We report a case of progressive carcinoid heart disease having caused readmission of a patient after excision of endocardium with arterial intima and multiple valve replacement. The data prove that in cases of combined valve heart disease of the right side it is necessary to assume the high probability of the carcinoid disease. Therefore, in spite of the high value of echocardiography, multispiral computed tomography, confirming the diagnosis, detecting metastases in different organs and determining the localization of the primary tumor, becomes particularly important. Open heart surgery can be effective only when accompanied by operative therapy for the primary tumor with the use of somatostatin analogues.

**Keywords:** carcinoid heart disease, excision of endocardium and arterial intima, multiple artificial heart valves, echocardiography, multispiral computed tomography of the heart.
shortness of breath at rest, a tendency to hypotension, intermittent "hot flashes" in the face and neck, leg swelling, weakness. She had considered herself as sick since 2009. The sharp deterioration — in April 2012. Single telangiectasia on the face. Severe pulsation of neck vessels. RR — 20 breaths per minute. Vesicular breathing, no wheezing. Heart sounds — rhythhmal, systolic murmur along the right edge of the sternum. HR — 80 heart beats per min. BP — 100/80 mmHg. Abdomen was soft and painless. Liver at the costal margin, painless. Normal stool. Urination — painless.

Echocardiography on 13.04.12: the aortic root — 2.9 cm, the left atrium (LA) — 3.7 cm, the right atrium (RA) — 4.2 cm; the left ventricle (LV): end-systolic volume (ESV) — 19 ml; end-diastolic volume (EDV) — 53 ml; ejection fraction (EF) — 64%. The thickness of the interventricular septum (IVS) — 1 cm posterior wall thickness — 1 cm. The size of the right ventricle (RV) — 4.2 cm Aortic (AV) and mitral (MV) valves without pathology. TV leaflets were thin with violation of their coaptation, regurgitation grade 3. No signs of the PA trunk narrowing. Diameter of the PA at the level of the valve — 2.8 cm, above the valve — 2.6 cm, the outflow tract (OT) of the RV — 2.5 cm. OT: indistinct intimal thickening with a floating structure of up to 6 mm, 8 mm beneath leaflets of the PA valve. The maximum gradient on the PA valve: 36—41 mmHg.

Coronary angiography on 13.04.12: no significant stenoses of the coronary arteries.

Doppler ultrasound (DU) of brachiocephalic arteries (BCA) on 12.04.12: atherosclerosis of BCA without hemodynamically significant stenosis.

MSCT with pulmonary angiography on 17.04.12: the heart has increased in size due to the right side; the RA— 6.5x5.5 cm; end-diastolic RV size of up to 4.5 cm; hypertrophy of the RV free wall spreading to the PA valve zone with the presence of funnel-shaped narrowing of up to 12 mm; the outflow tract of the RV and the initial parts of the PA had the form of a long channel with unevenly thickened walls (Fig. 1); the diameter of the right branch of PA — 20 mm, left — 17.5 mm. Conclusion: the symptoms of stenosis of the RV outflow tract, extended infundibular stenosis of the PA ostium, dilatation of the right heart chambers with severe hypertrophy of the RV.

Surgical intervention with cardiopulmonary bypass (23.04.12). The even endocardial thickening of up to 5—7 mm was observed in the RA cavity, the RV endocardium had the same structure as the one of the RA, the PA intima was thickened up to 3—4 mm, in the ostium of the PA — to 1 cm, with narrowing of up to 1 cm in diameter, simulating infundibular stenosis. The changed part of endocardium, including interatrial septum, was dissected from the inner surface of the RA in a blunt way. The changed endocardium of the RV outflow tract was excised off the parietal ridge and the area of the PA valve, and in the same way we removed the changed intima from the trunk of the PA and its major branches up to the level of the lobar arteries. TV leaflets were thickened up to 5 mm, their mobility was limited, the annulus fibrosus diameter — 5 cm, papillary muscles were dramatically thickened. The TV leaflets were excised together with the altered papillary muscles tissue, the xenopericardial bioprosthesis was implanted. The PA valve was degeneratively changed, the leaflets were deformed. The PA valve leaflets were excised and the xenopericardial bioprosthesis was implanted. In IVS, closer to the top, a defect of 5—6 mm diameter with callous edges was revealed. The defect was sutured.

