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An exceptional anomaly of the coronary venous drainage: anatomic description

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Abstract

Background: Anomalies of the coronary sinus are rare in the general population but are more frequent in patients with congenital heart defects. Whatever the cardiac anatomy, the coronary sinus is invariably located in the left atrioventricular sulcus, inferior to the wall of the morphologically left atrium. *Methods:* A complete morphological examination of a fetal cardiac specimen of the M3C collection, according to segmental analysis, was performed by two observers. *Results:* We report here for the first time a cardiac specimen with a venous channel receiving the majority of coronary veins, located in the right atrioventricular sulcus and therefore inferior to the wall of the morphologically right atrium, in a fetal cardiac specimen with congenitally corrected transposition in situs solitus. In addition, the anatomy of the venous drainage of the heart was mirror-imaged to that observed in a normal heart and different compared to that usually observed in congenitally corrected transposition. *Conclusion:* This very particular anatomy occurring in association with congenitally corrected transposition might be related with an additional disturbance in the laterality pathway.

Introduction

The interest of cardiologists for the coronary sinus has been renewed with the development of transcatheter techniques of pacing and resynchronisation therapies. Congenital anomalies of the coronary venous system have been extensively described, mostly from CT-scan or MRI examinations.[1–4] These anomalies are rare in the general population (0.33%,[3]) but are more frequent in patients with CHDs. However, whatever the morphology of the coronary sinus, it remains axiomatic that it is invariably located in the left atrioventricular sulcus, inferior to the wall of the morphologically left atrium.[5]

We report here for the first time, to the best of our knowledge, a venous channel located in the right atrioventricular sulcus and therefore inferior to the wall of the morphologically right atrium receiving the majority of the coronary venous drainage, in a fetal cardiac specimen with congenitally corrected transposition in situs solitus.

Anatomical description of the fetal cardiac specimen

A fetal screening ultrasound was performed at 33 weeks of gestation in a 40-year-old woman without significant medical history and revealed a congenitally corrected transposition. The parents chose to terminate the pregnancy at 35 weeks of gestation. Fetal pathologic examination did not reveal any extracardiac anomaly.

At fetal cardiac examination, the heart was in levocardia, and the diagnosis of congenitally corrected transposition was confirmed (Figure 1). The morphologically right atrium was right-sided and connected to the right-sided morphologically left ventricle, itself connected to a right-sided and posterior pulmonary artery. The morphologically left atrium was left-sided and connected to the left-sided morphologically right ventricle, which connected to a left-sided anterior aorta. The segmental analysis was therefore {S, L, L}: atrial situs solitus, ventricular L-loop, L-transposition of the great arteries.[6] The ventricular septum was intact and the tricuspid valve was anatomically normal. The oval foramen and the arterial duct were patent and of normal size.

The coronary arteries connected to the aorta in usual pattern for congenitally corrected transposition, following their respective ventricles[7]: the left-sided coronary artery was morphologically right and connected to the left-facing aortic sinus and the right-sided coronary artery was morphologically left, connected to the right facing aortic sinus and divided in anterior interventricular artery and circumflex artery. A large infundibular branch supplying the right ventricular outflow tract and crossing the subaortic infundibulum arose from the anterior interventricular artery (Figure 1).

The anatomy of the coronary sinus was unusual (Figure 2). The coronary sinus orifice opened as usual in the morphologically right atrium above the entrance of the inferior caval vein, bordered by a Thebesian valve. The left-sided coronary sinus coursed in an oblique fashion

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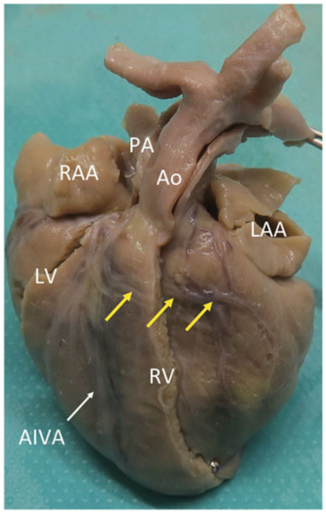

Figure 1. External anterior view of the cardiac specimen. Congenitally corrected transposition in atrial situs solitus {S, L, L}. The right-sided right atrium right atrial appendage (RAA) is connected to the right-sided left ventricle (LV), which ejects in the right-sided pulmonary artery (PA). The left-sided left atrium left atrial appendage (LAA), connects to the left-sided right ventricle (RV), which ejects in the left-sided aorta (Ao). AIVA, anterior interventricular artery; yellow arrows, large infundibular branch crossing the subaortic infundibulum.

