# Anatomical comparison of a defect in the hepatic segment of the inferior vena cava with the normal azygos system

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#### SUMMARY

During anatomical practice between 2001-2002 at the Tokyo Women's Medical University, a defect in the hepatic segment of the inferior vena cava was encountered in a 94-year-old male cadaver. Although this type of defect has often been observed clinically using imaging diagnosis, the topological changes in the small vessels and surrounding structures have not been described previously.

Here, the topological changes in small vessels and surrounding structures, in addition to the morphology of a defect in the hepatic segment of the inferior vena cava, are described in detail. Normal variations in the venous system were also examined using 26 human cadavers for comparison. The following results and interpretations were obtained:

The thoracic portion of the anomalous vein originated from the azygos system, judging from the topological relationships among the main venous trunk, small vessels, and the autonomic nerves.

Although a close relationship between the inferior vena cava and the azygos system was recognized, the development of each venous system should be regarded as independent, based on the normal variations observed in 26 human cadavers and the currently available medical literature.

The boundary between the cardinal vein (inferior vena cava) and the azygos system might be the lower border of the renal vein, since the renal vein drained into the ventral aspect of the inferior vena cava and the left renal artery ran superficially across the abdominal portion of the anomalous vein.

Examination of topological changes in the surrounding structures may provide important clues regarding the morphogenesis of defects in the hepatic segment of the inferior vena cava and normal venous systems.

**Key words:** Anomalous inferior vena cava – Azygos system – Humans – Gross anatomy – Morphogenesis – Hepatic segment

#### INTRODUCTION

A defect in the hepatic segment (d-hs) of the inferior vena cava (IVC) was observed in a male cadaver during anatomical practice at the Tokyo Women's Medical University School of Medicine. Although d-hs of the IVC is a very rare venous anomaly, advances in imaging diagnosis have led to an increase in the frequency of its clinical observation. Among clinicians, this anomaly is regarded as a defect of the IVC (Sato and Akita: 2000). The anomaly and its morphogenesis have been clearly described (Arey, 1965; Patten, 1968; Moore and Persaud, 1977; Larsen, 1997; Sadler, 2000). However, only a few anatomical cases have been reported (Kollmann, 1893; Dwight, 1900).

Recent clinical reports of this defect only contain sectional data from imaging diagnosis and do not discuss the topological changes in

Submitted: May 1, 2004 Accepted: June 18, 2004 Correspondence to: Tomokazu Kawashima. Department of Anatomy, School of Medicine, Tokyo Women´s Medical University, 8-1 Kawada-Cho, Shinjuku-ku, Tokyo 162-8666, Japan. Phone and Fax: +81-3-5269-7405. E-mail: kawa@research.twmu.ac.jp small vessels and surrounding structures that are associated with the defect. Furthermore, clinicians usually only refer to the hemi-/ azygos continuation by application of traditional embryology (Anderson et al., 1961; Muelheims and Mudd, 1962; Tsuzuki et al., 1964; Chapman and Crouch, 1967; Sakakibara et al., 1969; Nagashima et al., 1972; Chuang et al., 1974; Ohi, 1980; Pomeranz and Anthony, 1986; Mairesse et al., 1995; Vermeulen and Urk, 1996; Miyashita et al., 2000).

Here, the topography of the small vessels and surrounding structures associated with a dhs of IVC is described, and the boundary between the IVC and the azygos system (Az-s) is discussed using results obtained from the cadaver with the d-hs of IVC and 26 additional normal human cadavers.

#### MATERIALS AND METHODS

During anatomical practice dissection between 2001-2002 at the Tokyo Women's Medical University School of Medicine, a d-hs of the IVC was observed in a 94-year-old male who had died of pneumonitis. We also investigated the origin, course and drainage of the Az-s using 26 normal human cadavers. Each observation was made macroscopically, using a stereomicroscope if necessary.

#### RESULTS

#### 1. HEPATIC SEGMENT DEFECT OF THE INFERIOR VENA CAVA

In the cadaver with the d-hs of IVC, the abdominal aorta was situated on the right side, and the abdominal portion of the anomalous vein was located on the left side (Fig. 1).

#### Venous system

The abdominal portion of the anomalous vein was composed of the drainage of both common iliac veins. The abdominal portion of the anomalous vein ascended left to the vertebrate bodies, ran through the aortic hiatus along the aorta, and entered the thoracic cavity. Here, the anomalous vein was initially situated at the dorsolateral (left) side of the aorta, then gradually shifted to the medial (right) side, and finally drained into the dorsal aspect of the superior vena cava (SVC).

