Successful surgical treatment of yatagan syndrome in a child

A. A. Sloboda, Y. P. Truba, I. V. Dziuryi, V. V. Lazoryshynets
Amosov National Institute of Cardiovascular Surgery, Kyiv

Scimitar syndrome is a congenital heart disease characterised by anomalous drainage of the right pulmonary veins into the inferior vena cava (IVC) or various venous connections from the anomalous pulmonary vein to the IVC system. This is an infracardal form of partial anomalous pulmonary vein drainage (PAPVD) [1]. Drainage of blood from the right lung by the so-called scimitar vein varies from partial to complete [2].

The term "yataghan syndrome" refers to this congenital heart defect based on the characteristic shadow on the chest radiograph (CR) created by the abnormal flow of the right pulmonary veins into the RV system, which resembles a Turkish weapon – a yataghan [1, 3].

Yataghan syndrome occurs in two per 100,000 newborns [3]. It was first described in 1836 [2]. This syndrome is often combined with hypoplasia of the right lung, bronchus and pulmonary artery. The most common findings are heart dextrops with mediastinal displacement, absence of pericardium and/or absence of pleural sheets, and other malformations [4, 5].

Clinically, this form of ARF can manifest itself in different ways: frequent respiratory diseases, cyanosis, episodes of fainting, and dyspnoea [5]. Diagnosis of this syndrome is based on several methods. Usually, an X-ray of the chest can reveal a blackout characteristic of yatagana syndrome. Echocardiography allows to assess structural and functional changes, severity of pulmonary hypertension. However, three-dimensional CT or magnetic resonance imaging is of great importance in the diagnosis of this syndrome, as they are able to visualise an abnormal pulmonary vein and detect concomitant morphological pathology [6].

There are many variants of abnormal drainage of the pulmonary veins into the PFO or into the veins of the PFO system. Depending on this, several methods of correction of yatagana syndrome have been described in the literature, the main essence of which is reimplantation of the pulmonary veins into the PF. Implantation can be performed directly by suturing the pulmonary vein into the LV or by using vascular prostheses to form an anastomosis between the abnormally drained pulmonary vein and the LV [7]. The choice of surgical treatment tactics is primarily determined by anatomical features [8, 9]. The anatomical course of the so-called scimitar vein, the place of its drainage into the IJV, the presence of interatrial communication and other intracardiac anomalies determine the choice of surgical intervention. Here is a clinical observation.

Patient A., 6 years old, often suffered from respiratory inflammation, in particular bronchitis occurred on average 3 times a year, and suffered pneumonia in the third year of life. She complained of shortness of breath during physical activity.
Chest radiography. A shadow specific to yatagana syndrome was detected in the right lung field from the apex of the heart to the level of the IV rib with signs of atelectasis of the lower lobe of the right lung and possible dextrocardia (Fig. 1).

Echocardiography data. Situs solitus. Dextrocardia (left-sided right heart). Moderate dilatation of the right heart. Systolic pressure in the right ventricle (RV) 26 mm Hg. Pulmonary artery trunk diameter 19 mm, pressure gradient in the RV outflow tract 6 mm Hg. Indirect signs of PADLV. Pulsatility of blood circulation in the abdominal aorta. Left ventricular end-diastolic volume (LVEDV) 57 ml, LV end-diastolic index 64.7 ml/m², LV end-systolic volume 18 ml, LV stroke volume 39 ml, LV ejection fraction (EF) 69%.

Computed tomography (CT) scan of the PFO (Fig. 2): abnormal drainage of the upper right pulmonary vein into the ILD below the right atrium (RA) by 10 mm and the lower right pulmonary vein, which also flows into the upper part of the ILD. The oblique interlaminar, horizontal visceral lamina propria of the right pleura is not clearly visualised. The BVI of the right lung is represented by a short segment 3 mm long, subsegmental atelectasis of the CVII of the right lung. Pneumatisation of the rest of the lung parenchyma is preserved.

Tactics of surgical treatment

We performed the operation with artificial circulation. During the operation, the right pleural cavity was opened and the right pulmonary veins were isolated, with the following anatomical features: the upper right pulmonary vein entered the IJV to the right below the PP by 1 cm, and the lower right pulmonary vein was drained into the upper part of the IJV by a separate mouth (Fig. 3).

General cooling of the patient to 28 °C. Pharmacocold cardioplegia into the aortic root. Right atriotomy. The left atrium (LA) was drained through the atrial septal defect. The right pulmonary veins were cut off from the RV, and their distal ends were sutured. The pulmonary veins were connected by making a side-to-side anastomosis. The LV wall was dissected. An anastomosis was formed: the posterior wall of the pulmonary vein and the posterior wall of the LV, the roof of the collector was formed with an autopericardial patch (Fig. 4).

Plastic surgery of a secondary atrial septal defect with an autopericardial patch.

The duration of aortic clamping was 275 minutes, the perfusion time was 431 minutes, and the operation lasted 11 hours.

In the postoperative period, transudate was discharged for 10 days, mostly from the right pleural cavity, where the so-called jugular veins were isolated and a collector was formed. Support with mimetics: norepinephrine, dobutamine, dexdor, symdax with a gradual decrease in dosage and quantity. The patient spent 134 hours in the intensive care unit.

Six months later, the patient was examined: the general condition was good, active, and dyspnoea during physical activity disappeared. Auscultation: vesicular breathing, without peculiarities. Echocardiography: dextrocardia, residual enlargement of the right heart. LVEF 70%, mild tricuspid valve insufficiency. No fluid was found in the pericardial cavity and pleural cavities.

Conclusions

Yataghan syndrome is a very rare condition with a number of symptoms and symptom complexes. A plain radiograph of the OHP can help to establish the diagnosis, so a careful evaluation of the radiographs is very important. A CT scan plays a crucial role in the diagnosis of this syndrome. Timely diagnosis is very important, as it reduces the risk of chronic respiratory disorders when performing surgical correction.

Surgical treatment in the described observation consisted of radical correction of infracardiac type of CHADLV and
atrial septal defect repair with good immediate and long-term results.

Perioperative management included the use of adrenomimetic drugs with a gradual decrease in doses. The patient recovered quickly.

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**References**


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