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Case Report

CRISS - CROSS HEART (Report of four cases)

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Criss-cross heart is a rare congenital cardiac anomaly characterized by crossing of the inflow streams of the two ventricles. The anomaly seems to be due to abnormal twisting of the heart about its long axis and when the axis of the openings of the atrioventricular valves are not parallel. Thus in criss – cross heart the abnormality is on the atrioventricular relation, not connection, which happened due to clockwise or counter-clockwise ventricular rotation. We report four cases of criss-cross heart, three cases had levocardia - situs solitus with superior – inferior ventricles and one case had dextrocardia - situs inversus (mirror image) the ventricles were not in superior-inferior fashion. Subcostal and apical four-chamber views can be used to identify the ventricular morphology and position, the atrioventricular valves, the atrioventricular and ventriculo-arterial connection, and the characteristics of the great vessels. The artery connection can be seen more clearly in the parasternal window. All patients had concordance atrioventricular connection, ventricular septal defect and double outlet right ventricle with malposition of the great arteries. Other associated lesions included infundibular (subvalvular) and valvular pulmonary stenosis in three, atrial septal defect in two, and interrupted aortic arch with large persistent ductus arteriosus in one patient. The findings were confirmed by catheterization and angiography in three patients and multislice-CT in one.
Conclusions. patients with criss-cross heart can be easily diagnosed by a careful, systematic segmental study with two-dimensional color-coded transthoracic echocardiography. The failure to obtain a characteristic four - chamber view in any cut and the presence of crossed atrioventricular inflow bloodstreams with each atrium draining into the ventricle located contralaterally were diagnostic for recognition of this complex anomaly. The presence of superior-inferior ventricles although suggestive, should not be regarded as diagnostic for criss cross anatomy.

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Keywords: criss-cross, superior-inferior ventricle, sequential segmental

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Laporan Kasus

CRISS - CROSS HEART (Laporan empat kasus)

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Criss-cross heart merupakan anomali jantung bawaan yang langka, ditandai dengan aliran masuk ke kedua ventrikel yang saling berseberangan. Anomali ini mungkin disebabkan oleh salah putar jantung pada sumbu panjangnya dan ketika sumbu pembukaan katup atrioventrikular tidak parallel. Jadi, pada criss-cross heart kelainannya terletak pada relasi atrioventrikular, bukan koneksinya; akibat rotasi ventrikel searah atau berlawanan arah dengan jarum jam. Kami laporkan empat kasus criss-cross heart, tiga dengan levocardia - situs solitus dengan superior - inferior ventrikel, dan satu kasus lagi dengan dextrocardia - situs inversus (mirror image) - ventrikelnya tidak superior - inferior. Transthorakal echocardiografi pandangan subcostal dan apical four-chamber dapat dipakai untuk mengidentifikasi morfologi dan posisi ventrikel, katup atrioventrikular, koneksi atrioventrikular dan koneksi ventriculo-arterial serta karakteristik pembuluh-pembuluh darah utama. Koneksi arteri dapat lebih jelas kalau dilihat dari pandangn parasternal. Semua pasien ini mempunyai koneksi atrioventrikuler konkordans, defek septum ventrikel dan double outlet right ventricle disertai malposisi pembuluh-pembuluh darah besar. Kelainan penyerta lain adalah stenosis pulmonal infundibular (subvalvular) and valvular pada tiga kasus, defek septum atrium pada dua kasus, dan interupsi arkus aorta dengan duktus arteriosus persisten besar pada satu kasus. Temuan echocardiografi ini dikonfirmasi dengan kateterisasi dan angiografi pada tiga kasus dan multislice-CT pada satu kasus.
Kesimpulan. pasien dengan criss-cross heart dapat didiagnosis dengan mudah menggunakan pemeriksaan two-dimensional color-coded transthoracic echocardiography yang seksama dan sekuensial segmental analisis yang sistematis. Kegagalan memperlihatkan gambar khas pandangan four-chamber pada berbagai potongan, dan adanya aliran darah atrioventrikular yang saling menyeberang, dimana masing-masing atrium mengalirkan darah ke ventrikel yang lokasinya kontra-lateral, merupakan tanda diagnostik anomali yang kompleks ini. Ventrikel yang letaknya superior- inferior meskipun sugestif untuk criss-cross anatomy, tetapi tidak bisa dijadikan tanda diagnostik.

