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Clinical Recommendations of the Northwest Society for Enteral and Parenteral Nutrition, Interregional Association for Emergency Surgery, Russian Gastroenterological Association, Union of Rehabilitation Therapists of Russia and Russian Transplantation Society on Diagnosis and Treatment of Short Bowel Syndrome-Associated Intestinal Failure in Adults

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Aim. Current clinical recommendations address the epidemiology, causes, clinical manifestations and pathogenesis of possible immediate and long-term complications, as well as the problematic issues related to treatment and rehabilitation of adult short bowel syndrome patients.

Key points. Short bowel syndrome (SBS) is a symptom complex of impaired digestion caused by the reduction of small intestine absorptive surface and manifested by intestinal failure (IF) of various severity (maldigestion and malabsorption) developing into malnutrition and systemic somatogenic disorders. The vital strategic aspects of its treatment are the personalisation of liquid, macro- and micronutrients consumption as well as avoidance of intestinal failure- and parenteral nutrition-associated complications. Various nutritional support regimes and the indications for infusion therapy and maintenance parenteral nutrition are considered in this patient category, also in outpatient settings. To mitigate the dependence on intravenous fluid- and nutrient administration and attain enteral autonomy in SBS-IF patients, the use of recombinant glucagon-like peptide-2 (GLP-2) is justified as exerting a pronounced trophic effect on the epithelial regenerative potential as well as structural and functional adaptation of intestinal mucosa. The SBS-IF patients prescribed with home parenteral nutrition and/or their caregivers should be trained in a special programme that covers the catheter care, preparation of infusion solutions and nutrient mixture container, infusion pump operation as well as the prevention, recognition and management of complications. The main referral indications for small bowel transplantation (SBT) are: fast-progressing cholestatic liver disease-complicated irreversible intestinal failure; thrombosis of two or more central venous conduits used for parenteral nutrition; recurrent catheter-associated bloodstream infection.

Conclusion. Current recommendations on diagnosis and treatment as well as the developed criteria of medical aid quality assessment are applicable at different levels of healthcare.
Keywords: short bowel, intestinal failure, protein-energy malnutrition, nutritional status, nutritional support, metabolic therapy, parenteral nutrition, enteral nutrition
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Клинические рекомендации Северо-Западной ассоциации парентерального и энтерального питания, Межрегиональной ассоциации по неотложной хирургии, Российской гастроэнтерологической ассоциации, Союза реабилитологов России и Российского трансплантационного общества по диагностике и лечению синдрома короткой кишки с кишечной недостаточностью у взрослых

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Цель: В клинических рекомендациях рассматриваются эпидемиология, причины развития, клинические проявления, патогенез развития возможных ближайших и отдаленных осложнений, а также проблемные вопросы лечения и реабилитации взрослых пациентов с синдромом короткой кишки.
Основное содержание. Синдром короткой кишки (СКК) представляет собой симптомокомплекс нарушенного пищеварения, обусловленный уменьшением всасывательной поверхности тонкой кишки и проявляющийся кишечной недостаточностью (КН) различной степени выраженности (мальдигестия и мальабсорбция), что приводит к развитию недостаточности питания и системным соматогенным нарушениям. Наиболее важными аспектами лечебной стратегии являются персонализированное определение потребности в жидкости, макро- и микронутриентах, минимизация осложнений, связанных с кишечной недостаточностью и проведением парентерального питания. Рассматриваются различные варианты нутриционной поддержки, показания для инфузионной терапии и поддерживающего парентерального питания данной категории больных, в том числе в амбулаторно-поликлинических условиях. Для снижения или устранения зависимости от внутривенного введения жидкости и питательных субстратов и достижения энтеральной автономии у пациентов с СКК и КН возможно применение рекомбинантного аналога глюкагоноподобного пептида-2 (GLP-2), оказывающего выраженное трофическое воздействие на регенераторный потенциал эпителиоцитов и структурно-функциональную адаптацию слизистой оболочки кишечника. Пациенты с СКК и КН, которым планируется проведение

парентерального питания в домашних условиях, и/или лица, осуществляющие уход, должны пройти обучение по специальной программе, которая включает уход за катетером, этапы приготовления инфузионных растворов и контейнера с питательными субстратами, использование инфузомата, а также предотвращение, распознавание и устранение осложнений. Основными показаниями для направления пациентов на трансплантацию тонкой кишки (ТТК) являются: необратимая кишечная недостаточность, осложненная явлениями быстро прогрессирующего холестатического заболевания печени, тромбоз двух или более центральных венозных каналов, используемых для парентерального питания, и рецидивирующая катетерная инфекция кровотока.
Заключение. Представленные рекомендации по диагностике и лечению и разработанные критерии оценки качества медицинской помощи применимы на различных уровнях ее оказания.
Ключевые слова: короткая кишка, кишечная недостаточность, белково-энергетическая недостаточность, нутритивный статус, нутриционная поддержка, метаболическая терапия, парентеральное питание, энтеральное питание
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Terms and Definitions

Short bowel syndrome is a symptom complex of maldigestion caused by a decrease in the absorption surface of the small bowel and manifested by intestinal failure of varying severity (maldigestion and malabsorption), which leads to the development of malnutrition and systemic somatogenic disorders.

Intestinal failure is a decrease in the functional capacity of the small bowel, leading to disorders in the processes of intracavitary and parietal hydrolysis, as well as absorption of nutrients, water and electrolytes, which is accompanied by disorders of trophic and fluid and electrolyte homeostasis, progressive malnutrition, often requiring auxiliary therapy, including intravenous administration of water, electrolytes and nutrients.

Malnutrition is a heterogeneous syndrome complex, which can be caused by both a total or partial deficiency of various nutrients entering the body relative to its actual needs, and their impaired assimilation combined with increased spending, which is accompanied by persistent changes in trophic homeostasis, as well as structural (decrease in cell mass), and metabolic disorders, leading to a decrease in the functional reserves of the body and deterioration of clinical outcomes of the disease.

Protein-energy malnutrition is a condition characterized by a deficiency or imbalance, primarily in the energy and/or protein supply of the body relative to the available need, which leads to a body mass reduction with a disorder of its component composition and/or depletion of the visceral pool of proteins.

Nutritional status is a set of clinical, anthropometric and laboratory indicators reflecting the body state associated with the patient's nutrition.

Nutritional support is the process of providing patients with a substrate using special methods that differ from conventional nutrition and artificially created nutritional mixtures of various directions.

Enteral nutrition is the process of providing the body with the necessary nutrients by oral consumption or administration through a probe into the gastrointestinal tract of special artificially created nutrient mixtures.

Parenteral nutrition is a method of nutritional support, in which the nutrients necessary to ensure proper trophic homeostasis are administered into the body, bypassing the gastrointestinal tract.

1. Brief Information on the Disease

1.1. Mechanisms of Development and Epidemiology of Short Bowel Syndrome with Intestinal Failure in Adults

Short bowel syndrome (SBS) is a symptom complex of impaired digestion caused by a decrease in the absorption surface of the small bowel and manifested by intestinal failure of varying severity (maldigestion and malabsorption), which leads to the development of malnutrition and systemic somatogenic disorders [1]. The true prevalence of SBS in adults in the Russian Federation is not known, since there is no unified national register of this group of patients. In the USA, out of 40,000 adults receiving home parenteral nutrition (HPN), about 10,000 people receive it associated with SBS [2]. In Europe, the prevalence of SBS requiring HPN, according to various data, is 2–4 people per million urban population [3, 4]. Considering data that SBS patients in Europe represent 35 % of all patients receiving HPN, the prevalence of SBS is thought to be approximately 1.4 cases per million population [5]. The prevalence of SBS in Europe varies considerably by region: from 0.4 per million in Poland to 30 per million in Denmark [6]. The prevalence of SBS tends to be lower in regions where there are no rehabilitation centers for patients after enterectomies and HPN programs. Nevertheless, there has been an increase in the number of these patients. For example, a leading center for rehabilitation of patients with SBS in Denmark reported that the number of patients with SBS on HPN has doubled in a decade [7].

The main causes of SBS are: surgical removal of most of the jejunum and/or ileum; excluding of various parts of the small bowel from digestion and absorption processes, for example, when performing bariatric bypass surgery; jejunostomy or ileostomy as well as in cases of interintestinal fistula formation; application of anastomosis between proximal parts of the small and large bowel bypassing an ileocolic valve. Among the most frequent etiological factors of resection of different parts of the small bowel, mesenteric arteries and veins thrombosis, adhesive obstruction, abdominal trauma and wounds, multiple intestinal fistulas, Crohn's disease, small or large bowel cancer, small bowel lymphoma are considered the most frequent [1, 8].

Clinically significant symptoms of impaired digestion in the form of maldigestion and malabsorption occur in the majority of patients who have undergone resection of more than 60–70 % of the small bowel length. According to other data, intestinal failure can occur even when the remaining part of the small bowel is less than 200 cm long [9–11].

In a large multicenter study of 688 adults who received long-term HPN for nonmalignant chronic bowel failure, approximately 75 % of patients had SBS [12]. In this survey, the mean age of patients was 52.9 ± 15.2 years (18.5–88.0 years), the majority of patients were female (57 %), and the most common primary causes of SBS with IF were mesenteric ischemia (27 %), Crohn's disease (23 %), and radiation enteritis (11 %). Patient demographics can vary widely, depending on the region and the treatment center specifics. For example, a recently published study reported that the most common primary causes in SBS patients (268) were mesenteric infarction (43 %), radiation enteritis (23 %), surgical complications (12 %), Crohn's disease (6 %), and GI tumors (6 %). Most patients (67 %) underwent ileocolic anastomoses, 18 % had end jejunostomies, and 15 % had jejunoileoanastomoses [13, 14].

1.2. ICD-10 Coding

K91.2 Absorption disorder after surgical intervention not classified under other headings.

E40 Kwashiorkor. Severe malnutrition with nutritional oedema with dyspigmentation of skin and hair.

E41 Marasmus. Severe malnutrition with marasmus.

E42 Marasmic kwashiorkor.

E 43 Unspecified severe protein-energy malnutrition.

E44Protein-energy malnutrition of moderate and mild degree.

E 44.0 Moderate protein-energy malnutrition.

E 44.1 Mild protein-energy malnutrition.

E 46 Unspecified protein-energy malnutrition.

2. Diagnosis of Short Bowel Syndrome

2.1. Clinical Manifestations

There are three main categories of patients with SBS, who often have chronic severe intestinal failure, which requires long-term, sometimes lifelong, intravenous infusion therapy and parenteral nutrition (PN) for lifelong indications:

1) who underwent resection of most of the jejunum and the entire ileum with the jejunostomy (residual segment of 100 cm or less);

2) who underwent resection of the jejunum and/or ileum with formation of an ileocolic anastomosis bypassing the ileocolic valve (residual segment of the small bowel 60 cm or less);

3) who underwent extensive resection of the jejunum and ileum with complete preservation of the large bowel with ileocolic valve (ileocolic anastomoses with residual segment of 35–40 cm or less) [1, 8, 15].

Clinical manifestations in short bowel syndrome are predetermined:

A. The length of the resection and the remaining (functioning) part of the small bowel:

- extensive resection (residual segment of the small bowel < 200 cm);
- short bowel (preserved section of small bowel < 100 cm);
- super(ultra)short bowel (preserved section of small bowel < 50 cm).

B. The place of its resection:

- jejunum (proximal SBS) — disorders of hydrolysis of nutrient substrates and absorption of most nutrients prevail, which is manifested mainly by hypoabsorptive-osmotic diarrhea and progressive malnutrition;

• ileum (distal SBS) — disorders of absorption of water, electrolytes, bile acids and vitamin B_{12} prevail, which is manifested by development of mainly secretory-exudative diarrhea and fluid and electrolyte disorders with high risk of delayed (in 5–6 months) overlay of B_{12}-deficient megaloblastic anemia.

C. The presence or absence of ileocecal valve, which slows the passage of chyme through the intestine, prevents ascending bacterial colonization of the small bowel, aggravating the phenomena of enzymopathy, maldigestion and malabsorption, contributes to increased absorption of fluids, electrolytes and bile acids. When the ileocecal valve is preserved, the compensatory total absorption capacity of the small bowel can increase 8–10 times.

D. The ability of morphofunctional adaptation of the remaining part of the bowel, which to a certain extent depends on the patient's age, existing underlying and concomitant pathology, as well as properly selected and timely initiated therapy [1, 8, 15].

SBS manifests as persistent intestinal dyspepsia in the form of repeated diarrhea within 24 hours, caused most often by total maldigestion (creato-, steato- and amylorrhea), fluid and electrolyte disorders and progressive malnutrition due to the developing endogenously caused macro- and micronutrient deficiencies.

The severity of clinical symptomatology is mostly predetermined by the existing postoperative anatomical changes in the intestine.

The most favorable prognosis is observed in patients with jejunoileoanastomosis, preserved large bowel and ileocolic valve. In such patients, even with the remaining length of the small bowel just over 50–60 cm, its structural and functional adaptation and relative compensation of digestive processes over the next 6–12 months are possible. When the jejunum is resected with anastomosis between the jejunum and the large bowel, the severity of intestinal dyspepsia is predetermined not only by the length of its remaining part, but also by frequently recurrent ascending contamination of proximal parts of the jejunum by opportunistic colonic microflora, which aggravates the impaired processes of intracavitary and parietal digestion. When the length of the jejunum is less than 100 cm, the possibilities of its structural and functional adaptation are very limited and require a long time (many months, sometimes years). Patients have progressive body mass reduction, sarcopenia, anemia, hypoproteinemia and hypoalbuminemia, polyhypovitaminosis and immunosuppression. This leads to a decrease in performance capability and quality of life. Optimal oral nutrition in these patients is very problematic, since attempts to expand dietary restrictions are often accompanied by an increasing of intestinal dyspepsia. These patients experience a constant feeling of hunger and are tend to overeating, which further aggravates the phenomena of intestinal dyspepsia. Weight loss during the year in these patients can reach 20–30 % of its initial value, and sometimes more. In this regard, this category of patients often needs long-term maintenance infusion therapy and additional or complete parenteral nutrition.

When the ileum is resected with ileocolonastomosis bypassing the ileocolic valve, the clinical picture is initially dominated by fluid and electrolyte disorders caused by malabsorption of water and bile acids. A similar situation is observed in euno- or ileostoma, when the discharge of intestinal contents can reach several liters per day, which is accompanied by dehydration, dyselectrolycemia (hyponatremia, hypomagnesemia, hypocalcemia), and rapidly increasing malnutrition of patients due to the inability to take optimal oral food [1, 16, 17].

Three variants can be distinguished in the development and course of SBS according to the severity of its clinical manifestations:

1. Mild (relatively compensated) — with recurrent (most often with dietary errors) phenomena of transient intestinal dyspepsia (frequent up to 2–3 times/day loose stools, increased aerogenesis and hyperactive bowel sounds), moderate (up to 5 %) weight loss, relatively rapid effect of the therapy.

2. Moderate (subcompensated) — presence of daily diarrhea up to 3–5 times a day, despite compliance with dietary recommendations, weight deficit (more than 10 % of the initial value) with a continuing tendency to further decrease for more than 3 months, persisting absolute lymphopenia (less than 1,200 cells), and moderately marked hypoproteinemia (up to 60 g/L) and/or hypoalbuminemia (up to 30 g/L). Anemia, polyhypovitaminosis, transient edema join up. Possible phenomena of gastric dyspepsia (acid indigestion, belching, nausea), which may be caused by erosive-ulcerative lesions of the gastroduodenal mucosa. Correction of progressive malnutrition in such patients requires additional, often long-term, prescription of highly biologically valuable balanced polymeric or oligomeric enteral NM consumed by sipping or by adding them in powdered form to ready meals, and sometimes periodic courses of additional parenteral nutrition.

3. Severe (decompensated) — manifests as persistent intestinal dyspepsia in the form of repeated (more than 5 times a day), often watery diarrhea, recurrent syndrome of bacterial overgrowth in the proximal parts of the small bowel, aggravating manifestations of intestinal dyspepsia(increased aerogenesis and hyperactive bowel sounds, increased frequency of stools), total maldigestion (creature-, amylo- and steatorrhea) in coprogram, rapidly progressing asarcia (BM reduction during 3–6 months reaches 20–30 % or more of its initial value) against a background of constant hunger and frequent overeating, aggravating existing intestinal dyspepsia. Hypovolemia is often observed, manifested by marked general weakness, tachycardia, hypotension, orthostatic dizziness and dyselectrolycemia (hyponatremia, hypokalemia, hypomagnesemia, hypophosphatemia, hypocalcemia), as well as polyhypovitaminosis, increased seizure activity, mixed anemia, immunosuppression, severe hypoproteinemia (less than 45 g/L) and/or hypoalbuminemia (less than 25 g/L), oncotic edema. These patients have a high risk of gall bladder and/or kidney stones. This category of patients requires long-term (often lifelong) continuous intravenous correction of fluid and electrolyte disorders and maintenance parenteral nutrition for life indications [18].

