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Case reports

Cor triatriatum associated with complex heart defects

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Considered is one of the rarest congenital cardiac anomalies, *cor triatriatum sinistrum*, which has been identified in only 0.1% of children with congenital heart disease since the initial description of the lesion by Church in 1868. It is characterized by presence of a fibromuscular membrane that subdivides the left atrium into the proximal superior common pulmonary venous chamber and the distal true left atrial cavity. In most cases the proximal chamber receives blood from all four pulmonary veins and transmits it to the distal left atrial cavity (which contains both the mitral valve and atrial appendage) via one or more foramina in the intra-atrial membrane. Approximately in 75% of individuals there exists an atrial septal defect between the right atrium and the proximal venous chamber or the true distal atrium. Other major cardiac anomalies coexist in more than half of patients diagnosed with *cor triatriatum*.
Two paediatric patients (tree and eight years old) are presented. In one case the *cor triatriatum* was complicated with partial anomalous pulmonary venous drainage and a left accessory vena cava, in the second case a partial atrio-ventricular septal defect was present. In both cases the surgical repair was successful. Transthoracic and transesophageal echocardiography was used to confirm diagnosis and success of repair.

Key words: congenital heart disease, *cor triatriatum*, partial pulmonary venous drainage, atrioventricular septal defect, transesophageal echocardiography, surgical repair

INTRODUCTION

> "Division, or partitioning, of one of the atrial chambers is rare but important malformation in as much as severe symptoms or death may ensure if it is untreated. When recognized and treated surgically, life expectancy may be normal".
>
> ***R. H. Anderson (1)***

Apparently *cor triatriatum* has been first described by Church (2). The name "cor triatrium" was applied to the malformation by Borst (3). The angiographic diagnosis first have been made at the Mayo Clinic and described by Miller (4). The echocardiographic diagnosis of the cardiac anomaly was described by Ostman–Smith (5) and by Wolf (6). The first surgical correction is believed to have been performed by Vineberg and Galloreto (7) and shortly therefore by Lewis (8). Typically, in classical *cor triatriatum*, the common pulmonary venous chamber (proximal chamber) is somewhat larger than the left atrium (distal chamber). The common wall between them, which may have one or more openings in it, is usually rather thick and fibromuscular. Occasionally, rather than an aperture in the common wall or diaphragm, the connection may be tubular (9, 10). The "proximal" chamber is usually thick-walled whereas the "distal" left atrium containing the mitral valve and left atrial appendage is thin-walled. Despite the high pressure in the proximal chamber, the pulmonary veins are not dilated (11).

The right ventricle is usually enlarged but the enlargement depends on the presence and degree of left-to-right shunting at atrial level. The left ventricle is usually normal size or small.

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The *fossa ovalis* or atrial septal defect may be in the septum between the proximal chamber and the right atrium (12, 13). Rare cases have been described where the communication with the distal chamber was an atrioventricular rather than an atrial septal defect (1). The foramen ovale is usually patent and stretched (11).

In normal hearts with atrio-ventricular concordant connection and normally positioned ventricles the left atrium contains the left atrial appendage and empties into the left ventricle through the mitral valve. An atrial septum, with or without a foramen ovale, separates the left atrium from the right one (1, 11).

In *cor triatriatum* the right and left pulmonary veins can be considered as not joining the left atrium but rather as entering a chamber generally posterior and a little superior or medial to the left atrium, which is analogous to the anomalous pulmonary venous connection. Right-sided pulmonary veins can instead connect to and drain into the right atrium or in coronary sinus (1).

CLINICAL FEATURES AND DIAGNOSTIC CRITERIA

Infants with classical *cor triatriatum*, with a small opening between the proximal and distal left atrium, usually present with an evidence of low cardiac output, including pallor, tachypnoe, poor peripheral pulses, and growth failure (10). When there is associated left-to-right shunting because of an opening of the proximal chamber into the right atrium or because of associated partial anomalous pulmonary venous connection, evidence of increased pulmonary blood flow and venous obstruction may be present in the chest radiograph, and right ventricular enlargement is prominent (13, 14).

