Living with Short Bowel Syndrome

Information for Parents and Caregivers
of Children with Short Bowel Syndrome

Dear Parents and Caregivers,

You probably came across this booklet because your child or a child in your care has **short bowel syndrome with chronic intestinal failure**. This booklet is intended to help you better understand what this diagnosis means for your child's day-to-day life and what you can expect going forward.

Whenever someone is diagnosed with a severe disease, patients and their loved ones usually have many questions, especially: How will this condition affect my child, my family, and me? How can we learn to live with this condition? Who can help us manage our day-to-day lives? Where do we go from here?

It is not possible to answer all of these questions in one single booklet. But hopefully we can help guide you toward the answers by providing some basic information in a concise and easy-to-read manner. Feel free to come back to this booklet whenever you want or need to learn more about a specific topic in order to better help your child manage her or his condition. This booklet is also designed to help you prepare for the many conversations you will likely be having with your healthcare team in the near future.

This booklet contains:

+ Information about **short bowel syndrome**
+ **An introduction** to digestion and the bowel
+ **Real-life stories** from children with short bowel syndrome
+ Links to more information and **support resources**
+ A **Glossary** explaining the most important terms (written in italics in the booklet)
+ A **Checklist** for conversations with your healthcare team

We recommend you read this booklet all the way through at first to familiarize yourself with short bowel syndrome. If you are just interested in one specific topic, you can find it in the **Table of Contents**. The **Glossary** is designed as a reference in case you come across words you don't understand while reading other resources. Use the **Checklist** to prepare specific questions for your next doctor's appointment.

Table

What is short bowel syndrome?

Short bowel syndrome with chronic intestinal failure is a complex and sometimes life-threatening disease. Its name provides us some clues about its nature: the term “bowel” is another word for the intestines. When the **intestines are abnormally short**, they can lose the ability to carry out their vital function of delivering nutrients, minerals and fluids to the body – a situation called intestinal failure. Because having a short bowel is a persistent condition, the disorder it causes is chronic, or long-lasting.

With short bowel syndrome (SBS), the intestines are no longer able to digest nutrients eaten by mouth or taken by feeding tube enough to keep the body health and allow a child to grow. Instead, a type of nutrition called *parenteral support* is needed, which means that nutrients and fluids are provided through a thin tube called a *catheter*. This *catheter* delivers nutrition directly into the bloodstream rather than it being absorbed by the digestive system. *Parenteral support* can be used to replace some nutrients and fluid, or all of it when given in a form called *total parenteral nutrition* (TPN).

Short bowel syndrome can be caused by a number of different conditions. Its impact is determined by several factors, including:

+ How much intestine is missing,
+ What part(s) of the intestine are missing,
+ How old someone is when SBS starts, and
+ What the underlying cause of SBS is.

Your child may experience different symptoms and complications depending on what type of SBS they have and what originally caused it. Diarrhea and fluid loss are common problems at first, and often require extra medications in addition to *parenteral support*.

Even though short bowel syndrome is **a chronic (meaning long-term) condition**, that doesn't mean it will always stay the same. The digestive system is able to **adapt to a short bowel** over time, especially in children. **Medications** can help this process in some cases.

In many newborns with short bowel syndrome, their condition improves so much during the first year that they can go for extended periods or even entirely without *parenteral support*. The chances of becoming completely independent of *parenteral support* go up when the child's healthcare team develops a patient-specific, integrated care plan that balances nutritional, surgical and medical management. On the other hand, the likelihood that the bowel can adapt to normal food by itself decreases the older the child gets.

Your child's healthcare team will talk with you about how short bowel syndrome will affect your child, what you and your child can expect, and the outlook for your child's condition.

Short bowel syndrome is often abbreviated as *SBS*, while chronic intestinal failure is abbreviated as *CIF*.

How does the bowel work?

The human intestine, or bowel, is a tube-shaped organ that is divided into the small intestine and the large intestine.

The **small intestine** is the longer segment. It is 5 to 6 meters long in adults, and can be as long as 2 to 3 meters in healthy infants. The small intestine lies between the stomach and the large intestine in the abdomen and winds back and forth as shown in Figure 1. It is divided into 3 segments: the *duodenum,* the *jejunum* and the *ileum*. While these segments have slightly different functions, they are all key sites for absorbing vital nutrients from food and drink and supplying them to the body through the bloodstream. Although it may look like a smooth tube in pictures, in reality the lining of the small intestine is made up of many small folds that form finger-like structures called *villi*. On top of the villi are even smaller hair-like structures called *microvilli* that give the surface of the small intestine a brush-like appearance. All of these folds and bulges increase the surface area of the small intestine tremendously: in fact, it is 100 times larger than the surface of the skin! In people with a healthy small intestine, this large surface is what makes digestion possible.

The **large intestine** (also called the *colon*) is the shorter segment of the bowel. It is about 1.5 meters long and is located downstream of the small intestine. At the end of the large intestine is a segment called the *rectum*, which is the final part of the bowel and connects to the *anus*. Between the small and large intestines is a gate called the *ileocecal valve* that prevents the contents of the large intestine from flowing back into the small intestine. Although Figure 1 on the next page depicts the large intestine as a series of small segments, in reality these are not fixed units. Digested food forms clumps in the large intestine that are slowly pushed through as separate portions. As this happens, water and minerals are extracted from these clumps, making them ever more dense until they finally form the stool which is passed from the anus as a bowel movement.