Histological test of the valve leaflets and endocardium revealed unevenly thickened intima of the PA due to severe fibrosis and active fibroplastic reaction (Fig. 2); partial destruction of the endocardial elastic frame with preserved elastic structures of the small vessels of the myocardium; complete destruction of the elastic structure of the PA wall.

The postoperative period was complicated by hypotension that required prolonged use of sympathomimetics, persistent diarrhea, Frederick syndrome. The "ECS-electrode" pacemaker was implanted on 18.05.12.

The check echocardiographic study of the LV on 12.05.12: EDV — 70 ml; ESV — 33 ml; EF — 52%. The RA — 4 cm; The RV — 3.4 cm. The peak gradient of the PA valve prosthesis — 3.5 mmHg, regurgitation — grade 0—1. Average diastolic gradient of the TV prosthesis — 2.4 mmHg, regurgitation into the cavity of the RA — grade 0—1, pulmonary artery systolic pressure (PASP) — 25 mm Hg. The fluid in the pericardial and pleural cavities was not revealed.

The patient was discharged on the 55th day after surgery with the diagnosis: “Endocardial thickening of RA and RV, thickening of the PA trunk and intima of its branches. Valvular and subvalvular stenosis of the PA. IVS defect. The excision of endocardium and intima from RA, RV, PA and its branches, suture of the IVS defect, plastics of the RV outlet, replacement of the TV and the PA valve. Complete atrioventricular block grade III. The implantation of permanent electro-cardiostimulator with epidural electrodes NK;A in May 2012”.

Re-admission to the Emergency Cardiology Department occurred on 03.06.13 with the diagnosis of "Prosthetic PA valve thrombosis." Upon arrival: the growing dyspnea at rest, leg swelling, weight loss of 10 kg, transient series of pain in the right iliac region, spontaneous flushing of the face and neck, cough with scanty mucous expectoration, pain in the spine with limitation of motion. Puffy face, telangiectasia. Severe pulsation of neck vessels. Peripheral lymph nodes were not palpable. BR — 20 breaths per minute. Heart breathing in the lungs, dry rales. Heart sounds — rhythhmal, systolic murmur at Botkin’s area. Heart rate — 82 beats per min. BP — 115/80 mmHg. Liver was protruded from under the costal arch; the edge was thick and painless. Tendency to constipation. Urination — painless.

Echocardiography (03.06.13): compared with echocardiography sizes of 12.03.12 heart chambers were without changes. EF — 60%. The function of prosthesis was not impaired. The mass in the RV apex with the spread to the RV outflow tract of 35x23 mm without significant obstruction was detected (Fig. 3). The average gradient of the PA valve prosthesis — 16 mmHg. Dilatation of the inferior vena cava, its insufficient contraction at inhale. Pulmonary hypertension grade 2. (PASP — 53 mmHg). IVS flattening due to right ventricular overload.

Electrocardiography on 03.06.13: Rhythm of the ECS with heart rate — 77 beats per min.

Chest X-ray on 03.06.13: venous plethora. The structure of the right pulmonary root was insufficiently observed, the right root was expanded, the left root was shadowed by the heart. The heart was enlarged in diameter. Flat waist.

Ultrasound of the abdomen and kidneys on 05.06.13: diffuse changes of the liver, pancreas and kidneys.

Esophagogastroduodenoscopy on 12.06.13: superficial gastritis.

MSCT of the chest, abdomen, and head on 05.06.13: the soft tissue mass of 45x63x43 mm was detected in the outflow tract of the RV (Fig. 4), which accumulated contrast agent heterogeneously. The LA was enlarged; parietal overlay with decreased density of thickness up to 10 mm was revealed. Endocardial thickening of the RV, RA and LA of up to 4 mm. Thrombosis of small branches of the PA, areas of pulmonary fibrosis and infiltrative changes of different prescription. Pathological fractures of the vertebral bodies T7 12 and L2. Multiple focal masses in the liver. Multiple enlarged mediastinal lymph nodes (Fig. 5). Signs of abnormalities of the brain or the bones of the skull were not found.
Hemoglobin — 106.0 g/L erythrocytes — 3.8x10^{12}/L, hematocrit — 31.8%, thrombocytes — 140.0x10^{9}/L, leukocytes — 6.43x10^{9}/L, ESR — 37 mm/hour, INR — 2.27, Prothrombin — 27%, C-reactive protein — 44.8 mg/L, Anti-streptolysin O <13 IU/mL. Other parameters of blood biochemistry were normal. Blood somatostatin and 5-HIAA urine tests were not made due to lack of laboratory facilities.

