Short Bowel Syndrome (Paediatrics)

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Short bowel syndrome (SBS) is characterized by a decrease in the functional area of the intestine below the minimum required for digestion and absorption . Absorptive surface area of the intestine is reduced due to shortening of the length of the intestine, usually by massive resection of the small intestine. The severity of malabsorption also depends on which part of the intestine is missing. The most common cause of SBS in children is necrotizing enterocolitis and congenital intestinal defects such as volvulus , atresia , gastroschisis or aganglionosis . Among the most common causes in adults is Crohn's disease, trauma, malignancy, radiation and mesenteric ischemia. Among the negative consequences of SBS are excessive losses of fluids and electrolytes, inability to absorb enough energy, macronutrients (proteins, carbohydrates, fats), vitamins and minerals, failure to improve weight and growth.[1]

Intestinal failure is a condition where a malfunction of the digestive tract leads to a lack of nutrients and insufficient hydration of the body, therefore it is necessary to administer intravenous or enteral supplementation. SBS is the most common cause of chronic intestinal failure.[1]

Reduction of mucosal brush-line hydrolases, which are responsible for carbohydrate digestion and absorption, leads to carbohydrate malabsorption, energy losses in stool, and osmotic diarrhea. As a result of hypergastrinemia, there is an increased secretion of gastric acid and subsequently a decrease in the pH of the intraduodenal fluid, which inactivates pancreatic enzymes and worsens the malabsorption of fats and proteins. Carbohydrate malabsorption, absence of an ileocecal valve, and altered intestinal motility facilitate **bacterial overgrowth** . Resection of certain parts of the intestine leads to a deficiency of certain nutrients (terminal ileum - vitamin B12, proximal small intestine - iron, calcium and magnesium)[2]

The following factors affect bowel function:

- length of bowel resection (relative to age or body size);
- loss of ileum and ileocecal valve;
- loss of all or part of the colon;
- continuity vs. bowel incontinence.[1]

Further clinical course and nutritional management of SBS depend on the length of the remaining bowel and its function (proximal x distal small bowel).

Pathophysiology

Intestine length

In a full-term newborn, the small intestine measures about 240 cm and the colon about 40 cm. The length of the jejunum, ileum and colon doubles during the third trimester of pregnancy, so premature newborns have hope for a better outcome. At 1 year of age, the length of the small intestine is 380 cm on average. For premature newborns, nomograms are compiled based on the work of Walker-Smith and Touloukian: total length of small + large intestine in 19-27. week of gestation is 142 ± 22 cm and after the 35th week of gestation 304 ± 44 cm.[3][4][5] The length of the small intestine is about 70 cm (SE 6.3) at 24 to 26 weeks' gestation, 157.4 cm (SE 11.2) at term, and 423.9 cm (SE 5 ,9) at 49 to 60 months of age.[6]

A good clinical outcome is considered a condition where enteral nutrition covers all the nutritional and growth needs of the child. This can be achieved at 15 cm of jejunum and ileum with the presence of an ileocecal valve or at 40 cm without the presence of a valve. Of course, assuming normal function of the remaining intestine.[5] Children with a residual small intestine shorter than 75 cm are at risk of developing SBS.[7]

Resection site

The severity and extent of malabsorption and metabolic complications of short bowel syndrome (SBS) also depend on the site of resection. The proximal 2/5 of the small intestine is the jejunum and the distal 3/5 is the ileum. The prognosis of jejuno-colic anastomosis (resection of the entire ileum, ileocecal valve and part of the colon) depends on the length of the preserved jejunum. Jejuno-ileocolic anastomosis (resection of part of the ileum with preserved ileocecal valve) has the most favorable prognosis. End-jejunostomy (resection of the entire ileum and colon) has the worst prognosis.[1]

