CLINICAL CASE OF CORRECTION OF VENTRICULAR SEPTAL DEFECT COMBINED WITH AN ANOMALY OF THE SYSTEMIC VENOUS CONNECTION OF THE INFERIOR VENA CAVA

Kuatbekov K.N.[1], Egizekov A.L.[1], Mishin A.V.[1], Musagaliev D.T.[1], Baizhigitov N.B.[1], Suieubekov B.Y.[1], Botabekov T.E. [2]

[1]Center of Modern Medicine "Mediterra" Institute of Surgery LLP, Almaty, Kazakhstan,
[2]«S.D. Asfendiyarov Kazakh National Medical University», NC JSC Almaty, Kazakhstan

Abstract

Interrupted inferior vena cava is a rare condition that can occur either in isolation or in combination with asplasia or polysplasia syndromes. Abnormal development of systemic veins is closely related to atrial situs. In levocardia, there are signs of abdominal organ inversion, which is called visceral situs. The present paper describes a clinical case of a infant with a large interventricular septal defect combined with interrupted inferior vena cava with azygous continuation of visceral situs ambiguous heterotaxy. The defect plasty was performed at the operation, and the complete venous cannulation required for artificial circulation was performed by the correctly chosen method of drainage of the superior venous system - through the auricle of the right atrium and the inferior venous system - through a separate hepatic vein cannulation, with a good clinical result.

https://doi.org/10.35805/BSK2023III001

Kuatbekov K.N.
orcid.org/0000-0002-2679-5097
Egizekov A.L.
orcid.org/0000-0001-7979-2239
Mishin A.V.
orcid.org/0000-0003-2858-6181
Musagaliev D.T.
orcid.org/0000-0002-1914-8449
Baizhigitov N.B.
orcid.org/0000-0001-5041-4277
Suieubekov B.Y.
orcid.org/0000-0001-9683-8366
Botabekov T.E.
orcid.org/0000-0003-0159-6515

Author for correspondence:
Mishin A.V. *– cardiac surgeon, center of modern medicine "Mediterra" Institute of Surgery LLP, Almaty, Kazakhstan, e-mail: avm_mishin@mail.ru*

Conflict of interest:
The authors declare that they have no conflicts of interest

Keywords:
interrupted inferior vena cava, heterotaxy, case report, congenital heart disease, heart surgery

Төменгі қуысты көктамырының жүйелік көктамырлық қосылу аномалиясы мен қарыншааралық перденің ақауын түзетудің клиникалық жағдайы

Қуатбеков Қ.Н.[1], Егізеков А.Л.[1], Мишин А.В.[1], Мұсағалиев Д.Т.[1], Байжігітов Н.Б.[1], Сүйеубеков Б.Е.[1], Ботабеков Т.Е.[2]

[1]Зманауи медицина орталығы «Медитерра» «Хирургия институты» ЖШС, Алматы, Қазақстан,
[2]«С.Ж. Асфендияров атындағы Қазақ Ұлттық медицина университеті» КеАҚ, Алматы, Қазақстан

Аңдатпа

Төменгі қуысты көктамырдың үзілісі - бұл сирек кездесетін ауру, ол оқшауланған, аспления немесе полиспления синдромдарымен бірге пайда болуы мүмкін. Жүйелік тамырлардың қалыптан тыс дамуы жүрекшелік situs-пен тығыз байланысты. Левокардияда висцеральды ситус деп аталатын іш қуысының инверсиясының белгілері бар. Бұл жұмыста қарыншааралық перденің үлкен ақауы бар емшектегі нәрестенің клиникалық жағдайының сипаттамасы, төменгі қуыс көктамырдың үзілуімен және жалғасы azygos венасы висцеральды гетеротаксиямен situs ambiguous. Операцияда ақау пластикасы жасалады, ал жүрек-өкпе айналымы кезінде қажетті толық көктамырлық канюляция жоғарғы көктамырлы жүйені дренаждаудың дұрыс таңдалған әдісімен – оң жақ жүрекшенің құлақшасы және төменгі көктамырлық жүйе арқылы – бауыр көктамырының жеке канюляциясы арқылы, жақсы клиникалық нәтижемен жүзеге асырылады.

