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ЕЖЕМЕСЯЧНЫЙ НАУЧНЫЙ ЖУРНАЛ

Медицинские новости Грузии

საქართველოს სამედიცინო სიახლენი

GEORGIAN MEDICAL NEWS

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GMN: Georgian Medical News is peer-reviewed, published monthly journal committed to promoting the science and art of medicine and the betterment of public health, published by the GMN Editorial Board since 1994. GMN carries original scientific articles on medicine, biology and pharmacy, which are of experimental, theoretical and practical character; publishes original research, reviews, commentaries, editorials, essays, medical news, and correspondence in English and Russian.

GMN is indexed in MEDLINE, SCOPUS, PubMed and VINITI Russian Academy of Sciences. The full text content is available through EBSCO databases.

GMN: Медицинские новости Грузии - ежемесячный рецензируемый научный журнал, издаётся Редакционной коллегией с 1994 года на русском и английском языках в целях поддержки медицинской науки и улучшения здравоохранения. В журнале публикуются оригинальные научные статьи в области медицины, биологии и фармации, статьи обзорного характера, научные сообщения, новости медицины и здравоохранения. Журнал индексируется в MEDLINE, отражён в базе данных SCOPUS, PubMed и ВИНИТИ РАН. Полнотекстовые статьи журнала доступны через БД EBSCO.

GMN: Georgian Medical News – საქართველოს სამედიცინო სიახლენი – არის ყოველთვიური სამეცნიერო სამედიცინო რეცენზირებადი ჟურნალი, გამოიცემა 1994 წლიდან, წარმოადგენს სარედაქციო კოლეგიისა და აშშ-ის მეცნიერების, განათლების, ინდუსტრიის, ხელოვნებისა და ბუნებისმეტყველების საერთაშორისო აკადემიის ერთობლივ გამოცემას. GMN-ში რუსულ და ინგლისურ ენებზე ქვეყნდება ექსპერიმენტული, თეორიული და პრაქტიკული ხასიათის ორიგინალური სამეცნიერო სტატიები მედიცინის, ბიოლოგიისა და ფარმაციის სფეროში, მიმოხილვითი ხასიათის სტატიები.

ჟურნალი ინდექსირებულია MEDLINE-ის საერთაშორისო სისტემაში, ასახულია SCOPUS-ის, PubMed-ის და ВИНИТИ РАН-ის მონაცემთა ბაზებში. სტატიების სრული ტექსტი ხელმისაწვდომია EBSCO-ს მონაცემთა ბაზებიდან.

WEBSITE

www.geomednews.com

К СВЕДЕНИЮ АВТОРОВ!

При направлении статьи в редакцию необходимо соблюдать следующие правила:

1. Статья должна быть представлена в двух экземплярах, на русском или английском языках, напечатанная через **полтора интервала на одной стороне стандартного листа с шириной левого поля в три сантиметра**. Используемый компьютерный шрифт для текста на русском и английском языках - **Times New Roman** (**Кириллица**), для текста на грузинском языке следует использовать **AcadNusx.** Размер шрифта - **12**. К рукописи, напечатанной на компьютере, должен быть приложен CD со статьей.

2. Размер статьи должен быть не менее десяти и не более двадцати страниц машинописи, включая указатель литературы и резюме на английском, русском и грузинском языках.

3. В статье должны быть освещены актуальность данного материала, методы и результаты исследования и их обсуждение.

При представлении в печать научных экспериментальных работ авторы должны указывать вид и количество экспериментальных животных, применявшиеся методы обезболивания и усыпления (в ходе острых опытов).

4. К статье должны быть приложены краткое (на полстраницы) резюме на английском, русском и грузинском языках (включающее следующие разделы: цель исследования, материал и методы, результаты и заключение) и список ключевых слов (key words).

5. Таблицы необходимо представлять в печатной форме. Фотокопии не принимаются. **Все цифровые, итоговые и процентные данные в таблицах должны соответствовать таковым в тексте статьи**. Таблицы и графики должны быть озаглавлены.

6. Фотографии должны быть контрастными, фотокопии с рентгенограмм - в позитивном изображении. Рисунки, чертежи и диаграммы следует озаглавить, пронумеровать и вставить в соответствующее место текста **в tiff формате**.

В подписях к микрофотографиям следует указывать степень увеличения через окуляр или объектив и метод окраски или импрегнации срезов.

7. Фамилии отечественных авторов приводятся в оригинальной транскрипции.

8. При оформлении и направлении статей в журнал МНГ просим авторов соблюдать правила, изложенные в «Единых требованиях к рукописям, представляемым в биомедицинские журналы», принятых Международным комитетом редакторов медицинских журналов - http://www.spinesurgery.ru/files/publish.pdf и http://www.nlm.nih.gov/bsd/uniform_requirements.html
В конце каждой оригинальной статьи приводится библиографический список. В список литературы включаются все материалы, на которые имеются ссылки в тексте. Список составляется в алфавитном порядке и нумеруется. Литературный источник приводится на языке оригинала. В списке литературы сначала приводятся работы, написанные знаками грузинского алфавита, затем кириллицей и латиницей. Ссылки на цитируемые работы в тексте статьи даются в квадратных скобках в виде номера, соответствующего номеру данной работы в списке литературы. Большинство цитированных источников должны быть за последние 5-7 лет.

9. Для получения права на публикацию статья должна иметь от руководителя работы или учреждения визу и сопроводительное отношение, написанные или напечатанные на бланке и заверенные подписью и печатью.

10. В конце статьи должны быть подписи всех авторов, полностью приведены их фамилии, имена и отчества, указаны служебный и домашний номера телефонов и адреса или иные координаты. Количество авторов (соавторов) не должно превышать пяти человек.

11. Редакция оставляет за собой право сокращать и исправлять статьи. Корректура авторам не высылается, вся работа и сверка проводится по авторскому оригиналу.

