Comprehensive evaluation of complex congenital heart disease using the Van Praagh notation: step by step in MDCT

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Learning objectives

The educational objectives of this article are to convey major concepts of the Van Praagh notation system and illustrate the commonly encountered cardiac anomaly described in each step of analysis. By using this systemic approach, even beginners can evaluate complex congenital heart disease precisely and have no difficulties in communicating with other medical colleague.

Background

The Van Praagh segmental approach to the complex congenital heart disease was firstly developed in the 1960s and has been widely used "language" describing complex anatomy of congenital heart disease over the decades [1-3]. The Van Praagh notation demonstrates a group of three letters, with each letter representative for a key embryologic region of cardiac anatomy: the atria, ventricles and great vessels. We retrospectively review image findings of multi-detector CT scan of complex congenital heart diseases diagnosed in our hospital from 2005 to 2013. We comprehensively illustrate the typical image features according to Van Praagh notation system, including cases for demonstrating the visceroatrial situs orientation, the ventricular loop orientation, and the relation and anomaly of great vessels origin.

Findings and procedure details

There are three major steps in the Van Praagh notation: (1) determine the visceroatrial situs, (2) determine the ventricular loop orientation, (3) determine the relation and anomaly of great vessels origin.

Step 1: determine the visceroatrial situs. \{X, _, _\}

The visceroatrial situs is determined by the relationship of bilateral atria and adjacent thoraco-abdominal visceral organs. It would be much easier to observe the location of abdominal visceral organs at first. In the normal anatomical configuration (situs solitus, designated as\{S, _, _\}), the largest lobe of the liver is on the right side and the spleen and stomach are on the left. If the largest lobe of the liver is on the left side and the spleen and stomach are on the right side, situs inversus (designated as\{I, _, _\})(Figure 1) should be considered.
Second, determine the thoraco-abdominal situs via broncho-pulmonary anatomy since pulmonary sidedness usually coordinate with atria sidedness. The main bronchus of morphological left lung locates below the left pulmonary artery (hyparterial position) and its upper lobe bronchus originates from a more distal site compared to the right side. In contrast, the main bronchus of morphological right lung locates above the right pulmonary artery (eparterial position) and its upper lobe bronchus has a more proximal takeoff compared to left side (Figure 2). The number of pulmonary lobe also helps: the morphological right lung has three lobes while the morphological left lung has two lobes.

Third, determine the location of the morphological right atrium and left atrium. Identify the morphological right atrium according to image features of right atrial appendage (Figure 3): blunt, trapezoidal shape with a broad connection to the rest of the atrium. On the contrary, left atrial appendage (Figure 4), with a tubular, fingerlike shape, is usually narrower than the right and has a narrow connection to the rest of the atria. Sometimes the right and left atrial appendages are not well determined according to the radiologic image, we can observe the drainage chamber of systemic venous return (including inferior vena cava, superior vena cava and coronary sinus) in stead. Due to the venoatrial concordance, the drainage chamber of systemic venous return is usually the morphological right atrium.

At last, determine the visceroatrial situs according to the morphological features just mentioned. If the sidedness of thoraco-visceral structures (atria, bronchus, pulmonary lobes, liver, stomach and spleen) is as usual location, assign the letter S for situs solitus, \{S, _, _\}. If the sidedness of thoraco-visceral structures is all reversed, assign the letter I for situs inversus, \{I, _, _\} (Figure 5, 6). If the orientation of thoraco-visceral structures does not fit into either of condition, assign the letter A for situs ambiguous, \{A, _, _\}. Situs ambiguous is synonymous with Heterotaxy syndrome. Famous examples of heterotaxy syndrome are asplenic syndrome (Figure 7, 8) and polysplenic syndrome (Figure 9, 10) which demonstrates bilateral right-sidedness (right isomerism) and bilateral left-sidedness (left isomerism) of the thoraco-visceral structures respectively.

