

Dextrocardia and Situs Inversus with Incomplete Inversion: a Case Report

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Abstract : Situs inversus totalis is a very rare anatomical abnormality, which is characterized by dextrocardia and “mirror image” distribution of abdominal organs. Dextrocardia and situs inversus have important surgical implications due to altered anatomy. The aim of this study was to describe the anatomical structures of the situs inversus with dextrocardia in Korean.

Presence of situs inversus with dextrocardia was confirmed in a 72-year-old female cadaver during dissection in a gross anatomy course. It was observed by focusing on the positions of the thoracoabdominal structures.

In this cadaver, clearly inverted morpholoiges existed in the structures of the thoracoabdomen except the kidneys. The left-sided kidney was located at a slightly higher level than the right-sided kidney although the liver was located on the left side.

We reported herein a case of dextrocardia and situs inversus with incomplete inversion, which will provide useful information for accurate diagnosis and relevant surgical procedures.

Keywords : Dextrocardia, Incomplete inversion, Situs inversus

Introduction

Dextrocardia is defined as the right-sided embryologic development of the heart, with most of the cardiac mass positioned in the right hemithorax and a base-to apex pointing to the right [1]. Situs inversus totalis is a very rare anatomical abnormality occurring about 0.01% worldwide, which is characterized by dextrocardia and “mirror image” distribution of abdominal organs [2]. In pure cases the liver is on the left and the stomach, spleen and heart are on the right. In heterotaxy the abdominal viscera may be inverted, whereas the thoracic contents may be normal, or vice versa [3]. Dextrocardia may occur as situs solitus (normal viscer-

atrial arrangement), situs inversus (‘mirror image’ of normal visceratrial arrangement), or situs ambiguous (visceratrial isomerism) [1]. In situs inversus with dextrocardia, these individuals usually have a normal life expectancy, and the heart is structurally normal in 90~95% of cases, in contrast to dextroversion (dextrocardia with situs solitus), which has a high incidence of structural cardiac defects [4,5]. In complete situs inversus, the etiologic nature of transposition of the viscera is obscure, but this condition apparently does not influence normal health or life expectancy [6].

Dextrocardia and situs inversus have important surgical implications due to altered anatomy. Reversal of normal anatomy has obvious clinical implications, a typical example being a patient with mirror-image arrangements and appendicitis, who presents with an acute left lower quadrant pain [7]. The complex anatomy and altered fluoroscopic orientation can pose considerable challenge to transvenous permanent pacemaker implantation in dextrocardia [1]. In addition, the altered spatial orientation of the cardiac

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structures can present a challenge in patch closure of the ventricular septal defect. The surgeon approaches the ventricular septal defect at a different angle in a patient with dextrocardia than in a patient with levocardia [8].

The aim of this study was to describe the anatomical structures of situs inversus with dextrocardia in Korean, and thereby provides useful information for accurate diagnosis and relevant surgical procedures.

Case Report

Presence of situs inversus with dextrocardia was confirmed in a 72-year-old female cadaver during dissection in a gross anatomy course. All structures of the thorax were mirror-image in their distribution. The left-sided recurrent laryngeal nerve looped under the left-sided subclavian

artery and the right-sided recurrent laryngeal nerve looped under the right-sided aortic arch. The left-sided brachiocephalic vein descended almost vertically to join the right-sided brachiocephalic vein. The right-sided brachiocephalic vein descended obliquely to the left. And it joined the left-sided brachiocephalic vein to form the superior vena cava. The heart was right-sided in the thorax and the apex of the heart pointed to the right (Fig. 1). The left-sided atrium received the superior and inferior vena cavae. The ascending aorta began at the base of the right-sided ventricle and it ascended to be the aortic arch. From the convex aspect of the arch, three branches arose: the brachiocephalic trunk, right common carotid and right subclavian arteries. The pulmonary trunk arose from the base of the left-sided ventricle. The pulmonary trunk crossed the ventral side of the ascending aorta. After removal of the heart, the position of the sectioned large vessels at their cardiac ori-

