

# Congenital Anomalies and Anatomical Variants of the Coronary Sinus

Anomalías congénitas y variantes anatómicas del seno coronario

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Coronary sinus  
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 Coronary artery disease



## Palabras clave (DeCS)

Seno coronario  
 Tomografía computarizada multidetector  
 Enfermedad de la arteria coronaria

## Summary

The coronary sinus (CS) is an important vascular structure that allows access to the coronary veins in multiple interventional procedures such as mapping and ablation of arrhythmias, implantation of left ventricular electrodes in resynchronization therapy, mitral annuloplasty, stem cells therapy, and retrograde cardioplegia. The success of these procedures is facilitated by the knowledge of the CS anatomy, in particular the recognition of its variants and anomalies. The widespread availability of multislice CT (MSCT) allows non-invasive image acquisition with excellent representation of the CS anatomy, avoiding thus the need for invasive diagnostic procedures. MRI is also useful for the non-invasive evaluation of the coronary sinus, without radiation, allowing the morphological and functional assessment of various congenital heart abnormalities.

## Resumen

El seno coronario (SC) es la estructura vascular que permite el acceso a las venas coronarias en procedimientos intervencionistas como el mapeo y la ablación de arritmias, la implantación de electrodos izquierdos en terapia de resincronización, anuloplastia mitral, tratamiento con células madre y cardioplejía retrógrada. El éxito de estos procedimientos depende del conocimiento de la anatomía del SC, del reconocimiento de sus variantes y anomalías. La tomografía multicorte permite obtener imágenes de la anatomía del SC sin necesidad de procedimientos invasivos de diagnóstico. La resonancia magnética, por su condición no invasiva y sin radiación, también es útil en la evaluación del seno coronario porque permite la valoración morfológica y funcional de diversas anomalías congénitas del corazón.

## Objectives

To disclose the functionality of Magnetic Resonance Imaging (MRI) and multi-slice tomography (MS-CT) in the assessment and diagnosis of congenital anomalies and anatomical variants of the coronary sinus; to define the clinical significance of these entities and present representative cases of the proposed topic.

## Materials and methods

Patients diagnosed with an anomaly or coronary sinus variant by cardiac MRI and MS-CT.

## Results

Before practicing interventional procedures in electrophysiology or cardiac surgeries it is important to know the anatomy of the coronary sinus and coronary venous drainage. These range from simple incidental variants without clinical significance up to anomalies that require surgical management. The characteristic features of CT and MRI will be described for the following variants and abnormalities: Variants of caliber and coronary sinus shape, “unroofed” coronary sinus, persistence of left superior vena cava draining to the coronary sinus and stenosis of the ostium of the coronary sinus.



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## Conclusion

Cardiac MRI and MS-CT allow for a detailed evaluation of the normal anatomy, anatomical variants and congenital abnormalities of the coronary sinus in a non-invasive manner.

## Introduction

The coronary sinus (CS) is the vascular structure that allows access to coronary veins in multiple interventional procedures, such as mapping and ablation of arrhythmias, the implantation of left electrodes in resynchronization therapy, mitral annuloplasty, treatment with stem cells and retrograde cardioplegia. The success of these procedures depends on the knowledge of the anatomy of the CS, in particular the recognition of its variants and anomalies (1). The wide availability of the MS-CT allows to obtain images with excellent representation of the anatomy of CS, which avoids invasive diagnostic procedures. MRI is also useful in the non-invasive and radiation-free evaluation of CS, which allows the morphological and functional evaluation of various congenital heart abnormalities (1-3).

The CS is a venous duct located between the coronary veins and the right atrium. It has tributary drainage veins of the two ventricles and atria. It is posterior to the coronary sulcus, with its orifice located medially and anterior to the orifice of the vena cava and immediately above the atrioventricular groove. Usually the ostium of the CS is located in the right atrium and is guarded by a semicircular valve called the valve of Thebesium (1,3,4).

CS congenital anomalies cover a broad spectrum ranging from anatomical variants that are not pathologically meaningful to haemodynamically significant anomalies such as “unroofed” coronary sinus (CSS), anomalous venous drainage to CS, and coronary artery fistulas to the coronary sinus. Although some anatomical variants of CS are asymptomatic and do not represent pathology, they can create difficulties when performing procedures interventionists (1).