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Kata kunci: criss-cross, ventrikel superior-inferior, sekuensial segmental

Criss-cross heart (CCH) is a complex congenital cardiac malformation characterized by crossing of the inflow streams of the two ventricles. The term criss-cross heart was introduced by Anderson et al. in 1974,[1] but it was Lev and Rowlatt who described this anomaly for the first time in 1961.[2] The anomaly seems to be due to abnormal twisting of the apex of the heart while the base remains relatively fixed, the cause of and mechanism for the twisting remains unclear. This accounts for the crossing

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of the atrioventricular (AV) valve, the abnormal position of the ventricles and ventriculo-arterial (VA) discordance. Thus in CCH the abnormality is on the atrioventricular relation, not connection.[1]

Criss-cross heart is extremely rare, accounting for less than 0.1% of congenital cardiac malformation, so the frequency is not more than 8 per 1.000.000 life birth.[3] It is often associated with abnormalities of connection like transposition of great arteries, and usually accompanies other complex intracardiac anomalies[4] although it can also occur in simple congenital defect such as ventricular septal defect (VSD).[5] A large series of CCH study reported that situs solitus, AV concordance, VA discordance, followed by double outlet right ventricle (DORV), associated with pulmonal stenosis (PS) are the most common pattern.[6-8] Criss-cross heart is often associated with superior-inferior ventricles; this ventricular relationship can present in other cardiac anomalies.[8] In many cases, the size of the ventricles (small on many occasions) is conditioned by the size of the annulus of the atrioventricular valves.[9-10]

The physiology is determined by the discordant or concordant AV and VA connection and the associated cardiac defects, such as displaced and/ or straddling left AV valve, VSD (which always presents), valvular or subvalvular PS, small RV, and juxtaposition of the atrial appendages.[9,12-14] There is no specific clinical presentation for criss-cross heart, the patient presentation is depends predominantly on the hemodynamic consequences of the associated lesions. Accurate diagnosis of the condition is important as early palliative surgery has a favorable impact on functional status in adulthood.[15] Transthoracic echocardiography is an invaluable tool for screening and evaluation of CCH.

The aim of this presentation is to explore the value of echocardiography in the diagnosis of criss-cross heart.

Case Variations

Within twelve years we found four cases of congenital heart disease diagnosed as criss- cross heart associated with variable heart defects, three were female. The diagnosis was made by echocardiography and the findings were confirmed by catheterization and angiography in three cases and multislice-CT in one. Details of patients describe in Table 1.

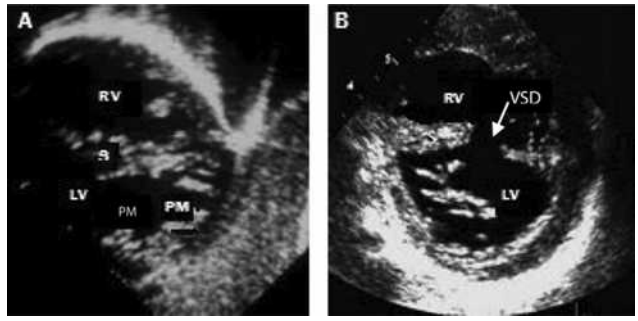

Figure 1. Two dimensional echocardiography in cases with situs solitus: subcostal short axis view (**A**) and parasternal short axis view (**B**) at mid - ventricular level showed superior RV - inferior LV (recognized by the presence of two papillary muscles) and horizontal alignment of interventricular septum.
RV - right ventricle, LV - left ventricle, PM – papillary muscle, S – interventricular septum, VSD – ventricular septal defect.

Table 1. Criss-cross heart case details.

Case no.	Age	Cardiac & apex position	Atrial situs	AV connection	VA connection	Associate Lesions	SIV
1.	9 years	levocardia	solitus	concordance	DORV	muscular inlet VSD, secundum ASD, valvular and sub valvular PS, malposition GA	+
2.	1 months	levocardia	solitus	concordance	DORV	muscular inlet VSD, secundum ASD, malposition GA, valvular and sub valvular AS, IAA, PDA, PH, severe TR	+
3.	3 years	levocardia	solitus	concordance	DORV	muscular inlet VSD, valvular and sub valvular PS, malposition GA	+
4.	2 years	dextrocardia	inversus	concordance	DORV	muscular inlet VSD, valvular and sub valvular PS, malposition GA	-

VSD – ventricular septal defect, ASD – atrial septal defect, DORV - double outlet right ventricle, PS – pulmonal stenosis, AS – aortic stenosis, GA – great arteries, IAA – interrupted aortic arch, PDA – patent ductus arteriosus, PH – pulmonary hypertension, TR – tricuspid regurgitation.

