

ROLE OF 128-MDCT SCAN IN DIAGNOSING CONGENITAL HEART DISORDERS USING 3 SEGMENT NOTATION IN SITUS INVERSUS, LEVO-LOOP, LEVO-MALPOSITION OF THE BLOOD VESSELS ACCOMPANIED WITH MORPHOLOGICAL DOUBLE OUTLET RIGHT VENTRICLE CASE

Ristaniah D. Soetikno^{*}, Vini Nilasari, Harry Galuh Nugraha.

Abstract: - *Situs inversus is an anatomical configuration in which the organ arrangement is upside down compared solitus sites. Situs inversus is a rare congenital disorder. CT scan is a modality of choice for a definite diagnosis of situs inversus. The diagnosis of situs inversus is carried out using segmental approach with 3 segments notation of Van Praagh, which includes determining the visceratrial sites, directions ventricular-loop and position toward large vessels. A case was reported that a male, age 19 years, with a chief complaint of shortness of breath, has been diagnosed with situs inversus and examined with MDCT. The test results showed the diagnosis of situs inversus, levo-loop, levo-malposition of great vessels (I, L, L-MGV) with atrioventricular concordance and morphologic double outlet on the right ventricle. Additionally, the image also showed subaortic ventricular septal defect and sub valvular pulmonic stenosis with dilatation of the stenotic post on the main pulmonary artery and the main branch of the pulmonary artery bilateral proximal portion. Several other abnormalities were also found, including pericardial effusion, dilation of the right hepatic vein, the media and the left hepatic vein as well as obstruction to the flow of the contrast in the superior vena cava and inferior and azygos veins that with dilatation of the morphologic right atrium and morphologic right ventricle which is likely due to heart failure, as well as anomalies of descendens aorta. Situs inversus imaging using 128-MDCT scans are important to be performed for intra-operative details such as visceral organs position, the apex of the heart, and the branches of the large blood vessels and error in identification of disordered structure can be avoided.*

Keywords: *Situs Inversus, 3 Segment notation, MDCT*

Introduction:

Situs inversus is an anatomical configuration in which the organ arrangement is upside down solitus sites; while the *situs solitus* is a normal anatomical configuration of the right atrium and the liver is located on the right side; left atrium, stomach, and spleen are located on the left side; right lung consisting of 3 main bronchus lobe has branches with branching at the proximal and the left lung has two lobes which have a main bronchi branch located more distal.^{1,2}

Situs inversus is a rare congenital disorder. *Situs inversus* is usually diagnosed using conventional radiography and ultrasonography. However, the CT scan is the main option for a definite diagnosis of *situs inversus*.³

The diagnosis of congenital heart defects, including *situs inversus*, is carried out using segmental approach firstly introduced by Van Praagh and has been widely used since then. Segmental approach with 3 segments notation is fairly easy to understand and very useful for the clinical importance.^{1,2}

This case report will discuss the role of MDCT in determining the diagnosis of *situs inversus* using segmental approach with three segments of notation system, which includes determining the viscera-atrial site, ventricular-loop direction, and the position of the great vessels.

Case Report:

A 19-year-old man with a chief complaint of continuous shortness of breath since 1 week before admission. Shortness of breath is not increased or decreased by a certain position. Complaints such as palpitations and fainting are denied. Patients also complain of tiredness in the last year and a long history of shortness of breath after light routine

activity since a year ago. Patient also sleeps with elevated head, and chief complaint accompanied by intermittent swelling in the legs,

Patient denied high blood pressure, history of diabetes melitus, high cholesterol, or heart disease in the family. Based on allo anamnesis, patient is known to have heart position upside down since the age of 6 months with a cyanotic history since childhood, especially after playing and feeding difficulty during infancy.

Plain chest X-Ray of the patient show cardiomegaly with dextrocardia, first echocardiogram show sinus rhythm, first degree AV block, enlargement of both atria and right ventricular enlargement. Echocardiography show dextrocardia and *Tetralogy of Fallot* (TOF), second echocardiography show *moderate circumferential pericardial effusion* without the sign of tamponade.