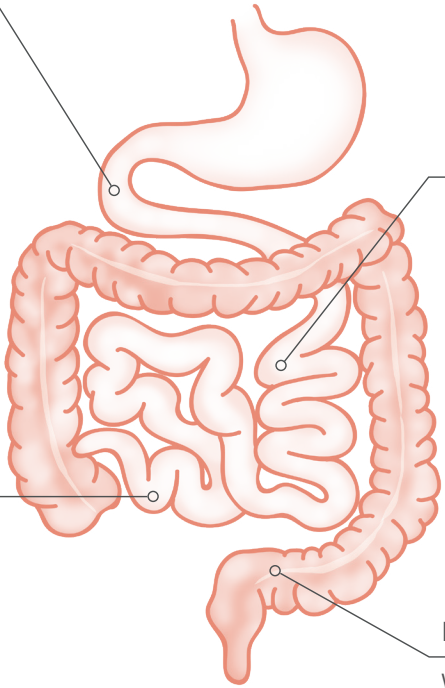
Figure 1 shows the structure of the small and large intestines and lists which nutrients are primarily absorbed in each segment. See the next section on digestion for more information.

Which segments of the intestine absorb which nutrients?

A selection of nutrients absorbed in the bowel is shown below.

Duodenum
First part of the small intestine

Fat, sugar, *protein*, iron, folate, calcium, trace elements, water



Jejunum
Middle part of the small intestine

Sugar, iron, calcium, *protein*, trace elements, fatty acids, fat-soluble vitamins, water

Ileum
Last part of the small intestine

Bile salts, vitamin B12, trace elements, electrolytes, water

Large intestine

Water, trace elements, *electrolytes*, *amino acids*, short and medium chain triglycerides, calcium

Figure 1

How does digestion work?

Digestion is the process by which the **food we eat** is broken down and supplied to the body for it to use as **energy** and **building blocks**. Our bodies need energy to carry out all of the processes important for living. The body is continuously building new cells and tissues, not only during childhood but across our entire lives since tissues are constantly being regenerated.

Digestion starts in the **mouth** when large bites of food are broken up by chewing and mixed with saliva. Saliva itself contains *enzymes* that are involved in the chemical reactions of digestion. The tongue then pushes a small lump of food into the back of the mouth. There, it presses against the gums, triggering a swallowing reflex that propels the food through the throat and into the stomach.

Once in the **stomach**, the lump of partly-digested food is mixed with stomach acid containing even more *enzymes*, transforming it into a soft mass. The muscles of the stomach then grind this mass further and gradually supply it to the **small intestine**, which is where the majority of digestion actually takes place. There, the individual nutrients from the food are absorbed into the bloodstream, which delivers them throughout the body.

The most important nutrients in the diet are energy sources like f**ats, *carbohydrates* and *proteins***, as well as ***essential amino acids*, fatty acids, vitamins, minerals** and ***trace elements***. All of these nutrients play crucial roles in metabolism, growth and tissue *regeneration*. Without them, life is not possible. And when digestion is restricted by a medical condition, some or all of the required nutrition must be supplied by a different route.

Once most of the nutrients have been absorbed in the small intestine, the mass of digested food passes into the **large intestine**. There, water as well as stomach acid and other substances produced by the body to aid digestion are absorbed back into the body. This turns the partly-digested food more solid, and allows the remaining undigested food to be eliminated from the body as stool. Stool also contains a great deal of dead **bacteria from the digestive tract**, which usually live in the small and large intestine and help promote digestion in healthy people.

Although this explanation of digestion is only a basic overview, it still illustrates how complex this process is. In order for nutrients to enter the body, it is not enough to eat the right amount of high-quality food: all of the digestive organs also need to work the way are supposed to! The body has to produce *enzymes* and stomach acid, and the right bacteria have to thrive in the intestines. There are a large number of medical conditions that can disrupt this complex process – including the rare and very severe condition of short bowel syndrome which is the topic of this booklet.

What causes short bowel syndrome?

Short bowel syndrome (SBS) can be caused by a number of different conditions. It can start at any age, including newborns, children and adults. The causes can include **conditions present at birth, illness** or even **trauma** when these conditions then require a section of the intestine to be surgically removed. SBS can have a wide range of severity and outlook depending on how much intestine was removed or is no longer working correctly, and at what age. This booklet will briefly describe some of the most common causes of SBS in newborns and children. Short bowel syndrome is a rare disorder in people of all ages. In infants, it is most frequently the result of ***necrotizing enterocolitis*** **(NEC)**, a condition when the lining of the intestinal wall dies in newborns (usually premature newborns).

Figure 2 shows some of the main causes of SBS in children and newborns, which will be described briefly in this section.

