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CHRONIC CONSTIPATION AND DEFECATION DISORDERS IN A NEUROLOGICAL PATIENT WITH LIMITED MOBILITY

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Abstract. Chronic constipation in neurological patients with limited mobility is a common problem among children and adults. About 98% of patients with limited mobility with cerebral palsy in the Russian Federation have constipation, and there are no data on its prevalence among children with other neurological pathologies in our country. The versatility of causes, ranging from impaired nerve conduction at different levels, ending with the psychological state of the patient and his family, only proves the need for a more attentive attitude to this problem and to find ways to solve it. This literature review presents the results of modern studies of compatriots and foreign colleagues on the problem of chronic constipation and defecation disorders among patients with limited mobility.

Key words: chronic constipation; limited mobility patient; cerebral palsy; Spina Bifida.

ХРОНИЧЕСКИЕ ЗАПОРЫ У МАЛОМОБИЛЬНОГО ПАЦИЕНТА НЕВРОЛОГИЧЕСКОГО ПРОФИЛЯ

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Резюме. Хронические запоры у маломобильных пациентов неврологического профиля являются часто встречающейся проблемой среди детского и взрослого населения. Около 98% маломобильных пациентов с детским церебральным параличом (ДЦП) в Российской Федерации имеют запор, а данные о его распространенности среди детей с иной неврологической патологией на территории нашей страны отсутствуют. Многогранность причин, начиная от нарушения нервной проводимости на разных уровнях, заканчивая психологическим состоянием пациента и его семьи, только доказывает необходимость в более внимательном отношении к данной проблеме и в поиске путей к ее решению. В литературном обзоре представлены результаты современных исследований соотечественников и зарубежных коллег проблемы хронических запоров и нарушения дефекации среди маломобильных пациентов.

Ключевые слова: хронический запор; маломобильный пациент; ДЦП; Spina Bifida.

Constipation is a disorder of intestinal function, which is manifested by a shortening (compared to the individual physiological norm) of the age-related rhythm of the defecation act, its difficulty, systematically insufficient emptying of the intestine and/or changes in the shape and nature of stool [1, 2–4]. Constipation affects about 12% of the world's population, with people in the Americas and Southeast Asia being twice as likely to be constipated as Europeans (17.3 and 8.75%, respectively) [1]. Studies published over the last decade have reported constipation rates in children ranging from 10 to 25%, with only sporadic data on the prevalence of chronic constipation (CC) in low-mobility patients (LMP), especially in children, with a high proportion of functional constipation [5, 6]. Patients with severe chronic diseases in the decompensation stage, neurological pathology, diseases of the muscular system, oncological diseases, morbid obesity, as well as with various traumas and terminal illnesses are usually considered to be LMP. It has been found that 98% of LMP with cerebral palsy have CC, and its incidence among oncological patients ranges from 32 to 87% and increases to 90% in the case of opioid use [7]. During normal evacuation the intestinal contents stretch the ampulla of the rectum and irritate baro- and mechanoreceptors. Through afferent pathways, the signal from them reaches the centre of involuntary defecation, which is located in the lumbosacral spinal cord. Under the influence of descending signals there is a relaxation of the internal sphincter of the anus. In turn, the arbitrary act of defecation occurs with the participation of the cortex: under the influence of efferent impulses, the external sphincter of the anus and the puborectal muscle relax [8]. The enteric nervous system regulates rhythmic contractions of the rectum, facilitating faecal movement [9]. Adequate control of the muscles of the anterior abdominal wall and pelvic diaphragm also contributes to proper and timely defecation by increasing intra-abdominal pressure. Disruption of nerve impulse transmission at each level can lead to disorders of the act of defecation (neurogenic bowel dysfunction — NBD) and CC. Chronic constipation and faecal incontinence often coexist; sometimes there is "overflow" diarrhea (when solid stools accumulated above the rectum allow only watery stools to pass by, resulting in incontinence of liquid faeces)

[10]. The term NBD implies autonomic and/or somatic denervation of the bowel. The problem of NBD in the paediatric literature is not sufficiently considered, and there is no standardised approach to therapy in national guidelines.

Causes of defecation disorders are tumours or organic lesions of the brain, pathology at the level of the spinal tract, infiltration of sacral nerves, disorder of autonomic innervation of the colon. The causes and manifestation of NBD in children and adolescents differ from adult forms. In most cases, paediatric NBD is caused by congenital problems such as cleft spine and cerebral palsy. Acquired forms caused by trauma, infection and other causes are similar to the clinical presentation in adult patients [11].

Cerebral palsy is defined as a congenital neurological condition due to non-progressive trauma (usually presumed post-hypoxic) or brain malformation occurring in the foetal or perinatal period [12]. The incidence of cerebral palsy is about 1,5 per 1000 births, making it the most common neurological condition encountered in paediatrics. Cerebral palsy encompasses a group of disorders of varying degrees of movement and postural development. Up to 90% of children with cerebral palsy suffer from constipation and 47% from faecal incontinence, although the majority suffer to a minor degree [13]. In patients with cerebral palsy, due to motor pathway damage at different levels, muscle hypertonicity occurs, limiting the development of motor function, which in some cases makes it difficult and almost impossible to control the abdominal muscles and pelvic diaphragm. There is evidence of lesions of the mesenteric and submucosal nerve plexuses as coordinators of intestinal peristalsis, controlled by the central nervous system (CNS), which is also a risk factor for the development of constipation in patients with cerebral palsy [14]. There is a direct correlation between the degree of motor skills development, according to the GMFCS classification, and the frequency of constipation in patients [15, 16]. About half of people with cerebral palsy are intellectually disabled [17, 18], which affects what treatments of NBD and CC can be used. In multiple sclerosis, lesions in the spinal cord and hypothalamic region of the brain will cause problems similar to reflex bowel emptying after spinal cord injury (Table 1) [19].

Another illustrative example of nerve conduction disturbance as a cause of NBD and CC development is the presence of a spinal hernia of various sizes in a child with *Spina Bifida*. A meningocele, meningoradiculocoele, myelomeningocele, and myelocystocele are distinguished depending on the contents of the hernial sac. Myelomeningocele (MMC) is the most common and myelocystocele is the most rare [20, 21]. The vast majority of MMC cases involve the lumbar spi-

nal cord and sacral roots that innervate the bladder, distal colon and their respective sphincters, so some degree of neurogenic bladder and bowel dysfunction is almost universal in this population [22]. The frequency of urethrovesical dysfunction in myelomeningocele is not completely known, but most studies suggest that it is very high [23]. Similarly, anorectal dysfunction is also common. In contrast, in meningocele, the dura mater protrudes through the spinal canal defect, but the nerve elements remain confined within the canal and therefore are usually not damaged either antenatally or postnatally. In *Spina Bifida occulta* the bony lesions are not open, so in most cases there are no obvious signs of neurological damage [24]. Bowel dysfunctions such as constipation and faecal incontinence have a significant impact on the quality of life and well-being of individuals with spina bifida, as well as their parents. The *Spina Bifida Association* in 2019 surveyed adult patients, parents and children with the condition to assess the extent to which bowel dysfunctions such as constipation and faecal incontinence affect quality of life; half of the parents surveyed rated it as the biggest problem [25–29].

Muscular dystrophies and mitochondrial disorders are also accompanied by symptoms of bladder and bowel dysfunction. Constipation in X-linked Duchenne muscular dystrophy can be life-threatening, but fecal incontinence is usually the most disabling [30, 31]. The cause of constipation in these patients is functional anorectal obstruction [30], altered gastrointestinal transport and possible sensory impairment due to expression of the DP116 dystrophin isoform in peripheral nervous tissue and autosomal DP116 homologues in sensory ganglia [30], as well as decreased myoelectric slow-wave activity along with decreased nitric oxide (NO) availability due to the absence of dystrophin acting as an anchor for NO synthase [30]. Loss of alpha-dystroglycan-laminin interaction due to defective glycosylation of alpha-dystroglycan underlies a group of congenital muscular dystrophies often associated with brain malformations called dystroglycanopathies [31]. Mitochondrial neurogastrointestinal encephalomyopathy is often associated with chronic intestinal obstruction. The pathophysiology leading to impaired peristalsis and movement of intestinal contents is related to impaired neuromuscular coordination due to the myopathy (affecting intestinal contraction), neuropathy (affecting coordination of intestinal reflexes) or mesenchymopathy (associated with abnormalities of interstitial cells of Cajal). In addition, mitochondrial abnormalities

may contribute to impaired homeostasis of the intestinal microbiota, which in turn may be involved in the manifestation of gastrointestinal dysmotility seen in neurogastrointestinal encephalomyopathy [32]. In Wolfram's syndrome, diabetes mellitus and optic atrophy debut in the first decade of life, the other symptoms appear later, may be severely delayed by non-suggestive diabetes, hearing impairment and usually bowel and bladder dysfunction [33]. In all these muscular disorders, muscular dystrophies and mitochondrial cytopathy, NBD and urinary incontinence symptoms may change as the disease progresses.