In all cases of SBS requiring continuous parenteral nutrition and supportive therapy, even in the presence of relatively satisfactory dimensions of nutritional status and functional parameters, they should be referred to a *severe variant* of the disease course.

Another prognostically significant factor in the further development of SBS is the underlying disease for which the small bowel resection was performed. For example, in patients with Crohn's disease after resection of the intestine section affected by a granulomatous process, the disease may recur, and these patients must receive long-term specific anti-relapsing therapy. In persons with malignant neoplasms or abdominal trauma, other abdominal organs are often involved in the pathological process, which can also affect the prognosis and adaptive capacity of the small bowel. In addition, the span of life and the need for additional nutritional support are influenced by the age of patients, the presence of concomitant disease, and baseline body mass index.

In young children, the ability of the small bowel to grow and recover is superior to that of adults, especially in elderly patients. Therefore, if all else were equal, older patients have a less favorable prognosis. According to recent studies, initial overweight is considered to be a favorable prognosis factor [19, 20].

Intestinal failure is a decrease in the functional ability of the small bowel, in which the processes of intracavitary as well as parietal hydrolysis of food chyme and the subsequent absorption of nutrients, fluid and electrolytes, which are necessary to maintain optimal body life activity, are impaired, requiring it intravenous administration. In all cases when patients who underwent small bowel resection in the postoperative period for 2–3 months have repeated diarrhea with detectable coprological signs of maldigestion (steatorrhea, amylorrhea, creatorrhea), which is accompanied by fluid and electrolyte disorders and progressive body weight loss of 5 % or more per month, they should be considered as patients with short bowel syndrome and existing intestinal failure [12, 21].

Consequentially, the clinical manifestations and somatogenic complications of SBS depend primarily on the severity and duration of the existing intestinal failure (IF), which is divided into 3 types:

- type I is an acute, short-term (days, less often weeks) and often self-resolving condition;
- type II is a prolonged acute condition, often in metabolically unstable patients, whose treatment requires a comprehensive multidisciplinary approach and mandatory intravenous infusion therapy for 4 weeks to several months. About 50 % of these patients move into the group of patients with type 3 IF;
- type III is a chronic condition in metabolically stable patients requiring prolonged intravenous infusion therapy and parenteral nutrition for months or years. It can be reversible or irreversible. The last one often requires lifelong maintenance parenteral nutrition.

Clinical manifestations of IF can develop when the length of the retained segment of the small bowel is less than 200 cm (40 % of the average length of the small bowel). Resection with preservation of less than 50 cm (10 % of the average length of the small bowel) is considered to be the most prognostically unfavorable situation with regard to the development of the most severe form of IF. Chronic IF can be associated with life-threatening complications and lead to disability of patients and deterioration of their quality of life. The main objectives of treatment of such patients are to maintain fluid and electrolyte homeostasis (FEH), optimum acid-base balance and proper substrate supply of the body with minimization of risks of adverse complications. The overall five-year survival rate for patients with benign disease on HPN is about 75 % and depends on the underlying disease, their age, and postresection anatomic bowel abnormalities [21, 22].

The highest risk of developing clinically significant intestinal failure syndrome is observed in patients with a remaining small bowel length of less than 100 cm with a jejunostomy, less than 60 cm when combined with right-sided hemicolectomy, and less than 35 cm even with a preserved large bowel with an ileocolic valve. Such patients, as a rule, need long-term and sometimes lifelong NS [16, 23].

2.2. The Most Common Consequences and Complications of Short Bowel Syndrome

Gastric hypersecretion of hydrochloric acid. It is most often expressed when the jejunum is removed, where most of the endocrine cells producing gastric inhibitory and vasoactive intestinal polypeptides are located, which inhibit the production of gastrin during active intraintestinal digestion. Due to elevated gastrin levels for 3–6 months, and sometimes more, there is hypersecretion of gastric juice hydrochloric acid, which develops within the next 24 hours after surgery. Due to increased acid-peptic aggression there is an increased risk not only of erosive and ulcerative lesion of gastroduodenal mucosa, but also of disorders of subsequent digestive processes, due to long-term decrease of pH in duodenum, which leads to inactivation of pancreatic lipase and deconjugation of bile acids located in intestinal lumen. Early and long-term (3–6 months) use of gastric secretion inhibitors improves digestion and absorption of nutrients in the bowel.

Cholelithiasis. The highest risk of cholelithiasis development is observed when the ileum is resected. This is caused by impaired absorption and enterohepatic recirculation of bile acids, a decrease in their concentration in bile and impaired cholate-cholesterol balance (lithogenic bile). Formation of cholesterol stones in the gall bladder is also promoted by its hypomotility and bile stasis caused by decreased cholecystokinin production on the background of a restricted sparing oral diet. The main direction of prevention of cholelithiasis in this category of patients is early prescription and subsequent split oral nutrition (food is a physiological stimulator of bile secretion), as well as periodic courses of ursodeoxycholic acid preparations with regard to their tolerability.

Hepatopathy can be a consequence of portal endotoxemia in patients with SBS in the absence of ileocecal valve (most often in small bowel resection combined with right-sided hemicolectomy), which contributes to the upward contamination of opportunistic microflora in the proximal parts of the remaining small bowel (syndrome of bacterial overgrowth) and translocation of bacterial toxins into the portal vein. Bacterial overgrowth also causes changes in bile acid metabolism in the bowel, resulting in increased formation of lithocholic acid, which contributes to cholestasis. The last one can also be induced by prolonged parenteral nutrition with long-term use of fat emulsions based only on soybean oil (long-chain triglycerides) in an amount ≥1 g/kg/day, which is more often observed in pediatric practice and is associated with the high content of phytosterols in this oil [24–26].

Nephrolithiasis and oxalic nephropathy. Under normal conditions, oxalates from food are bound to calcium in the small bowel to form an insoluble complex. In patients with SBS with preserved segmented intestine, calcium binds to unabsorbed fatty acids, resulting in increased absorption of oxalates in the large bowel. The resulting hyperoxalaturia, often combined with hypohydration and oliguria, can lead to the formation of oxalate kidney stones. Nephrolithiasis develops in 25 % of patients with SBS receiving parenteral nutrition for a long time. The main mean for the prevention of the nephrolithiasis in such cases is regular intake of calcium carbonate in amounts of 5–6 g per day (1 g before each meal).

D-Lactic acidosis. A rare complication based on excessive formation of D-lactic acid due to active bacterial fermentation of carbohydrates in the large bowel, which leads to metabolic acidosis. Clinically, this is manifested by increasing weakness, ataxia, and increased somnolence. Many patients note a certain correlation of this condition with the consumption of large amounts of carbohydrates, especially those with a high glycemic index (over 70). Consuming large amounts of mono- and oligosaccharides carries the risk of lactacidosis due to hyperproduction of lactic acid by small bowel lactobacilli and colonic microflora. Treatment includes restriction of simple carbohydrate intake and intestinal decontamination by prescription of nonabsorbable antibiotics (rifaximin, nifuroxazide) [27].

Osteoporosis. The probability of developing osteoporosis with prolonged HPN is quite high (30 %) due to malabsorption of vitamin D and calcium. Persistent chronic inflammation can increase osteoclastic activity, which aggravates bone damage. Chronic metabolic acidosis due to loss of bicarbonate with feces or in renal failure due to recurrent dehydration episodes, eventually reduces the buffering kidneys capacity and thus can decrease bone mineral content. Hypomagnesemia also plays an important role in the osteoporosis development. Magnesium is necessary both for parathyroid hormone (PTH) secretion and for its proper action on target organs, such as osteoblasts and kidney cells. Prolonged steroid use may also increase the risk of osteoporosis by decreasing osteoblast activity, increasing urinary calcium loss and further decreasing intestinal calcium absorption. Target levels for 25-OH vitamin D should be above 30 ng/mL. Periodic monitoring of vitamin D levels is necessary even in patients who regularly receive vitamin D in HPN. Testing serum PTH helps in the early detection of those patients who need more intensive correction of metabolic bone disease [1, 8, 28, 29]. The immediate and long-term complications of SBS are presented in a systematic way in Table 1.

2.3. Manifestations of Malnutrition in Patients with Short Bowel Syndrome and Intestinal Failure

The main consequences of the formation of SBS with IF are permanent fluid loss and tendency to hypovolemia, acid-base balance disorders, dyselectrolyemia (hyponatremia, hypokalemia, hypomagnesemia, hypophosphatemia, hypocalcemia), progressive weight loss, hypoproteinemia and hypoalbuminemia, and development of vitamin and micronutrient deficiency [30]. Progressive malnutrition can significantly worsen patients' quality of life and increase their risk of developing a number of severe visceral complications. Persistent deficit of free water and electrolytes (especially sodium and magnesium) causes postural hypotension, thirst, muscle spasms, tremors. It is not uncommon for renal dysfunction to develop against this background [31].

Pathophysiological changes after small bowel resection are mostly determined by the type of the resection.

At resection of the ileum and large bowel due to the disorder of the natural locking mechanism of the ileocolic valve, the rate of gastric emptying and passage through the small bowel significantly increases, which is caused by the hypoproduction of YY-peptide and glucagon-like peptide-2, which are normally secreted by the corresponding endocrine cells located mainly in their mucosa and play an important role in the regulation of appetite and intestinal motor activity [1, 6].

Significant jejunostomy resection of the jejunum involves significant losses of water and electrolytes. Under physiological conditions, passive secretion in the jejunum promotes isotonic equilibration between intestinal contents and plasma. If the length of the jejunum is less than 100 cm, fluid loss through the stoma usually exceeds the amount of fluid drunk. When patients consume a hypotonic solution with sodium content less than 90 mmol/L, additional sodium loss occurs due to its diffusion from plasma into the intestinal lumen along the concentration gradient, which may lead to hyponatremia.

After resection of more than 60–100 cm of the terminal ileum, malabsorption of fats, vitamin B_{12} and bile acids develops. Unabsorbed bile acids enter the large bowel and have a chemical effect on the mucosa, which is accompanied by increased secretion of water and electrolytes, and unabsorbed fatty acids bind magnesium ions. At the same time, due to secondary hyperaldosteronism often develops in these patients, urinary magnesium losses increase. Hypomagnesemia is accompanied by a decrease in parathormone activity, inhibition of D-1,25-diocholecalciferol production, and calcium uptake in the renal tubules and intestine.

Significant FEH abnormalities are rarely observed in preserved large bowel. If sodium is reduced throughout the day, it is recommended to take an oral isotonic glucose-saline solution in amounts determined by the degree of dehydration

Table 1. Complications of short small bowel syndrome

Immediate complications (up to 3 months)	Long-term complications (more than 3 months)
• Fluid and electrolyte disorders (hypovolemia, dyselectrolycemia) • Gastric hypersecretion and erosive-ulcerative lesions of gastroduodenal mucosa • Rapidly progressing polynutrient failure (rapid loss of BM with increasing sarcopenia, anemia, hypoproteinemia, hypoalbuminemia, immunosuppression, multiple organ dystrophy) • Infections	• Cholelithiasis (cholesterol stones) • Hyperoxalaturia and nephrolithiasis • Severe asarcia, multiple organ dystrophy, functional loss • D-Lactic acidosis • Recurrent infections • Hepatic fibrosis • B_{12}-deficient megaloblastic anemia • Peritoneal adhesions with episodes of dynamic intestinal obstruction

(the usual physiological requirement is about 30 mL/kg of weight) [1, 16].

In severe asarcia and hypovolemia, patients have general weakness, difficulty concentrating, apathy, somnolence, hypothermia, sarcopenia, orthostatic dizziness (sometimes fainting), palpitation and heart rhythm disorders on exercise, hypoalbuminemic edema (sometimes with the development of ascites and hydrothorax), amenorrhea, reduced libido. Patients often experience a constant feeling of hunger, which is often accompanied by poorly controlled large quantities of food, aggravating the phenomena of existing intestinal dyspepsia [4, 17, 32].

Recommendation 1. A dynamic assessment of nutritional behavior with assessment of food and fluid intake, as well as anthropometric, clinical, and laboratory parameters characterizing nutritional status, should be performed in all patients with short bowel syndrome [20]. Level of evidence — 1, grades of recommendation — A.

Comments. *This tactic allows timely assessment and diagnosis of signs of developing malnutrition both in the early postresection period and months after surgery (Appendix 5 of Order No. 330 of the Ministry of Health of the Russian Federation, August 5, 2003).*

The indications for prescribing various types of active NS to patients with SBS are:

1. Presence of rapidly progressive and significant body weight loss (BW) in the postoperative period of >2 % per week, >5 % per month, >7.5 % per quarter, or >10 % per 6 months.

2. Existing initial signs of hypotrophy: BMI <19 kg/m^2 height (<21 kg/m^2 in aged 60–75 years and <22 kg/m^2 in aged 75 years or more); hypoproteinemia <60 g/L and/or hypoalbuminemia <30 g/L; absolute lymphopenia <1.2×10^9 L.

3. The threat of rapidly progressing malnutrition due to inability to adequately ingest food naturally (do not want, should not, or cannot ingest food naturally) and/or inadequate digestion of food in the presence of persistent intestinal dyspepsia with phenomena of marked maldigestion and malabsorption [1, 8, 28].

Recommendation 2. Monitoring of signs and symptoms of fluid and electrolyte homeostasis disorders with clinical and laboratory assessment of daily fluid balance and serum electrolyte content should be performed regularly both in the early and periodically in the late postresection period [30]. Level of evidence — 2, grade of recommendation — B.

Comments. *One of the frequent manifestations of SBS is disorders of fluid and electrolyte homeostasis (FEH), which is most often observed during ileum resection, especially in combination with right-sided hemicolectomy, as well as in the presence of jejunostomy or ileostomy. Under physiological conditions, passive secretion in the jejunum promotes isotonic equilibration between intestinal contents and plasma. If the length of the jejunum proximal to the stoma is less than 100 cm, when oral consume a hypotonic solution with sodium content less than 90 mmol/L, additional sodium loss occurs due to its diffusion from plasma into the intestinal lumen along the concentration gradient. The most frequent FEH abnormalities are observed in patients with jejunostomies or proximal ileostomies. It should also be taken into account that in this category of patients in order to reduce the manifestations of intestinal dyspepsia in the form of repeated diarrhea is often forced to limit the oral fluid intake. In this regard, these patients should have a dynamic control of FEH, which is especially important in the early postoperative period (skin turgor, state of mucosae, fluid balance, serum sodium, potassium, magnesium and calcium content). FEH disorders can significantly reduce the effectiveness of patients NS. Fluid and sodium deficiency can lead to hypovolemia, which is manifested in patients by thirst, dry mucosae, low skin turgor, rapid BM reduction, hypotension, tachycardia and prerenal renal failure (oligouria, creatininemia). Daily body mass, accurate fluid balance (including stoma rate), determination of the above electrolytes in blood serum, and control of sodium content in a random urine sample less than 10 mmol/L (marked sodium deficiency) are the most important markers of FEH status [1, 8, 28, 33].*

3. Treatment

3.1. Conservative Treatment

The following stages of treatment of SBS with IF are distinguished:

1. Immediate postresection period (up to 10 days).
2. Early postresection period (10 days to 3 months).
3. Late postresection period (3 months to 2 years) [22, 23].

Treatment of patients with SBS and IF is complex process that requires an individualized and comprehensive step-by-step approach. The most important aspects of the treatment strategy are personalized determination of fluid, macro and micronutrient requirements, minimization of complications associated with intestinal failure and parenteral nutrition.

Recommendation 3. In the immediate postoperative period it is necessary to correct fluid and electrolyte homeostasis, taking into account the actual losses of fluid and electrolytes, as well as acid-base balance of the body as a prerequisite for effective follow-up nutritional support. In patients with jejunostoma or ileostoma, mandatory control of electrolytes in blood and additional administration of 100–120 mmol of sodium are necessary [34]. Levels of evidence — 3, grades of recommendation — C.

