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THE RESULTS OF A NEW ORGANIZATIONAL MODEL INTRODUCED FOR THE CARE OF NEWBORNS WITH CONGENITAL MALFORMATIONS: A CLINICAL CASE

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Abstract

An important aspect in providing surgical care to newborns with congenital disorders in the early neonatal period is a timely diagnosis, transportation of a little patient to the surgical hospital, nursing and care in the postoperative period. To date, the concern of transportation of newborn babies remains ambiguous and controversial.

The given study exemplifies a clinical case of a newborn with anorectal malformation, atresia of anus, rectum, an ivory form, in combination with a congenital malformation of the urinary system. On the example of this clinical case, the potential risk mitigation measures have been assessed to avoid any life-threatening complications with transportation excluded.

The timely diagnosis and treatment contributed to a favorable prognosis and the absence of complications. A special feature of this case is complex treatment in the conditions of the 3rd level perinatal center. The pre-operative and post-operative care was given by neonatologists. The patient was discharged with improvement at the age of 19 days in satisfactory condition.

Keywords: *malformations, newborns, transportation.*

Резюме

РЕЗУЛЬТАТЫ ВНЕДРЕНИЯ В ПРАКТИКУ НОВОЙ ОРГАНИЗАЦИОННОЙ МОДЕЛИ ОКАЗАНИЯ ПОМОЩИ НОВОРОЖДЕННЫМ С ПОРОКАМИ РАЗВИТИЯ: КЛИНИЧЕСКИЙ СЛУЧАЙ

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Важным звеном оказания хирургической помощи новорожденным с ВПР в ранний неонатальный период является своевременная диагностика, транспортировка пациента в хирургический стационар, выхаживание и уход в послеоперационном периоде. Вопрос транспортировки новорожденных детей остается неоднозначным и спорным до настоящего времени.

Представлен клинический случай новорождённого, с аноректальным пороком развития, атрезией ануса, прямой кишки, бессвищевая форма в сочетании с врожденным пороком развития органов мочевой системы. На примере

данного клинического случая проведена оценка возможности снижения рисков жизнеугрожающих осложнений при исключении транспортировки.

Своевременная диагностика и лечение способствовало благоприятному прогнозу и отсутствию осложнений. Особенностью данного случая является проведение комплексного лечения в условиях перинатального центра 3 уровня. Дооперационный и послеоперационный уход проводился неонатологами. Выписан с улучшением в возрасте 19 суток, в удовлетворительном состоянии.

Ключевые слова: *пороки развития, новорожденные, транспортировка.*

Түйіндеме

ДАМУ АҚАУЛЫҚТАРЫ БАР ЖАҢА ТУҒАН БАЛАЛАРҒА ЖАҢА ҰЙЫМДАСТЫҚ КҮТІМ ҮЛГІСІН ТӘЖІРИБЕГЕ ЕҢГІЗУ НӘТИЖЕЛЕРІ: КЛИНИКАЛЫҚ ЖАҒДАЙ

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Ерте неонаталдық кезеңде туа біткен ақаулары бар жаңа туған нәрестелерге хирургиялық көмек көрсетудің маңызды буыны дер кезінде диагностикалау, науқасты хирургиялық стационарға жеткізу, мейірбикелік күтім және операциядан кейінгі кезеңде күтім жасау болып табылады. Жаңа туған нәрестелерді тасымалдау мәселесі бүгінгі күнге дейін түсініксіз және даулы болып қала береді.

Жаңа туылған нәрестенің аноректальді даму ақауы, анус, тік ішек атрезиясы, фистулярлық нысаны зәр шығару жүйесі мүшелерінің туа біткен ақауымен біріктірілген клиникалық жағдайы ұсынылған. Осы клиникалық жағдайдың мысалында тасымалдауды алып тастай отырып, өмірге қауіп төндіретін асқынулардың қаупін азайту мүмкіндігін бағалау жүргізілді.

