Received: 02 April 2023 / Accepted: 28 May 2023 / Published online: 30 June 2023

DOI 10.34689/SH.2023.25.3.034

UDC 616.124.3-053.88

CLINICAL CASE OF A SINGLE RIGHT VENTRICLE IN A 52-YEAR-OLD FEMALE PATIENT

Svetlana Onichshuk[1], Assel Chinybayeva[2], Bolat Ashirov[3], Jamilya Mansurova[1*], Ludmila Karazhanova[1]

[1] Semey Medical University, Department of Therapy, Semey, Republic of Kazakhstan;
[2] Corporate Fund University Medical Center "Republican Diagnostic Center", Astana, Republic of Kazakhstan;
[3] South Kazakhstan Medical Academy, Department of Therapy and Cardiology, Shymkent, Republic of Kazakhstan.

Summary

A rare severe congenital heart disease is the single ventricle of the heart (SV), which is characterized by the presence of atria connected to SV. This publication describes a clinical case of a middle-aged patient with SV without surgical palliative operations.

This clinical case is unique it demonstrates such a gross congenital heart defect as SV and a large defect of the atrial septum, practically a "two-chamber heart", with mitral valve atresia, transposition of the main vessels, valvular and subvalvular pulmonary artery stenosis in combination with cardiac arrhythmia.

Keywords: *congenital heart disease, single ventricle of the heart, chronic heart failure, echocardiography.*

Резюме

ЕДИНСТВЕННЫЙ ПРАВЫЙ ЖЕЛУДОЧЕК У ПАЦИЕНТКИ 52 ЛЕТ. КЛИНИЧЕСКИЙ СЛУЧАЙ

Светлана Онищук[1], Асель Чиныбаева[2], Болат Аширов[3], Джамиля Мансурова[1*], Людмила Каражанова[1]

[1] НАО «Медицинский университет Семей», Кафедра терапии, г. Семей, Республика Казахстан;
[2] Корпоративный фонд Университетского медицинского центра «Республиканский диагностический центр», г. Астана, Республика Казахстан;
[3] Южно-Казахстанская медицинская академия, Кафедра терапии и кардиологии, г. Шымкент, Республика Казахстан.

Редким тяжелым врожденным пороком сердца является единственный желудочек сердца (ЕЖС), который характеризуется наличием предсердий, соединенных с ЕЖС. В данной публикации описывается клинический случай пациента среднего возраста с ЕЖС без хирургических паллиативных операций.

Этот клинический случай уникален, он демонстрирует такой грубый врожденный порок сердца, как ЕЖС с большим дефектом межпредсердной перегородки, практически "двухкамерное сердце" с атрезией митрального клапана, транспозицией магистральных сосудов, клапанным и подклапанным стенозом легочной артерии в сочетании с трепетанием предсердий.

Ключевые слова: *врожденный порок сердца, единственный желудочек сердца, хроническая сердечная недостаточность, эхокардиография.*

Түйіндеме

52 ЖАСТАҒЫ НАУҚАСТЫҢ ЖАЛҒЫЗ ОҢ ҚАРЫНШАСЫ. КЛИНИКАЛЫҚ ЖАҒДАЙЫ

Светлана Онищук[1], Асель Чиныбаева[2], Болат Аширов[3], Джамиля Мансурова[1*], Людмила Каражанова[1]

[1] «Семей медицина университеті» КеАҚ, Терапия кафедрасы, Семей қ., Қазақстан Республикасы;
[2] "Республикалық диагностикалық орталық" университеттік медициналық орталығының корпоративтік қоры "Республикалық диагностикалық орталық" университеттік медициналық орталығының корпоративтік қоры, Астана қ., Қазақстан Республикасы;
[3] Оңтүстік Қазақстан медицина академиясы, Терапия және кардиология кафедрасы, Шымкент қ., Қазақстан Республикасы.

Сирек ауыр туа біткен жүрек ақауы - жүректің жалғыз қарыншасы (ЖЖҚ) болып табылады, ол ЖЖҚ - мен байланысқан жүрекшелердің болуымен сипатталады. Бұл басылымда хирургиялық паллиативті отасыз ЖЖҚ бар орта жастағы науқастың клиникалық жағдайы сипатталады.