The jejunum, which has longer villi and a larger absorption surface, has a high concentration of digestive enzymes and transport proteins (enzyme carrier proteins) and is the place with the greatest absorption of nutrients. The duodenum and jejunum are the primary sites for the absorption of carbohydrates, proteins, fats, and minerals. Although the ileum has shorter villi and less overall absorptive capacity than the jejunum, it is the only site for absorption of vitamin B12 and bile salts via site-specific receptors. While the ileum is able to adapt and compensate for the loss of the jejunum, the duodenum and jejunum are not able to take over the absorptive functions of the ileum. Resection of the duodenum and jejunum leads to hypergastrinemia and gastric hypersecretion, as well as iron and folic acid deficiencies. Water is passively absorbed in the small intestine during

the transport of nutrients and electrolytes. Sodium transport creates an electrochemical gradient that controls nutrient absorption across the intestinal epithelium. In the jejunum, there are relatively large, leaky *junctions* between epithelial cells, which leads to rapid fluid and nutrient loss and inefficient fluid absorption. The mucosa of the jejunum is unable to thicken the contents of the intestine and sodium diffuses freely into the intestinal lumen. Sodium is absorbed against the concentration gradient in the jejunum and is tied to the absorption of glucose. [1]

Ileal resection leads to malabsorption of vitamin B12 (bound to intrinsic factor and absorbed in the distal 50-60 cm of the ileum), to disruption of enterohepatic circulation and loss of bile acids. The reduced amount of bile acids disrupts the normal formation of micelles and the efficiency of absorption of fats and fat-soluble vitamins. In addition, increased passage of bile acids can cause secretomotor diarrhea (biliary diarrhea). Malabsorption of bile acids can also lead to increased absorption of oxalates, followed by hyperoxaluria and thus to an increased risk of oxalate renal stone formation and an increased risk of cholelithiasis. In addition, resection of the ileum disrupts the normal regulation of intestinal motility because GI hormones are produced in the ileum, particularly those that slow gastric emptying and affect small intestinal motility (glucagon-like peptide-1 [GLP-1], enteroglucagon, peptide YY). The lack of these hormones due to ileal resection therefore leads to a faster transit time and impaired absorption of nutrients in the small intestine. Intercellular junctions are tighter in the ileum than in the jejunum, so less water and sodium flow there. In addition, active transport of NaCl takes place in the ileum, which enables fluid reabsorption and thickening of the contents of the ileum. Resection of a significant part of the ileum leads to impaired ability to absorb fluids and electrolytes, to intolerance of larger food boluses, and to intolerance of food with higher osmolarity (eg with a higher content of simple carbohydrates). The ileum has a greater ability to adapt than the jejunum. In addition, active transport of NaCl takes place in the ileum, which enables fluid reabsorption and thickening of the contents of the ileum. Resection of a significant part of the ileum leads to impaired ability to absorb fluids and electrolytes, to intolerance of larger food boluses, and to intolerance of food with higher osmolarity (eg with a higher content of simple carbohydrates). The ileum has a greater ability to adapt than the jejunum. In addition, active transport of NaCl takes place in the ileum, which enables fluid reabsorption and thickening of the contents of the ileum. Resection of a significant part of the ileum leads to impaired ability to absorb fluids and electrolytes, to intolerance of larger food boluses, and to intolerance of food with higher osmolarity (eg with a higher content of simple carbohydrates). The ileum has a greater ability to adapt than the jejunum.[5][1]

Clinical outcome is also influenced by the absence/presence of the **ileocecal valve** (IC valve) and the length of the remaining colon. In children, the absence of an IC valve is a negative predictor of the ability to wean the patient from parenteral nutrition . The ileocecal valve slows down the movement of chyme from the small intestine to the large intestine, so resection of this valve leads to accelerated emptying of the small intestine. Accelerated transit time leads to fluid and electrolyte losses. The ileocecal valve also creates a barrier that prevents reflux of colonic contents into the small intestine. Bacteria can migrate orally into the small intestine, leading to **bacterial overgrowth**(SIBO). Bacterial colonization of the small intestine leads to deconjugation of the remaining bile acids, which alters micelle formation and worsens steatorrhea. Bacterial overgrowth can lead to the formation of D-lactate, which humans are unable to metabolize. **D-lactate acidosis** causes neurological problems such as ataxia, dysarthria and confusion (D-lactate encephalopathy).[5][1]

The large intestine plays an important role in the absorption of water, electrolytes and short-chain fatty acids. Compared to the jejunum and ileum, it has the slowest transit, the tightest intercellular connections, and the highest water and sodium absorption efficiency. Nutrients are also absorbed in the columns, e.g. unabsorbed fermented carbohydrates. In patients with SBS and a diet rich in carbohydrates, the colon can absorb up to 50% of the required energy. In patients without colons, a diet rich in carbohydrates can, on the other hand, cause diarrhea due to its high osmolarity. Colonic resection results in fluid and electrolyte losses. However, it also accelerates gastric emptying and shortens intestinal transit time due to decreased secretion of peptide YY, glucagon-like peptide (GLP-1) and neurotensin (negative regulators of intestinal motility).[8][1]

