[1] Department of Pediatric Cardiology, University Children's Hospital, Muenster, Germany,
[2] Department of Thoracic and Cardiovascular Surgery, University Hospital, Muenster, Germany,
[3] Evelina Children's Hospital, St Thomas Hospital, London, UK.

Received: September 8, 2006; revision accepted: December 20, 2006

Key Words:
Cor triatriatum · Clinical symptoms · Congenital heart disease · Surgery

Herz 2007;32:506–10

DOI 10.1007/s00059-007-2882-6

Schlüsselwörter:
Cor triatriatum · Klinische Symptome · Angeborene Herzfehler · Chirurgische Therapie

Cor Triatriatum

Short Review of the Literature upon Ten New Cases

Zita Krasemann[1], Hans-Heinrich H. Scheld[2], Tonny D.T. Tjan[2], Thomas Krasemann[1, 3]

Abstract
Cor triatriatum is defined as a membrane within the left atrium, which might lead to restricted pulmonary venous return. Diagnosis is usually achieved by echocardiography, therapy of choice is excision of the membrane. Upon ten new cases, the association with other congenital heart diseases (CHDs), clinical symptoms and the surgical approach are discussed. Eight of ten patients were children, six of them aged < 1 year. Additional CHDs included atrial and ventricular septal defects, partial anomalous pulmonary venous return and complex CHD. Surgery was performed in all cases. Prognosis is related to associated CHD.

Cor triatriatum. Kurze Literaturübersicht anhand zehn neuer Fälle

Zusammenfassung
Das Cor triatriatum ist definiert als eine Membran innerhalb des linken Atriums, die zu reduziertem pulmonalvenösem Fluss führen kann. Die Diagnose wird üblicherweise echokardiographisch gestellt. Die Therapie der Wahl bei symptomatischen Patienten ist die chirurgische Exzision der Membran.

Anhand zehn neuer Fälle werden die Assoziation mit anderen angeborenen Herzfehlern, die klinischen Symptome und der chirurgische Zugangsweg diskutiert. Acht der zehn Patienten waren Kinder, von diesen wiederum sechs < 1 Jahr alt. Assoziierte Herzfehler lagen bei neun Patienten vor: sieben Vorhofseptumdefekte, ein Ventrikelseptumdefekt, eine valvuläre Pulmonalstenose, eine bikuspide Aortenklappe, ein singulärer Ventrikel, eine partielle Lungenvenenfehlmündung und eine persistierende linke obere Hohlvene. Die Therapie war in allen Fällen chirurgisch, wobei in neun von zehn Fällen bei assoziierten Herzfehlern ein rechtsatrialer Zugang gewählt wurde, während bei isoliertem Cor triatriatum vom linken Atrium aus operiert wurde.

Introduction

Despite the fact that more than 100 cases have been published since the first description, cor triatriatum is considered to be a rare congenital heart disease (CHD). It occurs in 0.1–0.4% of all patients with CHD [1–3]. The first report of this entity was a postmortem description by Church in 1868 [4].

In patients with cor triatriatum the left atrium is typically divided by a fibromuscular membrane, resulting in a posterior superior positioned proximal cavity and an anterior inferior positioned distal cavity. In most of the cases the pulmonary veins drain into the proximal cavity. The left atrial appendage is typically connected to the distal cavity, which drains into the left ventricle via the mitral valve [2]. The dividing membrane is frequently perforated by one or more orifices, connecting both parts of the left atrium. Some variants of cor triatriatum consist of the membrane and atrial septal defects (ASDs) connecting either the proximal or distal cavity (or both) with the right heart [2]. Anatomic variants are depicted in Figure 1.

The clinical symptoms depend on the size of the orifice (the classic case mimicking mitral stenosis) [5], the morphology of the defect including the presence and location of an ASD, and on associated CHD [2, 6–9]. The diagnosis "cor triatriatum" should be considered in patients with pulmonary congestion, pulmonary arterial hypertension and pulmonary venous obstruction [2]. Symptoms are usually not related to a specific anatomic subtype and include failure to thrive, tachypnea, dyspnea, and, occasionally, cyanosis [2, 6–9]. The latter can either be caused by the restricted pulmonary venous return (then associated with severe pulmonary symptoms like tachydyspnea), or in the presence of severe stenosis and ASDs, a right-to-left shunt might be present on atrial level [2, 8].

