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CASE REPORT

A Rare Case Report of Cor triatriatum with Atrial Septal Defect Diagnosed at an Adult Age

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Abstract:

Cor triatriatum dexter is a rare congenital heart defect divides the right atrium into two chambers with a varied clinical presentation ranging from asymptomatic to right heart failure. Accurate diagnosis is imperative as it may affect clinical decision making. This is a case of cor triatriatum dexter in a 53-year-old woman with Atrial septal defect with moderate tricuspid regurgitation which was diagnosed by Transthorasic Echocardiography (TTE) and later confirmed by Trans-esophageal Echocardiography (TEE).

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Introduction:

Cor triatriatum is an uncommon congenital heart malformation defined by an abnormal septation within the atrium (left or right) leading to inflow obstruction to the respective ventricles. It exists either in isolated classical form or may be associated with simple to complex congenital cardiac anomalies. Several anatomical variants exist even in the classical form, and therefore, it may require multimodal diagnostic modalities to characterize and differentiate for better percutaneous interventional or surgical planning. It commonly presents in infancy but may remain undetected till death. Symptomatology typically mimics mitral and tricuspid stenosis in sinister and dexter varieties, respectively. However, features of systemic embolization, heart failure, atrial fibrillation, cyanosis, cardiac asthma, syncope, and sudden cardiac arrest have also been reported in the literature. Surgical correction under cardiopulmonary bypass is the preferred treatment. Nevertheless, balloon dilatation may be considered in anatomically compatible variants and in special circumstances, such as heart failure, pregnancy, or as a bridge to definitive treatment.[1] Cor triatriatum dexter is a rare congenital heart malformation in which a persistent right sinus venosus valve divides the right atrium into two chambers. Before echocardiography, this anomaly has been rarely diagnosed before surgery or death. This is a case of cor triatriatum dexter in an adult with lifelong exertional cyanosis and dyspnea. A definitive diagnosis of cor triatriatum dexter with associated heart defects was best made by transesophageal echocardiography at 47 years of age. Subsequent surgical intervention confirmed all of the echocardiographic findings and successful correction of the defects was performed.[2]

Case report:

Mrs Begum , 53 years of age, muslim,housewife, hailing from Badda, Dhaka, admitted on 16/07/2023 at cardiology department, BangaBandhu Sheikh Mujib Medical University(BSMMU) with the complaints of Shortness of Breath and Palpitation for 6 months. Her shortness of breath was insidious onset 6 months back, gradually progressive initially felt during moderate to severe exertion (NYHA-II),now breathlessness occur even in mild exertion and sometimes even at rest(NYHA IV), awakening from sleep at mid night and gets relieved by staying in sitting position. SOB not associated with chest pain,cough,hemoptysis,no seasonal or diurnal variation, not aggravated by cold dust and fumes. She also complains of palpitation for last 6 month which was episodic , increased during exertion and occasionally on rest, persist for short period, relieved by rest and sometime subside spontaneously. But progressively increased

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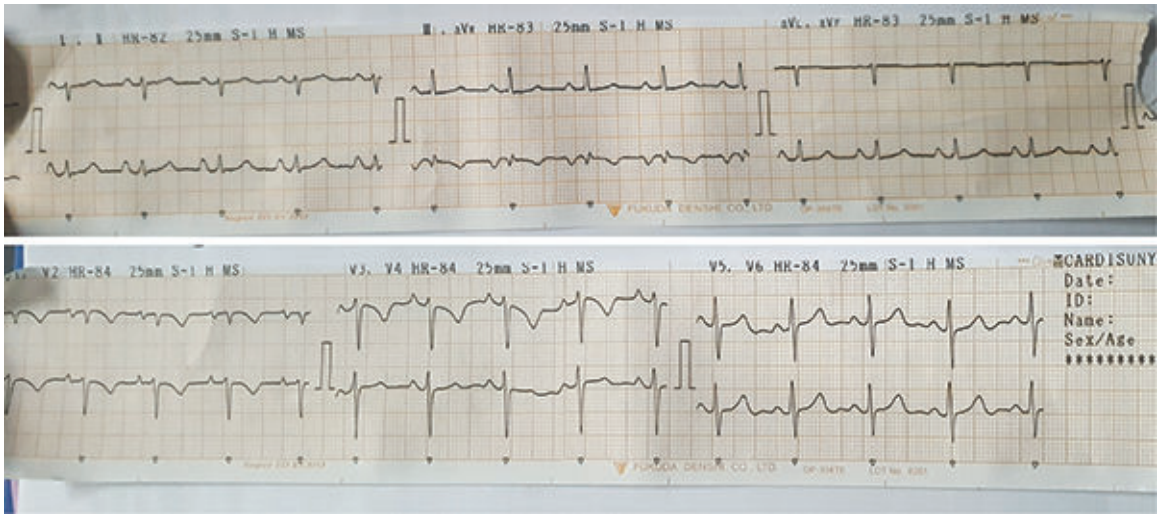