<u>Comments</u>. *Pre-correction and optimal support of fluid and electrolyte and acid-base balance is a prerequisite for effective implementation of subsequent nutrient and metabolic therapy. Optimal intracellular hydration is a prerequisite for successful intracellular metabolism. Patients' water requirements are determined on the basis of analysis of the body's water balance, taking into account, if possible, an accurate assessment of renal and extrarenal fluid losses. For this purpose, the volume of daily diuresis (the proper value is 1 mL/kg/h), fluid losses with vomit, stools and aspirated gastrointestinal contents, discharge through drains, losses by perspiration through skin and lungs, amounting to 10–15 mL/kg/day are summed. The loss of fluid that occurs when body temperature rises should also be considered — for every 1 °C rise in body temperature above 37 °C during the 24-hour period of hyperthermia, 2–2.5 mL/kg per day should be added. The baseline requirement for replenishing current fluid loss in patients aged 18–60 years is 35 mL/kg, and in those over 60 years of age it is 30 mL/kg per day [35]. In patients with SBS, especially in cases of jejunostomy or ileostomy, phenomena of extracellular dehydration due to increased loss of sodium and water with intestinal contents may be observed, which is manifested by hypovolemia (pallor, dryness and decreased skin turgor, dry tongue, tachycardia, nausea and vomiting, arterial hypotension, fall in diuresis rate, apathy, high hematocrit index, low urine density, normal serum sodium concentration with low urine sodium content). Consumption of plain water by patients with SBS may increase intestinal content and sodium loss. The clinical symptom of hyponatremia is an increased organoleptic need for salt, which requires its additional prescription as part of the consumed meals or enteral nutritional mixtures, in which the sodium content is usually less than 35 mmol/L. To increase the sodium content to 100 mmol/L, which is above the minimum critical level (90 mmol) at which sodium absorption occurs, at least 6 g of salt must be added to the mixture. Patients with high eunostomies have the highest sodium requirement, sometimes as high as 200 mmol/day. In this regard, for rehydration purpose, such patients should be recommended oral intake of chyme-like glucose-salt solutions, which is especially relevant for patients with jejunostoma or ileostoma. Optimal is the administration of rehydration isotonic glucose-salt solutions in 1 L of which contains at least 60 mmol (3.5 g) of sodium chloride. Fluid and electrolyte losses through the stoma or due to diarrhea can also be caused by dietary abnormalities, such as consumption of dairy products (lactose), sucrose, and/or fats. High losses of intestinal contents through the stoma may be due to overgrowth of opportunistic microflora in the remaining part of the small bowel (ascending colonization), which requires decontamination, and/or clostridial enteritis associated with antibiotics. Losses through the stoma increase after intake of large amounts of fluid (more than 1–1.5 L) or food. Note that each liter of intestinal secretion (especially in a jejunostomy) contains ≈100 mmol of sodium. Potassium losses are relatively small at ≈ 15 mmol/L, but they may increase due to hyperaldosteronism secondary to hyponatremia. Hypokalemia may also be a consequence of hypomagnesemia, which may be more often observed in the presence of a jejunostoma. Oral rehydration of patients with SBS is best performed with glucose-salt solutions with time intervals of 30–60 minutes after meals or between meals, which helps to reduce diarrhea.*

In this case, the daily diuresis should be at least 1–1.5 L. To reduce intestinal secretion in patients with a jejunostoma, oral intake of both hypotonic (water, tea, coffee, or alcohol) and hypertonic beverages (fruit juices, cola, mineral water) should be limited. If necessary, there is intravenous correction of the existing fluid and electrolyte deficit by administering balanced electrolyte and/or sodium-containing infusion crystalloid solutions [1, 14, 16, 17, 28].

Recommendation 4. Basic therapy of patients with SBS in the immediate postoperative period after correction of fluid and electrolyte homeostasis should include early (the first 24–48 hours) administration of minimal enteral nutrition in combination with parenteral administration on the 3–5 day of the required nutrient substrates, prescription of gastric secretion blockers and performance of (if the integrity of the ileocolic valve is compromised) intraintestinal decontamination. Glutamine currently has been shown to be an essential nutrient substrate for maintaining the structure of the integrity and function of the smallbowel [36]. Level of evidence — 3, grade of recommendation — C.

<u>*Comments.*</u> *The small bowel is the main place for digestion and absorption of nutrients. Epitheliocytes of the intestinal mucosa belong to the short-lived cells of the body, their composition is completely renewed within 3–7 days. About 285 g of intestinal epithelium exfoliates in the human intestine each day. Adequate regenerative trophism of intestinal mucosa cannot be fully provided by nutrients coming from blood. It was established that the regenerative potential of enterocytes of the small bowel mucosa depends on the presence of nutrients by about 50 % and colonocytes by 80 % in the intestinal lumen [1, 28]. Absence of nutrient substrates in intestine during starvation is accompanied by a relatively rapid decrease in the size and function of the intestinal mucosa and its atrophy. These morphological changes may be reversible under conditions of enteral, but not parenteral nutrition [36, 37]. To ensure structural integrity and polyfunctional activity of the gastrointestinal tract in the immediate postoperative period, early prescription of minimal enteral nutrition, which is aimed primarily at providing intraluminal trophism of mucosal epithelial cells, supporting their regenerative potential and maintaining the barrier function of the intestine, is of great importance [1, 28, 36]. Early minimal enteral nutrition does not provide the necessary substrate requirements of the body and is essentially aimed at intraluminal "nutrition of the intestine". If a minimum acceptable substrate supply of more than 50 % of the patient's needs is impossible, especially in patients with baseline hypotrophy (BMI less than 19 kg/m² or less than 21–22 kg/m² in the elderly patients) for 3 days additional parenteral nutrition in gradually increasing volume should be prescribed in order to achieve over the next 3–4 days an adequate (at least 80 % of need) energy and protein supply. Parenteral nutrition may be prescribed on day 5–7 in persons with an underlying euthrophic condition (BMI 20–25 kg/m²) or in the presence of excess BM (BMI over 25 kg/m²), as well as in obesity (BMI over 30 kg/m²) [38, 39]. Over the past 25 years, clinical medicine has accumulated quite a lot of experience in the use of the conditionally essential amino acid L-glutamine, which has a fairly wide range of pharmacological effects. L-glutamine, being the most important energy substrate for intestinal epithelial cells, prevents mucosal stress atrophy and increased intestinal permeability, reduces the frequency and severity of bacterial translocation, has a powerful antioxidant and cytoprotective effect. At the same time, it has a pronounced nitrogen-saving effect, enhances muscle anabolism and increases the activity of immunocompetent cells. These effects of L-glutamine allow us to classify it as a pharmaconutrient that has a direct effect on the structural and functional and metabolic processes of the body. The main "consumers" of glutamine, especially in critical patients, are epithelial cells of the small bowel mucosa (10–14 g/day) [1, 8, 28, 40, 41]. It has now been shown in experimental animals and humans that glutamine is an essential nutrient substrate for maintaining the integrity and function of the small bowel, especially when there is damage to its mucous and deterioration of its barrier function, which is accompanied by translocation of bacteria and their toxins into the bloodstream. Glutamine stimulates the growth of villi as well as the formation of organoids in crypt cells, their proliferation and differentiation, which can improve the absorption of nutrients [40, 41]. Glutamine supplementation has a favorable effect on the intestinal mucosal morphology of healthy volunteers and patients with gastrointestinal diseases and improves nutrient absorption [8, 15, 28, 40–43]. In other 8-week randomized, placebo-controlled, crossover study in 8 patients with SBS, there was*

no effect of glutamine on intestinal morphology, rate of food bolus transit, D-xylose absorption, or frequency of diarrhea [44]. It is to be noted that the effects of adding glutamine to enteral nutrition regimens have been studied less than the effects of adding it to parenteral nutrition regimens. Because L-glutamine is unstable in clinical practice, it is used in the form of glycine-glutamine dipeptides (enteral administration) and alanyl-glutamine (parenteral administration).

Recommendation 5. In the presence of gastrostasis for 48–72 hours after small bowel resection, it is possible to use the "two tubes" technique, for which a nasojejunal tube 25–30 cm distal to the Treitz ligament is installed using an endoscope, through which enteral therapy measures can be conducted, a part of which is minimal enteral nutrition, which provides early intraluminal trophicity of intestinal mucosa epitheliocytes [28]. Level of evidence — 3, grade of recommendation — C.

Comments. *Gastric motor-evacuation function disorder in patients after volumetric abdominal surgery is not uncommon in the early postoperative period. Developing gastrostasis significantly limits the possibility of prescribing enteral nutrition. Nasogastric tube administration into the stomach with boluses of chilled water (150–200 mL) activates its propulsive activity and contributes to earlier resolution of gastrostasis phenomena. If the latter phenomena persist for 48–72 hours, endoscopic installation of a second (nasojejunal) tube distal to 25–30 cm of the Treitz ligament is indicated. Administration of even a moderate amount of glucose-salt solutions (500 mL) and isocaloric nutrient substrates (300 mL) into the jejunum promotes activation of propulsive activity not only of the intestine, but also of the stomach. Early enteral support (therapy) aimed at preventing and minimizing the consequences of postaggressive effects on the GI tract an affordable and relatively fast method of its structural and functional rehabilitation in the early postoperative period [1, 8, 28].*

Recommendation 6. In the early postoperative period after resection of most of the jejunum (proximal SBS) when prescribing tube or oral nutrition is initially more preferable the use of easily digestible isocaloric isonitrogenic oligomeric (semi-elemental, oligopeptide) enteral nutritional formulas that contain hydrolyzed whey protein, at least 50 % medium chain triglycerides (easily absorbed in conditions of maldigestion with bile acid and lipase deficiency) and maltodextrin deep hydrolysis [45]. Level of evidence — 3, grade of recommendation — B.

Comments. *With resection of most of the jejunum, in which, as known, the most active hydrolysis and absorption of the vast majority of nutrients is carried out, in patients of this category rapidly progressive malnutrition (especially in the first months) may develop. The presence of pronounced maldigestion and malabsorption in these patients can be judged by the coprogram tests (creato-, amylo- and steatorrhea) and the increasing body mass reduction. Over time, the missed functions of the jejunum begin to be compensated by the adaptive structural and functional restructuring of the remaining ileum mucosa. Additional consumption of oligomeric (semi-elemental) NM, the features of the chemical composition of which provide their greatest bioavailability in conditions of maldigestion, contribute to a better maintenance of the nutritional status of these patients. Note that some oligomeric liquid mixtures, due to their poor organoleptic properties, must be administered through a probe and are unsuitable for oral consumption. The most acceptable for this purpose in terms of their taste properties are powdered oligomeric NMs, which can be added to ready meals or consumed by oral sipping in liquid form [1, 8, 28, 45].*

Recommendation 7. In distal SBS, polymeric isocaloric isonitrogenic isoosmolar enteral nutrient mixtures containing predominantly soluble dietary fiber with prebiotic (bifido- and lactogenic), trophic and bile acid sorbing effects can be initially prescribed. If the latter are poorly tolerated (intestinal dyspepsia), it is necessary to temporarily switch to the administration of oligomeric PM. In patients with baseline hypotrophy (BMI < 16 kg/m^2), regardless of the SBS variant, it is initially better to use oligomeric diets [46, 47]. Level of evidence — 3, grade of recommendation — B.

Comments. *After extensive resection of the ileum, fluid and electrolyte disorders initially dominate in the clinical picture of SBS in patients, which requires intravenous correction. In this case, the possibility of hydrolysis and absorption of nutrients in the remaining jejunum is preserved, which with a properly selected and organized diet of patients often avoids the need for additional parenteral nutrition. Prescription in the first days of the* *postoperative period of early enteral nutrition in the mode of continuous prolonged administration of diets with breaks every 4–6 hours for 30 minutes promotes activation of production of intestinal hormones that regulate the activity of the digestive-transport conveyor, as well as support regenerative activity of the mucosa. Balanced enteral diets have greater bioavailability and absorption capacity in the intestine compared with the traditional diet and contribute to accelerated structural and functional adaptation in the immediate postoperative period [1, 8, 28, 48]. In additional note that in patients with high jejunostoma, when initially there is accelerated transit of gastric contents and fluid through the small bowel, active use of liquid, especially hyperosmolar, enteral diets can cause hypersecretion and loss of intestinal discharge and electrolytes. With a preserved large bowel, this problem tends to be less marked [1, 16, 49]. A number of studies conducted in patients with SBS who have been treated with EN have revealed identical effectiveness of oligomeric and polymeric diets in terms of nutrient absorption, fluid and electrolytes loss [46, 47]. In patients with a high jejunostomy (90–150 cm remaining jejunum), better protein absorption was observed with an oligopeptide diet compared to whole protein mixtures. Despite the higher osmolarity of the peptide diet, fecal losses did not increase and lean body mass and electrolyte status remained constant [50]. Polymeric enteral nutrition diets are less expensive and less hyperosmotic than oligomeric mixes and are generally well tolerated. In a model of SBS in animals that had a preserved segmented intestine, it was shown that polymeric enteral diets can also effectively contribute to intestinal adaptation.*

A study conducted in 15 adults with SBS (3–130 months after the last surgery, 4 patients without segmented intestine) showed that drip enteral probe nutrition for 7 days in isolation or in combination with oral nutrition increased intestinal macronutrient absorption compared with isolated oral nutrition. Increasing the energy supply by about 400 kcal/day can be achieved by gradually increasing the oral intake of enteral diets to 1,000 kcal/day [51].

Recommendation 8. In the absence of encephalopathy, gastrostasis and preserved swallowing function on the 2–3 day postoperative period, the transition to split oral intake of enteral nutritional mixtures in small sips (sipping) with subsequent (4–5 days) prescription of gradually increasing volume of sparing medical diet with elements of diet with heat untreated food and separate fluid intake is possible. Powdered polymeric or oligomeric (semi-elemental) enteral solutions can be added to prepared meals to increase the biological value of the therapeutic diet of patients with SBS [34, 52]. Levels of evidence — 3, grades of recommendation — B.

Comments. Even if the patient is predicted to be unable to receive an acceptable substrate supply through the GI tract for 5–7 days (less than 50 % of need) and therefore parenteral nutrition is the mandatory method of choice for nutritional support, any opportunity for enteral administration of nutrients should always be considered. Specialized oligomeric diets are not always the only option for EN. Standard isocaloric isonitrogenic isoosmolar diets containing predominantly “fast” proteins (whey or plant proteins), which are relatively quickly evacuated from the gaster and easily hydrolyzed, can be used as starter nutrition in most patients. When choosing standard mixes, it is also important to consider the composition and fat content. Preference should be given to NMs with lower fat content or mixtures in which a certain part of it (15–50 %) is represented by medium chain triglycerides, which do not require bile acids and pancreatic lipase for rapid digestion. The indication for the prescription of oligomeric diets may be poor tolerance of isocaloric polymeric mixtures or the presence of patients with initial severe hypotrophy (BMI less than 16 kg/m²), which is often accompanied by phenomena of fermentopathy. Enteral nutrition has a trophic effect on the intestine and prevents mucosal atrophy, plays an important role in preserving the intestinal immune system, as well as in preventing ascending microbial colonization of the proximal small bowel and minimizing the risk of bacterial translocation. If patients with SBS are gradually transferred to a mechanically and chemically sparing diet to increase the biological value of the diet, additional prescription of ED by sipping or by adding them in powder form to ready meals is indicated [17, 28, 34, 52].

Recommendation 9. When determining the energy and protein requirements in the early postresection period in most patients with SBS an empirical approach is possible: energy — 25–30 kcal/kg, protein — 1,2–1,5 g/kg/day. Indirect calorimetry makes it possible to more accurately determine the energy requirements of patients, and determination of daily nitrogen loss makes it possible to most accurately

estimate the protein requirements of patients with SBS and IF [14, 53]. Level of evidence — 2, grade of recommendation — B.

Comments. The effect of malnutrition on the incidence of postoperative complications and mortality is well documented in both retrospective and prospective studies. Two systematic analyses shown that for hospitalized patients in general and for those undergoing surgery, PEM is an independent risk factor for complications and is also directly related to length of hospital stay, the cost of their care, and increased mortality [34, 51, 54]. The average energy and protein requirements of a stable surgical patient are most often 25–30 kcal/kg and 1.2–1.5 g/kg per day. There should be at least 90–100 non-protein kcal for every gram of nitrogen. The energy ratio of glucose/fat should not be less than 60/40 (70/30 is better), and lipids should not be administered more than 1 g/kg per day. With parenteral nutrition, it is necessary to strictly observe (not exceed) the prescribed rate of administration of nutrient substrates (amino acid no more than 0.1 g/kg/hour, fats no more than 0.15 g/kg/hour and glucose no more than 0.5 g/kg/hour). In overweight and obese patients, substrate requirements should be calculated by the recommended (ideal) body mass, and in the presence of severe hypotrophy (BMI <16 kg/m^2) by the actual BM + 20–30 %. Indirect calorimetry (metabolic monitoring) makes it possible to more accurately determine the energy requirements of patients, and determination of daily nitrogen loss makes it possible to most accurately estimate the protein requirements of patients [14, 28].

Recommendation 10. When the anatomical integrity of the ileocolic valve is impaired (distal SBS, jujuno- or ileocolonic anastomosis), as well as initially poor tolerance of oligomeric EDin the form of increased intestinal dyspepsia, intraintestinal decontamination is indicated for 5–7 days [55, 56]. Level of evidence — 2, grade of recommendation — B.