## Normal anatomy

The CS is the largest coronary venous structure. Its normal measurements are: average length between 45-63 mm; mean diameter of 7.05 mm  $\pm$  1.90; normal diameter of the ostium from 4  $\times$  5 mm up to 9  $\times$  16 mm. CS dilatation is established when the diameter is > 11 mm measured at 3 cm from the ostium (2).

The major part of the coronary venous circulation converges to the coronary sinus.

## The coronary sinus and its affluents (Figure 1) (1-6)

- » The anterior interventricular vein (AIV) ascends through the ascending interventricular sulcus and continues as the major coronary vein (MCV)
- » The major cardiac vein or major coronary vein (MCV) extends

along the left atrioventricular groove and ends in the CS. The transition between MCV and CS is defined by the origin of the Marshall oblique left atrial vein externally (Figure 2b) and by the Vieussens valve internally. The oblique vein runs along the back and left side of the left atrium and is located within a vestigial fold known as the Marshall’s ligament, which is the remnant of the development of the left superior vena cava (1-6).

- » The left ventricular marginal vein (VMV) runs along the lateral aspect of the left ventricle and leads to the MCV or directly towards the CS. VMV may be absent in patients with previous anterolateral myocardial infarction. This information is important in resynchronization therapy, since the left electrode should be located on the side wall of the left ventricle (the last area to be activated in patients with systolic dysfunction) where VMV is usually present. The VMV can be unique or multiple and information about their caliber, location, number and angle of origin is important before resynchronization therapy (figure 3). If there is no anatomy of the marginal vein, the left electrode of the resynchronization therapy must be surgically implanted (5).
- » The posterior vein (PV) drains the diaphragmatic and lateral wall of the left ventricle and in most cases leads to the CS. It can also drain into the MCV. The PV can be unique or multiple.
- » The middle posterior or cardiac interventricular vein runs through the posterior interventricular groove and ends in the inferior face of the CS, just proximal to its termination in the right atrium.
- » Minor coronary vein, when present, has its course through the inferior right atrioventricular groove, drains to the CS or directly into the right atrium (1-6).

## Variants and congenital anomalies of the coronary sinus

### 1. Variants of caliber, form and course (Figure 4)

The morphology of the CS is highly variable. The variants in morphology of the CS include the CS diverticulum, which is usually located in the inferior aspect of the union of the CS with the posterior interventricular vein. It has been reported that people with CS diverticulum may suffer from frequent arrhythmias, which it is required to do ablation. As for the form, they can be found in “wind sack”, filiform, varicose or bifid. Regarding the course, the most important variant is the high coronary sinus, in which the CS is found outside the atrioventricular groove (figures 4 b and d). This finding is important when planning a mitral annuloplasty because when the CS does not coincide with the mitral valve ring, the high CS, it is not favorable to perform this procedure through the coronary sinus (1,7,8).

### *Sub-Thebesian sac or bag (Keith sinus) (Figure 5).*

It is a sacular reinforcement area of the right atrium located inferior to the drainage ostium of the CS to the right atrium that can serve as a substrate for the development of reentrant atrial flutter. It is also associated with technical difficulties in interventional procedures of CS and the right ventricle (1).