Extensive small bowel resections lead to gastric **hypergastrinemia** and hypersecretion, probably due to disruption of the negative feedback loop that normally inhibits gastrin secretion and reduces gastric acid production. In this context, resection of the duodenum and proximal jejunum is the most risky. Treatment includes rehydration and H2 blockers and proton pump inhibitors, especially in the early phase of SBS.[1]

Patients with SBS and enterostomies tend to lose significant amounts of sodium in the stool, leading to **secondary hyperaldosteronemia** and significant urinary potassium losses. Therefore, it is important to supplement sodium (up to 8-10 mmol/kg/day) and monitor ostomy waste, electrolyte levels, dietary hydration and diuresis (at least 1-2 ml/kg/day and the amount of sodium in the urine (> 30 mmol/l).[9]

Gut adaptation

Intestinal adaptation includes macroscopic and microscopic changes that lead to an increase in the absorptive capacity of the intestine. Part of the adaptation is the improvement of intestinal absorption, increased secretion of intestinal hormones, development of hyperphagia, changes in the intestinal microflora. In adults, it takes place in the first two years after intestinal resection, in children even longer. Adaptive changes are most evident in the ileum. They are influenced by a whole range of external and internal stimuli, such as nutrients, hormones, growth factors and other biochemical and genetic factors.

Structural adaptive changes include an increase in the length of the remaining intestine, an increase in villi, an increase in microvilli, a deepening of the crypts and an increase in the number of enterocytes, and an increase in intestinal muscle. Functional adaptation includes changes in the activity of brush border enzymes, fluidity and permeability, up- or downregulation of carrier-mediated transport (upregulation of Na+/glucose cotransporters, Na+/H+ exchange, etc.) and slowing of transit time.

Intestinal adaptation is most effectively stimulated by the presence of nutrients in the intestinal lumen (disaccharides, long-chain fats stimulate better than monosaccharides and medium-chain fats). On the other hand, intestinal adaptation is inhibited by e.g. octreotide or inhibitors of prostaglandin synthesis (aspirin, NSAIDs, corticosteroids).[1]

Nutritional Management

Early stage

In the first months after resection, the main goal is to maintain a good nutritional status with the help of parenteral nutrition and prevent fluid and electrolyte losses. [10] Early initiation of enteral nutrition promotes intestinal adaptation. During long-term administration of parenteral nutrition, it is advisable to reduce intravenous lipid intake to reduce the risk of intestinal failure-associated liver disease (IFALD).

In the early phase after resection, large fluid losses (especially from the stomach and proximal small intestine) are frequent, so these patients need replacement of fluid losses, sodium, potassium, chloride and magnesium. Hedging is possible both parenterally and enterally (with oral rehydration solutions). Fluid and electrolyte losses through enterostomies or stools should be measured.

Hypersecretion of gastric acid is common in patients with SBS, which leads to a decrease in pH in other levels of the intestine and fat malabsorption, impaired enteral absorption of drugs and fluid loss. In the early phase after resection, it is therefore advisable to administer drugs suppressing the production of gastric acid (H2-receptor antagonists, proton pump inhibitors) and to continue this treatment for several months, longer if there are signs of gastroesophageal reflux/reflux disease of the esophagus or peptic ulcer.[11]

Practical Procedure

1. parenteral nutrition via a central venous catheter
    - initial hydration and electrolyte management to achieve hemodynamic stability (1-3 weeks)
    - glucose supply initially 5-7 mg/kg/min., increased by 1-3 mg/kg/min. up to 12-14 mg/kg/min. (enables a gradual response of endogenous insulin, serves to prevent hyperglycemia and glycosuria, prevents immune dysfunction, liver steatosis and excess CO2 production)
    - lipids initially 1 mg/kg/day, gradually increased by 1 mg/kg/day up to 3 mg/kg/day; lipids should not provide more than 30-40% of total energy to avoid immune dysfunction and hyperlipidemia;
    - amino acids initially 1.5-2 mg/kg/day and increase to target value over the next 1-2 days