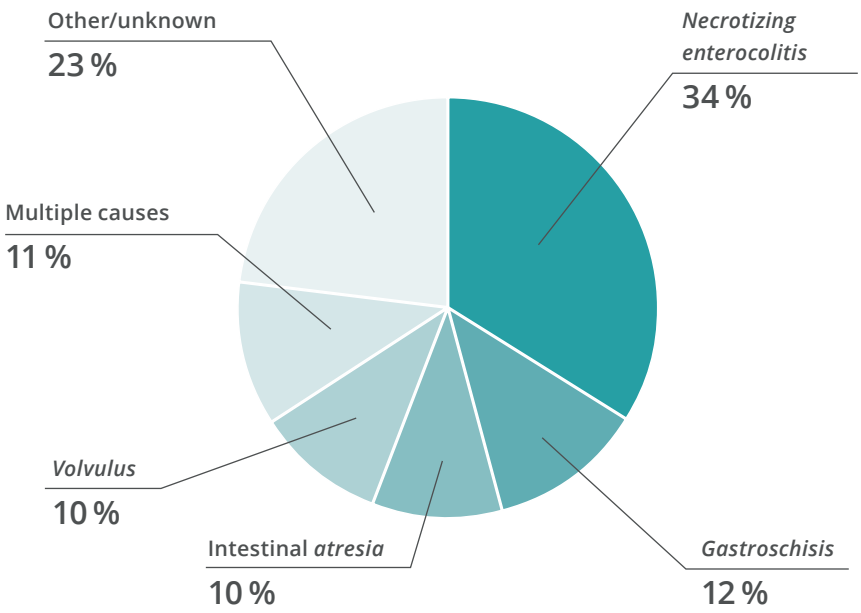


Figure 2

Necrotizing enterocolitis (NEC)

Necrotizing enterocolitis (NEC) is a type of inflammation at certain spots or along long stretches of the intestinal wall that causes intestinal tissue to die. Because the intestinal wall is no longer intact, bacteria and partly-digested food can escape into the abdomen, which can lead to a life-threatening situation called *sepsis*. About 1 in 1,000 newborn children develop NEC, especially premature babies and newborns with low weight at birth. Because NEC can be fatal in nearly one-third of all cases, surgery is often performed to remove the affected part of the intestine. This can result in SBS depending on how much of the intestine was removed in order to save the child's life..

Gastroschisis

Gastroschisis is a hole in the abdominal wall that develops before a child is born. During *fetal* development, the intestine passes through this hole in the abdomen and develop outside the body. There, it can become tied in knots, leading to blockage and possibly cutting off the blood supply to parts of the digestive tract. The unprotected part of the intestine is also prone to infections, inflammation and injuries. This type of birth defect can usually be detected during pregnancy, even though its cause is still not known. *Gastroschisis* is a very rare birth defect with a rate of 3 per 10,000 newborns. According to one study, 13% of children born with *gastroschisis* require surgical treatment, which may lead to SBS.

Malrotation and volvulus

Malrotation refers to a situation when the intestines in a developing fetus **do not rotate into their proper position**. It is not known what causes this to happen. Intestinal *malrotation* by itself is not always a problem and some people do not experience any symptoms. However, it can also lead to a very serious condition called ***volvulus,*** where the intestine twists around itself. This twisting can compress and block the intestine and prevent it from working properly. It can even cut off blood flow, causing the intestinal tissue to die. While a *volvulus* can occur anywhere along the bowel, it most often affects the small intestine in children. *Volvulus* is usually treated by surgery, often in an emergency operation that returns the intestine to a normal position. If the injury to the bowel is too great, it may be necessary to remove the damaged part of the intestine completely. This bowel removal can then create a risk of developing SBS.

Atresia and stenosis

The main causes of intestinal issues present at birth are **closure** *(atresia)* and **narrowing** *(stenosis)* of the intestines. These problems result from abnormal *feta*l development and can cause food to become stuck in the intestine and unable to pass through. Over time, this 15 blockage can cause the intestine to expand upstream of the closure. Newborns with intestinal *atresia* or *stenosis* frequently vomit up food they have just eaten. *Atresia* and *stenosis* can only be treated by surgery, which usually removes the unhealthy sections of the intestine. Once again, removal of part of the bowel can lead to SBS.

Other causes

Other causes of SBS in children include cancer or accidents which require large sections of the intestine to be removed by surgery.

Another condition that can cause SBS is Hirschsprung's disease, in which certain nerve cells to part of the intestine are missing. These nerve cells control *peristalsis*, which is the coordinated movement of the muscles around the intestine that propels digested food and stool forward through the gut. Like many other causes of SBS, Hirschsprung's disease is treated by removing the unhealthy part of the intestine, in this case where peristalsis is not working. SBS can develop if the section removed is too large.

Nutrition in short bowel syndrome

As shown in Figure 1, different segments of the bowel are specialized to absorb different nutrients. The table below provides a brief overview of which foods contain these nutrients and what roles they play in the body. Since this table can only serve as a rough guide, please talk with your healthcare team about your child's nutrition.

