SHORT BOWEL SYNDROME (SBS)

A little knowledge
can make a big difference.

INTERNATIONAL FOUNDATION FOR FUNCTIONAL GASTROINTESTINAL DISORDERS

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INTRODUCTION

Short bowel syndrome (SBS), or simply short gut, is broadly described as a condition in which nutrients are not properly absorbed because a large part of the small intestine is missing. This is most often due to defects existing at birth (congenital), or surgical removal of part of the small bowel. There may not be enough functioning bowel or surface area left in the remaining bowel to absorb needed water and nutrients from food. Sometimes, loss of normal function may occur even when the bowel length is intact. Typically a loss of half or more of the small bowel will result in SBS.

Managing short bowel syndrome is challenging for patients, families, and healthcare providers. In addition to dealing with multiple symptoms, special steps must be taken to be sure that fluid and nutrient requirements are met.

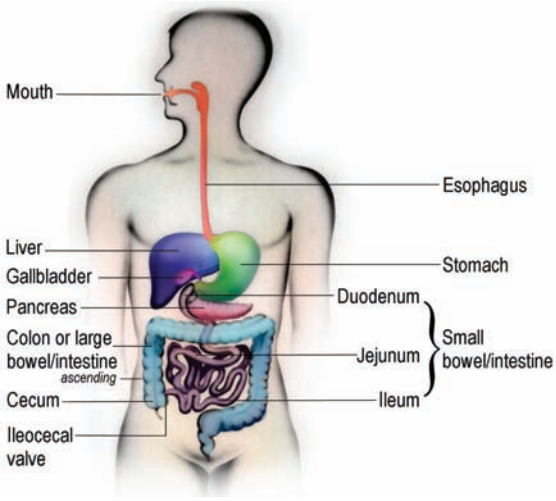
Treatments for short bowel syndrome are aimed at controlling symptoms and maintaining nutritional status. This involves special dietary measures and often use of medications. In some situations surgery is required. Many people with SBS are unable to take in adequate fluids and nutrients by oral diet alone and must depend on parenteral nutrition (through a vein) or enteral nutrition (through a feeding tube).

This publication is intended to help people with short bowel syndrome and their family members understand why symptoms occur and provide an overview on how SBS can be managed.

SHORT BOWEL SYNDROME

Short bowel syndrome is a condition characterized by malabsorption – difficulties absorbing both nutrients and fluids. Each year, many patients undergo surgical removal (resections) of large segments of their intestinal tract due to diseases, injuries, or congenital defects. Those patients can be left with too little intestinal absorptive surface areas. This then may lead to malabsorption and resulting malnutrition, diarrhea, and fluid and electrolyte imbalances. The severity of short bowel syndrome in an individual depends on several factors, including:

- length of the remaining bowel,
- site of the resection,
- presence of the ileocecal valve,
- presence of the colon,
- health of the remaining bowel, and
- ability of the remaining bowel to compensate (adapt).



THE DIGESTIVE SYSTEM

Food and liquid must be converted into forms that the body can use as nourishment. What we eat and drink is broken down into small molecules that can be absorbed into the blood to nourish cells. The process is normally carried out in the digestive system, which includes the mouth, esophagus, small intestine (or small bowel), and large intestine (colon or large bowel). The liver and pancreas produce digestive chemicals or juices important for digestion.

Mouth and Stomach

Digestion begins in the mouth where food is chewed, mixed with saliva, and swallowed. It moves down the esophagus to the stomach. In the stomach the food stimulates the release of digestive juices (secretions) like hydrochloric acid and digestive enzymes that chemically further break down and mix with the food. The mixture is referred to as chyme.

Small Intestine

The chyme then passes, in a regulated controlled manner, out of the stomach into the small bowel/intestine. The average total length of the normal small bowel in adults is about 7 meters/22 feet. The small intestine has 3 segments:

- the duodenum,
- the jejunum, and
- the ileum.

Each part or section performs an important role in nutrient absorption.

Duodenum – The chyme first enters into the duodenum where it is exposed to secretions that aid digestion. The secretions include bile salts, enzymes, and bicarbonate. The bile salts from the liver help digest fats and fat soluble vitamins (Vitamin A, D, E, and K). Pancreatic enzymes help digest carbohydrates and fats. Bicarbonate from the pancreas neutralizes the acid from the stomach.