12. Недопустимо направление в редакцию работ, представленных к печати в иных издательствах или опубликованных в других изданиях.

При нарушении указанных правил статьи не рассматриваются.

REQUIREMENTS

Please note, materials submitted to the Editorial Office Staff are supposed to meet the following requirements:

1. Articles must be provided with a double copy, in English or Russian languages and typed or compu-ter-printed on a single side of standard typing paper, with the left margin of **3** centimeters width, and **1.5** spacing between the lines, typeface - **Times New Roman** (**Cyrillic**), print size - **12** (referring to Georgian and Russian materials). With computer-printed texts please enclose a CD carrying the same file titled with Latin symbols.

2. Size of the article, including index and resume in English, Russian and Georgian languages must be at least 10 pages and not exceed the limit of 20 pages of typed or computer-printed text.

3. Submitted material must include a coverage of a topical subject, research methods, results, and review.

Authors of the scientific-research works must indicate the number of experimental biological species drawn in, list the employed methods of anesthetization and soporific means used during acute tests.

4. Articles must have a short (half page) abstract in English, Russian and Georgian (including the following sections: aim of study, material and methods, results and conclusions) and a list of key words.

5. Tables must be presented in an original typed or computer-printed form, instead of a photocopied version. **Numbers, totals, percentile data on the tables must coincide with those in the texts of the articles.** Tables and graphs must be headed.

6. Photographs are required to be contrasted and must be submitted with doubles. Please number each photograph with a pencil on its back, indicate author's name, title of the article (short version), and mark out its top and bottom parts. Drawings must be accurate, drafts and diagrams drawn in Indian ink (or black ink). Photocopies of the X-ray photographs must be presented in a positive image in **tiff format**.

Accurately numbered subtitles for each illustration must be listed on a separate sheet of paper. In the subtitles for the microphotographs please indicate the ocular and objective lens magnification power, method of coloring or impregnation of the microscopic sections (preparations).

7. Please indicate last names, first and middle initials of the native authors, present names and initials of the foreign authors in the transcription of the original language, enclose in parenthesis corresponding number under which the author is listed in the reference materials.

8. Please follow guidance offered to authors by The International Committee of Medical Journal Editors guidance in its Uniform Requirements for Manuscripts Submitted to Biomedical Journals publication available online at: http://www.nlm.nih.gov/bsd/uniform_requirements.html
http://www.icmje.org/urm_full.pdf
In GMN style for each work cited in the text, a bibliographic reference is given, and this is located at the end of the article under the title "References". All references cited in the text must be listed. The list of references should be arranged alphabetically and then numbered. References are numbered in the text [numbers in square brackets] and in the reference list and numbers are repeated throughout the text as needed. The bibliographic description is given in the language of publication (citations in Georgian script are followed by Cyrillic and Latin).

9. To obtain the rights of publication articles must be accompanied by a visa from the project instructor or the establishment, where the work has been performed, and a reference letter, both written or typed on a special signed form, certified by a stamp or a seal.

10. Articles must be signed by all of the authors at the end, and they must be provided with a list of full names, office and home phone numbers and addresses or other non-office locations where the authors could be reached. The number of the authors (co-authors) must not exceed the limit of 5 people.

11. Editorial Staff reserves the rights to cut down in size and correct the articles. Proof-sheets are not sent out to the authors. The entire editorial and collation work is performed according to the author's original text.

12. Sending in the works that have already been assigned to the press by other Editorial Staffs or have been printed by other publishers is not permissible.

Articles that Fail to Meet the Aforementioned Requirements are not Assigned to be Reviewed.

ავტორთა საყურადღებოდ!

რედაქციაში სტატიის წარმოდგენისას საჭიროა დავიცვათ შემდეგი წესები:

1. სტატია უნდა წარმოადგინოთ 2 ცალად, რუსულ ან ინგლისურ ენებზე,დაბეჭდილი სტანდარტული ფურცლის 1 გვერდზე, 3 სმ სიგანის მარცხენა ველისა და სტრიქონებს შორის 1,5 ინტერვალის დაცვით. გამოყენებული კომპიუტერული შრიფტი რუსულ და ინგლისურენოვან ტექსტებში - **Times New Roman (Кириллица)**, ხოლო ქართულენოვან ტექსტში საჭიროა გამოვიყენოთ **AcadNusx.** შრიფტის ზომა – 12. სტატიას თან უნდა ახლდეს CD სტატიით.

2. სტატიის მოცულობა არ უნდა შეადგენდეს 10 გვერდზე ნაკლებს და 20 გვერდზე მეტს ლიტერატურის სიის და რეზიუმეების (ინგლისურ, რუსულ და ქართულ ენებზე) ჩათვლით.

3. სტატიაში საჭიროა გაშუქდეს: საკითხის აქტუალობა; კვლევის მიზანი; საკვლევი მასალა და გამოყენებული მეთოდები; მიღებული შედეგები და მათი განსჯა. ექსპერიმენტული ხასიათის სტატიების წარმოდგენისას ავტორებმა უნდა მიუთითონ საექსპერიმენტო ცხოველების სახეობა და რაოდენობა; გაუტკივარებისა და დაძინების მეთოდები (მწვავე ცდების პირობებში).

4. სტატიას თან უნდა ახლდეს რეზიუმე ინგლისურ, რუსულ და ქართულ ენებზე არანაკლებ ნახევარი გვერდის მოცულობისა (სათაურის, ავტორების, დაწესებულების მითითებით და უნდა შეიცავდეს შემდეგ განყოფილებებს: მიზანი, მასალა და მეთოდები, შედეგები და დასკვნები; ტექსტუალური ნაწილი არ უნდა იყოს 15 სტრიქონზე ნაკლები) და საკვანძო სიტყვების ჩამონათვალი (key words).

5. ცხრილები საჭიროა წარმოადგინოთ ნაბეჭდი სახით. ყველა ციფრული, შემაჯამებელი და პროცენტული მონაცემები უნდა შეესაბამებოდეს ტექსტში მოყვანილს.

