Cor triatriatum sinister

Elatafy Elmetwaly, MD

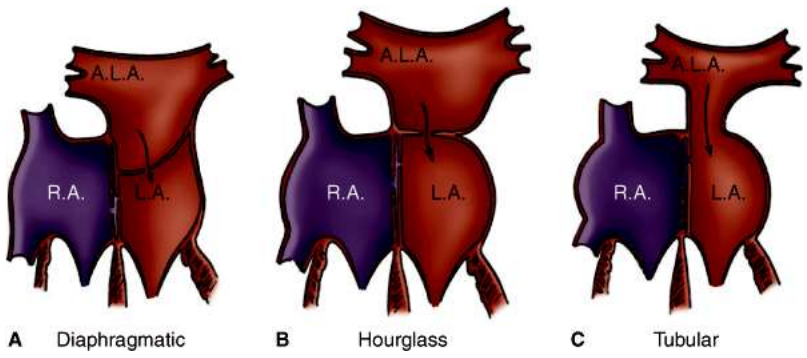
Introduction

That is a heart with 3 atria (triatrial heart), is a congenital anomaly in which the left atrium (cor triatriatum sinistrum) or right atrium (cor triatriatum dextrum) is divided into 2 compartments by a fold of tissue, a membrane, or a fibromuscular band.

Classically, the proximal (upper or superior) chamber receives venous blood, whereas the distal (lower or inferior) chamber is in contact with the atrioventricular valve and contains the atrial appendage and the true atrial septum that bears the fossa ovalis.

It may appear similar to a diaphragm or be funnel-shaped, bandlike, entirely intact (imperforate) or contain 1 or more openings ranging from small, restrictive-type to large and widely open.

Morphological types



A Diaphragmatic B Hourglass C Tubular

Church : **1st description in 1868**

Borst : **The term of cor triatriatum in 1905**

Miller et al : **Angiographic diagnosis in 1964**

Ostman-Smith : **Description by Echo. In 1984**

Vineberg & Gialloreto : **1st surgical correction in 1956**

Cor Triatriatum

Review of the Surgical Aspects with a Follow-up Report on the First Patient Successfully Treated with Surgery

By CHARLES R. JORGENSEN, M.D., RANDOLPH M. FERLIC, M.D., RICHARD L. VARCO, M.D., C. WALTON LILLEHEI, M.D., AND ROBERT S. ELIOT, M.D.

SUMMARY

A follow-up report on the first patient with cor triatriatum to undergo successful surgical treatment is presented. A second procedure was necessary 9½ years later because of stenosis of the initial surgically created orifice in the anomalous diaphragm. A review and ananalysis have been made of 17 cases from the literature reporting successful operations for this lesion. Total excision of the accessory septum utilizing cardiopulmonary bypass is presently the appropriate surgical treatment of this entity. Preoperative and postoperative hemodynamic data are discussed. Recognition and correction of this defect are emphasized in order to avoid permanent pulmonary vascular changes attended by a relatively fixed cardiac output.

Incidence

The incidence of cor triatriatum has been variously reported as 0.1-0.4%.

An incidence of 0.4% has been reported at autopsy of patients with congenital cardiac disease.

An incidence of 0.2% was reported among patients undergoing transesophageal echocardiography.

In high-volume echocardiographic laboratories, the incidence of cor triatriatum is less than 1 in 10,000.

However, this is expected to rise with the increasing use of cardiac diagnostic studies.

Congenital Heart Disease

Achievements in Congenital Heart Defect Surgery

A Prospective, 40-Year Study of 7038 Patients

Gunnar Eriksson, MD; Knut Liestøl, PhD; Egil Seem, MD; Sigurd Birkeland, MD; Kjell Johan Saatvedt, MD; Tom Nilsen Hoel, MD; Gaute Døhlen, MD; Helge Skulstad, MD; Jan Ludvig Svennevig, MD; Erik Thaulow, MD; Harald Lauritz Lindberg, MD

Miscellaneous Defects

The miscellaneous group (n=369) included double-outlet right ventricle (n=95), subvalvular and supravalvular aortic stenosis (n=65 and 21, respectively), Ebstein anomaly (n=11), congenitally corrected TGA (n=4), coronary artery anomalies (n=18), cor triatriatum (n=11), and vascular rings (n=48). A total of 97 patients (26.4% of all 369 patients in the miscellaneous group and 1.4% of all 7038 patients in the study) had defects lacking dominant features that justified classification into any of the previously mentioned categories.

Associated lesions

Tetralogy of Fallot tetralogy.

Double outlet right ventricle.

Coarctation of the aorta.

Partial anomalous pulmonary venous connection.

Persistent left superior vena cava with unroofed coronary sinus.