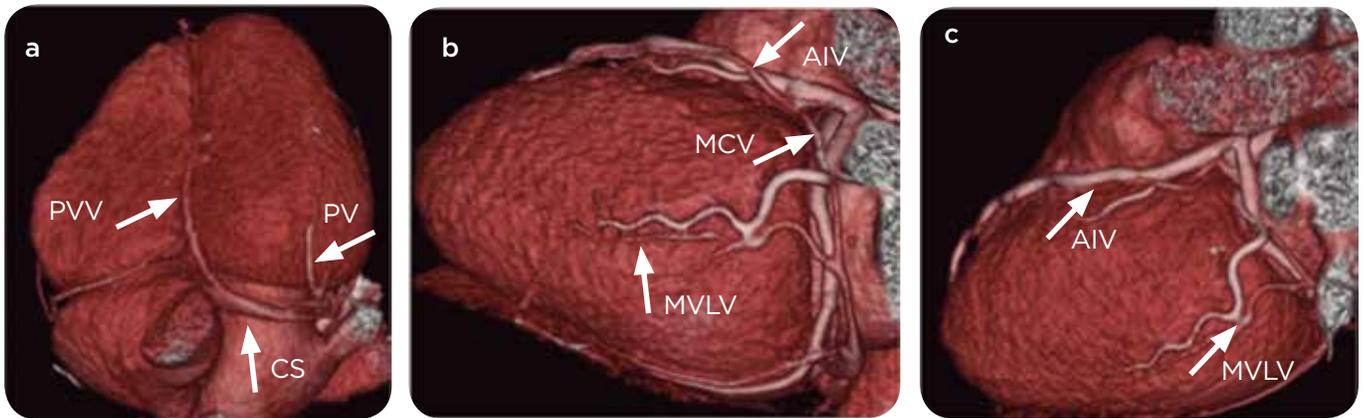


Figure 1. Normal coronary venous anatomy. Cardiac MS-CT of different patients. Three-dimensional reconstructions with projections of the heart a) inferior, b) left lateral and c) superior. Anterior interventricular vein (AIV), major coronary vein (MCV), marginal vein of the left ventricle (MVLV), posterior vein (PV), coronary sinus (CS), posterior ventricular vein (PVV).

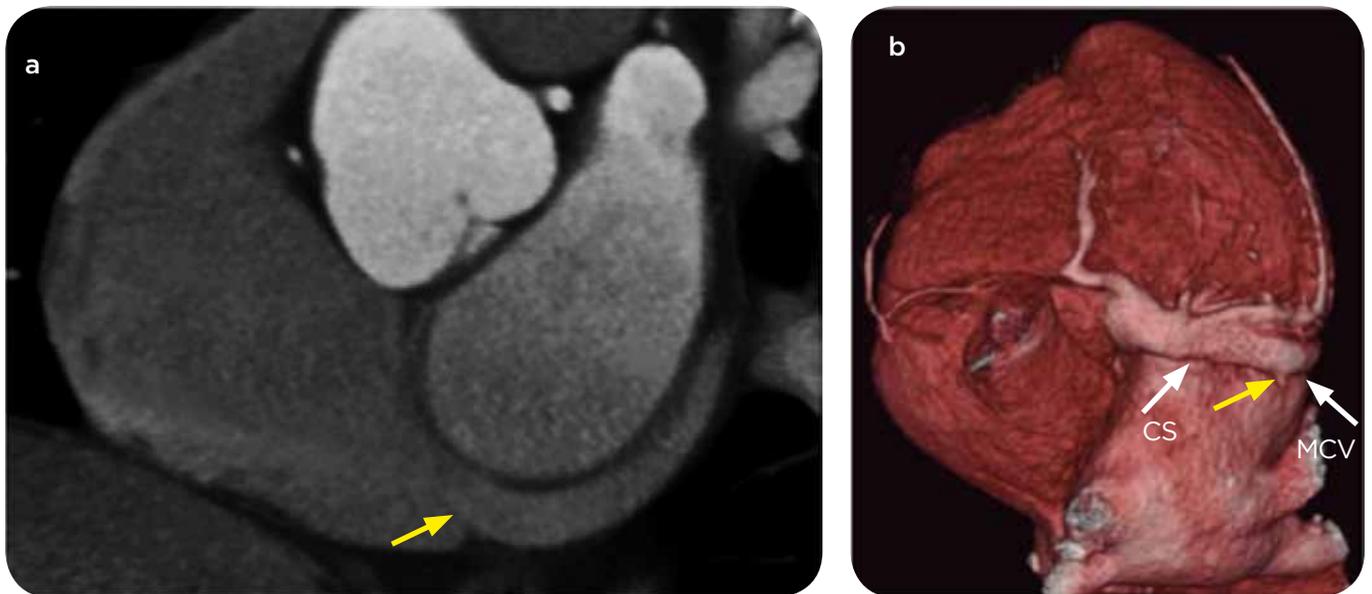


Figure 2. Cardiac MS-CT of different patients. a) Multiplanar normal reconstruction of the CS curve: Thebesian valve (arrow). b) Three-dimensional reconstruction of the inferior projection of the heart, transition between CS and major coronary vein (MCV) at the site of origin of the Marshall oblique vein (yellow arrow).