In children and young adults, the classic presentation is with the signs and symptoms of pulmonary venous hypertension. However, just as does mitral valve stenosis, *cor triatriatum* may present less classical symptoms (15).

The diagnosis can be strongly suspected and confirmed by examination with cross-sectional and colour Dopller echocardiography (5, 11, 15–18). Cardiac catheterisation and angiographic studies are no longer considered necessary for a precise diagnosis, unless the quantification of shunts and calculation of pulmonary vascular resistance are necessary (1, 11, 13).

Cardiac catheterisation and angiocardiography have been relegated to qualify pulmonary pressure and vascular resistance or/and, if non-invasive methods are not informative enough, to establish all complex heart defects (11, 19).

Hemodynamic abnormalities of this condition are similar to those of mitral valvae stenosis, in that both conditions produce pulmonary venous and pulmonary arterial hypertension. Important physical findings include dyspnoea, accentuated second heart sound and continuos murmur.

The electrocardiogram shows right axis deviation and severe right ventricular hypertrophy and occasional right atrial hypertrophy (10, 11).

The chest X-ray shows evidence of pulmonary venous congestion or pulmonary oedema, prominent pulmonary artery segment, and right-sided heart enlargement (1, 9, 11, 13, 14).

Echocardiography demonstrates a linear structure within the left atrial cavity and additional structural anomalies if they exist (5, 6, 13, 15, 17).

Surgical correction is always indicated. Pulmonary hypertension rapidly regresses in survivors if the correction is made early (11).

MATERIALS AND METHODS

1. Case report

An eight-years-old girl was hospitalised in our clinic for assessment following a 12-month history of dyspnoea and fatigue on exertion (curved anterior thoracic wall, pink nails). Her physical development was under normal age range. Anterior thoracic wall was slightly curved. There were signs of heart failure with slight tachycardia, tachypnoe and hepatomegaly, functional class II–III (NYHA). Her saturation on air was about 84%. Examination demonstrated the presence of 3/6-grade systolic murmur at the 2nd left

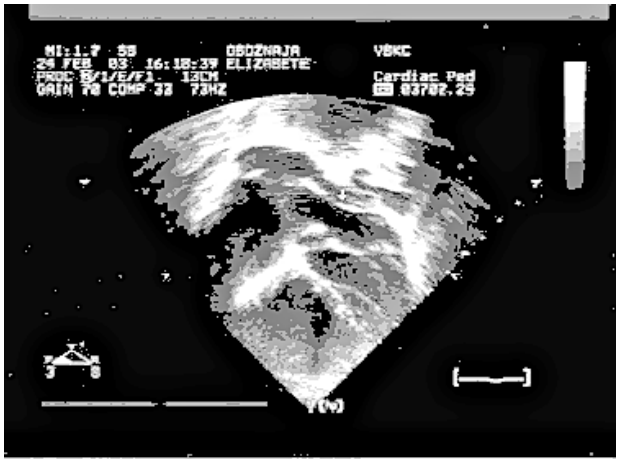

Fig. 1. A cross-sectional echocardiography in four chamber long axis plane showing (*arrow*) the dividing partition in the left atrium. Left ventricle is squeezed by the enlarged right heart side

intercostal space. The second heart sound was accentuated.

Chest X-ray showed pulmonary plethora and right-sided heart enlargement. The ECG showed sinus rhythm, right axis deviation +130°, right atrial and ventricular hypertrophy.

Transthoracic and transeosophageal echocardiography (Fig. 1.) demonstrated *situs solitus*, atrio-ventricu-

lar and ventriculo-arterial concordance, normal systemic venous drainage, patent ductus arteriosus, atrial septal defect (ASD). A linear structure within the left atrium cavity was visualized. There was an enlargement of pulmonary artery and squeezed left ventricle from the enlarged right side. The left ventricular function was normal.

