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Experience of separating conjoined twins in the Republic of Uzbekistan

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Aim – to present the clinical experience of the successful surgical separation of conjoined twins fused through the liver, with a description of the preoperative diagnostic assessment, surgical strategy, and postoperative management.

Clinical case. The article presents the successful separation of female Siamese twins from the Republic of Kyrgyzstan (omphalopagus), performed in Uzbekistan at the age of 6 months and 15 days. The examination results revealed that the children were fused through the left lobe of the liver. Each twin has a separate gallbladder. One of the girls exhibits a reversed arrangement of internal organs in the abdominal cavity (situs inversus visceralis). An electrocoagulator, ultrasonic scalpel, argon, and BloodSTOP iX hemostatic material were utilized during the operation. Liver tissue was dissected with an ultrasonic scalpel and electrocoagulator, followed by argon treatment of the exposed liver surface. The total blood loss was approximately 20–25 mL. The children were transferred from the intensive care unit to the neonatal surgery department on the 15th day post-operation. Thirty days after the operation, both children were discharged home in satisfactory condition.

Conclusion. Surgical separation of conjoined omphalopagus twins fused at the liver is feasible with thorough preoperative assessment of the anatomy of the fusion, meticulous surgical planning, and the use of effective hemostatic techniques. The surgical strategy employed in this case enabled successful separation of the twins with minimal blood loss, preservation of both children's lives, and a satisfactory postoperative outcome. This case represents the first independently performed successful separation of conjoined twins by pediatric surgeons in the Republic of Uzbekistan.

The study was conducted in accordance with the principles of the Declaration of Helsinki. Informed consent was obtained from the child's parents participating in the study.

The authors declare no conflicts of interest.

Keywords: conjoined twins, omphalopagus, hepatic fusion, surgical separation.

Досвід розділення сіамських близнюків у Республіці Узбекистан

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Мета – представити клінічний досвід успішного хірургічного розділення сіамських близнюків, зрощених у ділянці печінки, з описом передопераційної діагностики, хірургічної тактики та післяопераційного ведення.

Клінічний випадок. Описано випадок успішного розділення сіамських близнюків жіночої статі (омфалопагі) із Республіки Киргизстан, проведеного в Узбекистані у віці 6 місяців і 15 днів. Результати обстеження виявили зрощення дітей у ділянці лівої частки печінки. Кожна дитина мала окремий жовчний міхур. У однієї з дівчаток спостерігалось дзеркальне розташування внутрішніх органів черевної

порожнини (situs inversus visceralis). Під час операції використовували електрокоагулятор, ультразвуковий скальпель, аргон та гемостатичний матеріал Bloodstop iX. Розсічення тканин печінки здійснювали за допомогою ультразвукового скальпеля та електрокоагулятора з подальшою обробкою оголеної поверхні печінки аргонном. Загальна крововтрата становила приблизно 20–25 мл. На 15-ту добу після операції дітей перевели з відділення інтенсивної терапії до відділення неонатальної хірургії. Через 30 днів після операції обох дітей виписали додому в задовільному стані.

Висновок. Хірургічне розділення сіамських близнюків-омфалопагів, зрощених у ділянці печінки, є можливим за умови ретельного передопераційного оцінювання анатомії зрощення, детального планування операції та застосування ефективних методів гемостазу. Обрана хірургічна тактика дала змогу успішно розділити близнюків із мінімальною крововтратою, зберегти життя обох дітям і забезпечити задовільний післяопераційний результат. Цей випадок є першим прикладом успішного розділення сіамських близнюків, самостійно виконаного дитячими хірургами в Республіці Узбекистан.

Дослідження проведено відповідно до принципів Гельсінської декларації. Отримано інформовану згоду батьків дитини, яка брала участь у дослідженні.

Автори заявляють про відсутність конфлікту інтересів.

Ключові слова: сіамські близнюки, омфалопаги, зрощення печінки, хірургічне розділення.

It is well established that conjoined twins represent one of the rarest and most severe forms of congenital anomalies. The incidence of this condition ranges from approximately 1 in 50,000 to 1 in 100,000 live births.

