**CASE CONFERENCE**

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**Cor Triatriatum Sinister Presenting in Adulthood**

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**CASE REPORT**

A 43-year-old man was transferred to this institution for further evaluation and management of recurrent atrial tachycardia in the setting of newly diagnosed cor triatriatum and patent foramen ovale by transesophageal echocardiography. His presenting symptoms included palpitations, followed by chest pain and right jaw pain. He underwent Holter monitoring with no captured arrhythmias, followed by a nuclear stress test, which showed no electrocardiographic changes and no significant arrhythmia. Perfusion images showed no abnormality. He subsequently developed some prolonged episodes of atrial tachycardia that were captured on telemetry and treated with diltiazem. He had no history of arrhythmias before this presentation; his past medical history was significant only for hyperlipidemia, and there was no prior history of surgery or general anesthesia. Home medications included metoprolol, 50 mg daily. He had received coumadin at the transferring hospital, but was not anticoagulated at the time of presentation.

As part of his preoperative workup for surgical repair, he underwent cardiac MRI, which confirmed the diagnosis of cor triatriatum sinister (Fig 1). An extensive fenestrated left atrial membrane with attachment points at the fossa ovalis and at the junction of the left upper pulmonary vein and left atrial appendage was identified, with evidence of flow acceleration across the membrane. Left-heart catheterization was significant for two low-grade lesions (30% and 40%) in the left anterior descending coronary artery and a left-to-right interatrial shunt. The left ventricular end-diastolic pressure (LVEDP) was 16 mmHg. Simultaneous pulmonary capillary wedge pressure (PCWP) and LVEDP showed a PCWP – LVEDP gradient of approximately 3 mmHg, suggesting a mean gradient across the LA membrane of 3 mmHg. A right-heart catheterization showed the following oxygen saturations: superior vena cava, 70%; inferior vena cava, 70%; right atrium, 77%; right ventricle, 75%; and pulmonary artery, 77%. The shunt fraction (Qp:Qs) was calculated to be 1.4:1. The arterial saturation was 95%.

A transthoracic echocardiogram showed the left atrial membrane, a mildly dilated left ventricle (LVEF 50%), a dilated right ventricle with preserved systolic function, mild tricuspid regurgitation, 2+ mitral regurgitation, biventricular enlargement, and a right-to-left interatrial shunt (in contrast to the shunt direction noted at catheterization) consistent with a patent foramen ovale versus an atrial septal defect. The estimated right ventricular systolic pressure was 31 mmHg.

All preoperative laboratory values, including complete blood count, electrolytes, renal panel, and coagulation panel were within normal limits.

The patient underwent uneventful anesthetic induction with etomidate, midazolam, fentanyl, and intubation with rocuronium. Monitoring included standard ASA monitors, a left brachial arterial catheter, a right internal jugular introducer sheath with pulmonary artery catheter, and transesophageal echocardiography.

Intraoperative echocardiography showed a fenestrated membrane in the left atrium, extending from the interatrial septum to the junction of the left upper pulmonary vein and left atrial appendage (Fig 2, Video clips 1 and 2). A 1-cm secundum-type atrial septal defect was identified by color flow inferior to the atrial membrane (Video clip 3), with right-to-left shunting. There was 1-to-2+ mitral regurgitation, mitral annular dilatation, mild tricuspid regurgitation, normal left ventricular size and function, and a dilated right ventricle with normal systolic function.

The patient underwent resection of the atrial membrane, mitral valve annuloplasty, atrial septal defect (ASD) closure, and a MAZE procedure. The operation was performed through a median sternotomy with aortic and bicaval cannulation, delivery of antegrade and retrograde cardioplegia, and normothermia. A right atrial incision was made, with exploration of the left atrium through the interatrial septum. The left atrial membrane was visualized clearly and excised (Fig 3), and the left atrial appendage amputated. The mitral valve also was approached through the interatrial septum and repaired with a #36 Cosgrove annuloplasty band. The ASD was closed, and the patient was weaned from cardiopulmonary bypass.