Хат алысатын автор:
Мишин А.В. *– дәрігер-кардиохирург, заманауи медицина орталығы «Медитерра» «Хирургия институты» ЖШС, Алматы қ., Қазақстан, e-mail: avm_mishin@mail.ru*

Мүдделер қақтығысы:
Авторлар мүдделер қақтығысының жоқтығын мәлімдейді

Түйінді сөздер:
төменгі қуысты көктамырдың үзілісі, гетеротаксия, клиникалық жағдай, туа біткен жүрек ақаулары, жүрек хирургиясы

Клинический случай коррекции дефекта межжелудочковой перегородки в сочетании с аномалией системного венозного соединения нижней полой вены

Куатбеков К.Н.[1], Егизеков А.Л.[1], Мишин А.В.[1], Мусагалиев Д.Т.[1], Байжигитов Н.Б.[1], Суйеубеков Б.Е.[1], Ботабеков Т.Е.[2]

[1]Центр современной медицины «Mediterra» ТОО «Институт хирургии», Алматы, Казахстан,
[2]НАО «Казахский Национальный медицинский университет имени С.Д. Асфендиярова», Алматы, Казахстан

Мишин А.В. – врач-кардиохирург, центр современной медицины "Mediterra" ТОО "Институт хирургии", г. Алматы, Казахстан, e-mail: avm_mishin@mail.ru

Конфликт интересов:
Авторы заявляют об отсутствии конфликта интересов

Аннотация

Перерыв нижней полой вены – редкое заболевание которое может встречаться как изолированно, так и в сочетании с синдромами аспления или полиспления. Аномальное развитие системных вен тесно связано с предсердным situs. При левокардии обнаруживаются признаки инверсии брюшных органов, которое называют висцеральным ситусом. В настоящей работе представлено описание клинического случая грудного ребёнка с большим дефектом межжелудочковой перегородки в сочетании с перерывом нижней полой вены и продолжением в вена azygos висцеральной гетеротаксией situs ambiguous. На операции выполнена пластика дефекта, а необходимая при искусственном кровообращении полная венозная канюляция осуществлена правильно выбранным способом дренирования верхней венозной системы – через ушко правого предсердия и нижней венозной системы – через отдельную канюляцию печёночной вены, с хорошим клиническим результатом.

Ключевые слова:
перерыв нижней полой вены, гетеротаксия, клинический случай, врождённые пороки сердца, операция на сердце

Introduction

Inferior vena cava tear (IVC) is a rare disorder occurring in isolation or in combination with asplenia or polysplenia syndromes [1]. The subhepatic part of the IVC is absent, indicating a violation of the fusion of the yolk and subcardinal fetal parts of the IVC; it is replaced by a dilated unpaired or semi-unpaired vein that continues into the thorax, either into the superior vena cava or into the brachiocephalic veins.

Cardiac systemic venous return of blood is normally through the vena cava and coronary sinus. The most important anomalies of the vena cava appear to be an extra left superior vena cava (LSVC) and rupture of the LSVC with unpaired continuation, sometimes draining into the left atrium (LA) with clinical cyanosis. In all variants there is total abnormal drainage of hepatic veins directly into the right atrium (RA) or into the LA. In 60% of cases there are bilateral superior vena cava (SVC) draining into bilateral morphologically left atria. LSVC can drain into the coronary sinus. In 90% of patients there is an unpaired continuation of the IVC into the SVC. In situs inversus, the systemic venous return is a mirror image of the normal one. Unpaired continuation of IVC in left atrial isomerism significantly influences tactical and technical decisions in hemodynamic correction of unifocalizing malformations. Among many IVC anomalies, double and left IVC occur with the frequency of 3% and 0.5%, respectively. In patients with double IVC, the right and left IVC ascend from the iliac veins on both sides of the spine to the level of the hepatic veins. These anomalies are technically important for a surgeon.

