

COR TRIATRIATUM SINISTER AT 34 DAYS OLD BOY

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ABSTRACT

Cor triatriatum is a rare congenital cardiac anomaly. In this malformation, an anomalous septum divides the left or right atrium into two chambers. Cor triatriatum represent 0.1% of all congenital cardiac malformations and may be associated with other cardiac diseases in as many as 50% of cases. The natural history of this defect depends on the size of the communicating orifice between the upper and lower atrial chamber. If the communicating orifice is small, the patients is critically ill and may succumb at a young age (usually during infancy) due to congestive heart failure and pulmonary edema. If the connection is larger, patients may present in childhood or adolescent periode with a clinical pictures similar to that of mitral stenosis. We report a case of cor triatriatum in a 34 days old boy with chief complaint of shortness of breath and his lips became blue when crying since 2 week prior admission. The diagnosis was based on clinical features, chest radiography and echocardiography. The only treatment is surgical correction. Patient had total obstructive of pulmonary veins drain to left heart and partial AVSD so the prognosis is poor because he died one day after hospitalized. [MEDICINA 2014;45:65-70].

Keywords: *cor triatriatum, children*

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ABSTRAK

Cor triatriatum merupakan kelainan bawaan yang sangat langka. Pada kelainan ini jaringan ikat fibrus akan membagi atrium kanan atau kiri menjadi 2 bagian. Angka kejadian hanya sebesar 0,1% dari seluruh kelainan jantung bawaan dan biasanya ditemukan bersama-sama dengan kelainan jantung lainnya. Perjalanan klinis tergantung dari hubungan antara bilik atas dengan bilik bawah dari atrium yang terbagi. Bila lubang penghubungnya kecil, keluhan mulai muncul sejak lahir dan biasanya meninggal saat masa bayi. Bila penghubungnya besar, keluhan akan muncul pada usia anak atau dewasa dengan gejala klinis menyerupai stenosis mitral. Kami melaporkan kasus pada bayi usia 34 hari yang datang dengan keluhan sesak napas dan kebiruan pada bibir bila menangis. Diagnosis ditegakkan dari klinis, foto dada, dan ekokardiografi. Satu-satunya terapi adalah koreksi melalui pembedahan, tapi kasus meninggal sebelum pembedahan dilakukan. [MEDICINA 2014;45:65-70].

Kata kunci: *cor triatriatum, anak*

INTRODUCTION

Cor triatriatum is a rare congenital cardiac anomaly in which a fibromuscular membrane divides the left or right atrium into two chambers.^{1,2} In its most common form, about 54% are cor triatriatum sinister,³ the left atrium is divided into an upper chamber that receives the pulmonary veins and a lower

chamber that is related to the left atrial appendage and the mitral valve orifice. However, variable types of subtotal cor triatriatum are also noted, with only right or left pulmonary veins draining into the upper chamber. Cor triatriatum represent 0.1% of all congenital cardiac malformations and may be associated with other cardiac disease in as many as 50% of cases.^{5,6}

The natural history of this

defect depends on the size of the communicating orifice between the upper and lower atrial chamber.⁷ If the communicating orifice is small, the patients is critically ill and may succumb at a young age (usually during infancy) due to congestive heart failure and pulmonary edema. If the connection is larger, patients may present in childhood or adolescent periode with a clinical pictures similar to that of mitral stenosis.

Approximately 75% of patients die in infancy (generally from pulmonary hypertension) if the defect is unrepaired.⁷ If the communicating between the proximal and distal chambers is not restrictive or if an atrial septal defect allows decompression of the hypertensive left atrium, the prognosis is significantly improved.^{7,9,10}

The only treatment is surgical correction. Most postoperative deaths occur within the first 30 days. The early mortality rate in each large series was consistently 15-20%. Early deaths had a higher rate of associated severe cardiac anomaly.³ Long term results are excellent, with long term survival of 80-90% in patients surviving surgery.⁶

The purpose of this case report was to keep us more alert to this disease, to know the clinical symptoms, diagnosis, treatment, and prognosis.