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**Cor TRIATRIATUM** is an uncommon congenital cardiac anomaly that is characterized by division of the left atrium into two separate chambers by a fenestrated membrane. This condition typically presents in infancy or early childhood, and can be associated with other cardiac anomalies. A smaller number of adult cases have been reported in the literature. We present the case of a patient with cor triatriatum sinister and an associated atrial septal defect who presented in adulthood, along with a comprehensive review of the literature with a focus on adult patients.
without need for pharmacologic support. The post-pump TEE showed no residual mitral regurgitation and normal biventricular function. His postoperative recovery was uneventful.

DISCUSSION

Cor triatriatum, also known as cor triatriatum sinister, or triatrial heart, is an uncommon congenital cardiac condition that is characterized by separation of the left atrium into a proximal (superior, posterior) accessory chamber that receives inflow from all four pulmonary veins, and the distal (inferior, anterior) main left atrial chamber (including the left atrial appendage and mitral orifice) by a fenestrated fibromuscular septum. The insertion point of the membrane is at the margin of the fossa ovalis medially, and at the junction of the left upper pulmonary vein and left atrial appendage laterally.\(^1,2\) The proximal chamber has been described as a common pulmonary vein, or third atrium, where the 2 right and left pulmonary veins join together. The earliest description of cor triatriatum was by Church in 1868.\(^3\) The reported incidence is 0.1% to 0.4%,\(^4\) with a slight male preponderance (1.5:1).\(^5\) Developmentally, it is believed to result from incomplete incorporation of the common pulmonary vein into the left atrium, leading to a potential inflow obstruction with incomplete reabsorption of the common pulmonary vein.\(^6-8\) Other earlier proposed developmental mechanisms included abnormal growth of the septum primum,\(^9\) entrapment of the common pulmonary vein by the left horn of the sinus venosus, preventing incorporation into the left atrium,\(^10\) and development of a left atrial membrane by impingement of a persistent left superior vena cava on the developing left atrium.\(^11\) In 1949, a classification scheme was proposed by Loeffler,\(^8\) based on the number and size of openings in the left atrial membrane: Group I, no opening (ie, drainage is into the right heart by way of interatrial communication or abnormal pulmonary venous connection); Group II, \(\geq 1\) small opening (a relatively obstructive condition); and Group III, a single wide opening (generally little-to-no obstruction). The shape of the pulmonary venous chamber also has been used to classify this anomaly.\(^12\) The more common “diaphragmatic” type consists of a diaphragm of
thickened endocardial tissue dividing the left atrium into an accessory chamber and the true left atrium, and cannot be appreciated externally. The “hourglass” type demonstrates a constriction that can be seen externally at the junction of the accessory chamber and true left atrium. Lastly, the “tubular” type, which is visible on external inspection, lacks a membrane; however, a tubular channel receives all 4 of the pulmonary veins and is considered a common pulmonary vein, with its lower end joining the “true” left atrium. A second classification scheme was proposed by Lam et al in 1962 (Fig 4A-E): Type A, in which all pulmonary veins enter the proximal chamber, the distal chamber contains the left atrial appendage and mitral valve, and there is no ASD; Subtype A1, in which an ASD communicates with the proximal chamber; Subtype A2, in which an ASD communicates with the distal chamber, as in this patient; Type B, in which all pulmonary veins drain into the coronary sinus (a variant of total anomalous pulmonary venous connection); and Type C, in which the pulmonary veins

Fig 3. Surgical specimen from resected left atrial membrane. (Color version of figure is available online.)

Fig 4. (A) Type A defect; (B) Type A1 defect; (C) Type A2 defect; (D) Type B defect; (E) Type C defect. (Color version of figure is available online.)
do not communicate with the proximal chamber (rarest type).  

Other classification schemes also have been proposed and are described in Table 1.  

In “partial” or subtotal cor triatriatum sinister, a rarer entity, the left atrium is only partly divided, and one or more of the pulmonary veins may enter the true left atrial cavity. Unilateral pulmonary venous hypertension may result.

Cor triatriatum dexter, also a rarer entity, occurs when the right atrium is separated into 2 chambers by a membrane as a result of persistence of the right sinus venous valve. The proximal upstream chamber receives blood flow from the superior and inferior vena cavae, and the distal chamber includes the right atrial appendage. This anomaly frequently is associated with other right-sided cardiac abnormalities, though isolated cases also have been reported.