The girl had been on diuretic and ACE inhibitor medication for a couple of months.

Preoperative diagnosis: congenital heart disease, *cor triatriatum*, partial anomalous pulmonary venous drainage, atrial septal defect, patent ductus arteriosus. Functional impairment class II–III (NYHA).

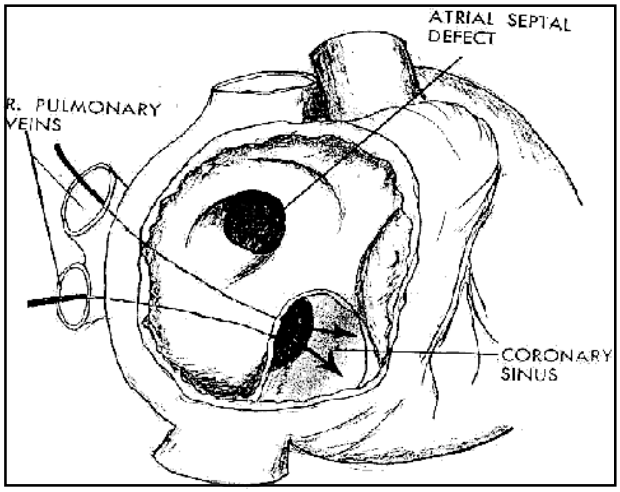

Fig. 2. Case 1. Right atrial view. Right anomalous pulmonary venous return into coronary sinus.
The communication between right pulmonary vein and the left atrium is enlarged

Surgical repair

After median sternotomy the pericardium was opened. The proximal left venous chamber was not enlarged, contrary to the right atrium, which was extremely large.

Before the preparation of cardiopulmonary bypass (CPB) the patent ductus arteriosus was dissected and ligated. After the usual preparations, a moderately hypothermic (+31, 8 °C R) CPB, using two venous cannulae (DLP right-angled, steel type) was established. Cold cardioplegia (St. Thomas Hospital Solution I, +4 °C, 15 ml/kg) and supplementary intrapericardial cooling with ice water were used. The surgical approach through a vertical incision in right atrial wall was preferred. Right-sided pulmonary veins were found drained in coronary sinus.

In the region of *fossa ovalis* a 20 mm Ø ASD was localised. The proximal left atrial chamber was opened through a vertical incision and the intraatrial septum was excised.

The orifices of the left pulmonary veins are located on the level of proximal left atrial cavity (Fig. 2.). The fibro-muscular partition with the central orifice 6 mm Ø between the proximal and the distal chambers was observed and secondary excised to make an opening as large as possible. The anterior wall of coronary sinus was dissected as large as possible for the optimal drainage of right pulmonary veins into the later formed left atrial cavity. By using a glutaraldehyde-pretreated autopericardium, separation of the right and left atrial cavities were performed. When the cardiotomy was closed, the heart having been filled with blood and evacuation air from the heart cavities and aorta, the crossclamping of aorta was realised (finished). A spontaneous sinus rhythm was established. The remainder of the operation was completed in the usual fashion.

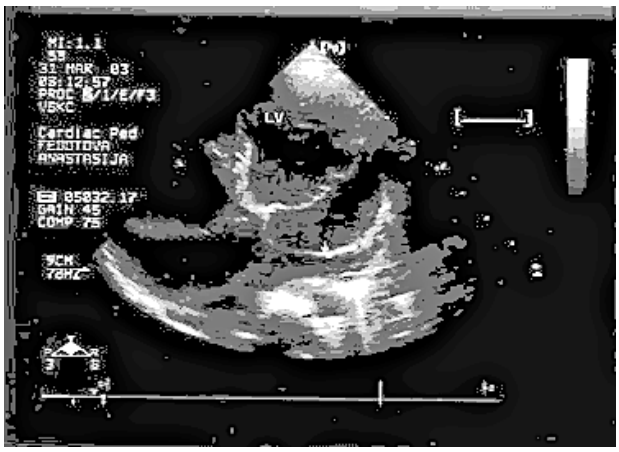

Fig. 3. Cross-sectional echocardiography in four chamber long axis plane showing (*arrow*) a dividing partition in the left atrium

Transesophegeal echocardiography showed normally divided atrial chambers, no signs of atrio-ventricular valvar regurgitation and obstruction of pulmonary veins.