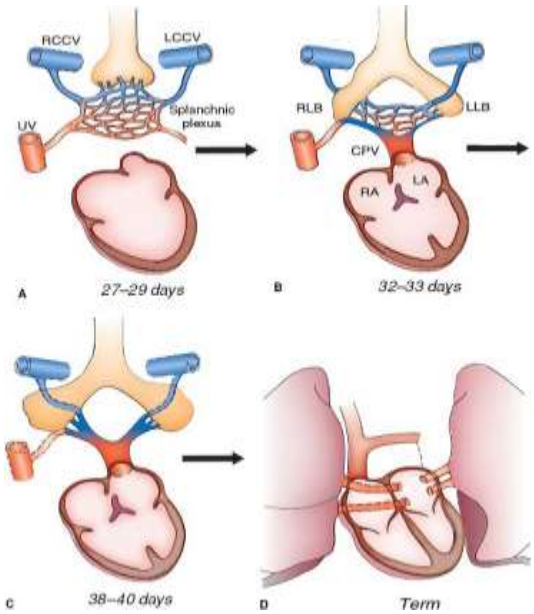
Ventricular septal defect.

Atrioventricular septal (endocardial cushion) defect.

Rarely, asplenia or polysplenia has been reported in these patients.

Development of pulmonary venous system

Embryology of PVs



Embryology of Cor Sx

Malincorporation theory

The most popular theory holds that cor triatriatum sinister occurs when the common pulmonary vein fails to incorporate the pulmonary circulation into the left atrium and the common pulmonary venous ostium remains narrow . The result is a septum-like structure that divides the left atrium into 2 compartments.

FIG. 7.—Hagenauer's theory. (1) Sinus venosus. (2) Septum primum. (3) Primitive auricle. (4) Pulmonary vein closed by expanding sinus venosus. (5) Septum secundum.

Embryology of Cor triatriatum Sx

Malseptation theory

Malincorpration theory fails to explain the presence of fossa ovalis and atrial muscle fibers within the walls of the proximal chamber where only a venous wall is supposed to be present In addition, several cases have been reported in which 1 or 2 pulmonary veins drain into the proximal (accessory) chamber and the others drain directly into the true left atrium. Others believe that the membrane dividing the left atrium is an abnormal growth of the septum primum

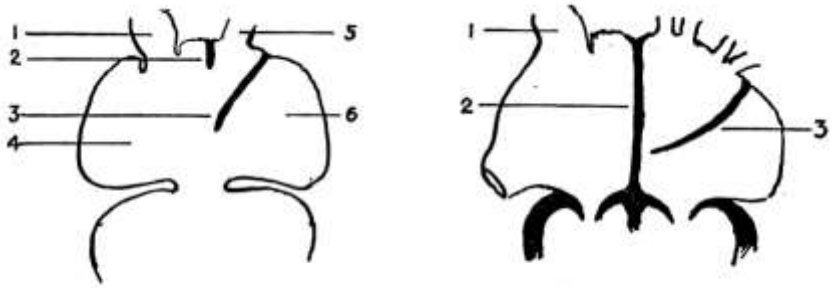
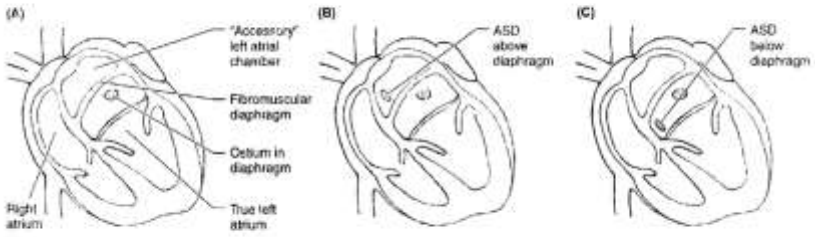


FIG. 6.—Borst's theory. (1) Sinus venosus. (2) Septum secundum. (3) Septum primum. (4) Right auricle. (5) Pulmonary vein. (6) Left auricle.

Entrapment theory

The right horn of the embryonic sinus venosus entraps the common pulmonary vein and thereby prevents its incorporation into the left atrium)..(***Van pragh theory***)

Classification of Cor triatriatum Sx



Loeffler classification

Based on the number and size of fenestrations in the fibro-muscular membrane:

Group 1: there is 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 2: there are one or few small openings in the intra-atrial membrane.

Group 3: The accessory chamber communicates widely with the true atrium by a large single opening.

