CASE REPORT

Coronary Artery Variations in Dextrocardia with Situs Inversus: Two Cadaveric Case Reports

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Abstract

Cadaveric studies on the anatomy of the coronary artery distribution in dextrocardia with situs inversus are rarely documented in literature due to the rarity of the condition.
These two case reports describe the anatomical variations in the coronary circulation in two male cadavers with dextrocardia and situs inversus. The cadavers were those donated to the Department of Anatomy, Faculty of Medical Sciences, University of Sri Jayewardenepura, Sri Lanka for Medial education and research. There are based on two independent incidental findings of dextrocardia with situs inversus during routine student dissection over 07 years. Special focus was placed on documenting the course and patterns of the coronary artery branches to detect any variations. In the first case, a rare variation was observed as a trifurcated right-sided left coronary artery with a ramus intermedius branch. The second case revealed a right-sided left coronary artery with tetrafurcation, accompanied by myocardial bridging. Another noteworthy finding was a slight alteration in the aortic sinus configuration, deviating from the typical mirror image seen in situs inversus with dextrocardia. Morphological differences were also noted in the external diameters of the right-sided left coronary artery branches. Understanding these coronary vessel anomalies and the myocardial bridging in dextrocardia with situs inversus is critical for optimizing therapeutic and surgical interventions. This study adds evidence to the scientific literature on morphological variations that can impact clinical outcomes.

Keywords: Situs Inversus; Dextrocardia; Coronary artery

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Introduction

The embryonic development of the heart begins with bilateral splanchnic mesoderm forming two cardiogenic heart tubes that fuse cranially and at the midline during the fourth week, creating a single cardiac tube (1). Subsequent dilations, looping, and rotations form the four-chambered heart (1). Dextrocardia, first described in the 17th century, is a rare congenital anomaly (1 in 12,000) where the heart points to the right side due to abnormal leftward looping of the cardiac tube. It may occur with heterotaxy, where only some organs are mispositioned (1,2). Situs inversus, an autosomal recessive disorder (0.001–0.01%), in which there is a reversal of internal organs, as a mirror image to the normal anatomical location. It includes situs inversus with levocardia, where the heart is in the normal anatomical position, but the abdominal organs are arranged as a mirror image to the normal anatomical position; or situs inversus with dextrocardia, where both the heart and abdominal organs are positioned, as mirror images to their usual location. Isolated dextrocardia occurs with normal abdominal organ positions (situs solitus), while situs inversus totalis (SIT) presents with mirrored thoracic and abdominal viscera (3,4). Left-right axis determination occurs in week 4 via gene cascades. The Paired-like homeodomain transcription factor 2 (PITX2) gene, a member of the RIEG (Rieger Syndrome 1)/PITX homeobox family encodes a transcription factor that plays a crucial role in embryonic development, regulates the left-sidedness, and its ectopic expression can lead to laterality defects like dextrocardia or situs inversus. 5HT signaling alterations also play a role (1). Coronary arteries arise from the aortic root, with orifices in the aortic sinuses (5). The right coronary artery typically arises at a 90º angle, and the left at a lower angle (6). Coronary anomalies (0.7%–2.4%) in general, may lead to clinical symptoms or sudden death, especially in young athletes (7).

A ramus intermedius is a common variant where the left main coronary artery trifurcates instead of bifurcating, supplying the lateral and inferior walls (7). A myocardial bridge, an intramyocardial course of a coronary artery, may compress the vessel during systole. Though often asymptomatic, it may rarely cause ischemia or angina. Prevalence ranges from 0.15%–25% angiographically and 5%–86% on autopsy (7). Its presence in dextrocardia with situs inversus remains unclear (8). Advanced imaging modalities aid in early detection of coronary anomalies (7). Surgical outcomes for coronary bypass in situs inversus and dextrocardia are favorable, with prognosis similar to normal anatomy. Our study presents two cadaveric cases of situs inversus with dextrocardia, analyzing their coronary circulation to enhance understanding and management of these rare anatomical variants.