At present, in cases where a favorable anatomical configuration of conjunction is present, such twins can undergo surgical separation, enabling each of them to subsequently lead a fully functional and relatively normal life [2,3].

In clinical practice, the classification of conjoined twins proposed by Potter is the most commonly and widely applied. According to this classification, the following types are distinguished: thoracopagus (40.0% of cases), omphalopagus (33.0%), pygopagus (19.0%), ischiopagus (6.0%), and craniopagus (2.0%), [4,6].

Aim – to present the clinical experience of successful surgical separation of conjoined twins fused through the liver, with a description of the preoperative diagnostic assessment, surgical strategy, and postoperative management.

The study was conducted in accordance with the principles of the Declaration of Helsinki. Informed consent from parents was obtained for the study.

Clinical case

The article presents a case of the successful separation of female Siamese twins from the Republic of Kyrgyzstan (omphalopagus) in Uzbekistan at the age of 6 months and 15 days. Ultrasound of internal organs and Multi-slice Computed Tomography (MSCT) with contrast of the vessels and gastrointestinal tract (GI) were employed for diagnosis. The examination results revealed that the children were fused through the left lobe of the liver. Fatima's left lobe of the liver is connected to Zukhra's left lobe, with separate gallbladders for each twin. Zukhra exhibits a reversed arrangement of internal organs in the abdominal cavity (Situs Inversus Visceralis). Electrocoagulator, ultrasonic scalpel, argon, and BloodSTOP iX

hemostatic material were utilized during the operation [5,6]. Liver tissue was dissected with an ultrasonic scalpel and electrocoagulator, followed by argon treatment of the exposed liver surface. The total blood loss was approximately 20–25 mL. The authors successfully employed a minimal number of instruments to bloodlessly separate the twins within 3 hours, completing the operation. The children were transferred from the intensive care unit to the neonatal surgery department on the 15th day post-operation. Thirty days after the operation, both children were discharged home in satisfactory condition. Fatima's weight at discharge was 6200 grams, and Zukhra's weight was 6050 grams.

Our case was classified as omphalopagus, meaning that the twins were conjoined at the abdomen, specifically at the level of the liver. On July 6, 2022, citizens of the Kyrgyz Republic contacted Professor Ergashev B.B. via telephone and Telegram with a request for assistance in the treatment of their conjoined daughters, named Fatima and Zukhra. At that time, the infants were 37 days old (Fig. 1).

We advised the parents to arrange Magnetic Resonance Imaging (MRI) or MSCT at a local medical facility and to forward the results to our center for evaluation. However, a few days later, they re-established contact and reported that these investigations could not be performed at the local clinic, as the physicians had declined to conduct the examinations. Subsequently, we monitored the twins remotely for a period of three months. In October 2022, they were invited to our center for comprehensive evaluation. Their first admission took place from October 7 to October 12, 2022.

Upon admission to the clinic, a comprehensive medical history was obtained.

According to the obstetric history, at 28 weeks of gestation, prenatal ultrasonography revealed a developmental anomaly diagnosed as conjoined twins. The pregnancy was complicated by maternal anemia and toxemia. At the place of residence, a medical council was con-

Clinical case



Fig. 1. Conjoined twins (37 days old)



Fig. 2. Conjoined twins (age 4 months 15 days, total weight 9150 g, length 67 cm)

vened; due to the lack of capacity to provide specialized care for such infants, it was decided to terminate the pregnancy for medical indications by cesarean section. Following the operation, female conjoined twins were delivered with a combined birth weight of 1,600 grams. The infants were from the mother's second pregnancy and represented a twin gestation. On the 10th day of life, the twins were discharged home with a poor prognosis, as it was assumed that their survival would be limited. However, their postnatal condition remained relatively satisfactory. No significant delays in growth or development were observed. In view of this, the parents decided to seek consultation with Professor Ergashev B.B., requesting further management and resolution of their condition.