Most of the cases are diagnosed in infant patients [10]. Asymptomatic cases have been described; in these patients the orifice was wide without a pressure drop between the proximal and distal chamber [7, 11]. Sometimes, diagnosis is delayed until late adult-

hood due to missing symptoms [1, 9, 12–15]. If the pulmonary blood flow is restricted (i.e., pulmonary valve stenosis), symptoms might be misinterpreted [7, 9, 16].

In 1955, Lewis et al. performed the first operation on this disease with partial resection of the membrane [13]. Since then, several reports of surgery for this entity have been published [2, 16].

Between 1992 and 2003, more than 2,000 patients were surgically treated at the University Hospital Muenster, Germany, for CHD. Ten of these were diagnosed as cor triatriatum.

Case Studies

Six of our ten patients were aged < 1 year, one was 6 years old. The remaining two were adults (39 years and 54 years). Associated CHD was present in nine of our cases. This included seven ASDs, one ventricular septal defect, one pulmonary valve stenosis, one bicuspid aortic valve, one single ventricle, one partial anomalous pulmonary venous return, and one persistent left upper caval vein. For demographic data see Table 1. Two of the smaller infants had extracardiac malformations (one dysmelia syndrome, one VACTERL). The children were transferred to our institution either because of cardiac murmurs or because of symptoms like tachydyspnea.

The clinical symptoms showed a broad variety: all of the children aged < 1 year had a failure to thrive. Signs of pulmonary congestion including tachydyspnea were present in six cases, intermittent or permanent cyanosis in two patients. In one of these, the additional CHD was a single ventricle; thus, the symptoms were probably related to this as well.

Both adult patients were asymptomatic concerning the cor triatriatum: one was diagnosed after revealing a stroke, the other had coronary artery stenosis and underwent echocardiography and catheterization for this reason. This patient had decreasing exercise tolerance.

Interestingly, a cardiac murmur was not present in all cases, but led to diagnostic procedures and diagnosis in three of the children.

All patients underwent transthoracic echocardiography and cardiac catheterization prior to surgical treatment, and transesophageal echocardiography intraoperatively.

In case 3 it was possible to reach the proximal chamber with a multipurpose catheter through the right atrium, the ASD and the orifice of the membrane. In this patient a paper-thin ruptured membrane was found during surgery; an excision was not necessary.

In all other patients surgical treatment consisted of excision of the membrane through a right or left atriotomy (depending on the presence of an additional ASD to be closed), closure of an ASD (if present) and surgery for any additional CHD. Cardiac surgery was performed with extracorporeal circulation and hypothermic circulatory arrest, if the associated CHD required this. Surgical procedures are depicted in Table 2.

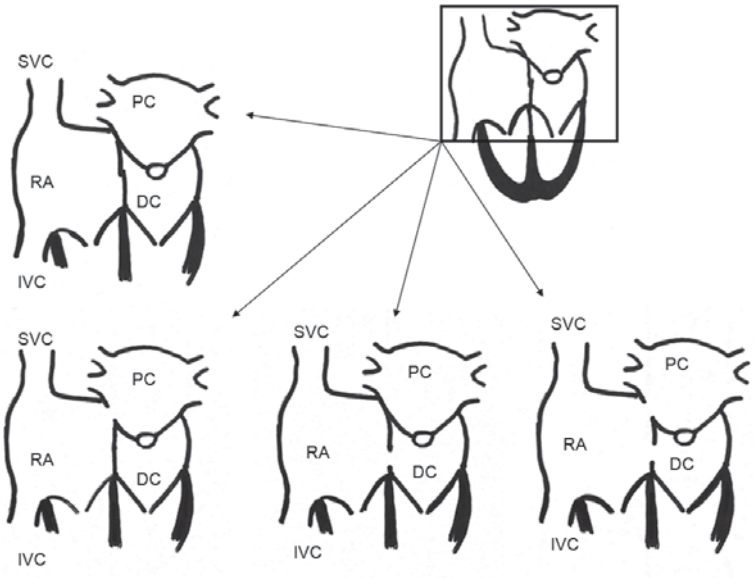

Figure 1. Cor triatriatum might be associated with interatrial defects either proximal or distal to the membrane, or with both. Additionally, anomalous pulmonary venous return has been described. DC: distal chamber; IVC: inferior caval vein; PC: proximal chamber; RA: right atrium; SVC: superior caval vein.