Patient undergoing thoracic angiography CT scan using 128-MDCT scanner (SOMATOM Definition Flash, Siemens Healthcare), with detector collimation, 128 x 0.6 mm; pitch, 1.2; slice thickness, 5.0 mm; and gantry rotation time, 0.5 seconds. This examination is performed with 120 kV, mAs value is 352 mAs, with a duration of 4.32 seconds examination. Contrast used is a nonionic contrast iodine levels 300 MGI/mlL. (Iopamirol® 370, Bayer Healthcare). The dose given was 70 ml with a flow rate of 3.5 ml/s. The examination was conducted on the level of thoracic inlet to the first lumbar vertebral bodies

Test result show *situs inversus, levo-loop, levo-malposition of great vessels* (I, L, L-MGV) dengan *atrioventricular concordance* dan *double outlet morphologic right ventricle*. (Fig.1 and Fig.2)

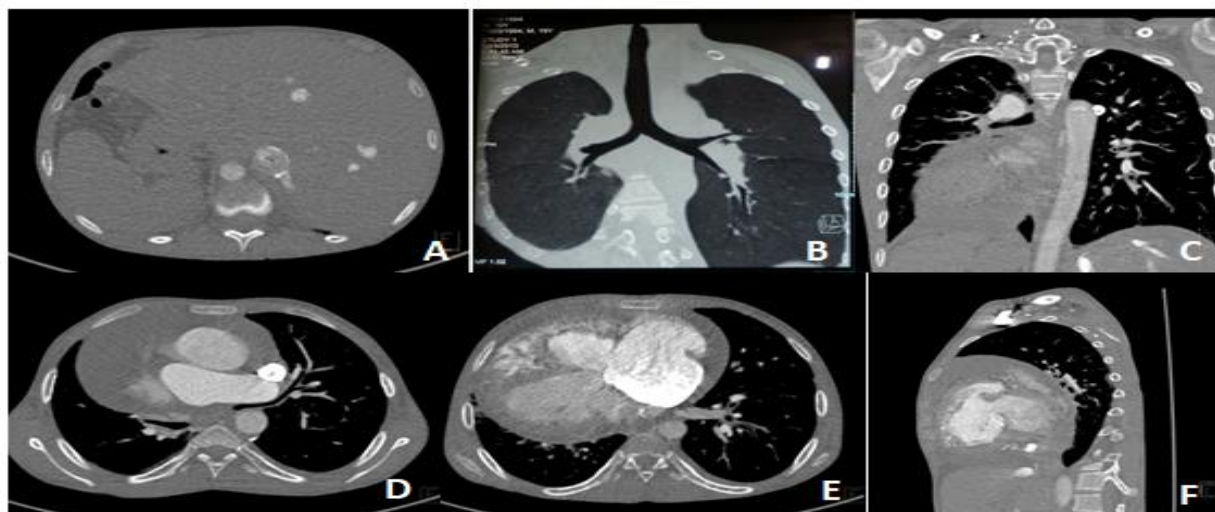


Figure 1: Determination of the site visceratrial (A) The biggest liver lobes are located on the left side and spleen is located on the right side (B) Superior lobe bronchus of morphologic right lung come from the more proximal than superior lobe bronchus of morphologic left lung (C) main bronchus of morphologic left lung positioned below the pulmonary artery (hiparterial) (D) main bronchus of morphologic right lung has a position behind the pulmonary artery (eparterial) (E) the board and blunt shape of trapezoidal appendix morphologic right atrium (F) 'fingerlike' formation of appendix morphologic left atrium are narrower and tubular.