The condition of the girls was relatively satisfactory, though they responded to examination with signs of

restlessness. On physical inspection, two bodies were observed, conjoined at the abdomen with their faces turned toward each other. Visually, Fatima appeared larger than Zukhra. The combined weight of the infants was 9,150 grams, with a length of 67 cm. The diameter of the fused area of the anterior abdominal wall measured 24 cm. Due to the described and constrained positioning, the twins were unable to lie in other postures. They remained in a lateral position, facing each other (Fig. 2).

The skin and visible mucous membranes of both girls were clean and pale pink in color. Subcutaneous adipose tissue was normally developed. Breathing was spontaneous and nasal. Auscultation over both lungs revealed harsh respiratory sounds. Heart tones were rhythmic but muffled. Peripheral pulses were of moderate amplitude. Intestinal peristalsis was audible. On palpation, the abdomen was soft and non-tender. The genitourinary organs and anus were separately formed for each twin. Bowel movements were regular in both infants, while urine output in Zukhra was slightly reduced compared to Fatima. No allergic reactions to medications were identified. The family history was unremarkable, although the parents were consanguineous. Information regarding routine vaccinations received was not available.

At our center, the twins underwent a comprehensive diagnostic evaluation, including complete blood count, urinalysis, and stool analysis. Biochemical blood tests were performed, along with ultrasonography of all internal organs, neurosonography (NSG), and echocardiography (EchoCG). Chest radiography was conducted, as well as MSCT with vascular contrast and MSCT with gastrointestinal contrast, performed on one of the twins (Fatima).

The results of the examinations, including MSCT with vascular and gastrointestinal contrast, confirmed that the twins are omphalopagus, joined via the left lobes of their livers. The left lobe of Fatima's liver was connected to the left lobe of Zukhra's liver, while the main hepatic hila were separate. Each twin had an individual gallbladder. Gallbladder dimensions on ultrasonography were 24×9.8×9.1 mm for Fatima and 25×8.3×8.6 mm for Zukhra. Zukhra was noted to have situs inversus visceralis, indicating a reversed arrangement of the abdominal organs. MSCT with gastrointestinal contrast performed on Fatima demonstrated that the stomach and intestines were separately developed, with no fusion between them. EchoCG revealed a patent foramen ovale (PFO) measuring 3 mm in both twins. NSG showed echographic signs of hypoxic changes. All other organs in both twins were anatomically separate.



Fig. 3. Sequential intubation of the twins



Fig. 4. Position of the twins on the operating table after intubation

Laboratory findings for Fatima: Hemoglobin (Hb) – 119 g/L, erythrocytes – $4.30 \times 10^{12}/L$, hematocrit – 0.94, leukocytes – $11.28 \times 10^9/L$, platelets – $301 \times 10^9/L$.

Biochemical analysis: total protein – 60 g/L, blood glucose – 5.4 mmol/L, alanine aminotransferase (ALT) – 37 U/L, total bilirubin – 8.1 $\mu\text{mol}/L$ (direct bilirubin – 0 $\mu\text{mol}/L$, indirect bilirubin – 8.1 $\mu\text{mol}/L$). Urinalysis and stool analysis were within normal limits. Blood type: B (III), Rh positive.

Laboratory findings for Zukhra: Hb – 107 g/L, erythrocytes – $4.10 \times 10^{12}/L$, hematocrit – 0.92, leukocytes – $11.28 \times 10^9/L$, platelets – $301 \times 10^9/L$.

Biochemical analysis: total protein – 60 g/L, blood glucose – 5.4 mmol/L, ALT – 37 U/L, total bilirubin – 8.1 $\mu\text{mol}/L$ (direct bilirubin – 0 $\mu\text{mol}/L$, indirect bilirubin – 8.1 $\mu\text{mol}/L$). Urinalysis and stool analysis were within normal limits. Blood type: B (III), Rh positive.

Based on clinical and instrumental examinations, the following diagnosis was established: Conjoined twins, omphalopagus (shared liver and abdominal cavity, ICD-10: Q89.4), with situs inversus visceralis in one of the twins.