Abbildung 1. Das Cor triatriatum kann mit interatrialen Defekten proximal oder/und distal der Membran vergesellschaftet sein. Zusätzlich wurden auch Fehlmündungen der Lungenvenen beschrieben. DC: distale Kammer; IVC: untere Hohlvene; PC: proximale Kammer; RA: rechter Vorhof; SVC: obere Hohlvene.

Postoperatively, continuous transcutaneous oxygen saturation and arterial blood pressure measurement and assessment of urine output were routinely applied in all patients. Pulmonary artery pressures were not obtained routinely. Mechanical ventilation was applied with a positive end-expiratory pressure (PEEP) of 4 cmH_2O and maximum inspiratory pressure as needed.

There were no intraoperative complications, but a pulmonary hypertensive crisis occurred in one patient postoperatively (# 1). In patient 7 on day 14 a thrombus in the right atrium was diagnosed on echocardiography and treated successfully with low molecular heparin. In patient 3 pulmonary venous thrombosis occurred about 1 month post surgery; this was treated with low molecular heparin and resolved partly.

The patient with complex CHD (single ventricle) died due to electromechanical dissociation on the 1st postoperative day.

Table 1. Demographic data. AoV: aortic valve; ASD: atrial septal defect; Cts: Cor triatriatum; FT: failure to thrive; LPSCV: left persistent superior caval vein; PAPVR: partial anomalous pulmonary venous return; PH: pulmonary hypertension; PS: pulmonary stenosis; SV: single ventricle; VSD: ventricular septal defect.

Tabelle 1. Demographische Daten. AoV: Aortenklappe; ASD: Vorhofseptumdefekt; Cts: Cor triatriatum; FT: Gedeihstörung; LPSCV: persistierende linke obere Hohlvene; PAPVR: partielle Lungenvenenfehlmündung; PH: pulmonale Hypertonie; PS: Pulmonalstenose; SV: singulärer Ventrikel; VSD: Ventrikelseptumdefekt.

Patient #	Sex	Age	Weight	Echo, catheterization	Extracardiac diagnosis	Symptoms
1	Female	1 month	3,350 g	Cts, bicuspid AoV		Dyspnea, FT
2	Male	5 months	4,400 g	Cts, VSD, PH, LPSCV	Dysmelia syndrome	Tachypnea, FT
3	Female	5 months	3,550 g	Cts, ASD II		Tachypnea, FT
4	Female	6 months	4,830 g	Cts, ASD II	VACTERL	Tachypnea, FT, cyanosis
5	Female	9 months	6,190 g	Cts, SV, ASD II		Tachydyspnea, FT, cyanosis
6	Male	10 months	8,900 g	Cts, PH		Dyspnea, intermittent cyanosis
7	Male	17 months	10.5 kg	Cts, ASD II, PS		Cardiac murmur
8	Female	6 years	19.8 kg	Cts, ASD II, PAPVR		Cardiac murmur
9	Female	39 years	90 kg	Cts, ASD II	Left-sided stroke	None
10	Male	54 years	78 kg	Cts, ASD I	Systemic hypertension	None

Table 2. Surgical approach and complications. AoV: aortic valve; ASD: atrial septal defect; BPI: bradycardia with subsequent pacemaker insertion; Cts: cor triatriatum; KDS: Kaye-Damus-Stensel palliation; LA: left atrium; LPSCV: left persistent superior caval vein; PAPVR: partial anomalous pulmonary venous return; PH: pulmonary hypertension; PS: pulmonary stenosis; RA: right atrium; RM: resection of membrane; SV: single ventricle; VSD: ventricular septal defect.