Methods

Two cadavers with Dextrocardia with Situs Inversus were detected accidently over two different independent occasions over the past seven years (case 01 – dissected in 2024 and case 02 – dissected in 2018) during routine anatomical dissections of adult donated cadavers by medical students at the Department of Anatomy, Faculty of Medical Sciences, University of Sri Jayewardenepura, Sri Lanka. The cadavers have been donated to the department with prior consent acquired before death for medical research and education. Annually, the Department of Anatomy uses 16 -18 cadavers for medical and dental (newly established from 2021 December onwards) undergraduate dissection-based teaching (9). According to available records, from year 2000 to 2024, the Department of Anatomy has utilized 388

donated cadavers for student dissections. These two case reports were found among the 388 dissected cadavers (0.51%). The age (at the time of donation of the cadaver) of each case was obtained from the available records at the Department of Anatomy. Both cases had dextrocardia with situs inversus totalis. A detailed dissection of the coronary vessels was conducted to examine the coronary circulation pattern and identify any anomalies. The variations were documented, with particular attention given to the course and patterns of the coronary arterial branching. The maximum diameter of the arteries was measured using a digital Vernier caliper by two different investigators independently and the mean value was taken. Throughout the process, special care was taken to preserve the integrity of the specimens to ensure accurate anatomical observations. These two hearts were not cut open to see the intracardiac defects. The hearts were mounted and kept in the anatomy museum display as an exhibit to be used for demonstration purposes of undergraduate medical students. Therefore, this article does not provide associated intracardiac defects.

Nomenclature

As the authors were unable to find a widely accepted nomenclature for the coronary arteries in cases of mirror image dextrocardia, the authors described the coronary arteries in this case report as the right-sided left coronary artery and the left-sided right coronary artery. This description is based on the side of the apex of the heart, similar to the approach used in a study conducted in Japan (10). However, in a study conducted by the Department of Anatomy, Faculty of Medicine, Chiang Mai University, from 2001 to 2015, which focused on the myocardial bridge in dextrocardia with situs inversus, the coronary arteries were described based on their anatomical position relative to the sides of the heart (8).

Case Report 01

A 68-year-old male cadaver (donated in 2023) showed dextrocardia, with the heart apex pointing rightward and abdominal organs reversed—liver on the left and stomach on the right—consistent with situs inversus totalis. The heart was removed to examine the coronary vasculature. A variant coronary pattern was identified, with the ramus intermedius branch arising from the right-sided left coronary artery, as previously reported.[7] The aortic cusps displayed a mirror-image arrangement (Figure 1 and Figure 3a).

The right-sided left coronary artery (diameter: 6 mm) originated from the right posterior aortic sinus and trifurcated after 13.2 mm into the anterior interventricular, circumflex, and ramus intermedius arteries. The anterior interventricular artery (4 mm) gave off a diagonal branch (2.2 mm) and descended in the groove. The circumflex (3.8 mm) circled to the back, and the ramus intermedius (2.6 mm) descended between the other two branches over the right-sided left ventricle. Coronary circulation was left dominant, as the posterior interventricular artery arose from the left-sided right coronary artery (10), which showed no variations. The left-sided right coronary artery (diameter: 4.7 mm) arose from the anterior aortic sinus, passed between the right auricle and right ventricular infundibulum, and descended in the atrioventricular groove. It gave off the conus and SA nodal arteries, followed by a right marginal artery (2.2 mm) which passed obliquely over the ventricle. It continued along the diaphragmatic surface before dividing into the posterior interventricular artery (3.9 mm) and posterolateral branch.

Case Report 02

A 74-year-old male cadaver (donated in 2017) was preserved as a teaching specimen for dextrocardia with situs inversus. Two anterior

aortic sinuses and one posterior sinus were observed, differing from the expected mirrored configuration. The left-sided right coronary artery (diameter: 5 mm) originated from the right anterior aortic sinus, coursing acutely over the infundibulum. It gave off the conus artery, from the left sided right coronary artery (diameter: 3.7 mm). Then the main artery trunk continued as the posterior interventricular and posterolateral branches. At the termination, the posterior interventricular artery anastomosed with the anterior interventricular artery of the left coronary artery (2.4 mm) (Figure 2).

The right-sided left coronary artery (6.3 mm) originated from the posterior coronary sinus (Fig. 3b), passed behind the pulmonary trunk, and emerged between the auricle and infundibulum. It divided into four branches: the largest was the anterior interventricular artery (7.1 mm), which anastomosed with the posterior interventricular artery. Other branches included D1, supplying the right ventricle; D2, which bifurcated and ended with myocardial bridging supplying the left ventricle; and the circumflex artery (3.8 mm), which passed around to the posterior side via the atrioventricular groove. A myocardial bridge was observed over the proximal third of the anterior interventricular artery. Proximal to the bridge, the vessel measured 7.1 mm—larger than typical dimensions.

Discussion

The study of coronary artery variations in Dextrocardia with Situs Inversus holds clinical significance for diagnosis and treatment. Our dissection of two such cadavers revealed key anatomical deviations in coronary circulation. Normally, the left coronary artery arises from the left posterior aortic sinus, bifurcating into the anterior interventricular and circumflex arteries, though trifurcation with a ramus intermedius occurs in 25–40% of cases (7).