As in other series reported, most of our cases had situs solitus, AV concordance, muscular inlet VSD, DORV and malposition of the great arteries (aorta anterior to the pulmonary artery). Three with valvular and sub-valvular PS protected from pulmonary vascular obstructive disease. One case had valvular and sub-valvular aortic stenosis (AS) with type B interrupted aortic arch (IAA), the descending aorta supplied by patent ductus arteriosus (PDA), the pulmonary trunk was dilated and hypertensive. All cases had superior – inferior ventricle, except in one who had mirror image with dilated RV located right slightly superior to the smallish LV.

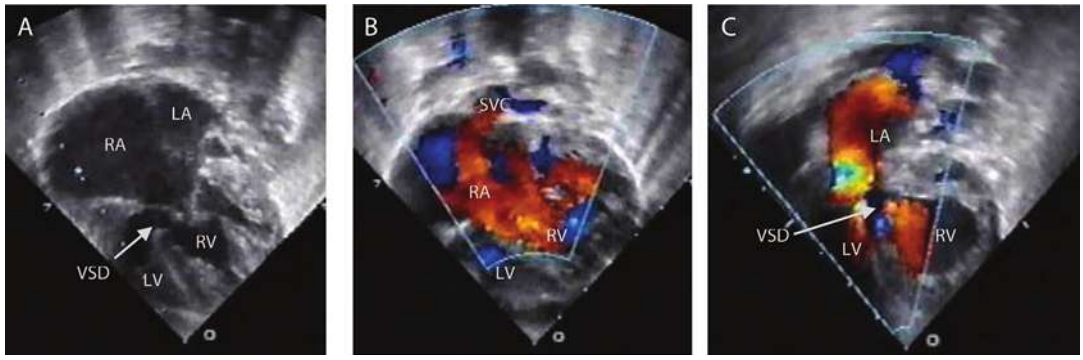

Figure 2. Subcostal 4 - chamber view in cases with situs solitus showed atrioventricular concordance connection with criss-cross atrioventricular relation: **A**. RA on the right side is connected to the rough trabecular RV on the left side, by color Doppler echocardiography the bloodstreams could be visualized : **B**. from superior vena cava (SVC) drained into RA - filled RV on the left side, **C**. LA on the left side filled LV on right side. *RA – right atrium, LA – left atrium, SVC – superior vena cava, RV – right ventricle, LV – left ventricle, VSD – ventricular septal defect.*

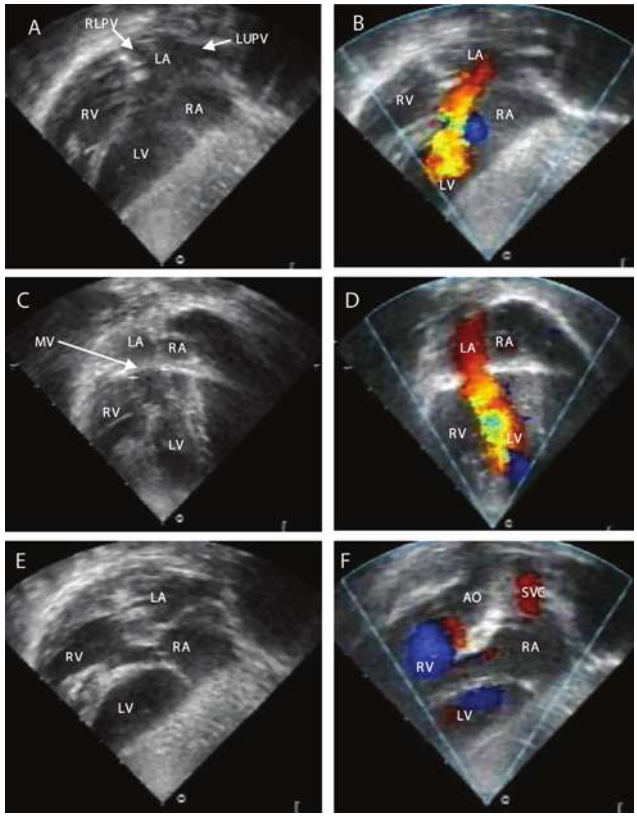

Figure 3. Subcostal 4 -chamber view in patient with dextocardia situs inversus, AV concordance connection with criss-cross AV relation : **A**. the pulmonary veins LVdrained into the LA on the right side which connected to the smooth trabecular LV on the left side, **B**. color Doppler clearly showed the direction of blood streams from LA on the right side to LV contralaterally through small mitral valve. It can also be visualized through apical 4 - chamber view (**C, D**).