Patients also have presented in adulthood. Surgical correction is indicated in symptomatic patients with significant obstruction.

In most instances, cor triatriatum is an isolated malformation, although it can be associated with a number of other congenital defects, the most common of which is atrial septal defect (ASD) or patent foramen ovale. A secundum ASD can communicate with either or both chambers. A patent foramen ovale almost always communicates with the true left atrium. Mitral regurgitation also has been reported in association with cor triatriatum, although no true mechanism has been defined. Other less common associations include anomalous pulmonary venous return, coarctation with bicuspid aortic valve, isolated left superior vena cava, left superior vena cava with unroofed coronary sinus, subpulmonary stenosis, subaortic stenosis, transposition of the great vessels, mitral atresia, interrupted aortic arch, hypoplastic left ventricle, ventricular septal defect, patent ductus arteriosus, tetralogy of Fallot, and Ebstein’s anomaly. If an ASD that communicates with the pulmonary venous chamber is present, partial decompression can occur, thereby potentially delaying presentation (until the ASD becomes symptomatic). Clinical symptoms can mimic those of anomalous pulmonary venous return.

Cor triatriatum generally is diagnosed in infancy and childhood, with age at presentation being dependent on the size of the membrane fenestration(s), the presence of an atrial septal defect, atrial fibrillation, and mitral regurgitation. Physiologically, this condition closely can mimic mitral stenosis, depending on the extent of obstruction to inflow. The distinguishing element is diastolic flow in mitral stenosis as opposed to flow throughout the cardiac cycle in cor triatriatum. Pressures in the common pulmonary vein typically are elevated, while pressures in the true left atrium are normal. Symptoms are highly dependent on the size of the septal opening(s), which generally are restrictive. Calcification and fibrosis of the openings later in life can precipitate symptoms. Because of a lack of well-defined features on clinical examination, in the absence of symptoms, the condition can go undetected for decades and into adulthood. In fact, the appropriate diagnosis has been mistaken for pulmonary disease in the absence of appropriate diagnostic studies. In one report, a patient with coexisting COPD, cor pulmonale, and only slightly elevated PCWP despite severe pulmonary hypertension, pulmonary venous obstruction was masked, and the diagnosis of cor triatriatum was not made until echocardiography ultimately was performed. Auscultative findings may include a heart murmur. Other associated symptoms have included dyspnea, orthopnea, and even hemoptysis. Atrial fibrillation and even cerebral embolism also have been presenting symptoms. It has been suggested that the abnormal septum may be a precipitating factor for the development of atrial fibrillation, rather than the associated hemodynamic disturbances. In one patient who underwent surgical correction of cor triatriatum, the presenting symptom of atrial fibrillation completely resolved postoperatively. Another patient with symptomatic drug-refractory atrial fibrillation incidentally was found to have cor triatriatum during routine TEE examination to rule out thrombus before planned catheter ablation. Intracardiac echocardiography facilitated catheter ablation by providing visualization of the membrane and passage of the catheter into the appropriate pulmonary vein chamber.

The natural history depends on the size of the membrane fenestrations and whether or not an ASD is present, as well as its location. When the fenestrations are small and in the absence of an ASD (or the presence of a small ASD), presentation is typically in infancy. Presentation will be delayed if the membrane fenestrations are less restrictive and if an ASD communicates with the proximal atrial chamber, thereby allowing decompression. In this latter situation, right-
sided volume overload eventually manifests. Decompression of the lower chamber by an ASD would be unlikely to affect potentially high pressures in the proximal chamber.

Congenital supravalvar mitral ring, an uncommon subtype of congenital mitral stenosis that typically presents early in infancy, also can have a similar presentation to cor triatriatum.\textsuperscript{31,42} It can occur in isolation, but in most cases (approximately 90%), it is associated with other congenital cardiac defects. With this anomaly, a thick fibrous connective tissue ridge partially or completely surrounds the mitral orifice and usually is associated with a normal mitral valve apparatus (although the ring may adhere to the mitral valve and cause leaflet restriction). The ridge originates at the base of the atrial surface of the mitral valve and protrudes into the mitral valve inlet. Echocardiography easily distinguishes this entity from cor triatriatum.