1. continuous enteral nutrition and reduction of parenteral nutrition (on the order of months)
    - if the patient on parenteral nutrition has stable electrolyte and water status and is growing, enteral nutrition can be initiated to promote intestinal adaptation and bowel growth
    - intestinal adaptation = cell hyperplasia, hypertrophy of villi, lengthening of the intestine, improvement of hormonal response -> increased absorption area
        - starts as early as 24-48 hours after bowel resection, but can last more than a year depending on a number of factors
    - the most suitable is continuous administration with a gradual increase in the administration rate - continuous nutrition tends to be better tolerated and enables better absorption of nutrients
    - an increased stool volume of more than 50% or a significantly positive presence of reducing substances in the stool is a contraindication to increasing the diet;
    - with good tolerance of a continuous diet, it is possible to switch to cyclic feeding with gradual weaning of parenteral nutrition (the weaning is extended by 2-4 hours) leaving 8-12 hours of night feeding
    - complex nutrients probably stimulate intestinal adaptation better than simple nutrients (amino acids, peptides, monosaccharides), but enteral nutrition is usually started with a protein hydrolyzate or an amino acid formula with a high fat content, mainly in the form of long-chain fatty acids; some children can also be fed standard polymeric infant formula; breast milk can also be given (intestinal adaptation is supported by immunoglobulins and growth factors such as growth hormone and epidermal growth factor)
    - long-chain fatty acids most support intestinal adaptation, while medium-chain triglycerides (MCT) are more soluble in water, better absorbed in children with SBS, have a higher osmotic effect and a lower trophic effect for the intestine;
    - excessive intake of simple carbohydrates can have a significant osmotic effect and worsen diarrhea;
    - saccharides should provide no more than 40% of total energy
    - monitoring of food tolerance, ostomy waste and growth parameters

1. period of complete adaptation of the intestine
    - enteral nutrition is well tolerated, oral nutrition is initiated[5]

Growth and energy needs

Resting *energy expenditure* in children with SBS is similar to that of healthy children[12], , however, due to malabsorption, they usually need 30-70% more calories with enteral nutrition than with parenteral nutrition.[13] Growth failure is a common problem in children with SBS, especially in children after catheter sepsis or necrotizing enterocolitis, as well as in children treated with corticosteroids and children very quickly weaned from parenteral nutrition (for fear of chronic complications).[11]

Enteral nutrition

Enteral nutrition should be started early, increased cautiously but gradually as rapidly as the patient tolerates. Oral rehydration solutions are a suitable supplement to enteral nutrition, which replenish the necessary fluids and electrolytes, especially in children with feeding tubes and large fluid losses.

The presence of nutrients in the intestinal lumen supports the adaptation of the intestine ("trophic doses" are enough), on the other hand, the absence of enteral nutrition leads to atrophy of the intestinal mucosa. Gradually increasing enteral nutrition allows reducing parenteral nutrition and preventing the development of IFALD, metabolic bone disease, and bacterial overgrowth. In older children, oral nutrition is important to prevent aversion to feeding.

Enteral nutrition should be started as soon as the child is stable, usually a few days after intestinal resection. The prerequisite is audible peristalsis and the absence of contraindications (ileus, bloody stool, radiological changes with suspected intestinal ischemia).[11]

It is optimal to start nutrition with breast milk, which has an optimal composition of macronutrients, trophic factors supporting intestinal adaptation, immunoglobulin and other immune factors supporting the function of the mucous barrier, is relatively hypoallergenic and low osmolar (compared to hypoallergenic formula). If breast milk is not available or well tolerated, it is advisable to give elemental nutrition (amino acid formula) because it is better absorbed and hypoallergenic (children with EBE have a higher risk of protein allergies or intolerance[14][15][16]. babies can be started on milk formula with intact protein because protein intolerance is less common in them than in young children. In addition, complex nutrients support intestinal adaptation.[17]

Enteral nutrition should first be administered continuously through a nasogastric or gastrostomy tube. Continuous administration maximizes diet tolerance with impaired functional intestinal capacity.[18] Furthermore, it makes it possible to permanently saturate the transport proteins (carrier transport proteins), fully utilize the available absorption surface and support the adaptation of the intestine. In patients with poor gastric emptying and intestinal dysmotility, post-pyloric Nasojejunal tube feeding may be beneficial.

