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Original Research Article

Surgical Repair Results of Cor Triatriatum Sinister in 5 Cases

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Abstract: As a rare congenital defect, cor triatriatum sinister represents only 0.1% -0.4% of congenital cardiac anomalies. Depending on the accompanying symptoms and the degree of obstruction, cor triatriatum sinister is usual for patients to present in infancy and early childhood, although some cases remain undetected until adult life. We described 5 patients with cor triatriatum sinister who underwent operation in our institution. Five patients underwent surgical correction of cor triatriatum sinister were seen at Affiliated Hospital of Guilin Medical University between January 2005 and December 2015. The demographic characteristics and surgical results of these patients are outlined in this retrospective study. There were 4 males and 1 female with a mean age of 11.4 years (age range, 1 year to 27 years). The surgical approach consists of right atriotomy, excision of the obstructing membrane, and repair of the associated intracardiac anomalies. After an uneventful postoperative hospital stay, all patients were discharged 6–8 days postoperatively. Patients were followed up from 15 months to 12 years and were symptom free. There were no recurrence after surgical repair of the cor triatriatum sinister. Surgical treatment of cor triatriatum sinister provides satisfactory early and mid-term survival with low risk for additional intervention.
Keywords: Cor triatriatum sinister, Congenital heart disease, Surgery

INTRODUCTION

Cor triatriatum is a rare congenital cardiac disease represented by the development of a fibro muscular membrane that divides the left atrium (sinister) or the right atrium (dexter) into two chambers. Cor triatriatum sinister occurs in approximately 0.1% to 0.4% of patients with congenital heart disease, whereas cor triatriatum dexter occurs in less than 8% of all cor triatriatum patients [1]. Cor triatriatum sinister is slightly more common among men than women with a 1.5:1 male-to-female ratio.

Cor triatriatum sinister is a cardiac anomaly in which a common pulmonary venous chamber is separated from the left atrium by a fibro muscular septum. The two chambers communicate through the opening in the sptum. This malformation is usually isolated, but it may be associated with other cyanotic or acyanotic congenital heart anomaly. The clinical manifestations of triatriatum sinister depend upon the accompanying symptoms and the degree of obstruction. In many patients, the opening is severely restrictive. In other cases, the onset of clinical manifestations may be delayed if the opening is large enough.

Cor triatriatum sinister is a surgically treatable congenital defect. We report 5 cases of cor triatriatum sinister who came to us with variable physical presentations and symptoms, along with other cardiac morphology. In these cases, surgical management differed due to varied heart abnormalities.

PATIENTS AND METHODS

A retrospective analysis was performed in order to determine the patients with a history of surgical intervention for cor triatriatum sinister following the approval of the ethical committee of Guilin Medical University Hospital. Clinical presentations, preoperative echocardiographic findings, perioperative course and follow-up records were evaluated. Five patients (4 male

and 1 female) with cor triatriatum sinister were operated between January 2005 and December 2015. The mean age was 21.4 years (age range, 1 year to 27 years) and median body weight was 28.8 kg(range 8–53 kg).

RESULTS

In our patient population, all of the patients had atrial septal defect (ASD), all of which were located between the proximal chamber and right atrium. Four of the patients had ASD and 1 of the patients had partial abnormal pulmonary venous connection (PAPVC). In one case with PAPVC, the abnormal pulmonary veins were draining into the right atrium. None of the cases with cor triatriatum sinister had cardiac catheterization.

The preoperative symptoms ranged from mild respiratory distress to exertional dyspnoea and fatigue. The surgical indication for intervention was predominantly determined by the accompanying symptoms and the concomitant heart defects. Two of the patients had preoperative pulmonary hypertension, which was detected by transthoracic echocardiographic evaluation.

A right atrial approach was performed in order to facilitate the surgical exposure. Cardiopulmonary bypass (CPB) with mild-to-moderate hypothermia was implemented in all cases. Mean aortic cross-clamping time and total CPB time were 50 ± 16 and 65 ± 36 min, respectively. Following the cross-clamping of the aorta, cold antegrade blood cardioplegia was administered. Our surgical procedure incorporated resection of the membrane at the left atrial cavity and a complete correction of the accompanying heart defects. In the case with PAPVC, the pulmonary venous return was rerouted to the left atrium.

No early postoperative mortality was encountered in all patients. The early postoperative results were evaluated using transthoracic echocardiography at the first postoperative day, before discharge and at the third postoperative month. Afterwards, annual transthoracic echocardiographic evaluations were recorded. None of the patients needed postoperative reintervention following the initial repair.