6. ფოტოსურათები უნდა იყოს კონტრასტული; სურათები, ნახაზები, დიაგრამები - დასათაურებული, დანომრილი და სათანადო ადგილას ჩასმული. რენტგენოგრამების ფოტოასლები წარმოადგინეთ პოზიტიური გამოსახულებით **tiff** ფორმატში. მიკროფოტოსურათების წარწერებში საჭიროა მიუთითოთ ოკულარის ან ობიექტივის საშუალებით გადიდების ხარისხი, ანათალების შეღებვის ან იმპრეგნაციის მეთოდი და აღნიშნოთ სურათის ზედა და ქვედა ნაწილები.

7. სამამულო ავტორების გვარები სტატიაში აღინიშნება ინიციალების თანდართვით, უცხოურისა – უცხოური ტრანსკრიპციით.

8. სტატიას თან უნდა ახლდეს ავტორის მიერ გამოყენებული სამამულო და უცხოური შრომების ბიბლიოგრაფიული სია (ბოლო 5-8 წლის სიღრმით). ანბანური წყობით წარმოდგენილ ბიბლიოგრაფიულ სიაში მიუთითეთ ჯერ სამამულო, შემდეგ უცხოელი ავტორები (გვარი, ინიციალები, სტატიის სათაური, ჟურნალის დასახელება, გამოცემის ადგილი, წელი, ჟურნალის №, პირველი და ბოლო გვერდები). მონოგრაფიის შემთხვევაში მიუთითეთ გამოცემის წელი, ადგილი და გვერდების საერთო რაოდენობა. ტექსტში კვადრატულ ფჩხილებში უნდა მიუთითოთ ავტორის შესაბამისი N ლიტერატურის სიის მიხედვით. მიზანშეწონილია, რომ ციტირებული წყაროების უმეტესი ნაწილი იყოს 5-6 წლის სიღრმის.

9. სტატიას თან უნდა ახლდეს: ა) დაწესებულების ან სამეცნიერო ხელმძღვანელის წარდგინება, დამოწმებული ხელმოწერითა და ბეჭდით; ბ) დარგის სპეციალისტის დამოწმებული რეცენზია, რომელშიც მითითებული იქნება საკითხის აქტუალობა, მასალის საკმაობა, მეთოდის სანდოობა, შედეგების სამეცნიერო-პრაქტიკული მნიშვნელობა.

10. სტატიის ბოლოს საჭიროა ყველა ავტორის ხელმოწერა, რომელთა რაოდენობა არ უნდა აღემატებოდეს 5-ს.

11. რედაქცია იტოვებს უფლებას შეასწოროს სტატია. ტექსტზე მუშაობა და შეჯერება ხდება საავტორო ორიგინალის მიხედვით.

12. დაუშვებელია რედაქციაში ისეთი სტატიის წარდგენა, რომელიც დასაბეჭდად წარდგენილი იყო სხვა რედაქციაში ან გამოქვეყნებული იყო სხვა გამოცემებში.

აღნიშნული წესების დარღვევის შემთხვევაში სტატიები არ განიხილება.

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CLINICAL FEATURES OF THE COURSE OF HIRSCHSPRING'S DISEASE INCHILDREN OF THE FIRST YEAR

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Abstract.

The purpose of the work: To investigate the peculiarities of the clinical course of Hirschsprung's disease in children of the first year of life and to determine the significance of symptoms in the verification of the disease.

Research materials and methods: From 1980 to 2021, at the pediatric surgery clinic of the National Medical University named after O.O. Bogomolets on the basis of the National Children's Specialized Hospital "OKHMATDYT" and in the pediatric surgery clinic of the Ivano-Frankivsk National Medical University on the basis of the Ivano-Frankivsk Regional Children's Clinical Hospital, 483 children of the first year of life suffering from Hirschsprung's disease were examined and treated.

Results: The clinical manifestation and course of aganglionosis varied in length at the time of hospitalization and depended on the time after birth. During the first month of life, 97 (20.08%) patients were hospitalized, of which 39 (8.07%) hadatypical clinical picture due to: colonic atresia in 15 (3.10%), colonic atresia + gastroschisis in 3 (0.62%), ileal atresia in 9 (1.86%), esophageal atresia in 3 (0 .62%), clefts of the hard and soft palate in 9 (1.86%). Depending on the age, there were 280 (57.97%) patients under 6 months, and 203 (42.03%) patients between 6 months and 1 year. The classic typical clinical picture was in 444 (91.93%) patients, which was characterized by the absence of meconium excretion, abdominal distension in 444 (91.93%), delayed physiological weight gain against the background of nutritional insufficiency with the development of hypotrophy in 327 (67.70%) , vomiting of stagnant gastric and intestinal contents in 417 (86.34%). On the other hand, enterocolitis in 315 (65.22%), toxic megacolon in 16 (3.31%), and anemia of various degrees occurred in 241 (49.89%) patients among the complications that arose during the examination of patients with Hirschsprung's disease. According to the results of a comprehensive examination, the following extent of aganglionosis was established: rectal in 100 (20.70%), rectosigmoid in 192 (39.75%), subtotal in 150 (31.06%) and total in 41 (8.49%) patients. Concomitant malformations were found in 98 (20.29%) patients: renal malformations were diagnosed in 7 (1.45%) patients, concomitant heart malformations in 18 (3.73%) patients. Associated intraoperative findings were Meckel's diverticulum in 5 (1.03%) and congenital cyst of the right ovary in 1 (0.21%) patient. The clinical course was affected by concomitant malformations: incomplete bowel rotation in 10 (2.07%) and internal abdominal hernia in 2 (0.42%).

Conclusions: The clinical manifestations and course of Hirschsprung's disease primarily depend on the presence of accompanying developmental defects, which may prevail during the examination due to vital disorders. In the clinical course of Hirschsprung's disease, it is necessary to distinguish between typical and atypical forms. Typical clinical symptoms were in 444 (91.93%), and atypical in 39 (8.07%).