1) Communication with LA (classic)

a. With intact atrial septum

b. CPVC communicating with RA

c. Atrial defect into lower chamber

2) No communication with LA (imperforate diaphragm)

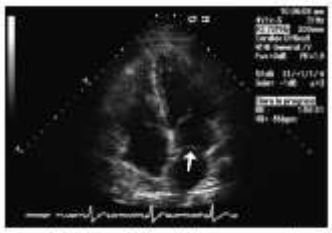
Drainage into RA

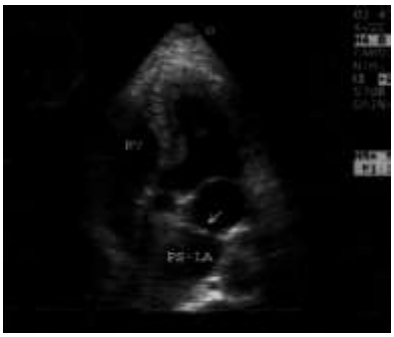
Drainage into coronary sinus

Drainage into systemic vein

Diagnostic work up

Echocardiography





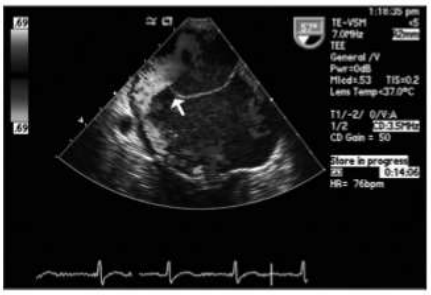
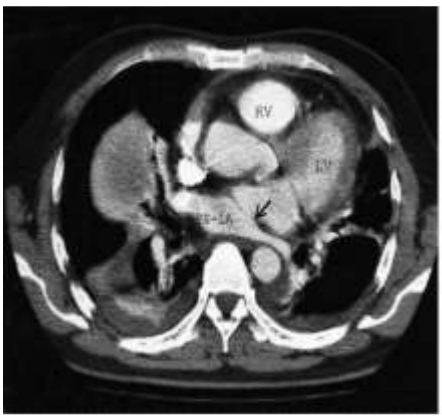
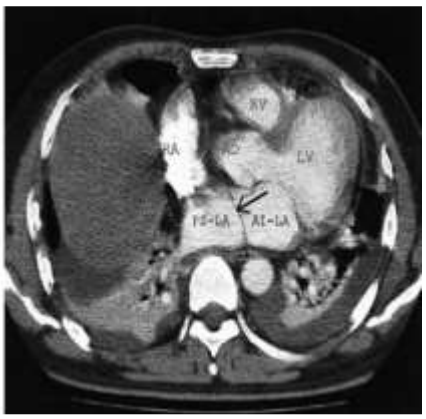


Figure 2) *Transesophageal echocardiography showing an isolated membranous cor triatriatum with a single orifice (arrow) and mitral regurgitation*





Cardiac Multislice Computed Tomography(MSCT)

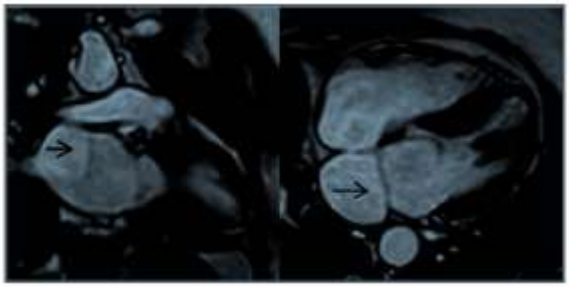


Figure 3. Cine magnetic resonance images using a steady-state free prec sion sequence clearly depict the mem brane within the left atrium (arrows); 2-chamber (left) and 4-chamber (right) views.

Cardiac Magnetic Resonance Imaging(MRI)

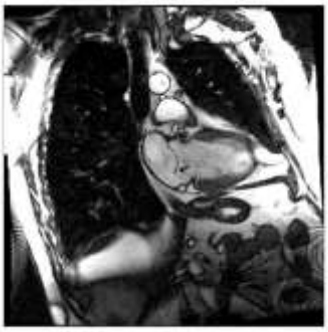


Figure 5 Magnetic resonance imaging of the heart showing cor triatriatum [arrow] with fenestration allowing for blood flow between the anterior and posterior chambers of the heart.

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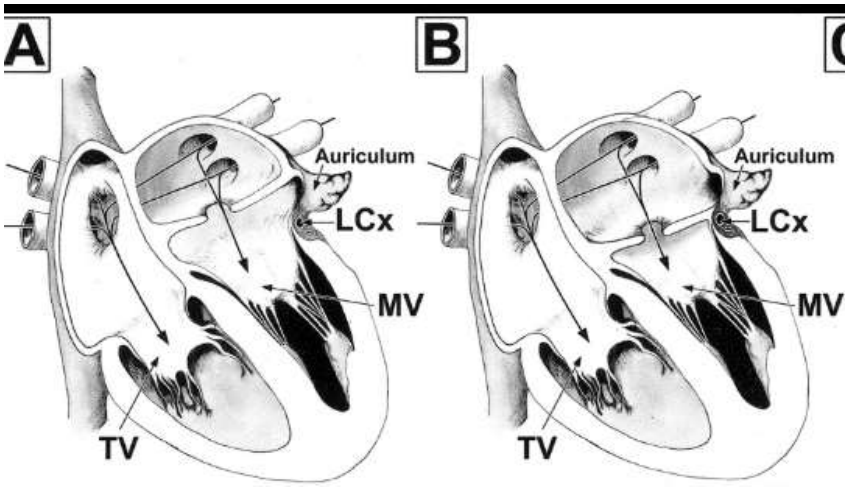
Editorial

Editorial: Cor triatriatum: An intraoperative diagnosis

What is difference between Cor and supramitral ring?