The most significant anomalies of IVC are subhepatic rupture of IVC with unpaired continuation and abnormal drainage of IVC into the left atrium, causing cyanosis. The rupture of the IVC with unpaired continuation being the most frequent anomaly of the IVC occurs in 0.6% of patients with congenital heart disease (CHD) and usually with left atrial isomerism. The hepatic part of IVC is absent, below the level of renal veins it is normal. Without taking the hepatic veins, the IVC is drained instead of the RA through the dilated unpaired vein into the right superior vena cava (RSVC) or occasionally into the RA. The hepatic veins drain directly to the RA. Neparate continuation of IVC is often combined with complex cyanotic CHD, less often with simple malformations, it is not registered in asplenia syndrome. This defect creates difficulties during cardiac catheterization and can complicate surgical correction of the underlying malformation. A wide RSVC is a sign of an unpaired continuation of the IVC into the SVC. This anomaly itself does not require correction. During surgery, the SVC should be cannulated with a wide catheter. It is necessary to avoid overlapping the site where the unpaired vein enters the SVC. When the IVC is drained through the left unpaired vein into the LSVC and the coronary sinus, cannulation of the vena cava system should be done through the coronary sinus. The LSVC can also be directly cannulated. When cannulation of abnormal systemic veins is technically difficult, hypothermic perfusion with a single

venous cannula in RA is advisable. Direct venous cannulation through the opened RA can be performed during hypothermic arrest of the artificial circulation. Abnormal drainage of hepatic veins can be partial or complete. Normally, veins from both lobes of the liver drain into the IVC. Partial drainage of hepatic veins directly into the right atrium is most common in right atrial isomerism, but also in situs solitus or situs inversus. Cases of partial drainage into the coronary sinus have been described. Total abnormal hepatic venous connection is observed in left atrial isomerism. Veins can drain into the heart in one or two trunks.

Abnormalities can be diagnosed on echocardiography (ECHO), CT, MRI [2]. Anomalies of hepatic vein inflow play a significant role in the immediate and long-term results of Fontan surgery, as they are not included in the pulmonary circulation. The positive effect consists in decompression of the systemic venous return, which is manifested by reduced duration and intensity of pleural exudation. The negative long-term effect consists in the development of intrahepatic venous connections and intrapulmonary fistulas, leading to increased right-left shunting. Interrupted inferior vena cava with azygotic continuation is associated with cardiovascular and extracardiac anomalies, the most frequent anomalies being cardiac malformations and visceral heterotaxy [3, 4]. Angiographic confirmation of the anatomy of the unpaired system is important when planning surgical correction of patients with left isomerism [5].

Case study

A one-year-old 9 kg baby was admitted to our clinic on 11.30.2022 with the diagnosis: Situs ambiguous. VSD. PFO. Interrupted inferior vena cava with azygous continuation. PH., with complaints of dyspnea and rapid fatigue when walking. Basic diagnostic tests were performed: ECG, ECHO, catheterization cardiopulmonoangiography. The defect was confirmed by ECHO data: Levocardia. Situs ambiguous. Left-sided location of the liver. IVC was not dilated, a break above the confluence of the renal veins with continuation in the unpaired vein and SVC. SVC in the RA, not dilated. LA 1.8 cm. The pulmonary veins flow into the LA. Valve apparatus, coronary arteries without pathology. Aorta: 1,1 cm. PA 1.2 cm; right branch 0.9 cm, left branch 0.8 cm. RV 73%. Average pressure in the RV was 25 mmHg. Interventricular septum: perimembranous aneurysm, aneurysm gate diameter 0.8 cm with defects of 0.2 cm and 0.36 cm. Interatrial septum: defect with a defect closer to the mouth of the PFO 0.1 cm. Cardiac cavities: left parts of the heart are moderately dilated.