**Step 2: Determine the ventricular loop orientation. \{_, X, _\}**

During early development of embryo, the primitive heart is a tubular structure. This primitive tubular structure is composed of truncus arteriosus, bulbus cordis, primitive left ventricle and primitive atria in a cranio-caudal direction (Figure 11)[4]. These segmental structures will develop into great vessel, right ventricle, left ventricle and atria in the future, respectively[4]. At about gestational age of 4\textsuperscript{th} week, this cardiac tube begins to elongate and fold on itself to right side or left side (Figure 11). In normal condition, the cardiac tube bends toward right side, making the future right ventricle right side to the future left ventricle and forming a D-loop orientation[5]. In contrast, if the cardiac tube folds toward
left side, positioning the truncus arteriosus left side to the future left ventricle, an L-loop orientation develops. This folding process is important in the embryologic development and determines the position and relationship of major structures of cardiac tube.

To describe the orientation of ventricular loop, we identify the morphological right and left ventricles first. There are several distinguishing features of morphological right and left ventricles. In morphological right ventricle, the trabeculae are coarse and the papillary muscles attach to both free wall and interventricular septum (Figure 12). It also contains the famous "moderator band" (Figure 13), a trabecular structure extending across the right ventricular apex from the anterior papillary muscle to the interventricular septum. On the other hand, the trabeculae of morphological left ventricle are thin and fine, with smooth superior septal surface (Figure 14). The papillary muscles in morphological left ventricle attached to the free wall only.

After identifying the morphological right and left ventricles, we can determine the ventricular loop orientation. If the morphological right ventricle is located right side to the morphological left ventricle, assign the letter D for D-loop orientation, {_, D, _}. If the morphological right ventricle is located leftward of the morphological left ventricle, it indicates a L-loop orientation and the letter L can be denoted.

In complex cases, such as single ventricle, superior-inferior ventricles or failure to determine morphological right and left ventricles, use the loop rule. With the loop rule, we predict the ventricular loop orientation based on the position and relation of the great vessels. In the presence of a right-sided aortic valve(Figure 15), the morphologic right ventricle is located rightward of the morphologic left ventricle (D-loop)(Figure 16)#In the presence of a left-sided aortic valve, the morphologic right ventricle is located leftward of the morphologic left ventricle (L-loop).

**Step 3: determine the relation and anomaly of great vessels origin. {_, _, X}**

There are a group of congenital heart disease with anomaly developing at the level of junction of great vessels and ventricles, known as conotruncal anomalies, such as L-TGA, D-TGA, DORV, TOF…etc. Normally we observe the connection between great vessel origin and ventricles for diagnosis of these malformations. However, in 1980, Van Praagh and Layton et al. proposed the relationship of conotruncal anomalies and the positions of great vessel origin [6]. In their concept, the orientation of great vessel origin can be demonstrated and categorized in a six-point diagram (depicted as Figure 17). Observe the level of aortic valve and pulmonic valve representing origin of great vessels. Look for the aortic root and main pulmonary trunk (MPA) instead if the aortic and pulmonic valves are difficult to identify due to previous operation.
In the normal orientation of great vessels origin, the aortic root is posterior and rightward of the MPA. We assign the letter "S" to denote this normal configuration, which is situs solitus of great vessels: \(\text{S}\)(Figure 18). If the aortic root is posterior and but leftward of the MPA, the letter "I" is assigned to represent situs inversus of great vessels: \(\text{I}\)(Figure 19). When malformation develops at connection of great vessels and ventricles, it may be either transposition or malposition of the great vessels. Transposition of great vessels means the aorta originates from morphological right ventricle and MPA originates from morphological left ventricle. Malposition of the great vessel means both great vessels originate from the same ventricle or are overriding.

In case of transposition, the aorta is usually but not always anterior to the MPA. If the aorta is anterior and right to the MPA, the anomaly is regarded as dextro-transposition of the great vessels, which is recorded as \(\text{D-TGV}\)(Figure 20). If the aorta is anterior and left to the MPA, the anomaly is described as levo-transposition of the great vessels or congenitally corrected transposition of great vessels, which is recorded as \(\text{L-TGV}\)(Figure 21). If the aorta is not anterior or posterior to the MPA, the condition is usually malposition of the great vessels. If the aorta is right side to the MPA, it is described as D-malposition and recorded as \(\text{D-MGV}\)(Figure 22, 23). If the aorta is left side to the MPA, it is described as L-malposition and recorded as \(\text{L-MGV}\)(Figure 24, 25).