Afterward, we temporarily discharged the twins home, as time was required for preparation and for a multidisciplinary consultation with our colleagues. Considering that the children were foreign nationals and that their stay in our clinic would incur significant costs, we offered the parents the option of a temporary discharge.

The twins were admitted to our center for the second time on December 3, 2022. At admission, their condition was of moderate severity, with symptoms including cough, nasal congestion, and elevated body temperature up to 38.5°C. According to the parents, the children developed a cold while crossing the border between the Kyrgyz Republic and Uzbekistan. The twins were examined by a pediatrician, who diagnosed acute bronchitis and initiated antibacterial therapy. Both children received airway clearance therapy and inhalation therapy.

After two weeks of therapy and complete somatic recovery of the twins, a multidisciplinary consultation was held. A detailed operative plan was developed, specifying the role and position of each surgeon involved in the procedure. It was decided that the surgery would be performed with the participation of four lead surgeons. Furthermore, it was planned that after separation, one surgical team of three surgeons would continue operating on Fatima, while a second team of three surgeons, working in a separate operating room, would complete the procedure for Zukhra. An anesthesia plan was jointly developed with the anesthesiology team to ensure perioperative support. The operation was scheduled for December 19, 2022.

Thus, on December 19, 2022, the twins were taken to the operating room. Intubation was performed sequentially according to the pre-established plan. One twin was placed in the supine position while the other was held manually. After intubation of the first twin, she was lifted and held in position, and the second twin was then intubated in the supine position (Fig. 3, 4).

No allergic reactions to medications were identified. The family history was unremarkable, although the parents were consanguineous. Information regarding previously received routine vaccinations was not available.

The surgery was performed as follows. After preparation and sterilization of the operative field (Fig. 5), a circumferential incision was made along the area of fusion, with slightly more skin preserved for Fatima than for Zukhra (Fig. 6).

The abdominal cavity was then opened layer by layer. Liver tissue was encountered at the wound site (Fig. 7). Upon careful inspection, two hepatic hila and two gallbladders were identified. The twins were joined by their left liver lobes; due to Zukhra's situs inversus visceralis, her liver was located on the left side, whereas Fatima's liver was in the normal anatomical position on the right.

Next, the liver was separated along a faintly visible groove between the left lobes. During the procedure, an electrocautery device, ultrasonic scalpel, argon beam,

Clinical case



Fig. 5. Preparation of the surgical field



Fig. 6. Circumferential skin incision at the site of twin fusion



Fig. 7. Abdominal cavity opened; liver tissue exposed



Fig. 8. Use of argon, ultrasonic scalpel, and electrocautery during the surgery



Fig. 9. Moment of liver separation

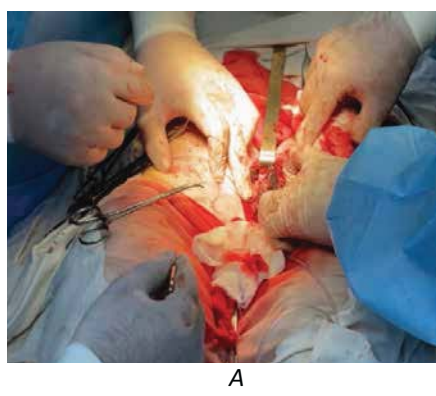


Fig. 10. Use of BloodSTOP iX during (A) and after liver separation (B)

and hemostatic material BloodSTOP iX were used (Fig. 8). Initially, the liver tissue was dissected with the ultrasonic scalpel and electrocautery, after which the exposed liver surface was treated with argon. The total blood loss was approximately 20–25 mL. Using a minimal set of instruments, the twins were successfully separated in a bloodless manner within three hours, and the operation was completed successfully.

After liver separation (Fig. 9), a single layer of BloodSTOP iX was applied to the exposed liver surfaces of both children (Fig. 10).

