issue affecting the NICU population. A guideline was necessary to provide clinicians with an evidence-based roadmap to assist in managing the complex nutritional needs of neonates with SBS. Over the past several months, in-depth literature reviews were conducted and content experts were consulted to develop the clinical practice guideline. Although the project is in its infancy, it is the author’s intention to advocate for its use in clinical practice. The effectiveness of the guideline will be evaluated with the intended goal being its adoption by PCMC. While the guideline may be a useful tool for clinicians, the ultimate goal is to improve patient outcomes by providing consistent, evidence based care.
References

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Appendix A

Nutritional Management of Neonates with Short Bowel Syndrome
- For use in neonates with < 25% contiguous bowel for gestational age or anticipated need for PN > 6 weeks

**Micronutrients**
- Zinc: 400 mcg/kg/day if less than 3 kg, 200 mcg/kg/day if > 4 kg. Check level at one month.
- Copper: Check level after one week on PN, supplement at 10-20 mcg/kg if level is less than laboratory normal.
- Magnesium: Do not supplement if PPI, enough exists as a contaminant to provide the nutrient.

**Initiate Maintenance PN**
- Obtain central venous access.
- Target fluid total: 150 mL/kg/day.
- Target Kcal: 80-100 Kcal/kg/day

**Fluid and electrolyte replacement**
- Monitor fluid loss from gastric tubes, secretary GI output, and enteral output 0.2-3 hours.
- Bayley’s Law: 1:1 if output > 10 mL/kg with ONS 0.45% NS with 20 mL/kg KCl. Consider additional replacement if signs of dehydration.

**Laboratory Monitoring**
- Electrolytes, BUN, creatinine, phosphorus, magnesium, albumin, triglyceride, and glucose with changes to PK, weekly until stable, then every 2 weeks.
- Copper and urine iodine after 7-10 days on PN.
- Selenium after two weeks on PN.
- Zinc, Vitamin A, and 25 OH D after one month of PN.
- Iron studies (Zinc, protoporphyrin and ferritin) in documented microcytic anemia.
- Vitamin B12 one week off PN or 3 months of PN if ileal fluid was resected. Consider sooner if macrocytic anemia develops.
- Direct and indirect bilirubin, AST, ALT, and SGOT every 1 to 2 weeks to monitor for PNH/D.

**Micronutrients**
- Glucose: Max GIR 15 mg/kg/min
- Protein: 2.5-3.5 g/kg/day
- Lipid: Minimum 0.5 g/kg/day, no more than 30% of total calories. If > 21 days of PN anticipated, limit to 1.5 g/kg/day

**Assess Readiness for Enteral Feeds**
- Resolution of post-surgical issues.
- Reassuring abdominal exam.
- Absence of GI symptoms/ileus.

YES

NO

**Initiate Enteral Feeding**
- Begin low volume triglycerides feeds (10-20 mL/kg/day).
- Continuous feeds once able to tolerate 10-15 mL/hour.
- Breast milk preferred.
- Donor breast milk < 34 weeks, < 1880 grams.
- Semi-Elemental formula if no BM available.
- Preterm, preterm, neonates, formula.
- Select formula with higher LCTs than MCTs.

**Do NOT Begin Feeds**
- Continue PN.
- Reassess readiness to begin enteral feedings daily.

YES

NO

**Assess Readiness to Advance Feeds**
- Tolerating feedings?

**Advance Enteral Feeds**
- Increase feeds as clinically tolerated, not to exceed 20 mL/kg/day.
- Re-feed enteral output into mucous fistula when cleared to do so by surgical team.
- Begin oral feeds when developmentally appropriate. Start with 5 to 10 mL per feeding 2-4x per day as clinically indicated.
- Begin condensing pump feeding times once full volume continuous feeds are tolerated.
- Total Kcal goal (30-70% > than PHN).
- Term: 120-130 Kcal/kg/day.
- Preterm: 110-160 Kcal/kg/day.

**Do NOT Advance Feeding**
- Decrease to previously tolerated volume.
- Consider NPO.
- Consider working up for SIBO.
- Reassess readiness to advance daily.
- Attempt to advance feedings once symptoms of intolerance subsided.
- If bolus feedings not tolerated, increase pump times or resume continuous feeds.
- If unable to tolerate full volume enteral feeds:
  - Target nutritional goal: 60% total calories from enteral feeds. 40% calories from PN. Concentrate PN to ensure 100% infusion once tolerating feeding goal.

