Short Bowel Syndrome (SBS)
(also known as short gut syndrome, short gut, intestinal failure)

Patient and family information, brought to you by the Education Committee of APSA

Overview - "What is it?"

Short Bowel Syndrome (SBS) is a condition in which the intestines, specifically the small intestine, are unable to digest and absorb the proper amount of nutrients from food to sustain life. This condition requires intravenous (IV) or enteral (oral) supplementation of nutrients to maintain continued growth and development for the child.

Short bowel syndrome can occur anywhere from birth into adulthood.

Causes:

- SBS can be caused by congenital (conditions present at birth) defects, malabsorptive diseases (diseases where the intestines cannot absorb adequate calories and nutrients) or surgical removal of long segments of intestines.
- The most common cause of SBS in infants and babies is necrotizing enterocolitis (NEC) which accounts for up to 40% of SBS patients. NEC is a severe infection of the intestines that is common in premature infants and may require removal of a long segment of intestine.
- SBS can also result from volvulus (twisted intestine leading to lost blood flow), atresia (something causes the intestine to have lost blood flow while the baby is developing in the uterus, causing segments of intestine to disappear) or complications of meconium ileus (impacted intestines commonly associated with cystic fibrosis), among other causes.
- Later in childhood, intestinal trauma, or inflammatory bowel conditions such as Crohn's disease can also cause SBS.

The normal length of the small intestine in a full-term infant is approximately 250 cm (about eight feet). In general, if most of the intestinal length is gone or removed, SBS can result. In some situations, although there can be adequate length of intestine, the intestine may not work well, therefore leading to inability to digest and absorb nutrients.

Signs and Symptoms - "What symptoms will my child have?"

Early symptoms:

- Diarrhea: Inability to absorb fluid in the intestine leads to diarrhea
- Failure to thrive: Since the baby is not able to digest or absorb food, it is difficult to grow and gain weight.
- Lack of energy and failure to meet milestones

Diagnosis - "What tests are done to find out what my child has?"

Physical examination by a physician is routinely performed with emphasis on growth charts. In addition, looking for symptoms of vitamin deficiencies during the exam is also done.

Stool tests to see if fat and carbohydrates are being digested.

Blood and urine tests will be ordered regularly to check your child's electrolyte, vitamin, and nutrition parameters.

Radiographic imaging such as **abdominal X-rays, ultrasound or CT scans** can be used to investigate the different causes of SBS.

Treatment - "What will be done to make my child better?"

Most children with SBS are managed either fully or partially with nutrition delivered by vein (Total Parenteral Nutrition or TPN).

The goal of treating SBS patients is to train the intestines gradually to digest and absorb nutrients and transition the child's nutrition from being given by vein to being given into the gut.

Medical strategies can take months to years to properly treat SBS. Principles of intestinal rehabilitation include:

- Early oral feeding is recommended as soon as your child is stable.
- Sometimes, feeding needs to be given continuously and your child may require a gastrostomy tube.
- Often special formula that has been partially digested is needed.
- Total Parenteral Nutrition (TPN) is a way to give nutrition through the vein. It consists of carbohydrates, protein, fat, vitamins, and minerals. Often, TPN is needed to supplement nutrition in patients with SBS.
- Long-term use of TPN use is associated with blood stream infections and liver disease.

- Antimotility (anti-diarrheal) agents may be used to slow down intestinal transit time and allow food to spend more time in the intestine, thus allowing more opportunities for nutrient absorption.
- Medicines to decrease acid production in the stomach are used because high acid content that reaches the intestine can make diarrhea worse.
- Antibiotics are selectively used if bacterial overgrowth is suspected.
- Vitamin/mineral supplementation and electrolyte repletion as needed

Surgery: In select patients, surgery can be used to lengthen the short intestine, taper dilated intestine, or slow down the movement of intestinal contents. Your surgeon will determine the best operation for your child. Some options include:

- **Longitudinal intestinal lengthening and tailoring (LILT) procedure** is also known as the "Bianchi" procedure. Originally described in the 1980s, LILT procedure can only be done once per patient. Currently, this procedure is rarely performed.
- **Serial Transverse Enteroplasty Procedure (STEP)** is an intestinal lengthening procedure which is now the preferred intervention at most SBS centers because it can be repeated or used after a prior Bianchi procedure. The STEP increases intestinal length and improves intestinal absorptive ability by using a special stapler to make intermittent transverse cuts in the intestinal wall that can then be stretched.
- **Interposition reversed intestinal segmentation** is a procedure where a section of intestine is removed and then connected in the reverse direction. This leads to decreased nutrient transit time with peristalsis (intestinal contraction) occurring in the opposite direction through that segment.

Combined small intestine/liver transplantation is indicated in patients who have irreversible intestinal failure-associated liver disease. Five-year survival rates are greater than 75% if a suitable donor can be found. Possible complications include graft rejection (condition where the transplant fails) and sepsis (infection) and patients require lifelong immunosuppression therapy.

Informed consent: A consent form is a legal document that states the tests, treatments or procedures that your child may need and the doctor or practitioner that will perform them. Before surgery, your doctor should tell you what the operation is, the goal of the surgery and other possible treatment options that are available. Your doctor should explain the risks and benefits of the surgery. You give your permission when you sign the consent form. You can have someone sign this form for you if you are not able to sign it. You have the right to understand your child's medical care in words you know. Before you sign the consent form, make sure all your questions are answered. It is important to know that during surgery, there are things that can happen that your doctor may have not predicted before going in. He or she will explain these to you after the surgery.

Home Care - "What do I need to do once my child goes home?"

Your child will have very close follow-up with the surgeon and gastroenterologist. There will also likely be a nutritionist who specializes in short bowel syndrome. Many of these clinicians work together in one group to help streamline your care. You will be given specific home instructions based on your child's needs. In general, call your surgeon if your child has a fever, decreased urination, belly pain, vomiting, or redness/draining from any wounds.

Long Term Outcomes - "Are there future conditions to worry about?"

Long-term use of TPN can lead to liver disease, which is a major cause of complications and death associated with SBS.

Depending on the original cause of your child's SBS, how much intestine is left, whether or not the valve between the small and large intestine (ileocecal valve) remains, the condition can improve, with or without surgery, as the intestine "adapts" and improves its ability to digest and absorb.

In most cases, babies start with a combination of TPN and feeds through the gut (by mouth and/or by G tube). Slowly increasing the food that the intestines process can make the intestines adapt and rehabilitate.

If no progress is made with advancing feeding through the gut, then intestinal lengthening operations are considered.

If all medical and surgical options have been tried and no progress is made with advancing on feedings through the gut, a transplant may be considered.

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