THE CASE

JN, a 34 day old boy was referred from another public hospital to the Pediatric Emergency Department at Sanglah Hospital on November 23th, 2010 due to shortness of breath. Mother noted that the patient had shortness of breath since 2 week prior to admission and worsen since 1 week prior to admission. The other noted that he was often became blue in his lips while he was crying since 2 week prior admission. Feeding difficulties also noted same time with shortness of breath, and as a result he had weight lost in two weeks. There was no cough, fever, and common cold within two weeks was noted.

He was born term with no cry after delivered. His birth weight was 3200 grams. No visible abnormalities were found.

Upon admission (November 23th 2010) according to physical examination, he was looked pallor and irritable. Respiration rate was 78x/minute with the heart rate

190x/minutes regular in rhythm. His axillary temperature was 36.2^o C. Body weight was 3.7 kg compares to ideal body weight patient's nutritional status was considered as mild malnutrition. The oxygen saturation was 64% in room air and increase to 90% with oxygen of 10 lpm via oxygen hood.

There were no pale conjunctiva nor icteric sclera were noted. His lips looked cyanosis. There was no abnormalities on ear. Nasal flare was noted in this patient. There was no enlargement of lymph nodes or nuchal rigidity. On chest examination, we found the chest wall shape was normal and symmetric. There was subcostals, intercostals and suprasternal chest indrawing. The breath sounds was broncovesicular, with rales in lung base without wheezing. There was no precordial bulging. Ictus cordis was palpable but not accentuated on the 3rd intercostals space on left mid-clavicular line. Thrill was not palpable. There was right ventricular (RV) heave. Auscultation revealed accentuation of the pulmonary second sound with gallop, systolic murmur on the 3th parasternal line sinistra and diastolic murmur on the 2nd-3rd intercostals space on

midclavicular line sinistra with grade III/6. Abdominal examination revealed no distended abdomen with normal bowel sound. Liver was palpable 1/2-1/2, with blunt edges and soft, spleen was not palpable. Capillary refill time was more than 3 minutes with cutis marmorata and cyanosis seen on his extremities.

The result of the laboratory investigation showed WBC 12.2 K/uL, Hb 13.8 g/dl, HCT 42%, PLT 162 K/uL, CRP 0.428 mg/dl, blood sugar 43 mg/dl, sodium 135 mmol/L, potassium 4.619 mmol/L, chloride 95.63 mmol/L, calcium 5.905 mmol/L. Arterial blood gas showed metabolic acidosis and respiratory acidosis with pH 7.04, pCO₂ 35 mmHg, pO₂ 67 mmHg, HCO₃ 9.50 mmol/L, TCO₂ 10.6 mmol/L, BE -20 mmol/L, SaO₂ 81%.

Chest radiography revealed cardiomegaly with cardiothoracic ratio (CTR) of 72%, enlargement of right atrium, right ventricle with prominent pulmonary segment and increased of pulmonary vascular marking (**Figure 1**).

Echocardiography revealed cor triatriatum (**Figure 2**) 2 chambers of left atrium (LA), with functionally intact intra LA septal, without persisten foramen ovale