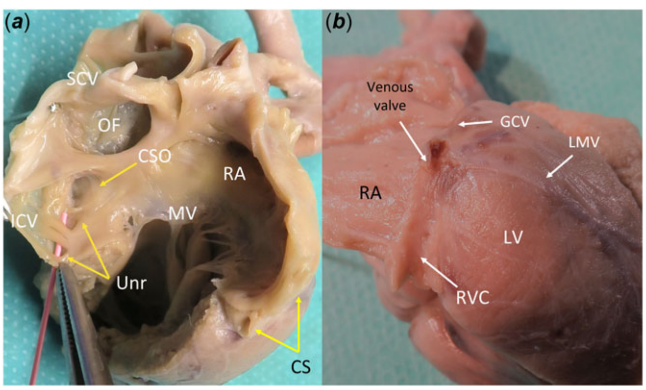

Figure 2. Anatomy of the coronary sinus. ***(A)*** The right-sided right atrium is open, showing the atrial septum (OF, oval fossa), the mitral valve (MV) and the left ventricle underneath. The coronary sinus orifice (CSO) opens between the inferior caval vein (ICV) and the oval fossa. The right-sided venous channel (RVC) courses in the right inferior atrioventricular groove, and is partially unroofed in two locations (Unr) near the atrial septum. RA, morphologically right atrium: SCV, superior caval vein. ***(B)*** External view of the right atrium (RA) and the left ventricle (LV). The right-sided venous channel is open. A venous valve is located at the junction between the great cardiac vein (GCV) and the coronary sinus, immediately after the connection between the left marginal vein (LMV) and the great cardiac vein.

behind the morphologically left-sided left atrium on a very short distance (4 mm) and received the oblique vein of Marshall, the inferior interventricular vein (or middle cardiac vein), and the small cardiac veins (Figure 3). Surprisingly, there was another venous structure in the right atrioventricular groove, which received the major coronary veins and extended to the right border of the heart. This second venous channel, right-sided, was partially unroofed at its extremity. It opened in the right-sided morphologically right atrium through two small defects within the wall separating the venous channel from the right atrium, near the connection of the inferior caval vein to the right-sided morphologically right atrium, and close to the opening of the left-sided coronary sinus (Figure 2). There was a small venous valve at the junction between the venous channel and the great cardiac vein (Figure 2).

The anatomy of the coronary veins is depicted in Figure 3. The great cardiac vein, or the anterior interventricular vein, coursed in the right-sided atrioventricular groove before joining the distal part of the right-sided venous channel, near the entrance of the right atrial appendage. The anterior lateral vein and the left marginal vein were located on the lateral side of the right-sided morphologically left ventricle and merged with the great cardiac vein, before joining the right-sided venous channel. Some small left inferior veins drained directly into it.

The inferior interventricular vein (middle cardiac vein) is connected to the left-sided coronary sinus, next to its orifice. The small cardiac vein located to the anterior and lateral surface of the right ventricle joined the left-sided coronary sinus at the junction with the inferior interventricular vein.

Discussion

We present here an anomaly of the coronary venous drainage never described so far in the literature, in which the main coronary veins, including the great cardiac vein, are connected to a right-sided venous channel located in the right atrioventricular groove, in a fetal heart specimen with congenitally corrected transposition {S, L, L} without any associated anomalies.

The coronary venous system in the normal heart

In the normal heart in atrial situs solitus, the coronary sinus is located into the inferior left atrioventricular groove, inferior and posterior to the morphologically left atrium (Figure 4). It opens into the right atrium in posteromedial fashion, just above and medial to the orifice of the inferior caval vein. The orifice of the coronary sinus is guarded by the Thebesian valve, which partially overlaps it. The coronary sinus drains blood predominantly from the left ventricle, the left atrium, part of the right ventricle, and the interventricular septum.[1]