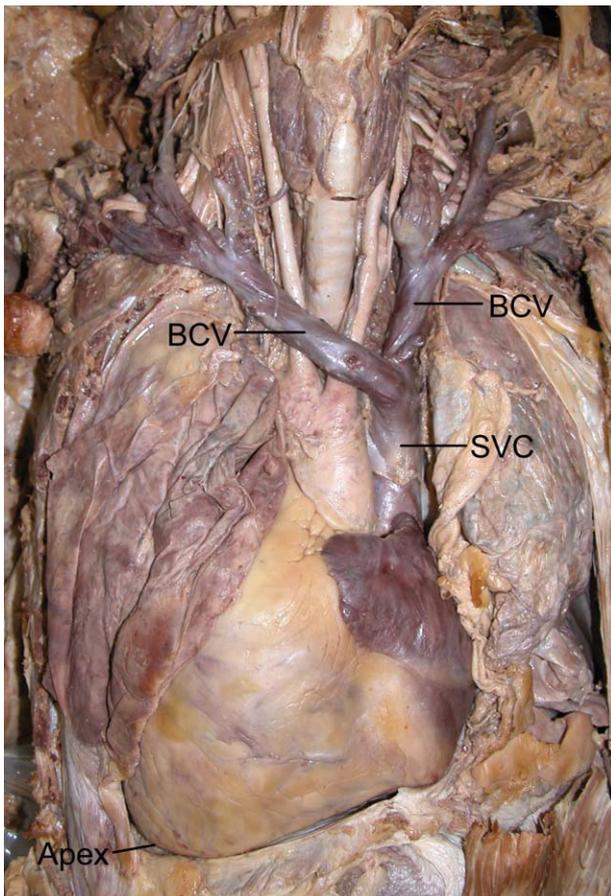


Fig. 1. A photograph showing the dextrocardia and the left-sided brachiocephalic vein (BCV) descending almost vertically and the right-sided BCV descending obliquely to the left. SVC, superior vena cava.

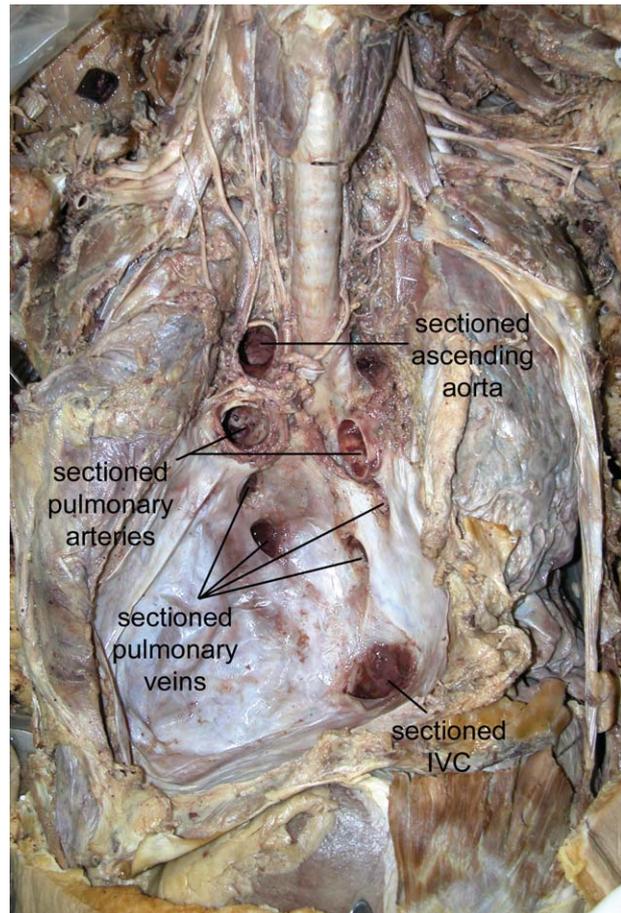


Fig. 2. A photograph showing the inverted position of the sectioned large vessels at their cardiac origin in the interior of the serosal pericardial sac. IVC, inferior vena cava.