Бұл клиникалық жағдай бірегей, ол ауыр туа біткен жүрек ақауын көрсетеді, оған біріктіріледі ЖЖҚ -сы жүрекше аралық қалқанның үлкен ақауы, іс жүзінде митральды қақпақшаның атрезиясы бар "екі камералы жүрек", магистральды тамырлардың транспозициясы, атриальды діріл мен клапан және ішкіклапанның өкпе стенозы.

Түйінді сөздер: *туа біткен жүрек ауруы, жалғыз жүрек қарыншасы, созылмалы жүрек жеткіліксіздігі, эхокардиография.*

Bibliographic citation:

Onichshuk S., Chinybayeva A., Ashirov B., Mansurova J., Karazhanova L. Clinical case of a single right ventricle in a 52-year-old female patient // *Nauka i Zdravookhranenie* [Science & Healthcare]. 2023, (Vol.25) 3, pp. 275-278. *doi 10.34689/SH.2023.25.3.035*

Онищук С., Чиныбаева А., Аширов Б., Мансурова Д., Каражанова Л. Единственной правый желудочек у пациентки 52 лет. Клинический случай // Наука и Здравоохранение. 2023. 3 (Т.24). С. 275-278. *doi 10.34689/SH.2023.25.3.035*

Онищук С., Чиныбаева А., Аширов Б., Мансурова Д., Каражанова Л. 52 Жастағы науқастың жалғыз оң қарыншасы**.** Клиникалық жағдайы // Ғылым және Денсаулық сақтау. 2023. 3 (Т.25). Б. 275-278. *doi 10.34689/SH.2023.25.3.035*

Introduction:

The prevalence of CHD is 8-10 per 1000 live births. Of these, the incidence of single ventricular malformation varies according to clinical data from 0.2% to 3.2% of all congenital heart defects. A rare severe congenital heart disease (CHD) is the single ventricle of the heart (SV), which is characterized by the presence of atria connected to SV. Single ventricle usually includes hypoplasia syndrome of the left heart, tricuspid valve atresia and a single ventricle with a double inflow. The most favorable cases occur when the only ventricle is represented anatomically by the left ventricle in combination with separate and full-fledged atrioventricular valves, in the absence of stenosis of the main arteries and transposition. This variant of a single ventricle with a double inflow is called the "Holmes heart".

In patients with SV, the clinical picture is represented by dyspnea, cyanosis, rhythm disturbances, chronic heart failure, secondary erythrocytosis, the development of thrombosis and thromboembolic complications. However, even patients who have undergone surgery can rarely live to adulthood and most often die from progressive heart failure, cardiac arrhythmias and sudden cardiac death.

This publication describes a clinical case of a middle-aged patient with SV without surgical palliative operations, which will be interesting for acquaintance and discussion among doctors of such specialties as therapy, cardiology, cardiac surgery and arrhythmology

Case report:

In 2022 the 52-year-old woman patient turned to the emergency room of the Semey Emergency Hospital with complaints of shortness of breath at the slightest physical exertion, feeling of lack of air, weakness, swelling of the lower extremities. The patient is hospitalized in the department of chronic heart failure.

When collecting anamnesis and examination, it is possible to find out that the patient has a history of CHD, SV diagnosed in childhood, then surgical treatment was not performed, because of the high risk of perioperative complications exceeding the expected benefit. At the age of 25, she gave birth to a full-term, healthy baby by C-section. In 2014, by the decision of a multidisciplinary consultation, taking into account the prescription of CHD, age, subcompensated condition of the patient, it was recommended to continue drug therapy with ACE-inhibitors, MRAs, diuretics, antiplatelet agents. In 2018, the patient develops atrial flutter, and anticoagulants are added to the treatment, but the patient has a low compliance to therapy. At the age of 52, she suffered a minor ischemic stroke.

Objective examination: acrocyanosis, the chest is not deformed, the boundaries of the heart are expanded, rough systolic noise is heard at all points of auscultation, rhythmic tones. In laboratory studies: secondary erythrocytosis, an increase in the level of pro-BNP 12 647.8 pg/ml.

The changes on the ECG and Chest X-ray is presented in Fig. 1.