Key words. Hirschsprung's disease, infants, clinical course, enterocolitis, anemia, hypotrophy, accompanying malformations.

Introduction.

Hirschsprung's disease (HG) or intestinal aganglionosis (AK) belongs to the group of severe malformations and is usually diagnosed shortly after delivery due to the absence of spontaneous excretion of meconium within 24 hours. In addition, the main clinical manifestations include abdominal distension, vomiting of stagnant gastric and intestinal contents, and as a result, the development of enteric insufficiency and hypotrophy [1-17,3]. The degree of aganglionosis varies from the lesion of a short segment covering the rectum and sigmoid colon (75%-80% of cases) to total aganglionosis of the colon (5%-7% of cases). From 10% to 15% of cases, which are called long segment disease, reveal aganglionosis proximal to the sigmoid colon [2,9].

Based on data from the literature and personal experience, the existence of various forms of CG and clinical variants of its manifestation leads to the fact that some patients are not diagnosed with the disease in the newborn period, until the development of complications [1,2,3,16]. This may be due to the lack of surgical vigilance among neonatologists in maternity hospitals and newborn departments, as well as incorrect interpretation of clinical and radiological symptoms [2,3,7,12].

In some patients, up to 15-20%, according to various researchers, AK occurs against the background of concomitant malformations, including those of the alimentary canal, which significantly affect the clinical manifestation of CG [10].

The most typical and severe complication of CG is enterocolitis (EC) and toxic megacolon, which is diagnosed from 57% to 68%, depending on the length of aganglionosis and the patient's condition at the time of hospitalization [9,15]. Meanwhile, with untimely diagnosis and inadequate medical measures, EC and toxic megacolon are the key to the development of sepsis, organ, and multi-organ failure with fatal consequences, which according to various data currently range from 5% to 50% [4,8,9,11,15].

Thus, after considerable development, and to this day, timely diagnosis, and verification of HG, taking into account the length of the aganglionic area and the course of the disease against the background of accompanying developmental defects, as well as the presence of EC, determine the relevance of this study.

The purpose of the work. To investigate the peculiarities of the clinical course of HCG in children of the first year of life and

to determine the significance of symptoms in the verification of the disease.

Materials and Methods.

From 1980 to 2021, at the pediatric surgery clinic of the National Medical University named after O.O. Bogomolets on the basis of the National Children's Specialized Hospital "OKHMATDYT" and in the pediatric surgery clinic of the Ivano-Frankivsk National Medical University on the basis of the Ivano-Frankivsk Regional Children's Clinical Hospital, 483 children of the first year of life who suffered from CKD were examined and treated. Based on the results of the anamnesis and physical examination, a general clinical laboratory examination of blood and urine, coprological and microbiological examination of feces, ECG, NSG, echocardiography, ultrasound of the abdominal cavity and retroperitoneal space, X-ray of the organs of the abdominal and chest cavities were performed.

Results.

When studying the features of the clinical picture of the course of various forms of AK in children of the first year of life, first of all, the family status was investigated. Careful study of the anamnesis established that 436 (90.27%) families were complete, and 47 (9.73%) were incomplete. The age of women who gave birth after 40 years was determined to be 87 (18.01%). The first child in the family was in 196 (40.58%), the second in 162 (33.54%), the third in 107 (22.15%) and the fourth or more in 18 (3.73%). In 36 (7.45%) cases, they were used to conceive a child assisted reproductive technology (ART). Harmful working conditions (chemical industry and others) occurred in 185 (38.30) women giving birth, and 93 (19.25%) pregnant women lived in environmentally polluted areas with family succession (parents and children - brothers and sisters) of HG was established in 24 families, which is 4.97%.

The study of the course of pregnancy showed that in 94 (19.46%) cases, the pregnancy occurred against the background of amniotic fluid, and in 76 (15.73%) - polyhydramnios. The course of pregnancy was monitored: anemia of various degrees - in 204 (42.24%) pregnant women, acute respiratory viral infection in various stages of pregnancy - 182 (37.68%) observations, preeclampsia of various degrees - 97 (20.08%) observations, urogenital infection - 72 (14 .91%) of observations, bleeding in different stages of pregnancy - 35 (7.25%) observations. Natural childbirth in 429 (88.82%), caesarean section - in 54 (11.18%). According to the term of gestation, 471 (97.51%) patients were full-term, and 12 (2.49%) patients were premature.

The clinical manifestation and course of aganglionosis varied in length at the time of hospitalization, to some extent depended on the time of hospitalization after birth, and when the diagnosis was established. Table No. 1 shows the term of hospitalization of children in surgical clinics. Research has established that 58 (12.00%) patients were referred by neonatologists from the maternity hospital, 26 (5.38%) from neonatal pathology departments, 116 (24.02%) by pediatricians and family doctors, 138 (28.57%) by pediatric by surgeons (after ineffective treatment), and 145 (30.03%) applied without referrals. Meanwhile, the presented monthly hospitalization of patients does not indicate the state of the problem in the region but indicates the state of awareness of parents and doctors with the identified problem, as 257 (53.21%) patients lived in different regions of Ukraine. Research has established that on the first and second days after birth, 39 (8.07%) patients with212 (43.89%) patients who were hospitalized for up to 6 months. The main reasons for hospitalization of children were accompanying malformations: esophageal atresia in 3 (0.62%), gastroschisis and intestinal atresia in 3 (0.62%), colon and ileal atresia in 24 (4.96%), and cleft of the dura and soft palate in 9 (1.86%) patients, whose clinical course prevailed over aganglionosis. During the first and second week of life, 31 (6.42%) patients were hospitalized for examination (on the referral of neonatal pathology departments) due to lack of emptying. At 3-4 weeks of life, 27 (5.59%) patients out of 212 (43.89%) were hospitalized for examination with complaints of lack of emptying and coagulated milk syndrome, of which 12 (2.48%) had pylorostenosis confirmed, and 15 (3.11%) had pylorospasm. Thus, 39 (8.07%) were hospitalized in the second month, 43 (8.90%) in the third, 35 (7.25%) in the fourth, 34 (7.04%) in the fifth, and 32 (6.62%) in the sixth month of life. patients Table No. 1 shows the terms of hospitalization of children in the first year of life, depending on the extent of aganglionosis.