Acquired brain injury (ABI) represents the leading cause of death and neurological disability in children after infancy. Today, the number of survivors of traumatic brain injury is increasing and these patients constitute a large proportion of patients in neurorehabilitation units. Functional impairments (motor, behavioural, learning and cognitive) including CC and NBD are common and may persist during all life [34].

Acquired damage to the nerves innervating the pelvic organs is iatrogenic in most cases, but rarely may also occur as a result of the impact. Any pelvic surgery in infants and children for anorectal malformation or Hirschsprung's disease [35, 36], neuroblastoma, ganglioneuroma, sacrococcygeal teratoma are theoretically capable of damaging the pelvic parasympathetic nerves of the rectum, anus, bladder and genitalia. In addition, pelvic irradiation can cause damage to adjacent nerve fibres, resulting in altered function, as can some cytotoxic drugs [36].

Acquired spinal cord injury leads to different types of defecation disorders: faecal incontinence, chronic constipation or a combination of both [37]. According to electromyography of the external anal sphincter, 25–33% of patients with spinal cord injury had bilateral or unilateral abnormalities of muscle action during defecation, and 88.5% had pelvic floor dysfunction [38]. The mean rectal volume for generating defecation urge was also elevated in them.

There are two models of bowel dysfunction are distinguished depending on the level of the conductive tract lesion relative to the *conus medullaris*: supraconal disorder, or "upper bowel motor neuron syndrome", or "hyperreflexic bowel", or "spastic bowel", and infraconal disorder, "lower motor neuron type", or "areflexic bowel" [39]. [39]. In the case of supraconal disorder, there is an increase in tone of the colon wall, pelvic floor and spasmodically constricted state of the external anal sphincter, which causes stool retention [39]. When the

anal sphincter cannot be relaxed arbitrarily, the signals between the colon and the brain become disconnected: the reflex that triggers bowel emptying is still working, but the child may not feel it coming, resulting in sudden unplanned passage of stool whenever the rectum is full. These disorders are characterised by high anal resting tone, anal and bulbocavernous reflex present. The situation is different in the case of an areflexic bowel: loss of colorectal tone and reduced amplitude of the rectoanal inhibitory reflex leads to a cyclic pattern of rectal filling and progressive distension of the rectum, eventually leading to faecal incontinence. In a situation of sluggish bowel, there is decreased movement in the colon, decreased peristalsis, and the anal sphincter is in a more relaxed state than normal. This can lead to constipation with frequent stool leakage. Typically, these patients have any or low anal resting tone and lack of anal and bulbocavernous reflexes [40].

The extent of symptoms also depends on the degree of injury: severe spinal injury has been shown to result in the most severe form of NBD with loss of control of the external anal sphincter [41].

Transverse myelitis is a rare immune-mediated disease resulting in spinal cord injury [42]. Approximately 20% of acute myelitis cases occur in children, in whom one of the most common initial symptoms is pain (60%). Other common symptoms in children include motor deficits, numbness, ataxic gait and loss of bowel or bladder control. Constipation can be severe and may be accompanied by a marked feeling of fullness in the left lower quadrant. Long-term autonomic sphincter dysfunction has been reported in 22–80% of children [43].

Multiple sclerosis (MS) is the most common progressive neurological disorder in young adults with a median age in the early 30s and a prevalence of 40–220 cases per 100,000 people in Europe [44], with similar rates in North America [45]. The incidence of childhood onset of multiple sclerosis is low, ranging from 0.3–0.9 per 100,000 people. The prevalence of childhood MS is 5–10% of all MS cases [46, 47]. Constipation in MS is observed in 31–54% of patients [48].

Defecation disorders have been described in 15% of patients in Guillain-Barré syndrome [49, 50], cauda equina syndrome (damage to nerve roots from L2 to S4) [51, 52], central lumbar disc prolapse, spinal cord tumour, spinal canal stenosis, spinal malformations and iatrogenic causes (during spinal surgery or spinal anaesthesia) [53].

Other rare paediatric neurological diseases affecting cells in the anterior horn of motor neurons

that control voluntary skeletal muscle activity (i.e. the external anal sphincter, not the bowel itself, have been described, although they can also indirectly affect bowel function due to weakened muscles and abdominal immobility). They are associated with a very poor prognosis as they are often progressive and there is currently no known cure (treatment is limited to symptomatic relief and support of basic vital functions such as breathing and feeding). Such diseases include spinal muscular atrophy, amyotrophic lateral sclerosis, progressive muscular atrophy, progressive bulbar palsy and primary lateral sclerosis [54], X-linked adrenoleukodystrophy [55], and Menkes disease [56]. Theoretically, any congenital or acquired disease that affects neurological and/or cognitive development and behaviour and results in limited mobility could also have secondary effects on the bowel (and bladder) of a child or adolescent.

Nutrition plays an important role in the development of constipation. Often people with neurological pathology have a reduced appetite and receive insufficient amounts of food and fluids [57]. Children on tube feeding or through a gastrostomy require the use of specialised high-calorie therapeutic mixtures enriched with dietary fibre [58]. However, constipation may occur even if the correct dietary regime is followed, if there is no possibility of comfortable defecation or if there is a negative experience associated with it. Thus, hard faeces when passing through the anus can traumatise it, causing pain. A vicious circle is created: the unwillingness to experience the painful sensation again makes the patient arbitrarily delay stool. Further, due to the reverse absorption of water by the intestine, the faecal masses become harder, which causes even more negative emotions in subsequent acts of defecation. There are other factors in the development of CC in LMP, such as metabolic and absorption disorders, side effects of certain medications (opioids, iron preparations, antacids, etc.), disruption of the intestinal microbiocenosis due to frequent antibiotics and a diet low in fibre, and lack of parental control and interest in the defecation of a child with low mobility [59–61].

DIAGNOSTICS

Diagnosis of CC begins with the complaints, but these may not be present due to various circumstances: disinterest of the patient or his/her representative, inability to assess the extent to which constipation affects quality of life, etc. When collecting anamnesis, it is necessary to assess whether the underlying disease, concomitant

Table 1. Possible bowel problems and associated neurological conditions [19]

Таблица 1. Возможные проблемы с кишечником и связанные с ними неврологические состояния [19]

Neurological pathology/ Неврологическая патология	Bowel and urinary problems / Проблемы с кишечником и мочеиспусканием
Spinal cord injury / Повреждение спинного мозга	<ul style="list-style-type: none"> • Loss of control and sensation of the need to defecate / Потеря контроля и ощущения потребности в дефекации. • Urinary incontinence and/or constipation / Недержание мочи и/или запор
<i>Spina Bifida</i>	<ul style="list-style-type: none"> • Constipation / Запор. • Stool incontinence / Недержание стула
Multiple sclerosis / Рассеянный склероз	<ul style="list-style-type: none"> • Constipation / Запор. • Stool incontinence / Недержание стула
Stroke and brain injury / Инсульт и черепно-мозговая травма	<ul style="list-style-type: none"> • Loss of conscious desire for defecation / Утрата осознанного желания дефекации. • Constipation / Запор. • Urinary incontinence / Недержание мочи
Cerebral palsy / Детский церебральный паралич	<ul style="list-style-type: none"> • Constipation / Запор
Parkinson's disease / Болезнь Паркинсона	<ul style="list-style-type: none"> • Constipation / Запор. • Less commonly urinary incontinence / Реже недержание мочи

pathology in LMP or previous gastrointestinal diseases influence the occurrence of constipation. It is necessary to find out when difficulties in defecation appeared, whether their appearance is connected with pelvic or spinal surgery, with trauma; what sensations the patient experiences against this background (a discomfort, its localisation, an increased gas formation, a feeling of incomplete emptying of the rectum after stool discharge, pain and other symptoms such as nausea, vomiting, decreased appetite, signs of dysuria) and how often stool is discharged. It should be clarified whether there are factors that improve or worsen the situation, such as a change in the amount of food or drink, taking certain medications, or changes in motor activity. Particular attention should be paid to the quality of the stool, its colour, density, odour, quantity and presence of abnormal impurities such as blood or mucus. When the last stool occurred, whether the patient has urges to defecate and whether he or she needs to push. During the discussion, the doctor should determine the patient's understanding of the importance of constipation management, as some patients and their parents may not be bothered by such difficulties. Attention should also be paid to social factors, such as how the patient and family feel about the problem, the importance the patient attaches to constipation, and whether there is privacy and a comfortable environment for defecation [62]. It is likely that the patient has already tried to control constipation, so it is necessary to clarify how this occurred and whether there was an effect.