The postoperative course was uneventful and the patient was discharged home on the 10th day.

Eleven months after surgery the patient had no cardiac symptoms or residual cardiac murmur. Her chest X-ray, electrocardiogram and transthoracic echocardiography were normal.

2. Case report

A three-year-old girl was hospitalised in our clinic for assessment following a last 3-month history of dyspnoea and slight cyanosis. The patient was under normal age range with presence of heart failure – dyspnoea, tachycardia and hepatomegaly. Functional impairment class III (NYHA). Examination demonstrated the presence of 3/6-grade non-specific systolic murmur at the 2nd left intercostal space widely auscultative on the anterior thoracic wall. The second heart sound was accentuated.

The *chest X-ray* showed mesocardia, pulmonary plethora and right-sided heart enlargement. The *electrocardiogram* showed sinus rhythm, left ventricular hypertrophy and left axis deviation, Š a –154°.

Transthoracic and transesophageal echocardiography (Fig. 3) demonstrated mesocardia, *situs solitus*, atrio-ventricular and ventriculao-arterial concordance, multiperforate atrial septum. A linear structure within the left atrial cavity was noted. Mitral valvae regurgitation (4, 8 m^2 /1 m^2 BSA). Estimated pulmonary artery pressure 45 mmHg (via acceleration time and tricuspid valvae regurgiation). Enlarged left atrium and good left ventricular function was noted.

Preoperative diagnosis: congenital heart disease, mesocardia, *cor triatriatum*, atrial septal defect, trivial mitral insufficiency, pulmonary hypertension. Functional impairment class III (NYHA).

Surgical repair

After the usual preparations, moderately hypothermic (+25, 8 °C R) CPB using two venous cannulae was established. Cold cardioplegia and supplementary intrapericardial cooling with ice water were used. The surgical approach through a vertical incision in the right atrial wall was performed. Intraatrial septum was multiperforated (Fig. 4). One of the ASDs was localised near the basis of the septal leaflet connecting the right atrium and the distal left atrial chamber. After overlooking the intraatrial septum was totally excised; the orifice of the left superior vena cava was noted and secondary the fibromuscular partition was observed with an eccentric 12 mm diameter orifice communicated with the distal left atrial chamber. Left- and right-sided pulmonary veins were found opened in the proximal left atrial chamber. The fibromuscular partition with eccentric orifice was excised to make an opening as large as possible. The cleft of the anterior mitral valve was observed and saturated. Hydraulic control accepted an optimal closing function of the mitral valve. Then the intraatrial septum was formed by using a glutaraldehyde-pretreated autopericardium patch. When the cardiotomy was closed, the operation was finished like in the previous case. The spontaneous sinus rhythm recovered.

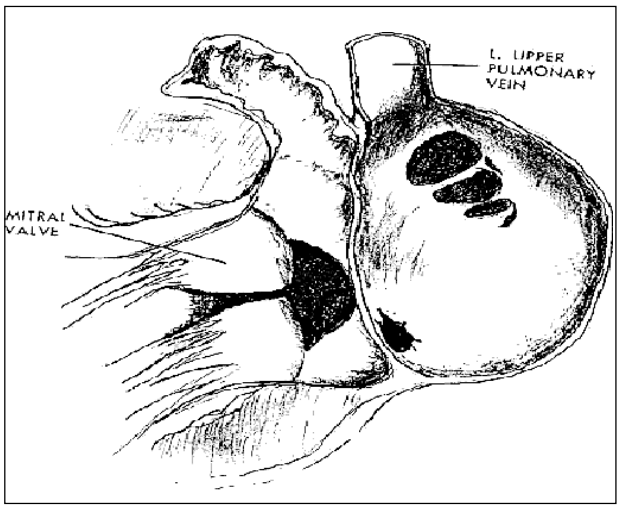

Fig. 4. Multiperforated atrial septum and partition with eccentric orifice communicated to the proximal and distal left atrial chamber.