The clinical course of different forms of aganglionosis depended to a certain extent, and sometimes to a large extent, on the presence of concomitant developmental defects. Table No. 2 presents the quantitative characteristics of patients with intestinal aganglionosis who had concomitant congenital malformations. Despite the fact that all patients had a delay in the passage of meconium for 48 hours or more, and 30 (6.2%) patients had accompanying malformations - esophageal atresia in 3 (0.62%) and intestinal atresia in 27 (5.58%), manifested by clinical aspiration syndrome and high intestinal obstruction. At the same time, 9 (1.86%) patients had problems with nutrition associated with various types of clefts of the hard and soft palate. Kidney malformations were diagnosed in 7 (1.45%) patients, including hydronephrosis in 5 (1.03%) and other defects: hypoplasia of the right kidney and polycystic left kidney in 2 (0.42%). Concomitant malformations of the heart: (atrial septal defect, gastric septal defect, open oval window, and others) were found in 18 (3.73%) patients. To some extent, the manifestation of clinical symptoms was caused by the presence of microcephaly in 4 (0.83%) patients, which was manifested by impaired swallowing and coughing during eating. Associated intraoperative findings were Meckel's diverticulum in 5 (1.03%) and congenital cyst of the right ovary in 1 (0.21%) patient. The clinical course of various forms of aganglionosis at the examination stages was affected by concomitant malformations, namely: incomplete bowel rotation in 10 (2.07%) and internal abdominal hernia in 2 (0.42%).

Studying the clinical course of the disease, the dependence of the development of clinical manifestations of the developmental defect on the extent of aganglionosis and the development of complications was established. Table No. 3 presents the quantitative characteristics of patients, in groups of the extent of aganglionosis and the development of clinical manifestations of both the disease and its complications.

Table 1. *The term of hospitalization of children after birth to clinics.*

The extent of aganglionosis / Age of patients	In total (n, %)	Rectal-on (n, %)	Rectosig-movidna (n, %)	Subtotal (n, %)	Total on (n, %)
1-2 days	39*	-	2	27	10
1st month	58	5	20	22	11
2nd month	39	3	10	12	14
3rd month	43	1	17	19	6
4th month	35	1	11	23	-
5th month	34	1	15	18	-
6th month	32	3	17	12	-
7th month	43	8	31	4	-
8th month	41	11	26	4	-
9th month	28	13	12	3	-
10th month	21	7	12	2	-
11th month	31	18	10	3	-
12th month	39	29	9	1	-
Together:	483 (100%)	100 (20.70%)	192 (39.75%)	150 (31.06%)	41 (8.49%)

* - *accompanying malformations that prevailed in the clinical course.*

Table 2. *Concomitant congenital malformations in patients with intestinal aganglionosis.*

No	Associated congenital malformations	Number	%
1.	Meckel's diverticulum	5	1.03
2.	Congenital heart defects	18	3.73
3.	Cleft of the hard and soft palate	9	1.86
4.	Pylorostenosis	12	2.48
5.	Microcephaly	4	0.83
6.	Hydronephrosis	5	1.03
7.	Other kidney malformations	2	0.42
8.	Atresia of the colon	15	3.10
9.	Colonic atresia + gastroschisis	3	0.62
10.	Atresia of the ileum	9	1.86
11.	Esophageal atresia	3	0.62
12.	Internal abdominal hernia	2	0.42
13.	Incomplete bowel movement	10	2.07
14.	Ovarian cyst	1	0.21
	Total:	98	20.29%

Table 3. *Dependence on the volume of aganglionosis lesions and the development of clinical manifestations ailments and complications.*

No n\n	Clinical manifestation of the disease	Age of patients								Together
		0-6 months (n, %)				6 - 12 months (n, %)				
		1	2	3	4	1	2	3	4	
1.	Absence of the first emptying more than 24 hours after birth	39	98	102	41	61	94	48	-	483
2.	Abdominal distension to contour loops of the intestine on the front abdominal wall	39 -	87 *11	86 *16	29 *12	61 -	94 -	48 -	- -	444 *39
3.	Delay in physiological weight gain against the background of alimentary insufficiency (hypotrophy).	34	45	85	41	20	41	61	-	327
4.	Vomiting masses at the beginning of the illness with stagnant contents, over time green, and with prolonged vomiting brown	39 **1 ***3	87 **2 ***3	86 **5 ***4	29 **4 ***5	61 - -	94 - -	48 - -	- - -	417 **12 ***15
5.	There are no physiological emptyings, and when the tube is inserted into the anus, there is an explosive emptying up to diarrhea.	18	50	-	-	51	49	-		168
6.	EC and toxic megacolon	21 -	48 -	102 -	41 -	10 -	45 7	48 9	- -	315 16
7.	Anemia of various degrees	8	29	102	41	4	9	48		241
	Total by groups:	280 (57.97%)				203 (42.03%)				
1 - rectal form of aganglionosis, 2 - rectosigmoid form of aganglionosis, 3 – subtotal form of agangiosis, 4 – total form of agangiosis.										
* - accompanying malformations, ** - pylorostenosis, *** - pylorospasm.										