In a review of 43 adult cases of cor triatriatum (age range 16 to 76 years) presenting as mitral stenosis between 1966 and 2004, Slight et al found that pulmonary capillary wedge pressure and mean pressure gradients were significantly greater in younger adults and that the incidence of atrial fibrillation and mitral regurgitation increased with age.\textsuperscript{32} Dyspnea was the most common presenting symptom (68%), with palpitations occurring next in frequency (27%).

Although cor triatriatum remains a rare condition, notably in adults, it appears to be diagnosed with increasing frequency with the greater use of imaging techniques. In 2005, fewer than 250 cases had been reported.\textsuperscript{27} Although cardiac catheterization has been used to establish the diagnosis of cor triatriatum,\textsuperscript{46–48} echocardiography is the simplest diagnostic tool and can identify the location of the fibromuscular septum, as well as measure gradients between the proximal and distal chambers (velocities less than 1.2 m/s are considered nonobstructive) and estimate pulmonary artery pressures using Doppler.\textsuperscript{43,44} With a highly obstructive septum, pulmonary hypertension can develop. Anatomically, the membrane generally begins at the superior aspect of the left lateral atrial wall, with variable insertion into the interatrial septum.

Three-dimensional transthoracic\textsuperscript{46–48} and transesophageal echocardiography\textsuperscript{46–49} have been used to better describe the anatomy of this condition, including the size and location of membrane fenestrations and membrane dimensions. The diagnosis also has been made incidentally with computed tomography.\textsuperscript{49} Three-dimensional multidetector row-computed tomography has been used as an adjunct to echocardiography to provide reconstruction images that further delineate the anatomy of this condition and accurately identify drainage of the pulmonary veins.\textsuperscript{5} Magnetic resonance imaging also has been useful in diagnosing this condition.\textsuperscript{30}

Treatment of cor triatriatum is generally surgical, either via median sternotomy or right thoracotomy, and consists of resection of the membrane separating the common pulmonary vein chamber from the distal left atrium.\textsuperscript{51,52} The obstructing membrane can be approached through either the right or left atrium. The right atrial trans-septal approach provides excellent exposure, especially in the presence of a small left atrium. The left atrial approach opens the proximal common pulmonary venous chamber, and provides direct exposure of the membrane. The first reports of surgical repair of this defect were in 1956.\textsuperscript{53,54} In earlier times, dilatation of the mitral orifices was performed using the surgeon’s finger to fracture the membrane through the left atrial appendage\textsuperscript{53} or using a valvulotome.\textsuperscript{54} Percutaneous balloon dilatation of the membrane fenestrations also has been performed successfully when the membrane anatomy/thickness was favorable.\textsuperscript{55–58} Recently, the robotic approach to repair of isolated cor triatriatum also has been described.\textsuperscript{59} Modern surgical results are generally excellent.\textsuperscript{28,60} In a review of 28 consecutive patients (predominantly pediatric population) over a 22-year span, postsurgical repair survival was 96% and 88% at 5 and 15 years, respectively, for both isolated defects and those associated with other congenital cardiac anomalies.\textsuperscript{28} Overall life expectancy postrepair is expected to be at or near that of the general population.\textsuperscript{61} The importance of intraoperative transesophageal echocardiography cannot be overemphasized, and particular attention should be paid to mitral valve function post-resection, because of the proximity of the membrane to the valve.\textsuperscript{62}

Timing of surgery can be based on the results of stress echocardiography to assess postexercise gradients and estimated pulmonary artery pressures,\textsuperscript{67} although there is little information about the relationship among clinical symptoms, measured gradients, and long-term prognosis.\textsuperscript{68} In the case of a 70-year-old patient being evaluated for worsening angina and dyspnea, a nonobstructive cor triatriatum was discovered at transthoracic echocardiography, with no specific treatment recommended for this abnormal finding.\textsuperscript{63}

Using the classification scheme developed by Lam et al, this patient had subtype A2 cor triatriatum, with the interatrial communication occurring between the right atrium and distal left atrial chamber. His late presentation was related to the size of the fenestrations in the left atrial membrane, and possibly to decompression of the inferior left atrial chamber (in the presence of mitral regurgitation) by an atrial septal defect. The presence of arrhythmias at the time of evaluation also may have been related to the diagnosis of cor triatriatum. Although this pathology has been described, this case adds to the existing reports of this uncommon entity, and highlights the need for adult cardiothoracic anesthesiologists to be familiar with congenital cardiac anomalies that can present in adulthood.