Nutrient	Major sources (examples)
Fats	Animal fats: butter, cream, fat-rich meat/bacon, fat-rich types of fish Plant oils: oils, margarine, avocado
Medium chain triglycerides (MCT)	Coconut oil, palm oil, special products for high-MCT diets
Carbohydrates (sugar)	Table sugar, honey, as well as starch breakdown products (potatoes, bread, rice, pasta), fruit, juices
Protein	Animal protein: meat, fish, eggs, yogurt Plant-based protein: beans, peas, tofu, oats
Iron	Animal sources: liver, some types of sausage Plant sources: some nuts, some types of mushrooms, spinach, chickpeas
Calcium	Animal sources: milk and dairy products Plant sources: broccoli, spinach, kale
Folate	Animal sources: liver, egg yolks Plant sources: wheat germ, lettuce, spinach, broccoli
Cholesterol	Animal sources: fat-rich meat, offals Plant sources: seed oils (corn oil, pumpkin seed oil)
Vitamin B12	Animal sources: dairy products, liver Plant sources: not found
Trace elements	Iodine, zinc, selenium, iron, etc.: various sources

Parenteral support

Patients with short bowel syndrome may need a type of *parenteral support* called *total parenteral nutrition* (TPN) as a short-term or long-term treatment, depending on how severe their condition is and what digestive functions are affected. *Total parenteral nutrition* ensures that the body receives all of the nutrients and fluids it needs. Unlike normal eating by mouth, *parenteral support* is not taken at fixed meals but is given all day long, or sometimes only at night. *Parenteral support* uses special mixtures that are tailored to each individual child and need to be updated regularly.

The role of eating in parenteral support

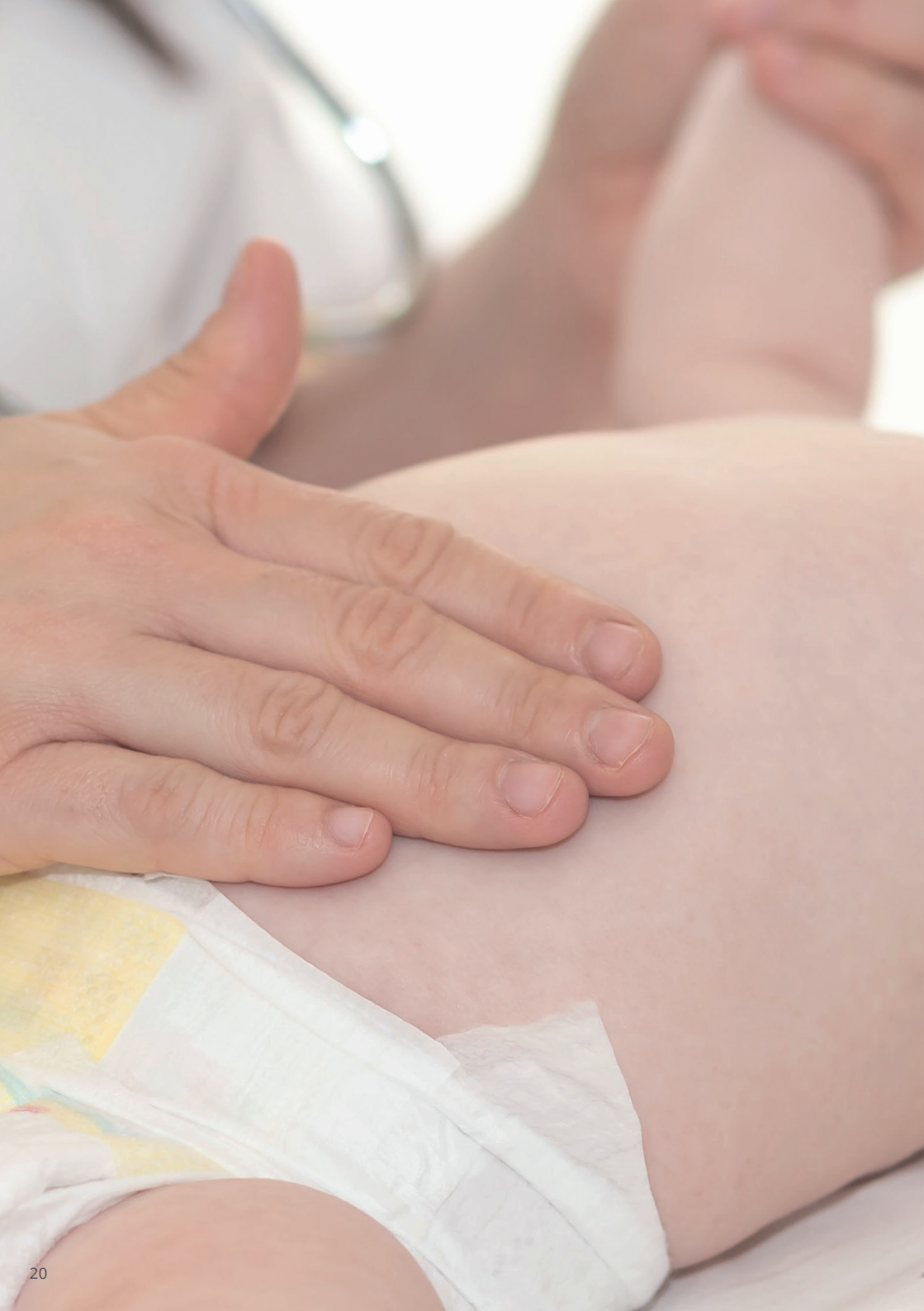
Many patients with SBS still have enough intestinal function to get some of their nutrition from eating normally or from feeding tubes. While *parenteral support* through a *catheter* in a vein is still the main source of nutrition, eating additional nutrition by mouth can help maintain and sometimes even increase the ability of the remaining intestine to absorb nutrients. The aim is to meet as much of the child's nutritional needs this way as possible. Engaging the digestive tract can also train the large intestine to take over some of the roles of the missing part of the small intestine. These beneficial effects may allow the amounts of fluids and nutrients in *parenteral support* required to be reduced, which also lowers the risks of complications. One of the risks with TPN is that the lining of the small intestine may shrink when not used, further decreasing the surface available for digestion and absorption of nutrients. Keeping the *colon* trained (when it is still present) also helps prevent painful constipation once the child is again able to eat more.