Уақытылы диагноз қою және емдеу қолайлы болжамға және асқынулардың болмауына ықпал етті. Бұл жағдайдың ерекшелігі 3-деңгейдегі перинаталдық орталықта кешенді ем жүргізу болып табылады. Операцияға дейінгі және операциядан кейінгі күтімді неонатологтар көрсетті. Жағдайы қанағаттанарлық, 19 тәулікте жақсарумен шығарылды.

Түйінді сөздер: *даму ақаулары, жаңа туған нәрестелер, тасымалдау.*

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Background

Maternal and child health is one of the most important and most challenging tasks for the national health system. In the strategic documents and Addresses to the people of Kazakhstan, the President highlighted the need to reduce maternal and infant mortality and increase life expectancy of the population.

In the structure of infant mortality in the Karaganda region, congenital malformations (CM) consistently occupy a leading place and account for 35.5%. An important aspect in providing surgical care to newborns (0-6 days) with congenital disorders in the early neonatal period is a timely diagnosis, transportation of a little patient to the surgical hospital, nursing and care in the postoperative period. To

date, the issue of transportation of newborn babies remains ambiguous and controversial.

Transporting newborns within the Karaganda region, which is the largest region in area, can take from 3 to 7 hours. This time factor affects the survival of newborn children with malformations due to the violation of the "heat chain" during transportation from hospital to hospital, which deteriorates the condition of the newborn and increases the chances of metabolic disorders and postoperative complications.

In order to improve the quality of the neonatal surgical service in the Karaganda region, a mobile team was arranged in November 2017 to conduct emergency surgical interventions in the premises of the regional perinatal center, in Zhezkazgan, Balkhash. Within the period of 2018-2022, 192 newborns with congenital malformations were operated on: gastrointestinal tract (GIT), urinary system organs, defective anterior abdominal wall, diaphragmatic hernias and other malformations [2].

For the reporting period from 2018 to 2022, the mortality rate of newborns with congenital malformations operated in the maternity hospitals of the Karaganda region decreased from 53.8% (2017) to 33.3% (2021). For 2020 and 2021, such severe defects as diaphragmatic hernias, large omphalocele, gastroschisis of large sizes, a rare pathology of complete gastric duplication, complete bladder exstrophy were operated with a favorable outcome [2]. Mortality in these pathologies varies within fairly wide limits – on average at 45-65%, while the mortality rate in the CIS countries is 23-55%.

Anorectal malformations of the gastrointestinal tract are the most common anomalies in the development of the gastrointestinal tract. According to various authors, the frequency of anorectal malformations (AMs) ranges from 1.66 to 9.94 per 10,000 children and does not tend to decrease [1].

In case of AMs, there are concomitant malformations in 40% - 70% of cases with the following pathologies: genitourinary system (42%), skeletal anomalies (30%,), other malformations of the gastrointestinal tract (18%) and cardiovascular anomalies (17%). Mortality in anorectal malformations reaches 17%-18% and is due to either concomitant malformations or postoperative purulent-septic complications, the frequency of which reaches 57-68% of all cases [5,6].

In the case of an antenatally detected malformation or suspected malformation, delivery is prescribed as the obstetric care, which includes specialized surgical care.

The aim of the study is to analyze an approach and strategies to be taken in case of congenital anorectal malformations with a clinical case of a newborn with anorectal pathology in combination with a congenital malformation of the urinary system.

Materials and methods of research. The child was born in the Regional Clinical Hospital in the Perinatal Center No. 2 of the Karaganda region, which is a level 3 institution according to the perinatal care categorization. A comprehensive examination was done, which included radiography of the chest and abdominal cavity in an upright position, ultrasound of the kidneys and heart, electrocardiography, consultation of specialty physician, surgical correction, early rehabilitation, conservative and symptomatic therapy. A written voluntary informed consent has been obtained from the patient's parents to publish this clinical case for scientific purposes, including the use of patient's medical data in both medical log and electronic version (results of examination, treatment and observation). When describing the case, data from the child's medical record were used.