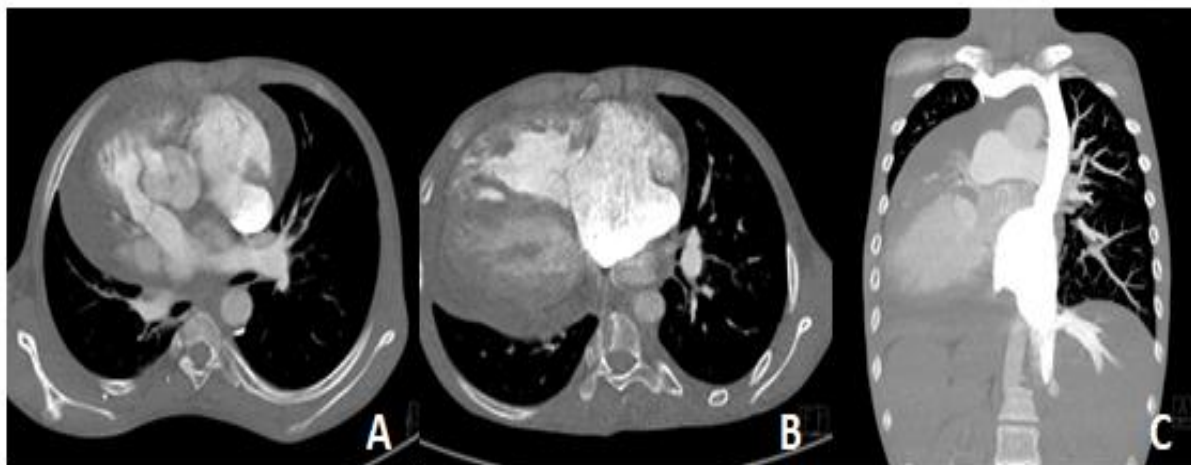


Figure 2: (A) The position of the aorta that is located on the left side of the main pulmonary artery showing the condition of levo-malposition of the great arteries (B) Atrioventricular concordance, it appears, the image of the band and trabecular moderator rough as a marker of the morphologic right ventricle (C) Venoatrial concordance

Additionally, the image of subaortic ventricular septal defect and Subvalvular pulmonic stenosis with dilatation of the stenotic post on the bilateral

main pulmonary artery and the proximal portion of main branch of the pulmonary artery (Fig. 3)

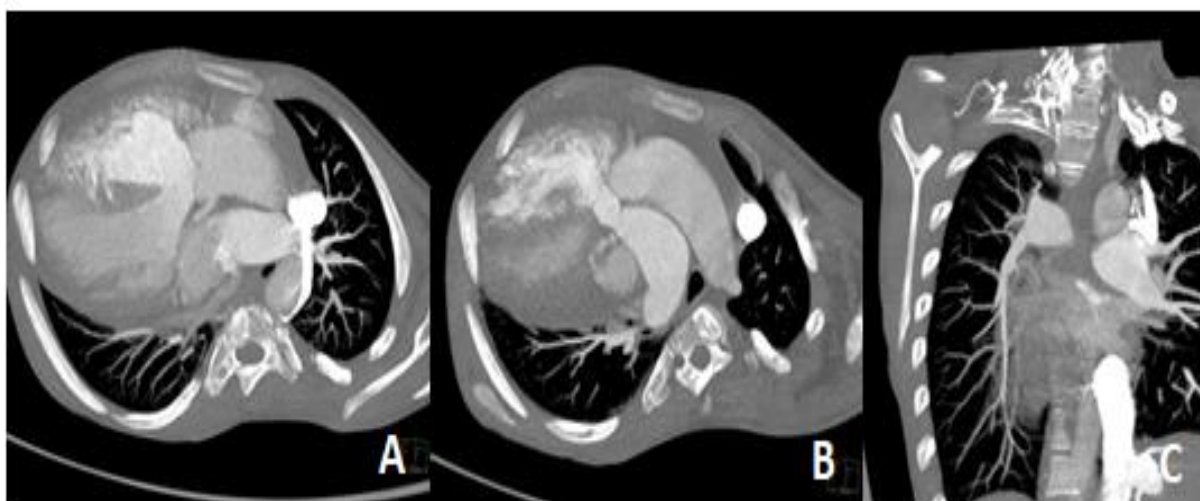


Figure 3: (A) Ventricular Septal Defect (VSD) and double outlet right morphologic ventricle, morphologic right ventricle that flow blood to ascending aorta and coronary artery came out from the aorta (B) Subvalvular pulmonic stenosis with dilatation of the stenotic post on the main pulmonary artery (C) post-stenotic dilatation of the main branch of bilateral proximal portion of pulmonary artery

Several other abnormalities were also found such as pericardial effusion, dilation of right hepatic vein, the media and the left as well as the flow resistance of the contrast in the superior and inferior vena cava

and the azygos veins with dilatation of morphologic right atrium and morphologic right ventricle are likely due to heart failure. (Fig. 4 and Fig. 5)

