

“PREURETERIC VENACAVA” EMBRYOLOGICAL BASIS AND ITS CLINICAL IMPORTANCE

Ganesh Elumalai and Anto Sicily Norbert

Department of Embryology, College of Medicine, Texila American University, South America.

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ABSTRACT

Pre-ureteric vena cava is a rare congenital disorder, usually presenting clinically with hydronephrosis and an “S or fishhook” deformity of the ureter at the lumbar vertebrae three and four. Unusual presentations do occur and can lead to misdiagnoses. Though it is a congenital disorder, patients do not normally present with symptoms until the 30 years of life with various complains, results due to back pressure changes lead to hydronephrosis (water in the kidney).

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Introduction

Retro-caval ureter also called as the circumcaval ureter or pre-ureteral vena cava. It is a rare congenital anomaly. The ureter passes posterior to the inferior vena cava. The ureter classically courses medially behind the inferior vena cava covering around it and then passes laterally in front of it then course distally to the urinary bladder.[1,2,3] Though it is a congenital disorder, patients do not normally present with symptoms until the 30 years of life with various complains, results due to back pressure changes leads to hydronephrosis [water inside kidney]. [4,5,6,7]The hydronephrosis may be due to curving of the ureter, a ureteric segment that is a dynamic or compression of inferior vena cava. It was finally considered as an aberration in ureteric development current studies in embryology have led to being considered as an aberration in the development of the inferior vena cava. Hence it is being suggested that the anomaly is referred to as a pre-ureteral vena cava.[8,9,10,11] Although this anomaly is known to urologists as a circumcaval or retrocaval ureter terms that are anatomically descriptive but it misleads about development—it is not the result of an abnormality in ureteral development, but it is an anomaly in the development of the inferior vena cava. The former embryologic theory was that normally the inferior vena cava develops below the kidney from the posterior cardinal, subcardinal, and supracardinal veins, which must undergo development, anastomosis, and regression to become the inferior vena cava and the azygos venous system.[12,13,14,15] Normally, the right sub cardinal vein forms the pre-renal inferior vena cava, the sub cardinal-supracardinal anastomosis forms the renal segment, and the right supracardinal vein forms the post-renal inferior vena cava. Typically, the retrocaval ureter etiology is assumed to be an abnormal embryologic development of the inferior vena cava as a result of atrophy [degeneration of cell] failure of the right sub cardinal vein in the lumbar portion.[16,17] When the inferior vena cava is formed by a sub cardinal vein that lies

ventral to the ureter, the ureter will develop in a “retrocaval” position[18,19,20].

The disorder predominantly involves the right ureter, which typically deviates behind (dorsal to) the inferior vena cava, winds about and crosses in front of the vena cava from a medial to a sidewise direction, and resumes a normal course, distally, to the bladder. A retrocaval ureter is a rare entity and its exact prevalence is still unknown.[21,22,23,24,25] Although the lesion is congenital, symptoms usually present in the third to fourth decade of life. In most patients who become symptomatic, symptoms are due to ureteral blockage and consequent hydronephrosis.[26,27,28] The patients usually present with right flank pain and discomfort. However, patients can also complain of urinary tract infections [UTI], gross haematuria, urolithiasis, or fever. In contrast, the hydronephrosis may be calm and fully asymptomatic. The main causes of hydronephrosis are compression by the psoas muscle, spinal column, and the IVC. In patients with a circumcaval ureter, ureteral obstruction is a frequent but not unavoidable complication.[29,30,31,32,33]

Incidence

An anomalous development of infrarenal inferior vena cava leads to circumcaval ureter or pre-ureteral vena cava, a rare congenital disorder with an autopsy incidence of 0.9 per 1000.[34] We present a case of pre-ureteric vena cava and review the literature with a special emphasis on the paradigm shift seen during the last decade, in diagnostic and management strategies.

Ontogenesis for normal development of IVC and Ureter

At the fifth week of development, the ureteric bud arises as a diverticulum from the mesonephric (Wolffian) duct. The bud grows laterally and invades the center of metanephric blastema, the primordial renal tissue. The touching of these two tissues causes the change in the bud and the metanephros.[35,36] The metanephric blastema forms glomeruli, proximal tubules, and distal tubules.