Indication for surgery

1. **Urgent indication for restrictive aperture**

2. **Operation could be necessary in the 1st year of life**

3. **In older patients with chronic symptom, operation is also urgently indicated.**

4. **In complex cor triatriatum, operation is indicated according to the associated lesion.**

Operative results

Surgical Repair of Cor Triatriatum Sinister: The Mayo Clinic 50-Year Experience

Pankaj Saxena, FRACS, PhD, Harold M. Burkhart, MD, Hartzell V. Schaff, MD, Richard Daly, MD, Lyle D. Joyce, MD, PhD, and Joseph A. Dearani, MD
Division of Cardiovascular Surgery, Mayo Clinic, Rochester, Minnesota

CONGENITAL HEART

Methods. Twenty-five patients underwent surgical correction of cor triatriatum between May 1960 and September 2012. There were 11 males and 14 females with a mean age of 27.4 years (age range, 1 day to 73 years).

Results. All patients underwent excision of cor triatriatum membrane using cardiopulmonary bypass. Twenty patients (80%) required concomitant cardiac surgical procedures. There was no early mortality. None

Conclusions. Surgical repair of cor triatriatum provides satisfactory early and long-term survival with low risk for additional intervention. Cor triatriatum with complex congenital anomalies may be associated with adverse outcome.

(Ann Thorac Surg 2014;97:1659–63)

Cor Triatriatum: Presentation, Diagnosis and Long-Term Surgical Results

Nelson Alphonso, MD, Martin A. Nørgaard, MD, Andrew Newcomb, MD, Yves d'Udekem, MD, Christian P. Brizard, MD, and Andrew Cochrane, MD

Victorian Pediatric Cardiac Surgery Unit, Royal Children's Hospital, Melbourne, Australia

Methods. A retrospective review of 28 consecutive patients diagnosed at the Royal Children's Hospital in Melbourne, Australia during a 22-year span from 1981 to 2003.

Results. There were 13 men (46%) and 15 women (54%). Fifteen patients were less than 1 year of age (neonates, [n = 7], 25%; infants, [n = 8], 29%). 17 patients (61%) had a communication between the right atrium and either the proximal or distal chamber. Fifteen patients (54%) had atypical cor triatriatum. Median age at presentation was 6 months (range, 0.6 to 240). Twenty-four patients (86%) had presented by 5 years of age. Five patients (18%) underwent emergency surgery. Median age at operation before treatment. Twenty-seven patients (96%) were treated surgically. The defect was approached through the right atrium in 26 patients (93%). There was 1 early death and 1 patient died 10 years after repair. Follow-up was 86% complete. At a median follow-up of 98 months (range, 0.2 to 284), all patients including those with atypical cor triatriatum were in New York Heart Association's function class 1. Post-repair survival was 96% and 88% at 5 and 15 years, respectively.

Conclusions. Surgery offers good early and long-term results for both classic and atypical cor triatriatum.

(Ann Thorac Surg 2005;80:1666–71)

Our cases

We have 7 cases operated on for resection of the membrane of cor triatriatum Sx.

3 cases with intact IAS.

Age ranged from 3 months to 7 years.

No operative or hospital mortality.

No major comorbidity.

Presentation of 2 particular cases

The 1st case

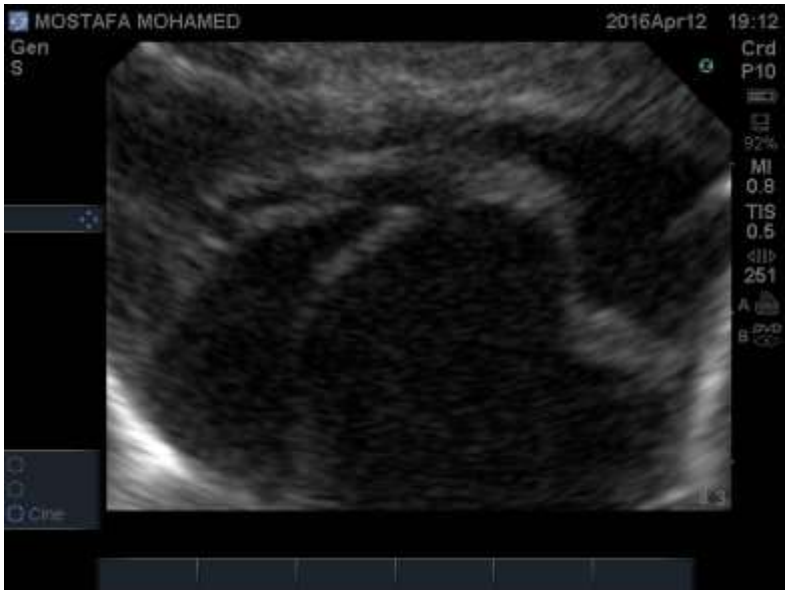
Male infant, aged 3 months.