Figure 1: *12 leads ECG reveals: Right axis deviation*

duration of episodes for last 1 month, not associated with any sweating , tremor, headache, nausea, syncope, increase

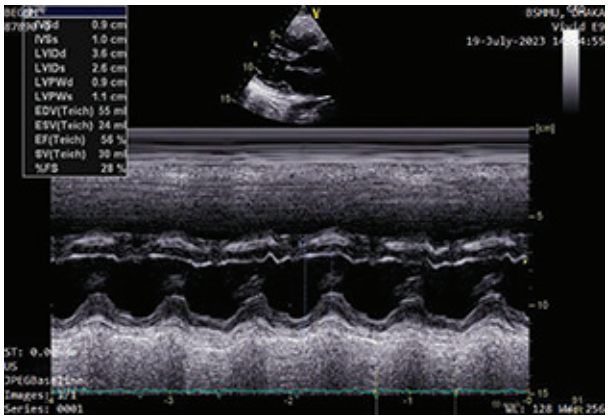

Figure 2: *2D and M-mode Echocardiography Para-sternal long axis view reveals: flattening of septum, dilated RV dimension, with normal LV systolic function.*

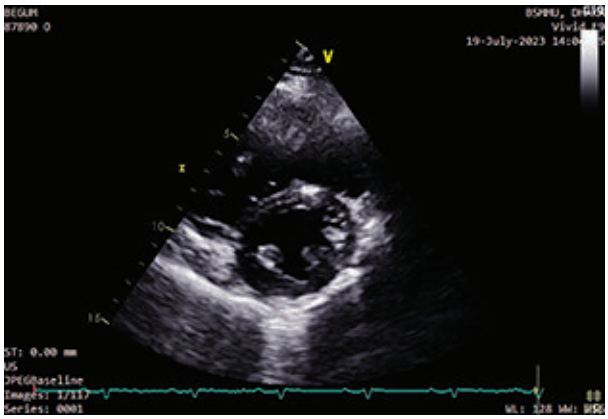

Figure 3: *Para sternal short axis view shows dilated RV.*

frequency of micturition, weight loss, pallor . No history of dysphagia, voice change, unilateral weakness,no history suggestive of GI and genitourinary blood loss or any other

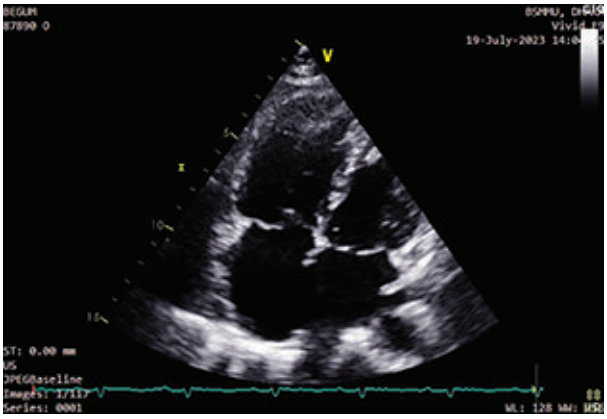

Figure 4: *Apical 4 chamber RV focus view shows echo drop out in mid inter atrial septum.*