Jejunum – The chyme is then further transited down into the second or middle part of the small intestine, the jejunum. Mainly in the first half of the jejunum, the majority (about 90%) of nutrient absorption occurs involving proteins, carbohydrates, vitamins, and minerals.

Ileum – The ileum is the last section of the small intestine and leads to the large intestine or colon. The ileum mainly absorbs water, bile salts, and vitamin B12.

The ileocecal valve is a one way valve located between the ileum and the cecum, which is the first portion of the colon. This valve helps control the passage of contents into the colon and increases the contact time of nutrients and electrolytes (essential minerals) with the small intestine. It also prevents back-flow (reflux) from the colon up into the ileum, and minimizes the movement of bacteria from the large intestine up into the small bowel.

Depending on what parts of the small bowel are removed or not functioning properly deficiencies of certain nutrients will result. These deficiencies may be many and complex. Examples include:

- Iron deficiency (duodenum)
- Vitamin and mineral deficiency; and malabsorption of carbohydrates, proteins, and fats (jejunum)
- Vitamin B12 deficiency and malabsorption of bile acids (ileum)
- Small intestinal bacterial overgrowth and increased fluid losses (ileocecal valve)

Large Intestine

The primary function of the large intestine or colon is to absorb fluids and electrolytes, particularly sodium and potassium, and to convert remaining luminal contents into more solid stool. The colon absorbs on average 1–1.5 liters (about 1–1.5 quarts) of fluid every day and has a capacity to adapt its fluid absorption to as much as 5 liters/quarts per day if needed. Another function of the colon is to break down (ferment) dietary fiber to produce short chain fatty acids – substances that can be absorbed and provide added nutrition.

The first portion of the colon, the cecum, is shaped like a pouch, and is the area of storage for the contents arriving from the ileum. The second portion is the ascending colon, where fluids are absorbed and where some stool formation begins.

SYMPTOMS OF SBS

Symptoms of short bowel syndrome result when fluids and nutrients are not properly absorbed. These will vary from person to person. Symptoms often include:

- diarrhea,
- fatigue, and
- pale greasy stools (steatorrhea).

Swelling of lower extremities (edema), foul smelling stools, weight loss as a result of dehydration, electrolyte losses, and malnutrition are often experienced.

Vitamin and mineral losses can lead to some symptoms. Depending on which vitamin or mineral is deficient, symptom examples include visual disturbances and excessive dryness of the eyes; prickling or tingling feeling on the skin; muscle spasms; loss of coordination; loss of bone mass; easy bruising and/or prolonged bleeding; lack of energy (lethargy), weakness, or difficulty breathing on exertion.

RISK FACTORS

Short bowel syndrome can occur in a person of any age. Risk factors for SBS include defects existing at birth and diseases of the small intestine that require extensive or recurrent surgery such as Crohn's disease or gastrointestinal cancers. In addition SBS can be caused by loss of function due to injury or disease in a normal length small intestine. Other explanations include emergency situations related to injury or trauma, perforated bowel, or blocked or restricted blood flow to the bowel.

WHAT HAPPENS WHEN SBS DEVELOPS

Immediately following surgical resection of the small intestine, with the resulting loss of absorptive surface area, the intestine begins to compensate on its own. It undergoes various phases to increase absorption and maintain balance (homeostasis). This process, known as adaptation, occurs through structural changes that increase surface area in the remaining bowel. These physiological changes and adaptations can be separated into 3 phases:

1. Acute phase
2. Adaptation phase
3. Maintenance phase

The *acute phase* occurs immediately after bowel resection and may last 3–4 months. This phase is associated with malnutrition, and fluid and electrolyte losses as high as 6–8 liters/quarts per day. Enteral nutrition may be needed during this phase. If a more significant length of small bowel is removed parenteral nutrition is required.

Next, *the adaptation phase* begins 2–4 days after bowel resection and lasts 12–24 months. During this phase the intestinal villi, the tiny finger-like projections within the small intestine, will grow in length and thickness, which increases surface area. In addition some increase in the diameter (dilation) of the bowel may occur.