Comments. The main function of the ileocecal valve is to allow portions of intestinal chyme from the ileum into the cecum, preventing the contents of the large bowel from flowing back into the small bowel. When anileocolonic anastomosis bypasses this valve (ileocolic valve), it creates real conditions for upward (reflux) contamination of fecal, including opportunistic microflora in the proximal small bowel (small intestinal bacterial overgrowth syndrome — SIBOS). Bacterial pool of colonic flora, which has changed its site, causes damage the small bowel mucous, inflammatory phenomena, disorders of its barrier function, as well as the activity of digestive enzymes and premature deconjugation of bile acids, that is accompanied by intensification of secretion processes, development or intensification of maldigestion, malabsorption and intestinal dyspepsia (tympanites, hyperactive bowel sounds, abdominal pain, watery diarrhea, steatorrhea, creatorrhea, amylorrhea). Catheter-associated infection may develop as a consequence of the small bowel mucosa barrier function disorder often presenting in SIBOS and translocation of opportunistic microflora into the systemic bloodstream [57–62]. The prevalence of bacterial overgrowth in the small bowel in various gastrointestinal diseases and in the consequences of digestive surgery is 40–99 % [63, 64]. The most common cause of increased intestinal dyspepsia in patients with SBS when using oligomeric ED is SIBOS. Protein peptides of oligomeric mixtures serve as a good breeding ground for growth and rapid development of opportunistic microflora located in the proximal parts of the small bowel, which is accompanied by hyperproduction of microbial toxins, increased intestinal permeability, intraintestinal secretion, its motility and increased diarrhea. Intestinoscopy with aspiration of small bowel contents and inoculation of the aspirate on nutrient media is considered to be the "gold standard" for diagnosis of SIBOS [59, 62, 64, 65]. 5 randomized studies shown the efficacy of antibacterial intraintestinal decontamination in the treatment of SIBOS. The most commonly used drugs for this purpose are metronidazole, rifaximin, nifuroxazide, and fluoroquinolones [55, 56].

Recommendation 11. With preservation of swallowing function, proper level of consciousness, stabilization of gastrointestinal motor function, presence of stool and good tolerance to ED, the volume of oral split (5–6 times/day) consumption of sparing therapeutic diets with elements of diet with heat untreated food and adding to ready meals powdered polymeric or oligomeric enteral nutritional solutions is expanded [34]. Level of evidence — 3, grade of recommendation — B.

Comments. An exception may be cases in which there are proximal fistulas with high production (500 mL/day or more) of intestinal *content, when temporary oral fasting, creating functional rest of the fistula area, promotes its healing. Enteral nutrition in such a situation is possible only when a nasointestinal probe is placed distal to the fistula area [34, 52].*

Recommendation 12. In short and especially super-short small bowel syndrome, separate intake of food and fluid is recommended. Fluid should not be consumed 30 minutes before and within 30–45 after meals. In patients with moderate dehydration or low serum sodium, it is reasonable to use hypo- or isoosmolar glucose-salt solution or saline supplements for oral rehydration and compensation of intestinal losses [62, 66]. Levels of evidence — 3, grades of recommendation — B.

Comments. On the one hand, fluid intake accelerates the evacuation of food consumed from the gaster and accelerates the transit of intestinal chyme, and on the other hand, causes a decrease in the concentration of intraintestinal digestive enzymes that perform intracavitary and partially intestinal hydrolysis of nutrient materials. This leads to a disorder of substrate and enzyme relationships, incomplete hydrolysis of nutrient materials, increased intraintestinal osmotic pressure and finally to the aggravation of intestinal dyspepsia. Adherence to a restrictive diet with elements of relative diet with heat untreated food and preferential hydration of patients with SBS during the interdigestive period helps to reduce the severity of intestinal dyspepsia [8, 28]. Correction of high discharge from the small bowel (ileostoma or high fistula) is best started by restriction of total oral intake of hypotonic fluids (water, tea, coffee) as well as hypertonic fluids (fruit juices, Coca-Cola and most commercial sipping enteral diets with osmolarity above 400 mosmol/L) to 500 mL per day. To compensate for the rest of the fluid need, the patient is advised to drink glucose-salt solutions with a sodium content of 90 mmol/L or more [67].

Many patients at home with significant loss of intestinal contents through the stoma (1–1.5 L) may benefit from a combination of restricting oral fluid intake (less than 1 liter per day) and adding salt to their diet. Patients with a loss of less than 1,200 mL per day can usually maintain sodium balance by adding extra salt (5–6 g per day) at meal times or during it preparation. When losses are in the 1,200–2,000 mL range, and sometimes more, the patient can maintain sodium balance by taking glucose saline or saline supplements [68].

Recommendation 13. Insoluble dietary fiber (soy polysaccharide, resistant starch, microcrystalline cellulose, lignin) should be limited or excluded from the diet of patients with SBS and IF. When the large bowel and especially the ileocolic valve are preserved, soluble dietary fiber (inulin, pectin, oligosaccharides, gum) can be used, taking into account their tolerability, which have prebiotic and trophic effects. Level of evidence — 3, grade of recommendation — C.

Comments. The effect of dietary fiber on diarrhea depends on which part of the intestine the patient has retained. If the large bowel has not been resected and most carbohydrates can be digested and absorbed in the small bowel, the addition of soluble dietary fiber can increase fluid absorption and decrease stool bulk. Moreover, soluble dietary fiber has a bifido- and lactogenic effect, since it is the main nutrient substrate for these bacteria. As a result of microbial hydrolysis of soluble dietary fiber by indigene microbiota, short chain fatty acids (butyrate, acetate, propionate) are formed, which have a trophic effect, primarily on epithelial cells of the large bowel, which improves fluid and electrolyte absorption. However, if amylorrhea, indicating incomplete hydrolysis and absorption of carbohydrates, is present in patients with SBS and IF, then soluble dietary fiber may increase intestinal dyspepsia [28, 50, 69].

Recommendation 14. Probiotics should not be added to the ED in order to force the adaptation of the small bowel. Metabiotics can be an effective method of controlling the intestinal microbiota and preventing small intestinal overgrowth syndrome, as well as trophic effects on the mucosa in patients with SBS and IF [70, 71]. Level of evidence — 3, grade of recommendation — B.

Comments. One of the frequent complications of SBS at extensive (over 50 %) resection of the small bowel with ileocolonic anastomosis, especially in combination with right-sided hemicolectomy, is SIBOS in the small bowel, resulting in aggravation of functional disorders of the digestive-transport conveyor and intestinal dyspepsia phenomena. The main mechanism of increased microbial contamination of the proximal small bowel in the absence of the ileocolic valve is reflux of colonic contents into the small bowel, which is usually accompanied by local inflammation and increased intestinal *permeability (IP), based on the impaired barrier function of the small bowel mucosa. IP is accompanied by episodes of transient translocation of opportunistic microflora and their toxins from the intestine into the systemic bloodstream, which can lead to various infectious complications, up to sepsis [72]. When relieving intestinal dyspepsia in patients with SBS, restoration and support of intestinal microbiocoenosis with pre-, meta- and probiotics is of great importance, along with dietary nutrition, antimicrobial decontamination and enterosorption [60, 62, 65, 73, 74]. The use of probiotics for rehabilitative purposes in SBS has not been evaluated. Several case-specific publications described the use of probiotics in SBS to treat D-lactate acidosis [75]. However, cases of probiotic bacteremia are described in adults and children due to their translocation into the systemic bloodstream against a background of increased intestinal permeability [70, 71]. In a systematic review of studies in pediatric patients, for example, the authors concluded that there are no sufficient data on the effects of probiotics in children with SBS and that the safety and effectiveness of probiotics in this high-risk cohort should be evaluated in subsequent large studies [76].*

In this regard, metabiotics, which are structural components of probiotic microorganisms and/or their metabolites that can optimize specific regulatory and metabolic intraintestinal processes aimed at supporting the barrier function of the intestine and preserving the indigene microbiota of the host with an antagonistic effect against opportunistic flora, have become widely used in high-risk groups of translocation-dependent probiotic infection [77]. Randomized studies to evaluate the effectiveness of metabiotics in patients with SBS and IF have not yet been conducted.

Recommendation 15. Patients with a preserved large bowel should receive a diet low in long chain triglyceride fats, and limit mono- and disaccharide intake [78]. Level of evidence — 3, grade of recommendation — B.

<u>*Comments.*</u> *A low-fat diet is preferable for patients with SBS and IF because long chain triglycerides (LCTs), when hydrolyzed by pancreatic lipase with the obligatory participation of bile acids, are absorbed mainly in the jejunum and proximal ileum. In patients with ileocolic anastomoses, unabsorbed LCTs entering the large bowel shorten intestinal chyme transit time and decrease fluid and sodium absorption, which can exacerbate diarrhea. In addition, LCTs are toxic to saccharolytic intestinal microflora and inhibit their growth, which reduces the processes of carbohydrate fermentation. They bind to calcium and magnesium, increasing stool loss, and increasing the absorption of oxalates, which predispose to kidney stone formation [78]. In this regard, consumption of fats based on long chain triglycerides is recommended to be limited to 20–25 % of the total energy requirement. A low-fat diet can increase the absorption of calcium, magnesium, and zinc, but increases essential fatty acid deficiencies. Since these fats are the most energy-intensive macronutrient (1 g — 9.3 kcal), the energy deficit of the daily diet can be compensated by carbohydrates (up to 60 % of the total energy requirement). In additional note t that the use of simple mono- or disaccharides (sucrose) in such patients is better to limit, because they, increasing osmolarity of intestinal chyme, on the one hand, may increase intraintestinal secretion and diarrhea, and on the other hand, being easily accessible substrate for intestinal microflora, cause development of D-lactate acidosis and nephrolithiasis. Such patients need a diet containing carbohydrates with a low glycemic index (polysaccharides) and low oxalate content [6]. If necessary, soluble and easily digestible hydrolyzed starch in the form of a maltodextrin module can be added to the diet of such patients [1]. The amount of energy consumed by patients with SBS can also be increased by including in their diet fats based on medium chain triglycerides (MCTs) in the amount of 0.3–0.5 g/kg per day, which even with impaired bile secretion and lipase deficiency are relatively easily absorbed and enter the portal vein, quickly included in energy metabolism (1 g MCT — 8 kcal). Remember, however, that MCTs do not contain polyunsaturated fatty acids. Therefore, if MCTs are the predominant fat energy substrate, at least 2 % of their total energy value must be provided by essential fatty acids (4–8 g per day) [28, 79].*

Recommendation 16. Early parenteral nutrition is prescribed to patients in the first 48–72 hours after massive resection of the small bowel in parallel with the ongoing enteral therapy, a part of which is minimal enteral nutrition, when initially it is obvious that the necessary substrate supply of patients through the GI tract is impossible for the next 5–7 days. Main condition for prescribing PN is restoration of

fluid and electrolyte balance [80, 81]. Level of evidence — 3, grade of recommendation — B.

Comments. *Infusion therapy to maintain proper FEH and parenteral nutrition in patients with SBS and IF are the basic methods of treatment in the immediate and early postoperative periods. Prognosis in the need for long-term intravenous support of fluid balance and providing patients with nutrient substrates is often difficult to predict, since the potential for structural and functional adaptation of the remaining part of the small bowel depends on many factors: the length of the residual intestinal segment, the place of its resection, presence or absence of ileocecal valve, age, underlaying and concomitant diseases, the initial state of patient nutrition. If the length of the remaining part of the small bowel is more than 150–180 cm, then even in the absence of ileocecal valve in most cases, if patients comply with the prescribed fluid intake and dietary regimen with additional consumption of enteral nutritional mixtures, there is no need for infusion therapy and PN. If the length of the small bowel is between 60 and 150 cm (with or without the segmented intestine), patients require PN at least in the early postoperative period (the next 3 months) and often longer. If the residual segment of the small bowel is less than 60 cm (regardless of the presence of the segmented intestine), long-term (years and sometimes lifelong) administration of intravenous infusion therapy and parenteral nutrition is usually required [10, 23, 68, 80]. Thus, the main indication for prescribing maintenance PN in this category of patients is the lack of necessary intestinal adaptation, which does not allow to achieve an acceptable enteral autonomy, allowing absorption of sufficient fluids, electrolytes and nutrients. Clinical criteria for the severity of intestinal failure in patients (type II or III) and the need to prescribe or continue intravenous infusion and nutritional support are: The presence of persistent intestinal dyspepsia, manifested by repeated diarrhea with detectable coprologic signs of maldigestion (steatorrhea, amylorrhea, creatorrhea), despite the observed dietary nutrition; recurrent fluid and electrolyte disorders (hypovolemia, dyselectrolymia) requiring intravenous correction; presence of progressive weight loss more than 2 % per week or 5 % per month, as well as persistent hypoproteinemia (hypoalbuminemia) [1, 8, 28, 80]. In a prospective study and follow-up of patients with SBS and IF of non-oncological etiology (n = 124) over 5 years, 55 % of them achieved enteral autonomy and complete cancelled PN. The great majority of them (49 %) had their PP cancelled for 2 years. The probability of PN cancellation in patients with chronic intestinal failure at a later date was only 6 %. The key factors determining the course of chronic IF were the length of the residual small bowel segment < 100 cm and the presence of an end jejunostomy or ileocolic anastomosis. The substitutable amino acid citrulline produced by enterocytes can serve as a marker of the severity of existing intestinal failure. Plasma citrulline content < 20 umol/L tended to correlate with PN dependence more than 2 years after small bowel resection [1, 8, 81–84].*

Recommendation 17. Parenteral nutrition in some patients with SBS and baseline normal weight or overweight with moderately severe intestinal dyspepsia and maldigestion (body mass reduction less than 2 % per week or 5 % per month with preserved or moderately reduced visceral proteins) may be prescribed delayed, after several weeks, if indicated. Such patients, along with oral dietary nutrition, are recommended an additional administration of enteral nutritional mixtures in the amount of 500–600 kcal and 20–40 g of protein per day. In all cases of continued body mass reduction of more than 10 % of its initial value and/or the development of hypoproteinemia less than 60 g/L (hypoalbuminemia less than 30 g/L) on the background of persistent intestinal dyspepsia should consider the need for additional parenteral nutrition. Level of evidence — 3, grade of recommendation — C.

Comments. *In some patients with SBS and IF parenteral nutrition can be started at a later stage, sometimes after several weeks or even months of initially moderate manifestations of intestinal dyspepsia, if at the initial stage of their treatment dietary nutrition including enteral NMs (sipping), as well as pharmacological therapy (antisecretory agents, decontamination, enzymes, sorbents, etc.) allow to partially control impaired digestive processes at an acceptable level (moderate BM reduction, hypoproteinemia and/or hypoalbuminemia, orally maintained by FEH). However, over time, some patients may experience further progression of intestinal failure (most often in elderly patients), which prevents the achievement of sustained intestinal autonomy. In these patients, nutritional deficiencies continue to progressively increase, which is an indication for prescribing (most often temporary) additional PN [81–84].*

Recommendation 18. For parenteral nutrition of patients with SBS and IF, especially in dispensary health settings, it is recommended to use "all-in-one" containers [82, 83]. Level of evidence — 3, grade of recommendation — B.

Comments. According to the recommendations of the European Society of Clinical Nutrition and Metabolism (ESPEN), parenteral nutrition mixtures, especially at home, should be administered using "all-in-one" containers. Standardization of parenteral nutrition components allows the physician to quickly select the type of three-in-one system by knowing the amount of protein, glucose, fat, and energy value of the finished system. This "standardization" of PN allows to reduce the frequency of complications and the cost of its implementation. According to experts, three-in-one containers should be used for PN in MPI in 80 % of cases, and only 20 % of patients require individual selection of nutritional mixtures using the separate vial option. For parenteral nutrition at home, three-in-one systems are the only safe way to provide macronutrients and micronutrients to patients. In cases of poor tolerance of fat emulsions (hypertriglyceridemia, hepatopathy) two-in-one containers containing amino acid and glucose solutions can be used. In PN without fat emulsions, a deficiency of essential polyunsaturated fatty acids will develop after 2–6 months. To relieve their deficiency is recommended twice a week soybean oil at the rate of 1.2–1.5 g/kg body mass. The daily requirement for essential fatty acids is 7–10 g/day, which corresponds to 15–20 g of LCT from soybean oil (1st generation fat emulsions) or 30–40 g of LCT from fat emulsions of the 2nd and 3rd generations [79, 85]. A deficiency of essential fatty acids can be prevented by administering about 500–1,000 mL of 20 % fat emulsions per week. Deficiency of essential fatty acids can be avoided with regular oral intake [39, 86, 87].

Recommendations 19. If there is a need for prolonged (more than 10 days) parenteral nutrition as the main method of nutrient substrates administration, the prescription of specialized multivitamin and micronutrient complexes designed for intravenous administration is indicated [86]. Level of evidence — 3, grade of recommendation — B.