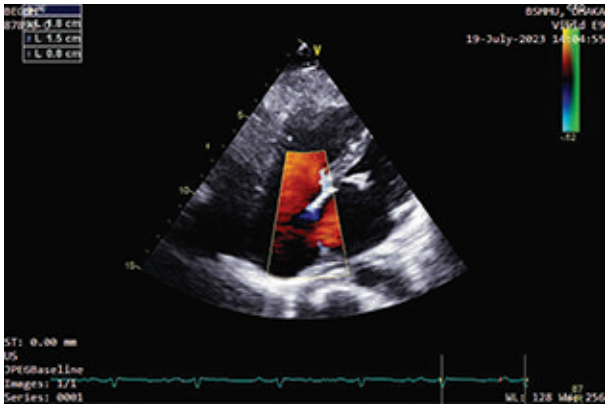

Figure 5: *Atrial septal defect, secundum type with Left to right shunt.*

bleeding manifestation, leg swelling or abdominal swelling. she did not give any history of joint pain, swelling or fever in childhood. She is mother of 5 childrens, all delivered by NVD, she does not report any complications during and after her pregnancies. She had her menopause at the age of 46 yr. She has 6 siblings, no history of heart disease in family. With this complain she visited to several local physician and received several medication(could not mention the name but with no improvement). Now she got admitted through OPD for further evaluation and management. On examination, she was ill looking and anxious, Decubitus in sitting position, mildly Anaemic, Pulse: 110 beats/min with drop beat, low volume, all peripheral pulses are palpable and condition of vessel wall is normal, JVP : raised about 5 cm, Blood pressure: about 5 cm, Blood pressure was 110/70 mm of Hg no postural drop, respiratory rate: 16 breaths/ min, Apex beat is found on left 5th intercostal space, 9 cm from midsternal line ,no specific character noted, Left parasternal heave is present, palpable p2 and epigastric pulsation is present.