Tabelle 2. Chirurgischer Zugang und Komplikationen. AoV: Aortenklappe; ASD: Vorhofseptumdefekt; BPI: Bradykardie mit anschließender Schrittmacherimplantation; Cts: Cor triatriatum; KDS: Kaye-Damus-Stensel-Palliation; LA: linker Vorhof; LPSCV: persistierende linke obere Hohlvene; PAPVR: partielle Lungenvenenfehlmündung; PH: pulmonale Hypertonie; PS: Pulmonalstenose; RA: rechter Vorhof; RM: Membranresektion; SV: singulärer Ventrikel; VSD: Ventrikelseptumdefekt.

Patient #	Diagnosis	Surgery	Approach	Intraoperative findings	Complications
1	Cts, bicuspid AoV	RM	RA	Diagnosis confirmed	Pulmonary hypertensive crisis
2	Cts, VSD, PH, LPSCV	RM, VSD patch	RA	Diagnosis confirmed	Sepsis
3	Cts, ASD II	ASD patch	RA	Membrane already ruptured	Pulmonary venous thrombosis, BPI
4	Cts, ASD II	RM, ASD patch	RA	Diagnosis confirmed	
5	Cts, SV, ASD II	RM, KDS, ASD patch	RA	Diagnosis confirmed	Death
6	Cts, PH	RM	LA	Diagnosis confirmed	
7	Cts, ASD II, PS	RM, ASD patch, commissurotomy	RA	Diagnosis confirmed	Thrombus RA
8	Cts, ASD II, PAPVR	RM, ASD patch	RA	Diagnosis confirmed	
9	Cts, ASD II	RM, ASD patch	RA	Diagnosis confirmed	
10	Cts, ASD I	RM, ASD patch	RA	Diagnosis confirmed	BPI

Discussion

In our institution the incidence of cor triatriatum was 0.5% of all patients with CHD; 80% of these were children. All the children aged < 1 year showed a failure to thrive. Thus, this is the most common symptom, but, of course, this is not specific.

If the patients do not show any symptoms, diagnosis of cor triatriatum might be missed [12]. Typically, if diagnosis is made during adulthood, the orifice is not too narrow and does not cause a high degree of restriction of pulmonary venous flow [14–17]. Most of these patients do not suffer from additional CHD. Both our adult patients had an additional ASD, which was hemodynamically insignificant. In one of them, closure of the defect was favored after a stroke to prevent possible right-to-left embolic events, and the membrane was resected during ASD repair. The other one had nonspecific symptoms (de-

creased exercise tolerance) leading to the diagnostic and, subsequently, to the surgical procedures.

Resection of the membrane is easily performed and resolves the related symptoms like pulmonary congestion as well as, in the long run, the failure to thrive. This is in accordance with the findings of Oglietti et al., who described a very good postoperative course in 20/21 patients without associated complex CHD [18].

Preoperative pulmonary hypertension typically is secondary to the restricted pulmonary venous return, comparable to mitral stenosis [1, 2].

Interestingly, one of our patients (# 1) showed the typical signs of pulmonary hypertensive crisis immediately after surgery, which was treated successfully. After resection of the membrane with now unobstructed pulmonary venous return this might have been caused by a reperfusion injury. This patient did not have any sequelae.

In patient 3 the left atrial membrane was probably destroyed during cardiac catheterization. Intraoperatively, only residuals of the paper-thin membrane were found. Treatment without open heart surgery has been described with the use of an Inoue balloon during cardiac catheterization [19].

In this patient (# 3) later pulmonary venous obstruction due to thrombosis occurred; but it is as well possible that mild pulmonary venous obstruction was the cause of the thrombosis. Pulmonary venous obstruction might be masked in the presence of cor triatriatum [7]. This might have been the case in this patient as well.

One of our patients died immediately after surgery. This patient (# 5) had additional complex CHD. This, of course, is associated with a higher risk of surgery [18].

Surgery for isolated cor triatriatum is safe; the perioperative risk is comparable to surgery for ASDs [2, 11].

Tuccillo et al. recommended transesophageal echocardiography for the diagnosis of cor triatriatum [20]. All of our patients were diagnosed by transthoracic echocardiography. Intraoperative transesophageal echocardiography confirmed the diagnosis as did catheterization. In our institution all patients undergo cardiac catheterization including angiography prior to CHD surgery for proper surgical planning. Cardiac magnetic resonance imaging (MRI) would be an alternative [17], but was not available.