Percussion to look for bowel bloating may be performed, as well as abdominal palpation and palpebral rectal examination. The patient's examination should form an opinion about the nature of the constipation: functional and/or organic [63].

An additional diagnostic method is the research of intestinal transit using X-ray contrast markers [64]. The colonic transit time can be used only in extremely unclear cases as a differential sign between functional constipation and functional faecal incontinence without constipation.

TREATMENT

The approach to the treatment of constipation in LMP varies according to the pathogenetic basis of constipation. In patients with hyperreflexic bowel, stimulation of the rectum (chemically or mechanically) results in evacuation of any rectal stool. The goal in hyperreflexic bowel is to achieve a relatively soft stool consistency to stimulate evacuation. In these patients, stool softeners and stimulant laxatives with mechanical stimulation of the anorectal area can provide stool relief. Patients with areflexic bowel may require abdominal muscle exercises and manual evacuation of stool. In patients who have low anal sphincter tone at rest, more formed stools may help reduce episodes of incontinence, so excessive use of stool softeners and stimulant laxatives should be avoided [65]. In patients with lesion levels at T6 or above any treatment that results in rapid rectal emptying carries the risk of precipitating life-threatening autonomic dysreflexia [66]. Patients at risk or carers

should be made aware of this risk and informed of appropriate emergency treatment (nifedipine).

In general, the treatment of CC in LMP should be staged, with the aim of finding the least invasive intervention that normalises stool consistency and frequency. The treatment approaches proposed by foreign researchers are presented in Figure 1. Treatment should be carried out for at least two weeks consecutively before considering further modification of the program.

First of all, dietary adjustments are required: an increase in the amount of fibre or other bulking agents in the diet and optimization of water balance [63, 68, 69]. For LMP with cognitive impairment, diet should be treated very carefully: an abdominal bloating and subsequent pain associated with flatulence cause them to scream and become uncontrollably anxious. Increased motor activity and traditional positioning during defecation also cannot be used in LMP. The use of technical rehabilitation devices (TRD) for daily positioning can improve bowel peristalsis and resolution of constipation with nutritional therapy. Rehabilitative aids are individually selected by orthopaedists and/or occupational therapists, and may include sitting supports, standing supports, walkers with additional body support, including for patients with cerebral palsy [70]. Abdominal massage activates intestinal stretch receptors, which causes increased contraction of the bowel and rectum, excites waves of contraction of the rectus abdominis muscles, decreases colon transit time, and stimulates the parasympathetic nervous system, thereby leading to increased intestinal secretion and motility and relaxation of sphincters in the digestive tract [71]. Mechanical effects may also be observed in lean LMP. Abdominal massage in children is usually performed starting at the right iliac region, using a gentle, squeezing, kneading motion in an inverted "U" direction around the top of the umbilicus to the left iliac fossa and then deep into the suprapubic region to help move gas and stool along the course of the colon to the rectum [72].

Anal/rectal stimulation [73] is a well-established technique used in LMP with constipation to facilitate bowel evacuation. The LMP caregiver inserts a lubricated finger (in medical glove) into the rectum and performs a rotary motion, dilating the anal canal and relaxing the pubic muscle, resulting in a reduction of the anorectal angle. Both of these effects result in reduced resistance to the passage of stool, thereby promoting bowel emptying [73]. This method of stimulation is very different from manual evacuation, where stool is extracted di-

rectly with the finger and which is generally not suitable as a regular treatment for the older child. Some studies have demonstrated the efficacy of non-invasive nerve stimulation, such as percutaneous electrical nerve stimulation [74], posterior tibial nerve stimulation [75] in the treatment of constipation.

If there is no effect from dietary changes, oral laxatives may be used, aimed at changing the rate of fecal passage. However, the side effects of the drugs used should be taken into account: a mineral oil, an osmotic laxative (polyethylene glycol preparations, lactulose) can aggravate fecal incontinence. Intestinal peristalsis stimulants such as Senna extract and bisacodyl are widely used. Rectal-sigmoid emptying can be carried out with a small-volume enema, glycerin suppositories. Sodium phosphate enemas should be avoided in children with renal impairment [3]. If there is no effect, the use of a larger volume enema administered through a conical tip or a special catheter, which provides transanal irrigation, is recommended [76]. The final step is the use of an antegrade enema administered through commercially available transanal irrigation systems. For the LMP and carers daily suppositories are more convenient and comfortable to use than transrectal irrigations, because with constant use it avoids fecal blockage and the need for 'high' enemas.

Surgical treatment of constipation in LMP should be considered taking into account all of

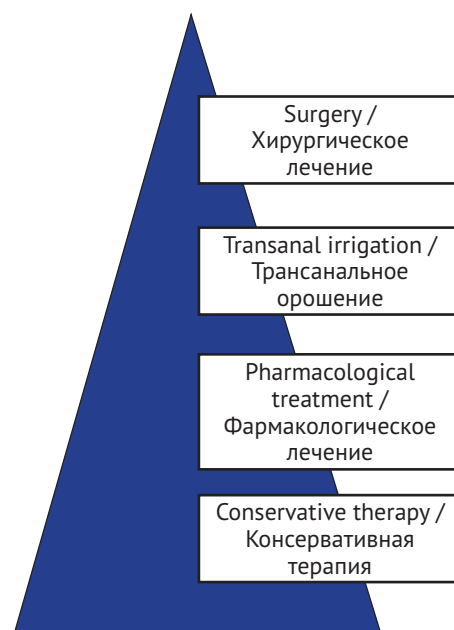


Fig. 1. Pyramid of recommendations for the treatment of neurogenic bowel dysfunction (adapted from [67])

Рис. 1. Пирамида рекомендаций по лечению нейрогенной дисфункции кишечника (адаптировано по [67])

Table 2. Guidelines for bowel care and function for people with spina bifida in different age periods [77]

Таблица 2. Руководящие принципы по уходу и функции кишечника для людей с расщелиной позвоночника в разные возрастные периоды [77]

Age group / Возрастная группа	Guidelines / Руководящие принципы
0-11 months / 0–11 месяцев	<ol style="list-style-type: none"> 1. Monitor stool frequency, consistency and quantity / Контролируйте частоту стула, консистенцию и его количество. 2. Use dietary treatment, in particular breastfeeding if possible, as breast milk is easier to digest and provides a better recovery of the microbiome after surgery / Используйте диетическое лечение, в частности грудное вскармливание, если это возможно, так как грудное молоко легче усваивается и обеспечивает лучшее восстановление микробиома после операции. 3. Consider dietary treatment (fibre and fluids) before pharmacological supplements and/or rectal stimulants (glycerine suppositories) to treat constipation / Рассмотрите диетическое лечение (клетчатка и жидкости) перед фармакологическими добавками и/или ректальными стимуляторами (глицериновые суппозитории) для лечения запоров. 4. Use barrier creams to protect the perineal area as needed / Используйте барьерные кремы для защиты области промежности по мере необходимости.
1–2 years 11 months / 1–2 года 11 месяцев	<ol style="list-style-type: none"> 1. Discuss toilet training and habits with parents / Обсудите с родителями обучение туалету и привычкам. 2. Set a goal to work towards correcting stool incontinence / Установите цель работы в направлении коррекции недержания стула. 3. Use fibre, sufficient fluids by mouth, exercise and a chronobiological approach (defecation in the morning after meals) / Используйте клетчатку, достаточное количество жидкости через рот, физические упражнения и хронобиологический подход (дефекации утром после еды). 4. Consider prescribing oral and rectal interventions for constipation / Рассмотрите назначение пероральных и ректальных вмешательств для борьбы с запорами. 5. Use dietary treatment (fibre and fluids), pharmacological supplements (sennoside, polyethylene glycol) and/or rectal stimulants (glycerin, docusate sodium or bisacodyl suppositories) to treat constipation and faecal incontinence / Используйте диетическое лечение (клетчатка и жидкости), фармакологические добавки (сеннозид, полиэтиленгликоль) и/или ректальные стимуляторы (глицерин, докузат натрия или бисакодил-суппозитории) для лечения запоров и недержания кала. 6. Use barrier creams to protect the perineal area as needed / Используйте барьерные кремы для защиты области промежности по мере необходимости. 7. Consult a <i>Spina Bifida</i> clinic or a specialist with expertise in bowel management / Обратитесь в клинику <i>Spina Bifida</i> или к специалисту с опытом в области управления кишечником при <i>Spina Bifida</i>.
3–5 years 11 months / 3–5 года 11 месяцев	<ol style="list-style-type: none"> 1. Discuss the consequences of constipation and bowel incontinence (including shunt malfunction, urinary tract infections (UTIs), skin maceration, social isolation) / Обсудите последствия запоров и недержания кишечника (включая неисправность шунтов, инфекции мочевыводящих путей (ИМП), мацерацию кожи, социальную изоляцию). 2. Set a treatment goal and establish a bowel control programme, using the recommendations given / Установите цель лечения и установите программу контроля за работой кишечника, используя приведенные рекомендации. 3. Use fibre, adequate oral fluids, exercise, and a chronobiological approach, exercise and a chronobiological approach (defecation in the morning after meals) / Используйте клетчатку, достаточное количество жидкости через рот, физические упражнения и хронобиологический подход (дефекации утром после еды). 4. Consider prescribing oral and rectal interventions to control management of constipation / Рассмотрите назначение пероральных и ректальных вмешательств для борьбы с запорами. 5. Use dietary treatment (fibre and fluids), pharmacological supplements (sennoside, polyethylene glycol) and/or rectal stimulants (glycerin, sodium docusate or bisacodyl suppositories) to treat constipation and faecal incontinence / Используйте диетическое лечение (клетчатка и жидкости), фармакологические добавки (сеннозид, полиэтиленгликоль) и/или ректальные стимуляторы (глицерин, докузат натрия или бисакодил-суппозитории) для лечения запоров и недержания кала. 6. Use barrier creams to protect the perineal area from maceration as needed / Используйте барьерные кремы для защиты области промежности от мацерации по мере необходимости. 7. Consult a <i>Spina Bifida</i> clinic or a specialist with experience in bowel management for <i>Spina Bifida</i> / Обратитесь в клинику <i>Spina Bifida</i> или к специалисту с опытом управления кишечником при <i>Spina Bifida</i>