Both renal veins drained into the ventral aspect of the abdominal portion of the anomalous vein.

Although the right renal vein ran ventral (superficial) to the aorta, the left renal artery crossed over the upper portion of the abdominal portion of the anomalous vein, compared to the position of the left renal vein (RV, shown by the star in Fig. 1). Both testicular veins drained into each RV on the same side.



Fig. 1.- Photograph showing the hepatic segment defect of the inferior vena cava. The abdominal aorta was situated on the right side, and the abdominal portion of the anomalous vein was situated on the left side shown by the single asterisk. This vein ran through the aortic hiatus along the aorta and was located on the dorso-lateral (left) side of the aorta after entering the thoracic cavity; then, it gradually shifted to the right side and drained into the superior vena cava (shown by the two asterisks). The star shows the left renal artery running ventral to the anomalous vein. A, common venous trunk; Ao, aorta, B, common trunk B; D, diaphragm; E, esophagus; IC, iliac artery; IM, inferior mesenteric artery; LRV, left renal vein; Ph, phrenic nerve; RRV, right renal vein; SM, superior mesenteric artery; SVC, superior vena cava.



**Fig. 2.-** Diagram of the defect in the hepatic segment of the inferior vena cava and the surrounding structures shown in Fig. 1. The renal vein opened into the ventral aspect of the abdominal portion of the anomalous vein, and the left renal artery (arrowheads) ran superficially across the upper portion of the anomalous vein. The intercostal arteries ran from a ventro-medial position to a dorso-lateral position against the anomalous vein.

#### Lumbar veins and arteries

Each fifth lumbar vein drained into the ipsilateral common iliac vein, and the majority of the right fourth lumbar vein drained directly into the abdominal portion of the anomalous vein. The right first to third lumbar veins, a portion of the right fourth lumbar veins, and the left second to fourth lumbar veins became one venous trunk and drained into the abdominal portion of the anomalous vein from a dorsal aspect.

On the other hand, the right and left first to fifth lumbar arteries branched segmentally. The left lumbar arteries ran dorsally to the left lumbar veins and the abdominal portion of the anomalous vein.

#### Intercostal veins and arteries

The venous trunk (A), comprising the right twelfth to eighth intercostal veins in the lower portion, and the venous trunk (B), comparing the left second to fifth intercostal veins, the right seventh to fifth and the right third to fourth common venous trunk, and the left twelfth to sixth intercostal veins drained directly into the thoracic portion of the anomalous vein.

The right second to twelfth intercostal arteries branched from the thoracic aorta and ran from the ventro-medial side to the dorso-lateral side against the venous trunk (A) and the thoracic portion of the anomalous vein. The left seventh to twelfth intercostal arteries ran from the ventromedial side to the dorso-lateral side against the lower thoracic portion of the anomalous vein.

## *Liver and intrahepatic distributions of Glisson's sheath and hepatic veins*

The hepatic artery, hepatic duct and the portal vein entered from the *porta hepatis*, as usual (Fig. 3). Glisson's sheath bifurcations from the segment 1 to 9 branches were also observed. Although the right, middle, and left hepatic veins were observed, these three hepatic veins gathered as one trunk and drained directly into the right atrium because of the d-hs of the IVC. The small branches of the dorsal segments drained directly into the three hepatic veins (Fig. 4). Any small branches between the liver and the right inferior phrenic vein were not clearly observed.



Fig. 3.- Photographs showing the shape of the liver. (A) Ventral view. (B) Dorsal view. (C) Cranial view. The hepatic segment of the inferior vena cava is not visible in the dorsal portion. An asterisk shows the trunk of three hepatic veins. C: caudate lobe, GB: gall bladder, L: left lobe, PV: portal vein, Q: quadrate lobe, R: right lobe, RL: round ligament.





Fig. 4.- Diagrams showing the intrahepatic distribution of Glisson's sheath and the three hepatic veins. The numbers indicate the hepatic segment. (A) Ventral view. (B) Dorsal view. The hepatic segment of the inferior vena cava is not visible in the liver. CBD, common biliary duct; HA, hepatic artery; LHV: left hepatic vein; MHV, middle hepatic vein; RHV, right hepatic vein; PV, portal vein; S1-9, hepatic segment 1-9.

#### Heart

Although the heart exhibited a right coronary dominance, characterized by a well-developed right coronary artery with distal distribution after the posterior interventricular branch, no anomalies were observed in the coronary blood system (Figs. 5 and 6). On the inner surface of the heart, the ostium of the coronary sinus and the *fossa* ovalis appeared normal (Fig. 5C).