Comments. All-in-one containers do not contain vital micronutrients (vitamins and trace elements), which are cofactors of all biochemical processes occurring in the human body. Studies showed that low intake and micronutrient deficiencies are associated with increased morbidity. On the contrary, restoration of their adequate intake led to normalization of nutritional status and reduction of morbidity. Studies conducted on healthy elderly patients who received individualized vitamin and micronutrient supplementation showed a reduction in the incidence of infectious diseases for more than a year. The authors attributed such an effect to an improvement in the body's protective functions. A large number of studies are devoted to the antioxidant defense of the body. Normalization of vitamins C and E help to reduce oxidative damage, which is associated with the restoration of enzymatic and non-enzymatic antioxidant systems, as well as improving the functional stability of lipids in cell membranes. Vitamin D accelerates the absorption of calcium and phosphorus in the intestine, necessary for normal bone mineralization processes, has a regulatory effect on calcium transport through biomembranes. Folic acid is involved in protein and nucleic acid biosynthesis, methylation reactions and metabolism of several amino acids (serine, glycine, histidine, methionine), is especially important for growth, development and differentiation of cells and tissues with high rate of renewal (blood formation, intestinal mucosa), has a lipotropic effect. Micronutrients also act as cofactors in most of the biochemical processes occurring in the body. For example, iron is a component of almost all respiratory enzymes, hemoglobin and myoglobin, takes part in the synthesis of DNA and thyroid hormones, and supports immunoreactivity. Zinc is involved in protein and nucleic acid synthesis, affects bone calcification processes, contributes to cell membrane stabilization and immunogenesis adequacy, and selenium has a marked antioxidant effect, prevents genetic disorders in DNA, promotes their differentiation, stimulates immunogenesis and enhances reparative processes [28, 87, 88].

Recommendation 20. Dipeptide glutamine solutions should be used for total parenteral nutrition in patients with SBS and intestinal failure in the early postresection period [89, 90]. Level of evidence — 2, grade of recommendation — B.

Comments. L-glutamine, being the most important energy substrate for intestinal epithelial cells, prevents mucosal stress atrophy and increased intestinal permeability, reduces the frequency and severity of bacterial *translocation, has a powerful antioxidant and cytoprotective effect. At the same time, it has a pronounced nitrogen-saving effect, enhances muscle anabolism and increases the activity of immunocompetent cells. The main "consumers" of glutamine, in critical patients, are epithelial cells of the intestinal tract mucosa (10–14 g/day) [1, 8, 28]. In 7 randomized clinical studies (2009) conducted with surgical patients who received only PN with the addition of glutamine dipeptide in a standard dosage of about 0.5 g/kg/day, its effect on the course of the pathological process and its outcome was analyzed [88–91]. Six studies studied patients who underwent elective surgery and one emergency surgery. All studies showed significant benefits of glutamine supplementation with regard to reducing the duration of postoperative hospital stay (5 studies) and reducing the incidence of complications (2 studies). In an earlier meta-analysis (2002), the authors also noted significant positive benefits of glutamine addition with regard to both the incidence of infectious complications (10 studies) and the reduction of patient treatment duration (8 studies) [89]. In experimental models of SBS combined with resection of the ileocecal angle and the large bowel, the use of glutamine as part of PN prevented the development of atrophy of the remaining intestinal mucosa, led to a decrease in translocation of intestinal flora, increased secretory IgA levels [90]. Contraindications to intravenous administration of glutamine solutions are: severe hepatic and renal failure (creatinine clearance less than 25 mL/min), and severe metabolic acidosis.*

Recommendation 21. Patients with SBS and IF receiving long-term PN (months, years) are recommended to use fatty emulsions (FE) of the second and third generation. Prolonged use of a soybean oil-only (first-generation) FE of more than 1 g/kg/day is associated with a higher risk of hepatic complications. The use of FE containing fish oil can contribute to the reduction of the resulting hepatopathy [92]. Level of evidence — 2, grade of recommendation — B.

Comments. Liver damage, which is often present in patients with intestinal failure, can be exacerbated by prolonged parenteral nutrition. Over the past 20 years, experimental and clinical studies showed that the use of traditional lipid emulsions based only on soybean oil represents an increased risk factor for liver damage in patients with SBS and IF [93]. The role of phytosterols (plant cholesterol-like compounds that are found in large quantities in soybean oil-based FEs and can interrupt bile acid homeostasis with the development of cholestasis is actively discussed) [24, 79]. Doses of intravenous FE based on soybean oil alone ≥1 g/kg/day have been strongly associated with an increased risk of hepatopathy in mixed cohorts of adults and children receiving HPN [25]. Pure soybean fat emulsions are not recommended in routine clinical practice for long-term (>6 months) HPN. MCT/LCT and emulsions containing fish oil demonstrate greater safety [79, 94]. The use of the latter minimizes the risk of hepatic complications. According to the latest (2020) expert consensus statements of the international summit "lipids in parenteral nutrition" in cases of development of hepatic complications during long-term use of FE on the basis of soybean oil it is recommended to transit to the use of mixed emulsions containing fish oil, which can contribute to the reduction of cholestasis and/or cytolysis phenomena [92].

Recommendation 22. To reduce or eliminate dependence on intravenous administration of fluid and nutrient substrates and to achieve enteral autonomy in patients with SBS and IF, it is possible to use recombinant analog of glucagon-like peptide-2 (GLP-2), which has a marked trophic effect on the regenerative potential of epithelial cells and structural and functional adaptation of intestinal mucosa [98–100]. Level of evidence — 2, grade of recommendation — B.

Comments. After the small bowel resection there is a relatively long process of its structural and functional adaptation. Structural adaptation affects all layers of the intestinal wall and involves proliferation of cells in the crypts, an increase in the height of the villi, the ratio of crypt length to villi length, mucosa absorption surface area and mass, as well as an increase in the lumen diameter and thickening of the intestinal wall. Functional adaptation consists of an increase in the rate and volume of absorption, delayed gastric emptying and increased transit time of intestinal contents, increased rate of transport of nutrients through mucous cells, as well as changes in the composition of pancreatic and bile secretions. The process of adaptation begins almost immediately after an extensive bowel resection and can last more than 2 years. Enteral autonomy through natural structural and functional adaptation of the remaining intestinal fragment is not achieved in allpatients *with SBS and IF [16, 19, 29, 80]. Patients with SBS and IF type I or II according to the ESPEN functional classification can achieve enteral autonomy by natural adaptation of the remaining intestine. The minimum residual length of the small bowel required to achieve possible enteral autonomy and wean patients off parenteral nutrition is about 100 cm in end jejunostomy, 60 cm in ileocolic anastomosis and 35 cm in ileocolic (jejuno-ileo) anastomosis. Patients with preserved large bowel are less dependent on parenteral nutrition and usually have a better prognosis [97]. In type III SBS-IF, irreversible intestinal failure occurs in 50 % of cases, requiring prolonged, often lifelong, intravenous support through regular administration of fluids, electrolytes and nutrient substrates. In order to reduce or eliminate the dependence of stable patients with SBS and IF on parenteral fluids and nutrition, conservative and surgical methods can be used at various stages of their treatment. For conservative therapy of SBS-IF, a synthetic recombinant analogue of glucagon-like peptide-2 (teduglutide) is used, which can be prescribed for patients aged 1 year and older. To evaluate its clinical efficacy, 17 randomized, placebo-controlled clinical trials were conducted. The studies included 595 patients with SBS and IF who were dependent on PN for 12 months at least 3 times a week. Teduglutide was administered by subcutaneous injection at a dose of 0.05 mg/kg/day for an average of 21.8 weeks. Against the background of its regular use there was a significant increase in plasma citrulline (an indirect marker of mucosal villous growth) compared with baseline and compared with the placebo control group of patients (20.6 ± 17.5 umol/L and 0.7 ± 6.3 umol/L respectively), which led to a significant improvement in intestinal absorption of nutrients [6, 98]. The requirement for PN when GLP-2 was used for 24 weeks or more decreased from the mean at baseline from 13.4 to 3.7 L/week, and the mean number of days of infusion decreased from 5.7 to 2.7 days. It was possible to completely overcome dependence on maintenance parenteral nutrition during the period of treatment lasting 7–18 months in 15% of patients, at the end of the 24-month continuous course of treatment — in more than 20% [99, 100]. The effectiveness of teduglutide therapy should be evaluated no earlier than after 6 months of treatment. Limited research data indicate that some patients may respond to therapy after a longer period of time. If overall improvement is not achieved after 12 months of therapy, then the likelihood of continuing treatment should be reassessed. In patients with SBS and IF against the background of long-term use of GLP-2 there was a decrease in the incidence of various complications, improved nutritional status and quality of life [101, 102]. The indication for teduglutide is the continued need for parenteral nutrition for at least a year or the impossibility of its proper implementation due to complications (recurrent catheter-associated bloodstream infection, multiple vascular occlusions, severe liver damage). The PN requirement increased from 4.0 to 5.5 L within 4 weeks of GLP-2 cancellation, while plasma citrulline content decreased by 20 %, indicating the need for continued use. Some studies showed a higher efficacy of combined use of growth hormone and glutamine in patients with SBS and IF, which have a synergistic effect on the structural and functional adaptation of the remaining part of the small bowel. Byrne T.A. et al. observed that SBS patients receiving growth hormone, glutamine and modified diet simultaneously for 3 months had the statistically significant ($p < 0.005$) highest reduction of PN volume (7.7 ± 3.2 L/week), calories (5,751 ± 2,082 kcal/week) and infusions (4.0 ± 1.0 inf/week) compared to the group on diet and growth hormone alone (volume 5.9 ± 3.8 L/week calories 4,338 ± 1,858 kcal/week; infusions 3.0 ± 2.0 weeks) or the group receiving diet combined with glutamine (volume 3.8 ± 2.4 L/week; calories 2,633 ± 1,341 kcal/week; infusions 2.0 ± 1.0 week) [43, 103, 104]. Surgical treatment can be aimed at slowing intestinal transit and increasing the absorption surface of nutrients [105, 106].*

Recommendation 23. For long-term parenteral nutrition, the safest venous accesses (subclavian or internal jugular vein) and long-term venous catheters (peripherally inserted central venous PICC catheters), implantable ports and tunneled catheters) are used, which reduce the risk of infectious complications [105, 106]. Levelof evidence — 3, grade of recommendation — B.

Comments. Home PN requires a well-functioning central venous access. When considering which type of central venous device is best, several aspects must be considered: the number of weekly infusions, duration of therapy (temporary or lifelong), diagnosis of underlying disease (benign or not), previous history of central venous access, and existing experience. The patient's age and daily activities, as well as their own wishes regarding the type of catheter/port should be taken into account. Upper *cava catheterization with a tunneled catheter is the most commonly used method in many countries for long-term, longstanding parenteral nutrition. Most centers use silicone Hickman or Broviac catheters with an open distal end and a dacron cuff in its proximal part, which is placed directly under the skin. Within 3–4 weeks after placement under the skin, the cuff is hermetically fused to the subcutaneous tissue and the skin peculiarly for a month, thereby preventing microbial colonization of the catheter tunnel along its entire length from the skin inlet to the vein, which minimizes the risk of local infectious complications. The catheter can have from 1 to 3 lumens. The advantages of tunneled catheters are that they can remain in place and be used for many years, and their connection to nutrition solution containers can be relatively easily performed by the patient himself, as it does not require skin puncture as it is necessary with an implantable port and allows the patient to use two hands, which extends his ability to implement PN at home by himself, which, for example, is impossible with the PICC catheter. Installation of such catheters is indicated when there is a daily need to administer nutrient substrates and fluids. The disadvantage of this catheter is its protruding external part at the place of its exit from the subcutaneous tunnel [107, 108]. Another option is to use a fully implantable port for parenteral nutrition that includes a silicone or polyurethane catheter connected to a compact one- or two-lumen chamber made of titanium or durable plastic, through which nutrient substrates and fluid are injected. The port chamber is implanted into a subcutaneous pocket on the front wall of the ribcage, located 5–10 cm from the central vein puncture site. The silicone diaphragm of the chamber is suitable for repeated (up to 2,000–3,000) punctures with the obligatory use only of a special Huber needle, the tip of which is sharpened in a special way, which allows not to damage the diaphragm at the moment of puncture. The needle in the port system reservoir can stay up to 7 days, after which it must be replaced. The catheter, departing from the camera, is placed in a subcutaneous tunnel running from the place of its insertion into the subclavian (jugular) vein with the tip placed on the border of the superior vena cava and the right atrium. The advantage of the port system is that it is completely covered by the skin, which makes it virtually invisible, as it does not change the appearance of the patient's body surface [105, 106]. Studies in intensive care units showed that catheterization of the subclavian vein is associated with a lower incidence of infection compared with catheter insertion into the jugular vein. The use of the HPN port is more indicated when periodic administration of nutrient substrates and fluids is required 2–3 times per week. Peripherally inserted central venous catheter Groshong (PICC catheter) is intended for short-term use and cannot be recommended for long-term (more than 6 months) PP at home. Single-lumen catheters are preferred to minimize the risk of CLABSI [35]. In patients with superior vena cava thrombosis, femoral vein catheterization is required, but the risk of mechanical complications and thrombosis is about 10 times higher than in the case of subclavian access [109]. In 289 patients, complications from the use of various long-term central venous catheters over 50,000 days were studied and found that the incidence of catheter-associated infections was lowest (0.35/1,000 days of catheter use) with the use of implantable ports (0.19/1,000) [110]. Antimicrobial-coated catheters have the potential to reduce central catheter colonization, but no benefit has been identified with respect to clinically diagnosed sepsis or associated mortality [109]. Adherence to a clear catheter/sports care protocol helps to minimize the incidence of angiogenic sepsis in patients with SBS and IF with long-term parenteral support [111]. The main indications for the removal of a long-term use CVC/port are: disruption of its integrity; inflammatory changes in the skin at the place where the CVC exit from the skin and intractable tunnel infection; catheter infection with no effect of systemic antibiotic therapy; obstructive catheter lumen thrombus or signs of thrombophlebitis in the place of its localization.*

Recommendation 24. Regardless of the type of catheter used, the location of the catheter tip using internal jugular or subclavian access should be near the junction of the superior vena cava and the right atrium, which reduces the risk of thrombosis [112, 113]. Level of evidence — 2, grade of recommendation — B.

Comments. A retrospective analysis of patients receiving PN for a long time showed that catheters with "adequately positioned" tip had the lowest relative risk of thrombosis (0.26 %) compared to poorly positioned CVCs [112]. A retrospective review of 428 randomly selected CVCs noted that only 2.6 % of patients had thrombus formation when the catheter was placed at the border of the right atrium and *superior vena cava (SVC). Thrombus formation was detected in 5.3 % of cases when the catheter tip was located in the middle of the SVC and in 41.7 % of cases when it was located in the proximal third of the SVC [113]. Patients with right-sided insertion had a lower risk of cardiovascular thrombosis (relative risk ≥0.39) compared with patients with left-sided insertion [112]. In all cases of planned contact with the CVC, hands should be treated with appropriate disinfectants. For patients with a stoma or fistula, it is important that stoma and fistula care be separated in time from catheter care. Care of the catheter exit site, including treatment (cleaning) of the catheter itself, should be performed regularly, at least once every 7–10 days, and every time the dressing becomes wet or contaminated [114].*

Recommendation 25. Adding heparin or antibiotics to the central venous catheter flushing solution and ensuring catheter blockage is not necessary. In parenteral nutrition of patients through long-term catheters, the lowest incidence of catheter bloodstream infection is observed when physiological saline is used as a postinfusion catheter lock, and taurolidine or 70 % ethanol is used in the presence of CLBSI risk factors [115, 116]. Levelof evidence — 2, grade of recommendation — B.