By rotating and tilting the transducer on subcostal 4- chamber views we can visualized : **E**. RA on the left side connected to RV on the right side, **F**. color Doppler showed the blood streams from the SVC to RA, and then filled the RV.

E RLPV – right lower pulmonary vein, LUPV – left upper pulmonary vein, LA – left atrium, RA – right atrium, LV – left ventricle, RV – right ventricle, MV- mitral valve

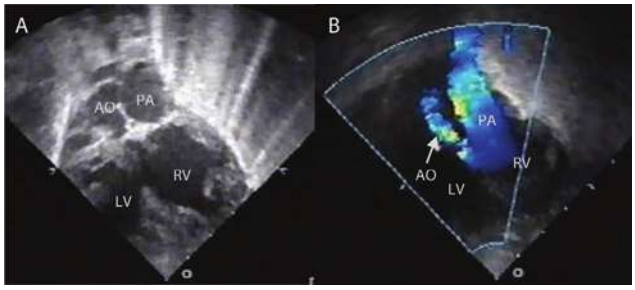

Figure 4. Subcostal 4-chamber views in case no. 2 : **A**. double outlet right ventricle, the aortic valve is small with sub-valvular and valvular aortic stenosis, PA hypoplatic ascending aorta positioned on the right side of pulmonary trunk. **B**. with color Doppler the flow could be visualized.
LA – left atrium, LV– left ventricle, RV – right ventricle, PA – pulmonary artery, A0- aorta.

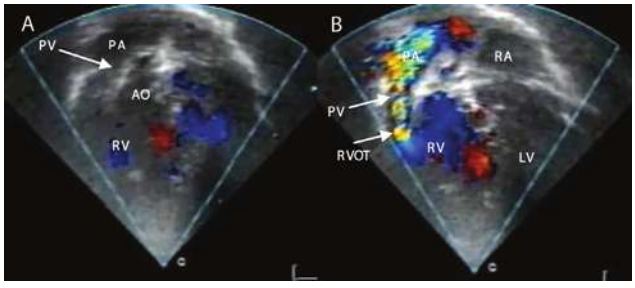

Figure 5. Apical 4-chamber view in case no. 4 : A. Double outlet right ventricle, the RV filled pulmonary artery through a narrow RVOT and PV annulus, B. turbulence was seen in pulmonary trunk - valvular and sub-valvular pulmonary stenosis.
LA – left atrium, RA – right atrium, LV – left ventricle, RV – right ventricle, PA – pulmonary artery, PV – pulmonary valve, RVOT – right ventricle outflow tract.

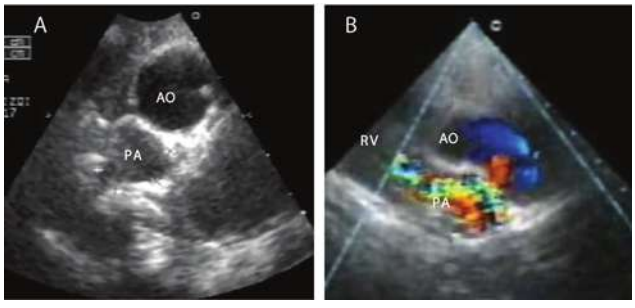

Figure 6. Parasternal short axis view: **A**. aorta left anterior to the pulmonary artery (malposition of the great arteries), **B**. Para- sternal long axis view : aorta anterior to pulmonary artery, turbulence in the pulmonary trunk due to pulmonary stenosis.
RV – right ventricle, PA – pulmonary artery, AO – aorta.

Echocardiography

Two-dimensional (2D) echocardiographic examinations of atrial situs were performed at abdominal short and long axis views as standard, three cases had situs solitus and one situs inversus. At mid-ventricular subcostal short-axis view : all cases with situs solitus had superior– inferior ventricles (SIV). The presence of SIV was defined as: (1) horizontal alignment of interventricular septum and (2) right ventricle stacking over left ventricle (figs. 1 A). SIV could also be assessed through parasternal short axis view at mid-ventricular level (figs. 1B).

Evaluation of the left ventricle, right ventricle, AV and VA connections and relations were determined by segmental analysis at subcostal and apical four-chamber views with 2D color Doppler echocardiography. Documentation of crossed ventricular inflow streams typically requires rotation and tilting of transducer because both inflows invariably cannot be visualized clearly in same imaging plane. Crossed ventricular inflow streams was defined as : (1) failure to visualized both AV valves and four cardiac chambers at one imaging plane and (2) the presence of crossed AV inflow bloodstreams with each atrium draining into the ventricle located contra- laterally (Figs. 2A, 2B, 2C in situs solitus and 3A, 3B, 3C, 3D, 3E, 3Fin situs inversus).