Table 4. *Dependence of the extent of aganglionosis and EC development.*

The extent of aganglionosis	Total patients in the group (n=, %)	All who had enterocolitis (n=, %)	Of them	
			0 - 6 months	6 - 12 months
Rectal	100	31	21	10
Rectosigmoid	192	93	48	45
Subtotal	150	150	102	48
Total	41	41	41	-
Total:	483 (100%)	315 (65.22%)	212 (43.89%)	103 (21.33%)

Table 5. *Dependence of the severity of the course of aganglionosis and the development of anemia in patients.*

The extent of aganglionosis	Total patients in the group (n, %)	All that had anemia (n, %)	Among them, anemia		
			mild degree (n, %)	moderate severity (n, %)	severe (n, %)
Rectal	100	12	2	9	1
Rectosigmoid	192	38	3	31	4
Subtotal	150	150	13	121	16
Total	41	41	3	30	8
Total:	483 (100%)	241 (49.89%)	21 (4.35%)	191 (39.54%)	29 (6.00%)

Table 6. *Dependence of the extent of aganglionosis and the development of hypotrophy.*

Anatomical forms of aganglionosis	Total patients (%)	Degrees of hypotrophy (n, %)			
		1	2	3	In total
Rectal	100	**27**	**21**	**6**	54 (11.18%)
Rectosigmoid	192	**20**	**56**	**10**	86 (17.80%)
Subtotal	150	**19**	**112**	**15**	146 (30.23)
Total	41	**23**	**18**	**-**	41 (8.49)
Total:	483 (100%)	89 (18.42)	207 (42.86%)	31 (6.42%)	327 (67.70%)

Research has established that 39 (8.07%) of 280 (57.97%) patients in the first 6 months of life did not have abdominal distension before the contouring of the intestinal loops on the anterior abdominal wall due to concomitant malformations: colonic atresia in 15 (3 .10%), colonic atresia + gastroschisis in 3 (0.62%), ileal atresia in 9 (1.86%), esophageal atresia in 3 (0.62%), cleft of the hard and soft palate in 9 (1.86%), and therefore had an atypical clinical picture in relation to different forms of aganglionosis. On the other hand, 241 (49.89%) patients under 6 months and 203 (42.03%) - older than 6 months had a classic pattern of abdominal distension with contouring of intestinal loops on the anterior abdominal wall, and in the presence of hypotrophy, the contouring was even more pronounced. When analysing the nature of vomitus, 27 (5.59%) patients had vomited milk, among which - 12 (2.48%) children had pylorostenosis, and 15 (3.11%) had pylorospasm.

All patients had a delay in physiological weight gain against the background of nutritional insufficiency, but 327 (67.70%) had varying degrees of hypotrophy. The development of alimentary insufficiency and the development of hypotrophy were significantly influenced by concomitant diseases of the digestive tract in 63 (%) patients, of which 46 (9.53%) - in the first weeks of life, which required surgery according to favourable indications. In our clinical observations, a cleft of the hard and soft palate was established in 9 (1.86%) patients who, on the background of concomitant pathology, had swallowing disorders, and as a result, enteral insufficiency, which was corrected by tube feeding. Further violations of the passage through the intestines for the presence of stagnant contents in the stomach and constipation were the basis for conducting irrigography.

Studies have established that the appearance of stasis in the stomach due to the presence of vomiting was correlated with the extent of aganglionosis and the presence of concomitant malformations of the digestive tract in 63 (13.04%) patients. However, the appearance of this symptom depended in a certain way on the speed of filling the intestines with food and air. Vomiting masses at the beginning of the disease with stagnant stomach contents, with time when the stomach and duodenum are full - with impurities of bile, which changes to green color over time with stagnation. With a long-term violation of the passage - stagnation of brown color and thin and thick intestinal. It was found that 417 (86.34%) patients had stasis and pathological vomiting, and 27 (5.59%) had coagulated milk at the beginning, in which 12 (2.48%) had pylorostenosis and 15 (3.11) pylorospasm %).

Physiological bowel movements were absent in all 483 patients. In addition, 383 (79.30%) patients had a violation of the release of intestinal gases, which was associated with the extent of aganglionosis, of which 41 (8.49%) had a total form, 150 (30.23%) had a subtotal form, and rectosigmoid - 192 (17.80%) patients. On the other hand, in 100 (11.18%) patients who had a rectal form of aganglionosis, emptying (leakage) of feces and intestinal gases occurred against the background of total bowel filling in 37 (7.66%), and 63 (13.04%) required staging intestinal tube. However, over time (7-11 days) the

involuntary leakage became absent. Progression of abdominal distension and lack of emptying required placement of an intestinal tube, and when the tube was inserted into the anus, explosive emptying occurred up to diarrhea 168 (34.78%). On the other hand, the lack of emptying and stasis in the intestine served as the cause of EC in 315 (65.22%), of which 16 (3.31%) had toxic megacolon.

Usually, the normal body temperature in children of the first year of life can be extremely labile under the influence of such factors as mental or emotional excitement, physical exertion, sleep, the nature of clothing and the type of feeding. However, having an ailment - aganliosis of the intestine of varying length with a violation of the transit of the food bolus, and as a result, stagnation in the lumen of the intestine with the development of putrefactive processes and intra-intestinal hypertension leads to the translocation of toxins and microorganisms into the lymphatic and circulatory system, and as a result - the body's reaction in the form hyperthermia. Research has established the following indicators of body temperature in children, depending on the degree of complication of the illness with enterocolitis and accompanying developmental defects. So normal temperature (up to 37.0°C) was established in 168 (34.78%), subfebrile (37.1–38 C) in 142 (29.40%); moderate febrile (38.1–39 C) in 94 (19.46%); highly febrile (39.1–41 C) in 63 (13.04%); and hyperpyretic (more than 41°C) in 16 (3.31%) who had toxic megacolon. Normal and subfebrile temperature was established in 310 (64.18%) patients who underwent intestinal irrigation, and it was successful - it was possible to adequately wash out the intestinal contents. On the other hand, in 157 (32.50%) patients with moderate to high febrile temperature, irrigation at the first attempt had no effect and required further attempts, while intestinal distension progressed, and the digestion process turned into the process of putrefaction. In 16 (3.31%) patients, EC was complicated by toxic megacolon due to the fact that there was obturation of the narrowed aganglionic intestine, and irrigation and insertion of the tube above the narrowed aganglionic area had certain difficulties due to the presence of a long form of aganglionosis and required more effective tactics - imposing a colostomy behind urgent indications.