Consultation of gynecologist on 05.06.13. Ovaries were not palpable. Asymptomatic uterine fibroid.

Consultation of oncologist on 10.06.13. Additional examination in city Oncology Center was recommended.


According to the results of further examination in Oncology Center multiple metastases in the liver, mediastinum, spine and ischium were confirmed. The primary focus was not revealed. The patient died on 22.10.13. The autopsy had not been performed.

From the data performed we may conclude that the pathological condition, which served as an indication for heart surgery using a heart-lung machine, was the CHD. This was indicated by the typical endocardial lesion of the right heart with forming TV leaflets failure and PA stenosis, not occurring in other diseases. In addition, there were other manifestations of CHD, transforming into crisis with hemodynamic instability that caused the prolonged use of sympathomimetics during the postoperative period, various arrhythmias and conduction abnormalities of the heart with the development of the Frederick syndrome which required implantation of an artificial pacemaker.

The diagnosis of CHD was established upon re-hospitalization after echocardiography and confirmed by MSCT data. The latter revealed previously missing multiple enlarged mediastinal lymph nodes, focal changes in the liver and spine. We failed to find localization of the primary site. If we take into account that the patient had no liver metastases before the heart surgery, then at first glance, the localization of the primary site in the gastrointestinal tract becomes problematic. Multiple enlarged lymph nodes in the mediastinum, liver metastases and spine findings upon readmission, might be associated with the primary tumor localized in the airways. However, neuroendocrine neoplasms originating from the ileum are considered to be the most malignant ones [14—15]. It is considered that in their small size, slow growth without infiltration and compression of surrounding tissues, local symptoms of carcinoid tumor can be masked for a long period and the first manifestation of the disease may be diffuse thickening of the endocardium of the right heart without signs of malignancy [3, 6, 8, 18—22]. This unusual way of metastasis makes difficult to find the primary lesion, which does not allow predicting its location and the further development of the CHD. The latter is considered the main cause of death in patients with malignant neuroendocrine tumors. Therefore, in the majority of patients, as illustrated in our observation, the treatment is initiated with the heart surgery instead of verification of the tumor [21—22, 36]. Preference is given to the use of bioprostheses which do not require lifelong anticoagulation therapy increasing the risk of bleeding in patients with liver metastases [37—39]. Surgical correction of the defect has a positive effect on the remodeling process [30—32, 34—39]. In the perioperative period patients often die due to the development of complications caused by the carcinoid crisis [21, 37]. The latter ones are often provoked by anesthesia [40], the use of sympathomimetic drugs lyzing the tumor, as well as by the intervention itself. There has been a tendency of decrease in the early postoperative mortality over the last decade [28, 34]. This is considered to be related to advances in cardiac surgery combined with surgical treatment of the primary tumor and the use of somatostatin analogues [38—39, 41—42]. In the present case, a large amount of surgery, including excision of endocardium and arterial intima of the right heart, multiple valve replacement and suturing the IVS defect, despite the development of carcinoid crisis, helped compensate for the circulatory failure and extend the life of the patient for 1.5 year. Effectiveness of cardiac surgery was limited due to unrecognizable primary carcinoid lesion.

The presented data confirm that in cases of lesions of the endocardium of the right heart with developing TV failure and ostéal stenosis of the PA we should mind high probability of CHD. In this regard, along with other manifestations of CHD, echocardiography and MSCT allowing to confirm the diagnosis, identify metastases in other organs, as well as to determine the localization of the primary lesion become especially useful. Method of choice for the treatment of CHD is the heart surgery, which increases the effectiveness of the surgical treatment of the primary lesion and the use of somatostatin analogues.

![Multispiral computed angiopulmonogram. Massive wall thickening, papillary muscles, and the outflow tract of the right ventricle with the presence of stenosis at the level of pulmonary valve are being revealed.](image)
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