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Cor Triatriatum Associated with Partial Anomalous Pulmonary Venous Connection to the Coronary Sinus: Echocardiographic and Angiocardiographic Features

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SUMMARY. **An infant girl is described who had cor triatriatum and partial anomalous pulmonary venous connection of the left pulmonary veins to the coronary sinus, the first report of this combination of lesions. The infant also had a Dandy-Walker malformation and multiple facial and intrathoracic hemangiomas. The cardiac diagnosis was made by two-dimensional echocardiography. Cardiac catheterization and angiography confirmed the findings and also demonstrated a persistent left superior vena cava draining to the coronary sinus. The infant underwent successful surgical repair. Partial anomalous pulmonary venous connection and left superior vena cava not infrequently are associated with cor triatriatum. Although two-dimensional echocardiography is sensitive for the detection of cor triatriatum, preoperative cardiac catheterization is necessary to identify unequivocally systemic and pulmonary venous connections.**

KEY WORDS: **Cor triatriatum — Partial anomalous pulmonary venous connection — Doppler echocardiography — Two-dimensional echocardiography — Dandy-Walker malformation**

Cor triatriatum is a rare defect, initially, described in 1868 comprising 0.1%–0.4% of congenital heart disease [7, 8, 10]. In its usual form, an oblique fibromuscular membrane containing a single orifice divides the left atrium into a proximal chamber (also termed accessory chamber) which receives the pulmonary veins and a distal chamber which contains the left atrial appendage and mitral valve [8, 16]. An atrial communication or patent foramen ovale joining the right atrium and distal chamber is often present. Patients typically have symptoms of pulmonary venous obstruction.

Many variations of cor triatriatum have been described including those with no or multiple openings in the membrane [8, 15], an atrial communication joining the right atrium and proximal chamber [7–9, 15, 16], drainage of some pulmonary veins to the distal left atrial chamber [7, 15] or anomalously to the right side of the circulation, or associated with other complex cardiac defects [6, 9, 15, 16]. Symptoms can vary and include those caused by excessive pulmonary blood flow [9] and those caused by associated defects [7, 15]. There may be no symptoms if the opening in the membrane is large or multiple [8, 15].

Partial anomalous pulmonary venous connection (PAPVC) is present in 9%–25% of patients with cor triatriatum [1, 2, 9]. Previous reports have described direct anastomosis of pulmonary veins to the right atrium [7, 15] or drainage to the superior vena cava either directly [7, 17] or via the vertical and innominate veins [2, 7, 9, 16]. The diagnosis of PAPVC associated with cor triatriatum can be difficult and even missed at surgery [17] and has not been made previously by echocardiography. In this report, we describe a patient with cor triatriatum and a new associated variety of PAPVC, namely, drainage of the left pulmonary veins to the coronary sinus. The diagnosis was made by echocardiography, confirmed by cardiac catheterization and angiography which facilitated surgical repair.

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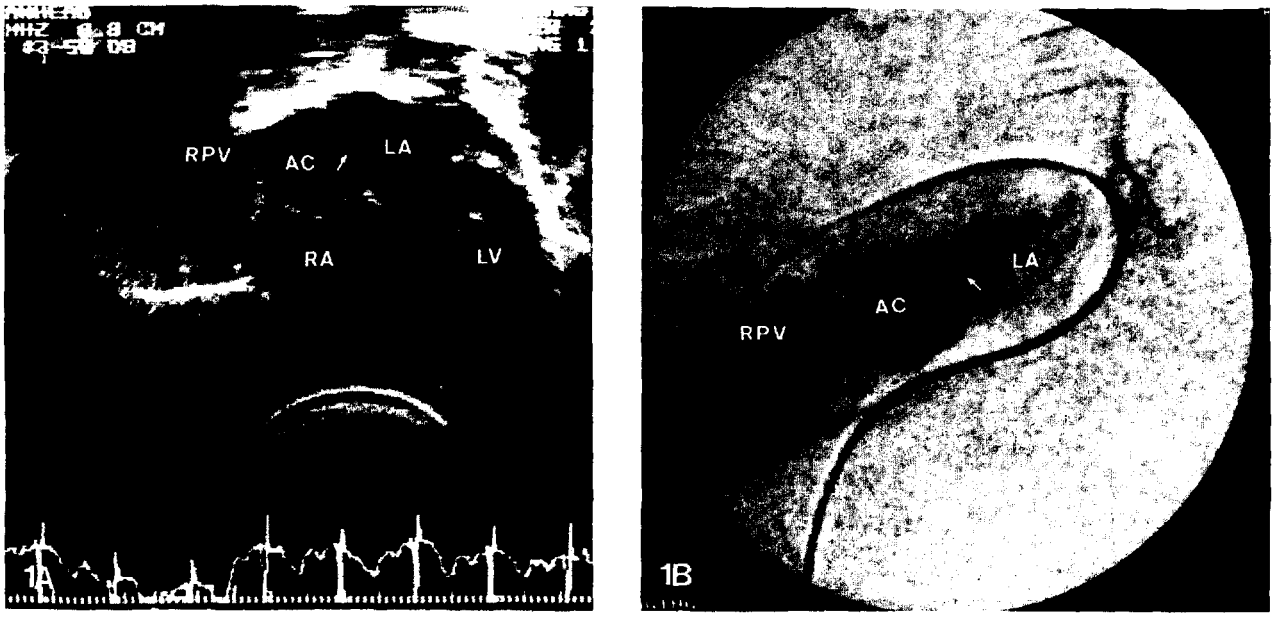