the patient's problems, including general prognosis, mental function, and trophological status, not just defecation problems that are not resolved by therapeutic methods.

There are no consensus documents on the management of constipation and defecation disorders in LMP; only for children with spina bifida has a standardized approach to the management of bowel dysfunction been developed [77]. Guidelines for bowel care and function for people with spina bifida at different ages are summarized in Table 2. For older age, the recommendations are the same.

In summary, the management of constipation and defecation disorders in pediatric LMP is challenging and has significant psychosocial implications for both the patient and carers. Clinical guidelines are needed to provide a proactive, systematic and rational approach to the management of bowel dysfunction, including fecal incontinence and constipation. Collaborative efforts between multidisciplinary specialists are needed to overcome research barriers and provide innovative solutions.

ADDITIONAL INFORMATION

Author contribution. Thereby, all authors made a substantial contribution to the conception of the study, acquisition, analysis, interpretation of data for the work, drafting and revising the article, final approval of the version to be published and agree to be accountable for all aspects of the study.

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REFERENCES

1. Minushkin O.N. Khronicheskiy zapor (opredeleniye, epidemiologiya, diagnostika): sovremennaya

medikamentoznaya terapiya. [Chronic constipation (definition, epidemiology, diagnosis): modern drug therapy]. MS. 2015; 13. URL: <https://cyberleninka.ru/article/n/hronicheskiy-zapor-opredelenie-epidemiologiya-diagnostika-sovremennaya-medikamentoznaya-terapiya> (data obrashcheniya: 29.04.2023). (in Russian).

2. Khavkin A.I., Fayzullina R.A., Bel'mer S.V. i dr. Diagnostika i lecheniye funktsional'nykh zaporov u detey (fragment Proyekta klinicheskikh rekomendatsiy po diagnostike i lecheniyu funktsional'nykh zabolevaniy organov pishchevareniya u detey Rossiyskogo obshchestva detskikh gastroenterologov, gepatologov i nutritsiologov). [Diagnosis and treatment of functional constipation in children (a fragment of the Draft Clinical Guidelines for the Diagnosis and Treatment of Functional Diseases of the Digestive Organs in Children of the Russian Society of Pediatric Gastroenterologists, Hepatologists and Nutritionists)]. Farmateka. 2020; 27(2): 60–8. DOI: <https://dx.doi.org/10.18565/pharmateka.2020.2.60-68>. (in Russian).
3. Mel'nikova I.Yu., Novikova V.P., Dumova N.B. Zapory u detey. [Constipation in children]. Rukovodstvo. 3-ye izd., pererab. i dop. Moskva: GEOTAR-Media Publ.; 2020. (in Russian).
4. Bel'mer S.V., Khavkin A.I., Pechkurov D.V. Funktsional'nyye narusheniya organov pishchevareniya u detey. [Functional disorders of the digestive system in children]. Printsipy diagnostiki i lecheniya (v svete Rimskikh kriteriyev IV). Moskva: GEOTAR-Media Publ.; 2018. (in Russian).
5. Zapory u detey. [Constipation in children]. Pod redaktsiyey S.V. Bel'mera, A.Yu. Razumovskogo, A.I. Khavkina, R.A. Fayzullinoy. Moskva: Medpraktika-M Publ.; 2016. (in Russian).
6. Tabbers M.M., DiLorenzo C., Berger M.Y. et al. Evaluation and treatment of functional constipation in infants and children: evidence-based recommendations from ESPGHAN and NASPGHAN. *J Pediatr Gastroenterol Nutr.* 2014; 58 (2): 258–74.
7. Palliativnaya pomoshch' bol'nym s khronicheskimi progressiruyushchimi zabolevaniyami v inkurabel'noy stadii, soprovozhdayushchimisya zaporami. [Palliative care for patients with chronic progressive diseases in the incurable stage, accompanied by constipation]. *Klinicheskiye protokoly MZ RK. Kazakhstan;* 2019. (in Russian).
8. Andrews Ch.N., Storr M. Patofiziologiya khronicheskogo zapora. [Pathophysiology of chronic constipation]. *Can J Gastroenterol.* 2011; 25: 17B–23B.
9. Galeysya Ye.N., Lychkova A.E. Nervnaya regulyatsiya tolstoy kishki. [Nervous regulation of the colon]. *EiKG.* 2013; 8. (in Russian).