The two major components of the coronary venous system, present in 90% of humans, are the great cardiac vein and the middle cardiac vein.[5] The great cardiac vein courses adjacent to the left anterior descending artery in the anterior interventricular sulcus, then in the left atrioventricular groove adjacent to the circumflex artery (left circumflex vein), and finally drains into the coronary sinus just before the connection of the left atrial oblique vein or vein of Marshall (Figure 4). The junction between the great cardiac vein and the coronary sinus is bordered by the venous valve of Vieussens, that is present in 87% of the population.[1,4] Three major veins drain into the great cardiac vein or the coronary sinus. The left ventricular marginal vein is located along the left side of the heart and connects with the great cardiac vein before its junction with the coronary sinus. The posterior vein of the left ventricle drains into the coronary sinus.[4,5] The inferior interventricular vein, or middle cardiac vein, runs in the inferior interventricular grove and joins the coronary sinus just before its opening in the right atrium. However, if the coronary sinus is always located in the left atrioventricular sulcus, the morphology of its tributaries is highly variable.[5] The coronary venous system of the right ventricle is less developed. The small cardiac vein (or right circumflex vein) courses in the right atrioventricular groove and joins the coronary sinus to the right of the middle cardiac vein. The anterior wall of the right ventricle is drained via the Thebesian

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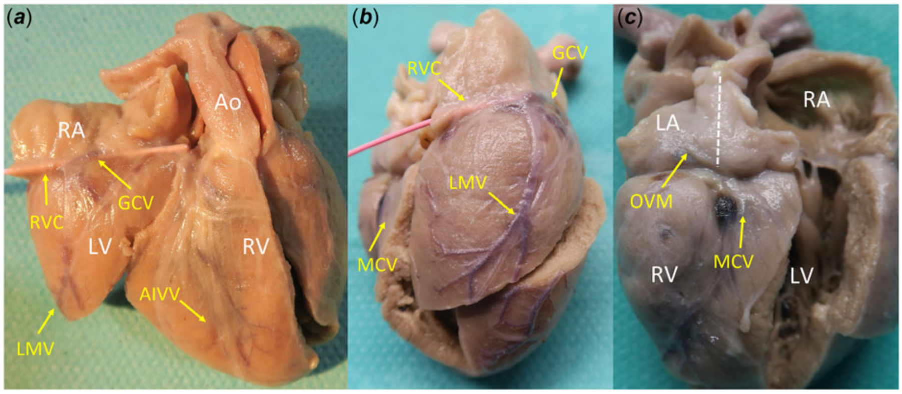

Figure 3. Anatomy of the coronary venous drainage. (***A***) The anterior interventricular vein (AIVV) courses in the anterior interventricular sulcus, next to the anterior inter ventricular artery, and becomes the great cardiac vein (GCV) as it courses in the right-sided atrioventricular groove, before joining the right-sided venous channel (RVC) at the junction with the left marginal vein (LMV). (***B***) The left marginal vein (LMV) joins the distal part of the great cardiac vein (GCV) before its junction with the right-sided venous channel (RVC). MCV, middle cardiac vein. (***C***) The inferior interventricular vein or middle cardiac vein (MCV) courses in the inferior interventricular groove and connects the right-sided venous channel (RVC) at its proximal part. The oblique vein of Marshall (OVM) is short and opens in the right atrium via the orifice of a very short left-sided coronary sinus. LA = left atrium. LV = left ventricle; RA = right atrium; RV = right ventricle.