This was sufficient to achieve complete hemostasis and bile stasis, with total intraoperative blood loss not

exceeding 25 mL. Following liver separation, the skin and muscular layers were incised, thereby fully separating the twins. One surgical team of three remained with Fatima, while the other team of three, together with anesthesiologists and critical care specialists, moved to a separate operating room to continue with Zukhra. Closure and reconstruction of the anterior abdominal wall presented no significant difficulty. Fatima had no viscerovisceral or viscerocostal disproportion, whereas Zukhra had a minor viscerocostal disproportion, which was corrected by mobilizing the skin – aponeurotic flaps laterally. For Zukhra, BloodSTOP iX was additionally applied to ensure reliable hemostasis and bile stasis.



Fig. 11. Day of discharge

The duration of surgery was 2 hours 45 minutes for Zukhra and 3 hours for Fatima. Both twins were extubated on postoperative day 3. Fatima's postoperative course was relatively smooth, and she was extubated on day 4. In contrast, starting from postoperative day 3, Zukhra developed oliguria, which progressed to oligoanuria by day 4. Considering these findings alongside elevated serum creatinine and urea levels, a single session of hemodialysis was performed on postoperative day 5. The hemodialysis was successful, as evidenced by a gradual increase in urine output and normalization of creatinine and urea levels thereafter.

On postoperative day 15, the children were transferred from the intensive care unit to the neonatal surgery ward. Thirty days after surgery, both children were discharged home in satisfactory condition. At discharge, Fatima weighed 6200 grams, and Zukhra weighed 6050 grams. This day was a major celebration for all medical staff in Uzbekistan, particularly the personnel of the Republican Perinatal Center (Fig. 11).

Thus, our case of separated «Siamese twins» represents the first independently successful surgery performed by pediatric surgeons in the Republic of Uzbekistan, resulting in the survival of both children.

Discussion

Conjoined twins represent a rare congenital developmental anomaly in which the feasibility of surgical separation is determined primarily by the anatomical location of the fusion, the nature and extent of shared organs, and the presence of common vascular structures. According to the literature, the incidence of conjoined twins is approximately 1 in 50,000 births, with a substantial proportion of cases resulting in intrauterine or early postnatal death. A favorable prognosis for surgical treatment is primarily associated with the absence of a shared heart and other vital organs.

The presented clinical case involved omphalopagus twins, one of the anatomical types of conjoined twins in which the fusion is predominantly localized to the anterior abdominal wall. In this case, the liver was the principal organ involved in the fusion. A distinctive feature of this case was the presence of separate gallbladders and independently developed GIs in each child, while the fusion involved portions of the hepatic parenchyma. The anatomical pattern of organ fusion, together with the absence of significant involvement of other vital organs, provided favorable anatomical conditions for surgical separation.

Clinical case

One of the key factors contributing to the successful management of conjoined twins is a detailed preoperative assessment of the anatomy of the fusion. Current studies emphasize the importance of comprehensive imaging to determine organ arrangement, the pattern of vascular communication, the anatomy of the biliary system, and the extent of shared hepatic parenchyma. Contrast-enhanced computed tomography and MRI are considered important modalities for preoperative planning, as they provide essential anatomical information required for selection of the surgical strategy.

In the present case, comprehensive preoperative assessment included ultrasonography of the internal organs, echocardiography, and contrast-enhanced multislice computed tomography of the vascular structures and GI. The findings demonstrated that the twins were predominantly fused through the left hepatic lobes, while the hepatic hila and gallbladders were separate. An additional anatomical feature was the presence of *situs inversus visceralis* in Zukhra. Thus, preoperative imaging not only confirmed the type of fusion but also identified critical anatomical features that had to be taken into consideration during the surgical procedure.

Similar findings have been reported in the literature. In some cases of omphalopagus twins, hepatic fusion has also been described in the presence of two separate gallbladders and independently developed GIs. However, the extent of vascular communication between the twins may vary considerably, emphasizing the need for individualized anatomical assessment in each case. In published cases of successful separation of twins sharing a portion of the liver, the surgical strategy was based on preoperative delineation of the vascular anatomy and assessment of the feasibility of safe division of the hepatic parenchyma.