To clarify the cardiac anatomy and extracardiac vascular structures, we performed inferior cavography, angiopulmonography, tonometry from the pulmonary artery trunk: the common femoral vein was catheterized with the following pressure records in mmHg: in the LA trunk 25/11, mean 17; in the RV 25/16 mean 14; in the RA 11/4. There is patency throughout the entire length of the IVC, and there is an infiltration of the IVC into the SVC (Figure 1). Angiopulmonography: the trunk and branches of the PA are of normal diameter, passable, the end branches of the PA can be traced throughout (Figure 2), the parenchymal phase is uniformly enhanced on both sides. The left and right pulmonary veins are drained into the LA in the phase of the levogram. In the phase of the left-side diagram, there is an intensive discharge of contrasting blood from the LA cavity into the RA cavity due to the ASD.

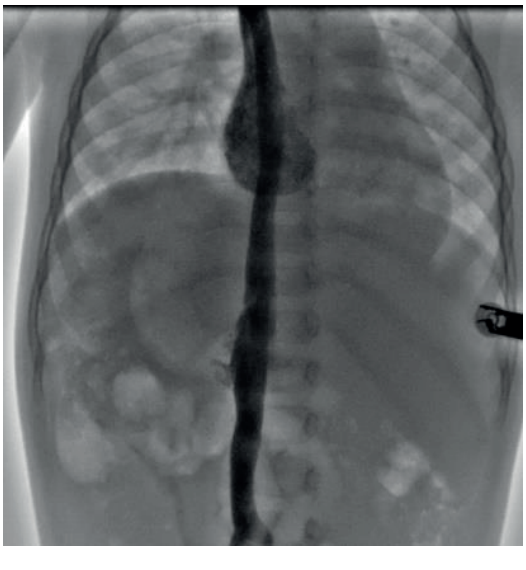
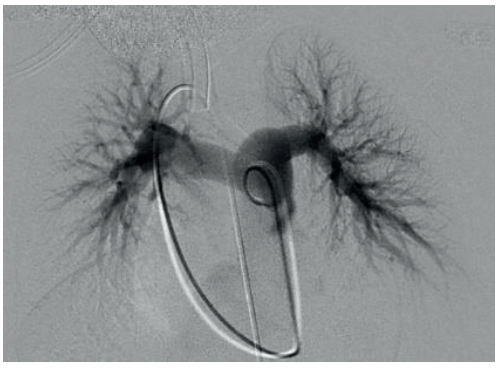


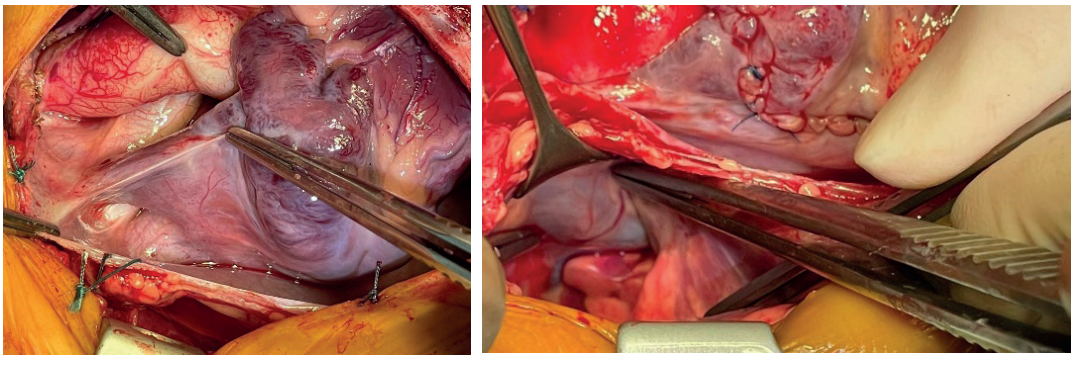
Figure 1.
Angiogram. Interrupted inferior vena cava with azygous continuation