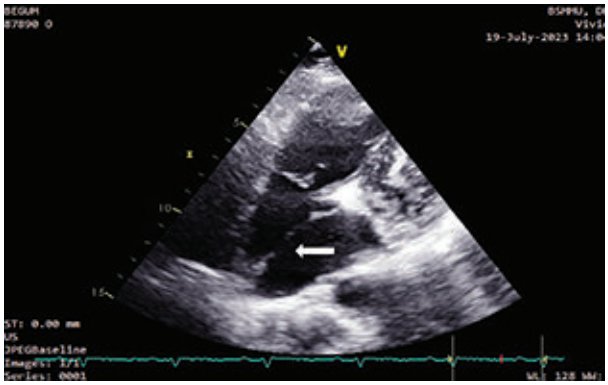

Figure 6: *Modified Apical 4 chamber view. Arrow shows abnormal septation in RA*

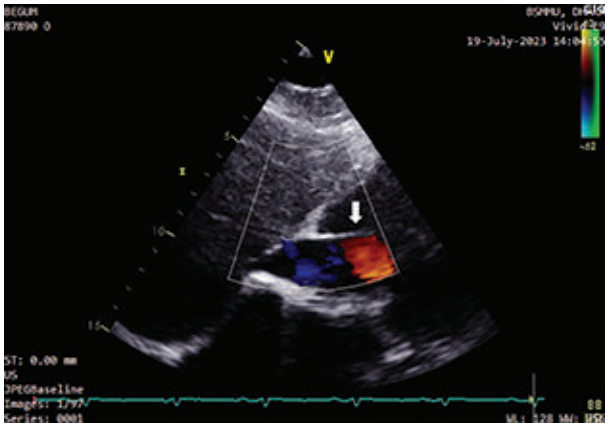

Figure 7: *Subcostal view. Arrow shows Cor triatriatum dexter*

First heart sound is mostly loud in in Tricuspid area, P2 component of second heart is loud in pulmonary area. There is wide and fixed splitting of second heart sound in pulmonary area PAN systolic murmur present GRADE 3/ 6 in left lower sternal area, best heard on breath hold after inspiration. Lung base: fine crepitation bilaterally (admission day). Provisional diagnosis was Atrial Septal Defect, pulmonary Hypertension, Heart failure (NYHA-IV). CBC (17.07.2023) Hb: 12.4 g/dl, ESR: 25 mm/ 1st hour, RBC: 4.32× 10^{12}/L, Platelet Count: 250×10^{9}/L, WBC: 8×10^{9}/L, N : 77 %, L : 25 %, M : 05 %, E : 04 %. RBS-5 mmol/l, HbA1c-5.8%, S creatinine-0.81 mg/dl, Serum electrolyte: Na-140mmol/l, k-4 mmol/l, Cl-104mmpl/l, T-CO2-30.0 mmol/l, Urine R/M/E: normal, S cholesterol: 165 mg/dl, S HDL: 23 mg/dl, S LDL: 125 mg/dl, TG: 84 mg/dl, PT with INR: 12 sec (1.04), TSH: 2.76 micro Lu/ml, Xray chest P/A view shows Cardiomegali with RV type apex and pulmonary conus was full. ECG -right axis deviation with right atrial enlargement. TTE shows dilated Right Atrium and Right ventricle, Normal LV systolic function (LVEF- 58%), Mild RV systolic dysfunction (TAPSE-13 mm), ASD (Secondum), Moderate pulmonary Hypertension, Cor triatriatum Dexter (Incomplete). Later, we confirm the diagnosis by TEE.

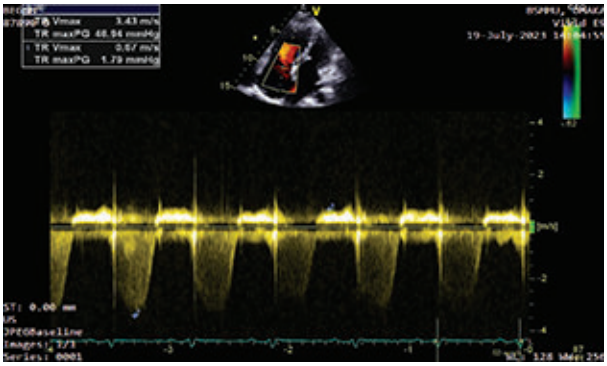

Figure 8: *Moderate Tricuspid regurgitation.*

Discussion:

Cor triatriatum is a rare condition occurring when a child is born with a thin, fibro-muscular membrane subdividing either the left or the right atrium into 3 chambers. The condition is also classified as a congenital heart defect. Cor triatriatum sinister (CTS) is the most common form. The left atrium divides via an atrial appendage into an upper and a lower chamber. The upper chamber receives blood from the pulmonary veins, while the lower chamber is attached to the left atrial appendage blocking the mitral valve orifice creating a significant left ventricular inflow

obstruction. The presence of the left atrial appendage differentiates cor triatriatum from another congenital heart defect, supravalvular mitral stenosis. Another, rarer form of cor triatriatum is cor triatriatum dexter (CTD). With CTD the right valve of the sinus venosus persists dividing right atrium into 2 chambers.[3] Our case is the rare variety of Cor triatriatum dexter which is a incomplete septum divides RA into 2 chambers. I Malciæ et al. found a Cor triatriatum dextrum as an incidental echocardiographic finding in a 5 year old boy and a 9 year old girl, who were evaluated for the presence of a heart murmur. Apart from slightly enlarged right atria, they had otherwise normal hearts and were symptomless. Most of the patients reported until then had been symptomatic due either to the persistent right sinus venosus valve or to commonly associated structural heart anomalies. Therefore, the clinical significance of asymptomatic cor triatriatum dextrum remains coniectural, but might lie in the possibility of development of arrhythmia, progressively worsening interference with the systemic venous return and thrombus formation.[4] Our case is similar like these cases. CTD was in some cases associated with a partial anomalous pulmonary venous return, and ASD of ostium secundum and sinus venosus type[5] in our case, however, the CTD associated with ASD type was ostium Secundum. Failure of the right venous valve regression is considered to be the etiopathogenetic mechanism for the generation of CTD. This results in an anterolateral and posteromedial subdivision of the right atrium. A diagnosis of CTD can be made at any age, often accidentally.[6] Clinical signs of CT depend on the size of the membranes, their exact position and any fenestrations therein. The symptoms are due to the level of obstruction of the vessels opening into the right atrium by the membrane and the presence or absence of ASD.[7] CTD could thus manifest as an obstruction of the right atrial inflow[6] or cyanosis.[7,8,9] Echocardiography complemented by a CT scan is used in diagnosis, but it could be still missed.[5] Cardiac catheterization with angiography is also used as a complementary method,[10] but even a combination of these modalities may not suffice to confirm the diagnosis [11]. Treatment is dependent mostly on the associated defects and is performed mostly through a surgical [12,13] or percutaneous [14,15] disruption of the membrane. Our patient remains asymptomatic for many years as CTD was incomplete. After incidental finding of CTD, we advised the patient of surgical correction of ASD with resection of CTD membrane and our earlier decision of device closure of ASD secundum was cancelled.