**Signs of Feeding Intolerance**
- Vomiting greater than 20% of daily feeding volume or bilious emesis.
- Fecal production greater than 30 mL/kg/day.
- Stool pH less than 5.5.
- Presence of fecal reducing sugars > 2.
- Increased abdominal girth.

**Pharmacologic Therapy**
- H2 Blocker: PPI for hypersecretion of stomach acid during the first few months.
- Cholestyramine: For diarrhea secondary to bile salt malabsorption. Monitor weight gain.
- Pectin: Consider in patients with contiguous colon.
- Antimotility agents: Avoid in neonates, use cautiously in older infants and children.
- Others: Glutamine, epidermal growth factor. GIP-2. None of these have sufficient evidence to date to support their use.

**Assess Daily for Adequate Nutrition**
- Tolerance of feeding?
- Appropriate growth for age.
- < 25 mL/kg/day (<2.5 kg).
- 25-35 mL/kg/day (2.5 kg).
- Laboratory studies.
- Presence of complications.

**Clinical Judgment Supersedes Guideline**
- Feedings should not be advanced faster than recommendations but may be advanced slower per patient’s clinical status.

**Complications**
- Watery diarrhea (>40 mL/kg/day).
- Parenteral Nutrition Associated Liver Disease (PNALD).
- Small intestine bacterial overgrowth (SIBO).
- Central line infection.
- Hypertrophic kidney stones.
- Nutrient deficiencies.
Appendix B

Nutritional Management of Infants with Short Bowel Syndrome

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PROBLEM STATEMENT
• Short Bowel Syndrome (SBS) is a significant contributor to newborn morbidity and mortality
• Long term dependency of parenteral nutrition (PN) leads to complications
• A clinical practice guideline (CPG) is needed to address: (1) ideal composition of PN to minimize the risk of intestinal failure associated liver disease (IFALD), (2) timing for initiating and advancing enteral feedings, (3) use of breast milk, polymeric, or semi-/elemental formula, and (4) laboratory monitoring
• An evidenced-based CPG will promote consistency of care with the goal of improving patient outcomes
• Currently no CPG for nutritional management of this population exists at Primary Children’s Hospital

OBJECTIVES
1. Develop a CPG to promote nutritional management of neonates with short bowel syndrome.
2. The proposed guideline will reflect the most current evidence and expert opinion in the field of neonatal nutrition for children with SBS.
3. Present the guideline to the Division of Neonatology at Primary Children’s Hospital for approval to incorporate into the division's guidelines and protocols for practice.
4. Prepare a manuscript for submission to an appropriate peer reviewed journal for dissemination to a broader audience of neonatal practitioners.

REVIEW OF LITERATURE
• Definition and Etiology of SBS
• Pathophysiology
• Location of the intestinal resection affects nutrient, vitamin, mineral, and fluid absorption
• Parenteral Nutrition
• Fluid and electrolyte management
• Glucose and protein
• Lipid infusions
• Trace minerals
• Vitamins
• Enteral Feedings
• Timing
• The type of feeding
• How to provide enteral feedings and when to advance
• How to assess tolerance
• Laboratory monitoring
• Medications

CLINICAL SIGNIFICANCE
• Incidence of SBS: 2.45/100,000 live births
• Incidence is 100 times greater for preterm versus term infants
• Mortality is as high as 37.5%
• Other Complications: Bacterial overgrowth, IFALD, diarrhea, lactic acidosis, increased gastric acid, gall stones, kidney stones, sepsis, nutritional deficiencies

METHODS
• Development of a clinical algorithm using the most current evidence.
• Research studies were evaluated for level of evidence.
• Recruitment of a neonatologist and dietitian to provide guidance and content expertise during development of the guideline.
• Revisions were made based on content expert feedback.
• Final CPG content and format approved by content experts.

EVALUATION/RECOMMENDATIONS
• Invited to present the CPG to the Department of Neonatology at a June 2013 Division meeting.
• Manuscript to be drafted following a planned research study to assess clinical outcomes of patients during implementation of the guideline in the NICU setting.
• The impact of the guideline on the system and health care providers will need to be evaluated during a follow up period with revisions being made based on stakeholder input.