The ureteric bud divide and branches forming the renal pelvis, infundibulum, calyces, and collecting tubules which will provide a conduit for urine drainage in the kidney.[37] This process is known as the induction of the kidney, from 28 to 35 days of development, the ureter is patent, probably as a result of the mesonephros which producing urine which fills the tube.[38,39] From 37 to 40 days of development, the ureter loses its lumen. At 40 days of development, the ureter regains a lumen. Starting at the midpoint and progressing in both sides toward the non-develop kidney and the urogenital sinus, the lumen of the ureter reforms.[40,41,42,43] The last segments of the ureter to gain a lumen are at either end (kidney or urogenital sinus)[44].

When the development of the bladder progresses the mesonephric duct and the attached ureter are incorporated into the proximal urethra and base of the urinary bladder. In the male, the mesonephric duct drains into the prostatic urethra as the ejaculatory duct.[45,46,47] In females, the mesonephric duct regresses and the ureter alone remains. As the ureter and the mesonephric duct which is absorbed into the base of the bladder, they rotate so that the ureter meets the bladder cephalad to the point at which the mesonephric duct meet the urethra.[48]

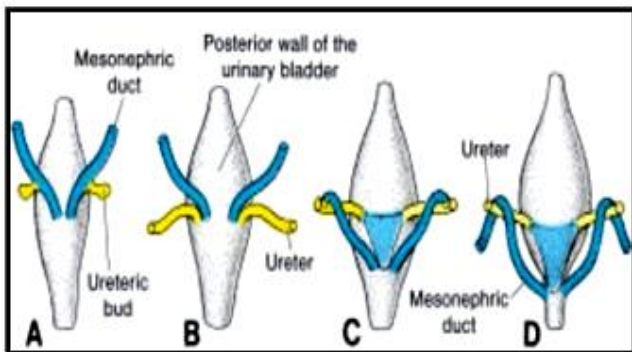


Fig 1. Mesonephric duct rotates as the ureter meets the bladder.

At the point where the ureter joins the urogenital sinus, a thin membrane (Chawalla's membrane) develops which separates the two lumens.[49] This membrane then ruptures allowing passage of fetal urine into the urogenital sinus. At nine weeks of development the metanephros, which will become the true kidney, starts to produce urine. When this fetal urine drains into the kidney, patency of the ureter is stabilized. Smooth muscle develops in the ureteric wall. Later, this muscle will generate and propagate peristaltic contractions to conduct urine from the kidney to the bladder.[50,51,52,53].

The inferior vena cava is a sequence of venous networks take part in the formation of the inferior vena cava. Each predominates temporarily, then retreats, and remains only partially in the final definitive system. [60,61,62,63,64] The mesonephros grows significantly and becomes very highly vascularized throughout week four. Although it is drained initially only by the posterior cardinal veins, a new classification takes over after week 4, the subcardinal network, which is made by the internal veins of the wolffian body. [65,66,67] The internal veins of the wolffian body are extensively anastomosed with the first posterior cardinal network and with each other, making the median subcardinal network, which soon predominates. [68,69] It takes over the posterior cardinal system, which vanishes in the central region of the embryo. The subcardinal sinus continues as the left renal vein. The anterior section of the left subcardinal vein disappears, but its posterior section will forms the left gonadal vein .

The right subcardinal vein will forms the right gonadal vein and the pararenal portion of the definitive inferior vena cava. Cranially, it continue with the mesenteric segment and the hepatic segment resulting from the hepatic vein (proximal right vitelline) and hepatic sinusoids[70,71,72,73,74] During weeks six and eight a extra dorsal network will form, called the supracardinal system, it will run parallel to the paravertebral sympathetic chain and opens into the proximal segment of the posterior cardinal veins. Anastomoses are made in between the two supracardinal veins, supracardinals and the subcardinals, on the right side between the extremities of the posterior cardinal vein. [75, 76] The left supracardinal vein becomes the hemiazygos vein and is drained toward the right by the transverse anastomosis which will make an interazygos communication. The right supracardinal vein becomes the azygos vein, which opens into right anterior cardinal vein. Under, the azygos vein drains the 2 iliac veins, thus become the prerenal portion of the definitive inferior vena cava. [77,78,79]