Additionally, since the situs solitus, detroposition and dextro-malposition of great vessels result from rightward folding of the primitive cardiac tube (the aorta is rightward of the MPA), these conditions usually concur with D type ventricular loop. On the other hand, situs inversus, levoposition and levo-malposition of great vessels (the aorta is leftward of MPA) often accompany with L type ventricular loop due to leftward folding of the primitive cardiac tube. These phenomena verify the "loop rule" mentioned before.

Step by step, the systemic approach of the Van Praagh notation for complex congenital heart disease has now been completely introduced and demonstrated via various cases. The major concepts and each condition mentioned in each step are summarized in a flowchart (Figure 26).

**Images for this section:**
Fig. 1: This 56-year-old female has her largest lobe of liver (L) at the left side and her stomach (G) and spleen (S) at the right side, which is regarded as situs inversus, {L, G, S}. 


Fig. 2: Normal pulmonary sidedness of broncho-pulmonary anatomy: (1) Hyparterial position of left main bronchus and eparterial position of right main bronchus (black arrow: right main pulmonary artery; black dashed arrow: left main pulmonary artery). (2) Proximal take-off of the right upper lobe bronchus (white arrow) and distal take-off of the left upper lobe bronchus (white dashed arrow).
Fig. 3: Typical appearance of right atrial appendage (white arrow): blunt, trapezoidal shape with a broad connection to the rest of the atrium.
**Fig. 4:** Typical appearance of left atrial appendage (white arrow): tubular, fingerlike shape, narrower than right atrial appendage.
Fig. 5: This 18 year old male has the morphological left appendage (white arrow), which is tubular, narrow and finger-like shape, on the right side. Situs inversus is considered and is designated as \{I,\_,\_\}. 
Fig. 6: This male (same case as Figure 5) has hyparterial location of right main bronchus with distal take-off of the right upper lobe bronchus (arrowhead). On the other hand, the left upper lobe bronchus takes off from a relative proximal location (white arrow). These findings also confirm situs inversus.
Fig. 7: A 1 month old girl with huge transverse liver and right side stomach (white arrow). No definite spleen can be found in the abdomen. Situs ambiguous is suspected and \{A,\_,\_\} is assigned.
**Fig. 8:** This 1 month old girl (same case as figure 7) has eparterial location of bilateral main bronchi with proximal take-off of the bilateral upper lobe bronchi. Bilateral right sidedness of broncho-pulmonary anatomy is confirmed, and splenic syndrome is considered.
Fig. 9: This 5 day old boy has largest lobe of liver(L) at right side with stomach(G) and spleen(S) at left side, which is as normal configuration. However, multiple lobes of the spleen are noted.
**Fig. 10:** This 5 day old boy (same case as figure 9) has hyparterial location of bilateral main bronchi with relative distal take-off of bilateral upper lobe bronchi. Bilateral left sidedness of broncho-pulmonary anatomy is observed, as combined with multiple lobes of spleen shown in figure 9, polysplenic syndrome is considered and {A, _, _} is assigned.
Fig. 11: Diagram illustrates bending of primitive cardiac tube. Cardiac tube is demonstrated from anterior view. Cardiac tube is composed of atrium (A), ventricle (V), bulbus cordis (B) and truncus arteriosus (T). Cardiac tube normally bends to right side, forming a D-bulboventricular loop. Rarely, cardiac tube may bend leftward, forming a L-bulboventricular loop.
Fig. 12: In morphological right ventricle, the trabeculae are coarse (white arrow) and the papillary muscles (black arrow) attach to both free wall and interventricular septum. In contrast, the papillary muscles in morphological left ventricle attach to free wall only.
**Fig. 13:** The morphological right ventricle contains "moderator band" (black arrow), a thick trabecular structure extending from anterior papillary muscle to interventricular septum.
**Fig. 14:** The morphological left ventricle has fine and thin trabeculae (white arrow) with smooth septal surface (S) as compared with thick and coarse trabeculae (black arrow) in morphological right ventricle. Also note the ventricular septal defect (*).  