10. Coggrave M. Neurogenic continence. Part 3: Bowel management strategies. *Br J Nurs.* 2008; 17(15): 962–8. DOI: 10.12968/bjon.2008.17.15.30698.
11. Krogh K., Christensen P., Laurberg S. Colorectal symptoms in patients with neurological diseases. *Acta Neurol. Scand.* 2001; 103: 335–43. DOI: 10.1034/j.1600-0404.2001.103006335.x.
12. Richards C.L., Malouin F. Cerebral palsy: Definition, assessment and rehabilitation. *Handb. Clin. Neurol.* 2013; 111: 183–95. DOI: 10.1016/B978-0-444-52891-9.00018-X.
13. Ozturk M., Oktem F., Kisloglu N., et al. Bladder and bowel control in children with cerebral palsy. *Croat. Med. J.* 2006; 47: 264–70.
14. Kamalova A.A., Rakhmayeva R.F., Malinovskaya Yu.V. Morfofunktsional'nyye osobennosti organov pishchevareniya pri detskom tserebral'nom paraliche. [Morphofunctional features of the digestive organs in children with cerebral palsy]. *EiKG.* 2020; 1: 173. (in Russian).
15. Erkin G., Culha C., Ozel S. et al. Feeding and gastrointestinal problems in children with cerebral palsy. *Int. J. Rehabil. Res.* 2010; 33(3): 218–24.
16. Afanas'yeva A.A., Chuynyshena S.A. Fizicheskoye razvitiye, nutritivnyy status i gastroenterologicheskiye problemy u detey s DTSP. [Physical development, nutritional status and gastroenterological problems in children with cerebral palsy]. *Forcipe.* 2019. (in Russian).
17. Von Wendt L., Similä S., Niskanen P., Järvelin M.R. Development of bowel and bladder control in the mentally retarded. *Dev. Med. Child Neurol.* 1990; 32: 515–8. DOI: 10.1111/j.1469-8749.1990.tb16977.x.
18. Bohmer C.J., Taminiau J.A., Klinkenberg-Knol E.C., Meuwissen S.G. The prevalence of constipation in institutionalized people with intellectual disability. *J. Intellect. Disabil. Res.* 2001; 45: 212–8. DOI: 10.1046/j.1365-2788.2001.00300.x.
19. Pellat G.C. Neurogenic continence. Part 1: pathophysiology and quality of life. *Br J Nurs.* 2008; 17(13): 836–41. DOI: 10.12968/bjon.2008.17.13.30534.
20. Prenatal'naya diagnostika nasledstvennykh i vrozhdennykh bolezney. [Prenatal diagnosis of hereditary and congenital disease]. *Pod red. E.K. Aylamazyan, V.S. Baranova.* Moskva: MEDpress-inform Publ.; 2006: 416. (in Russian).
21. Rocque B.G., Hopson B.D., Blount J.P. Caring for the Child with Spina Bifida. *Pediatr Clin North Am.* 2021; 68(4): 915–27. DOI: 10.1016/j.pcl.2021.04.013.
22. Barker E., Saulino M., Caristo A.M. Spina bifida. *RN.* 2002; 65(12): 33–8; quiz 39.
23. Agnarsson U., Warde C., McCarthy G. et al. Anorectal function of children with neurological problems. I: Spina bifida. *Dev Med Child Neurol.* 1993; 35(10): 893–902. DOI: 10.1111/j.1469-8749.1993.tb11566.x.
24. Nejat F., Radmanesh F., Ansari S. et al. Spina bifida occulta: is it a predictor of underlying spinal cord abnormality in patients with lower urinary tract dysfunction? *J Neurosurg Pediatr.* 2008; 1(2): 114–7. DOI: 10.3171/PED/2008/1/2/114.
25. Sawin K.J., Bellin M.H. Quality of life in individuals with spina bifida: a research update. *Dev Disabil Res Rev.* 2010; 16(1): 47–59. DOI: 10.1002/ddrr.96.
26. Bakaniene I., Prasauskiene A., Vaiciene-Magistris N. Health-related quality of life in children with myelomeningocele: a systematic review of the literature. *Child Care Health Dev.* 2016; 42(5): 625–43. DOI: 10.1111/cch.12367.
27. Choi E.K., Im Y.J., Han S.W. Bowel management and quality of life in children with spina bifida in South Korea. *Gastroenterol Nurs.* 2017; 40(3): 208–15. DOI: 10.1097/SGA.000000000000135.
28. Rocque B.G., Ralee' Bishop E., Scogin M.A. et al. Assessing health-related quality of life in children with spina bifida. *J Neurosurg Pediatr.* 2015; 15(2): 144–9. DOI: 10.3171/2014.10.
29. Burns A.S., St-Germain D., Connolly M. et al. Phenomenological study of neurogenic bowel from the perspective of individuals living with spinal cord injury. *Arch Phys Med Rehabil.* 2015; 96(1): 49–55. DOI: 10.1016/j.apmr.2014.07.417.
30. Lo Cascio C.M., Goetze O., Latshang T.D. Gastrointestinal Dysfunction in Patients with et al. Duchenne Muscular Dystrophy. *PLoS ONE.* 2016; 11: e0163779. DOI: 10.1371/journal.pone.0163779.
31. Crockett C.D., Bertrand L.A., Cooper C.S. et al. Urologic and gastrointestinal symptoms in the dystroglycanopathies. *Neurology.* 2015; 84: 532–9. DOI: 10.1212/WNL.0000000000001213.
32. Yadak R., Breur M., Bugiani M. Gastrointestinal Dysmotility in MNGIE: From thymidine phosphorylase enzyme deficiency to altered interstitial cells of Cajal. *Orphanet J. Rare Dis.* 2019; 14: 33. DOI: 10.1186/s13023-019-1016-6.
33. Barrett T.G., Scott-Brown M., Seller A. et al. The mitochondrial genome in Wolfram syndrome. *J. Med. Genet.* 2000; 37: 463–6. DOI: 10.1136/jmg.37.6.463.
34. Chiminello R., Mosiello G., Castelli E. Bladder and bowel dysfunctions in children with cerebral palsy and acquired brain injury: Are we able to define incidence and to predict them? *Eur. Urol. Suppl.* 2018; 17: e1179–e1180. DOI: 10.1016/S1569-9056(18)31662-2.
35. Berger M., Heinrich M., Lacher M. et al. Postoperative bladder and rectal function in children with sacro-coccygeal teratoma. *Pediatr. Blood Cancer.* 2011; 56: 397–402. DOI: 10.1002/pbc.22887.

36. Mosiello G., Gatti C., De Gennaro M. et al. Neurovesical dysfunction in children after treating pelvic neoplasms. *BJU Int.* 2003; 92: 289–92. DOI: 10.1046/j.1464-410X.2003.04326.x.
37. Ng C., Prutt G., Rutkowski S. et al. Gastrointestinal symptoms in spinal cord injury: Relationships with level of injury and psychologic factors. *Dis. Colon Rectum.* 2005; 48: 1562–8. DOI: 10.1007/s10350-005-0061-5.
38. Silveri M., Salsano L., Pierro M.M. et al. Pediatric spinal cord injury: Approach for urological rehabilitation and treatment. *J. Pediatr. Urol.* 2006; 2: 10–5. DOI: 10.1016/j.jpuro.2005.05.004.
39. Rasmussen M.M., Krogh K., Clemmensen D. et al. Colorectal transport during defecation in subjects with supraconal spinal cord injury. *Spinal Cord.* 2013; 51(9): 683–7. DOI: 10.1038/sc.2013.58.
40. Valles M., Vidal J., Clave P., Mearin F. Bowel dysfunction in patients with motor complete spinal cord injury: Clinical, neurological, and pathophysiological associations. *Am. J. Gastroenterol.* 2006; 101: 2290–9. DOI: 10.1111/j.1572-0241.2006.00729.x.
41. Qi Z., Middleton J.W., Malcolm A. Bowel Dysfunction in Spinal Cord Injury. *Curr. Gastroenterol. Rep.* 2018; 20: 47. DOI: 10.1007/s11894-018-0655-4.
42. Pidcock F.S., Krishnan C., Crawford T.O. et al. Acute transverse myelitis in childhood: Center-based analysis of 47 cases. *Neurology.* 2007; 68: 1474–80. DOI: 10.1212/01.wnl.0000260609.11357.6f.
43. Wolf V.L., Lupo P.J., Lotze T.E. Pediatric transverse myelitis. *J. Child Neurol.* 2012; 27: 1426–36. DOI: 10.1177/0883073812452916.
44. Kingwell E., Marriott J.J., Jetté N. et al. Incidence and prevalence of multiple sclerosis in Europe: a systematic review. *BMC Neurol.* 2013; 13: 128. DOI: 10.1186/1471-2377-13-128.
45. Evans C., Beland S.G., Kulaga S. et al. Incidence and prevalence of multiple sclerosis in the Americas: a systematic review. *Neuroepidemiology.* 2013; 40(3): 195–210. DOI: 10.1159/000342779.
46. Mah J., Thannhauser J.E. Management of multiple sclerosis in adolescents — current treatment options and related adherence issues. *Adolesc Health Med Ther.* 2010; 1: 31–43. <https://doi.org/10.2147/AHMT.S7594>.
47. Kuntz N.L., Chabas D., Weinstock-Guttman B. et al. Network of US Pediatric Multiple Sclerosis Centers. Treatment of multiple sclerosis in children and adolescents. *Expert Opin Pharmacother.* 2010; 11(4): 505–20. DOI: 10.1517/14656560903527218.
48. Munteis E., Andreu M., Téllez M.J. et al. Anorectal dysfunction in multiple sclerosis. *Mult Scler.* 2006; 12(2): 215–8. DOI: 10.1191/135248506ms1254oa.
49. Sawai S., Sakakibara R., Uchiyama T. et al. Acute motor axonal neuropathy presenting with bowel, bladder, and erectile dysfunction. *J. Neurol.* 2007; 254: 250–2. DOI: 10.1007/s00415-006-0336-3.
50. Zochodne D.W. Autonomic involvement in Guillain-Barre syndrome: A review. *Muscle Nerve.* 1994; 17: 1145–55. DOI: 10.1002/mus.880171004.
51. McCarthy M.J., Aylott C.E., Grevitt M.P., Hegarty J. Cauda equina syndrome: Factors affecting long-term functional and sphincteric outcome. *Spine.* 2007; 32: 207–16. DOI: 10.1097/01.brs.0000251750.20508.84.
52. Podnar S. Bowel dysfunction in patients with cauda equina lesions. *Eur. J. Neurol.* 2006; 13: 1112–7. DOI: 10.1111/j.1468-1331.2006.01423.x.
53. Podnar S. Epidemiology of cauda equina and conus medullaris lesions. *Muscle Nerve.* 2007; 35: 529–31. DOI: 10.1002/mus.20696.
54. McDermott C.J., Shaw P.J. Diagnosis and management of motor neurone disease. *BMJ.* 2008; 336: 658–62. DOI: 10.1136/bmj.39493.511759.BE.
55. Silveri M., De Gennaro M., Gatti C. et al. Voiding dysfunction in x-linked adrenoleukodystrophy: Symptom score and urodynamic findings. *J. Urol.* 2004; 171: 2651–3. DOI: 10.1097/01.ju.0000110885.26017.b0.
56. Kim M.Y., Kim J.H., Cho M.H. et al. Urological Problems in Patients with Menkes Disease. *J. Korean Med. Sci.* 2019; 34: e4. DOI: 10.3346/jkms.2019.34.e4.
57. Pak L.A., Makarova S.G., Fisenko A.P. i dr. Opyt organizatsii pitaniya patsiyentov s detskim tserebral'nym paralichom v khode kompleksnoy reabilitatsii na baze federal'nogo tsentra. [Experience in catering for patients with cerebral palsy in the course of complex rehabilitation on the basis of the federal center]. *Nevrologicheskiy zhurnal imeni L.O. Badalyana.* 2020; 1(2): 100–11. <https://doi.org/10.46563/2686-8997-2020-1-2-100-111>. (in Russian).
58. Afanas'yeva A.A., Chuynyshena S.A. Otsenka fizicheskogo razvitiya i nutritivnogo statusa u detey s detskim tserebral'nym paralichom, gospitalizirovannykh v mnogoprofil'nyy pediatricheskiy statsionar. [Assessment of physical development and nutritional status in children with cerebral palsy hospitalized in a multidisciplinary pediatric hospital]. *Forcipe.* 2020. URL: <https://cyberleninka.ru/article/n/otsenka-fizicheskogo-razvitiya-i-nutritivnogo-statusa-u-detey-s-detskim-tserebralnym-paralichom-gospitalizirovannykh-v> (data obrashcheniya: 19.05.2023). (in Russian).
59. Rendeli C., Paradiso V.F., Bucci V. et al. Gut microbiota and pediatric patients with spina bifida and neurogenic bowel dysfunction. *Childs Nerv Syst.* 2023; 39(3): 633–45. DOI: 10.1007/s00381-022-05688-0.