Regardless of the severity of SBS, it is necessary to give a small amount of food orally (taking into account the age of the child) and thus enable the development of sucking and swallowing, to prevent aversion to feeding.

Enteral nutrition should be increased slowly and continuously, parenteral nutrition should be adequately reduced and fluid losses should be compensated. The speed of increasing is individual. It is important to monitor output from the stoma/number or weight of stool, diuresis and hydration status, possibly condition of the perineal skin. With satisfactory parameters, it is possible to increase the diet by 10-20 ml/kg/day.[19]

Enteral tolerance limit criteria:

- enteral fluid losses: 2-3 ml/kg/day through stoma, 10-20 g/kg/day through stool. Lower fluid losses indicate a good tolerance of the diet and an indication to increase it, on the contrary, higher fluid losses show that the tolerance threshold has been exceeded.
- reducing substance: 1% in stool or stoma waste.

Faecal alpha-1-antitrypsin, faecal elastase, and *spot tests for fecal fat* are not suitable markers of dietary tolerance because they are often abnormal in children with SBS even with good dietary tolerance. However, they can help adjust enteral nutrition, e.g. in case of steatorrhea, give a formula with MCT oil, etc.

Transition to bolus feeding. It is usually customary to initiate bolus feedings at a dietary tolerance covering at least half of total energy requirements, with parenteral nutrition providing the remainder. It is possible, for example, to feed bolus during the day and continuously during the night. Smaller doses given more often are better tolerated. Bolus feeding allows for the cyclic release of gastrointestinal hormones.

An indication for continuing parenteral nutrition is failure to thrive or excessive fluid and electrolyte losses that cannot be compensated enterally. In borderline cases, supplemental parenteral nutrition can be administered at night (cyclic administration), thus providing the patient with greater freedom during the day, encouraging interest in oral nutrition, and reducing the hepatotoxic effect of parenteral nutrition (IFALD).[11]

Fats

Small children benefit from a diet rich in fats, when fats provide 40-50% of the total daily energy intake (the composition of breast milk in infant formulas also corresponds to this). Fat has a relatively low osmolarity and high energy content. It supports intestinal adaptation and improves food tolerance. LCTs stimulate intestinal adaptation, MCTs are better absorbed because they pass directly through the enterocyte membrane. For infants with SBS, their fat needs are covered by breast milk or infant milk formula.[11]

Carbohydrates

Most children with SBS, especially those without a large intestine, cannot tolerate high concentrations of carbohydrates due to their higher osmolarity compared to fat and protein. Complex carbohydrates have a lower osmolarity and are therefore better tolerated than simple sugars.[11]

Dietary supplementation

n children with SBS, there is a risk of lack of certain nutrients, especially when parenteral nutrition is discontinued and after its termination, when the level of intestinal adaptation and nutrient absorption is unpredictable. The type of nutritional deficit depends on the extent and location of intestinal resection. The most common is a deficiency of

fat- soluble vitamins (A, D, E, K), calcium , iron and vitamin B12 . Furthermore, a deficit of copper , selenium and zinc . Micronutrient deficits can occur even with adequate energy intake and somatic growth, therefore regular laboratory monitoring is important.[20]

Loss of function of the terminal ileum is associated with a deficiency of fat-soluble vitamins (A, D, E, K), vitamin B12 and zinc. Deficiency usually begins to manifest several months after stopping parenteral nutrition (in which these vitamins and minerals are contained). A child with SBS on full enteral nutrition should have serum levels of vitamin A, 25-OH D3, E, B12, and zinc measured 1 and 3 months after stopping parenteral nutrition (note that serum zinc reflects only albumin-bound zinc, not total body zinc zinc reserves[4]).

Fat soluble Vitamins

- risk of deficit in resection of terminal ileum;
- Vitamin K is produced by intestinal bacteria, so its deficiency rarely develops during long-term treatment with broad-spectrum antibiotics. Coagulation tests (PT, PTT, and INR) indirectly indicate vitamin K status. If PT, PTT, and INR do not improve after vitamin K injection, IFALD may be the cause.
- vitamins A, D and E should be supplemented; it is recommended to monitor the level of these vitamins (once a year) for their potential toxicity.