Fig. 1. **(A)** Subxiphoid long-axis echocardiogram demonstrating the intraatrial membrane containing a single orifice (*arrow*) that subdivides the left atrium into a proximal accessory chamber (*AC*) receiving the right pulmonary veins (*RPV*) and a distal left atrial chamber (*LA*). *LV*, left ventricle; and *RA*, right atrium. **(B)** Levophase of right lower pulmonary artery angiogram demonstrating drainage of right pulmonary vein (*RPV*) to accessory chamber (*AC*) and subsequent filling of distal left atrial chamber (*LA*) via stenotic opening in intraatrial membrane (*arrow*).

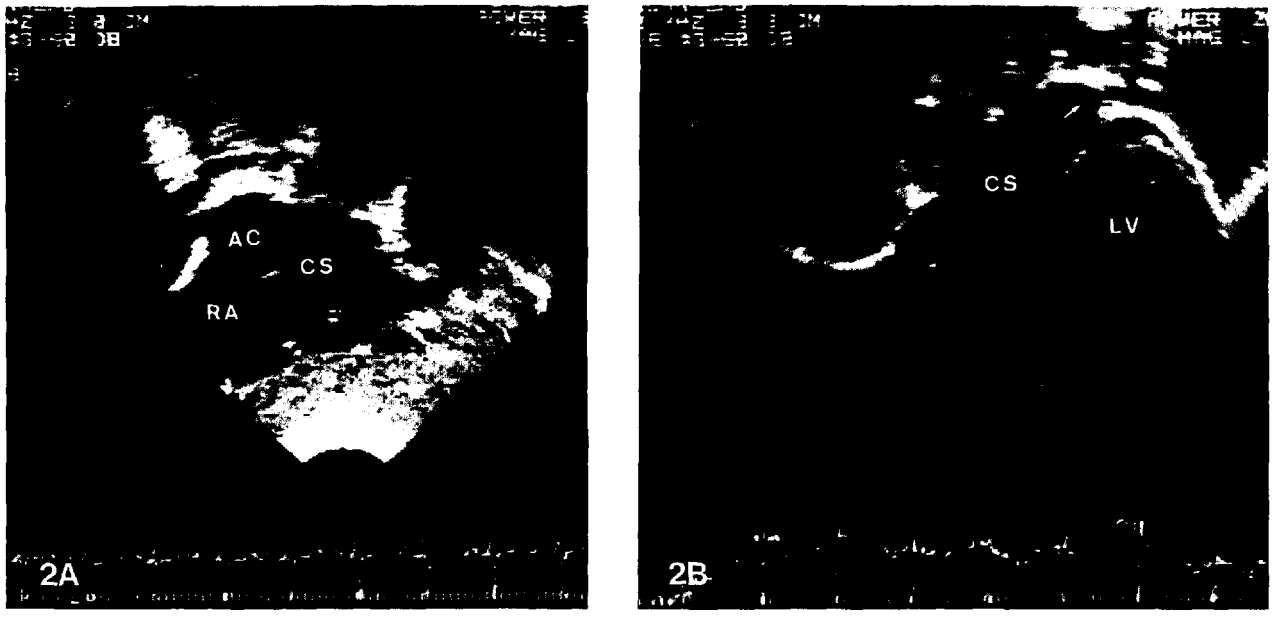

Fig. 2. **(A)** Subxiphoid long-axis echocardiogram with more posterior angulation than used in Fig. 1A demonstrating a greatly enlarged coronary sinus (*CS*) draining into the right atrium (*RA*). *AC*, accessory chamber. **(B)** With additional posterior angulation, a left pulmonary vein (*arrow*) draining into the dilated coronary sinus (*CS*) is visualized. *LV*, left ventricle.

Case Report