Children who received *TPN* at birth, or for longer periods later in life, may not feel the sensation of hunger as often as other children, or may not interpret the sensation of hunger correctly. Infants and toddlers who have eaten little to nothing by mouth for most of their lives may develop abnormal eating patterns since they have never learned "normal" eating habits. Ask your care team for advice and don't hesitate to request counseling by trained and experienced mental health professionals.

Children on *parenteral support* who also eat some food by mouth or feeding tube must plan their nutrition carefully and adjust it regularly. Your care team can support you with this task. Some general tips include:

+ Eat small but frequent meals (about every 2 hours)
+ Always eat slowly; encourage older children to chew well
+ Each meal should contain fats (you may need to determine which fats your child can tolerate and buy special groceries)
+ SBS patients tolerate fruit and vegetables better when cooked or blended (like applesauce)
+ Eat lots of meals with potatoes and noodles
+ Pay special attention to the child's food intolerances (like lactose intolerance)

Eating is a central activity in life and is closely linked to quality of life and physical well-being. Even healthy children often have issues when it comes to eating, so don't be discouraged if it takes a great deal of empathy, patience and knowledge on your part to find the "right" nutrition for your child with SBS. Instead, seek out the advice of a dietician or other nutritional specialist, as they can play a key role in managing your child's condition.



Symptoms and complications

Chronic malabsorption: Healthy development requires sufficient nutrients

When one or more sections of the bowel are missing or do not function properly, the body cannot receive the nutrients that would normally be supplied by those sections – a condition called *malabsorption*. The bowel also cannot absorb healthy volumes of fluids when itis abnormally short, raising the risk of *electrolyte* imbalance. In order to compensate for this lack of nutrients and fluid, SBS patients require *parenteral support*, sometimes even *total parenteral nutrition*.

Signs and symptoms of chronic *malabsorption* include:

+ Frequent, runny **diarrhea**
+ Constant *dehydration*, meaning **not enough fluids in the body**
+ **Weight loss** or insufficient weight gain during growth phases
+ **Growth and developmental delays**
+ **Bone density issues and fractures**
+ **Apathy** and depression
+ **Wound healing** issues

Short bowel syndrome with chronic intestinal failure is a complex condition in children