On Echocardiogram (Fig.2): SV, single atrium, large defect of the atrial septum 37 mm wide, rudimentary left ventricle, single atriovetricular valve, trunkus insufficiency 1.5+, mitral valve atresia, supravalvular moderate aortic stenosis, pulmonary artery stenosis, contractility of the myocardium of the SV is reduced. The ejection fraction is 52%. The average pressure gradient on the pulmonary artery is 33 mm Hg

The patient received in-hospital treatment according to the latest CHF therapy recommendations of the European Society of Cardiology, including loop diuretics, calcium channel blockers, beta blockers, SGLT2 inhibitors, sacubitril/valsartan, given the high risk of thrombosis - vitamin K antagonists, and pulmonary hypertension therapy. During her stay in the hospital, the patient's condition improved. Her exercise tolerance has increased, shortness of breath has disappeared at rest, swelling of the lower extremities has significantly decreased.

The patient was discharged from the hospital with recommendations and is currently under the supervision of a cardiologist at the polyclinic

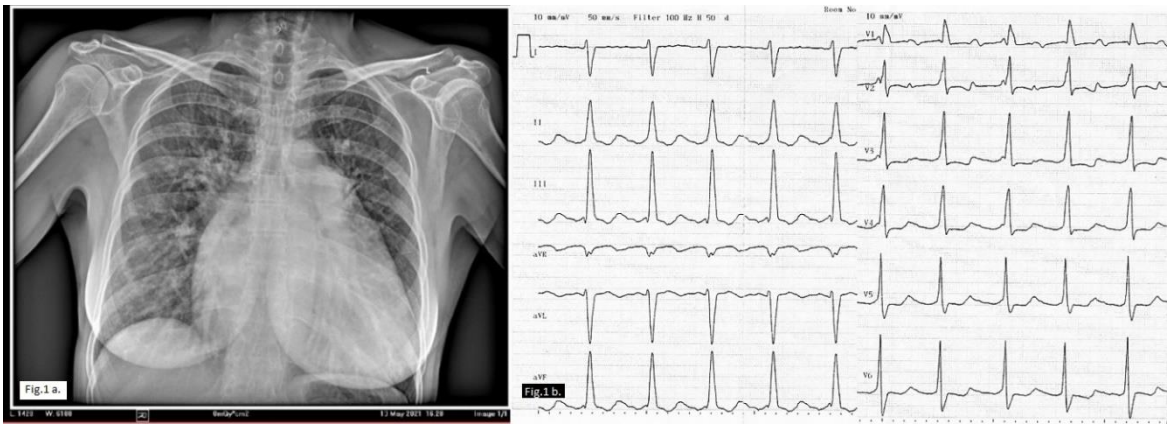

Figure 1 ECG and Chest X-ray.

Fig. 1a. - Plain chest x-ray in direct projection at the time of admission. An increase in the lung pattern and its enrichment in the lower sections were revealed. The heart is expanded in diameter, mitral configuration, cardio-thoracic index - 70%. atherosclerosis of the aorta.

Fig. 1b. - The patient's ECG at admission demonstrates the correct form of atrial flutter with a ventricular rate of 2:1. The axis of the heart is deviated to the right. Complete blockade of the right leg of the bundle of His.