Vitamin B12

- risk of deficit in terminal ileum resection
- risk of deficiency during acid suppression - disruption of the secretion of the *intrinsic factor* necessary for the absorption of vitamin B12 in the ileum
- elevation of serum methylmalonic acid (MMA) is a sensitive indicator of vitamin B12 status, however, MMA can be elevated in bacterial overgrowth (SIBO) without B12 deficiency being present, so it is not a reliable indicator of vitamin B12 deficiency [21][22]

- in SBS patients with bacterial overgrowth, vitamin B12 levels may be falsely normal, so bacteria may produce its biologically inactive analog. A more accurate assessment of vitamin B12 deficiency is made possible by the serum level of methylmalonic acid and homocysteine.[23]
- supplementation: cyanocobalamin (vitamin B12) IM once a month (0.5 mg for children < 10 years, 1 mg for children > 10 years).[11]

Sodium

- in children with high ostomy/faecal waste, it is advisable to monitor the level of sodium in the urine;
- low total body sodium contributes to failure to thrive (despite adequate caloric intake) and does not correlate with serum sodium, as renin and aldosterone keep it within the normal range;
- infants may need up to 4-8 mmol Na/kg/day to achieve satisfactory growth; [5]

- dietary supplementation with sodium (oral rehydration solution) to maintain urinary sodium above 20 mmol/L;[24]
- interpretation of urinary sodium is difficult in cirrhosis -induced hyperaldosteronism and/or use of loop diuretics .[11]

Zinc

- zinc losses are more pronounced with diarrhea and excessive waste from the ostomy. Zinc deficiency is manifested by failure to thrive, diarrhea, impaired wound healing, perinanal and perioral exanemia, and alopecia.[25][26]
- severe zinc deficiency manifests as acrodermatitis enteropathica, characterized by rash on the face, hands, feet and genitals.[23]

Pharmacotherapy

Children with SBS often have abnormal pharmacokinetics of enterally administered drugs due to intestinal malabsorption.

Antisecretory

- H2 receptor antagonists (famotidine) and proton pump inhibitors (omeprazole, lansoprazole, pantoprazole, esomeprazole) to inhibit excessive gastric acid secretion, side effects of long-term administration include bacterial overgrowth and vitamin B12 deficiency and H2RA tachyphylaxis .
- bile acid sequestrants (cholestyramine, colesevelam) in patients with diarrhea due to the presence of bile acids in the large intestine (after resection of the ileum and malabsorption of bile acids), adverse effects include malabsorption of fat-soluble vitamins and irritation of the digestive tract.

Anti-motile

- loperamide to relieve chronic diarrhea with high stoma/faecal wastes, side effects include bacterial overgrowth and acute gastrointestinal infection (especially Clostridium difficile) is contraindicated for risk of toxic megacolon.

Adaptive Agent

- teduglutide (glucagon-like peptide 2 analog) is used in some cases in PN-dependent adults with SBS to promote intestinal adaptation; given that it promotes the proliferation of the mucosa, it is advisable to first undergo a colonoscopy to rule out the presence of polyps; neoplasms of the digestive tract are a contraindication.[11]

Surgical options

When conservative procedures fail to discontinue parenteral nutrition, surgical solutions can be considered in individual cases.

- autologous intestinal reconstructive surgery - surgical lengthening of the intestine, narrowing or retraction of the dilated intestine to improve motility;
  - Bianchi procedure (longitudinal intestinal lengthening and tailoring, LILT);
  - STEP procedure (serial transverse enteroplasty procedure, STEP);
- small intestine transplantation - in SBS with life-threatening complications.[11]

Acute complications

Watery diarrhea
Electrolyte imbalance
Dehydration
Obstruction
Complications associated with central venous catheters
Infections associated with central venous catheters

Watery Diarrhea

- the most common early complication of SBS, but it can become a chronic recurrent problem
- it often appears when there is an excessive osmotic load of the intestine - when the absorption capacity is exceeded
- the volume of fluid in the intestinal lumen is involved in the development
- occasionally with bile acid malabsorption
- leads to excessive fluid and electrolyte losses
- therapy:
  - replacement of fluid losses - parenterally/enterally (oral rehydration solutions; also physiological solution, Ringer's lactate)
  - reduction of the osmotic load in the intestine - slowing down the rate of administration of enteral nutrition, transition to low-carbohydrate/high-fat enteral nutrition, transition from bolus feeding to continuous feeding.
  - antimotility (loperamide 0.8 mg/kg/day divided into 3 doses) to slow transit and enable absorption, contraindicated in acute infectious diarrhea and constipation
  - antisecretory drugs (proton pump inhibitors) improve the function of pancreatic enzymes and the absorption of nutrients; chronic use leads to electrolyte and bone abnormalities, a higher incidence of respiratory and gastrointestinal infections, changes the stomach microbiome and predisposes to bacterial overgrowth.