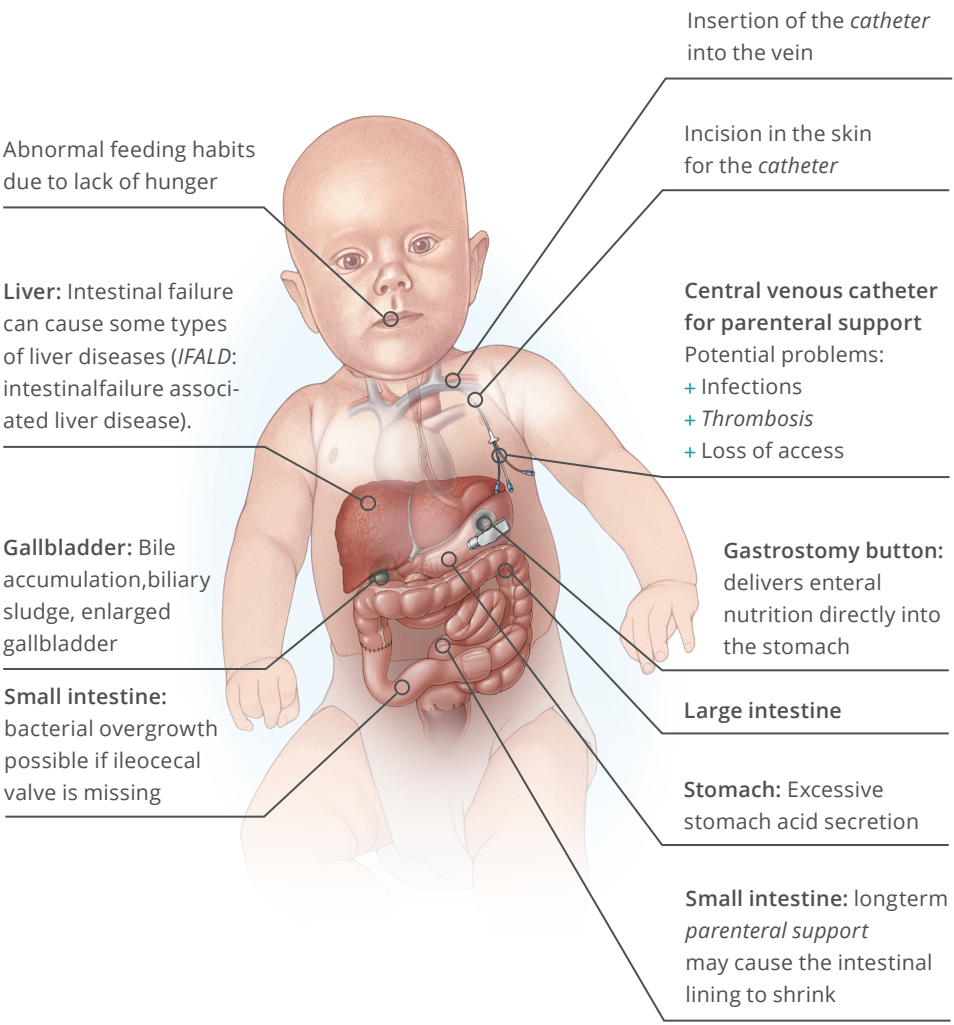


Abbildung 3

Chronic *malabsorption* is a state of ongoing nutrient deficiency. When sections of the bowel are removed, some or even all nutrients are no longer absorbed at sufficient levels. The direct impacts of *malabsorption* include diarrhea (especially when the large intestine has been removed), *dehydration*, weight loss as well as growth and developmental delays. There are also some indirect effects that can lead to complications of short bowel syndrome. Some of these complications are related to the *catheter* that must be inserted and left in place for long-term *parenteral support*. This *catheter* can trigger *thrombosis* (blood clots) or become infected, which can then lead to sepsis. *Parenteral support* can also cause problems in the liver, gallbladder and kidneys. People with no ileocecal valve are prone to bacterial overgrowth of the small intestine. Other complications include *enterocolitis*, intestinal blockage and slowed transit of food.

The combination of an underlying disease and its complications can have a major impact on patients' quality of life. Children with SBS often have to battle not only their physical symptoms but also emotional issues. These issues stem from both their physical condition as well as the resulting difficulties in participating in social activities at kindergarten, school and in other groups. Disrupted digestion and fluid imbalance can make toilet training a challenge. Because SBS breaks the normal links between hunger, eating, feeling full, digesting food and going to the bathroom, infants and toddlers often have problems feeding while older children have difficulty eating without support. Older children may also be embarrassed about the visible signs of their condition. And of course parents worry about the well-being of their child, and are often worn down from the extra care they need to give their child.

Parenteral support: A blessing, but also a curse

There is no question that *parenteral support* is a blessing for anyone with SBS. It is quite often the only short-term or long-term option for providing vital nutrients and fluids to the body and ensuring patients' survival. However, these benefits also come with their own drawbacks.

+ Because *parenteral support* is delivered through a **central venous *catheter***, a foreign object (the *catheter*) is in continuous contact with the bloodstream. This poses an ongoing risk of infections that require antibiotics and could even become life-threatening.
+ **Blood clots** (*thrombosis*) can form in the *catheter* and in the vein the *catheter* is placed in. These clots block the vein and can make access to it difficult or even impossible.
+ The nutrient solutions and fluids used must be measured precisely and continuously adjusted to the child's needs. This requires close coordination within the child's healthcare team.
+ The equipment required for *parenteral support* must always be nearby, even when the child is not at home. As a parent or caregiver, you will need to learn how to manage *parenteral support* and ensure a continuous supply of nutrition. This will be a difficult task at first.
+ Children who receive most of their nutrition from *parenteral support* might not feel the sensation of hunger, which can lead to abnormal feeding habits in infants.

Medications for the symptoms of SBS

Most of the symptoms described in previous sections can (and sometimes must) be treated with medications. The table below provides a brief overview of which drugs may be prescribe and what effects they have. Some of these medications are intended for long-term use while others will only be prescribed for acute issues. Medications also require close coordination within your child's healthcare team, since the doses of each medicine will need to be adjusted often. Your team will explain to you how to give your child each drug, what to look out for, and what to do if a dose is missed.

Medications	Purpose
Antibiotics	Stop the growth of bacteria in an infection
Antidiarrheal drugs	Used to treat diarrhea
Stomach acid blockers	Reduce stomach acid production, fluid loss and diarrhea
Gallensalzbinder	Wirken gegen von der Galle ausgehenden Durchfall
Bauchspeicheldrüsenenzyme	Improve the cleavage of fats and prevent fatty stool
Hormone therapy	Can improve the ability of the bowel to absorb nutr

Living with short bowel syndrome

Just as short bowel syndrome can take a wide range of forms, there is also a wide range of journeys among the people living with SBS. Here are a pair of stories from Lilly and Friedrich, two children who have been living with the condition since birth. Both stories are proof that a full life is possible with SBS, despite all of the hurdles, challenges and concerns it presents.

Lilly's story

Lilly is 6 years old and has been living with SBS her entire life. She was diagnosed with ***gastroschisis*** along with a ***volvulus*** while she was still in the womb, and underwent her first surgery only 2 days after she was born. After this operation, Lilly was left with only about 70 centimeters of her small intestine and 20 centimeters of her large intestine. This is much less than half of what would be normal in a healthy child of her age.