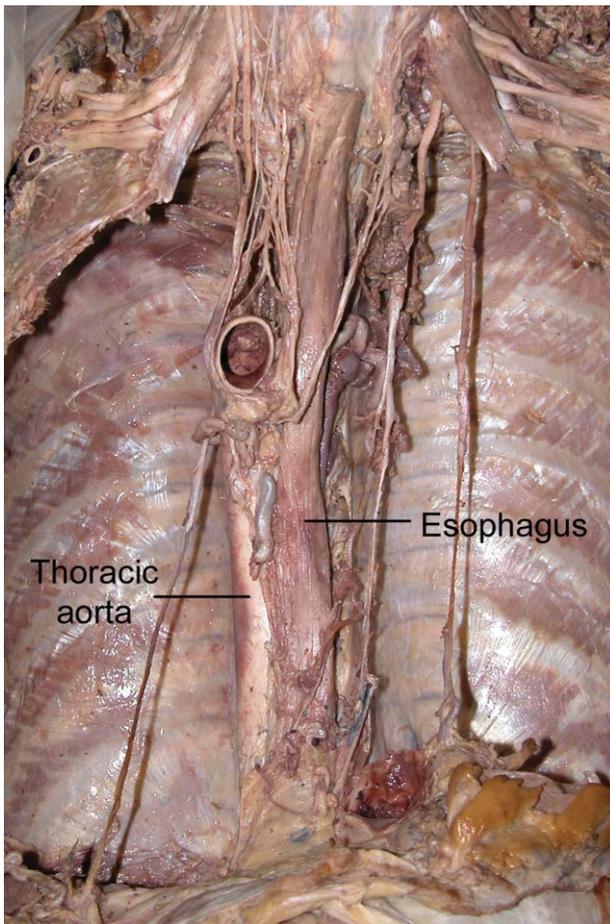


Fig. 3. The thoracic esophagus descending in the mediastinum, along the left side of the descending thoracic aorta.

gin was confirmed to be inverted in the interior of the serosal pericardial sac (Fig. 2). The thoracic esophagus descended in the posterior mediastinum, along the left side of the descending thoracic aorta (Fig. 3). The esophagus was posterior to the trachea as normal position. The left-sided principal bronchus was wider, shorter and more vertical than the right. The left-sided lung had three lobes and the right-sided lung had two lobes with the lingula. The accessory hemiazygos vein and the hemiazygos vein lay to the right of the vertebral column. The azygos vein lay leftward and anterior to the bodies of the lower thoracic vertebrae. The stomach and spleen were on the right side whereas the liver on the left side. The accessory spleen or asplenia was not observed in this cadaver. The cecum and the vermiform appendix were located in the left iliac fossa (Fig. 4). The kidney did not exhibit inverted morphology. Although the liver was located on the left side, the left-sided kidney was

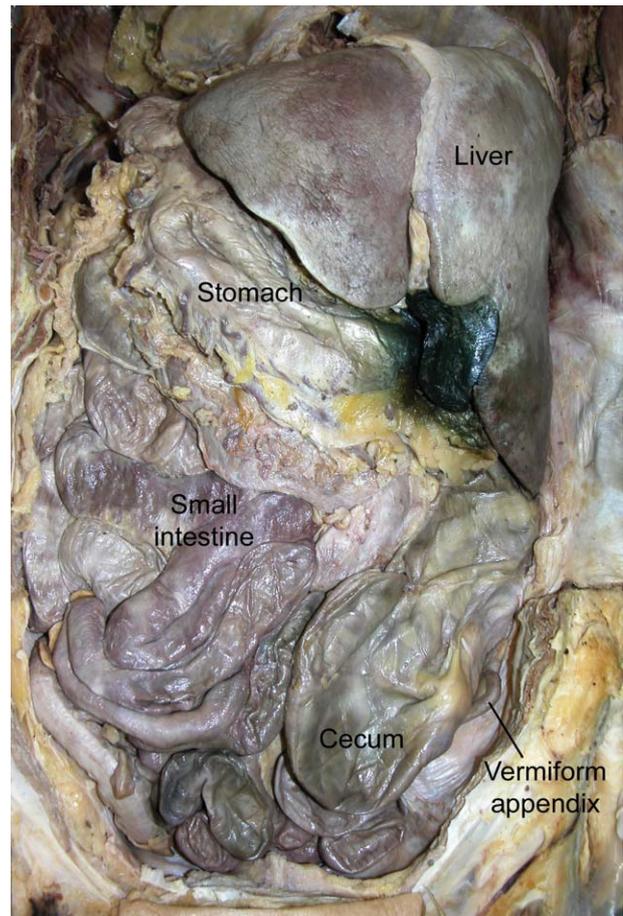


Fig. 4. Inverted position in the structures of the abdomen. The stomach is on the right side whereas the liver on the left side. The cecum and the vermiform appendix are located in the left iliac fossa.

located at a slightly higher level than the right-sided kidney (Fig. 5). The renal veins lay anterior to the renal arteries and the renal veins entered the inferior vena cava. The inferior vena cava was located to the left side of the abdominal aorta.

Discussion

This case report described about dextrocardia and situs inversus with incomplete inversion. In this cadaver, clearly inverted morphologies existed in the structures of the thoracoabdomen except the kidneys.