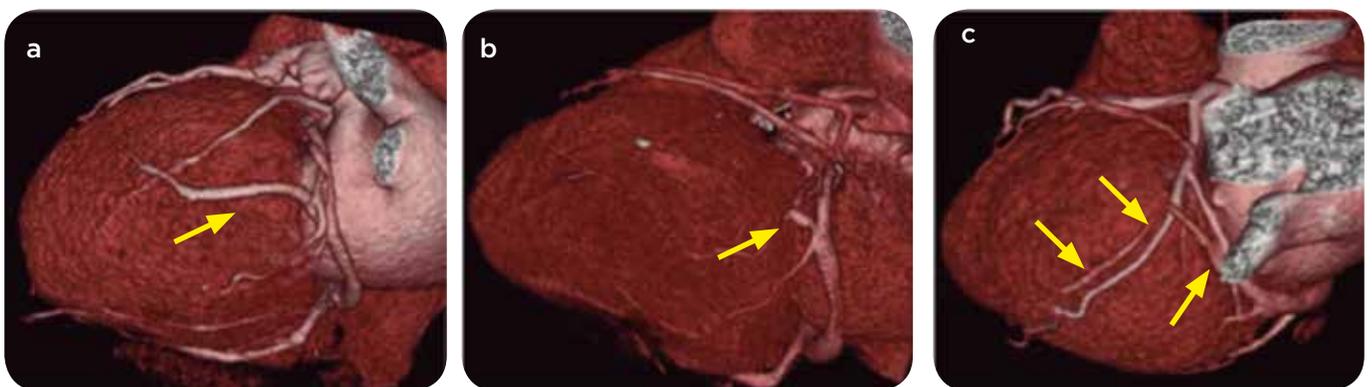


Figure 3. Cardiac MS-CT. Three-dimensional reconstructions in left lateral oblique projection of the heart. Anatomy for implant of left ventricular resynchronization therapy electrode in the marginal vein of the left ventricle (lateral wall of the left ventricle). a) Positive anatomy with left marginal vein (arrow) of good caliber and favorable angle of origin. a and c) Non-propitious anatomy, with marginal vein occluded (arrow in b) and angled marginal vein of low caliber (arrows in c).

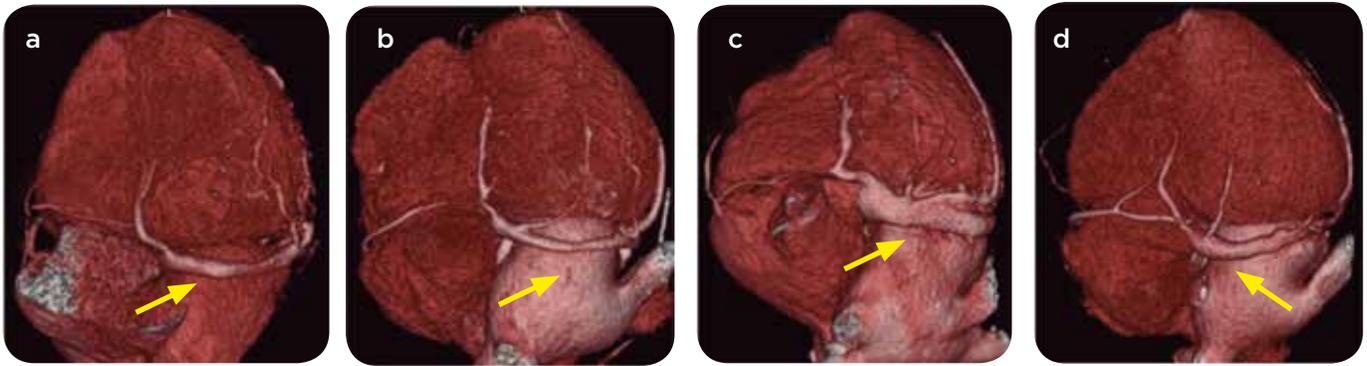


Figure 4. Variants of caliber, shape, and coronary sinus pathway. Cardiac MS-CT of different patients. Three-dimensional reconstructions in inferior projections (diaphragmatic face of the heart). a) Tubular CS (most common) (arrow). b) Filiform CS and high path (arrow). c) Varicose CS (arrow) and d) CS in "wind bag" and high path (arrow). Source: Modified from the article by Shah et al. (1) to illustrate the frequent variants of CS in the cases studied.