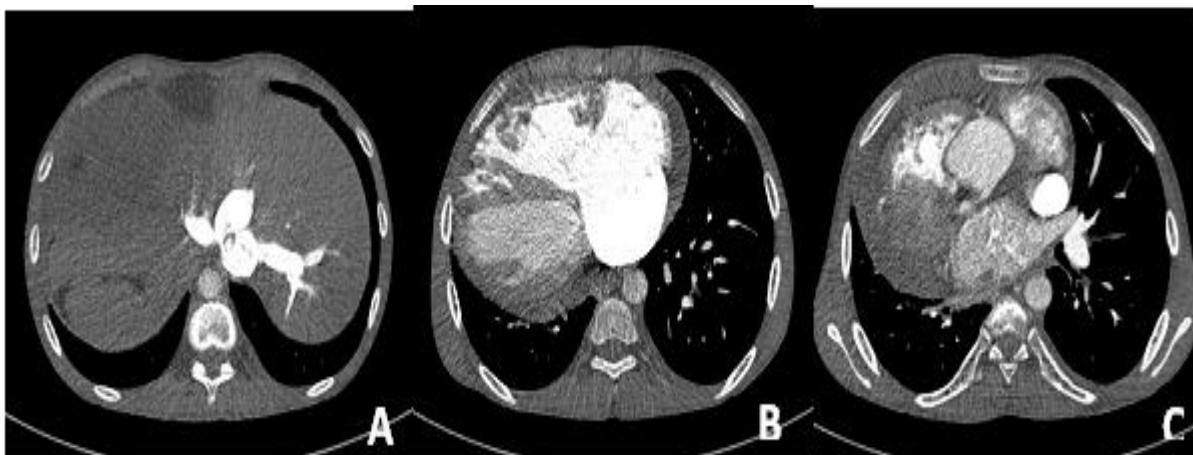


Figure 4: (A) Widening of right hepatic vein, the media and the left as well as the flow resistance of the contrast in the inferior vena cava, (B) dilation of morphologic right atrium and morphologic right ventricle, and (C) pericardial effusion which showed signs of heart failure

Another noticeable abnormality is an anomaly in which the ascending aorta outing located on the right anterior side and continues to be aortic arch turning from right anterior to the left posterior side. Thus, the descending aorta is in left paravertebral.

Aorta descenders then crossed the center line at the height of the diaphragm so that the abdominal aorta was on the right paravertebral. (Fig. 5)



Figure 5: Anomalies of aorta outing, azygos vein dilation appearance also located on the left side of the aorta

Discussion:

Situs inversus is a rare congenital disorder, prevalence of only 1:10,000 live births. Situs inversus is usually associated with dextrocardia events with an incidence of only 3-5% of all congenital heart defects and about 20% is part of Kartagener syndrome.^{3,4,5}

Situs inversus is usually diagnosed using conventional radiography and ultrasonography. However, the CT scan is the main option for a definite diagnosis of situs inversus. CT scan will provide good imaging in determining the position of

visceral organs, the apex of the heart, and the branches of the large blood vessels. In addition, the CT scan is also faster and easier to do.^{3,6}

Single-source MDCT rapid rotation time make better temporal resolution than CT single slice, although the radiation exposure becomes greater. Better temporal resolution and the ability of MDCT to generate image data free of cardiac motion artifacts broad range of heart rate, an advantage of MDCT in the evaluation of cardiac morphology. The image data are free from motion artifacts that

can show a clearer image data of ventricular cavity and endocardial (trabeculation and moderator band). Hence, the characteristics of each ventricle will be clearer.⁶

Imaging of situs inversus is very important to the interests of the operative where fault identification disordered structure can cause fatal errors.³

Situs inversus is an anatomical configuration in which the organ arrangement upside down compared with solitus sites; the site solitus a normal anatomical configuration of the right atrium and the liver is located on the right side; left atrium, stomach, and spleen is located on the left side; right lung which consists of 3 lobe with main bronchus and branches at proximal and the left lung has two lobes with a main bronchi branch located more distal

There are 3 types of sites, sites solitus, situs inversus, and the ambiguous sites. Site type is determined by the relationship between the atrium and surrounding organs and the types of sites is only valid for one person.^{1,2}

In determining a congenital heart defects, several approaches can be done. One system is already widely known is a system by Van Praagh with a notation system with 3 letters separated by commas and written in parentheses. Steps to assess the anatomy of the heart is based on the current segmentation of cardiac embryology formed namely 1) to determine viscera-atrial sites, 2)

determine the ventricular-loop, and 3) determine the position and the relationship between the great vessels.^{1,2}

The first step, determining the site visceratrial {X, _ _}

The first step is to determine the position of the liver, stomach and spleen which are on the right or left side of the patient.^{1,2}

Then determining the thoraco-abdominal site by determining the position of the morphologic right main bronchus and lung in epiretinal position and morphologic left main bronchus and lung position hiparterial position. Morphologic right superior lobe bronchus and lung are in the more proximal regions of morphologic left superior lobe bronchus and lung.^{1,2}

The amount of the pulmonary lobes also help in determining the side. Right lung has three lobes and the left lung has two lobes, although the state of the right and left isomerism, pulmonary circumstances cannot be a determinant of the side of the right atrium.^{1,2}