Figure 2.
Angiogram. The trunk and branches of the PA



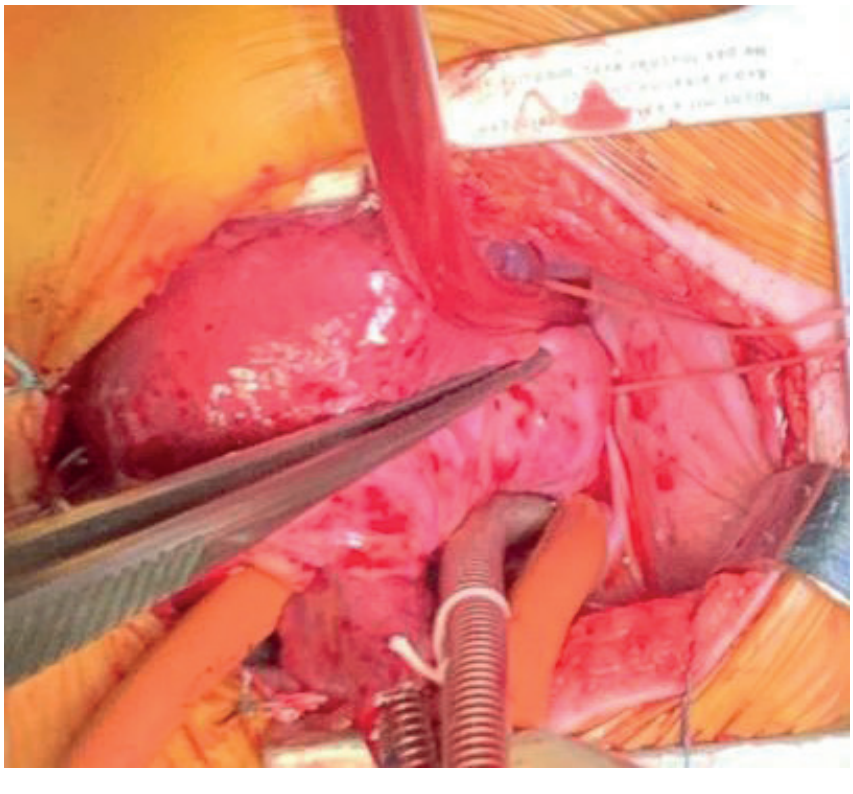
12/09/2022 a planned surgical correction was performed under the conditions of cardiopulmonary bypass: VSD plasty with an autopericardial patch, suturing PFO. Intraoperatively: The right heart sections were enlarged in size. The PA trunk was identified as Ao in diameter, unstressed, the vena azygos (Figure 3A) was dilated, on the right pleural cavity side there was an infiltration of the IVC - rupture of the IVC with azygos-continuation (unpaired continuation) into the SVC (Figure 3B), the hepatic vein mouth was identified in the place of its normal location in the RA.

Figure 3.
Intraoperative photo:
A - IVC dilated at the site of vena azygos inflow;
B - interrupted inferior vena cava with azygous continuation



The aorta was cannulated, separate cannulation of IVC through the RA ear, separate cannulation of the hepatic vein (Figure 4) with access to complete artificial circulation.

Figure 4.
Intraoperative photo.
Cannulated hepatic vein



Standard VSD with autopericardial patch was performed. There were no complications, the patient was admitted to the Intensive Care Unit. On control ECHO: interventricular septum was tight, left ventricular ejection fraction was 71%. First degree tricuspid valve insufficiency. The right parts of the heart were moderately dilated. No fluid was detected in the pericardium. On the first postoperative day the child was transferred to a specialized department, a week later he was discharged.