**Fig. 15:** This 3 year old girl bears aortic root (A) leftward to the main pulmonary trunk (white arrow). According to the loop rule, she would have L-loop ventricular orientation.
**Fig. 16:** This 3 year old girl (the same case as figure 15) has her morphological right ventricle (mRV) located leftward of the morphologic left ventricle (mLV), a L-loop ventricular orientation as the loop rule predicts, assigned as \(_L_\). Notice thick moderator band (black arrow) extending from ventricular wall to septum.
Fig. 17: This simple diagram demonstrates the relationship of great vessel origins on cross sectional image in all six condition, including: situs solitus, situs inversus, D-TGA, L-TGA, D-MGA, L-MGA. A=aorta, PA= pulmonary trunk, TGA= transposition of the great arteries, MGA= malposition of the great arteries.
Fig. 18: Normal configuration of the great vessels: the aortic root (A) is located posterior and leftward to the main pulmonary trunk (PA), at the level of the valves, which is designated as "{_,_,S}".
**Fig. 19:** Inversion of the great vessels: the aortic root (A) is located posterior and rightward to the main pulmonary trunk (PA), at the level of the valves, which is designated as "{_,_,l}".
Fig. 20: This 1 day old boy with D-TGA has aorta(A) arising from morphological right ventricle(mRV) and pulmonary atresia. The aortic root(white arrow) locates anterior and rightward of the pulmonary trunk(PA), which is designated as "[___.D-TGA]".
**Fig. 21:** This 1 week year old girl with L-TGA has aortic root(A) locates anterior and leftward to the main pulmonary trunk(PA), the configuration as shown in figure 17. This is assigned as "{a,a,L-TGA}".
Fig. 22: This 6 year old girl with double outlet right ventricle has aortic root(A) and main pulmonary trunk(PA) arising in a parallel plane with aorta at right side and main pulmonary trunk at left side, which is designated as {_, _, R-MGA}. 
**Fig. 23:** This 6 year old girl with double outlet right ventricle (same case as figure 22) has aortic root (A) and main pulmonary trunk (PA) arising in a parallel plane from morphological right ventricle (mRV), which has thick and coarse trabeculae.

**Fig. 24:** This 10 year old boy with double outlet left ventricle has aortic root (A) and main pulmonary trunk (PA) arising in a parallel plane with aorta at left side and main pulmonary trunk at right side, which is designated as \(_-, _, \text{L-MGA}\).
Fig. 25: This 6 year old girl with double outlet left ventricle (same case as figure 24) has aortic root (A) and main pulmonary trunk (PA) arising in a parallel plane and both from morphological left ventricle (mLV).
Segmental approach of the Van Praagh notation for congenital heart disease

**Step 1:** Determine the viscerocordial situs orientation according the location of liver, stomach, spleen and morphology of lung, bronchus and atria.

- **Situs solitus**  \( \{S, _, _\} \)
- **Situs inversus**  \( \{I, _, _\} \)
- **Situs ambiguous**  \( \{A, _, _\} \)

**Step 2:** Determine the ventricular loop orientation according the location of morphological right ventricle and left ventricle.

- **Dextro-loop**  \( \{_, D, _\} \)
- **Levo-loop**  \( \{_, L, _\} \)
- **Uncertain**  \( \{_, X, _\} \)

**Step 3:** Determine the position and relation of aorta and main pulmonary artery at the level of aortic valve and pulmonary valve.
**Fig. 26:** This flowchart demonstrates the three-step approach of the Van Praagh notation for complex congenital heart disease. Notice the major concepts of each step and each condition mentioned in every step.
Conclusion

The Van Praagh notation, as a step-by-step approach, is a clear-cut and concise expression system for evaluation of complex congenital heart disease and for communication and discussion between the medical colleagues. With practice, even complicated cases can be evaluated correctly and expressed precisely in a short period of learning. Cardio-radiologist should be familiar with this systemic analysis notation for understanding complex congenital cardiac anomaly and communicating with cardiologists, surgeons and pediatricians.

Personal information

References


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