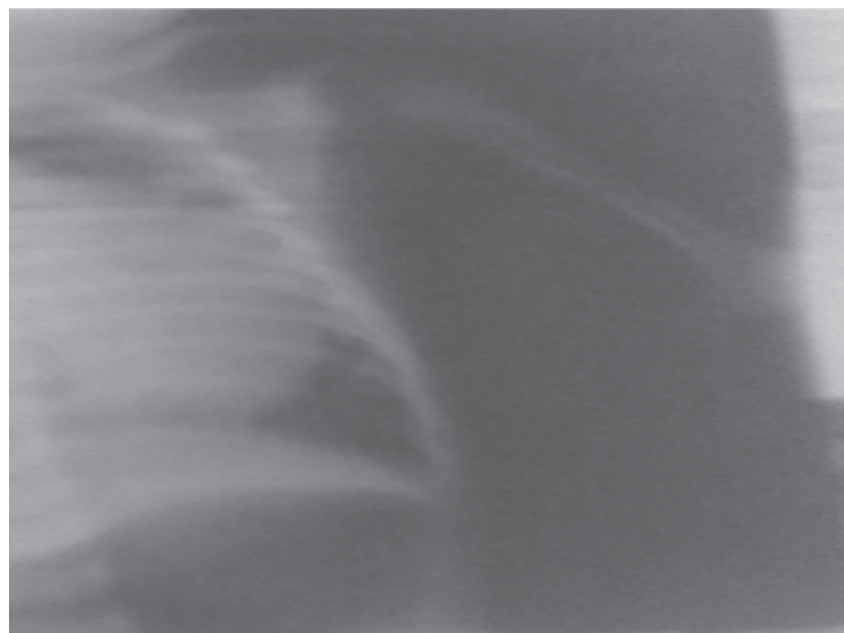


Figure 1. Chest radiography.

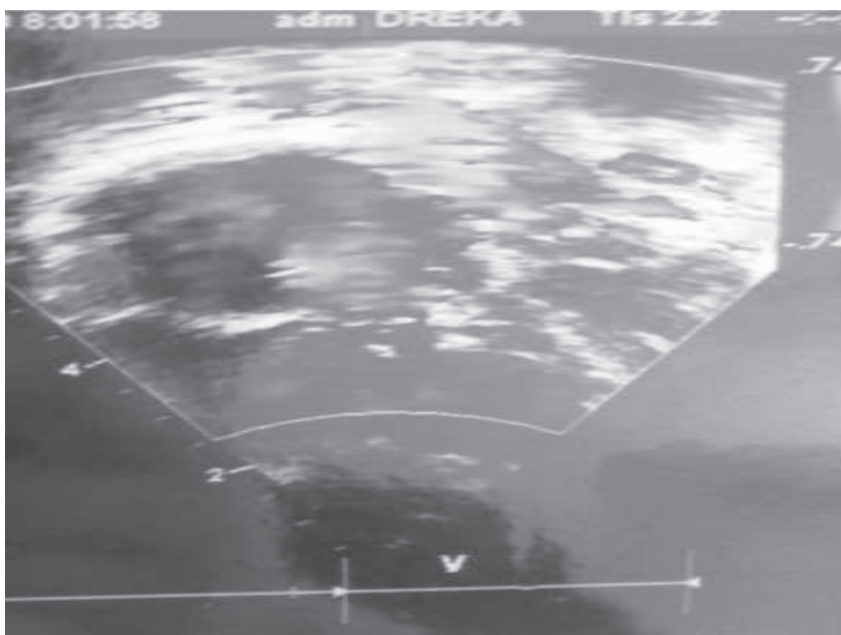


Figure 2. Echocardiography showed cor triatriatum, dilated RV, dilated RA, smallish LV.