60. Furuta A., Suzuki Y., Takahashi R. et al. Effects of Transanal Irrigation on Gut Microbiota in Pediatric Patients with Spina Bifida. *J Clin Med.* 2021; 10(2): 224. DOI: 10.3390/jcm10020224.
61. Zav'yalova A.N., Novikova V.P., Klikunova K.A. Yunosheskaya sarkopeniya u patsiyentov s det-skim tserebral'nym paralichom i prediktory yeye razvitiya. [Juvenile sarcopenia in patients with cerebral palsy and predictors of its development]. *Forcipe.* 2022; 52. (in Russian).
62. Tsapina S.Yu. Problema komplaentnosti sem'i rebenka s DTSP. [The problem of family compliance of a child with cerebral palsy]. *Vestnik fizioterapii i kurortologii.* 2015; 2 URL: <https://cyberleninka.ru/article/n/problema-komplaentnosti-semi-rebenka-s-dtsp> (data obrashcheniya: 19.05.2023). (in Russian).
63. Bel'mer S.V., Khavkin A.I., Pechkurov D.V. Funktsional'nyye zapory. [Functional constipation]. *Meditsinskaya sestra.* 2017; (7): 28–31. (in Russian).
64. Kazuyulin A.N., Kucheryavyy Yu.A. Prakticheskiye i klinicheskiye aspekty sindroma khronicheskogo zapora. [Practical and clinical aspects of chronic constipation syndrome]. Moskva; 2005: 21. (in Russian).
65. Mosiello G., Safder S., Marshall D. et al. Neurogenic Bowel Dysfunction in Children and Adolescents. *J Clin Med.* 2021; 10(8): 1669. DOI: 10.3390/jcm10081669.
66. Kewalramani L.S. Autonomic dysreflexia in traumatic myelopathy. *Am. J. Phys. Med.* 1980; 59: 1–21.
67. Mosiello G., Marshall D., Rolle U. et al. Consensus review of best practice of transanal irrigation in children. *J. Pediatr. Gastroenterol. Nutr.* 2017; 64: 343–52. DOI: 10.1097/MPG.0000000000001483.
68. Korenyuk Ye.S., Ivanchenko V.I., Alekseyeva O.G. Primeneniye poluelementnykh smesey v enteral'nom pitanii detey s tyazhelymi nevrologicheskimi zabolevaniyami. [The use of semi-elemental mixtures in enteral nutrition of children with severe neurological diseases]. 2016; 5 (73): 83–5. DOI: 10.22141/2224-0551.5.1.73.1.2016.78947. (in Russian).
69. Komarova O.N. Enteral'noye pitaniye u detey s khronicheskimi zabolevaniyami: optimal'nyy sposob dostavki. [Enteral nutrition in children with chronic diseases: the optimal method of delivery]. *Meditsinskiy Sovet.* 2020; 18: 87–93. <https://doi.org/10.21518/2079-701X-2020-18-87-93>. (in Russian).
70. Prikaz Ministerstva truda i sotsial'noy zashchity RF ot 13 fevralya 2018 g. № 86n "Ob utverzhdenii klassifikatsii tekhnicheskikh sredstv reabilitatsii (izdeliy) v ramkakh federal'nogo perechnya reabilitatsionnykh meropriyatiy, tekhnicheskikh sredstv reabilitatsii i uslug, predostavlyayemykh invalidu, utverzhdenogo rasporyazheniyem Pravitel'stva Rossiyskoy Federatsii ot 30 dekabrya 2005 g. N 2347-p" (in Russian).
71. Turan N., Aşt T.A. The Effect of Abdominal Massage on Constipation and Quality of Life. *Gastroenterol. Nurs.* 2016; 39: 48–59. DOI: 10.1097/SGA.0000000000000202.
72. Wald A. Constipation in the primary care setting: Current concepts and misconceptions. *Am. J. Med.* 2006; 119: 736–9. DOI: 10.1016/j.amjmed.2006.03.025.
73. Shafik A. Recto-colic reflex: Role in the defecation mechanism. *Int. Surg.* 1996; 81: 292–4.
74. Veiga M.L., Lordêlo P., Farias T., Barroso U., Jr. Evaluation of constipation after parasacral transcutaneous electrical nerve stimulation in children with lower urinary tract dysfunction — A pilot study. *J. Pediatr. Urol.* 2013; 9: 622–6. DOI: 10.1016/j.jpuro.2012.06.006.
75. Mentés B.B., Yüksel O., Aydın A. et al. Posterior tibial nerve stimulation for faecal incontinence after partial spinal injury: Preliminary report. *Tech. Coloproctol.* 2007; 11: 115–9. DOI: 10.1007/s10151-007-0340-3.
76. Patel S., Hopson P., Bornstein J., Safder S. Impact of Transanal Irrigation Device in the Management of Children With Fecal Incontinence and Constipation. *J Pediatr Gastroenterol Nutr.* 2020; 71(3): 292–7. DOI: 10.1097/MPG.0000000000002785.
77. Beierwaltes P., Church P., Gordon T., Ambartsumyan L. Bowel function and care: Guidelines for the care of people with spina bifida. *J Pediatr Rehabil Med.* 2020; 13(4): 491–8. DOI: 10.3233/PRM-200724.

ЛИТЕРАТУРА

1. Минущкин О.Н. Хронический запор (определение, эпидемиология, диагностика): современная медикаментозная терапия. МС. 2015;13. URL: <https://cyberleninka.ru/article/n/hronicheskiiy-zapor-opredelenie-epidemiologiya-diagnostika-sovremennaya-medikamentoznaya-terapiya> (дата обращения: 29.04.2023).
2. Хавкин А.И., Файзуллина Р.А., Бельмер С.В. и др. Диагностика и лечение функциональных запоров у детей (фрагмент Проекта клинических рекомендаций по диагностике и лечению функциональных заболеваний органов пищеварения у детей Российского общества детских гастроэнтерологов, гепатологов и нутрициологов). Фарматека. 2020; 27(2): 60–8. DOI: <https://dx.doi.org/10.18565/pharmateca.2020.2.60-68>.
3. Мельникова И.Ю., Новикова В.П., Думова Н.Б. Запоры у детей. Руководство. 3-е изд., перераб. и доп. М.: ГЭОТАР-Медиа; 2020.