<u>Comments.</u> *Vascular catheters are a major cause of primary bloodstream infections. There are 2 main mechanisms of CLBSI development: colonization by bacteria living on the skin or external surface of the catheter through the skin entry wound, when there are signs of soft tissue inflammation with spread of infection to its internal (intravascular) end, and infection of the catheter internal surface due to contamination of the catheter-infusion system junction. CLBSI is established in the presence of systemic clinical manifestations of infection (febrile hyperthermia with algidity and/or hypotension) with primary bacteremia or fungemia in the absence of other obvious sources of infection and isolation of the same microorganism from the catheter surface by quantitative or semiquantitative methods as from blood [117]. Numerous methods of preventing CLBSI have been tested and implemented, including the use of several variations of flushing solutions and different types of catheter blocking. Antibiotics, singly or in combination, and antiseptics such as ethanol, taurolidine, and trisodium citrate were used for this purpose. These drugs are often combined with an anticoagulant such as heparin or EDTA. The results of some meta-analyses showed that heparin flushing of CVCs that are not used for blood flow does not prolong their use compared to normal saline [115, 118]. Preservative-free heparin at concentrations <6,000 U/mL has no antibacterial properties and may even promote catheter colonization and biofilm growth [116, 119]. A randomized study comparing the effect of a low-dose heparin (300 U/3 mL) with 0.9 % saline conducted with 750 cancer patients with a newly installed port showed no significant differences between the groups in terms of primary outcomes (ease of injection, possibility of aspiration), but the rate of catheter infection was significantly lower in the saline group (0.03/1,000 days versus 0.10/1,000 days in the heparin group). There is also no evidence that preventative antibiotic use reduces the incidence of catheter infection in patients receiving HPN, while this strategy carries an inherent risk of developing microbial resistance, especially in patients who require long-term HPN [120]. According to a literature review by Wouters Y. et al. (2019), taurolidine (0.13 per 1,000 catheter-days) had the highest 1-year survival rate (97 %) in patients with short bowel and central venous port without CLBSI. Taurolidine can prevent colonization of the inner surface of catheters by a wide range of microbial pathogens and prevent the development of life-threatening cases of catheter-associated sepsis. When saline and heparin was used the frequency of CLBSI was 0.74 and 2.01, per 1,000 catheter days consequently. Studies show that saline may be the second best option (after taurolidine) for catheter or port filling solution [121, 122]. In patients with intestinal failure on HPN, taurolidine-citrate-heparin catheter lock showed clinically significant and cost-effective reductions in catheter-associated bloodstream infections in high-risk groups compared to heparin [123]. There is evidence of the preventive efficacy of CLBSI in patients receiving long-term home ethanol lock (EL) PN. In 87 patients receiving 5–7 times per week home PN through a Hickman tunnelling silicone catheter using EL by daily injection of 2 mL of 70 % ethanol into the catheter lumen after flushing it with saline at the end of parenteral administration of nutrient solutions, the incidence of CLBSI was retrospectively studied over a 14-month period. The total follow-up period was 13,386 days of catheterization. Patients were compared with a clinically similar group of patients (n = 22) receiving home PN under the supervision of the same institution who had heparin lock prior to implementation of the ethanol protocol. Only 5*

of 87 patients (5.7 %) with the EL protocol CLBSI was diagnosed (0.45/1,000 catheter-days) during follow-up. In the control group, one or more CLBSI episodes (8.7/1,000 catheter-days) were diagnosed significantly more frequently (10 cases, 8.7 %, $p < 0.001$) during the follow-up period. According to the results of this study, it is concluded that the incidence of CLBSI when using the EL was 19 times less frequent [124]. A meta-analysis on the effect of catheter EL showed its effectiveness in reducing CLBSI in patients with tunneled central venous catheters on hemodialysis [125]. If intraluminal thrombus is suspected, an ultrasonography should be performed to rule out ongoing thrombosis outside the catheter, which requires removal of the CVC. If the thrombus is parietally localized, it is recommended to flush the catheter more thoroughly before and after use. If the catheter lumen is completely obstructed by a thrombus, an attempt should be made to use a lock with heparin, or better yet, a lock with urokinase (5,000 U in 1.0 mL of saline). If possible, a pharmacopoeial preparation containing a taurolidine solution, citrate and urokinase in the amount corresponding to the internal volume of the catheter (port) can be used. If there are thrombomasses around the catheter without disruption of the CVC function, systemic heparinization can be performed in the absence of contraindications. Frequent thrombotic complications should alert in terms of the presence of thrombophilia in a patient [117].

Recommendation 26. Each patient with SBS and IF who is indicated and planned to receive parenteral nutrition at home and/or a caregiver should be trained in a special program, which includes care of the catheter, the steps of preparation of infusion solutions and a container with nutrient substrates, the use of an infusomat, as well as prevention, recognition and management of complications [126]. Levels of evidence — 3, grades of recommendation — B.

Comments. *HPN conducting a is a complex task. It is important to assess a patient's (caregiver's) cognitive and physical abilities before beginning a HPN training program. The rehabilitation potential of a patient, as well as home environment, is assessed. Training should only be provided by dedicated employees. A variety of training methods, including printed handouts, manuals, and videos, are used to prepare patients for HPN [127]. The training program should include catheter care, the basics of preventing and recognizing complications related to vascular access, fluid imbalances, hyperglycemia or hypoglycemia, the most common errors, container storage and handling, adding vitamins and micronutrients, and pump use and care. HPN training must take place in an inpatient setting before patient's discharge. In additional, no time restrictions should be set for training [128, 129]. Before discharge, a patient should be given detailed written instructions on the use of central venous access and the sequence of implementation of infusion therapy, as well as PN at home, after which he/she signs an informed consent. Since this category of patients may have various questions and problems in the course of infusion therapy and PN, they should be able to have continuous telephone and, if necessary, in-person support from a well-trained team. Patients with a connection to such a specialized team have been shown to have better outcomes and a lower incidence of sepsis [126, 130].*

Recommendation 27. The efficacy and safety of long-term home parenteral nutrition should be monitored as needed for specific indications, but at least once every 3 months [127]. Levelof evidence — 3, grade of recommendation — B.

Comments. *The purpose of clinical and laboratory monitoring of patients with SBS and IF receiving parenteral nutrition at home is to control the correctness and evaluate its effectiveness. After being instructed and learning how to implement PN, patients will be able to recognize the initial stages of potential complications, including infections, mechanical catheter problems, venous thrombosis, and metabolic disorders. Psychological monitoring is also important in connection with long-term home PN and its potentially adverse effect on the patient's mood. The evidence-based literature on monitoring in home PN is insignificant. An observational study examining the monitoring status of patients receiving home PN in 42 centers in Europe showed that all patients had the dynamics of somatometric and some laboratory parameters assessed at least once every 3 months, 88 % of centers recorded the status offluid balance, and 74 % received information on oral intake [127]. For laboratory monitoring of stable patients, the following tests are recommended every 3 months: complete blood count, routine urine analysis, liver function tests (ALT, AST, bilirubin, alkaline phosphatase and gamma GTP when indicated), total protein, albumin, urea, creatinine, triglycerides,* *glucose, and sodium, potassium, calcium, magnesium, phosphate and iron. Determination of microelements, vitamins A, D, B_{12} and folic acid should be carried out at intervals of 12 months. Annual measurement of bone mineral density is also recommended [8, 28, 127].*

Recommendation 28. For successful implementation of home clinical (enteral and parenteral) nutrition, it is recommended to create a nutritional support team (NST), which prepares protocols, performs record keeping, training and follow-up of patients. Level of evidence — 3, grade of recommendation — B.

Comments. *The need for the introduction of HPN as an inpatient replacement technology is quite large and continues to increase steadily. In this regard, the creation of NST is important for the optimization, efficiency, and safety of treatment of patients receiving clinical, especially parenteral, nutrition at home. All patients needing this comprehensive treatment should have coordinated care from HNP specialists, who should provide both physical and psychological support to all patients who are discharged from the hospital and make the transition to home clinical nutrition. Team tasks should include patient record keeping, minimizing enteral and parenteral nutrition complications by ensuring compliance with treatment protocols (especially catheter/port care), and monitoring complications, including, catheter-associated problems (CLBSI and central vein thrombosis) and metabolic complications such as liver and bone disease and micronutrient imbalances [127].*

3.2. Surgical Treatment of Short Bowel Syndrome

Autologous intestinal reconstructions (AIRs) currently occupy one of the leading positions in the scheme of non-transplantological treatment of patients with SBS and chronic intestinal failure. The main goal of surgical treatment of SBS is to restore enteral autonomy with cancellation of PN or to increase enteral tolerance to reduce dependence on intravenous support, which has undeniable clinical and economic value [131, 132].

Operations aimed at increasing the absorptive surface of the intestinal tube have proven effective, butare used in pediatrics much more frequently than in adult practice, being the prerogative of large multidisciplinary centers dealing with intestinal rehabilitation. The main types of enteroplasty used to achieve enteral autonomy in patients with SBS and IF are:

- Longitudinal Intestinal Lengthening and Tailoring (LILT — Bianchi operation);
- Serial Transverse Entero Plasty (STEP);
- spiral intestinal lengthening and tailoring (SILT).

The term “autologous intestinal reconstructions” is more correctly used not as a generalizing concept for different types of enteroplasty, but as its combination with one-stage amelioration of other surgical intestinal diseases and/or complications, leading in the aggregate to the creation of optimal conditions for the digestive system. For example, the combination of enteroplasty with restoration of intestinal tube integrity or closure of intestinal stomas with formation of interintestinal anastomoses increases the chances of restoration of enteral autonomy. The small number and heterogeneity of patients with SBS limits the conduct of randomized controlled studies, but a number of scientific papers have published statistically proven predictors of enteral autonomy recovery in adult patients after AIR [133, 134].

These include: anatomy of the reconstructed intestine (length and width of the reconstructed small bowel, condition of the large bowel), duration and composition of PN before surgery, bilirubin level as the main marker of liver disease progression associated with intestinal lesions [135]. If a patient with SBS and ID is treated comprehensively as part of an intestinal rehabilitation program, the probability of achieving enteral autonomy after AIR can be up to 83 % [136]. At the same time, in patients with ultra-short variants of SBS, the observed reduction of infusion days and PN volume after AIR is also a good result of treatment and is possible in 40 % of cases. A comparative analysis of patients with SBS and IF who underwent AIR and transplantation showed that long-term survival and quality of life were significantly higher among patients after intestinal reconstruction surgery than among patients who underwent various types of visceral transplants [137, 138]. It is necessary to emphasize the clinical, socioeconomic, and ethical advantages of reconstruction surgery in amelioration SBS and IF after bariatric surgery compared with long-term PN and transplantation [139, 140].

Recommendation 29. Autologous intestinal reconstructions for patients with SBS are recommended in clinics specializing in the treatment of this pathology. Level of evidence — 3, grade of recommendation — B.

Comments. *In order to obtain a positive result of surgical treatment, the technical aspects of AIR must be clearly worked out. Surgical complications of enteroplasty (intestinal suture failure, intestinal tube stenosis in the reconstruction* *site, reconstructed intestine ischemic damage, adhesions) require not only repeated surgical treatment, but may entail secondary reduction of the residual intestine length. It is an indisputable fact that the results of surgical treatment of patients with SBS (duration and quality of life, enteral autonomy restoration) are significantly higher with interdisciplinary care of patients within the intestinal rehabilitation program of one center [141–144].*

Recommendation 30. It is recommended to choose the type of enteroplasty individually in each specific case, depending primarily on the anatomical and functional characteristics of the residual intestinal segment. Levels of evidence — 3, grades of recommendation — B.

Comments. *The development of the AIR scheme requires a personalized approach that primarily takes into account the postresection anatomy. LILT requires dilation of the residual small bowel over 4 cm. SILT, a relatively recently introduced spiral enteroplasty technique, is suitable for lengthening a moderately dilated segment of the residual small bowel, but is safe when performed on a reconstructed bowel segment up to 10 cm. STEP can be performed throughout the residual intestine and on the intestine with varying degrees of dilatation [145–147].*

Recommendation 31. Surgical treatment is recommended for SBS-IF patients with no tendency to restore enteral autonomy for one and a half to two years after initial resection. Level of evidence — 3, grade of recommendation — C.

Comments. *Early reduction of parenteral nutrition and/or obtainment full enteral autonomy as a result of lengthening the small bowel and recreating acceptable gastrointestinal anatomy avoids a number of complications associated with long-term parenteral nutrition. First of all, we are talking about infectious and thrombotic catheter-associated complications, which are leading in patients with IF and are one of the main causes of lethal outcomes [141, 143, 148].*

Recommendation 32. Surgical treatment is recommended for SBS-IF patients with the development of life-threatening complications due to residual small bowel redilatations. Level of evidence — 3, grade of recommendation — B.

Comments. *Drug-resistant bacterial overgrowth syndrome and recurrent translocation of opportunistic intestinal microflora and their toxins into the systemic bloodstream are the causes of sepsis and one of the main factors aggravating the course of intestinal failure-associated liver disease (IFALD). Significant dilatation of the residual small bowel that supports these complications, regardless of its length, requires an AIR [148, 149].*

Recommendation 33. Patients with SBS-IF with continued dependence on parenteral nutrition after autologous intestinal reconstructions, but with the potential benefit of repeated enteroplasty, are recommended as candidates for subsequent staged surgical treatment. Levels of evidence — 3, grades of recommendation — C.

Comments. *Repeated lengthening of the intestinal tube is technically possible after any of the most common enteroplasty procedures (STEP and LILT). Repeated AIRs conducting in stable patients without complications with clear indications for transplantation (first of all, secondary biliary cirrhosis against IFALD) allows to achieve enteral autonomy in more than half of them [150, 151].*

Recommendation 34. It is recommended to clearly indicate the section and length of the remaining small bowel, the preserved section of the large bowel, and the presence or absence of ileocecal valve in the operation protocols of the initial and subsequent intestinal resections, as well as after each AIR. Level of evidence — 2, grade of recommendation — B.

Comments. *Initially, the basic information about the SBS variant is based on the surgical protocols, which should clearly indicate the cause of the initial and subsequent bowel resections, the sections and length of the resected bowel, and the sections and length of the residual bowel. It is recommended to measure the length of the residual small bowel with a tape placed along the antimesenteric border, starting from the Treitz ligament or in its absence in case of incomplete bowel turn from the duodeno-jejunal junction. When describing the residual large bowel, the presence or absence of the ileocecal angle and the preserved parts of the large bowel should be clearly indicated [138, 141, 152].*

3.3. Small Bowel Transplantation

Recommendation 35. The main indications for referral of patients for small bowel transplantation (SBT) are: irreversible intestinal failure complicated by the phenomena of rapidly progressing cholestatic liver disease, thrombosis of two or more central venous conduits used

for parenteral nutrition and recurrent catheterized bloodstream infection. Level of evidence — 2, grade of recommendation — B.

Comments. *Small bowel transplantation is possible in patients with SBS and severe IF. Currently, more than 1,200 such operations have been performed worldwide [138]. Unlike renal failure, where transplantation is preferable to long-term extracorporeal support, intestinal transplantation cannot yet be recommended as an alternative therapy for patients who stably maintain their homeostasis and nutritional status with intravenous therapy. This is due both to the good results in general on long-term parenteral nutrition and to the serious problems encountered in intestinal transplantation [112]. Isolated intestinal transplantation is performed in patients with SBS and severe IF in the absence of concomitant liver disease. Quality of life after SBT is considered to be higher or equal to that after long-term PN [153]. Hepatointestinal transplantation (HIT) is considered in recipients with irreversible IF and end-stage liver disease. A large database study showed that patients with PN-dependent liver disease who underwent a combined hepatic and intestinal transplantation had significantly worse outcomes than patients who underwent an isolated SBT. Infectious (especially bacterial) complications remain the main cause of death after SBT. Contraindications to SBT are: the presence of active infection, malignant tumor, multisystem organ failure, cerebral edema, HIV infection in the stage of active AIDS [154, 155].*

Recommendation 36. The small bowel transplantation is performed by a multidisciplinary team that includes a transplant physician, hepatologist/gastroenterologist, clinical pharmacologist, infectious disease specialist, cardiologist, nutritionist, psychologist, social worker and financial coordinator. Level of evidence — 2, grade of recommendation — B.

Comment. *The pre-transplantation assessment of the required scope of intervention is mandatory: isolated or multivisceral transplantation. The stages of pre-transplant assessment are given in Appendix G5 [156–158].*

4. Medical Rehabilitation of Patients with SBS and IF

Implementation of medical rehabilitation of patients with SBS and IF should be carried out at all stages of medical care: hospital stage — day hospital — outpatient observation. Rehabilitation measures should include an assessment of the patient's rehabilitation potential, followed by a program for the initial stage and the final goal of each stage of rehabilitation. Rehabilitation subsequent stages conducting, if the previous stage goal is not achieved, is inexpedient.

Patients with SBS and IF are a heterogeneous group, which requires a differentiated approach to their rehabilitation.

Group 1 — patients with residual segment of the small bowel amounting to 30–40 % (<200 cm) of its average length (500 cm). These patients may have transient (type 1) intestinal failure, requiring dietary restrictions and some pharmacological support to maintain enteral autonomy. Intravenous support is usually not required.

Group 2 (includes 2 categories of patients who will have temporary or lifelong intravenous dependence):

- patients with residual segment of the small bowel amounting to 10–20 % (50–100 cm). In this situation, there is almost always prolonged severe intestinal failure (type 2), requiring months (up to a year, sometimes more) of intravenous support (hydration + parenteral nutrition). Adaptive enteral autonomy can occur in 50 % of patients within 1 to 2 years of the postresection period;
- patients with residual segment of the small bowel amounting to 10 % (ultra-short intestine less than 50 cm). Type 3 intestinal failure develops, requiring lifelong intravenous support (hydration + parenteral nutrition).

Patient categories with low rehabilitative potential, which in the vast majority of cases will require intravenous support:

- presence of jejunostoma with the residual part of the intestine less than 100 cm;
- presence of an ileocolic anastomosis with right-sided hemicolectomy and residual small bowel less than 60 cm;
- presence of ileocolic anastomosis with residual segment of small bowel less than 35 cm even with preserved large bowel and ileocolic valve.

If the goal of rehabilitation is not achieved at any stage, the reasons for its failure must be analyzed:

- initially incorrectly assessed clinical and functional status of a patient;
- incorrectly assessed rehabilitation potential at the beginning of the stage;
- an inadequately designed program;

- low compliance in the implementation phase of the rehabilitation program.