In case report 01, the right-sided left coronary artery trifurcated, giving rise to a ramus intermedius, a pattern also observed in previous reports (8). In Case Report 02, this artery tetrafurcated into the right-sided left interventricular artery (7.1 mm), D1, D2, and the circumflex artery, with myocardial bridging over the proximal anterior interventricular artery. The D2 branch resembled a marginal artery, while the D1 course was unlike any previously documented. Another notable observation was the larger diameter of the anterior interventricular artery (7.1 mm) compared to the right-sided left coronary artery (6.3 mm) in Case 02. While Case 01 had a mirror image ostial pattern, Case 02 exhibited aortic root axis rotation, with altered origins of coronary arteries. Though rare, coronary artery disease occurs in dextrocardia patients at rates similar to the general population in the absence of structural abnormalities (11). However, coronary bypass surgery is more technically demanding in these individuals (11). Myocardial bridges (MB)—intramyocardial courses of coronary arteries—are typically benign and detected in 23–60% of autopsies. In some cases, they can cause systolic compression, delayed diastolic filling, and even ischemia (12). Associations with arrhythmias, cardiomyopathy, and sudden cardiac death have also been reported (12). MBs cause proximal arsitusterial dilation and intimal thickening (12). Few studies have addressed MB presence in dextrocardia with situs inversus (13).

Our cases emphasize the need for careful anatomical evaluation in such rare presentations. Importantly, this report adds novel morphometric data on coronary arteries in dextrocardia with situs inversus, although a limitation is that both specimens were formalin-fixed and not fresh.

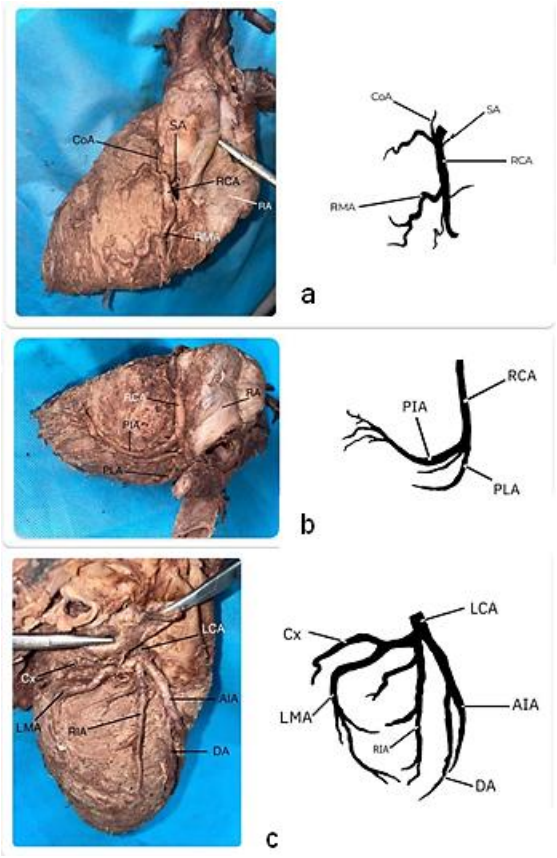

Figure 1: Case report 01: Anterior (a), Inferior (b), and Posterior (c) views of the heart and schematic representation of the coronary arteries and their branches. RCA: (Left sided) Right Coronary Artery; SA: SA Nodal Artery; RMA: Right Marginal Artery; CoA: Conus Artery; RA: Right Atrium; PIA: Posterior Interventricular Artery; PLA: Posterolateral Branch; LCA: (right sided) Left Coronary Artery; AIA: Anterior Interventricular Artery; DA: Diagonal Artery; Cx: Circumflex Artery; LMA: Left Marginal Artery; RIA: Ramus Intermedius Artery