#### Sympathetic system and surrounding structures

Both sympathetic trunks were situated laterally to the anomalous vein. The right greater splanchnic nerve ran superficially across the



Fig. 5.- Photographs showing the morphology of the heart. (A) Ventral view. (B) Dorsal view. (C) Inner surface of the right atrium and ventricle. The asterisk shows the drainage portion of the trunk of the three hepatic veins. AI: anterior interventricular branch, Ao: Aorta, C: circumflex branch, AV: anterior cardiac vein, CS: coronary sinus, FO: fossa ovalis, GV: great cardiac vein, MV: middle cardiac vein, OS: ostium of coronary sinus, OV: oblique vein of the left atrium, PI: posterior interventricular branch, PT: pulmonary trunk, PV: pulmonary vein, RC: right coronary artery, SVC: superior vena cava.



Fig. 6.- Diagrams of the heart shown in Fig. 5. (A) Ventral view. (B) Dorsal view.

venous trunk (A), and the left greater splanchnic nerve ran superficially across the thoracic anomalous vein (Fig. 7).

Three lobes in the right lung and two lobes in the left lung were observed as usual. However, the spleen was composed of five lobes.



Fig. 7.- Photograph showing the positional relationships between the left greater splanchnic nerve and the anomalous vein (\*). The left greater splanchnic nerve ran superficially across the anomalous vein. D: diaphragm, GS: greater splanchnic nerve; ST, sympathetic trunk.

2. VARIETY IN THE ORIGIN, COURSE AND DRAINAGE OF THE AZYGOS SYSTEM

The normal morphology of the Az-s was investigated using 26 human cadavers. Since the morphology of the Az-s was difficult to classify, only the origin, course, and drainage points of the Az-s will be described.

#### Origin

Regarding the origin of the Az-s, the connection between the IVC/RV and the Az-s was examined to obtain information about the boundary between the IVC/RV and the Az-s. The results are as follows; the connection between the IVC/RV and the Az-s was absent on 31 sides (overall 59.6%; right, 12 sides, 46.2%; left, 19 sides, 73.1%) and present on 21 sides (overall 40.4%; right, 14 sides, 53.8%; left, 7 sides, 26.9%). The participants of the Az-s connection were the IVC on 4 sides with only the azygos vein, the RV on 3 sides with only the hemiazygos vein, the posterior RV on 2 sides with the hemiazygos vein, and the common trunk of the ascending lumbar vein on 12 sides (right, 10 sides; left, 2sides). Examples of the normal connections between the azygos vein and the IVC are shown in the Figs. 8-A and 9-A, between the hemi-azygos vein and the left RV in Figs. 8-B and 9-B, and between the hemiazygos vein and posterior venous ring in the Fig. 9-C.

#### Course

Variations in the course of the Az-s, such as a separate Az-s course or joining of the Az-s with the hemi- / azygos veins in the lower or upper region, were observed. Importantly, the majority



Fig. 8.- Photographs showing normal variations of the azygos system. (A): The connection between the azygos vein and the inferior vena cava is shown by the arrowheads. (B): The connection between the azygos vein and the left renal vein is shown by the arrowheads. (C): The venous ring in the course of the azygos system is shown by the asterisk. Az, azygos vein; GS, greater splanchnic nerve; hAz, hemiazygos vein; IVC, inferior vena cava; LK, left kidney; LRV, left renal vein; SR, suprarenal gland; Td, thoracic duct.



Fig. 9.- Diagrams showing the various connections between the azygos system and the IVC/RV. (A): The connections between the IVC and the azygos/hemiazygos veins. (B): The connection between the hemiazygos vein and the left renal vein is indicated by the asterisks. (C): The connection between the hemiazygos vein and the posterior portion of the circum-aortic venous ring is highlighted by the asterisks. In normal cases, the intercostal arteries run from the ventro-medial to the dorso-lateral sides to the Az-s, and the greater splanchnic nerve always run superficially across the Az-s.

of the intercostal arteries ran across the Az-s from a ventro-medial position to a dorso-lateral position, as shown in fig. 9.

Throughout the course of the Az-s, the number of venous rings (Fig. 8-C) between the sympathetic trunks was counted to evaluate the complexity of the Az-s. Venous rings were observed in 13 of the 26 cases (50.0%).

#### Drainage

Regarding the drainage points, all of the intercostal veins except for the *vena intercostalis suprema* were collected by the Az-s and drained into the SVC in 20 cases (76.9%; Figs. 9B and 9C), whereas the azygos vein drained into the SVC and the hemiazygos drained into the left brachiocephalic vein in 6 cases (23.1%; Fig. 9-A).