In identifying the right atrium, it is important to consider (Table 1), starting with the use of the intrinsic characteristics of the atrium. Broad and blunt right atrium appendix is often called a trapezoid shape. Left atrium appendix is usually narrower than the right and more tubular in shape, with a configuration resembling a finger (finger-like).^{1,2}

Table 1: Characteristics of Cardiac Atrium²

Right atrium morphology	Appendix of right atrium is trapezoid with a broad base Pectinatus muscle extend from appendix to the atrioventricular canal Septum surface has superior and inferior limbic band Myocardial components includes terminal crest and sagittal tenia Usually receive blood flow from the coronary sinus and superior and inferior vena cava
Left atrium morphology	Appendix of left atrium is tubular with narrow ties Pectinatus muscle inside a tubular appendix

The presence of other signs, such as an external anatomical structure that is also known to be an indicator of the position of the atrium. Veno-atrial concordance condition is a normal condition, the room where the inferior vena cava and coronary sinus leads are usually a morphologic right atrium.^{1,2}

After comparing the position of the right atrium with

the visceral organs and determines the notation, we can write 'S' for solitus sites, showing normal position called visceratrial sites {S, _ _}. If the orientation is reversed, write 'I' for situs inversus, {I, _ _}. If the position visceratrial does not fit any of the categories above, it should be written by 'A' to the site of ambiguous, {A, _ _}, also known as heterotaxy which means 'different position', which formed the right side of bilateral or left side of

bilateral.^{1,2}

The second step is determining the ventricular-loop {_, X, _}.

In embryological development, the primitive heart is tube-shaped. Under the direction of craniocaudal, the components of the tube is truncus arteriosus (the future large blood vessels), bulbus cordis (going to the right ventricle), embriogenic left ventricle, primitive atrium and sinus venosus.^{1,2}

In 23-28th day (week IV) of gestation, the heart tube lenght increase and fold forward and backward. In normal condition, the tube curved to the front and right side, placing future right ventricle to right side of left ventricle, known as D-loop. If the tube curve to the elft, right ventricle with be on the left side of left ventricle, known as L-loop. The fold of this tube will determine the heart morphology and position of each heart tube component (Fig 6).²

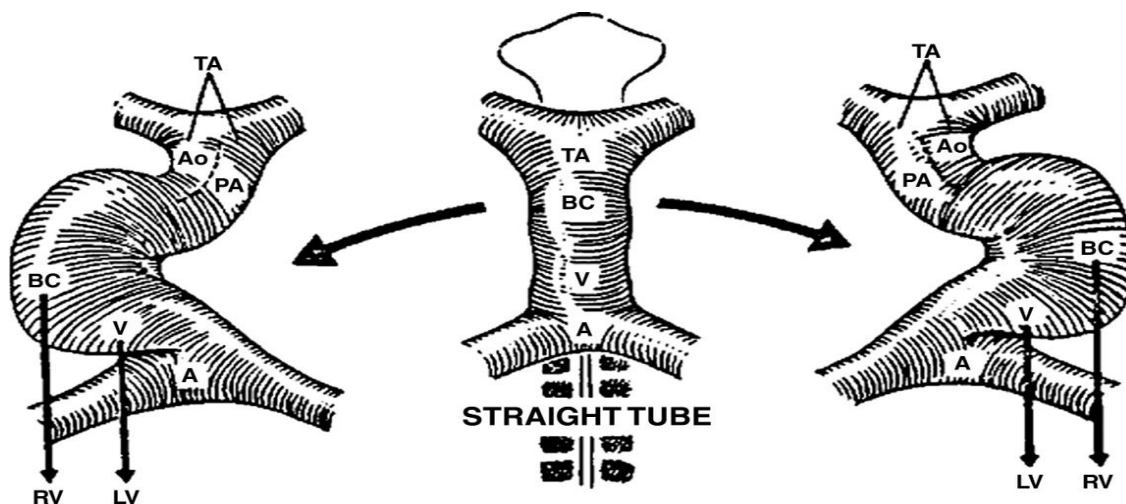


Figure 6: Diagram showing D-loop formation (left) dan L-loop (right) from heart embriological tube (centre) A=atrium, BC=bulbus cordis, LV=left ventricle, PA=pulmonary artery, RV=right ventricle, TA=truncus arteriosus, V=embryonic left ventricle¹

First we have to identify the morphologic right ventricle and morphologic left ventricle based on its intrinsic character (Table. 2) in order to determine the orientation of the ventricular-loop. When the morphologic right ventricle located on the right side morphologic left ventricle, then write 'D' to indicate

the dexro-loop. If the morphologic right ventricle located on the left side of the left ventricle showed Levo-loop, then write 'L'. In complex cases, for example supero-inferior ventricle, AV alignment crisscross, and single ventricle, then the orientation of the ventricular-loop will be difficult to evaluate.^{1,2}