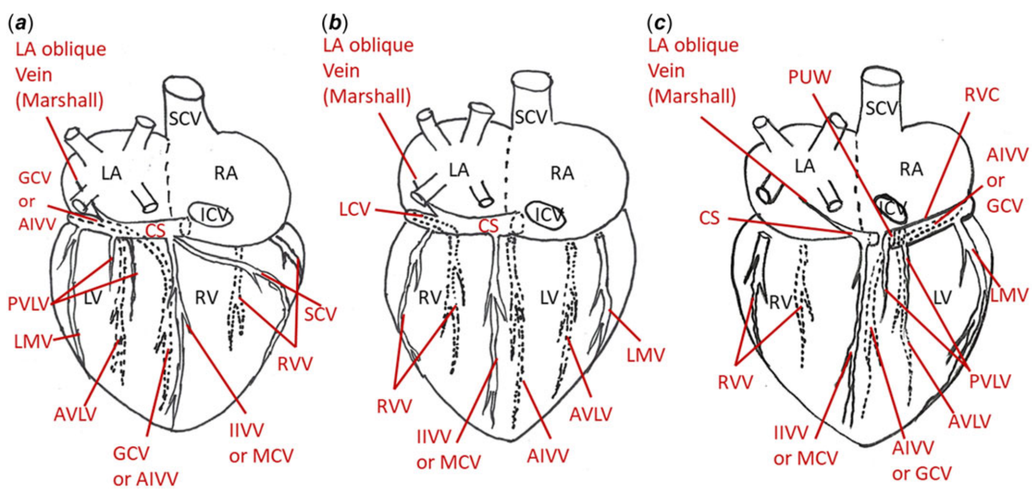

Figure 4. Diagram indicating the distribution of the coronary venous system in posterior view of the heart in Valentine position. (***A***) In a normal heart. (***B***) In congenitally corrected transposition {S, L, L}. (***C***) In our cardiac specimen. AIVV = anterior interventricular vein; AVLV = anterior vein of the left ventricle; CS = coronary sinus; GCV = great cardiac vein; ICV = inferior caval vein; IIVV = inferior interventricular vein; LA = left atrium; LCV = left circumflex vein; LMV = left marginal vein; MCV = middle cardiac vein; PUW = partially unroofed wall; PV = pulmonary veins; PVLV = posterior veins of the left ventricle; RA = right atrium; RV = right ventricle; RVC = right-sided venous channel; RVV = right ventricular veins; SCV = superior caval vein.

veins which open directly in the right atrium or in the right ventricle.[1,5]

Embryology of the coronary sinus

Embryologically, the sinus venosus (or venous sinus) is at first a medial structure, which receives the blood of the hepatocardiac channels, formed by the fusion of the umbilical and vitelline veins, and later on of the common cardinal veins.[8] The right part of the systemic venous sinus is growing more rapidly than the left. The appearance of myocardium on the epicardial side of the asymmetrically expanding systemic venous sinus delineates the right and left sinus horns.[8] Transcription factor Tbx18, expressed in the sinus horns but not in the atrial walls, is required for differentiation of the myocardium of the sinus horns from mesenchyme.[8,9] With the establishment of left-right asymmetry, the right horn and the right common cardinal vein progressively incorporate into the right atrium. The left cardinal vein gradually obliterates, like the distal part of the left sinus horn that becomes the ligament of Marshall, while its proximal part remains as the coronary sinus.[8,10] The caudal part of the left cardinal vein can persist as the left superior caval vein draining into the coronary sinus.[8,10]

Embryology of the coronary veins

Since the work of Red-Horse et al. in mouse embryos,[11] it is established that the coronary vessels (arteries, capillaries, and veins) all derive from an immature vascular plexus made of endothelial cells originating from the sinus venosus. This vascular plexus appears first at the dorsal surface of the heart, near the sinus venosus, and expands all over the surface of the heart, in the inferior then anterior interventricular groove, and around the atrioventricular junction up to the developing outflow tract.

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The vascular plexus is thus initially composed of venous cells. As some of the vascular sprouts gradually invade the underlying myocardium, they lose their venous identity to express arterial markers (*EphrinB2*) and form coronary arteries within the myocardium. The vascular network at the surface of the heart expresses venous markers and forms the coronary veins. Arteries emerge at the surface of the heart, and coronary veins appear, at the confluence of the dorsal interventricular and atrioventricular grooves. The two other sources of endothelial coronary cells are the endocardium of the ventricles, and for a very small part the proepicardium.[12–14]

In addition to the venous markers *EphB4* and *COUP-TF2*, the surface coronary veins need a specific factor, *Ang1* (myocardium-derived angiopoietin-1) for their development.[14] The development of the coronary veins is intimately related to that of the coronary arteries, as both develop together in the interventricular and atrioventricular grooves. Why their final connection takes place in two so distant regions of the heart is still unknown.[15]

The coronary venous system in hearts with congenitally corrected transposition

Similar to that of the coronary arteries, the anatomy of the coronary venous system is different in congenitally corrected transposition compared to the normal heart (Figure 4).