Lilly needed additional operations over the coming months and years. After her small and large intestine were reconnected, multiple *STEP* operations were performed. *STEP* stands for serial transverse enteroplasty, and is used to lengthen the intestine. In this procedure, a number of small cuts are made in the intestine in a zig-zag pattern and then sewn back together, which slows the transit of food through the bowel and allows more nutrients to be absorbed.

Her *Broviac catheter* had to be re-inserted several times to maintain *parenteral support*, and she has had to deal with numerous infections and leaks stemming from the *catheter*. In addition to her *catheters*, Lilly also has a gastrostomy button (sometimes called a G-button), which is a short feeding tube that leads directly into her stomach. She has to battle bacterial overgrowth in her small intestine from time to time. Lilly's delayed growth is also apparent just by looking at her: at 108 centimeters tall and weighing just 16 kilograms, she is noticeably shorter and lighter than the other children in her kindergarten group.

Friedrich's story

Friedrich's story also begins before he was born, when doctors diagnosed a ***stenosis*** in his small intestine that may have been triggered by an infection while his mother was pregnant. Friedrich's first operation was performed 1 day after being delivered by Caesarean section in order to clear the narrow section of his intestine. His doctors hoped his bowel would be able to develop normally after this procedure, but it turned out to already be too damaged. His mother had to pump the milk she fed him back out of his stomach since it simply refused to travel down the rest of the digestive tract. He then needed a second operation which also removed part of his large intestine. So much of his bowel had been removed by this second round of surgery that he developed SBS and required *total parenteral nutrition* 24 hours a day.

Unfortunately, Friedrich's TPN didn't start well. He lost a lot of fluids, especially at night. These issues kept Friedrich and his mother in the hospital until he was 6 months old. After that he was able to go home, where a home care team helped him take his TPN through his *Broviac catheter*. Despite this professional support, Friedrich was prone to infections that often required him to return to the hospital. One factor in these infections appeared to be repeated blood draws through the *catheter*, which were taken at a clinic without a great deal of expertise in TPN. Despite these setbacks, Friedrich was still able to enroll in an inclusive day care program at the age of two and a half. Since entering day care, he has made great progress: learning how to speak with the help of a speech therapist, then discovering how to make friends with people outside of his family.

Although he has undergone several *STEP* operations to lengthen his bowel, Friedrich (now in the third grade) still requires *parenteral support*. Even his medical specialists at the university hospital can't explain why he still needs *parenteral support*, which just shows how complex the bowel can be. Despite needing *parenteral support*, Friedrich lives a full life: he goes to school where he is assisted by developmental therapists, he learns judo, he likes breakdancing, and he is now learning to play the drums. Managing his *catheter* is just as routine for him as brushing his teeth. Even though he doesn't feel hungry and occasionally needs to be reminded to eat, he has learned to enjoy steak at family dinners. His dedicated parents are optimistic that he will be able to make up any remaining developmental setbacks in the coming years.

Finding help and support

Online information and connection

Being able to find and talk with people who are dealing with similar problems can be very helpful when facing a chronic condition. Because SBS is such a rare disorder, you might not know anybody nearby who has it. There are several online platforms for finding and connecting with others: for example, visit **leben-mit-kds.de** to discover a wide range of information and inspiring stories from other people with SBS. Join the Facebook community **"Leben mit KDS"** (https://www.facebook. com/lebenmitkds), an initiative of Takeda Pharma Vertrieb GmbH & Co. KG, to read the latest about living with SBS and to connect with other SBS patients.

Specialized centers

Because SBS is so rare and yet so complex, it is important to seek medical care from specialists with experience treating the condition. This can be in addition to your local healthcare team if there is no specialized center in your area. Visit **www.leben-mit-kds.de** to find a list of centers in Germany that specialize in treating and caring for children with SBS.

Scan this QR code to jump directly to the list.

Self-help groups

A severe illness places a heavy strain on an entire family – especially when the patient is a child. While medical care from your healthcare team, dieticians and further specialists is essential, connecting with other families going through a similar situation can also provide comfort and community. Ask your doctors whether they can point you to a self-help group.

Glossary

Term	Definition
Amino acids	Amino acids are the building blocks the body uses to make proteins. When proteins are eaten in food, they are broken down by the digestive system into amino acids.
Atresia	Atresia refers to a condition in which the passage in a hollow organ like the bowel is closed or blocked. This condition can be congenital or acquired.
Broviac catheter	A long-term catheter that is placed into a large vein, such as the vein below the collarbone, and is frequently used for parenteral support in children since it is less prone to infection and is less likely to slip.
Carbohydrates	A group of biologically-important substances including sugars, starches, glycogen and cellulose. Carbohydrates are important sources of energy in the diet.
Catheter	A catheter is a thin tube than can be inserted into blood vessels and other hollow organs in the body. Parenteral support is delivered through a catheter that is inserted into a vein.
CIF	*Abbreviation for "chronic intestinal failure".*
Colon	Another name for the large intestine
Congenital	A congenital disorder is one that is present at birth. It can be caused by genetics or by certain events or conditions during pregnancy.
Dehydration	When the body does not have enough fluids, either because a person did not drink enough liquids or loses liquids too quickly, for example by diarrhea.
Duodenum	The first segment of the small intestine downstream of the stomach.