4. Бельмер С.В., Хавкин А.И., Печкуров Д.В. Функциональные нарушения органов пищеварения у детей. Принципы диагностики и лечения (в свете Римских критериев IV). М.: ГЭОТАР-Медиа; 2018.
5. Запоры у детей. Под редакцией С.В. Бельмера, А.Ю. Разумовского, А.И. Хавкина, Р.А. Файзуллиной. М.: Медпрактика-М; 2016.
6. Tabbers M.M., DiLorenzo C., Berger M.Y. et al. Evaluation and treatment of functional constipation in infants and children: evidence-based recommendations from ESPGHAN and NASPGHAN. *J Pediatr Gastroenterol Nutr.* 2014; 58 (2): 258–74.
7. Паллиативная помощь больным с хроническими прогрессирующими заболеваниями в инкурабельной стадии, сопровождающимся запорами. Клинические протоколы МЗ РК. Казахстан; 2019.
8. Andrews Ch.N., Storr M. Патология хронического запора. *Can J Gastroenterol.* 2011; 25: 17B–23B.
9. Галейся Е.Н., Лычкова А.Э. Нервная регуляция толстой кишки. *ЭиКГ.* 2013; 8.
10. Coggrave M. Neurogenic continence. Part 3: Bowel management strategies. *Br J Nurs.* 2008; 17(15): 962–8. DOI: 10.12968/bjon.2008.17.15.30698.
11. Krogh K., Christensen P., Laurberg S. Colorectal symptoms in patients with neurological diseases. *Acta Neurol. Scand.* 2001; 103: 335–43. DOI: 10.1034/j.1600-0404.2001.103006335.x.
12. Richards C.L., Malouin F. Cerebral palsy: Definition, assessment and rehabilitation. *Handb. Clin. Neurol.* 2013; 111: 183–95. DOI: 10.1016/B978-0-444-52891-9.00018-X.
13. Ozturk M., Oktem F., Kisloglu N., et al. Bladder and bowel control in children with cerebral palsy. *Croat. Med. J.* 2006; 47: 264–70.
14. Камалова А.А., Рахмаева Р.Ф., Малиновская Ю.В. Морфофункциональные особенности органов пищеварения при детском церебральном параличе. *ЭиКГ.* 2020; 1: 173.
15. Erkin G., Culha C., Ozel S. et al. Feeding and gastrointestinal problems in children with cerebral palsy. *Int. J. Rehabil. Res.* 2010; 33(3): 218–24.
16. Афанасьева А.А., Чуйнышена С.А. Физическое развитие, нутритивный статус и гастроэнтерологические проблемы у детей с ДЦП. *Forcipe.* 2019.
17. Von Wendt L., Similä S., Niskanen P., Järvelin M.R. Development of bowel and bladder control in the mentally retarded. *Dev. Med. Child Neurol.* 1990; 32: 515–8. DOI: 10.1111/j.1469-8749.1990.tb16977.x.
18. Bohmer C.J., Taminiau J.A., Klinkenberg-Knol E.C., Meuwissen S.G. The prevalence of constipation in institutionalized people with intellectual disability. *J. Intellect. Disabil. Res.* 2001; 45: 212–8. DOI: 10.1046/j.1365-2788.2001.00300.x.
19. Pellat G.C. Neurogenic continence. Part 1: pathophysiology and quality of life. *Br J Nurs.* 2008; 17(13): 836–41. DOI: 10.12968/bjon.2008.17.13.30534.
20. Пренатальная диагностика наследственных и врожденных болезней. Под ред. Э.К. Айламазяна, В.С. Баранова. М.: МЕДпресс-информ; 2006: 416.
21. Rocque B.G., Hopson B.D., Blount J.P. Caring for the Child with Spina Bifida. *Pediatr Clin North Am.* 2021; 68(4): 915–27. DOI: 10.1016/j.pcl.2021.04.013.
22. Barker E., Saulino M., Caristo A.M. Spina bifida. *RN.* 2002; 65(12): 33–8; quiz 39.
23. Agnarsson U., Warde C., McCarthy G. et al. Anorectal function of children with neurological problems. I: Spina bifida. *Dev Med Child Neurol.* 1993; 35(10): 893–902. DOI: 10.1111/j.1469-8749.1993.tb11566.x.
24. Nejat F., Radmanesh F., Ansari S. et al. Spina bifida occulta: is it a predictor of underlying spinal cord abnormality in patients with lower urinary tract dysfunction? *J Neurosurg Pediatr.* 2008; 1(2): 114–7. DOI: 10.3171/PED/2008/1/2/114.
25. Sawin K.J., Bellin M.H. Quality of life in individuals with spina bifida: a research update. *Dev Disabil Res Rev.* 2010; 16(1): 47–59. DOI: 10.1002/ddrr.96.
26. Bakaniene I., Prasauskiene A., Vaiciene-Magistris N. Health-related quality of life in children with myelomeningocele: a systematic review of the literature. *Child Care Health Dev.* 2016; 42(5): 625–43. DOI: 10.1111/cch.12367.
27. Choi E.K., Im Y.J., Han S.W. Bowel management and quality of life in children with spina bifida in South Korea. *Gastroenterol Nurs.* 2017; 40(3): 208–15. DOI: 10.1097/SGA.000000000000135.
28. Rocque B.G., Ralee' Bishop E., Scogin M.A. et al. Assessing health-related quality of life in children with spina bifida. *J Neurosurg Pediatr.* 2015; 15(2): 144–9. DOI: 10.3171/2014.10.
29. Burns A.S., St-Germain D., Connolly M. et al. Phenomenological study of neurogenic bowel from the perspective of individuals living with spinal cord injury. *Arch Phys Med Rehabil.* 2015; 96(1): 49–55. DOI: 10.1016/j.apmr.2014.07.417.
30. Lo Cascio C.M., Goetze O., Latshang T.D. Gastrointestinal Dysfunction in Patients with Duchenne Muscular Dystrophy. *PLoS ONE.* 2016; 11: e0163779. DOI: 10.1371/journal.pone.0163779.
31. Crockett C.D., Bertrand L.A., Cooper C.S. et al. Urologic and gastrointestinal symptoms in the dystroglycanopathies. *Neurology.* 2015; 84: 532–9. DOI: 10.1212/WNL.0000000000001213.
32. Yadak R., Breur M., Bugiani M. Gastrointestinal Dysmotility in MNGIE: From thymidine phospho-