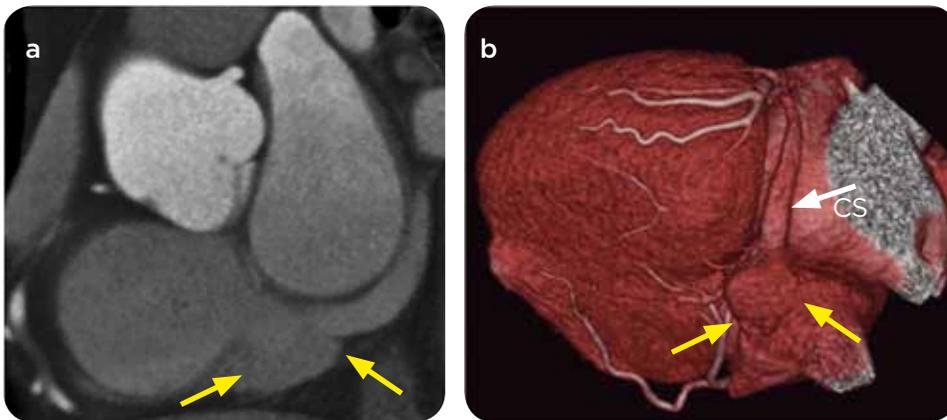


Figure 5. Sub-Thebesian bag (Keith sinus). Cardiac MS-CT. a) Multiplanar reconstruction of the coronary sinus curve (arrows). b) Three-dimensional reconstruction, oblique lower projection of the heart where there is a prominent sub-Thebesian bag (yellow arrows)



Figure 6. Cardiac MDCT. Multiplanar reconstruction of the coronary sinus curve. Persistence of the left superior vena cava (arrows) draining to CS with central venous line in its interior.

## 2. Increase in size

There are several entities that can be found with increase of the CS size associated with a dilated right atrium, for example: tricuspid stenosis, tricuspid insufficiency, dysfunction of the right ventricle and pulmonary hypertension, among others.

Congenital structural abnormalities that can occur with increase in CS size can be divided into anomalies without short circuit and anomalies with a left-right short circuit.

**Anomalies without short circuit:** The CS expands when receiving a larger volume of the systemic venous circulation, usually by persistence of the superior left vena cava (SLVC) (Figure 6) draining to the CS. It is estimated that persistent SLVC has a prevalence of 0.3 % of the general population. The SLVC originates by the confluence of the internal jugular vein and the left subclavian vein, passes lateral to the aortic arch and usually drains to the CS. Persistent SLVC has no haemodynamic or short-circuit repercussion when the coronary sinus roof is intact (1,7-10).

Other causes of CS dilatation without short-circuitry are the continuation of the azygos vein or vein with the SLVC with interruption of the inferior vena cava (common in syndromes of heterotaxia) and direct drainage of the suprahepatic veins into the CS.

