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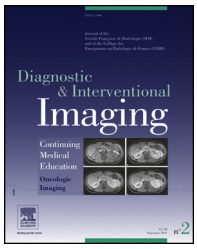

CONTINUING EDUCATION PROGRAM: FOCUS...

Cardiac CT and MRI of cardiac malformations: How to interpret them?

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Abstract A segmental sequential approach is widely used for the description of congenital heart disease abnormalities in routine reports of computed tomography and magnetic resonance imaging examinations. This consists of three stages as follows: (a) the anatomical description of each segment (viscero-atrial situs, the bulboventricular loop and the position of the great vessels); (b) the relationship between each segment at the atrioventricular and ventriculoarterial levels; and (c) related intra- and intersegmental abnormalities. This article describes the interpretation of computed tomography and magnetic resonance imaging examinations in patients with cardiac malformations using a structured plan.

The development of cardiac computed tomography (CT) and magnetic resonance imaging (MRI) has increased radiologists' interest in the diagnosis of congenital heart disease (CHD). In order to play a role in the diagnosis and monitoring of these diseases, radiologists need to use the same language as cardiologists and cardiac surgeons in classifying and describing these diseases. Introduced by Van Praagh et al. and then Anderson et al., the segmental sequential approach of cardiac anatomy, which is widely used throughout the world, provides a succinct, accurate and unambiguous description of all of the possible combinations of cardiac malformations [1—10]. The strength of this approach lies in the fact that it is easy to understand and can be used in cross-sectional imaging.

The goal of this article is to describe the different stages of the segmental sequential approach in analyzing CHD, review the important concepts of cardiac embryology associated with this approach, and demonstrate the practical value of the segmental approach in assessing CHD on cross-sectional imaging.

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The segmental sequential approach

The segmental sequential approach is based on three stages:
- anatomical description of each segment;
- the relationship between each segment;
- related intra- and intersegmental abnormalities [11—13].

Anatomical description of each segment

Identification of cardiac anatomy forms the basis of Van Praagh's segmental approach [1]. Through this approach, the heart is separated into three different segments, the viscero-atrial situs, the ventricles and the position of the great vessels. With this analysis, Van Praagh developed an annotation system made up of a series of three letters separated by commas, in parentheses. This will be explained together with the description of the segments.

Viscero-atrial situs

Three types of viscero-atrial situs exist: solitus (S,-,-), inversus (I,-,-) and ambiguous (A,-,-) [14,15]. By definition, the situs is determined by the relationship between the atria and the abdominal viscera. The atria are identified from the morphological examination of the atrial appendages, which are their extensions. The right atrial appendage is wide and triangular in shape, whereas the left is narrow and tubular, and is finger shaped (Fig. 1).

Situs solitus is the normal anatomical configuration, where on the right of the patient, an atrium of right morphology, the liver, a lung with three lobes and an early origin of the upper lobe bronchus are found, together with an anterior path of the right pulmonary artery to the right mainstem bronchus (epiarterial bronchial position). On the left, there are an atrium of left morphology, the stomach, the spleen, a bilobed lung with a distal origin for the upper lobe bronchus and the left pulmonary artery passes superiorly to the left mainstem bronchus (hypoarterial bronchial position). The anatomical configuration is reversed in situs inversus.

If situs is neither solitus nor inversus, the term situs ambiguous is used (heterotaxia). Situs ambiguous is often associated with congenital asplenism, less commonly with polysplenism, and occasionally with a normal spleen. Many abnormal viscero-atrial configurations may be found although many authors separate situs ambiguous into asplenism (right isomerism) and polysplenism (left isomerism).

Classically with asplenism, two atria of right atrial morphology are found, with two lungs having three lobes, a liver in the central symmetrical position, no spleen, and total anomalous pulmonary venous return (TAPVR). The abnormalities seen with polysplenism include two atria of left atrial morphology, two lungs with two lobes, interruption of the inferior vena cava (IVC), multiple spleens, and pulmonary veins draining both into the right and left atria.

Practical aspects

It may be difficult to determine the morphology of the atrial appendages on imaging and the position of extracardiac organs helps to determine situs. A few tips for these are shown below:
- On a thoracic level, the bronchial anatomy and relationship between the pulmonary arteries and the mainstem bronchus are excellent indicators of atrial configuration (Fig. 2).
- According to the venoatrial concordance rule, the IVC usually drains into an atrium of right configuration [1,5,12] and the configuration of this atrium can therefore be categorized.
- On an abdominal level:
  - Ultrasound can show the position of the liver and stomach and determine the number and position of spleens, which are major factors in identifying the type of situs.
  - A liver, which is increased in size lying centrally is often associated with asplenism [13] (Fig. 3).
- Although the position of the cardiac apex does not form part of the description of situs in general, cardiac and situs abnormalities are found with a discordant position for the cardiac apex, stomach, and aortic arch. However, situations in which the heart is displaced secondary to a chest and/or lung abnormality, such as pulmonary hypoplasia [14], should not be included.

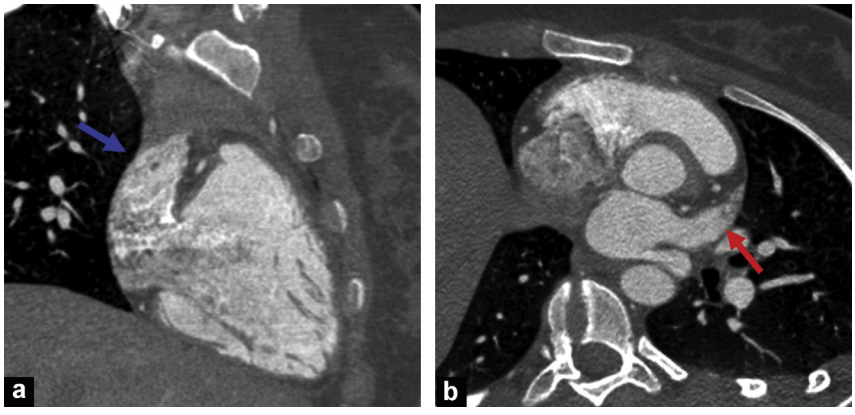

Figure 1. Normal atrial appendage morphology. Thoracic computed tomography (CT) angiography: a: image in the coronal plane shows triangular trabeculated appearance of the right atrial appendage (blue arrow); b: image in the transverse plane shows tubular smooth appearance of the left atrial appendage (red arrow).