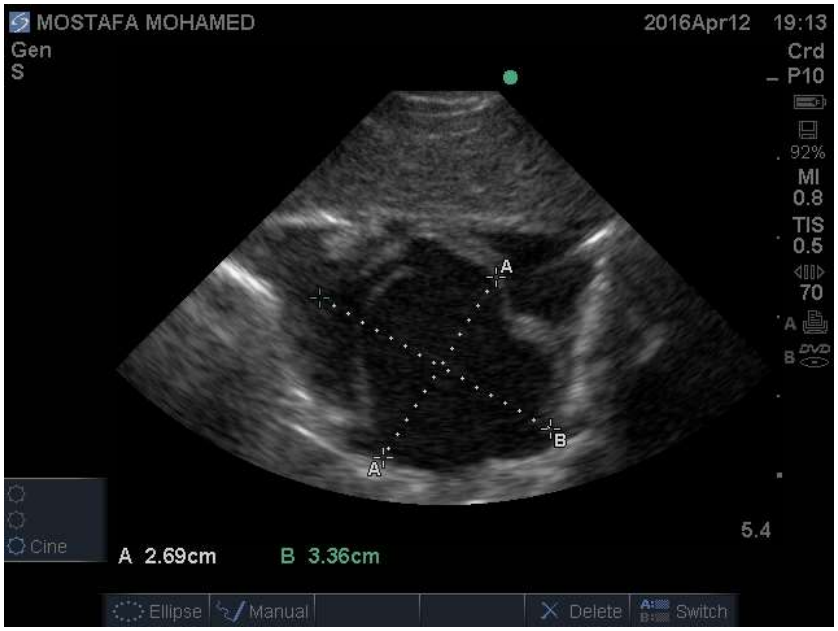
Weight 4 kgm

Presented with frequent chest infection and lung congestion.

The patient was on antifailure drugs.

Preoperative echocardiography

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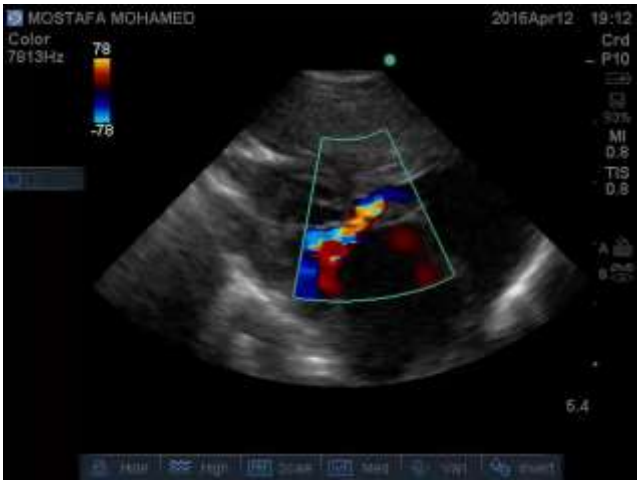


Surgery and postoperative course

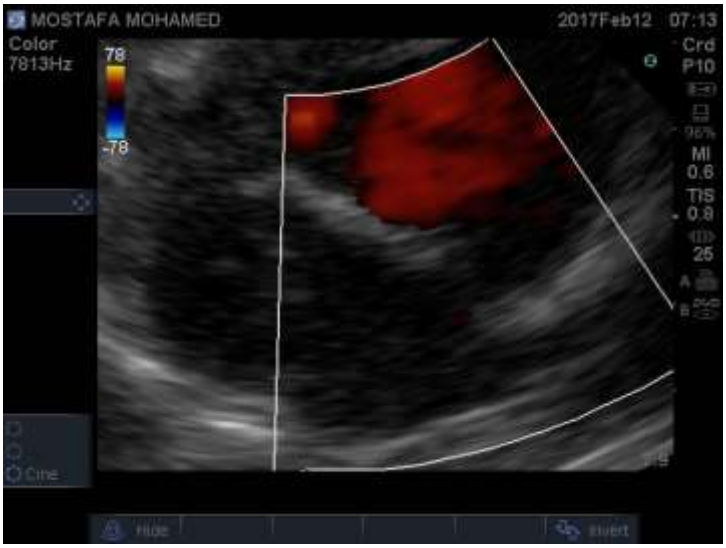
Conventional surgery was done for resection of the membrane.