**COMMENTARY 1: COR TRIATRIATUM SINISTER PRESENTING IN ADULTHOOD†**

This report describes a 43-year-old man who underwent open surgical repair of cor triatriatum, closure of a secundum ASD and a MAZE procedure to manage atrial fibrillation. Cardiac surgery certainly was indicated in this situation. The patient had a standard approach—median sternotomy—which provides excellent surgical visualization and enables manageable of all three conditions. But could the same result be achieved less invasively using a surgical robot? There are limited reports of the robotic approach for cor triatriatum, largely because of the rarity of this condition in the adult population.

Today, patients often seek a less invasive approach for correction of structural heart disease. Familiar with stenting to open coronary arteries and the rapid evolution of percutaneous

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aortic valve replacement, patients desire minimally invasive solutions to their heart problems. From a surgeon’s perspective, although the authors have not encountered a report describing a minimally invasive approach for this combination of lesions, it is likely that a robotic approach would have been effective for this patient.

Robotically assisted heart surgery enables the surgeon to perform exactly the same operation on the heart with smaller incisions and less tissue trauma. This means less blood loss, faster recovery, and a superior cosmetic result. It is important to understand that use of the robot entails no compromises in the operative procedure.

For this case, peripheral perfusion would be used for cardiopulmonary bypass. The femoral artery and the femoral vein would be cannulated under direct vision, and a right internal jugular central venous catheter placed by the anesthesiologist would be exchanged over a guidewire for a second venous cannula. Using an intravascular balloon to occlude the aorta, antegrade cardioplegia would be used for myocardial protection.

After robotic resection of the left atrial membrane, mitral valve exposure would be standard. A running suture technique is employed to create the lesions of the MAZE procedure. The orifice of the left atrial appendage is oversewn from the endocardial aspect in the left atrium. The secundum atrial septal defect is closed from the left atrium if primary suture closure is feasible. If the defect is large, an autologous pericardial patch is used; this is placed from the right atrial side of the defect.

After robotically-assisted surgery, patients typically spend 1 night in the intensive care unit and generally 2 to 4 additional days in the hospital. Because there is no sternotomy, return to full activity is generally possible in 2 to 4 weeks.

COMMENTARY 2: COR TRIATRIATUM SINISTER PRESENTING IN ADULTHOOD†

Capdeville et al present a case of a rare congenital cardiac defect, cor triatriatum sinister (CTS), in a 43-year-old gentleman who presented with palpitations, chest pain, right jaw pain, and recurrent atrial tachycardia. As previously described, CTS is defined as a membrane within the left atrium that might lead to restricted pulmonary venous return. Asymptomatic cases have been described in patients without a pressure drop between the proximal and distal left atrial chambers with reports of incidental diagnosis in the elderly. Although this case report details an adult presentation of cor triatriatum sinister, this anomaly often presents during infancy with symptoms and signs that are different from adults.

Humpel et al investigated the largest group of patients with CTS over the longest period of time (50 years). The most common symptoms or clinical findings of the 82 cases were congestive heart failure (CHF), respiratory complications, failure to thrive, and pulmonary hypertension. Congestive heart failure also was the most common presentation in 12 patients aged 1 month to 7 years as reported by Gheissari et al. With significant pulmonary venous obstruction, the lungs become stiffer because of venous distention and increased interstitial fluid due to elevated capillary pressure. The decreased lung compliance causes increased oxygen consumption, already double that of the adult, and increased cardiac work when infants exert themselves when crying or feeding. The dyspnea and feeding difficulty result in poor weight gain and failure to thrive. In a review of 10 cases of CTS, Krasemann et al found the most common presentation in children < 1 year of age to be failure to thrive. The association of recurrent lower respiratory tract infection, wheezing, and congenital heart disease previously has been described. In some cases of congenital heart disease, the only presenting symptom may be in the lung. In CTS, symptoms of wheezing and rhonchi can be caused by the raised bronchial venous pressure causing edema of the bronchial mucosa and increased bronchial secretions. Mucosal swelling also may cause chronic cough and hemoptysis.