The fundamental factors of rehabilitation of patients with SBS and IF are: individual assessment of the current clinical condition, the degree of "loss" of anatomical volume and function, as well as the expected timing of achieving the goal of each stage of rehabilitation.

Recommendation 37. For patients with intestinal failure, for optimal development of individual rehabilitation programs at all stages of treatment, it is advisable to formulate the degree of structural and functional dysfunction as early as possible. Level of evidence — 2, grade of recommendation — B.

Comments. A detailed clinical assessment of the patient at the initial stage of treatment should include data on the volume of previous surgery, condition and size of the residual small bowel, nutritional status at the time of surgery, as well as the main indicators characterizing the activity of gastrointestinal tract and other organs and body systems (FEH, acid-base balance, hemodynamics, respiratory parameters). Rehabilitation of patients with SBS and IF begins against the background of basic treatment and is supplemented by monitoring and correction of the patient's quality of life with the formation of aphysical activity program aimed at stabilizing body mass. The main goal of physical activity is to maintain muscle mass. Exercises are performed according to generally accepted methods using the definition of intervals of the maximum and minimum allowable heart rate during loads (physical exercises) for a particular patient [159]. When assessing the impact of long-term PN on quality of life, most studies use two popular methods — the Short Form 36 Health Survey Questionnaire (SF-36) and the Euro QoL Index [129]. These tools are well adapted for patients with chronic diseases, are universal methods of quality of life assessment, but are not specific for patients receiving PN for a long time. In all quality of life studies, a patient or a close family member answers the questionnaire, and since perceptions of health and suffering are subjective life values, assessments of the patient and family, especially over time, provide the most valuable information. The patient's quality of life at any stage of treatment and rehabilitation is an essential tool for assessing their adequacy [160].

Recommendation 38. All patients with SBS and IF, in whom during the hospital phase of their treatment there was a need to prescribe intravenous infusion therapy to support FEH and acid-base balance, as well as to connect additional PN, should, if possible, pass the sanatorium stage of rehabilitation, whose main objectives are personalized optimization (selection) of the patient's medical diet to achieve possible relative enteral autonomy and restore his physical activity.

Comments. In the immediate postresection period after extensive resection of the small bowel, a personalized selection of a water schedule and optimal therapeutic diet is necessary for most patients to reduce intestinal dyspepsia phenomena as well as dependence on intravenous infusion therapy and PN. At the end of the sanatorium stage, the further routing of these patients at the dispensary health stage of their further rehabilitation is determined.

5. Prevention and Follow-Up Medical Care, Medical Indications and Contraindications to the Use of Methods of Prevention

The main activities for the prevention of SBS-IF progression and the development of secondary complications are shown in Table 2.

Follow-up medical care of patients with SBS-IF is performed in accordance with Order No. 404н of the Ministry of Health of the Russian Federation of April 27, 2021, "On Approval of the Procedure for Preventive Medical Examination and Health Examination of Certain Groups of the Adult Population". In accordance with this order, patients must undergo preventive medical examinations and health examinations in the medical organization where they receive primary health care. A general practitioner or a general practice doctor (family physician) is responsible for organizing and conducting preventive medical examinations and health examinations. Based on the results of the examination, the patient's health group is established, which can be changed. Patients with SBS-IF belong to health group III-a, which requires specialized medical care. For examination and dynamic assessment of their health status, as well as determining the further need for infusion therapy and parenteral nutrition of patients at home, specialists from centers with nutritional support groups and providing specialized medical care to this category of patients should be involved. The frequency of medical examinations and the list of necessary investigations in the follow-up medical care of patients with SBS-IF are presented in Table 3.

Table 2. Short bowel syndrome and intestinal failure complications prevention

Complications	Preventive activities
Erosive-ulcerative lesions of gastroduodenal mucosa	• Compliance with prescribed dietary restrictions and a split dietary regimen. • Prescription of gastric secretion blockers during the first 6 months of the postresection period and thereafter on demand
Worsening phenomena of maldigestion and malabsorption	• Split sparing diet (5–6 times a day) in small portions, taking into account dietary restrictions. • Water intake separated from meals (30 min before or 45 min after meals). • Coprogram control. • Intraintestinal decontamination with prescription of non-absorbable antibiotics (rifaximin, nifuroxazide), which is most relevant in the presence of an ileocolic anastomosis bypassing the ileocolic valve. • Prescription of gastric secretion blockers. • Prescription of pancreatin in microgranules or microtablets
Protein-energy malnutrition	• Optimization of therapeutic dietary nutrition of patients, taking into account the actual possibility of digestion and assimilation of various foods and dishes (severity of the phenomena of maldigestion and malabsorption). • Regular dynamic assessment in early and late postoperative periods of somatometric, clinical and laboratory (hemoglobin, lymphocytes, total protein, albumin, urea, glucose, triglycerides, electrolytes, nitrogen losses, etc.) indicators reflecting the state of the patient's nutritional status. • In case of progressive BM reduction phenomena of 5 % or more per month, supplement the patients' nutritional therapy with balanced polymeric and, if poorly tolerated, oligomeric ENMs, taking into account their individual tolerance and sensory preferences. • In case of continuing BM reduction and inability to provide patients with proper substrate supply through the gastrointestinal tract, additional and, if necessary, complete parenteral nutrition should be prescribed. • As digestion processes stabilize and enteral autonomy expands, allowing to maintain FEH and substrate provision of the body with energy and protein not less than 75–80 % of the need, parenteral nutrition should be gradually reduced under control of the dynamics of the main indicators of the nutritional status of patients. • PN is canceled when sufficient enteral autonomy (including pharmacological support) is achieved, when an orally consumed nutritional therapy will stabilize patients' nutritional status and ensure good quality of life
Cholelithiasis	• Split sparing diet (5–6 times a day) in small portions. • Course administration of preparations of ursodeoxycholic acid, taking into account individual tolerance and contraindications
Nephrolithiasis and oxalic nephropathy	• Water intake of 30 mL/kg body mass, taking into account intestinal tolerance (the amount of fluid consumed during the day should not lead to increased frequency of stools). If there are clinical signs of hypovolemia (thirst, dry mucosa, decreased skin turgor, hypotension, tachycardia, diuresis less than 1,000 mL/day, decreased central venous pressure) — intravenous infusion correction of FEH. • Prescription of calcium carbonate 1 g before each meal (5–6 g per day)
D-Lactic acidosis	• Simple carbohydrates consumption restrictions. • Intraintestinal decontamination by prescribing non-absorbable antibiotics (rifaximin, nifuroxazide)
Anemia	• Administration of foods high in heme-bound iron and vitamin B_{12} into the diet. • Monitoring of hemoglobin, erythrocytes, iron and vitamin B_{12} content in the blood, TIBC and transferrin. • Pharmacological correction of iron deficiency. • Periodic intramuscular injections of vitamin B_{12} and folic acid oral administration
Osteoporosis	• Periodic monitoring of vitamin D levels (target level of at least 30 ng/mL). • A course of vitamin D and calcium preparations

6. Health Care Delivery Organization

When providing medical care to patients with SBS-IF and determining their subsequent routing at the dispensary health stage of their treatment, 2 groups should be distinguished:

1. Patients with intestinal failure type 1 with relatively mild postresection course of the disease and positively predicted morphofunctional adaptation of the residual small bowel segment with achievement of optimal enteral autonomy for 6–12 months.

2. Patients with postresection intestinal failure types 2 and 3 who have long-term (more than 12 months, sometimes lifelong) dependence on intravenous hydration and nutritional support.

The first group includes patients with residual segment of the small bowel amounting to 30–40 % (200 cm) of its average length (500 cm). These patients may have transient intestinal failure, requiring certain dietary restrictions, additional oral administration of balanced ENMs and some pharmacological correction of impaired digestive processes, allowing to achieve proper enteral autonomy during the first year. Wherein intravenous support is not required.

The second group includes 2 categories of patients who will have temporary or lifelong intravenous dependence:

A. Patients with residual segment of the small bowel amounting to 10–20 % (50–100 cm). In this situation, there is almost always prolonged severe intestinal failure (type 2), requiring months (up to a year, sometimes more) of intravenous support (hydration + parenteral nutrition). Adaptive enteral autonomy can occur in 50–60 % of them within 1 to 2 years of the postresection period.

B. Patients with residual segment of the small bowel, which usually develops type 3 intestinal failure, requiring most often lifelong intravenous support (hydration + parenteral nutrition). The most frequently such a need arises:

- in the presence of jejunostoma with the residual part of the intestine less than 100 cm;
- in the presence of an ileocolic anastomosis with right-sided hemicolectomy and residual small bowel less than 60 cm;
- in the presence of ileocolic anastomosis with residual segment of small bowel less than 35 cm even with preserved large bowel and ileocolic valve.

Patients with enteral autonomy (group 1) may be followed up by a general practice doctor (family physician, general practitioner) or an outpatient gastroenterologist (where available). If necessary, they are referred for consultation to a gastroenterologist, nutritionist, surgeon.

Patients requiring parenteral nutrition and intravenous hydration must be supervised not only by the general practice doctor, but also by specialists specially trained in enteral and parenteral nutrition (Home Clinical Nutrition Center).

In accordance with Order No. 543н of the Ministry of Health of the Russian Federation on May 15, 2012 “On Approval of the Provision of Primary Health Care to the Adult Population”, group 2 patients can receive infusion therapy and parenteral nutrition as part of a “home care”. Such patients should be cared for by specialists trained in enteral and parenteral nutrition.

Table 3. The frequency of medical examinations and the list of necessary investigations in the follow-up medical care of patients with SBS-IF

Frequency of examinations	Obligatory studies list*
Monthly for the first 3 months after hospital discharge and then at least once a quarter	• Physical examination with assessment of subjective symptoms • Anthropometry (body mass, BMI) • Full blood count • Clinical urine analysis • Coprogram • Blood chemistry (ALT, AST, bilirubin, urea, total protein, albumin, glucose, cholesterol, triglycerides, potassium, sodium, calcium, magnesium, iron, phosphates) • Vitamins B_{12} and D — once every 6 months • ECG • Densitometry — once a year

Notes: other tests are conducted in accordance with the requirements of Order No. 404 of the Ministry of Health of the Russian Federation of April 27, 2021;
discharge of patients with SBS-IF from the dispensary registration is possible not earlier than after 12 months and only when full enteral autonomy is achieved.

7. Health Care Delivery Quality Assessment Criteria

No.	Quality criteria	Level of evidence	Grades of recommendation
1	Assessment of nutritional status indicators in the early postoperative period in patients with SBS and IF: total protein, albumin, blood lymphocytes, body mass and BMI dynamics	1	A
2	Early enteral nutrition in the absence of contraindications	1	A
3	Parenteral nutrition in the presence of contraindications to enteral nutrition or inability to properly implement enteral nutrition	2	B
4	Dynamic assessment of the main indicators of nutritional status at least once a week during the first month of the postoperative period: body mass, BMI, hemoglobin, lymphocytes, total protein, albumin, blood lymphocytes	2	B
5	Infusion therapy and parenteral nutrition (if possible, at home) in patients with SBS and IF with low efficiency of oral dietary nutrition with additional intake of enteral nutritional mixtures by sipping	2	B
6	Regular monitoring of somatometric (body mass, BMI) and laboratory (full blood count, ALT, AST, bilirubin, urea, total protein, albumin, electrolytes) indicators reflecting the dynamics of nutritional status at least once every 3 months in patients receiving HPN	2	B

Note. The criteria apply at all three levels of care.

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Annex A3. Linked documents

Current clinical recommendations are prepared in accordance with the following legal documents:

1) Federal Law No. 323-FZ of 21 November 2011 “On the bases of health protection in the Russian Federation”;

2) Procedure of healthcare provision by the Order of the Ministry of Health of the Russian Federation No. 919n of 15.11.2012 “On approval of the Procedure of healthcare provision to adults in specialty “anaesthesiology and resuscitation””;

3) Order of the Ministry of Health and Social Development of the Russian Federation No. 543n of 15 May 2012 “On approval of the Regulation on primary healthcare provision to adults”;

4) Order of the Ministry of Health of the Russian Federation No. 520n of 15 July 2016 “On approval of criteria for medical aid quality assessment”;

5) Order of the Ministry of Health of the Russian Federation No. 279n of 05 May 2016 (revised 21.02.2020) “On approval of the Procedure for organising therapeutic resort treatment”;

6) Order of the Ministry of Health of the Russian Federation No. 395n of 21 June 2013 “On approval of norms for therapeutic nutrition”;

7) Order of the Ministry of Health of the Russian Federation No. 796n of 02 December 2014 “On approval of the Regulation on providing specialty healthcare, including high-technology aid”;

8) Order of the Ministry of Health and Ministry of Labour and Social Protection of the Russian Federation No. 345n/372n of 31 May 2019 “On approval of the Regulation on palliative care provision, including the interaction procedure between medical, social service institutions, public associations and other non-profit organisations involved in healthcare”;

9) Order of the Ministry of Health of the Russian Federation No. 348n of 31 May 2019 “On approval of the list of home-use medical appliances to sustain human organ and systemic functions”;

10) Order of the Ministry of Health of the Russian Federation No. 1008n of 23 September 2020 "On approval of the Procedure for patient provision with therapeutic nutrition';

11) Order of the Ministry of Health of the Russian Federation No. 559n of 09 June 2020 "On approval of the Procedure for population provision with medical aid in specialty "surgery (combustiology)"";

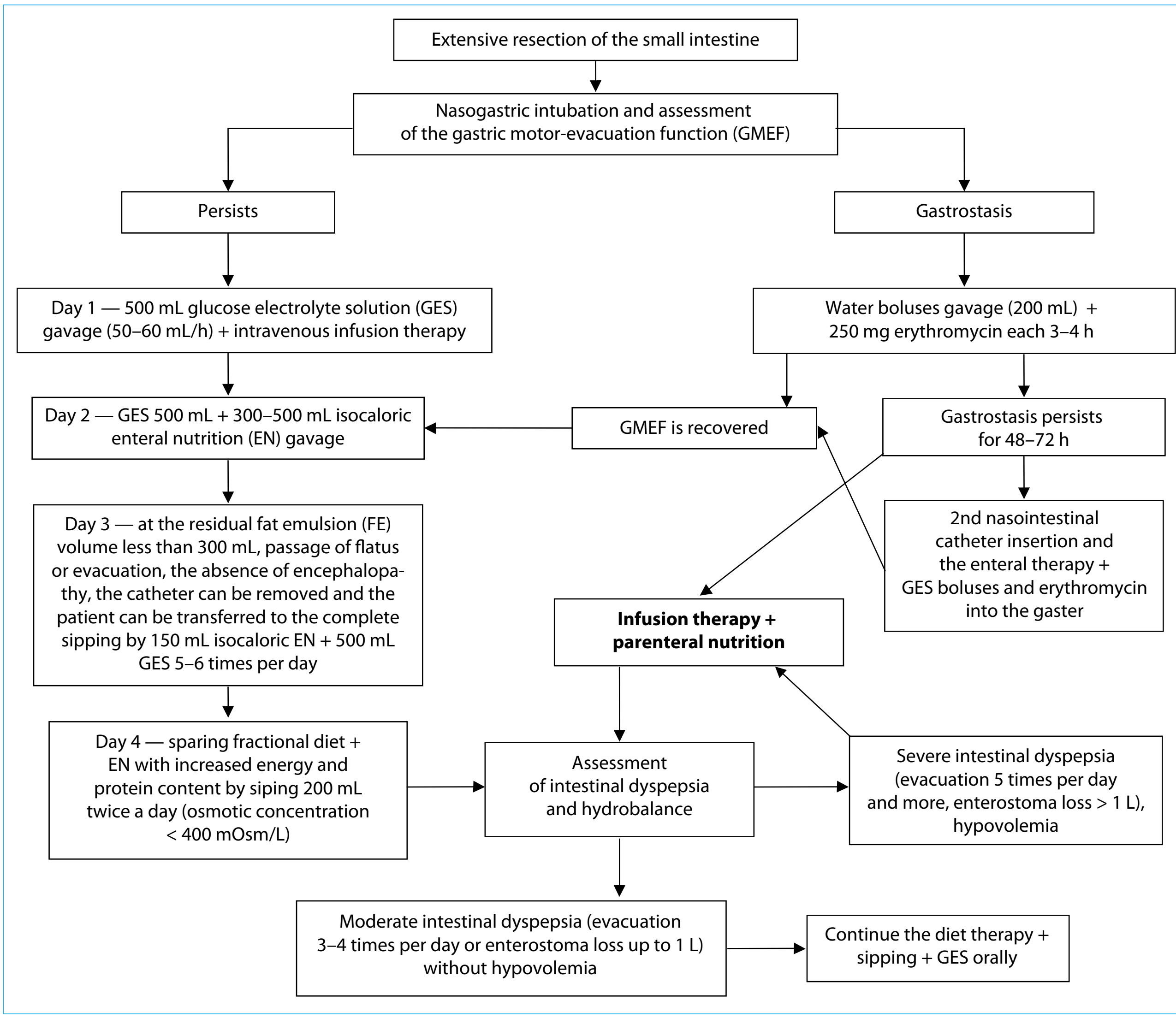
12) Order of the Ministry of Health of the Russian Federation No. 788n of 31 July 2020 "On approval of the Procedure for organising medical rehabilitation in adults";