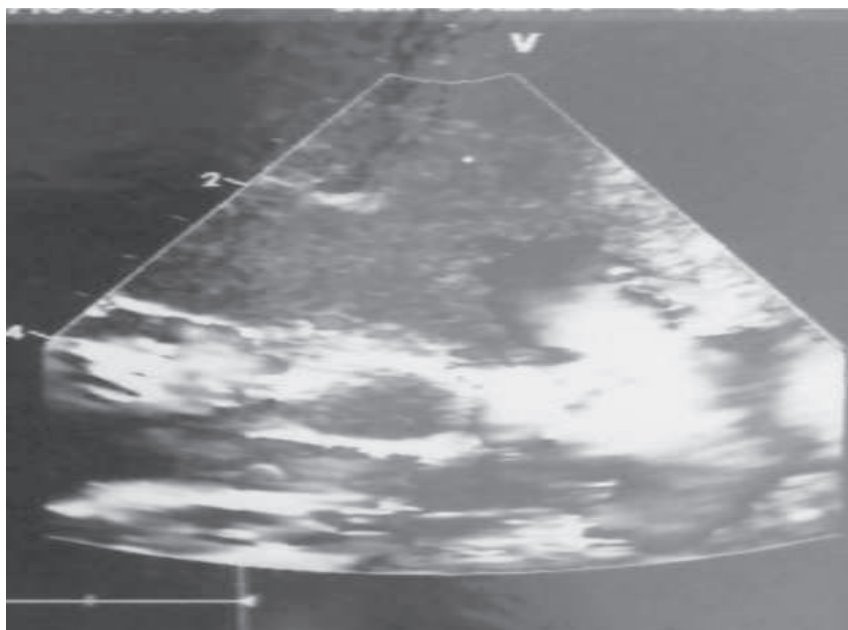


Figure 3. Echocardiography showed, small aorta and large RV.

(PFO)/atrium septal defect II), dilated right atrium (RA) & right ventricle (RV), smallish left ventricle (LV), small Ao (**Figure 3**), no persistent ductus arteriosus (PDA) severe pulmonary hypertension (PHT) (pulmonal arteri pressure estimation 99 mmHg), tricuspid regurgitation and partial atrioventricular septal defect (AVSD). Left ventricular

systolic function was normal with ejection fraction (EF) 77.2%.

Based on clinical manifestation, radiologic, and echocardiography investigation, the final diagnosis was **congestive heart failure functionally Ross IV + cor triatriatum with total obstructive of pulmonary veins drain to left heart + partial AVSD**

He was hospitalized in Pediatric Intensive Care Unit and treated with oxygen using continous positive airway pressure (CPAP), IVFD D51/4 NS 320 ml/day with the rate 12 micro drips/minute, intravenous furosemide at 4 mg every 12 hours, dopamine 5 micrograms/kg/minute. And we planned to refer to cardio-thoracic-vasculer surgeon for intra-atrial septectomi procedure. Unfortunately the patient died on November 24 2010, one day after hospitalisation.

DISCUSSION

Cor triatriatum sinister are more common form than cor triatriatum dextrum.³ In cor triatriatum sinister the left atrium is divided into an upper chamber that receives the pulmonary veins and a lower chamber that is related to the left atrial appendage and the mitral valve orifice.^{1,2} Cor triatriatum represents 0.1% of all congenital cardiac malformation,^{2,3} and may be associated with other cardiac defect in as many as 50% of cases. Examples of associated cardiac defects include atrial septal defect, persistent left superior vena cava with an unroofed coronary sinus, partial anomalous pulmonary venous connection, ventricular septal defect, and more complex cardiac lesions, such as tetralogy of Fallot, atrioventricular canal, and double outlet right ventricle.³ Depending on the presesence and localization of an atrial septal defect, there are four types of cor triatriatum (**Figure 4**). Type 1 (intact inter-atrial septum), type 2 (ASD to upper atrial chamber), type 3 (ASD to lower atrial chamber), and type 4 (ASD to both chamber). **In our case**, the diagnosis was cor triatriatum sinister with total obstruction of pulmonary veins drain to left heart, and its present during infancy (1 month). We also found partial AVSD and large ASD I in our case, which the anatomical was type 3 with ASD to lower atrial chamber.