A full-term infant girl was considered normal at birth except for a small red facial nevus. By two months of age, she was hypotonic and had extensive right-sided hemifacial orbital capillary hemangiomas with occlusion amblyopia and exophthalmos. At four months of age, computed tomographic scans of the head and chest demonstrated this lesion with extension into the orbital space and cavernous sinus, a Dandy-Walker malformation with mild hydrocephalus, small cerebellar hemispheres (right more hypoplastic than the left), a retropharyngeal hemangioma with extension toward the mediastinum, and two separate intratho-

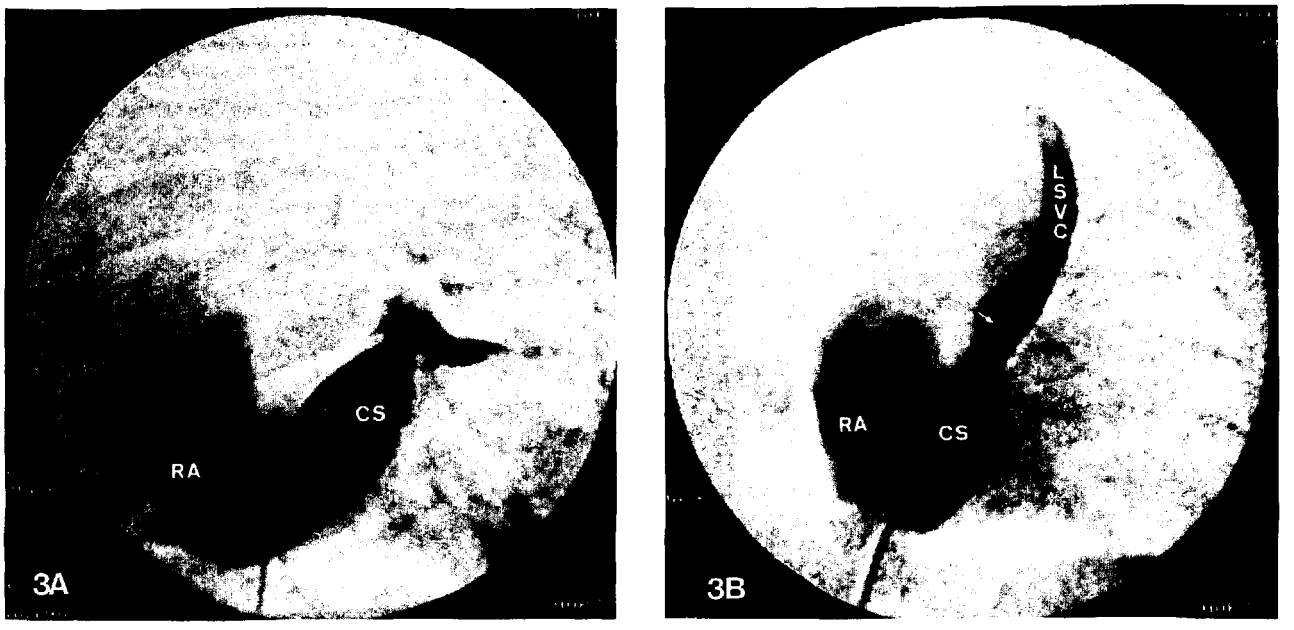

Fig. 3. Anterior–posterior angiogram with catheter positioned in left pulmonary vein (*LPV*) demonstrating drainage into the dilated coronary sinus (*CS*). *RA*, right atrium. **(B)** Anterior–posterior angiogram with catheter positioned in left superior vena cava (*LSVC*) demonstrating drainage of this structure into the coronary sinus (*CS*). Entry site of unopacified left pulmonary venous return can be seen (*arrow*). *RA*, right atrium.

racic hemangiomas. At four months of age, she developed tachypnea considered secondary to the vascular malformation. She received systemic prednisone and aminocaproic acid with no improvement. Nasogastric tube feedings were initiated and maintained to provide adequate caloric intake.