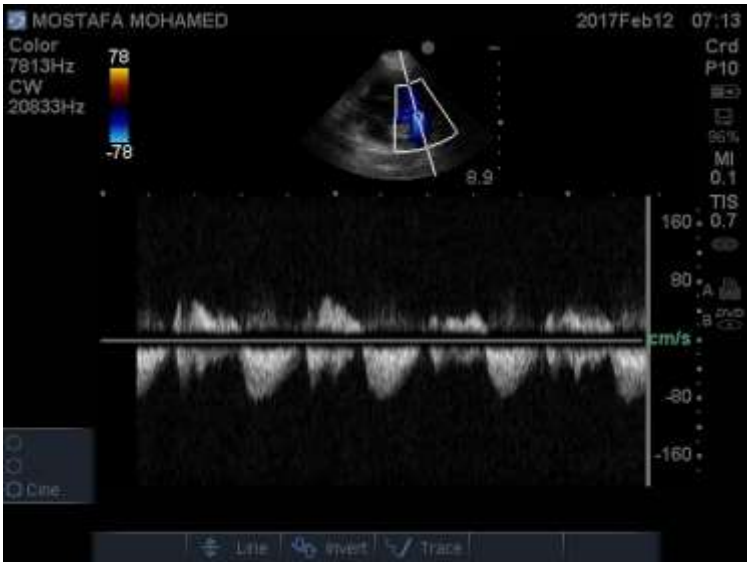
2 days of postoperative ventilation.

4 days in postoperative ICU.

Discharge after 8 days of hospitalization.

Postoperative echocardiography





The 2nd case

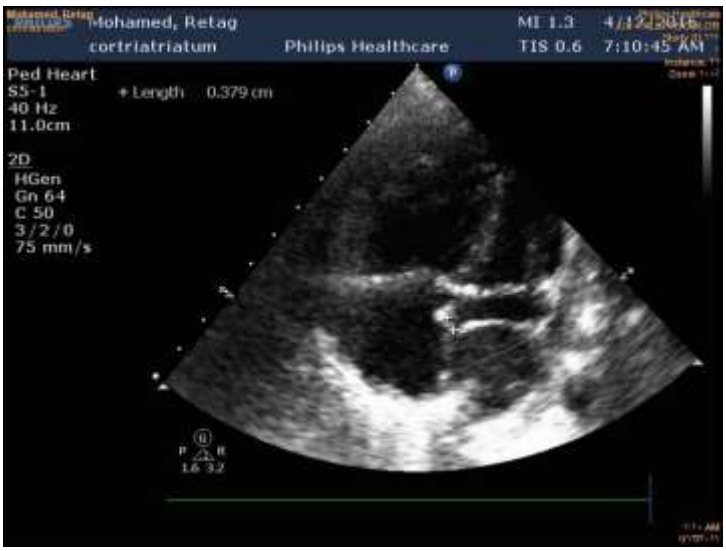
Female child, aged 2 years.

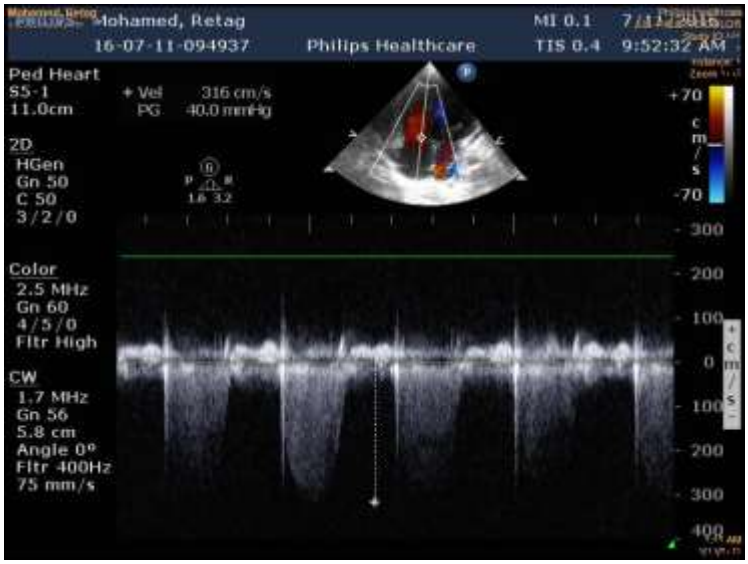
Weight 8 kgm.

Frequent hospitalization since birth.

The last hospitalization for 2 months with severe lung congestion and on maximal antifailure drugs.