Chronic complications

Liver damage (*Intestinal failure-associated liver disease, IFALD)* or *parenteral nutrition-associated liver disease, PNALD*) with the risk of progression to cirrhosis, portal hypertension and liver failure
Complications associated with central venous catheters
Dysmotility
Diarrhea
Electrolyte imbalance
Bacterial overgrowth in the small intestine
Nutritional deficits
Growth retardation
Metabolic bone disease
Enteric hyperoxaluria
Eosinophilic gastrointestinal distress
Bleeding from the digestive tract
Skin complications
Oral aversion
Oral complications
Adhesion/obstruction
Gallstones[27]

Bacterial Overgrowth (SIBO)

Bacterial overgrowth (SIBO) is traditionally understood as an excessive amount of bacteria in the small intestine (> 10 5 CFU/ml). However, SIBO also involves the presence of inappropriate microbial species in certain parts of the small intestine. It is therefore a quantitative and qualitative change in the microflora of the small intestine. Under

normal circumstances, the proximal small intestine mainly contains Gram-positive aerobic bacteria, while the distal small intestine mainly contains facultative anaerobes and the colon almost exclusively anaerobic bacteria. Fungal overgrowth (SIFO, *small intestinal fungal overgrowth) can also cause problems.*[28]

Up to 60% of children with SBS develop **bacterial overgrowth** (SIBO). Impaired intestinal motility and intestinal dilatation create ideal conditions for abnormal bacterial proliferation. The negative consequences of bacterial overgrowth include: abdominal pain, impaired intestinal motility, mucosal ulceration and bleeding, deconjugation of bile acids and the formation of toxic products such as D-lactate. Bacterial overgrowth appears to facilitate translocation and thus septicemia.[23]

Patients with SBS tend to have an overgrowth of bacteria in the small intestine, especially anaerobic bacteria. An excessive population of bacteria in the small intestine can lead to small intestine bacterial overgrowth (SIBO), worsening malabsorption, flatulence and food intolerance. An excess of bacteria causes deconjugation of bile acids, thereby impairing the absorption of monoglycerides and fatty acids in the intestine. The inflammatory response caused by bacterial overgrowth damages the absorption surface and worsens malabsorption by impaired absorption of carbohydrates and proteins. Bacteria in the lumen of the small intestine compete for vitamin B12.[1]

For this purpose, stool culture from the stoma can be quantitatively examined.[23]

The gold standard in diagnostics is the **quantitative culture of aspirate from the small intestine** (however, it has a number of limitations). Another widely used option is a **breath test** after eating a defined amount of carbohydrates (even this examination has a number of limitations). Due to the limited informative value of the above-mentioned tests, empirical antibiotic treatment is sometimes considered (diagnosis is carried out on the basis of the treatment effect), however, the disadvantage is often unindicated treatment, the risk of C. difficile selection and the development of bacterial resistance.[28]

Suspected bacterial overgrowth is treated empirically with enterally administered antibiotics (targeting anaerobes or Gram-positive organisms).[23] E.g. rifaximin is preferred, or a combination of neomycin and rifaximin. It is also advisable to optimize the level of glycemia (diet with a low content of FODMAPs, fermentable oligo-, di- and monosaccharides and polyols), to discontinue drugs that slow down peristalsis and suppress gastric acid secretion. Probiotics (*Lactobacillus, Bifidobacterium, Saccharomyces boulardii*) are also used in the treatment.[28]

D-lactic acidosis may develop in SBS with bacterial overgrowth. During the normal anaerobic metabolism of mammals, L-lactate is produced , which is easily converted to pyruvate and further processed. D-lactate is produced by bacteria and people cannot process it well, so it accumulates in the body. D-lactate acidosis in the newborn manifests as acidosis with anion gap and convulsions. In older children, in addition, confusion, slurred speech and slowed cognitive functions. The diagnosis can be confirmed by the detection of D-lactate in the serum. It is treated by rehydration and eradication of bacterial overgrowth with enteral antibiotics.[23]

Links

Related Articles

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