The course of malformations in 315 (65.22%) patients was complicated by EC, and in 16 (3.32%) - by toxic megacolon. Table No. 4 presents the quantitative characteristics of EC patients depending on the extent of aganglionosis. EC was confirmed clinically by swelling of the abdominal cavity; lack of emptying, and when the tube is placed, explosive emptying, up to diarrhea; hyperthermia, which indicated an inflammatory process, and when the gas outlet tube was placed and irrigation was performed, the temperature normalized; leukocytosis and anemia. Excretions had the character of vidyanist with a putrid smell, and during coprological examination the main indicators were leukocytes (32.53±2.18), erythrocytes (12.18±2.18) and mucus (in a significant amount). It is appropriate to point out that all patients had digestive disorders, and therefore had hypotrophy of various degrees, which was established in 327 (67.70%) patients.

Clinical examinations and monitoring of laboratory parameters during treatment revealed anemia in 241 (49.90%) of 483 patients. Table No. 5 presents quantitative characteristics of patients with different degrees of anemia and extent of aganglionosis. When determining the degree of anemia, the following indicators were used: mild anemia — hemoglobin 110–90 g/l, erythrocytes - up to 3.5×1012/l; moderate anemia — hemoglobin 90–70 g/l, erythrocytes - up to 2.5×1012/l; severe anemia — hemoglobin less than 70 g/l, erythrocytes less than 2.5×1012/l. Regarding leukocytosis, moderate leukocytosis (9-15×109/l) and high level of leukocytosis (15-50×109/l) were distinguished [17].

When interpreting the obtained indicators, we took into account the fact that 39 (8.07%) had accompanying malformations, and 30 (6.21%) patients had primary operations due to esophageal atresia in 3 (0.62%), gastroschisis with colonic atresia in 3 (0.62%) and ileal and colonic atresia in 24 (4.96%) patients.

The study of hemoglobin and erythrocyte indicators showed the following data: mild anemia in 21 (4.35%), moderate anemia in 191 (39.54%), severe anemia in 29 (6.00%). On the other hand, 242 (50.11%) hemoglobin and erythrocyte indicators were within the age norm. The degree of anemia in the study was correlated with the severity of the course of aganglionosis, or rather - with its length and EC development in 315 (65.22%), as well as against the background of various degrees of enteric insufficiency - hypotrophy in 327 (67.70%). The study of the number of leukocytes indicated the following, that the number of leukocytes was within the normal range in 168 (34.78%), moderate leukocytosis was established in 209 (43.27%) and a high level of leukocytosis in 106 (21.95%) patients. It was established that leukocytosis was also correlated with the severity of complications of aganglionosis - EC and toxic megacolon.

Clinical examinations and monitoring of physical data revealed hypotrophy of various degrees in 327 (67.70%). The first degree of hypotrophy included children with a body weight deficit of 10-20%, the second - 20-30%, and the third - more than 30%. A lag in body weight gains up to 10% was considered insignificant. The first degree of hypotrophy was found in 89 (18.42%) children, the second - in 207 (42.86%) and the third - in 31 (6.42%) patients. Table No. 6 presents quantitative characteristics of patients with different degrees of hypotrophy and extent of aganglionosis. Research has established that all 238 (49.28%) patients who had the second and third degree of hypotrophy in the clinical course and had enterocolitis and enteric insufficiency in 36 (7.45%) due to transferred operations for concomitant malformations that were corrected first with causes of welcome disorders. Thus, in the first two weeks of life, patients who underwent surgery for esophageal atresia in 3 (0.62%), small and large intestine atresia in 24 (4.96%), gastroschisis and colonic atresia had nutritional deficiency in the first two weeks of life. (0.62%), as well as internal hernia in 1 (0.21%) and incomplete bowel rotation in 6 (1.24%). In 12 (2.48%) patients, the cause of intestinal insufficiency was pylorostenosis. Meanwhile, in the second month of life, 5 (1.03%) patients had a clinic of small bowel obstruction, and the cause was internal hernia in 1 (0.21%) and incomplete bowel rotation in 4 (0.83%). It was established that among 54 (11.18%) patients with concomitant developmental defects and

who underwent operations on favourable indications against the background of the main developmental defect - intestinal aganglionosis, peristalsis was restored within 3-4 days in 14 (2.90%), during 5-7 days in 29 (6.00%), 10-12 days in 11 (2.28%), and parenteral nutrition was provided all this time.

So, clinical features of the course of CG in infants depends on the presence of concomitant malformations established in63 (13.04%) patients. In the clinical picture of the course of the disease, the main symptoms prevailed: the absence of meconium discharge and physiological emptying in 483 (100%), abdominal distension and vomiting with stagnant gastric and intestinal contents in 444 (91.93%), and in case of complications, the development of EC in 315 (65, 22%), toxic megacolon in 16 (3.31%) and hyperthermia (moderate and high febrile temperature) in 173 (35.82%). The consequences of the course of the disease are nutritional deficiency and hypotrophy in 327 (67.70%), anemia in 241 (49.89%).

It was established that the clinical course of CG differed in groups depending on the extent of aganglionosis. 64 (13.25%) patients with rectal form, 72 (14.91%) with rectosigmoid, 150 (31.05%) with subtotal and 41 (8.49%) with total form of aganglionosis had such an acute form of the disease. On the other hand, 36 (7.45%) patients with rectal and 120 (24.85%) with rectosigmoid form of aganglionosis had a subacute form of the disease.

Thus, according to the results of a comprehensive physical examination among 483 newborns with CG, the following extent of aganglionosis was established: rectal form in 100 (20.71%), rectosigmoid in 192 (39.75%), subtotal in 150 (31.05%) and total in 41 (8.49%) patients.