Term	Definition
Electrolytes	Electrolytes are substances which have an electrical charge when dissolved in a liquid (like blood). The most important electrolytes in the human body are sodium, potassium and magnesium and are required for many metabolic processes.
Enteral	The term enteral means "in the gut" and is used to describe any type of nutrition taken in through the stomach and/or intestines. For SBS patients, it often refers to enteral nutrition (see below).
Enteral nutrition	While enteral nutrition technically means any type of nutrition that goes through and is absorbed by the bowel, including food eaten by mouth, in the setting of SBS treatment it usually refers to nutrition delivered by a feeding tube (sometimes called a gastrostomy button or G-button) directly into the stomach.
Enterocolitis	Inflammation in the digestive tract.
Enzymes	Enzymes are a type of protein that carry out a wide range of functions in the body by accelerating biochemical reactions. During the digestive process, enzymes from the human body and from bacteria living in the bowel play important roles.
Essential amino acids	These are amino acids that the human body needs but cannot make by itself and therefore must obtain from the diet. Examples of essential amino acids: isoleucine, leucine and lysine.
Fetal	A fetus is another term for a baby as it develops in the womb of a pregnant mother.
Gastroschisis	A congenital hole in the abdominal wall
Hormone therapy	Treatment with hormones that are normally produced by the intestine may help reduce the need for parenteral support.

Term	Definition
IFALD	Abbreviation for "intestinal-failure associated liver disease", an occasional complication of SBS.
Ileocecal valve	The ileocecal valve acts as a gate between the small and large intestines, preventing partly-digested food from flowing back into the small intestine. This is important because the bacteria that live in the colon should not enter the small intestine.
Ileum	The last segment of the small intestine before the large intestine.
Jejunum	The middle segment of the small intestine.
Malabsorption	When nutrients are insufficiently absorbed in the bowel.
Malrotation	When the small and/or large intestine do not rotate into their proper position during fetal development.
Microvilli	Hair-like protrusions that help expand the surface area of cells in the intestine, improving the exchange of substances into and out of the gut.
Necrotizing	Necrotizing refers to the death of a small area of tissue.
Oral	When something is eaten or taken by mouth.
Parenteral	Parenteral literally means "to bypass the intestines". Parenteral nutrition bypasses the digestive tract by being delivered directly into the bloodstream.
Parenteral support	Parenteral support refers to any type of nutrition delivered through a catheter into a vein, and thus directly into the bloodstream. This nutrition can be a partial replacement of enteral nutrition, for example fluids or some nutrients, or it can be a way to supply a person's complete nutritional needs (see *Total parenteral nutrition*).

Term	Definition
Peristalsis	The coordinated movement of muscles around the intestines to propel partly-digested food through the digestive tract.
Proteins	Proteins are complex biological molecules made of amino acids which serve many functions in the body, including as enzymes and structural molecules. In the diet, they can also serve as a source of energ
Rectum	The last segment of the large intestine upstream of the anus.
SBS	*Abbreviation for "short bowel syndrome".*
Sepsis	A life-threatening situation which can occur when bacteria or other microorganisms get into the bloodstream.
Stenosis	An abnormal narrowing in a hollow organ such as the bowel or a blood vessel.
STEP	STEP is an abbreviation of "serial transverse enteroplasty", and is a surgical procedure used to slow down the transit of food through the bowel by reshaping the intestine in a zig-zag pattern.
Thrombosis	A blood clot.
Total parenteral nutrition	When a person's entire nutritional needs are delivered by catheter directly into the bloodstream.
Villi	Villi are finger-like folds in the lining of the small intestine. They are densely packed inside the small intestine, which expands the surface area of the intestine a great deal.

Checklist for doctors' appointments

Short bowel syndrome is a complex condition, and there will always be many topics to discuss at each visit with the doctor or other members of your healthcare team. It can be helpful to prepare for these conversations by writing down a few questions ahead of time and taking notes during the visit itself. The following pages provide some sample questions you might like to ask your doctor.

You might also find it helpful to bring along another adult relative or good friend to help you process the wide range of information discussed with the doctor.

Some important questions might include:

+ What segments of the bowel are affected?

+ What type of nutrition will my child need: is *parenteral support* necessary?

+ What is the expected outlook? Is there a possibility that our child's intestines will recover enough to be free of *parenteral support*?

+ How often will our child need to see the doctor/go to the hospital? Where can families who live far away spend the night?

+ Should we expect a need for more surgery?

+ Does our child need a *catheter*? If yes, how will it be maintained?

+ Is there a home care service that can support us?

+ What medications does our child need and how are they taken?

+ What signs or complications do we need to look out for? When is it okay to call the doctor's office, when is urgent care needed, and when do I need to call an ambulance?

+ Do you know of any self-help groups or other resources for families of SBS patients?

The information presented here does not replace a medical consultation.
Should you require in-depth consultation, please speak with your doctor.

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Takeda SBS

Short bowel syndrome: info for kids!

Takeda Pharma Vertrieb GmbH & Co. KG
Potsdamer Str. 125
10783 Berlin
www.takeda.de

EXA/DE/SBS/0092_07.2022