DISCUSSION

The first case of an abnormal membrane in the left atrium was described by Church in 1868 [2]. In cor triatriatum sinister, the membrane within the left atrium results from failure of resorption of the common pulmonary vein. The upper atrium chamber receives the pulmonary veins and the lower atrium chamber contains the atrial appendage and the mitral valve. The two atrium chambers communicate through an opening in the membrane. Cor triatriatum sinister is often associated with other congenital cardiac defects. The membrane can cause obstruction of pulmonary venous return, which leads to pulmonary hypertension and overloading of the right ventricle [3, 4]. Most frequently, the upper chamber communicates with the right atrium through a patent foramen ovale (PFO) or ASD, and the clinical symptoms simulate anomalous pulmonary venous return. Less commonly, the PFO communicates with the distal chamber and the clinical features mimic mitral stenosis. When cor triatriatum sinister is the only abnormality, the clinical findings are also similar to mitral stenosis with development of pulmonary hypertension and subsequent right ventricular hypertrophy and atrial enlargement [5]. Cor triatriatum sinister is usually diagnosed in childhood; however, some cases, may rarely present in adulthood when the membrane contains large opening [6-8].

Various anatomical variations exist and were categorized by Loeffler in 1949 [9], It is based on the number and size of openings in the fibro-muscular membrane and it distinguishes three groups. Group one is defined by the absence of connection between the two chambers, the accessory chamber might connect with the right atrium or some of the pulmonary veins might drain in anomalous fashion. Group two is defined by one or few small openings in the intra-atrial membrane. Group three is defined by the accessory chamber communicates widely with the true atrium though a large single opening. While the group one and group two are usually diagnosed in highly symptomatic infants and children and are associated with increased mortality at a younger age, the group one is mostly found in the adult population having this abnormality,

Earlier surgical series required cardiac catheterization to establish the diagnosis of cor triatriatum sinister. Since the 1990s echocardiography has become the main diagnostic choice for classic cor triatriatum sinister [10]. Our patients had transthoracic echocardiograms as their initial evaluation, which revealed abnormal structures in the left atrium. When compared with 2D imaging, 3D transesophageal echocardiography enables a more accurate data about the anatomy and the structure of the membrane and its fenestrations [11]. Real-time 3D imaging was utilized to more completely define the pathology. Real-time 3-D transesophageal echocardiography offered a comprehensive anatomic and functional evaluation of cor triatriatum sinister [12]. Further investigations, such as cardiac catheterization or MRI were recommended for the diagnosis of cor triatriatum sinister. All of our

patients were diagnosed by transthoracic echocardiography. In our institution, no patients undergo cardiac catheterization including angiography prior to surgery for proper surgical planning.

The cor triatriatum sinister may be associated with other congenital heart disease in 24–80% of the cases [10]. Partial or total anomalous pulmonary venous connection is the most commonly associated cardiac pathology, which has been reported in nearly one-third of the patients with cor triatriatum sinister [10]. We encountered PAPVC and ASD as the main associated congenital heart diseases in our patient population.

Since the first surgical approach in 1956 [13], surgery is the treatment of choice in symptomatic patients with cor triatriatum sinister. Surgical correction is indicated in patients with obstructive symptoms regardless of age [10].The surgical approach can be through either a median sternotomy or a right thoracotomy [14]. We preferred the median sternotomy approach. Surgical approach consists of left or right atriotomy, depending on the presence of an ASD, and excision of the membrane. Of our patients all cases were operated through a right atriotomy. When this opening is significantly obstructive, it causes restriction of pulmonary venous return and possibly pulmonary hypertension, mimicking the pathophysiology of mitral stenosis. In these cases, surgical resection of the membrane is indicated, with excellent long-term outcome. In our surgical experience, we feel that a transseptal approach provides an excellent view while remaining technically easier in all cases. The left atrial approach can be utilized in older children and adults with a dilated left atrium.

A successful outcome depends on complete excision of the diaphragm between the proximal atrial and distal atrial chambers. Most reported deaths occurred in children who presented in a critically ill condition, those with complex associated anomalies or after a misdiagnosis. We have had no mortality related to surgery in our series in either the classic or atypical cor triatriatum sinister. The results are encouraging, the outcome is more dependent on associated lesions than on the cor triatriatum sinister itself. In the case series by Salomone and his colleagues, three of the 15 patients died, two of them had additional complex congenital heart dise [15]. In recent years, reported long-term results of surgery are excellent and patient's life expectancy can be expected to approximate that of the general population [16].

CONCLUSION

Pulmonary venous congestion might be caused by cor triatriatum sinister. As surgical treatment is easy, searching for cor triatriatum sinister in patients showing these symptoms should be considered. Diagnosis nowadays can easily be achieved per echocardiography. Surgery resolves all related symptoms at once. The long-term results are excellent.

CONFLICT OF INTERESTS

None declared.

AUTHOR'S CONTRIBUTION

Haiyong Wang and Xianzhu Liang wrote the paper. Fugui Ruan, Qiong Wang, Feng Lin, Jianbin Sun, Zhenzong Du, Jianfei Song supervised the composition of the paper. All authors read and approved the final paper.

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