Medical History. A full-term male, 3680.0 grams, 55 cm, Apgar score – 8/9 points from woman born in 1992, 1 pregnancy, 1 urgent birth, subject to regular D checks from the 8th week + 1 day. The pregnancy proceeded with some mild anemia, which was treated from the second half of pregnancy. During pregnancy, the fetus was scanned twice by UT. The second ultrasound screening at 31 week and 2 days antenatally showed a congenital malformation of the fetal urinary system: aplasia of the left kidney, hydrocele. It was recommended to deliver a baby in the level 3 perinatal center.

Anamnesis. The condition of the baby at birth is of moderate severity. The severity of the disease is due to malformation of the anus. The newborn is transferred after birth to the intensive care unit for newborns. The cry is loud. Independent breathing is regular without any auxiliary muscles engaged. Blood saturation is 100%, no need in additional oxygenation. The skin is clean and pink. Body temperature is 36.6 ^{o}C. Visible mucous membranes are clean and pink. Umbilical cord is clamped. The head is moderately configured, the sutures are closed, the anterior fontanel is 0.5 * 0.5 cm, the bones of the skull are dense. The eyes open freely. There are no focal symptoms. Reflexes of congenital automatism are active; muscle tone is physiological. In the lungs, puerile breathing, no wheezing, no shortness of breath. Heart sounds are loud, rhythmic, heart rate is 123. Pulsation on peripheral vessels is satisfactory. Hemodynamics is stable. The abdomen is of regular shape, symmetrical, not swollen, soft, painless on palpation. The liver and spleen are not enlarged. External genitalia of the male type. The baby cannot urinate and defecate.

Status localis: On examination, the anal orifice is missing in a typical location, there are no perianal folds.

The baby was examined by a neonatal surgeon on the first day after birth. To clarify the level of rectal atresia and the type of defect, Wangenstein's invertography was performed. The distance from the cecum rectal sac to the skin was 10 cm.

To exclude combined congenital malformations, the organs of the urinary system were UT tested to diagnose the echography of the megaureter on the right and agenesis of the left kidney.

Preliminary diagnosis: Congenital malformation of the gastrointestinal tract: anorectal malformation, atresia of the anus, rectum, spineless form (anal agenesis). Pre-operative preparation was started.

On the second day of life, taking into account multiple congenital malformations, including potentially undiagnosed ones, minimizing the possible consequences, surgical intervention, namely, laparotomy was performed in-situ in the perinatal center. Further on, abdominoscopy and sigmostomy were completed. No technical difficulties arise during the operation. The baby received a course of antibiotic therapy postoperatively. The first 2 days after the

operation, respiratory support by ventilation continued. On the 3rd day, stimulation of the gastrointestinal tract began by enteral nutrition in the amount of minimal trophic nutrition. On the 4th day, the baby began to absorb enteral nutrition. On the 12th day of life, the baby was given his mother's breast milk. On the 14th day of life, it was transferred to the second stage of nursing newborns.

The baby was discharged home at the age of 19 days (18th day of the postoperative period) under the supervision of the local pediatrician, pediatric surgeon at the place of residence until the II stage of hospitalization.

The condition is satisfactory. The abdomen is not swollen, not tense on palpation, painless. Peristalsis is satisfactory. The baby defecates down the sigmostome.

Status localis: Peristomal area with no signs of inflammation.

Weight at discharge was 3580 gr. (+112,0 gr from birth). Anorextoplasty is recommended at the age of 3 months. At the age of 2 months, excretory urography was performed to reveal the following: Agenesis of the kidney on the left. The megaureter on the right was removed.

A special feature of this case is the combination of anus atresia and congenital malformation of the urinary system together with some complex treatment provided at the 3rd level perinatal center. The preoperative and postoperative care was given by neonatologists. Based on this clinical case, the possibility of reducing the risks of life-threatening complications with the exclusion of transportation was assessed.