After the infancy period, clinical symptoms present as exertional dyspnea, exercise intolerance or easy fatigability, and cyanosis with strenuous exercise. Unusual presentations of CTS in children include recurrent syncopal episodes, hemolytic anemia related to turbulence and shear stress produced by flow through stenosed orifices, and incidental finding of an abnormal P wave on an ECG secondary to pacemaker action from specialized tissue in the abnormal septum.

Most patients (77%) in the Humpl et al study presented with associated cardiac lesions, of which atrial septal defect (53%), total or partial anomalous pulmonary venous drainage (27%), and patent ductus arteriosus (18%) were the most common defects. More than one-third of patients (35%) also presented with associated noncardiac lesions, mainly chromosomal abnormalities (12%) and respiratory issues (11%).

Congenital division of an atrial chamber as described in this case report is a very rare congenital malformation that more commonly affects the left atrium. Cor triatriatum dexter (CTD), division of the right atrium, is even rarer. The CTD membrane represents the right-sided valve of the embryonic sinus venosus. Normally, this valve regresses early in fetal life between 9 and 15 weeks gestational age; however, remnants are not uncommon and are recognized as the normal eustachian and thebesian valves and the crista terminalis. The purpose of this valve in the fetus is to deflect preferentially more highly oxygenated blood from the umbilical vein and inferior vena cava through the foramen ovale to the left atrium. Therefore, organs of the highest oxygen consumption, the fetal brain and heart, are perfused with blood with the highest oxygen content in utero. Persistence of the right sinus valve results in a separation of the right atrium into two separate chambers: The superior portion receives systemic venous return from the inferior and superior vena cava, whereas the inferior portion contains the right atrial appendage and the orifice of the tricuspid valve.

Similar to CTS, the clinical manifestations of CTD are dependent on the degree of right atrial obstruction. However, moderate obstruction in neonates with CTD usually is diagnosed rapidly because of the severe cyanosis from right-to-left shunting across the atrial septum, triggering a neonatal cardiac work-up, as opposed to CTS, which presents with a myriad of symptoms,
including failure to thrive, feeding intolerance and respiratory complications that might not indicate further work-up until the patient has more cardiovascular symptoms. With minor degrees of obstruction, patients may remain clinically silent for some time or detected incidentally by echocardiography at surgery or at autopsy. The obstructed right atrium also may present with symptoms consistent with increased portal venous pressure resulting in ascites or weight loss. During fetal life evidence of CTD may be picked up on routine obstetric scans. Marked fetal nuchal edema may be associated with significant obstruction of the right ventricular inflow. Also, significant obstruction may lead to the underdevelopment of the right heart structures: Tricuspid valve abnormalities including Ebstein’s anomaly, hypoplastic right ventricle, and pulmonary stenosis or atresia. Diagnostically, the remnant valves appear as a “windsock” within the right atrium on transthoracic or fetal echocardiography. Surgical relief is recommended only in the symptomatic patient.

In summary, both cor triatrium sinister and dexter are rare congenital cardiac defects that could present anytime from the neonatal period, adulthood or necroscopy. Cor triatriatum presenting as pulmonary congestion, pulmonary edema, cardiogenic shock or severe cyanosis in infancy requires urgent surgery. However, cardiac malformations involving low-pressure chambers (ie, either of the atria) in general are more difficult to diagnose. For example, atrial septal defects or partial anomalous pulmonary vein connections are diagnosed more commonly later in life than lesions that involve high-pressure systems, such as ventricular septal defects. The diagnosis of primary cardiac disease requires a very high index of suspicion on the part of the physician when there are no significant cardiac findings on physical examination. In cor triatriatum, asthma or wheezing or pulmonary hypertension of unknown etiology may constitute the only evidence of underlying heart disease.

**APPENDIX A. SUPPLEMENTARY DATA**

Supplementary data associated with this article can be found in the online version at [http://dx.doi.org/10.1053/j.jvca.2013.08.018](http://dx.doi.org/10.1053/j.jvca.2013.08.018)

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