Conclusion:

In our patient, the recognition of cor triatriatum played a key role in the multidisciplinary discussion of a transcatheter versus surgical approach regarding clossure treatment of Secundum type of Atrial septal defect.

References:

1. Ajay Kumar Jha 1, Neeti Makhija . Cor Triatriatum: A Review. Seminors in Cardiothoracic and Vascular Anaesthesia. 2017;21(2):11-13.
2. A Dobbertin 1, C A Warnes, J B Seward. Cor triatriatum dexter in an adult diagnosed by transesophageal echocardiography: a case report. J Am Soc Echocardiogr. 1995 Nov-Dec;8(6):952-7.
3. Binish Ather , Amanda Meredith , Waqas J. Siddiqui. Cor Triatriatum: StatPearls 2024;3(2):27.
4. I Malciæ , L Tambiæ, D Richter. Case report: Cor Triatriatum Dextrum, Acta Med Croatica; 1992;46(1):63-5.
5. Goel, A.; Viswamitra, S.; Reddy, B.N.; Gaduputi, J. Computed tomography features of cor triatriatum: An institutional review. Br. J. Radiol. 2021, 94, 20201252.
6. Simsek, Z.; Koza, Y.; Tas, H. Cor triatriatum dexter, atrial septal defects, and pulmonary stenosis—A rare association. Echocardiography 2014, 31, E124–E127.
7. Mazzucco, A.; Bortolotti, U.; Gallucci, V.; Del Torso, S.; Pellegrino, P. Successful repair of symptomatic cor triatriatum dexter in infancy. J. Thorac. Cardiovasc. Surg. 1983, 85, 140–143.
8. Hoye, D.J.; Wilson, E.C.; Fyfe, D.A.; Guzzetta, N.A. Cor triatriatum dexter: A rare cause of neonatal cyanosis. Anesth. Analg. 2010, 110, 716–718.
9. Zainudin, A.R.; Tiong, K.G.; Mokhtar, S.A. Cor triatriatum dexter: A rare cause of childhood cyanosis. Ann. Pediatr. Cardiol. 2012, 5, 92–94.
10. Saxena, P.; Burkhart, H.M.; Schaff, H.V.; Daly, R.; Joyce, L.D.; Dearani, J.A. Surgical repair of cor triatriatum sinister: The Mayo Clinic 50-year experience. Ann. Thorac. Surg. 2014, 97, 1659–1663.
11. Rozema, T.K.; Arruda, J.; Snyder, C.S. Cor Triatriatum: A Tale of Two Membranes. CASE 2019, 3, 25–27.
12. Humpl, T.; Reineker, K.; Manlhiot, C.; Dipchand, A.I.; Coles, J.G.; McCrindle, B.W. Cor triatriatum sinistrum in childhood. A single institution's experience. Can. J. Cardiol. 2010, 26, 371–376.
13. Kardos, M.; Culen, M. Cor Triatriatum and Unusual Pulmonary Venous Drainage. Rev. Esp. Cardiol. (Engl. Ed.) 2018, 71, 391.
14. Savas, V.; Samyn, J.; Schreiber, T.L.; Hauser, A.; O'Neill, W.W. Cor triatriatum dexter: Recognition and percutaneous transluminal correction. Cathet. Cardiovasc. Diagn. 1991, 23, 183–186.
15. Juhas, S.; Gbur, M.; Kirsch, P.; Grendel, T.; Gibarti, C.; Jankajova, M.; Valocik, G. Catheter based therapy of cor triatriatum sinister—Case report. Cor et Vasa 2021, 63, 509–512.