- rylase enzyme deficiency to altered interstitial cells of Cajal. *Orphanet J. Rare Dis.* 2019; 14: 33. DOI: 10.1186/s13023-019-1016-6.
33. Barrett T.G., Scott-Brown M., Seller A. et al. The mitochondrial genome in Wolfram syndrome. *J. Med. Genet.* 2000; 37: 463–6. DOI: 10.1136/jmg.37.6.463.
34. Chiminello R., Mosiello G., Castelli E. Bladder and bowel dysfunctions in children with cerebral palsy and acquired brain injury: Are we able to define incidence and to predict them? *Eur. Urol. Suppl.* 2018; 17: e1179–e1180. DOI: 10.1016/S1569-9056(18)31662-2.
35. Berger M., Heinrich M., Lacher M. et al. Postoperative bladder and rectal function in children with sacro-coccygeal teratoma. *Pediatr. Blood Cancer.* 2011; 56: 397–402. DOI: 10.1002/pbc.22887.
36. Mosiello G., Gatti C., De Gennaro M. et al. Neurovesical dysfunction in children after treating pelvic neoplasms. *BJU Int.* 2003; 92: 289–92. DOI: 10.1046/j.1464-410X.2003.04326.x.
37. Ng C., Prott G., Rutkowski S. et al. Gastrointestinal symptoms in spinal cord injury: Relationships with level of injury and psychologic factors. *Dis. Colon Rectum.* 2005; 48: 1562–8. DOI: 10.1007/s10350-005-0061-5.
38. Silveri M., Salsano L., Pierro M.M. et al. Pediatric spinal cord injury: Approach for urological rehabilitation and treatment. *J. Pediatr. Urol.* 2006; 2: 10–5. DOI: 10.1016/j.jpuro.2005.05.004.
39. Rasmussen M.M., Krogh K., Clemmensen D. et al. Colorectal transport during defecation in subjects with supraconal spinal cord injury. *Spinal Cord.* 2013; 51(9): 683–7. DOI: 10.1038/sc.2013.58.
40. Valles M., Vidal J., Clave P., Mearin F. Bowel dysfunction in patients with motor complete spinal cord injury: Clinical, neurological, and pathophysiological associations. *Am. J. Gastroenterol.* 2006; 101: 2290–9. DOI: 10.1111/j.1572-0241.2006.00729.x.
41. Qi Z., Middleton J.W., Malcolm A. Bowel Dysfunction in Spinal Cord Injury. *Curr. Gastroenterol. Rep.* 2018; 20: 47. DOI: 10.1007/s11894-018-0655-4.
42. Pidcock F.S., Krishnan C., Crawford T.O. et al. Acute transverse myelitis in childhood: Center-based analysis of 47 cases. *Neurology.* 2007; 68: 1474–80. DOI: 10.1212/01.wnl.0000260609.11357.6f.
43. Wolf V.L., Lupo P.J., Lotze T.E. Pediatric transverse myelitis. *J. Child Neurol.* 2012; 27: 1426–36. DOI: 10.1177/0883073812452916.
44. Kingwell E., Marriott J.J., Jetté N. et al. Incidence and prevalence of multiple sclerosis in Europe: a systematic review. *BMC Neurol.* 2013; 13: 128. DOI: 10.1186/1471-2377-13-128.
45. Evans C., Beland S.G., Kulaga S. et al. Incidence and prevalence of multiple sclerosis in the Americas: a systematic review. *Neuroepidemiology.* 2013; 40(3): 195–210. DOI: 10.1159/000342779.
46. Mah J., Thannhauser J.E. Management of multiple sclerosis in adolescents — current treatment options and related adherence issues. *Adolesc Health Med Ther.* 2010; 1: 31–43. <https://doi.org/10.2147/AHMT.S7594>.
47. Kuntz N.L., Chabas D., Weinstock-Guttman B. et al. Network of US Pediatric Multiple Sclerosis Centers. Treatment of multiple sclerosis in children and adolescents. *Expert Opin Pharmacother.* 2010; 11(4): 505–20. DOI: 10.1517/14656560903527218.
48. Munteis E., Andreu M., Téllez M.J. et al. Anorectal dysfunction in multiple sclerosis. *Mult Scler.* 2006; 12(2): 215–8. DOI: 10.1191/135248506ms1254oa.
49. Sawai S., Sakakibara R., Uchiyama T. et al. Acute motor axonal neuropathy presenting with bowel, bladder, and erectile dysfunction. *J. Neurol.* 2007; 254: 250–2. DOI: 10.1007/s00415-006-0336-3.
50. Zochodne D.W. Autonomic involvement in Guillain-Barre syndrome: A review. *Muscle Nerve.* 1994; 17: 1145–55. DOI: 10.1002/mus.880171004.
51. McCarthy M.J., Aylott C.E., Grevitt M.P., Hegarty J. Cauda equina syndrome: Factors affecting long-term functional and sphincteric outcome. *Spine.* 2007; 32: 207–16. DOI: 10.1097/01.brs.0000251750.20508.84.
52. Podnar S. Bowel dysfunction in patients with cauda equina lesions. *Eur. J. Neurol.* 2006; 13: 1112–7. DOI: 10.1111/j.1468-1331.2006.01423.x.
53. Podnar S. Epidemiology of cauda equina and conus medullaris lesions. *Muscle Nerve.* 2007; 35: 529–31. DOI: 10.1002/mus.20696.
54. McDermott C.J., Shaw P.J. Diagnosis and management of motor neurone disease. *BMJ.* 2008; 336: 658–62. DOI: 10.1136/bmj.39493.511759.BE.
55. Silveri M., De Gennaro M., Gatti C. et al. Voiding dysfunction in x-linked adrenoleukodystrophy: Symptom score and urodynamic findings. *J. Urol.* 2004; 171: 2651–3. DOI: 10.1097/01.ju.0000110885.26017.b0.
56. Kim M.Y., Kim J.H., Cho M.H. et al. Urological Problems in Patients with Menkes Disease. *J. Korean Med. Sci.* 2019; 34: e4. DOI: 10.3346/jkms.2019.34.e4.
57. Пак Л.А., Макарова С.Г., Фисенко А.П. и др. Опыт организации питания пациентов с детским церебральным параличом в ходе комплексной реабилитации на базе федерального центра. *Неврологический журнал имени Л.О. Бадаляна.* 2020; 1(2): 100–11. <https://doi.org/10.46563/2686-8997-2020-1-2-100-111>.
58. Афанасьева А.А., Чуйнышена С.А. Оценка физического развития и нутритивного статуса у детей с детским церебральным параличом,

- госпитализированных в многопрофильный педиатрический стационар. *Forcipe*. 2020. URL: <https://cyberleninka.ru/article/n/otsenka-fizicheskogo-razvitiya-i-nutritivnogo-statusa-u-detey-s-detskim-tserebralnym-paralichom-gospitalizirovannyh-v> (дата обращения: 19.05.2023).
59. Rendeli C., Paradiso V.F., Bucci V. et al. Gut microbiota and pediatric patients with spina bifida and neurogenic bowel dysfunction. *Childs Nerv Syst*. 2023; 39(3): 633–45. DOI: 10.1007/s00381-022-05688-0.
60. Furuta A., Suzuki Y., Takahashi R. et al. Effects of Transanal Irrigation on Gut Microbiota in Pediatric Patients with Spina Bifida. *J Clin Med*. 2021; 10(2): 224. DOI: 10.3390/jcm10020224.
61. Завьялова А.Н., Новикова В.П., Кликунова К.А. Юношеская саркопения у пациентов с детским церебральным параличом и предикторы ее развития. *Forcipe*. 2022; S2.
62. Цапина С.Ю. Проблема комплаентности семьи ребенка с ДЦП. *Вестник физиотерапии и курортологии*. 2015; 2 URL: <https://cyberleninka.ru/article/n/problema-komplaentnosti-semi-rebenka-s-dtsp> (дата обращения: 19.05.2023).
63. Бельмер С.В., Хавкин А.И., Печкуров Д.В. Функциональные запоры. *Медицинская сестра*. 2017; (7): 28–31.
64. Казюлин А.Н., Кучерявый Ю.А. Практические и клинические аспекты синдрома хронического запора. М.; 2005: 21.
65. Mosiello G., Safder S., Marshall D. et al. Neurogenic Bowel Dysfunction in Children and Adolescents. *J Clin Med*. 2021; 10(8): 1669. DOI: 10.3390/jcm10081669.
66. Kewalramani L.S. Autonomic dysreflexia in traumatic myelopathy. *Am. J. Phys. Med*. 1980; 59: 1–21.
67. Mosiello G., Marshall D., Rolle U. et al. Consensus review of best practice of transanal irrigation in children. *J. Pediatr. Gastroenterol. Nutr*. 2017; 64: 343–52. DOI: 10.1097/MPG.0000000000001483.
68. Коренюк Е.С., Иванченко В.И., Алексеева О.Г. Применение полуэлементных смесей в энтеральном питании детей с тяжелыми неврологическими заболеваниями. 2016; 5 (73): 83–5. DOI: 10.22141/2224-0551.5.1.73.1.2016.78947.
69. Комарова О.Н. Энтеральное питание у детей с хроническими заболеваниями: оптимальный способ доставки. *Медицинский Совет*. 2020; 18: 87–93. <https://doi.org/10.21518/2079-701X-2020-18-87-93>.
70. Приказ Министерства труда и социальной защиты РФ от 13 февраля 2018 г. № 86н «Об утверждении классификации технических средств реабилитации (изделий) в рамках федерального перечня реабилитационных мероприятий, технических средств реабилитации и услуг, предоставляемых инвалиду, утвержденного распоряжением Правительства Российской Федерации от 30 декабря 2005 г. N 2347-р».
71. Turan N., Aşt T.A. The Effect of Abdominal Massage on Constipation and Quality of Life. *Gastroenterol. Nurs*. 2016; 39: 48–59. DOI: 10.1097/SGA.0000000000000202.
72. Wald A. Constipation in the primary care setting: Current concepts and misconceptions. *Am. J. Med*. 2006; 119: 736–9. DOI: 10.1016/j.amjmed.2006.03.025.
73. Shafik A. Recto-colic reflex: Role in the defecation mechanism. *Int. Surg*. 1996; 81: 292–4.
74. Veiga M.L., Lordêlo P., Farias T., Barroso U., Jr. Evaluation of constipation after parasacral transcutaneous electrical nerve stimulation in children with lower urinary tract dysfunction — A pilot study. *J. Pediatr. Urol*. 2013; 9: 622–6. DOI: 10.1016/j.jpuro.2012.06.006.
75. Mentis B.B., Yüksel O., Aydin A. et al. Posterior tibial nerve stimulation for faecal incontinence after partial spinal injury: Preliminary report. *Tech. Coloproctol*. 2007; 11: 115–9. DOI: 10.1007/s10151-007-0340-3.
76. Patel S., Hopson P., Bornstein J., Safder S. Impact of Transanal Irrigation Device in the Management of Children With Fecal Incontinence and Constipation. *J Pediatr Gastroenterol Nutr*. 2020; 71(3): 292–7. DOI: 10.1097/MPG.0000000000002785.
77. Beierwaltes P., Church P., Gordon T., Ambartsumyan L. Bowel function and care: Guidelines for the care of people with spina bifida. *J Pediatr Rehabil Med*. 2020; 13(4): 491–8. DOI: 10.3233/PRM-200724.