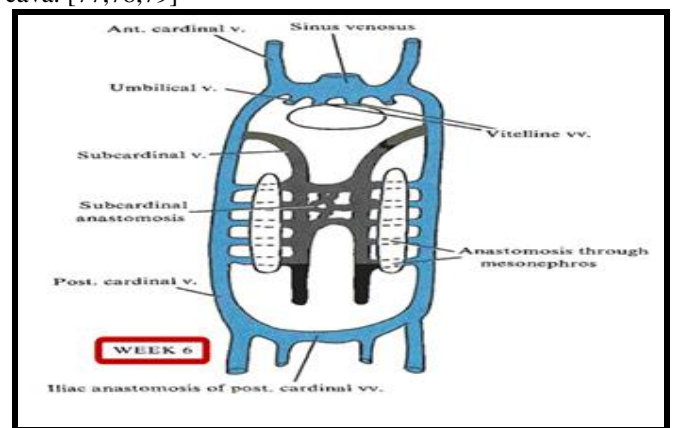


Fig 2. Development of inferior venacava in week 6.

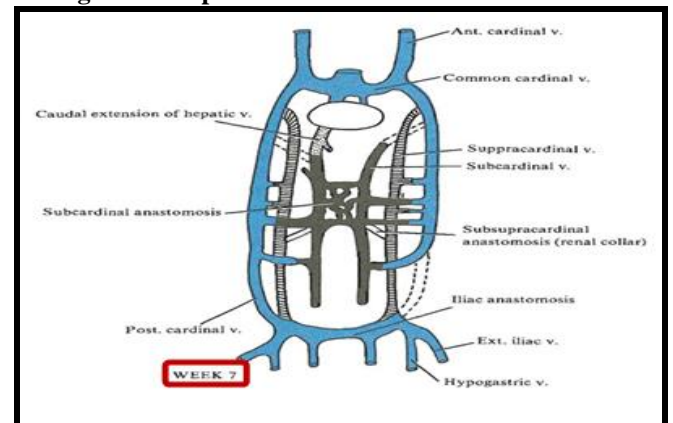


Fig 3. Development of inferior venacava in week 7.

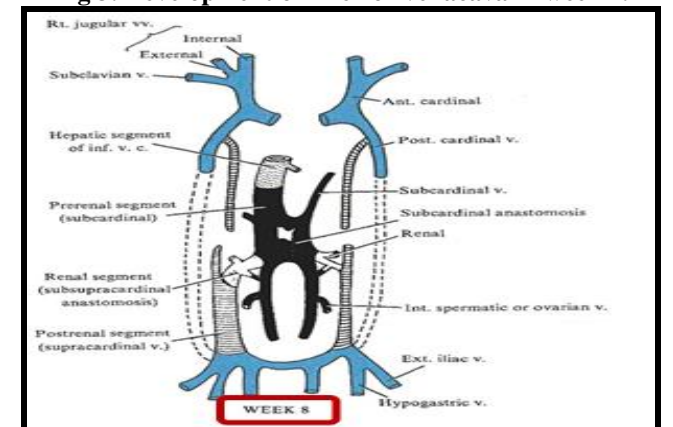


Fig 4. Development of inferior venacava in week 8.

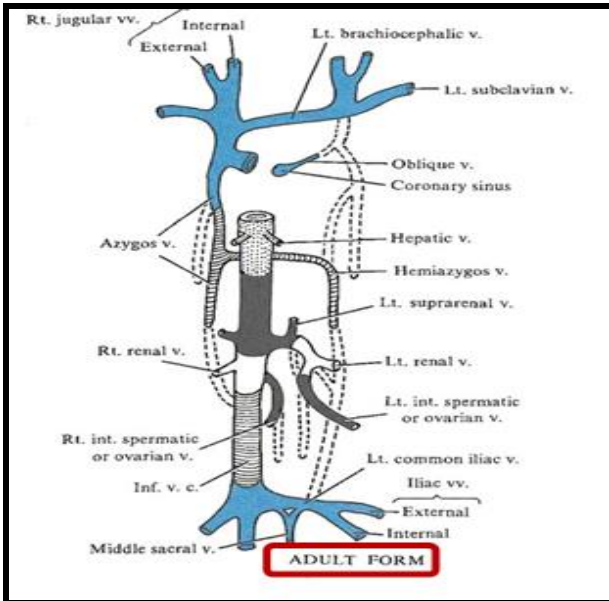


Fig 5. Development of inferior venacava in adult stage.