The mitral valve cleft and typical ASD characterise the anatomy of partial atrioventricular septal defects

Also *transesophageal echocardiography* confirmed surgical success, as it showed normally divided atrial chambers, no signs of atrio-ventricular valvar regurgitation, residual septal defect or pulmonary venous obstruction. The postoperative course was uneventful and the patient was discharged home on the 9th day. She was on diuretics and ACE inhibitor medication for a month after heart surgery.

Ten months after surgery the patient had no cardiac symptoms or residual cardiac murmur. Her chest X-ray, electrocardiogram and echocardiography were normal.

DISCUSSION

The age at presentation and the clinical manifestation of *cor triatriatum* are the factors that influence the pathophysiologic features of this defect. Patients with a restrictive membrane opening are seen in infancy with signs and symptoms of pulmonary venous obstruction and severe congestive heart failure. In such infants, the diagnosis is often confused with other cardiac anomalies, causing pulmonary venous hypertension, obstructed total anomalous pulmonary vein drainage, congenital mitral valve stenosis, and pulmonary vein stenosis (11). Individuals with a large partition ostium may remain symptom-free for many years before developing congestive heart failure like in our patients. The procedure of choice for diagnosing *cor triatriatum* has become two-dimensional echocardiography with colour flow Doppler (5, 6, 11, 13, 15, 17, 19). This non-invasive modality is associated with fewer complications than cardiac catheterisation and angiocardiography, especially in critically ill neonates with severe pulmonary venous obstruction, and is more accurate in providing anatomic definition of the intra-atrial membrane and excluding other differential diagnosis. For cases in which transthoracic views provide insufficient anatomical details, transesophageal echocardiography has emerged as an invaluable adjunctive diagnostic procedure. Cardiac catheterisation and angiocardiography have been relegated to qualify pulmonary pressure and vascular resistance or/and if non-invasive methods are not informative enough to establish all complex heart defects (11).

The classical *cor triatriatum* with a restrictive aperture in the partition between the proximal common pulmonary venous chamber and the distal left atrium is an urgent indication for operation. Since 75% of patients with such malformations die in infancy, symptoms usually develop early, and operation is necessary in the first year

of life. When older patients present with chronic symptoms, operation is also urgently indicated (11).

In a complex *cor triatriatum*, more significant is to perform the repair as early as possible. Before the operation, the surgeon must try to determine with reasonable certainty the connections and drainage of all the pulmonary and systemic veins, including the possible presence and connection of a left superior vena cava. The largest "atrial" chamber appearing on the right side of the heart should be opened initially at operation (11). If it is not the right atrium, the right atrium often should be opened subsequently to complete a thorough examination of the morphologic details, all of which must be verified at operation.

The repair itself will be some combination of the repair of typical *cor triatriatum*, of partial and total anomalous pulmonary venous connections and of unroofed coronary sinus syndromes. For patients with *cor triatriatum* postoperative care is as usual.

RESULTS

In isolated cases of *cor triatrium* in which a timely diagnosis is made, the surgical mortality is less than 1%. The higher mortality rates reported by many centres are rather a reflection of the natural history of frequently associated complex heart defects than triatrial heart *per se*. An excellent long-term prognosis and quality of life are to be expected in uncomplicated cases.

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COR TRIATRIATUM IR KOMPLIKUOTOS ÐIRDIES YDOS

Santrauka

Cor triatriatum yra viena ið reèiausiai aptinkamø ágimtø ðirdies anomalijø. Autoriai pateikia du atvejus (3 ir 8 metø amþiaus vaikai), kai tokie ligoniai buvo sëkmingai operuoti ir ðiuo metu jauèiasi gerai.