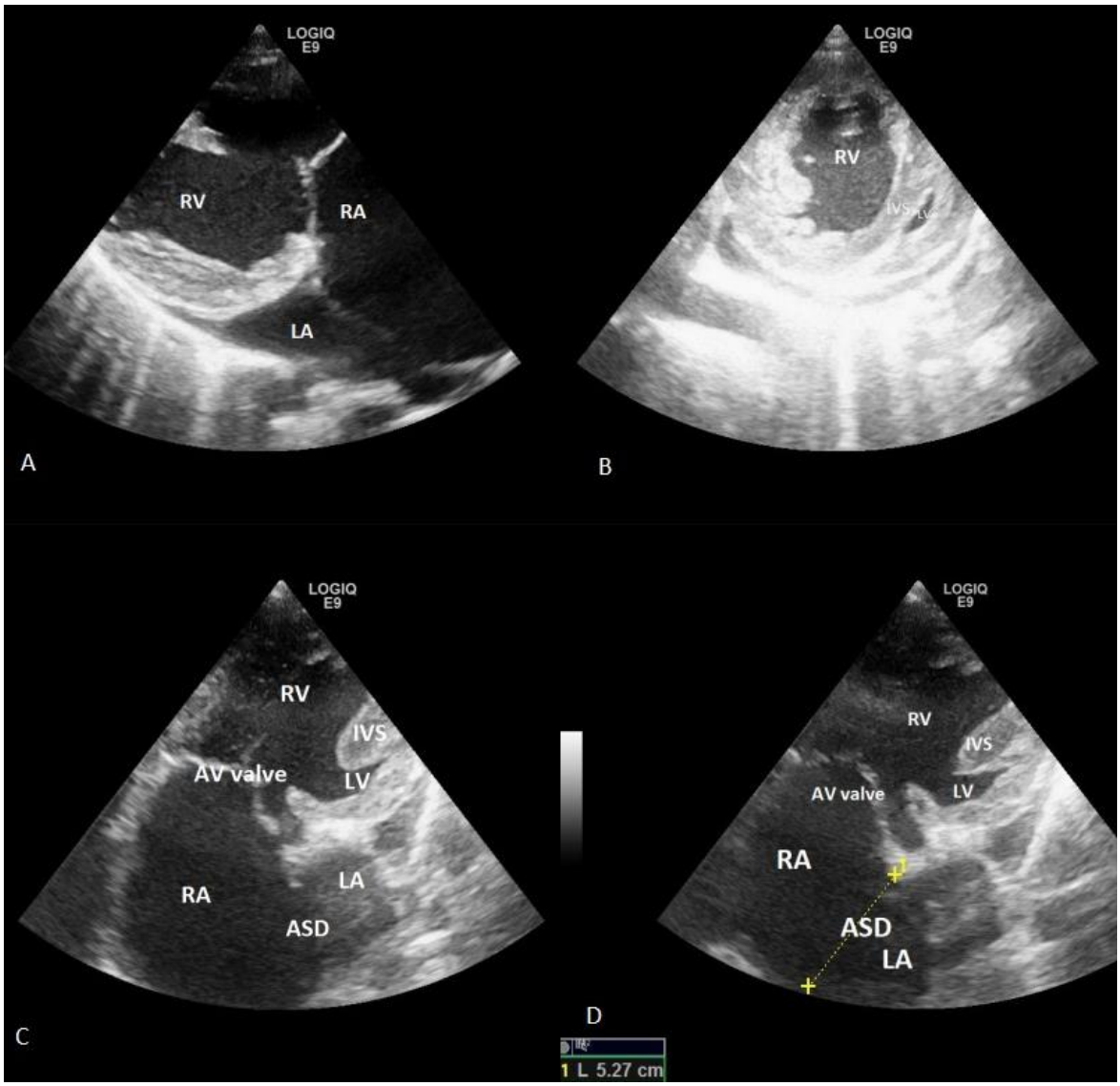

Figure 2. Echocardiography.

A - Parasternal long axis position; **B** - Parasternal position along the short axis;
C - Apical four-chamber position; **D** - Apical four-chamber position size of the atrial septal defect (ASD).
RV - right ventricle, **LV** - left ventricle, **RA** - right atrium, **LA** - left atrium,
IVS - interventricular septum, **AV** valve - atrioventricular valve.

Discussion:

SV is a defect that has an unfavorable course with mortality at an early age [4; 7]. According to literature sources, the survival rate of patients with SV without surgery is 6-7%, if patients underwent surgery at an early age, then 85% of children born with a dominant left ventricle survive to adulthood, and children with a dominant right ventricle in 65% of cases. [1; 5; 8; 9].

SV refers to the defects of blue color, therefore, the clinical picture should be dominated by the characteristic symptoms of cyanosis, shortness of breath, lag in physical and mental development, secondary erythrocytosis, thromboembolic complications and other typical symptoms [6]. Although the prognosis is unfavorable, according to literature data, if the blood circulation is "well balanced", then the patient has the opportunity to live to adulthood with a relatively small number of symptoms. Usually, a positive flow occurs in patients in whom the SV is anatomically left and the inflow into it is carried out through separate, properly formed mitral and tricuspid valves, and the outflow occurs in normally located main arteries. It is known that the survivability of individuals with pulmonary artery stenosis is higher due to the slower development of stagnation in the small circle of blood circulation [4].