At seven months of age, the family relocated to Boston. On initial evaluation, the infant was acyanotic with weight 7.2 kg (25th percentile), length 66 cm (25th percentile), respirations 100/min, and pulse 130/min. Extensive hemifacial hemangiomas were present. There were diffuse bilateral rhonchi. S1 and S2 were single. There was no S3, S4, or ejection click. There was a grade 2/6 harsh systolic murmur along the left sternal border. Diastole was clear. The liver was palpable 2 cm below the right costal margin.

A chest radiograph demonstrated situs solitus, cardiomegaly (cardiothoracic ratio 0.62), increased pulmonary vascular markings and mild interstitial edema slightly more prominent in the right hemithorax, hyperinflation, and a widened superior mediastinum. In addition, a right upper lobe posterior density and a paravertebral mass in the lower thoracic spine consistent with hemangiomas detected on previous computed tomographic studies were present. An electrocardiogram showed normal sinus rhythm, right axis deviation, right atrial enlargement, and right ventricular hypertrophy. The hemoglobin was 12.5 gm/dl and hematocrit 38%.

A two-dimensional echocardiogram (ATL Ultramark 8) in the subxiphoid long-axis view (Fig. 1A) demonstrated a membrane in the left atrium containing a single orifice and which subdivided this chamber into a proximal portion receiving the right pulmonary veins and a distal chamber containing the left atrial appendage and mitral valve. With posterior angulation in the subxiphoid long-axis view (Fig. 2A and B), a greatly enlarged coronary sinus was visualized into which the left pulmonary veins drained. There was normal drainage of the hepatic veins to the right atrium and no coronary sinus septal defect. Doppler interrogation of the left atrial membrane estimated a 7 mmHg gradient between the proximal and distal chambers.

Cardiac catheterization (Table 1) and angiography (Figs. 1B and 3A and B) confirmed the echocardiographic findings. Right-sided oxygen saturations were elevated because of anomalous drainage of the left pulmonary veins into the coronary sinus; no pulmonary vein connected to the right superior vena cava or right atrium. The mean pulmonary arterial wedge pressure was normal in the left lung and elevated in the right. The distal left atrial chamber was entered from the right atrium via an anatomically patent foramen ovale. A persistent left superior vena cava (LSVC), but no left innominate vein, was detected (Fig. 3B). The LSVC was identified on retrospective interpretation of the echocardiogram.

At surgery, the patient was placed on cardiopulmonary bypass and the LSVC was cannulated and occluded. The right atrium was incised and the atrial septum and left atrial membrane, which contained a single orifice, excised. The coronary sinus was incised longitudinally so that it emptied into the left atrium. An atrial septal patch was placed that occluded the right atrial orifice of the coronary sinus. The mean pressure in the LSVC was initially 14 mmHg, rose to 30 mmHg immediately after occlusion, and was 15 mmHg after intracardiac repair was complete, a pressure identical to that of the right SVC. The LSVC was ligated. A hypoplastic left innominate vein was present. In the postoperative period, the patient did not develop a SVC syndrome. Cranial computed tomographic studies performed four weeks later demonstrated no cerebral edema and no change in the degree of hydrocephalus.