Ontogenesis for abnormal development

A circumcaval ureter is a rare congenital disorder usually associated with upper urinary tract stasis and an “S” or “fishhook” disformality of the ureter, in which the ureter itself passes behind the inferior vena cava [IVC]. [54] A circumcaval ureter results from the posterior cardinal vein persisting as the renal segment of the inferior vena cava [IVC] during development. Normally, the inferior vena cava formed from the vitelline vein, subcardinal vein and supracardinal veins, which must undergo step by step development, anastomosis and regression to become the inferior vena cava [IVC]. [55,56] Normally, the right vitelline vein forms the pre-renal or hepatic segment of the inferior vena cava, the right subcardinal vein forms the kidney segment and the right supracardinal vein forms the post-renal vena cava. Typically, the circumcaval ureter etiology is assumed to be abnormal embryologic formation of the vena cava as a result of atrophy failure of the right posterior cardinal vein in the lumbar portion. Whether the renal segment of inferior vena cava is made from the right posterior cardinal vein that lies ventral to the ureter, then the ureter will develop in a “circumcaval” position. [57,58,59]

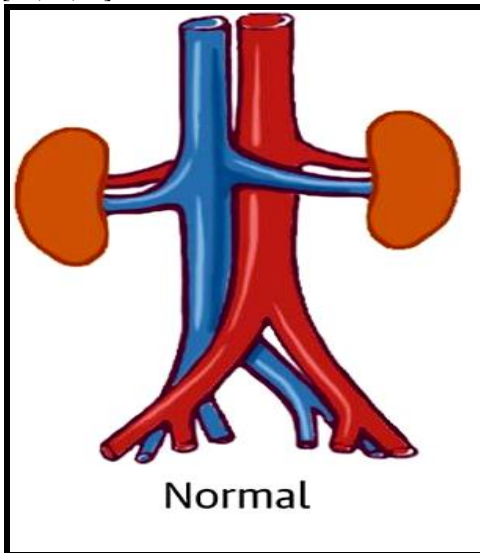


Fig 6. The schematic representation shows the normal blood system development.

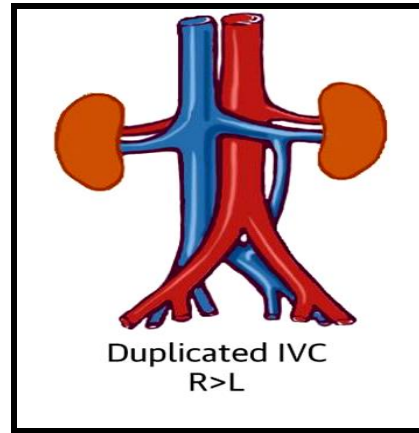


Fig 7. The schematic representation shows duplicated inferior vena cava R>L.

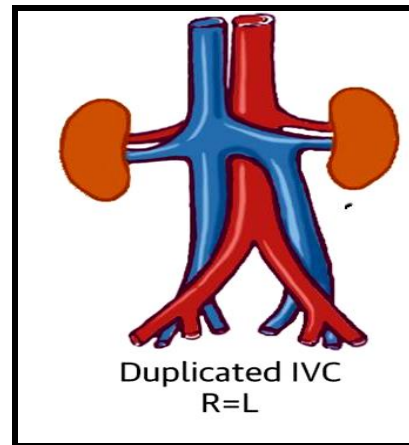


Fig 8. The schematic representation shows duplication of IVC with R=L.

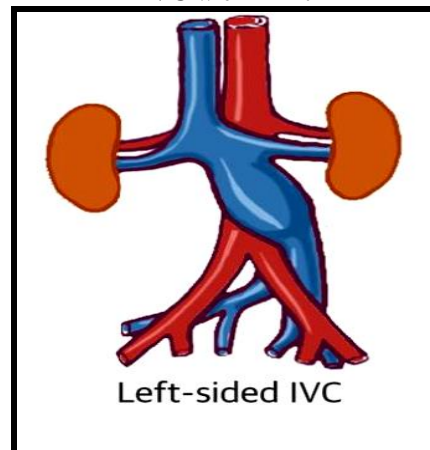


Fig 9. The schematic representation shows left sided IVC.