The *maintenance phase* is the last change, and here the absorptive capacity of the remaining bowel will be maximized. Some patients will still have dependence on parenteral or enteral nutrition. Others will meet their nutritional needs with oral meals, nutrition supplements, and vitamins and minerals, with or without supplements delivered enterally or parenterally.

TREATMENT

The aims of treatment for short bowel syndrome are to promote adaptation and get the best use out of the existing bowel, maintain adequate nutritional status, and manage symptoms and complications. Complications can arise not only as a result of the underlying condition, but also in connection with treatments.

The ultimate goal is for the patient to resume daily life as well as possible. Treatment involves some combination of:

- Nutrition strategies
- Managing gastric acid secretions, bacterial overgrowth, bile salt malabsorption, and diarrhea
- Medications
- Surgery

Treatment of short bowel syndrome begins immediately after bowel loss. It starts with restoring fluid and electrolyte balance and quickly progresses to nutritional support. Nutritional support includes individualized meal plans and may involve the use of supplements, oral rehydration solutions, enteral nutrition, or parenteral nutrition. Treatment will often include medications, and in some instances surgery. In situations where all other treatment approaches have failed, intestinal transplant is considered. The course of treatment will depend on how well the bowel is able to support individual fluid and nutrient needs.

DECIDING ON THE TREATMENT

Deciding on the treatment requires knowledge about the remaining length of small bowel. This may require diagnostic testing, which usually begins with x-rays and/or an enteroscopy (using an endoscope, a thin, flexible tube with a light and a lens on the end) to examine the esophagus, stomach, and small bowel.

In the average adult, the minimal length of small bowel required so that some nutrient absorption can be anticipated is 4 feet. Usually this is counted as 4 feet of jejunum beyond the point where the duodenum ends (ligament of treitz). Parenteral nutrition is always required for remaining small bowel of less than 4 feet.

The length required for adequate fluid absorption and avoidance of dehydration will be influenced by whether or not the colon is intact. Parenteral nutrition is still required on many days for remaining bowel length of less than 10 feet.

Nutrition Strategies

Nutrition strategies aim to prevent malnutrition and dehydration, and maintain the best possible nutrition status. This may include oral eating, enteral nutrition, and parenteral nutrition. All methods may be used by patients in their homes. Nutrition support specialists (registered dietitians, nurses, doctors) play a key role in helping manage short bowel syndrome. Nutrition specialists will tailor the approach used to each person according to their individualized needs and provide detailed dietary guidelines. Nutritional needs may change over time.

Oral eating is preferred whenever possible. Recommended diet components and balance of nutrients will vary in each person with SBS. A number of factors influence this including the resection site(s) and remaining bowel.

In the majority of people with short bowel syndrome the colon is intact. The general dietary guidelines for those with a colon involve a low fat, high carbohydrate diet. A diet that includes chopped, ground, or well chewed nutrient rich foods, with small frequent meals (up to 6–8 per day) is recommended. Fluids should be taken in between meals rather than with meals.

Concentrated sweet foods and liquids should be avoided; and foods high in oxalate limited to avoid kidney stones. Lactose restriction may help some individuals, as well as limiting alcohol, and caffeine.

With greater stool losses recommendations are for beverages formulated to replace fluid and electrolytes (isotonic). Oral rehydration solutions contain specific amounts of sodium, carbohydrate, and water, which increase fluid absorption in the small bowel.

Salty meals and/or snacks, plus a soluble fiber supplement can be helpful when the absorptive colon segment is present. In addition, a probiotic supplement and multivitamin and mineral supplements may also be advised. A supplement of pancreatic enzymes is often used to aid in digestion and help prevent gas and passing fat in stools (steatorrhea) when other measures are not effective. The type and dose of supplement advised will depend on the site and extent of surgical resection.

Enteral nutrition is used when oral eating does not supply adequate nutrition. Enteral nutrition involves the delivery of liquid food to the stomach or small intestine through a feeding tube. While not without risks, it is associated with fewer complications than parenteral nutrition.

Both enteral nutrition and oral eating stimulate the remaining intestine to function better (adaptation). This may allow patients to avoid or discontinue parenteral nutrition.