Discussion.

Own research correlates with data from review publications Gershon EM, Rodriguez L, Arbizu RA. (2023) and Philip K. Frykman, Scott S. Short (2012), that there is a predominance of male patients over female patients in a ratio of 4:1 [1,3,9,15]. The degree of aganglionosis varies from the lesion of a short segment covering the rectum and sigmoid colon (75%-80% of cases) to total aganglionosis of the colon (5%-7% of cases). From ten to 15% of cases, which are called long segment disease, reveal aganglionosis proximal to the sigmoid colon [9]. In our study, the extent of aganglionosis was as follows: rectal form - in 11.18%, rectosigmoid form - in 17.80%, subtotal form - in 30.23%, total form - in 8.49%.

Hirschsprung's disease as an isolated developmental defect is diagnosed in 70% of patients, and in about 30% it is associated with additional congenital anomalies in 18-20% and chromosomal anomalies in 10%-12% of cases. In a review publication, Jeanne Amiel & Stanislas Lyonnet (2023) indicate combined malformations of other organs and systems in 18% of patients, and in our study these indicators were 20.29% [10].

According to H. Lampus (2023) family transmission (parents and children - brothers and sisters) of HG ranges from 5% to 20%, and in our study, it was 4.97% in 24 families [13,15].

Despite considerable progress regarding complications in the course of CG, to this day EC associated with CG is a serious and potentially fatal complication. Thus, according to EM Gershon et al. (2023), EC can have different clinical manifestations, but usually manifests itself with fever, intoxication, lethargy, hypovolemia, abdominal distension, unpleasant odor and explosive diarrhea, sometimes with blood impurities with a preoperative morbidity ranging from 6% up to 60% [9]. Mortality associated with enterocolitis according to various researchers ranges from 5% to 50%, with a higher prevalence and frequency in the neonatal period [7,8, 9,14,15]. And therefore, a justified method of providing emergency care for EC against the background of EC is the imposition of a colostomy [1,6,12,16]. In our study, EC associated with HCG was established in 315 (65.22%) patients.

The most serious and unpredictable complications in the course of Hirschsprung's disease include toxic megacolon. According to R. Khasanov et al. (2015) [11] and Shiwani R Garg et al. (2016) [8]. Voloxic megacolon is defined as an acute expansion of the large intestine accompanied by clinical signs of toxemia. The leading symptoms are abdominal distension, constipation, decreased intestinal motility, and intoxication symptoms such as fever, tachycardia, or hypotension, as well as radiologically confirmed distended bowel loops [4,8,11]. Our own research and data from the literature indicate that the most effective method of treatment, including when providing emergency care, is also the imposition of a colostomy [1,6,7,8,9,12,14,16]. In our study, toxic megacolon was diagnosed in 16 out of 483 patients, which is (3.31%).

It is well known that the result of timely diagnosis and treatment of CG, regardless of the length of aganglionosis and the development of EC complications and toxic megacolon, is an indicator of mortality. In our study, all the children who were admitted to the clinics remained alive.

Thus, summing up, it was established that 444 (91.93%) had a typical classic clinical picture of various forms of aganglionosis. On the other hand, 39 (8.07%) patients out of 483 (100%) had an atypical clinical picture, which was caused by accompanying developmental defects that prevailed in terms of the severity of the course at the time of hospitalization. In our clinical study, the causes of an atypical clinical picture for CH were colonic atresia in 15 (3.10%), colonic atresia + gastroschisis in 3 (0.62%), ileal atresia in 9 (1.86%), esophageal atresia in 3 (0.62%), cleft hard and soft palate in 9 (1.86%). During follow-up examination, EC occurred in 313 (65.22%) patients, of which 212 (75.71%) of 280 children were patients under 6 months of age. Meanwhile, the severity of the course of the disease in the presence of alimentary and enteral insufficiency, as well as against the background of enterocolitis contributed to the development of hypotrophy in 327 (67.70%) and anemia in 241 (49.89%) patients.

Conclusion.

Clinical manifestations and course of CG in children of the first year of life primarily depends on the presence of accompanying malformations, which, depending on the clinical condition and vital disorders, may prevail with varying lengths of intestinal aganglionosis. Therefore, in the clinical course of CG in children of the first year of life, it is necessary to focus on typical and atypical clinical symptoms. In our study, typical clinical symptoms from birth were found in 444 (91.93%), and atypical - in 39 (8.07%).

39 (8.07%) of 483 (100%) patients had an atypical clinical picture, and the reasons were: colonic atresia in 15 (3.10%), colonic atresia + gastroschisis in 3 (0.62%), atresia ileum in 9 (1.86%), esophageal atresia in 3 (0.62%), cleft of the hard and soft palate in 9 (1.86%).

An important point in the timely diagnosis of HCG in children of the first year of life should be greater awareness of neonatologists of maternity hospitals with the disease, to minimize objective complications associated with both typical and atypical clinical picture, and for surgical alertness in maternity hospitals and district pediatricians and family doctors.

The most objective criteria that allow for a timely diagnosis are the absence of meconium excretion, abdominal distension in 444 (91.93%), delayed physiological weight gain against the background of nutritional insufficiency with the development of hypotrophy in 327 (67.70%), stagnant vomiting gastric and intestinal contents in 417 (86.34%). Instead, the complications that arose during the examination of patients with CG based on our study include EC in 315 (65.22%), toxic megacolon in 16 (3.31%) and anemia of various degrees - in 241 (49.89%) patients.

According to the results of a comprehensive examination, the following extent of aganglionosis in children of the first year of life was established: rectal form in 100 (20.70%), rectosigmoid in 192 (39.75%), subtotal in 150 (31.06%) and total in 41 (8.49 %) of the patient. Depending on the age, there were 280 (57.97%) patients under 6 months, and 203 (42.03%) patients between 6 months and 1 year. Associated malformations were found in 20.29% of patients.

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