#### DISCUSSION

Although d-hs of the IVC is a very rare venous anomaly, advances in imaging diagnosis have led to an increase in the frequency of its clinical observation. Among clinicians and anatomists, this anomaly is regarded as a defect of the IVC (Sato and Akita, 2000). The anomaly and its morphogenesis have been described in many embryological texts (Arey, 1965; Patten, 1968; Moore and Persaud, 1977; Larsen, 1997; Sadler, 2000). However, only a few anatomical cases have been reported (Kollmann, 1893; Dwight, 1900).

Recent clinical reports of this defect only contain sectional data from imaging diagnosis and do not discuss the topological changes in small vessels and surrounding structures that are associated with the defect. Furthermore, many clinicians only refer to the hemi-/ azygos continuation by application of traditional embryological theory. According to traditional embryological theory, our case would be classified as a hemiazygos continuation.

In this study, the morphology and topography of the small vessels and surrounding structures in addition to the d-hs of the IVC were examined; the normal Az-s, and conventional descriptions were reviewed. This strategy provided important clues regarding the morphogenesis of the venous system anomaly.

#### Venous system

Although much anatomical research on the Az-s has been reported (Adachi, 1940; Aoki et al., 1943; Batson, 1940; Falla et al., 1963; Hojo, 1979; Kino, 1939; Mutel et al., 1923; Murakami et al., 1980; Nathan, 1960; Rokutanda, 1959; Seib, 1934; Stauffer, 1951), the numerous Az-s variations are still difficult to classify. In the present observations, the number of classifications would be the same as the number of subjects if all important factors in the conventional classification system plus the intercostal arterial position in relation to the IVC and Az-s, etc. were considered. Therefore, only the initial part of the connection, the course, and the drainage point of the Az-s will be discussed here. The present results showed a variety of Az-s and a close relationship between the Az-s and the IVC/RV.

Okamoto (1990) explained the relationship between the venous system and the renal venous system using detailed observations of 270 human cadavers. Among his observations, some interesting cases with relevance to morphogenesis of the venous system were described. Okamoto observed a high frequency of a posterior RV (right in 82 cases, left in 90 cases), compared with the conventional frequency.

The present findings, Okamoto's findings (1990), conventional descriptions, and the venous drainage from the IVC to the Az-s when co-lateral pathways are created by hepatopathy suggest a close relationship between the IVC and the Az-s.

According to Yano and Sato (1980), two theories on the morphogenesis of the venous system have been proposed. One theory regards both the azygos and the caval systems as being derived from the cardinal system (Hochsttler, 1893, 1906; Lewis, 1902; Huntington- McClure-Butler school, 1903, 1906, 1907, 1920, 1925, 1927, 1929, 1950; van Gelderen, 1927; Aray, 1965). The other theory also regards the caval system as being derived from the cardinal system. However, this second theory considers that the Az-s originate from the external vertebral venous plexus, in a manner that is relatively independent of the caval system's derivation from the caval veins (Regan, 1919, 1927; Gladstone, 1929; Seib, 1934; Hamilton and Mossman, 1972; Ura-School (Abe, 1960; Inui, 1960; Miura, 1960); Yano and Sato, 1980). The difference between these two theories is an important issue.

Although the morphogenesis of the venous system cannot be clarified using macroscopic observations, the boundary between the IVC and the Az-s may provide an important clue regarding the morphogenesis of the d-hs of the IVC.

#### Morphogenesis of the defect in the hepatic segment of the IVC based on a macroscopic observations

While considering the morphogenesis of the venous system, the intercostalis veins were observed to drain segmentally into the anomalous vein; the intercostalis arteries ran from a ventro-medial position to a dorso-lateral position, relative to the anomalous vein, and the greater splanchnic nerve ran superficially across the anomalous vein in the thoracic region. These observations led to the speculation that the thoracic portion of the anomalous vein may have derived from the Az-s.

We do not know whether the hepatic segment anomaly was caused by a congenital defect or may be a secondary disappearance. However, the boundary between the cardinal vein (main anlage of the IVC) and the Az-s would be the lower border of the renal vein, because the RV drained into the ventral aspect of the IVC and the left renal artery ran superficially across the abdominal portion of the anomalous vein.

The topological data on the small vessels and surrounding structures in the present case may provide important clues regarding the morphogenesis of the d-hs of the IVC and the normal venous system, from the viewpoint of macroscopic anatomy.

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