The length of remaining and functioning small intestine is a key factor. The length needed for adequate absorption will be influenced by whether or not the colon is intact. When nutritional goals cannot be met by other means, the medical management will likely rely on long-term use of parenteral nutrition.

Removal of up to 50% of total small bowel is generally well tolerated from the standpoint of maintaining nutritional requirements. If greater than 50% of small bowel is removed the amount of functional small bowel remaining and whether the colon is still present are the determining factors if parenteral nutrition can be avoided.

Parenteral nutrition bypasses the digestive system. It involves the delivery of fluids, electrolytes, and liquid nutrients into the bloodstream through a tube placed in a vein (intravenous or IV). It is often needed short-term after resection while the remaining bowel adapts. It may be needed long-term depending on the bowel's ability to absorb nutrients. If there is greater than 4 feet of small bowel remaining, then attempts to go from daily parenteral nutrition to a less frequent use can be tried.

Parenteral nutrition is a complex therapy. The long-term use of parenteral nutrition significantly impacts quality of life issues such as loss of sleep, mobility, and social interactions. It also increases the risk for infections and other complications. Some complications can be life-threatening, including liver failure, vein thrombosis (blood clot), and sepsis (bloodstream infection). A specialist in nutrition support will provide detailed instructions on how to use and maintain parenteral, or enteral, nutrition.

Managing Gastric Acid Hypersecretion

Gastric hypersecretion happens when the stomach produces too much acid. It must be addressed after significant resection of the small bowel. It will increase the volume of secretions entering the small bowel and increase acid in the upper gut.

Loss of segments in the small bowel results in a change in the levels of hormones involved with digestion (cholecystokinin or CCK, secretin, and gastrin) that results in continued acid secretion. This increased acid load causes erosion of the gut lining and an increased stool volume contributing to diarrhea, and electrolyte losses. In addition it alters pancreatic enzymes, and compromises bile salt function making them less effective. The treatment for gastric hypersecretion is acid suppression through H2 blockers, and proton pump inhibitors (PPIs).

Managing Bacterial Overgrowth

When the surgical resection results in loss of the ileocecal valve, bacteria from the colon can enter the small bowel. Small intestinal bacterial overgrowth (SIBO) occurs when there are too many bacteria in the small intestine. Symptoms occur after eating because the bacteria in the intestine begin to consume the food in the small intestine before it can be absorbed. These bacteria give off hydrogen and other gases, which cause bloating and diarrhea. These bacteria can also contribute to malabsorption and loss of nutrients. Here there is a treatment role for cycles of antibiotics, and a long-term maintenance approach with probiotics.

Managing Bile Salt Malabsorption

Bile is a fluid produced in the liver and stored in the gallbladder. It is released into the duodenum during a meal to help digest fats and is reabsorbed in the ileum. When part of the ileum is lost, it results in malabsorption of bile acids which can lead to diarrhea. This may be treated by medications that bind bile, such as cholestyramine (Questran, Cholybar). However, when too much ileum is removed (greater than 100 cm/3.3 feet of the terminal ileum), more severe bile salt malabsorption may occur, and the liver is unable to compensate. When that happens cholestyramine may actually worsen steatorrhea, or undigested fat in the stools.

Managing Diarrhea

Current supportive medical treatments include the use of anti-motility agents that reduce fluid loss, and

hence decrease diarrhea. Reducing motility slows transit time and increases intestinal absorption. Examples of these agents include loperamide (Imodium), diphenoxylate with atropine (Lomotil), opium, and codeine. Octreotide is another drug that may help increase absorption time and decrease diarrhea. It reduces bile and pancreatic secretions and gastric acid production, while inhibiting fluid and electrolyte secretion from the small bowel. In addition it slows stomach emptying allowing increased transit time.

Other Medications

In the recent years pharmacological hormonal therapy has been introduced aiming to stimulate intestinal adaptation after intestinal resections. There are studies involving growth hormone, glutamine, and glucagon-like peptide 2 growth hormone (GLP-2).