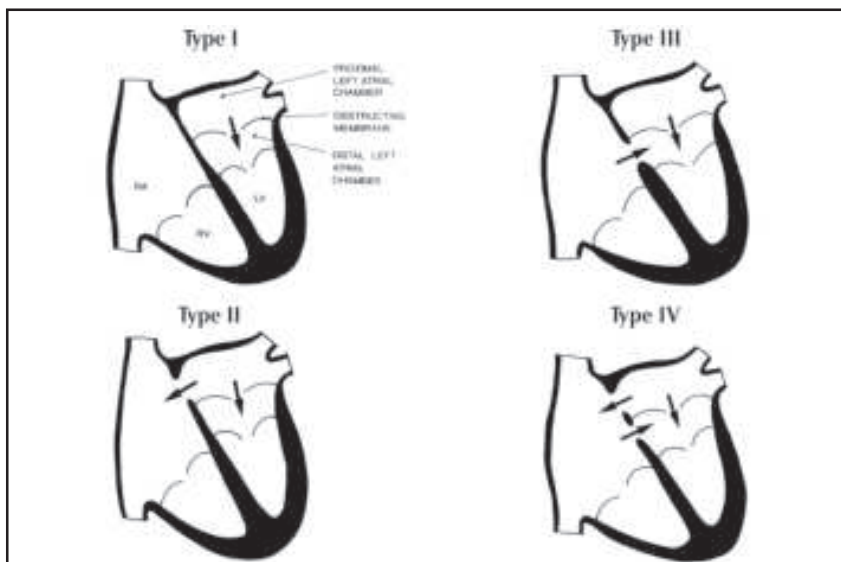


Figure 4. Anatomical types of cor triatriatum.

The natural history of this defect depends on the size of the communicating orifice between the upper and lower atrial chambers. If the communicating orifice is small, the patient is critically ill and may succumb at young age (usually during infancy) due to congestive heart failure and pulmonary edema. If the connection is larger, the symptoms will present during childhood or adolescents with a clinical picture similar to that of mitral stenosis. Cor triatriatum may also be an incidental finding when it is non obstructive. **In our case**, the symptom appeared at 1 month of age, with congestive heart failure and pulmonary edema because there was total obstruction of pulmonary vein drain to left heart, it means there is functionally no communicating orifice between upper and lower atrial chamber. Meaning, may be there was restrictive communicating but the flow can not be seen on color dopler echocardiography.

Approximately 75% of patients die during infancy (generally from pulmonary hypertension) if the defect is unrepaired. If the communication between the proximal and distal chambers is not restrictive or if an atrial septal defect allows decompression of the

hypertensive left atrium, the prognosis is significantly improved. **In our case**, the patient can not survive from cor triatriatum with total obstructive pulmonary veins drain to left heart and died at 35 day old, just one day after hospitalization.

Cor triatriatum is essentially a form of left atrial inflow obstruction and presents with signs and symptoms of pulmonary venous obstruction. Most patients presents during infancy with a restrictive opening in the membrane. These infants is usually present with evidence of low cardiac output, including pallor, diminished peripheral pulses, and tachypnea and tachycardia. Feeding difficulties, poor weight gain, and respiratory distress are common. Presentation later in life is less classic; however, when the patient becomes symptomatic, evidence of pulmonary venous obstruction predominates. In these patients, the membrane may become calcified with its orifice becoming smaller or the patient may develop mitral valve insufficiency. These patients may also present with arrhythmias secondary to an enlarged, hypertensive atrium. **In our case**, patient referred from another public hospital to the Pediatric Emergency Department

at Sanglah Hospital due to shortness of breath. Mother noted that the patient's have shortness of breath since 2 week prior to admission and worsening since 1 week prior to admission. The other noted that he was often became blue in his lips only while he was crying since 2 week prior admission. He had feeding difficulties since the shortness of breath occur, and as a result he had weight lost in two weeks. There was no cough, fever and common cold within two weeks were noted.

Despite the intracardiac defect, no characteristic murmur or pathognomonic physical characteristics are present. Signs of pulmonary venous obstruction and pulmonary hypertension are present. A right ventricular lift and accentuation of pulmonary second sound are frequent and may be accompanied by an early diastolic murmur of pulmonary insufficiency. Rales may be present in the lung base. A soft continous murmur may be present due to flow across the membrane. A murmur at the left sternal border is heard in patients with an atrial septal defect and a left-to-right shunt. A diastolic rumble of mitral stenosis at the apex is generally not heard with cor triatriatum. **In our case**, there was subcostal, intercostals, and suprasternal chest indrawing. The breath sounds were bronchovesicular, with rales in lung base without wheezing. Ictus cordis was palpable but not accentuated with impulse on the 3rd intercostals space in left mid clavicular line. Thrill was not palpable but there was RV heave. Auscultation revealed there was accentuation of the pulmonary second sounds with gallop, systolic murmur on the 3th parasternal line sinistra and diastolic murmur on the 2nd-3rd intercostals space on left parasternal line grade III/6.