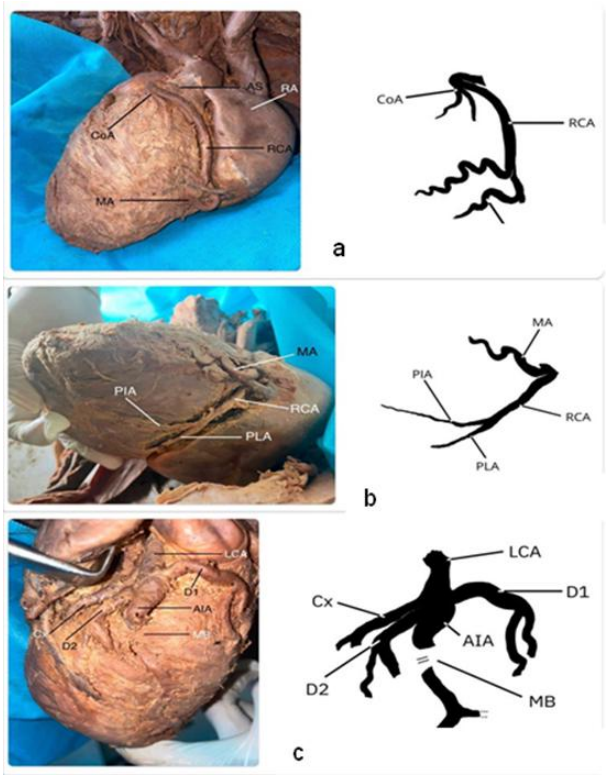

Figure 2: Case report 02Anterior(a), Inferior(b), and Posterior(c) views of dextrocardia and schematic presentation of the coronary arteries and their branches. RCA: (Left sided) Right Coronary Artery; AS: Aortic Sinus (Right Anterior); MA: Right Marginal Artery; CoA: Conus Artery; RA: Right Atrium; PIA: Posterior Interventricular Artery; PLA: Posterolateral Branch; LCA: (right sided) Left Coronary Artery; AIA: Anterior Interventricular Artery; Cx: Circumflex Artery; MB: Myocardial Bridging; D1: Branch; D2: Branch

Conclusion

Our findings contribute to the limited but growing knowledge on coronary artery anatomy in dextrocardia with situs inversus. The presence of unique variations, such as the ramus intermedius, tetrafuracation of right sided left coronary artery and myocardial bridging, along with alterations in aortic sinus configuration, underscores the complexity of this congenital condition. Future research and clinical studies are needed to further elucidate these variations and improve the management strategies for patients with dextrocardia and situs inversus.

The detailed anatomy knowledge in coronary arteries in Dextrocardia with Situs Inversus variations and their variations may significantly lower the complication risks and improve patient management.

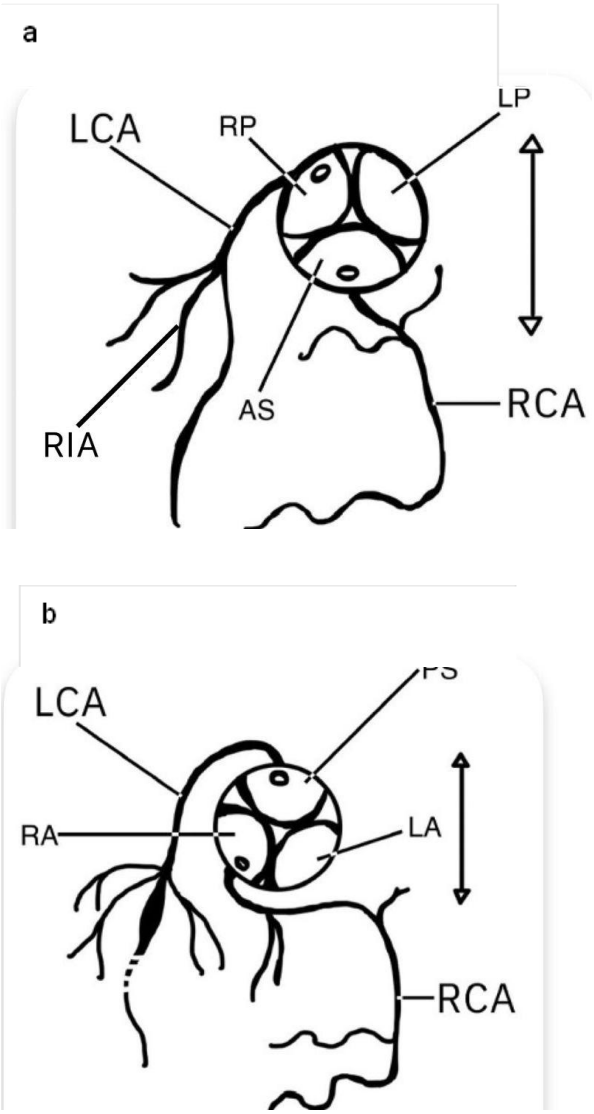

Figure 3: Illustration of coronary artery distribution of case 01(a) and case 02 (b)
RCA: (Left sided) Right Coronary Artery; LCA: (right sided) Left Coronary Artery; AS: Anterior Aortic sinus; LP: Left posterior Aortic Sinus; RP: Right Posterior Aortic sinus; PS: Posterior Aortic Sinus; LA: Left Anterior Aortic Sinus; RA: Right Anterior Aortic sinus; RIA: Ramus Intermedius Artery

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