Table 2: Characteristics of Cardiac Ventricle

Morphologic right ventricle	<ul style="list-style-type: none"> • Moderator band • Rough irreguler trabecula • Papillaris muscle that connects the interventricular septum to the free wall section • AV tricuspoid valve • Septal and parietal band
Morphologic left ventricle	<ul style="list-style-type: none"> • Flat septum • Smooth trabecula • Papillaris muscle is only attached to the free wall • AV bikuspoid valve • Superior part of septum has smooth surface

The third step is determining the position and the relationship between the great vessels {_, _ X}

Aorta dan MPA (Main Pumonary Artery) are classified based on their position and their

relationship to the level of aortic valve and pulmonary valve.^{1,2}

Aorta will supply at least one coronary artery and systemic arteries, while MPA are usually supplying

the right and left pulmonary artery. Normal configuration of large blood vessels, the aorta is located more posteriorly and to the right of the MPA. This configuration is written with 'S' to indicate the site solitus or normal configuration of large blood vessels {_, _, S}. When the aorta lies in posterior of MPA but on the left side, it is written 'I' to show situs inversus {_, _, I}.

When large blood vessels have abnormal position, such as transposition (TGV) or malposition (MGV), such as the case when both arteries originate from the same ventricle or overriding, for example DOLV or DORV.^{1,2}

In the aortic position of the MPA usually, although

not always, indicates transposition. If the aorta is located in the anterior and right of the MPA, this anomaly is called dextro-transposition or D-transposition of the great vessels, {_, _, D-TGV}. If the aorta is located in the anterior and left of the MPA called levo-transposition or L-transposition or congenitally corrected transposition, written {_, _, L-TGV}.^{1,2}

If the aorta is not located in the anterior or posterior of MPA often called malposition. If the aorta is located right next to the MPA, this anomaly is called D-malposition {_, _, D-MGV}. If the aorta is located on the left of the MPA is called L-malposition, written {_, _, L-MGV}.^{1,2}