All vertebrates have characteristics asymmetries along the left/right axis. The positioning of asymmetric visceral organs is highly conserved evolutionarily and disruptions in left/right patterning can lead to several morphological

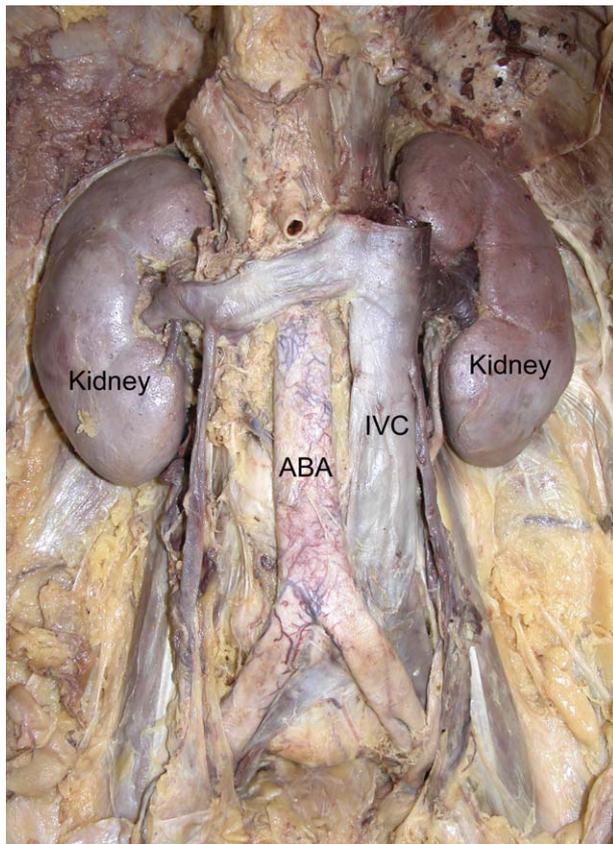


Fig. 5. A photograph showing the normal position of the kidneys in situs inversus. Although the liver is located on the left side, the left-sided kidney is located at a slightly superior level than the right-sided kidney. ABA, abdominal aorta; IVC, inferior vena cava.

defects, demonstrating the importance of regulation of left/right development asymmetries [9]. Several studies suggest that left-right asymmetry defects are likely to be due to genetic abnormalities in the left, nodal, i.v., *HAND*, *ZIC3*, *Shh*, *ACVR2B* and/or *Pitx3* genes [4]. It has been stated that multiple molecules or genes determine laterality and act in a region- or organ-specific manner [10].

Several studies reported about situs inversus totalis that all thoracic and abdominal viscera were completely transposed [2,4,8,11-16]. However, Mano et al. (2006) [17] reported that situs inversus with unclearly inverted morphology, neutral laterality (i.e. positioned midsagittally) or even usual morphology in the posterior midsagittal and retroperitoneal structures. One of them is the left-sided kidney situated at a slightly higher level than the right-sided one. They stated that each organ seems to have its own cue to determine its side.

In the present study, the left-sided kidney was located

superior to the right-sided kidney although the liver was located on the left side. Both kidneys appeared to undergo normal cranial migration and rotation during development. The position of the right kidney is usually a little bit inferior than the left kidney, reflecting its relationship to the liver [7]. Therefore it is thought that regardless of the inversion of the liver, the kidney underwent normal development due to a gene or molecule for kidney asymmetry.

Situs inversus totalis might be associated with multiple abnormalities such as accessory spleen, asplenia, intestinal malrotation and so on [18]. In this cadaver, the spleen was normal in the inverted position and there was no intestinal malrotation.

In spite of situs inversus with dextrocardia, some structures can be located in the normal position as incomplete inversion. Therefore it should be recognized for accurate diagnosis and relevant surgical procedures.

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오른심장증과 불완전한 좌우바뀔증의 증례보고

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간추림 : 좌우바뀔증은 매우 드물게 나타나는 비정상적 형태로, 오른심장증과 함께 배의 장기가 거울에 비친 모양으로 좌우가 완전히 바뀌어져 있는 것이 특징이다. 오른심장증과 좌우바뀔증은 뒤바뀐 해부학적 구조를 하고 있으므로 중요한 임상적 의미를 가진다. 이 연구의 목적은 오른심장증과 좌우바뀔증의 해부학적 구조들을 관찰하는 데 있다.

오른심장증과 좌우바뀔증은 72살 여자 시신에서 확인되었다. 가슴과 배의 장기 위치에 초점을 맞추어 관찰하였다.

이 시신에서는 콩팥을 제외하고는 가슴과 배의 구조들이 완전히 좌우 자리가 바뀐 형태였다. 간은 왼쪽에 위치하였지만, 왼쪽에 위치한 콩팥이 오른쪽에 위치한 콩팥보다 위쪽에 위치하였다.

이 결과는 오른심장증과 불완전한 좌우바뀔증에 대한 사례를 보고함으로써, 정확한 진단 및 관련 수술에 유용한 자료를 제공할 것으로 기대된다.

찾아보기 낱말 : 오른심장증, 불완전한 바뀔, 좌우바뀔증