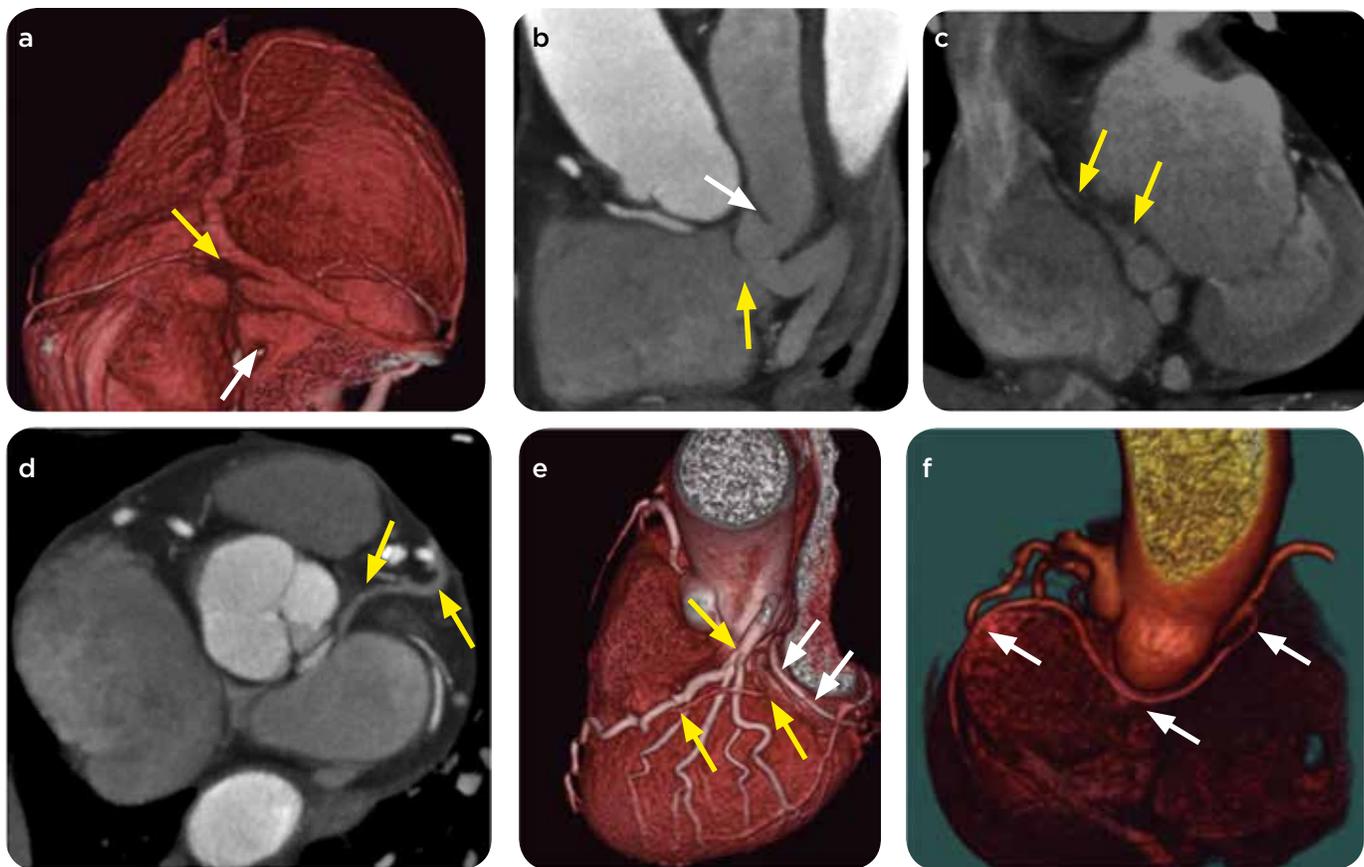


Figure 7. Cardiac MS-CT. a) Three-dimensional reconstruction in inferior projection. b and c) Curved reconstructions of the coronary sinus. d) Reconstruction curve of the anterior interventricular vein. e and f) Oblique upper three-dimensional reconstructions. Ostium stenosis of the CS (yellow arrows in a and b) with diameters of  $3 \times 3$  mm, without persistence of SLVC, with tortuous and dilated CS, and communication with the left atrium / “unroofed” (white arrows in a and b). Collateral accessory vein that communicates the CS with the upper part of the right atrium (arrows in c). Anterior interventricular vein (AIV) draining to the left atrium and not communicating with MCV (arrows in d and e). The patient also has a circumflex artery that originates in the right Valsalva sinus with a benign retroaortic path (white arrows in e and f). Note in (e) the lack of continuity of the circumflex artery with the common trunk of the left coronary artery due to its anomalous origin and the lack of continuity of the anterior interventricular vein with the major coronary vein by the direct anomalous drainage of the AIV to the left atrium. In e) the circumflex artery and the anterior interventricular vein have a parallel path extending in the direction of the retroaortic space.

**Anomalies with short circuit:** The oxygenated blood from the lungs can return to the right atrium through the CS by three routes: from the left atrium (LA) by an “unroofed” CS, or by the pulmonary veins in the abnormal venous returns that drain to the CS, these are low-pressure short-circuits or high-pressure fistulas between the coronary arteries to the CS (1,11). Scaling CS is a rare congenital anomaly that produces an internal short circuit due to a wall defect between the CS and the LA. In 63 % of cases it is associated with persistent SLVC; as well it is associated with interventricular communication, right ventricular outflow tract obstruction (including tetralogy of Fallot) and the cor triatriatum. Patients with unroofed CS may have the potential risk of cerebral abscesses and paradoxical emboli when they are associated with persistent SLVC, because intermittent right-to-left short-circuit may appear in episodes where right cardiac cavity pressure increases. MRI in the phase contrast sequence can define the direction of the short circuit (1).

### 3. Absence

The absence of the CS is usually associated with heterotaxia syndromes.

### 4. Atresic Ostium

In CS with atresic or stenotic ostium the coronary venous flow can have two possibilities: persistent SLVC or without persistent SLVC (1,8).

**Atresic or stenotic ostium with persistent SLVC:** It is a rare anomaly where the SLVC allows retrograde circulation of the venous flow Venous to the right superior vena cava through a bridge vein and from there to the right atrium, so that there is no short circuit or haemodynamic compromise; This anomaly is important because it contraindicates the ligation of the SLVC.

**Atresic or stenotic ostium without persistent SLVC:** In these cases, the coronary venous flow has communication with the left atrium from the coronary sinus through single or multiple communication. Clinically, this anomaly behaves as a small right-left short circuit, generally insignificant (Figure 7) (1,9).

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