The preoperative preparation of the surgical team was also of considerable importance in the present case. A multidisciplinary consultation was conducted before surgery, during which the sequence of operative steps was established, responsibilities among the surgeons and anesthesiologists were allocated, and the subsequent management of each child following separation was planned. This approach is consistent with current principles of conjoined twin surgery, according to which successful intervention requires multidisciplinary collaboration, meticulous preoperative planning, and the ability to provide simultaneous independent care for each twin after separation.

Particular attention should be paid to the technique of hepatic separation. In the present case, an ultrasonic scalpel and electrocautery were used to divide the hepatic tissue, followed by argon treatment of the hepatic

surface and application of the hemostatic agent Blood-STOP iX. This multimodal approach enabled hepatic separation with an overall blood loss of approximately 20–25 mL. Comparable reports in the literature demonstrate the feasibility of successful division of a hepatic bridge using modern surgical technologies; however, the amount of blood loss depends substantially on the pattern of vascular communication and the extent of shared hepatic parenchyma. In one published case, separation of omphalopagus twins with a shared portion of the liver using a harmonic scalpel lasted approximately 2.5 hours, after which both children were successfully separated and discharged.

Recent publications emphasize that surgical separation of twins with shared hepatic tissue is technically challenging because of the variability of vascular anatomy and the risk of substantial blood loss. Accordingly, accurate visualization of vascular structures, careful planning of the sequence of tissue division, and the availability of effective methods for rapid hemostasis are of major importance. In specialized centers, three-dimensional reconstructions and physical models are also used to facilitate a more detailed understanding of the anatomy and improve the accuracy of preoperative surgical planning.

In the present case, the absence of a shared heart and other critically important organs represented a major favorable factor for successful separation. The literature similarly indicates that the extent of involvement of vital organs is one of the principal determinants of the feasibility and prognosis of surgical separation. The most unfavorable anatomical configurations are those involving a shared heart and severe cardiovascular anomalies, whereas omphalopagus twins with predominantly hepatic fusion and favorable anatomy have substantially greater potential for successful separation.

The postoperative course of the presented twins was generally favorable. Both children were extubated during the early postoperative period and were transferred from the intensive care unit to the neonatal surgery department on postoperative day 15. One of the twins developed oliguria progressing to oligoanuria, accompanied by elevated serum creatinine and urea levels, and required a single session of hemodialysis. Following the procedure, a favorable clinical response was observed, with restoration of urine output and normalization of laboratory parameters. Thirty days after surgery, both girls were discharged home in satisfactory condition.

Thus, the present clinical case supports the findings reported in the literature regarding the feasibility of successful surgical separation of omphalopagus twins in the absence of a shared heart and other critically im-

portant organs. At the same time, it demonstrates that even in cases with a relatively favorable anatomical pattern of fusion, a successful outcome requires comprehensive preoperative assessment, detailed operative planning, coordinated efforts of multiple surgical and anesthesiology teams, and effective control of intraoperative bleeding. This experience is of particular importance for the development of pediatric and neonatal surgery in the Republic of Uzbekistan, as the procedure represented the first independently performed successful separation of conjoined twins by pediatric surgeons in the country.

Conclusion

Surgical separation of omphalopagus conjoined twins with hepatic fusion is a complex, multistage procedure whose success depends on thorough preoperative assessment, detailed evaluation of the anatomy of fusion, individualized surgical planning, and comprehensive postoperative management. In the present clinical case, comprehensive preoperative imaging, sequential division of the hepatic parenchyma, and the use of modern hemostatic techniques enabled successful surgical separation with minimal blood loss and preservation of the lives of both children. The outcome confirms the

feasibility of successful surgical treatment of omphalopagus twins with a favorable anatomical pattern of fusion and underscores the importance of a multidisciplinary approach to the management of these patients. This clinical experience has important practical implications for the further development of specialized pediatric and neonatal surgical care for patients with conjoined twins in the Republic of Uzbekistan.

The authors declare no conflicts of interest.

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