Preoperative echocardiography

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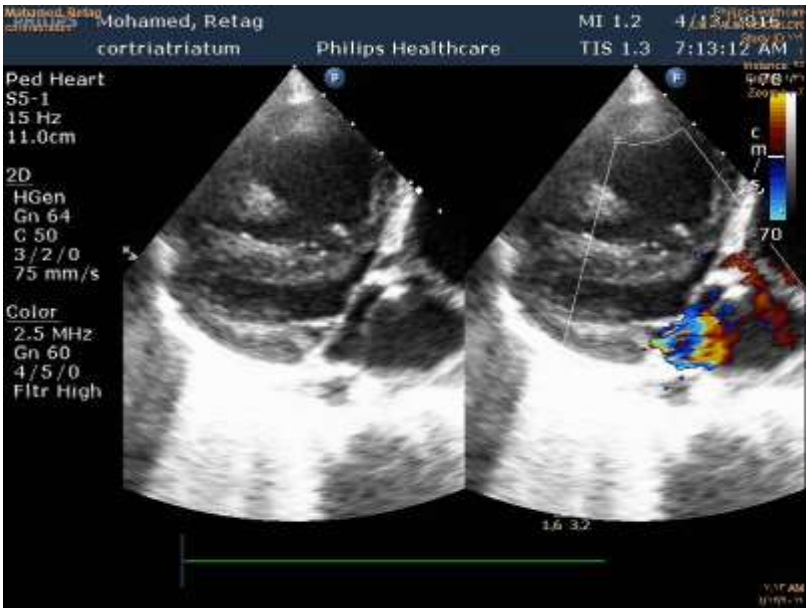
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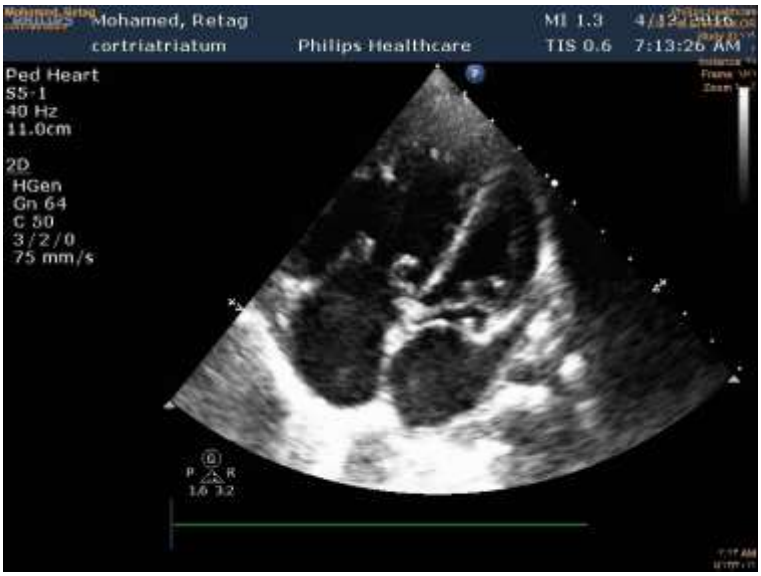


Surgery and postoperative course

Conventional surgery for resection of the membrane.

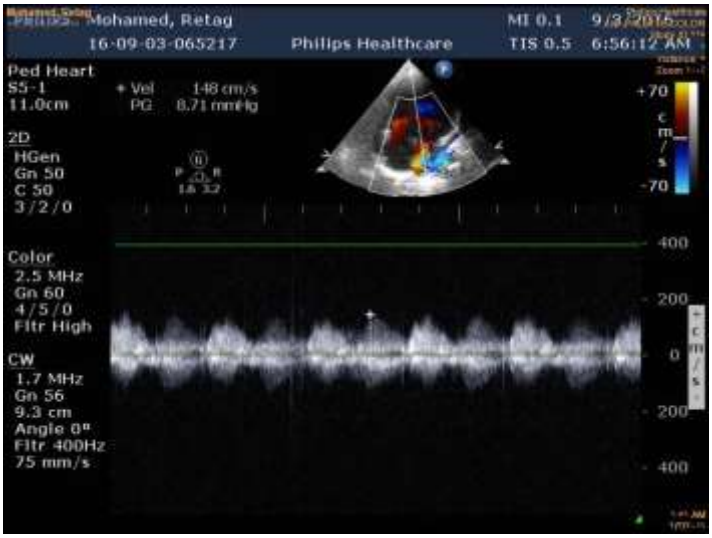
3 days on mechanical ventilation.