All of our patients had VSD with abnormal VA connections - double outlet right ventricle (DORV). Evaluation of the VA connections and relations were determined by subcostal and apical 4-chamber (figs. 4A, 4B, 5A, 5B). Case no. 2 had significant valvular and sub-valvular aortic stenosis, the others had valvular and sub valvular pulmonary stenosis (PS).

The great arteries connection and spatial relation can be seen more clearly in the parasternal window (figs. 6A, 6B). All patients had double outlet right ventricle with malposition of the great arteries.

Two patients (case no. 3 and no. 4) received palliative procedures bidirectional cavopulmonary shunt (BCPS) to relieve cyanosis due to the presence of DORV with valvular and sub-valvular PS; Fontan-type operation was plan for both cases because the left ventricle was small. Case no 1 refused surgery, case no. 2 died before any intervention was done.

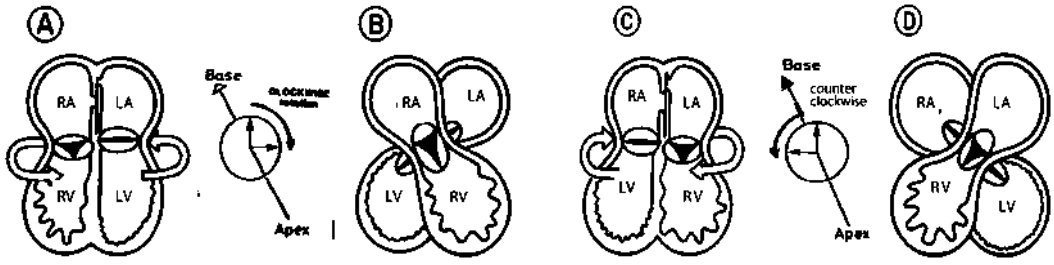

Figure 7. Looking from apex toward the base of the heart, this diagram illustrating the etiologic mechanism (ventricular twist) that causes criss-cross heart. **A**: in atrioventricular concordance - ventricular twist is clockwise. **B**: atrioventricular concordance connection with discordant spatial relationship; right-sided morphologic right atrium (RA) connected to a left-sided morphologic right ventricle (RV) and left-sided morphologic left atrium (LA) connected to a right-sided morphologic left ventricle (LV). **C**: in atrioventricular discordance - ventricular twist is counter clockwise. **D**: atrioventricular discordance with concordant spatial relationship: right-sided morphologic RA connected to a left-sided morphologic LV and left-sided morphologic LA connected to a right-sided morphologic RV.

Discussion

Criss-cross heart can be found in hearts with any biventricular atrioventricular connection, as well as with any ventriculo-arterial connection and be anticipated with any type of atrial situs[4]; three of our cases had situs solitus and one situs inversus. Criss-cross heart illustrates the important concepts that atrioventricular connection is not always predictive of atrioventricular alignments, that both are important, and both must be elucidated and described independently.

The direction of rotation required to produce criss-cross heart is different according to the atrioventricular connection. Looking toward the base of the heart, the rotation required to produce criss-cross hearts is clockwise rotation with a concordant atrioventricular connection and counter-clockwise with a discordant atrioventricular connection (figs. 7).[4]

The previously labeled equivalence of crisscross heart to superior inferior ventricle[16] is not correct. Some CCH cases exhibited vertical interventricular septal alignment,[7,13,17,18] as seen in case no.4, although horizontal alignment remains the most prevalent. Criss-cross heart comprised only 41–63% of reported series of superior inferior ventricle.[19,20]

Detection of the associated defects: apex position, situs of the atrium, atrioventricular and ventriculo-arterial relationships, valves and aortic arch were determined by segmental analysis as standard.[18,21]

Conclusions

Patients with criss-cross heart can be easily diagnosed by a careful, systematic segmental study with two-dimensional color-coded transthoracic echocardiography. The failure to obtain a characteristic four chamber view in any cut and the presence of crossed atrioventricular inflow bloodstreams with each atrium draining into the ventricle located contralaterally were diagnostic for recognition of this complex anomaly. The presence of superior-inferior ventricles although suggestive, should not be regarded as diagnostic for criss cross anatomy.

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