13) Order of the Ministry of Health of the Russian Federation No. 1008n of 23 September 2020 "On approval of the Procedure for patient provision with therapeutic nutrition";

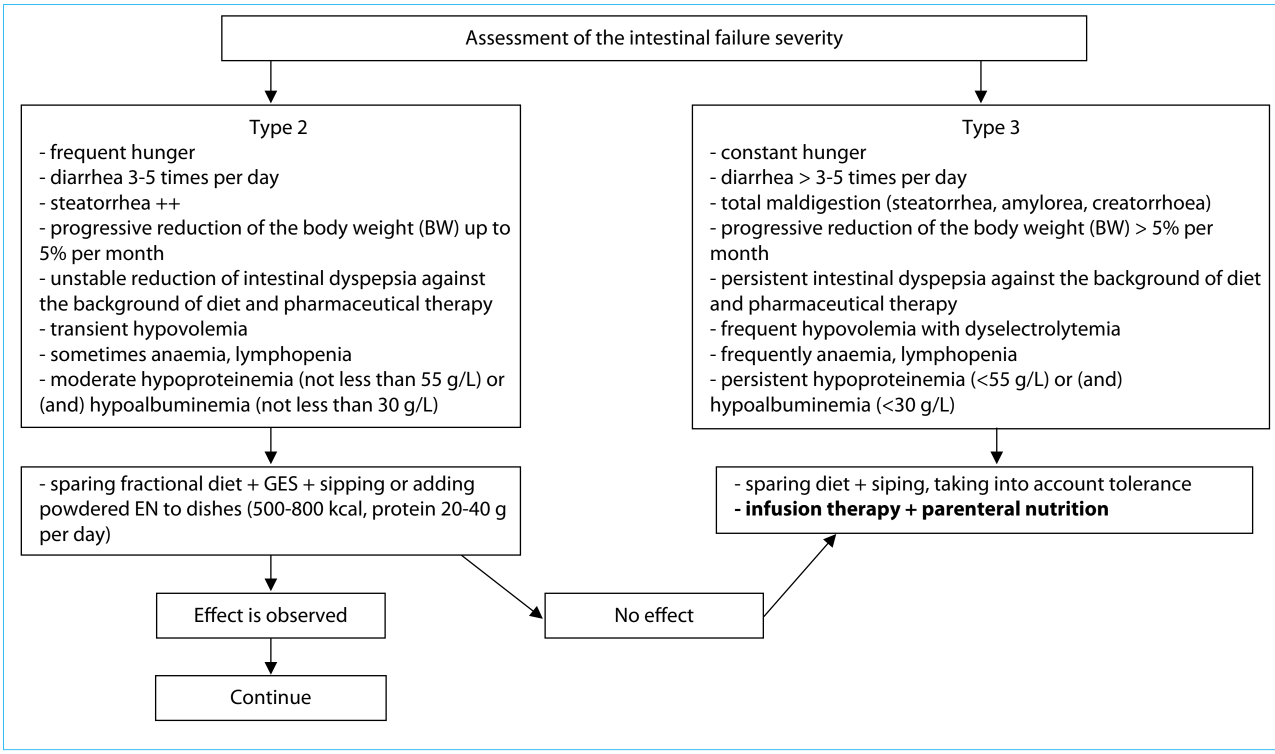
14) Order of the Ministry of Health of the Russian Federation No. 404n of 27 April 2021 "On approval of the Procedure for conducting preventive medical and clinical examinations in selected adult categories".

Annex B. Patient management algorithms

Algorithm 1. Nutritional support procedure in immediate and early post-resection periods



Algorithm 2. Nutritional support procedure in late post-resection period



Notes:
• post-resection conditions likely requiring long-term (possibly lifelong) home parenteral nutrition — jejunal tube in bowel length <100 cm, anastomosis between jejunum (<60 cm length) and transverse colon, jejunoileal anastomosis (<35 cm total length) with preserved ileocecal valve and colon;
• final decision on further needs and volume of parenteral nutrition, as well as infusion therapy, is made in 2 years.

Annex C. Catheter use and care, nutrient mixture infusion procedure

Dressing change

1. Consumables:

A. Sterile self-adhesive dressings:

• Cosmopor I.V. (6×8 cm) — dressing change frequency: once a day. Similar products: Cosmopor Antibacterial (7.2×5 cm, 10×8 cm), Cosmopor ESteril (7.2×5 cm);

• Tegaderm™ I.V. (7×8.5 cm, item 1633). Dressing change frequency: once every 5 days;

• 3M™ Tegaderm CHG chlorhexidine antimicrobial dressing for catheter fixation (item 1657R or 1658R).

Dressing change frequency: once every 7–10 days.

B. TauroLock™-HEP500 antimicrobial catheter lock solution (10 ampoules, 5 mL) or 70° alcohol.

2. Prepare a dressing kit (preferably on a separate table or tray): gloves, anatomical forceps or Billroth clamp, gauze balls or tissue (2–3 pieces), antiseptic (70° alcohol or any skin antiseptic liquid like Akhdez), any sterile self-adhesive dressing available (Tegaderm, Cosmopor).

3. Wear surgical gloves (non-sterile gloves to be thoroughly treated with an antiseptic like alcohol, Akhdez, etc.).

4. Carefully remove the self-adhesive dressing from the side to catheter exit point (even minor catheter disturbance is undesirable).

5. Re-sterilise gloves.

6. Using an antiseptic-soaked gauze ball (or tissue; 70° alcohol, Akhdez, etc.), use forceps (or clamp) to slowly work out the catheter exit site, catheter itself (3–4 cm) and surrounding skin by gentle rotating and sponging stirs to disinfect and degrease (within the area of intended self-adhesive dressing).

7. Unpack the self-adhesive dressing.

8. Treat the gloves with an antiseptic.

9. Apply the self-adhesive dressing so that the catheter exit point from skin is completely covered by the dressing.

Nutrient mixture intravenous infusion procedure (three-in-one container)

1. Wash, dry and treat your hands thoroughly.
2. Unpack the container.
3. Stir the container cells content by tearing the inter-bridges.
4. Wear surgical gloves (non-sterile gloves to be thoroughly treated with an antiseptic like 70° alcohol, Akhdez, etc.).
5. Administer the necessary medication (Addamel, Vitalipid, Soluvit) via the container inlet port (red).
6. Stir the container several times.
7. Hang the container on a rack.
8. Unpack the intravenous infusion kit (dropper).
9. Attach the intravenous infusion kit (dropper) to the container outlet port (white) and fill it with nutrient mixture (avoiding air bubbles, until a mixture drop forms under in the dropper cap).
10. Mount the kit silicone piece into infusion pump and enter the intended infusion parameters (volume and infusion rate).
11. Treat surgical gloves with an antiseptic (70° alcohol, Akhdez, etc.).
12. Using an antiseptic-soaked gauze ball (or tissue; 70° alcohol, Akhdez, etc.), carefully (for 1–1.5 min) wipe (clean) the catheter cover plug.
13. Remove the cap into a prepared antiseptic container (70° alcohol, Akhdez, etc.).
14. Using an antiseptic-soaked gauze ball (or tissue; 70° alcohol, Akhdez, etc.), carefully clean (wipe) the uncapped end of catheter connector.
15. Attach a syringe to a 10 mL 0.9% sodium chloride catheter.
16. Open the catheter clamp, flush catheter and close the clamp.
17. Dismount the kit (dropper) cap and store it in the same antiseptic container (if the nutrient mixture container is used for 2 days, a sterile cap will be re-used to plug the dropper upon the infusion end).
18. Attach the kit (dropper) cannula tightly to the catheter connector, open the roller and catheter clamp, switch on the infusion pump and commence infusion.

End of infusion

1. Fill a 10 or 20 mL sterile syringe with 0.9% sodium chloride. Take 2.5 mL TauroLock™-HEP500 or 70° alcohol into a 2.5 or 5 mL syringe.
2. Switch off the infusion pump and close the catheter clamp.
3. Wear surgical gloves (non-sterile gloves to be thoroughly treated with an antiseptic like 70° alcohol, Akhdez, etc.).
4. Using an antiseptic-soaked gauze ball (or tissue; 70° alcohol, Akhdez, etc.), carefully (for 1–1.5 min) wipe (clean) the kit-catheter junction.
5. Dismount the kit (dropper) cannula from the catheter connector and attach a 0.9% sodium chloride syringe.
6. Open the catheter clamp and flush it impulsively with 10–20 mL 0.9% sodium chloride (do not use heparin).
7. Close the catheter clamp and disconnect syringe.
8. Attach a TauroLock™-HEP500 or 70° alcohol syringe.
9. Open the catheter clamp.
10. Inject 2.5 mL TauroLock™-HEP500 or 70° alcohol.
11. Close the catheter clamp.
12. Screw a new sterile (or stored in antiseptic) plug (injection cap) tightly onto the catheter connector.
13. If the nutrient mixture is left in the container for a later infusion, use an antiseptic-soaked gauze ball (or tissue; 70° alcohol, Akhdez, etc.) to thoroughly clean (wipe) the kit (dropper) cannula.
14. Screw a new sterile (or stored in antiseptic) plug tightly onto the kit (dropper) cannula.

Notes:

- The catheter should be flushed with 10–20 mL 0.9% sodium chloride (do not use heparin) daily (if not used) and after each infusion.
- After a nutrient mixture infusion, blood sampling and drugs administration, the catheter must always be flushed impulsively with 10–20 mL 0.9% sodium chloride (do not use heparin), followed by adding 2.5 mL TauroLock™-HEP500 antimicrobial lock solution or 70° alcohol.

Annex D. Assessment scales, questionnaires and other patient scoring tools provided in clinical recommendations

Annex D1. Assessment of malnutrition severity in SBS patients

No.	Indicators	Reference 3 points	Malnutrition		
			light	moderate	heavy
			2 points	1 point	0 points
1	RBM from BMI deviation, %	100–90	90–80	80–70	<70
2	BMI, kg/m^2				
	18–25 y.o.	23.0–18.5	18.5–17.0	16.9–15	<15
	>25 y.o.	26.0–19.0	19.0–17.5	17.4–15.5	<15.5
	>60 y.o.	27.0–21.0	21.0–19.0	18.9–16	<16
3	Shoulder girth, cm				
	men	29–26	26–23	23–20	<20
	women	28–25	25–22.5	22.5–19.5	<19.5
4	TSFT, mm				
	men	10.5–9.5	9.5–8.4	8.4–7.4	<7.4
	women	14.5–13.0	13.0–11.6	11.6–10.1	<10.1
5	Shoulder muscle girth, cm				
	men	25.7–23.0	23.0–20.5	20.5–18.0	<18
	women	23.5–21.0	21.0–18.8	18.8–16.5	<16.5
6	Total protein, g/L	≥65	65–55	55–45	<45
7	Albumin, g/L	≥35	35–30	30–25	<25
8	Transferrin, g/L	≥2.0	2.0–1.8	1.8–1.5	<1.5
9	Lymphocytes, ths.	≥1.2	1.2–1.0	1.0–0.8	<0.8
	Total score	27	27–18	18–9	<9

Annex D2. Water and electrolyte dosage determination for total parenteral nutrition in SBS patients

Elements	Per kg body weight / day	Average daily dosage
Water*	20–30 mL	1000–2000 mL
Sodium	1–1.5 mM	60–150 mM
Potassium	1–1.5 mM	40–100 mM
Chlorides	1–1.5 mM	40–100 mM
Phosphates	0.3–0.5 mM	10–30 mol
Magnesium	0.1–0.15 mM	4–12 mM
Calcium	0.1–0.15 mM	2.5–7.5 mM

Note: * allowing for macronutrient oxidation-produced metabolic water: proteins — 41 mL/100 g, lipids — 107 mL/100 g, carbohydrates — 55 mL/100 g.

Annex D3. Micronutrient dosage determination for parenteral nutrition in SBS-IF patients

Microelements	Dosage, mM
Zinc	38–100
Cuprum	8–24
Selenium	0.4–0.9
Ferrum	18–20
Manganese	3–5
Chromium	0.2–0.3

Microelements	Dosage, mM
Molybdenum	0.2–0.3
Iodine	0.01–1.0
Fluorine	50–79
Vitamins	
A, mg	50–79
E, mg	1000 mg
K, mg	10
D, mg	150
B_1, mg	5
B_2, mg	3–3.5
B_6, mg	3.6–4.9
Niacin (B_3), mg	4–4.5
Folic acid (B_9), mg	40–46
B_{12}, ug	5–6
Biotin (B_7), mg	60–69
C, mg	100–125

Annex D4. Parameters and frequency of clinical and laboratory monitoring in home clinical nutrition patients

Control parameters	Stable condition, year 1	Stable condition, year 2 and onwards
General examination (skin turgor, swelling, dry mucous membranes, etc.)	Once a month	Once a quarter
Stool properties and frequency	Upon prescription	Upon prescription
Water balance	Once a week	Upon prescription
Oral nitrogen and energy intake	Upon prescription	Upon prescription
Body weight	Twice a week	Once a week
Shoulder girth	Once a month	Once a quarter
Shoulder muscle girth	Once a month	Once a quarter
TSFT	Once a month	Once a quarter
Laboratory values:		
General blood panel	Once a month for first 3 months and onwards upon prescription, but at least once a quarter	Once a quarter
Clinical urinalysis		
Acid-alkaline state	Upon prescription	Once a quarter
Glucose	Upon prescription	Upon prescription
Urea	1–2 times a week	Once a month
Creatinine	Once a month for first 3 months and onwards upon prescription, but at least once a quarter	Once a quarter
Potassium, sodium, chlorides		-//-
Magnesium, calcium, phosphates		-//-
Total protein	-//-	-//-
Albumin	-//-	-//-
ALT, AST, bilirubin	-//-	-//-
Triglycerides	-//-	-//-
Urine biochemistry:		
Urea	Upon prescription	Upon prescription
Creatinine	Upon prescription	Upon prescription
Estimated values:		
Nitrogen balance	Upon prescription	Upon prescription
Creatinine-growth index	Upon prescription	Upon prescription

Annex D5. Patient assessment prior to intestinal transplantation

Medical records evidence	Analysis of medical and surgical history, therapy received, current enteral and parenteral nutrition
Anamnesis and check-up	Thorough physical and history examination
Laboratory tests	Blood type
	Tissue typing (HLA)
	Pre-existing antibodies (PRA)
	Serology (CMV IgGandIgM, EBV IgGandIgM, HIV, HCV, HBeAg, HBsAg, HBsAb)
	General blood and biochemical panels, inflammation factors
Imaging examination	Chest X-ray
	Doppler liver scan
	Upper and lower limb veins ultrasound
	Abdominal and pelvic CT
	Stomach and bowel examinations
	Barium X-ray
Endoscopy	OGDS
	Colonoscopy
Intestinal passage examination	Oesophageal-gastric
	Small-intestinal
	Colonic
Liver	Liver biopsy
Cardiac assessment	ECG
	Echocardiography
	Stress test and/or cardiac catheterisation if patient is >50 y.o., risk factors (HTN, DM)
Nephrological assessment	Renal ultrasound
	24-h creatinine clearance
Additional check-ups	Neurologist
	Infectionist
	Anaesthetist and resuscitator
	Nutritionist
	Pulmonologist
Clinical examination	Dentist
	Mammography
	Cervical oncology smear (Papanicolaou)
	Vaccination
	Hepatitis A
	Hepatitis B
	Pulmonary category
	SARS-nCoV-2
Inter-specialty assessment	Transplant surgeon, surgeon, gastroenterologist, nutritionist

Annex D6. Venous access comparison for parenteral nutrition in outpatient settings

Access type	Expected indwelling time	Usage	Potential complications
Peripherally inserted central venous catheter (PICC)	Maximum length of stay in vein up to 12 months	Applicable in acute treatment as well as short- and medium-term PN in children and adults	Associated with a higher risk of deep vein thrombosis. Antecubital puncture may hamper self-care and activity. Easily removable in suspected infection or if PN discontinued
Tunnelled central venous catheter	6 months — several years	Applicable in long-term and frequent PN; cuff inhibits microbial migration and reduces the catheter displacement risk	Upper limb activity unrestricted Position on chest facilitates catheter care Applicable for self-care
Implantable port system	6 months — several years	Applicable for recurrent PN; low CRBSI risk	Applicable in selected PN settings Motivated patients can be trained in access care PN may provoke CRBSI and occlusions in oncology children

Appendix D7. Medical and social contraindications for parenteral nutrition in outpatient settings

- Terminal incurable disease
- Indications for inpatient treatment
- Patient or family members inability to train in parenteral nutrition techniques
- Inadequate housing and social conditions

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