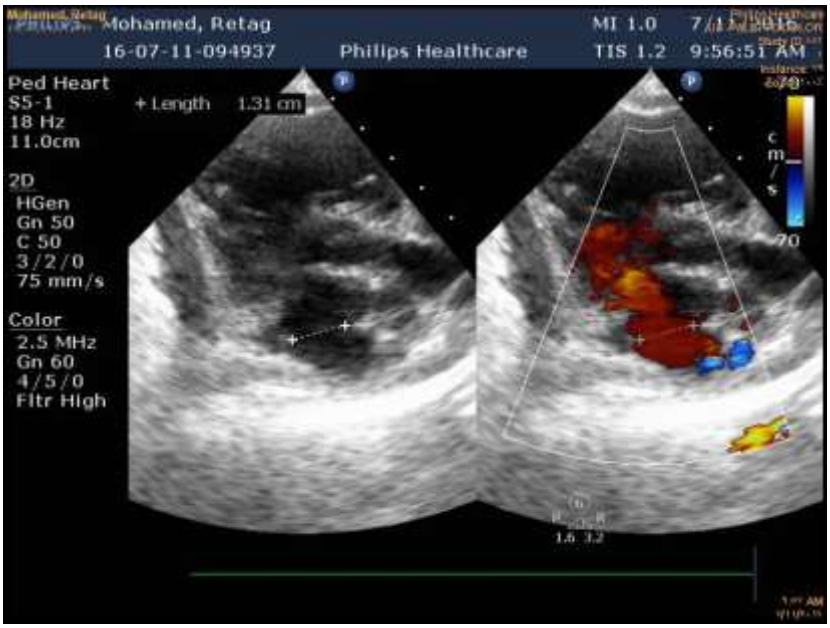
7 days in the postoperative ICU.

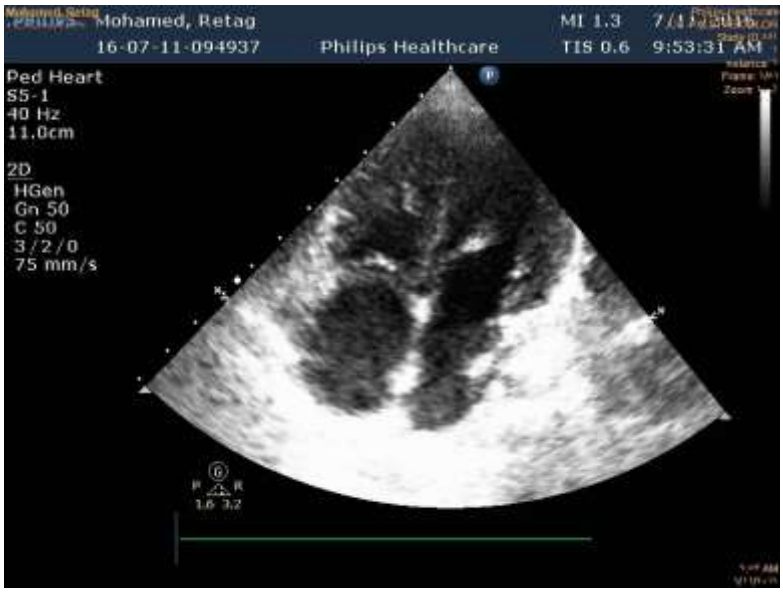
Postoperative fits with normal CT brain.

Discharged after 18 days from the hospital.

Postoperative echocardiography



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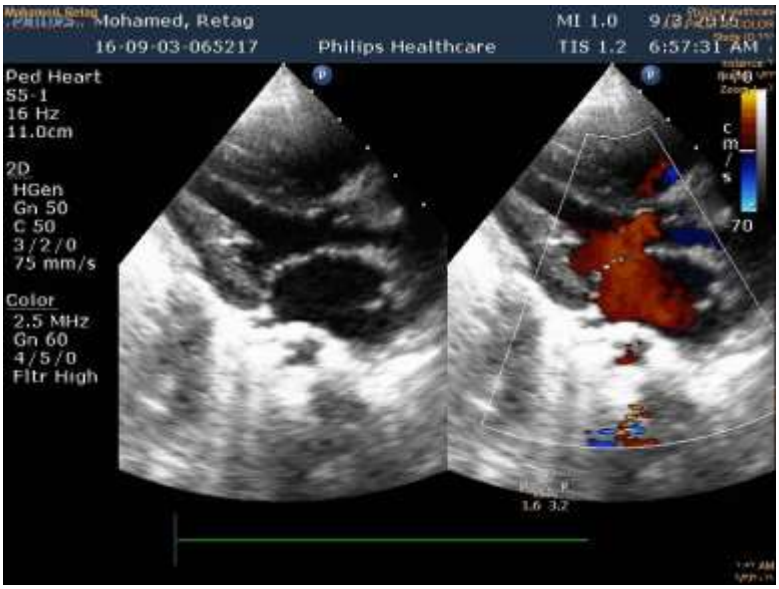
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Fltr High
cm/s
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Conclusion

Cor triatriatum is a very rare anomaly.

Surgical resection has excellent results.

Surgery should be carried on in symptomatic critical cor triatriatum as early as possible to avoid complications of pulmonary venous hypertension.

Intraoperative TEE may be helpful in assessing complete resection of the membrane.

THANK YOU