Discussion

Two-dimensional echocardiography is considered the most sensitive method for diagnosis of cor tri-

Table 1. Cardiac catheterization data[a]

Source	Oxygen saturation (%)	Pressure (mmHg)
IVC	81	—
RSVC	86[b]	$\overline{8}$
LSVC	77	$\overline{7}$, $\overline{23}$[c]
CS	87	—
RA	85	$\overline{5}$
RV	84	55/8
RPA	85	45/15 $\overline{28}$
LPA	84	55/20 $\overline{32}$
RPA wedge	—	$\overline{17}$
LPA wedge	—	$\overline{7}$
LA proximal	—	Not entered
LA distal	94	$\overline{8}$
LV	95	110/10

[a] CS, coronary sinus; IVC, inferior vena cava; RA, right atrium; RPA, right pulmonary artery; RSVC, right superior vena cava; RV, right ventricle; LA, left atrium; LPA, left pulmonary artery; LSVC, left superior vena cava; and LV, left ventricle.
[b] Angiography demonstrated reflux of blood from the RA to the RSVC; there was no pulmonary vein–RSVC connection.
[c] Pressure recorded after 15 s of occlusion by balloon-tipped end-hole catheter positioned superior to the left pulmonary veins entry sites.

atriatum [10]. M-mode echocardiographic features are nonspecific and subtle [12, 14]; angiography may fail to demonstrate the membrane [1, 9, 10]. Previous reports have outlined the two-dimensional echocardiographic features utilizing standard [10] and transesophageal [12] views. The present report describes a case of cor triatriatum illustrating for the first time the echocardiographic detection of PAPVC associated with cor triatriatum, the association of PAPVC to the coronary sinus with cor triatriatum, and the Doppler estimation of severity of stenosis of the obstructing membrane. The catheterization and angiographic information complimented the echocardiographic findings, documented the suspected unilateral pulmonary venous obstruction, and demonstrated the persistent LSVC draining to the coronary sinus.

Several development theories for cor triatriatum have been proposed [4, 7, 8, 15, 16]. Of these, the entrapment theory [16] is consistent with most anatomic observations. This view contends that cor triatriatum is produced by an abnormality of the endothelium of the right horn of sinus venosus leading to incomplete incorporation of the left atrial ostium of the common pulmonary vein to the posterior portion of the primitive atrium. The obstructing membrane is composed of left atrial and common pulmonary vein tissue [16]. When PAPVC is associated with cor triatriatum, the obstructing membrane is usually very stenotic. The elevated pressure in the proximal left atrial chamber contributes to retention of some primitive channels arising from the cardinal or umbilicovitelline systems [15]. In patients with cor triatriatum who have PAPVC, involvement of the left pulmonary veins is more common [17], which contrasts with the usual pattern of isolated PAPVC in which the right pulmonary veins are involved ten times more frequently than the left [13]. The coexistence of PAPVC with cor triatriatum is suggested by detecting lobar or unilateral variation in pulmonary congestion on a chest radiograph [2] or pulmonary wedge pressures at catheterization [2], features present in our patient. These findings are not specific and could be present in other conditions such as unilateral pulmonary vein stenosis, obstructive left atrial tumor or thrombosis, or extrinsic compression of the pulmonary veins.

Coronary sinus anomalies associated with cor triatriatum have been previously described, including total anomalous pulmonary venous connection [9], atresia of the right atrial ostium of the coronary sinus [15, 16], coronary sinus septal defect [6], and persistent LSVC draining to the coronary sinus. A persistent LSVC occurs in 0.5% of the general population [14], 3%–10% of patients with congenital heart disease [14], and 9%–32% of patients with cor triatriatum [1, 9, 16].

The preoperative detection of a persistent LSVC is helfpul in planning cardiopulmonary bypass techniques. Approximately three-quarters of patients with LSVC have agenesis or hypoplasia of the left innominate vein [3]. Ligation of a LSVC without adequate collateral drainage may result in central nervous system injury or the SVC syndrome [3]. Preoperative balloon occlusion of the LSVC with an end-hole catheter has been proposed to evaluate the capacity of the collateral venous system. A mean pressure rise greater than 10 mmHg after 15–20 min of occlusion has been considered a contraindication to ligation [3, 6]; in such cases, a baffle can be placed to direct the LSVC solely to the right atrium or the LSVC can be directly reimplanted into the right atrium. The duration of balloon occlusion at catheterization seems to be important. The high LSVC pressure observed during acute occlusion during cardiac catheterization may have decreased if balloon inflation was maintained for a longer period. Since the pressures in the left and right SVC were identical after the patient was weaned from cardiopulmonary bypass, the LSVC was ligated. No deleterious effects were detected. Because the right atrial ostium of the coronary sinus was occluded and a coronary sinus–left atrial communication made for repair of the anomalously draining left pulmonary veins, persistent drainage

of the LSVC into the coronary sinus would have been associated with cyanosis and risk of embolization and brain abscess [11].