Conclusion:

This clinical case is unique it demonstrates such a gross congenital heart defect as SV and a large defect of the atrial septum, practically a "two-chamber heart", with mitral valve atresia, transposition of the main vessels, valvular and subvalvular pulmonary artery stenosis in combination with cardiac arrhythmia. As is known, pulmonary stenosis is of decisive importance in the hemodynamic balance of blood flow in patients with SV. This is due to the fact that it restricts blood flow through the pulmonary vessels and prevents their damage, it also limits the flow of blood from left to right, thereby also preventing an increase in pulmonary blood flow and overloading the ventricle. Patients with SV who are adequately oxygenated and have a balanced ventricular load have a chance of long-term survival, as observed in our patient. Interesting is the fact of carrying a pregnancy to full term, and the birth of a healthy child [2; 3]. In the literature, such cases are rare, the patients were under strict control during the entire pregnancy, their condition worsened, accompanied by complications from the mother and child. The optimal anatomical structure of the heart for patients with SV is: normal morphology of the left ventricle, transposition of large arteries, no significant obstruction of the outflow tract, full functioning of the atrioventricular valves, and moderate pulmonary stenosis. These anatomical features allow patients to experience mild to moderate symptoms and have a good quality of life.

Informed Consent: *Written informed consent was taken from the patient for publication of this case report and accompanying images.*

Contribution of authors. *All authors were equally involved in the writing of this article.*

Conflict of Interest. *The authors declare no conflicts of interest*

Funding: *No funding was provided.*

References:

1. *Ambastha C., Bittle G.J., Morales D. [et al.]* Regenerative medicine therapy for single ventricle congenital heart disease // Translational Pediatrics. 2018. Vol.7. №2. P.176-87.

2. *Canobbio M.M., Warnes C.A., Aboulhosn J. et al.* Management of Pregnancy in Patients with Complex Congenital Heart Disease: A Scientific Statement for Healthcare Professionals from the American Heart Association // Circulation. 2017. Vol.135. №8. P.e50-e87.

3. *Delsol-Gyan D., Aniteye E., Oppong S. et al.* Pregnancy in non-palliated functionally single ventricle: Challenges of management in resource-poor settings // Pan African Medical Journal. 2020. Vol.35. P.6.

4. *Garcia A.M., Beatty J.T., Nakano S.J.* Heart failure in single right ventricle congenital heart disease: Physiological and molecular considerations // American Journal of Physiology - Heart and Circulatory Physiology. 2020. Vol.318. №4. P.H947-H965.

5. *Goldberg H.L., Sniderman K., Devereux R.B., Levin A.* Prolonged survival (62 years) with single ventricle / // The American Journal of Cardiology. 1983. Vol.52. №1. P.214-15.

6. *Hallidie-Smith K.A., Webb-Peploe M.M.* Presentation and prognosis of single ventricle // British Heart Journal. 1972. Vol.34.№9. P.962.

7. *Karaduman B.D., Bayram H., Kasapkara H.A.et al.* Long-term survival in a case of unoperated single ventricle // Turk Kardiyoloji Dernegi Arsivi. 2016. Vol.44. №4. P.338-41.

8. *McConville P.* Toward a phenomenology of congenital illness: a case of single-ventricle heart disease // Medicine, Health Care and Philosophy. 2021. Vol.24. P.587–95.

9. *Tseng S.Y., Siddiqui S., Di Maria M.V. et al.* Atrioventricular valve regurgitation in single ventricle heart disease: A common problem associated with progressive deterioration and mortality / // Journal of the American Heart Association. 2020. Vol.9. №11. P.e015737.

Corresponding Author:
Jamilya Mansurova, PhD, MD, cardiologist, Semey Medical University
Mailing address: 99A – 118, Mamay Batyr st., Semey, Kazakhstan, 071402
E-mail: mansurova_dzhami@mail.ru
Mob: 8 777 330 51 04