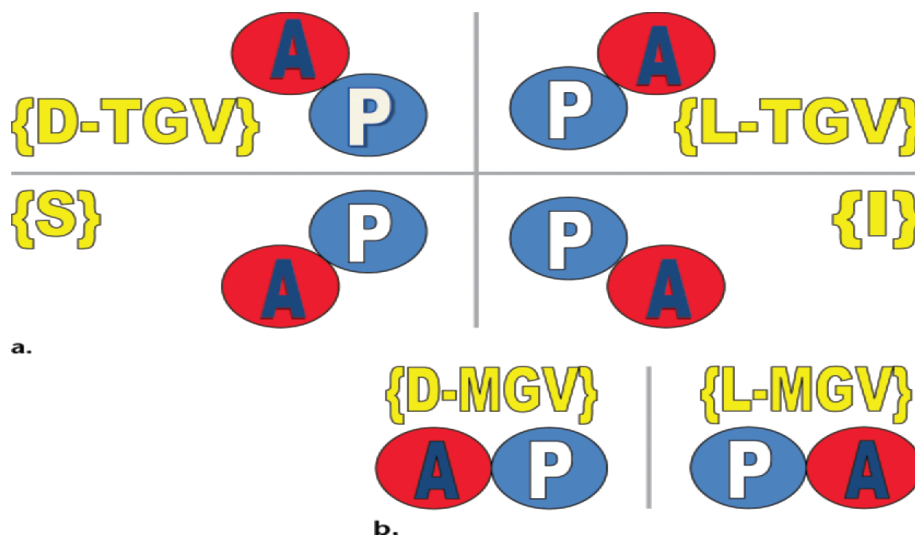
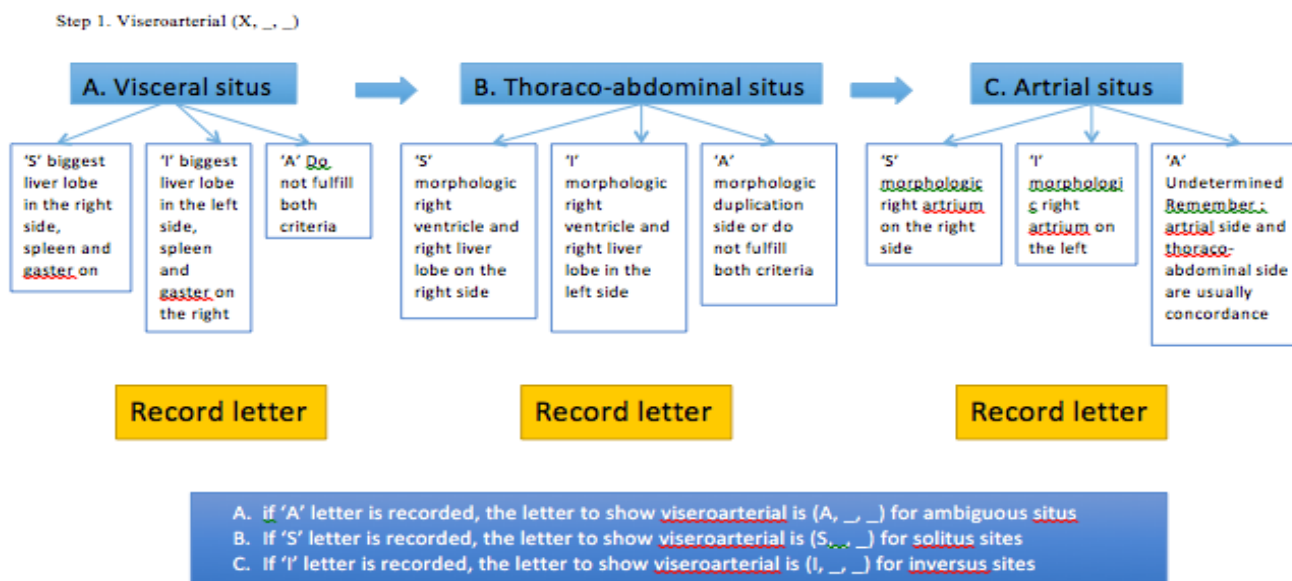
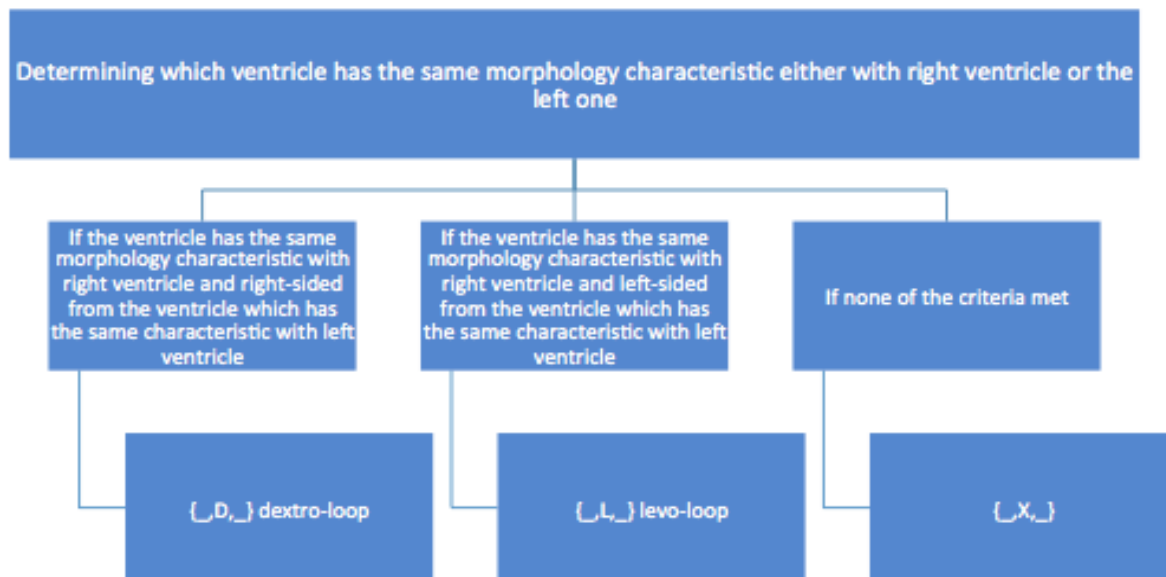