The U.S. Food and Drug Administration (FDA) has approved both growth hormone and glutamine as drugs to be used in the management of SBS. Most recently in 2012, teduglutide (Gattex), a recombinant analog of human glucagon-like peptide 2, was FDA approved for the treatment of adults with short bowel syndrome who are dependent on parenteral support. Gattex works by regeneration of cells in the intestinal lining, improving intestinal absorption of fluids and nutrients, and helping reduce the frequency and volume of parenteral nutrition.

Surgical Options

The main goal of surgery for short bowel syndrome is to increase the capacity for absorption by the existing bowel. Surgery is considered in patients who are dependent long-term on parenteral nutrition when medications have failed and where the goal is to try to avoid intestinal transplantation.

Several different surgical procedures have been devised depending on the existing bowel length and function. The goal of these procedures is to improve function of the existing bowel by slowing transit or increasing surface area. Different methods include reconnecting remaining small bowel to the colon when continuity has been lost, attempting to increase transit time by reversing a segment of bowel, and intestinal lengthening procedures.

Intestinal transplantation becomes a necessary option when required life-long parenteral nutrition begins to fail. This is defined as significant evidence of liver injury, loss of central venous access, frequent line bloodstream infections (sepsis) which are becoming life threatening, and inability to keep up with excessive fluid loss (dehydration).

Isolated intestinal transplant is performed as the preferred procedure in patients who have adequate liver function. However, a combined liver and intestinal transplant is required for those with liver failure. Management strategies for intestinal transplantation, both before and after the operation, require careful consideration involving a multidisciplinary team of specialists available at only a few centers.

WORKING WITH A HEALTHCARE TEAM

Managing short bowel syndrome requires the patient and often family members working together with a team of healthcare professionals. Members of the healthcare team may include primary care physicians (for example, family doctor, pediatrician, or gastroenterologist), surgeons, nutritional specialists, nursing specialists, and pharmacists. The primary care physician will take the lead in managing and coordinating the patient's care. If intestinal transplant becomes necessary, other specialists may be brought in including social workers, psychologists, and financial counselors to help deal with the complexities of organ transplants.

The most important member of the healthcare team is the person with short bowel syndrome. Family members or parents of children with SBS play essential roles as caregivers. Patients and caregivers need to have a thorough understanding of the condition and how it may best be managed in light of individual needs. This will include recurring contact with healthcare providers, and most likely use of outside resources, all aimed at helping navigate the complexities of managing SBS long term. A list of several resources is provided at the end of this publication.

MANAGE BOTH RISK AND BENEFIT OF TREATMENTS

All drugs and procedures have inherent risks. Some are unavoidable, while others can be avoided and managed. It is important for patients and families to talk to doctors and healthcare team members about both benefit and risk of any treatment. Working together helps make treatment decisions that best fit individual needs. Here are some questions to consider:

- *What are the possible benefits of the treatment*
- *How much benefit can reasonably be expected*
- *What are the possible side effects of the treatment*
- *What are the chances of experiencing a side effect*
- *How can the chances of a side effect be reduced*
- *What action should be taken if a side effect occurs*

RESOURCES

International Foundation for Functional Gastrointestinal Disorders (IFFGD), www.iffgd.org A nonprofit organization whose mission is to inform, assist, and support people affected by gastrointestinal disorders of function and motility, like short bowel syndrome.

A Patient's Guide to Managing a Short Bowel, by Carol Rees Parrish, M.S., R.D. An easy to read and comprehensive book aimed at patients and family members that helps understand the workings of the GI tract and how patients can get the most from their own short gut. Includes dietary guidelines, sample meal plans, as well as descriptions of treatments and management issues. Available free of charge in the U.S. and Canada at: *www.shortbowelsupport.com.*

The Short Bowel Syndrome Foundation, www.shortbowelfoundation.org. A nonprofit organization whose mission is to educate, support, and empower patients who live with the condition and the healthcare providers who help patients to manage short bowel syndrome.

The Oley Foundation, www.oley.org. A nonprofit organization whose mission is to enrich the lives of patients dependent on home intravenous (parenteral) and tube feeding (enteral) through education, outreach, and networking.

Transplant Living, www.transplantliving.org. A project of the nonprofit United Network for Organ Sharing (UNOS), Transplant Living provides resources to help people before and after organ transplantation.

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