Other clinical manifestation is caused by low cardiac output such as: pallor, tachypnea, tachycardia,

and diminished peripheral pulses. Children with cor triatriatum are typically small, suffering from poor weight gain. Patient may be dyspneic with a history of frequent pulmonary infections. They may have sign of right-sided heart failure, including distended peripheral veins, increased jugular venous pressure (JVP), and hepatomegaly. Signs and symptoms of pulmonary hypertension may be severe. **In our case**, patient revealed tachypneu with the the respiration rate was 78x/minutes, low peripheral oxygen saturation, it was 64% in room air and increase to 90% with oxygen 10 lpm via oxygen hood. Patient also had poor weight gain, because the body weight was 3.7 kg, compares to ideal body weight 3.9 kg, patient's nutritional status was considered as mild malnutrition. Liver was palpable 1/2-1/2, with blunt edges and soft consistency, spleen was not palpable. The capillary refill time was more than 3 minutes with cutis marmorata and cyanosis seen on his extremities.

From chest radiography, findings are usually nonspecific but may include pulmonary congestion with pattern of acute pulmonary edema in hilar areas. Patients may have mild RV enlargement and prominence of the pulmonary arterial segment. The dilated proximal chamber may produce the appearance of left atrial enlargement. Presence of an atrial septal defect or of an associated partial anomalous pulmonary venous connection adds pulmonary overcirculation to the pulmonary venous obstruction. Radiographic may then revealed significant right ventricular enlargement. **In our case**, the chest radiography showed cardiomegaly with CTR 72%, enlargement of right ventricle, right atrium with prominent pulmonal segment. There also seen increased of pulmonary vascular marking as a pattern of acute pulmonary edema.

Echocardiography is often sufficient for diagnosis and is the diagnostic modality of choice. Demonstration of the membrane by two-dimensional imaging, evaluation of turbulent flow on colour Doppler and determination of any gradient across the membrane on continuous wave Doppler can be performed with echocardiography. Coexisting congenital anomalies can also be well characteristic by echocardiography. **In our case**, we found the membrane dividing the left atrium as the characteristic of cor triatriatum sinistra. It can also showed large atrial septal defect, partial AVSD as a coexisting cardiac congenital anomalies in cor triatriatum.

Surgery is the treatment of choice. Open correction is currently preferred over closed (percutaneous) procedure. The procedure is performed on cardiopulmonary bypass through an atrial incision with complete resection of the diaphragm. For the medical care the goal in cor triatriatum is to reduce the symptom caused by pulmonary venous congestion until definitive surgical therapy can be performed. It is continued in the postoperative period until the pulmonary resistance falls and right ventricular performance improves. Mainstay of treatments are inotropic agents and diuretics. **In our case**, we treated the patient with oxygen using CPAP, IVFD D51/4 NS 320 ml/day with the rate of 12 micro drips/minute, intravenous furosemide at 4 mg every 12 hours, dopamine 5 micrograms/kg/minute. And we planned to refer to cardio-thoracic-vascular surgeon for intra-atrial septectomy procedure. Unfortunately the patient died on November 24 2010, one day after hospitalization.

Prognosis in symptomatic infants without surgical repair was poor with a mortality rate of 75%. Presence of associated cardiac anomalies adversely affect

prognosis. If the connection between the proximal and distal chamber is not restrictive or if an atrial septal defect is present, prognosis is improved. **In our case**, the patient had total obstructive of pulmonary veins drain to left heart and partial AVSD so the prognosis is poor because he died one day after hospitalized.

SUMMARY

We report a case of Cor Triatriatum at 34 days old boy. The definitive diagnosis was established by clinical feature, chest radiography and echocardiography. Surgical is the only treatment, but unfortunately the case died one day after hospitalization.

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