In congenitally corrected transposition in atrial situs solitus, the coronary sinus is as usual located in the left atrioventricular groove, adjacent to the postero-inferior wall of the morphologically left atrium.[16–18] However, because of the discordant atrioventricular connections, the coronary sinus drains blood predominantly from the morphologically right ventricle, instead of from the left as in the normal heart. Indeed, the coronary veins follow the coronary arteries in the interventricular grooves as in the normal heart. However, while the coronary arteries develop with their respective ventricles, so that the right-sided coronary artery divides in anterior interventricular and circumflex artery and the morphologically right coronary artery is left-sided,[7,17] the coronary sinus follows the disposition of the atria. The anterior interventricular vein drains the parietal wall of the morphologically right ventricle, mostly directly into the right atrium (Thebesian veins) or in the right atrial appendage, or in 15% of the cases via a "circumflex vein" in the right atrioventricular groove before ending in the right atrium or, more rarely, in the coronary sinus.[16,18] The inferior interventricular vein (or middle cardiac vein) drains into the coronary sinus. The left ventricular veins connect directly to the right atrium or drain via the coronary sinus. And mostly, all the right ventricular veins drain in the coronary sinus directly or via the left circumflex vein in the left atrioventricular groove.[16–18] The anatomy of the coronary sinus itself is often abnormal: shorter course is frequent, oblique course above the left atrioventricular sulcus is found in 62% of cases, and the orifice is atretic in 20% of cases.[18]

The coronary venous system in our cardiac specimen

In the fetal specimen with congenitally corrected transposition that we describe here, there are two venous structures receiving all the coronary veins: a very short left-sided coronary sinus opening as usual in the right atrium with an orifice bordered by a Thebesian valve, and a right-sided venous channel that receives the main cardiac veins for the right-sided left ventricle. This right-sided venous channel is partially unroofed at its left extremity and opens in the right-sided morphologically right atrium. Therefore, the drainage of the coronary veins appears to be mirror-imaged compared to the normal heart, following the mirror-imaged disposition of the ventricles and the coronary arteries. Interestingly, it is also different, but not completely mirror-imaged, compared to the usual coronary venous drainage in congenitally corrected transposition in situs solitus.

How we should define this right-sided venous structure is worth thinking about. To the best of our knowledge, there is no report in the literature of bilateral coronary sinuses. The presence of a venous valve in this venous structure at its junction between the great cardiac vein and the coronary sinus does not necessarily indicate that this structure could be a coronary sinus.[5,16] From the embryological perspective, the coronary sinus is a remnant of the left horn of the sinus venosus, and is thus located in the left atrioventricular groove in the heart with usual atrial arrangement.[8] In our specimen, the left-sided coronary sinus, that receives the vein of Marshall, the inferior interventricular vein and the small cardiac vein, are severely hypoplastic. In addition, the connection of the caval veins to the right atrium is normal, which indicates that the right horn of the sinus venosus has been successfully incorporated to the right atrium, as usual in atrial situs solitus. Uemura et al. described in 15% of hearts with congenitally corrected transposition a "circumflex vein" draining the anterior interventricular vein to the coronary sinus.[16] The right-sided venous channel present in our heart specimen could correspond to the description of such a circumflex vein, except that it does not drain within the coronary sinus but directly in the right-sided right atrium.

This exceptional anomaly of the coronary venous drainage, never described so far, is a developmental enigma. The fact that this rare anomaly occurred in our patient in association with congenitally corrected transposition might not be a mere coincidence, as congenitally corrected transposition is known as a laterality defect occurring very early in heart development. This mirror-imaged coronary venous system could be related to a problem of laterality as well, some laterality genes like Pitx2c being involved in the development of the atria, sinus horns, and pulmonary veins.[19]

Conclusion

We describe here for the first time a fetal heart with the main coronary veins, including the great cardiac vein, connected to a right-sided venous channel located in the right atrioventricular groove, in the setting of congenitally corrected transposition in atrial situs solitus. In addition, the anatomy of the venous drainage of the heart was different compared to that usually observed in congenitally corrected transposition, and mirror-imaged compared to the normal heart in situs solitus. This very particular anatomy occurring in association with congenitally corrected transposition might be related to an additional disturbance in the laterality pathway. This underlines the importance of assessing the coronary venous anatomy in patients with congenitally corrected transposition before interventional catheterisation procedures.[17]

Institutional review board statement

The study was conducted according to the guidelines of the Declaration of Helsinki, and approved by the Institutional Review Board of AP-HP (protocol code MR-004, no 20231215170355, 15/12/2023).

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