Discussion

Abnormal development of systemic veins is closely related to atrial situs. In situs solitus and situs inversus, the spectrum of anomalies is limited and predictable. The coronary sinus is always present, and the probability of significant venous return anomalies is low. However, in atrial heterotaxy and isomerism, there are marked abnormalities of atrial anatomy (common atrium) and venous return. In these conditions they are frequent, bizarre, but relatively standard depending on situs. If there is normal levocardia, no obvious CHD and at the same time there are signs of abdominal organ inversion, this condition in practice is also called visceral situs, assuming that the inversion should be complete, mirror, by the type of isolated dextrocardia. In visceral situs, even in the absence of obvious CHD, there is a significantly increased risk of abdominal heterotaxy and, accordingly, of such formidable complications as biliary atresia, gastrointestinal stenosis and atresia, incomplete bowel turn, intestinal volvulus [6].

In our case, the child in combination with CHD showed levocardia, situs ambiguous with inversion of internal organs (liver on the left). Interrupted inferior vena cava with azygous continuation was determined during the operation, it was decided to drain the superior venous return through the RA ear, and considering a satisfactory hepatic vein mouth flowing into the RA, we managed to cannulate it in isolation, which allowed us to perform total venous return of the body.

Conclusion

In our clinical case we encountered a rare anomaly of combined congenital heart pathology and interrupted inferior vena cava with azygous continuation of visceral situs ambiguous heterotaxy. The task we had to accomplish during VSD radical correction to drain the entire venous system as completely as possible during the artificial circulation was successfully achieved due to the correctly chosen method of drainage of the superior venous system - through the right atrial auricle and the inferior venous system - through separate cannulation of the hepatic vein, with a good clinical result.

References

1. Zafar SI, Halim A, Khalid W, Shafique M, Nasir H. Two Cases of Interrupted Inferior Vena Cava with Azygos/Hemiazygos Continuation. J Coll Physicians Surg Pak. 2022 Aug;32(8):S101-S103. doi: 10.29271/jcpsp.2022.Supp2.S101.
2. Batouty NM, Tawfik AM, Sobh DM, Razek AAKA. Computed tomography and magnetic resonance imaging of congenital thoracic systemic venous anomalies. Pediatr Radiol. 2023 Jan 5. doi: 10.1007/s00247-022-05570-w.
3. Xu Q, Sun HX, Xie JS, Wang JL, Pei QY, Zhang XH. Characteristics and prognosis of interrupted inferior vena cava with azygous continuation. Zhonghua Fu Chan Ke Za Zhi. 2018 Mar 25;53(3):149-154. doi: 10.3760/cma.j.issn.0529- 567X.2018.03.002.
4. Dang HQ, Tran MT, Nguyen NM, Le HT. Totally endoscopic atrial septal defect repair in a patient with interrupted inferior vena cava with azygos-hemiazygos continuation. J Card Surg. 2021 Dec;36(12):4814-4817. doi: 10.1111/jocs.16036.
5. Liu Y, Guo D, Li J, Zhang X, He J, Huang M, Dai J, Cai H. Radiological features of azygos and hemiazygos continuation of the inferior vena cava: A case report. Medicine (Baltimore). 2018 Apr;97(17):e0546. doi: 10.1097/MD.0000000000010546.
6. Комарова И.В., Никифоренко А.А., Гартунг Т.С., Винокурова Е.А. Левопредсердный изомеризм как вариант висцерального ситуса: анализ случая пренатальной диагностики с относительно благоприятным исходом и проблемы верификации диагноза после рождения. Пренатальная диагностика. 2017. Т.16, №4, С.340-346. [Komarova I.V., Nikiforenko A.A., Gartung T.S., Vinokurova E.A. Levopredserdnyj izomerizm kak variant visceral'nogo situsa: analiz sluchaya prenatal'noj diagnostiki s otnositel'no blagopriyatnym iskhodom i problemy verifikacii diagnoza posle rozhdeniya. Prenatal'naya diagnostika. 2017. T.16, №4, S.340-346.]