Most of our patients had a simple associated CHD (ASD, patent foramen ovale, bicuspid aortic valve, pulmonary valve stenosis), while more complex lesions occurred in only two of ten patients. This is comparable to the findings of Gheissari et al., who reported ASDs in 70–80% of their patients [21]. Bernhardt et al. underlined this thesis [11]. Rodefeld et al. reported twelve cases, of which four had complex CHD [2]. Interestingly, this workgroup found a left persistent upper caval vein in 50% of their patients, while we only had one patient with a left persistent upper caval vein.

According to the literature it seems that anomalies of the pulmonary venous return are quite common as well [2, 16, 22]. An embryologic relation to anomalous connection of the pulmonary veins has been assumed [6, 22–24]. Embryology has been discussed extensively [6, 22, 24] and is not subject of this article. We identified only one patient with additional partial anomalous pulmonary venous return.

Other reports found an association with complex CHD in almost half of their cases [16]. Interestingly, the more recent reports seem to have more simple than complex associated CHD [2].

Older studies like those of Niwayama [25] or van Praagh & Corsini [22] have the disadvantage that their anatomic descriptions are based on postmortems alone. This implies a bias to more complex CHD. Cor triatriatum alone is not lethal by all means, depending on the size of the orifice between the proximal and distal cavity.

Either the type of the dividing membrane (total/incomplete), the size of the connecting orifice, or the relation to the pulmonary veins were used to classify the different types of cor triatriatum [7, 8, 14, 23, 26]. We doubt that in such a rare disease (all in all about 150 cases in the literature) a classification makes sense. This might be interesting from the embryologic point of view, but for the treatment proper communication of the diagnostic team with the surgeons concerning the anatomic features is most important.

Clinical symptoms seem not to be associated with the anatomic subtype. Thus, we did not use any of the available classifications, but relied on good anatomic descriptions after noninvasive and invasive diagnostics for proper planning of surgery.

Since the first surgical approach in 1956 [13], surgery is the treatment of choice in symptomatic patients with cor triatriatum [2, 11]. If no obstruction over the membrane is patent, close follow-up with echocardiography, perhaps with pharmacologically induced stress, might be considered [11].

Surgical approach consists of left or right atriotomy, depending on the presence of an ASD, and excision of the membrane [2]. Of our patients only one was operated through a left atriotomy; this one had no additional CHD at all.

The results are encouraging, the outcome is more dependent on associated lesions than on the cor triatriatum itself. In our patients the only one who died had an additional single ventricle physiology. In the case series by van Son et al., two of the 13 patients died, both had additional complex CHD [16]. Rode-

feld et al., in their series of twelve patients, reported three deaths postsurgically; all had complex CHD [2]. Salomone et al. reported on 15 patients, three of whom died. Two of them had additional complex CHD, while the last one was in extremely poor condition and needed emergency surgery [8].

Conclusion

Pulmonary venous congestion might be caused by cor triatriatum. As surgical treatment is easy [2], searching for this diagnosis in patients showing these symptoms should be considered. Diagnosis nowadays can easily be achieved per echocardiography. Further investigations like transesophageal echocardiography and cardiac catheterization or MRI allow adequate therapeutic planning. Surgery resolves all related symptoms at once.

Conflict of interest: None. The authors declare that they have no financial or personal relations to other parties whose interests could have affected the content of this article in any way, either positively or negatively.

Address for Correspondence

Thomas Krasemann
Evelina Children's Hospital
St Thomas Hospital
Lambeth Palace Road
London SE1 7EH
UK
Phone (+44/207) 188-4562, Fax -4556
e-mail: Thomas.Krasemann@gstt.nhs.uk