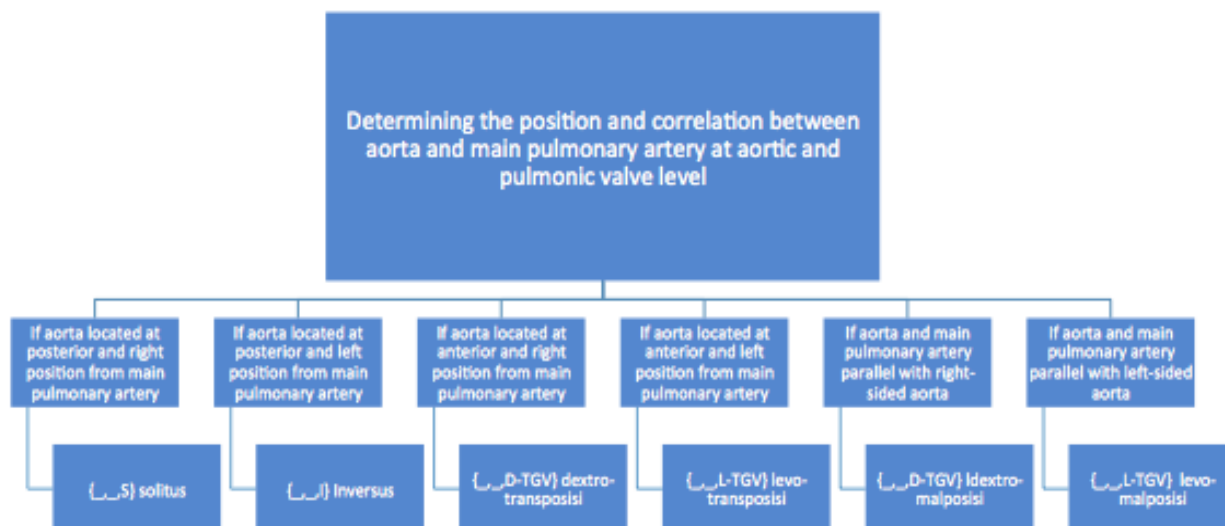
Figure 7: (A) Schematic image showing the configuration of large blood vessels on the axial cuts, normal position, inversion, transposition (B) image shows a schematic D-malposition or L-malposition of the great vessels.²



Step 2: Pointing The Ventricular Loop {D, L, X}



Step 3: Position and Correlation of Great Arteries {S, I, D-TGV, L-TGV, D-TGV, L-TGV}



Conclusion:

Van Praagh segmental classification system can be applied universally to interpret and report the findings in cardiac imaging with CT, MRI and ultrasound. In addition, the 3 segments notation can be understood by all clinical from various

disciplines.

A case with a diagnosis of situs inversus, levo-loop, levo-malposition of great vessels (I, L, L-MGV) with atrioventricular concordance and double outlet

morphologic right ventricle, which is confirmed by examination of the 128-MDCT scan and uses a system of 3 segments notation Van Praagh. Examination using a 128-MDCT scan, generate image data to better evaluate the morphology of the heart, to show clearly the picture of sub aortic ventricular septal defect and subvalvular pulmonic stenosis with dilatation post-stenotic on the main pulmonary artery and the main branch of the pulmonary artery bilateral proximal portion, and other sign due to heart failure, as well as anomalous descending aorta.

By using a 3 segment notation system, significant findings can be made with quick and neat format. In

addition, a thorough understanding of the fundamental concepts of segmental classification helps identify and determine abnormal AV and ventriculo-arterial relations and other related anomalies. Descriptions of all findings must be put into 3 segments notation on radiology report.

By using 3 notation system of this segment, significant findings can be made with quick and neat format. In addition, a thorough understanding of the fundamental concepts of segmental classification will help in identifying and determining abnormal AV and ventrikuloarterial relations and other related anomalies. Descriptions of all findings must be put into 3 segments notation on expertise radiology.

References:

- [1] Lapierre C, Dery J, Guerin R, Viremouneix L, Dubois J, Garel L. Segmental Approach to Imaging of Congenital Heart Disease. *Radiographics*. 2010; 30:397-411.
- [2] Schallert EK, Danton GH, Kardon R, Young DA. Describing Congenital Heart Disease by Using Three-Part Segmental Notation. *Radiographics*. RSNA. 2013; 33:E33-E46.
- [3] Wilhelm A, MD. Situs Inversus Imaging. Tersedia di <http://emedicine.medscape.com/article/4136>
- [4] Weerakkody Y, Gaillard F, et al. Situs inversus. Tersedia di http://radiopaedia.org/articles/situs_inversus. Diunduh tanggal 3 Januari 2014.
- [5] Ghosh, S. Yarmish, G. Godelman A. Haramati, LB. Spindola-Franco, H. Anomalies of Visceroatrial Situs. *AJR* 2009; 193:1107–1117.
- [6] Seidensticker PR, Hofmann LK. *Dual source CT Imaging*. Springer. 2008:91,97