Discussion

The diagnosis and treatment of children with anorectal anomalies currently remain the most urgent and unresolved problem of pediatric surgery. The duration of its study is correlated with the entire history of the development of congenital malformations' surgical correction. However, the issues of timely diagnosis and the choice of the optimal treatment depending on the anatomical form of malformation are still the subject of study and discussion and remain in the focus of attention of researchers and practitioners [3,4].

The relevance of the problem in modern conditions is due to the lack of a single strategy to conduct a preoperative examination and prepare a patient for radical surgical intervention. There is no consensus on the timing for such surgical corrections of various anatomical malformations [5,6]. The question of selecting an appropriate surgical correction method remains controversial. It is well known that newborns with anorectal malformations should be placed in specialized departments for children with congenital malformations, where specialists can perform such medical procedures such as examination, surgical intervention, treatment and care in the postoperative period, further dispensary observation, other medical appointments and repeated courses of rehabilitation. However, despite the development of minimally invasive surgery and the improvement of surgical treatment in recent decades, the functional results of operations do not always satisfy surgeons [10,12].

According to the recent studies, in order to reduce postoperative complications, it is recommended to adhere to the approximate timing of treatment: colostomy in the neonatal period, radical anorectoplasty at the age of 2-3 months (with an anogenital cleft at 4-6 months), colostomy closure at 1-2 months after surgery [7,8].

In the postoperative period, it is necessary to conduct antibiotic therapy with cephalosporin drugs with the addition of metronidazole to the therapy within 5-7 days and anesthesia for 2-4 days [7]. Enteral feeding is usually prescribed from the second day in the absence of severe concomitant pathology. When restoring the passage through the gastrointestinal tract (discharge passage, absence of stagnant discharge from the stomach, absence of intestinal paresis signs), the volume of enteral nutrition consistently increases. The child's parents are taught to take care for a little patient (cleansing the skin around the stoma, changing a colostomy bag) [9].

Anorectal malformations require surgical intervention in the early neonatal period in order to preserve the gastrointestinal tract functions, which is associated with the frequent development of complications. Most of the complications occur in the late postoperative period. Mostly often these complications include chronic colonic stasis, fecal incontinence as well as urological problems or sexual dysfunctions in adulthood [11,13]. These complications lead to disability and serious psychosocial maladaptation of patients with anorectal malformations. Clinical examination of patients with anorectal malformations should continue for a long period.

Having analyzed the available literature reviews and with own treatment results obtained in recent years in mind, one might conclude that surgical treatment of newborns at the place of birth in the conditions of the perinatal center increases the effectiveness of the measures provided [2]. At the same time, a number of adverse factors that have a negative impact on the outcome of the operation are eliminated. These factors include hypothermia, loss of time for transportation and adaptation of newborns, a risk of infection and aspiration.

Conducting complex rehabilitation therapy in the postoperative period ensures a full-fledged stable functional result. In case of applying standardized approaches in diagnosis, determining the indications for surgery, intraoperative strategies, postoperative management, it is possible to minimize complications. The presence of combined congenital malformations of the urinary system in the form of aplasia of the left kidney confirms the genetic nature of the pathology requiring further molecular genetic examination and significantly reducing the quality of life of a child with incapacitating consequences.

Thus, as a result of a new organizational model introduced for the care of newborns with malformations, the stage of transportation and the adverse factors associated with it were excluded: time costs, risks of hypothermia, infection, while this contributed to ensuring the continuity of the treatment process, reducing the frequency of complications at the stage of the preoperative period as well as the mortality rate for congenital malformations by 20.5%.

Findings

The presented study and clinical observation indicate that the delivery and surgical treatment of newborns at the place of birth in the conditions of the 3rd level perinatal center excludes adverse risk factors and increases the effectiveness of the measures taken.

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