References

1. Buchholz S, Jenni R. Doppler echocardiographic findings in 2 identical variants of a rare cardiac anomaly, "subtotal" cor triatriatum: a critical review of the literature. J Am Soc Echocardiogr 2001;14:846–9.
2. Rodefeld MD, Brown JW, Heimansohn DA, et al. Cor triatriatum: clinical presentation and surgical results in 12 patients. Ann Thorac Surg 1990;50:562–8.
3. Wolf WJ. Diagnostic features and pitfalls in the two-dimensional echocardiographic evaluation of a child with cor triatriatum. Pediatr Cardiol 1986;6:211–3.
4. Church WS. Congenital malformation of the heart; abnormal septum in the left oricle. Trans Pathol Logic Soc 1868;19:415–22.
5. Baweja G, Nanda NC, Kirklin JK. Definitive diagnosis of cor triatriatum with common atrium by three-dimensional transesophageal echocardiography in an adult. Echocardiography 2004;21:303–6.
6. Gharagozloo F, Bulkley BH, Hutchins GM. A proposed pathogenesis of cor triatriatum: impingement of the left superior vena cava on the developing left atrium. Am Heart J 1977;94:618–26.
7. Jacobstein MD, Hirschfeld SS. Concealed left atrial membrane: pitfalls in the diagnosis of cor triatriatum and supravalve mitral ring. Am J Cardiol 1982;49:780–6.
8. Salomone G, Tiraboschi R, Bianchi T, et al. Cor triatriatum. Clinical presentation and operative results. J Thorac Cardiovasc Surg 1991;101:1088–92.
9. Thilenius OG, Bharati S, Lev M. Subdivided left atrium: an expanded concept of cor triatriatum sinistrum. Am J Cardiol 1976;37:743–52.
10. Kacenelenbogen R, Decoodt P. Biplane transesophageal echocardiographic diagnosis of cor triatriatum. Chest 1994;105:601–2.
11. Bernhardt C, Seiler T, Kaulitz R, et al. Asymptomatic cor triatriatum in a 2¼ year old patient. Z Kardiol 2003;92:758–62.
12. Hogue CW Jr, Barzilai B, Forstot R, et al. Intraoperative echocardiographic diagnosis of previously unrecognized cor triatriatum. Ann Thorac Surg 1992;54:562–3.
13. Lewis FJ, Varco RL, Taufic M, et al. Direct vision repair of triatrial heart and total anomalous pulmonary venous drainage. Surg Gynecol Obstet 1956;102:713–20.
14. Loeffler E. Unusual malformation of the left atrium: pulmonary sinus. Arch Pathol 1949;48:371–6.
15. O'Murchu B, Seward JB. Images in cardiovascular medicine. Adult congenital heart disease. Obstructive and nonobstructive cor triatriatum. Circulation 1995;92:3574.
16. Van Son JA, Danielson GK, Schaff HV, et al. Cor triatriatum: diagnosis, operative approach, and late results. Mayo Clin Proc 1993;68:854–9.
17. Rumancik WM, Hernanz-Schulman M, Rutkowski MM, et al. Magnetic resonance imaging of cor triatriatum. Pediatr Cardiol 1988;9:149–51.
18. Oglietti J, Cooley DA, Izquierdo JP, et al. Cor triatriatum: operative results in 25 patients. Ann Thorac Surg 1983;35:415–20.
19. Huang TC, Lee CL, Lin CC, et al. Use of Inoue balloon dilatation method for treatment of cor triatriatum stenosis in a child. Cathet Cardiovasc Interv 2002;57:252–6.
20. Tuccillo B, Stumper O, Hess J, et al. Transoesophageal echocardiographic evaluation of atrial morphology in children with congenital heart disease. Eur Heart J 1992;13:223–31.
21. Gheissari A, Malm JR, Bowman FO Jr, et al. Cor triatriatum sinistrum: one institution's 28-year experience. Pediatr Cardiol 1992;13:85–8.
22. Van Praagh R, Corsini I. Cor triatriatum: pathologic anatomy and a consideration of morphogenesis based on 13 postmortem cases and a study of normal development of the pulmonary vein and atrial septum in 83 human embryos. Am Heart J 1969;78:379–405.
23. Marin-Garcia J, Tandon R, Lucas RV Jr, et al. Cor triatriatum: study of 20 cases. Am J Cardiol 1975;35:59–66.
24. Webb S, Kanani A, Anderson RH, et al. Development of the human pulmonary vein and its incorporation in the morphologically left atrium. Cardiol Young 2001;11:632–42.
25. Niwayama G. Cor triatriatum. Am Heart J 1960;59:291–317.
26. Arciniegas E, Farooki ZQ, Hakimi M, et al. Surgical treatment of cor triatriatum. Ann Thorac Surg 1981;32:571–7.