The Dandy-Walker malformation is associated with a 10% incidence of facial angiomas and 5% incidence of congenital heart disease [5]. Cardiac conditions include ventricular septal defect, patent ductus arteriosus, tetralogy of Fallot, aortic coarctation, and dextrocardia [5]. There are no previous associations with cor triatriatum.

The preoperative diagnosis of cor triatriatum and associated anomalies is important for successful surgical outcome [1]. In patients with cor triatriatum, PAPVC and persistent LSVC are relatively frequent and the possible presence of these defects needs to be investigated. While some patients have been referred to surgery directly after echocardiographic studies [10, 12], we feel that cardiac catheterization is necessary if systemic and pulmonary venous connections are not clearly identified.

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References

1. Arcinicgas E, Farooki ZQ, Hakimi M, Perry BL, Green EW (1981) Surgical treatment of cor triatriatum. *Ann Thorac Surg 32*:571–577
2. Case Records of the Massachusetts General Hospital (Case 47-1969) (1969) *N Engl J Med 281*:1178–1187
3. Freed MD, Rosenthal A, Bernhard WF (1973) Balloon occlusion of a persistent left superior vena cava in the preoperative evaluation of systemic venous return. *J Thorac Cardiovasc Surg 65*:835–839
4. Gharagozloo F, Bulkley BH, Hutchins GM (1977) A proposed pathogenesis of cor triatriatum: impingement of the left superior vena cava on the developing left atrium. *Am Heart J 94*:618–626
5. Hirsch J, Pierre-Kahn A, Renier D, Sainte-Rose C, Hoppe-Hirsch E (1984) The Dandy-Walker malformation: a review of 40 cases. *J Neurosurg 61*:515–522
6. Lee ME, Sade RM (1979) Coronary sinus septal defect: surgical considerations. *J Thorac Cardiovasc Surg 78*:563–569
7. Lucas RV Jr (1983) Anomalous venous connections, pulmonary and systemic. In: Adams FH, Emmanouilides GC (eds) *Heart disease in infants, children, and adolescents*, 3rd edn. Williams and Wilkins, Baltimore, pp 458–490
8. Niwayama G (1960) Cor triatriatum. *Am Heart J 59*:291–317
9. Oglietti J, Cooley DA, Izquierdo JP, Ventemiglia R, Muasher I, Hallman GL, Reul GL Jr (1983) Cor triatriatum: operative results in 25 patients. *Ann Thorac Surg 35*:415–420
10. Ostman-Smith I, Silverman NH, Oldershaw P, Lincoln C, Shinebourne EA (1984) Cor triatriatum sinistrum: diagnostic features on cross sectional echocardiography. *Br Heart J 51*:211–219
11. Schick EC Jr, Lekakis J, Rothendler JA, Ryan TJ (1985) Persistent left superior vena cava and right superior vena cava drainage into the left atrium without arterial hypoxemia. *J Am Coll Cardiol 5*:374–378
12. Schluter M, Langenstein BA, Thier W, Schmiegel W, Krebber H, Kalmar P, Hanrath P (1983) Transesophageal two-dimensional echocardiography in the diagnosis of cor triatriatum in the adult. *J Am Coll Cardiol 2*:1101–1015
13. Snellen HA, van Ingen HC, Hoefsmit ECM (1968) Patterns of anomalous pulmonary venous drainage. *Circulation 38*:45–63
14. Snider AR, Ports TA, Silverman NH (1979) Venous anomalies of the coronary sinus: detection by M-mode, two-dimensional and contrast echocardiography. *Circulation 60*:721–727
15. Thilenius OG, Bharati S, Lev M (1976) Subdivided left atrium: an expanded concept of cor triatriatum sinistrum. *Am J Cardiol 37*:743–752
16. Van Praagh R, Corsini I (1969) Cor triatriatum: pathologic anatomy and a consideration of morphogenesis based on 13 postmortem cases and a study of normal development on the pulmonary vein and atrial septum in 83 human embryos. *Am Heart J 78*:379–405
17. Wolf WJ (1986) Diagnostic features and pitfalls in the two-dimensional echocardiographic evaluation of a child with cor triatriatum. *Pediatr Cardiol 6*:211–213