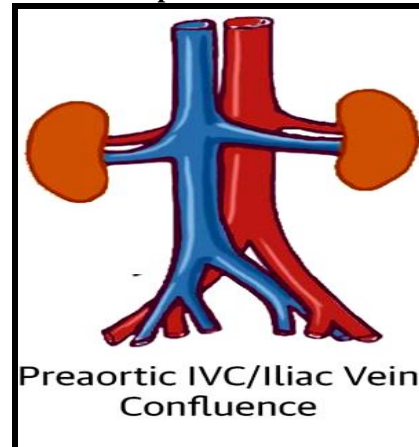


Fig 10. The schematic representation shows preaortic IVC.

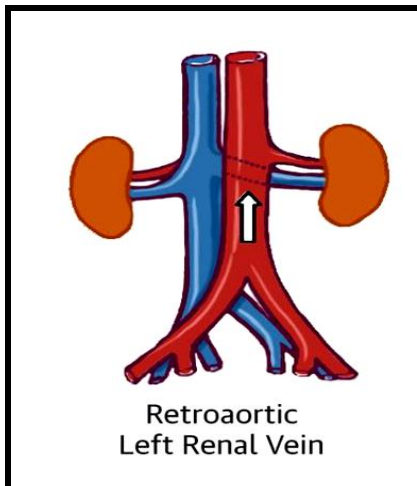


Fig 11. The schematic representation shows retroaortic left renal vein.

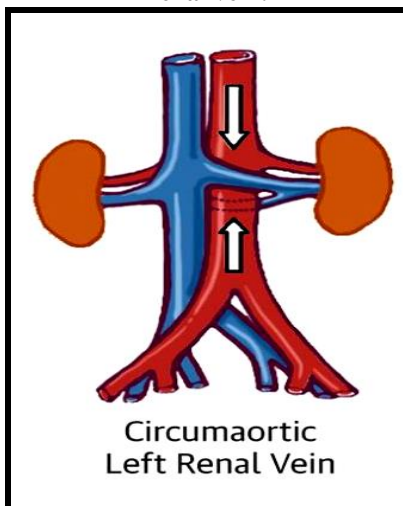


Fig 12. The schematic representation shows circumaortic left renal vein.

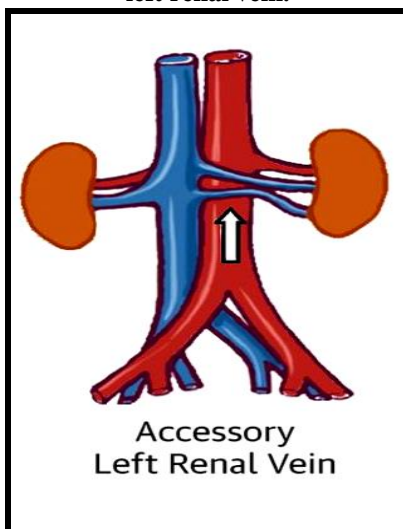


Fig 13. The schematic representation shows accessory left renal vein.

Discussion

A retrocaval ureter is a rare congenital disorder which occurs predominately in the male with a ratio of 2.8:1.3. Although it is a congenital disorder, patients rarely report before 2nd or 3rd decade. [80,81] The common presentation includes right flank pain, recurrent urinary tract infections and varying the degree of haematuria. Retrocaval ureter can be classified into two varieties according to radiological appearances.[82] In type-I ureter crosses behind the inferior vena cava at the level

of 3rd or the 4th lumbar vertebra and intravenous urography shows typical fish hook or S-shaped deformity, the extreme medial deviation with moderate to severe hydronephrosis. In type II which is less common, there is mild hydronephrosis, less medial deviation of the ureter and sickle-shaped abnormality at the level of obstruction. Both CT scan and magnetic resonance imaging are efficient methods of confirming the diagnosis. Surgical intervention is often required to alleviate the symptoms. [83,84,85] Open surgical exploration is commonly used it is being replaced by minimally invasive laparoscopic technique with advantages of minimal post-operative pain and shorter convalescence. Although this anomaly is known to urologists as a circumcaval or retrocaval ureter terms that are anatomically descriptive but it misleads about development—it is not the result of an abnormality in ureteral development, but it is an anomaly in the development of the inferior vena cava[86]

Conclusion

Retro-caval ureter is a rare congenital anomaly that presents clinically late in the second and third decades of life. Very few clinically symptomatic cases have been reported all over the world. Treatment is surgically allowing for correction of the anomaly with the resolution of symptoms. There is the need to research whether it is developmental anomaly of ureter of inferior vena cava.

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