Diagnosis and Treatment of the Circumcaval Ureter


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1. Topographic anatomy

The incidence of circumcaval ureter develops predominantly on the right side, but when it is reported on the left side, this congenital anomaly is associated with either partial or complete situs inversus or duplication of the inferior vena cava (IVC) [1–3]. This

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latter anomaly is a not uncommon variation that is caused by the persistence of all or part of the supracardinal vessels and presents an incidence of 1%–3% [4]. Recently Wang et al. [5] reported the case of a 21-year-old man who presented with right flank pain for about one month and had the radiological finding of moderate hydronephrosis and a right upper ureteral obstruction. The ureter was tapered and bent superiorly at the level of approximately L4. Due to the complete absence of stones or other significant abnormal findings, a circumcaval ureter was strongly suspected. At surgery the authors found that the ureter was kinked by an aberrant vessel, which drained laterally into the IVC. The inferior venocavography demonstrated one duplicated vessel about 0.8 cm in diameter and 6 cm long that originated from the ventral side of the IVC and subsequently drained laterally into the cava. Pierro et al. [6] reported two cases of a left-sided circumcaval ureter secondary to persistent left subcardinal vein without situs inversus or caval duplication. Bilateral circumcaval ureters have also been reported [7]. Several variants in the anatomic and topographic presentation of the anomalous circumcaval ureter have been observed in humans. Figs. 1–10 show few variants between those described in the literature [8]. Circumcaval ureters have been associated with several types of embryologic anomalies; recently an unusual presentation was reported in a 20-year-old woman with the notable findings of a circumcaval right ureter, Ladd’s bands, and incomplete intestinal malrotation that resulted in a six-month history of intermittent right lower quadrant pain [9].

Circumcaval ureters have been classified into two clinical types across the literature, in accordance with the common radiological appearance (Fig. 11) [10–12]. Type 1 (also named “low loop”) has been reported as the more common form; it is characterized by the so-called “typical S” or “fish-hook” deformity of the ureter to the level of the obstruction, with the point of obstruction placed some distance from the lateral margin of the IVC at the level of the third lumbar vertebra [13]. In the type 2 variant (also called “high loop”), the ureter has a “sickle-shaped” curve, with the point of obstruction at the lateral margin of the IVC. This second variant is rare, and represents around 10% of the known cases.
Type 1 is usually associated with moderate to severe hydronephrosis in 50% of the patients and type 2 with mild or no hydronephrosis [10–12]. Indeed, in the latter form, the upper ureter is non-kinked but passes behind the IVC at a higher level; the renal pelvis and upper ureter lies almost horizontal before it encircles the IVC with a smooth curve. The ureter is compressed against the perivertebral tissue.

Horseshoe kidney is the most common fusion anomaly of the kidney. It occurs in one in 353 cases to one in 1800 cases [14,15]. A great variety of associated congenital anomalies have occurred with horseshoe kidney, but horseshoe kidney accompanied by a totally circumcaval ureter has rarely been reported in the same patient (Fig. 12) [16–22]. Moreover, both features are well recognized as uncommon anomalies of the genitourinary system and rarely is the diagnosis of both anomalies made before surgical intervention [18].

2. Clinical diagnosis

As previously introduced, patients who suffer from both types of circumcaval ureter usually present in
their third to fourth decade of life with right flank pain and discomfort caused by ureteral obstruction and resulting hydronephrosis [8,12,23–29]. Pain associated with the hydronephrosis can be intermittent, dull, and aching. Patients may also complain of urinary tract infections, gross haematuria, or fever [8,12,23–26,29]. Haematuria in varying degrees is often present. Calculi occasionally co-exist with a circumcaval ureter. Fillo et al. [29] reported a case of gross haematuria in a patient with a carcinoma of the retrocaval ureter.

Kenawi and Williams [12] reported that in their cohort of patients, 52.5% complained of flank pain, 17.9% had significant kidney colic, 23.5% suffered from urinary tract infections, and 21.6% had haematuria.

The hydronephrosis in patients with a retrocaval ureter is mainly described as a consequence of the compression by the psoas muscle, spinal column, and IVC on the anomalous ureter [24,30]. In patients who suffer from retrocaval ureter, the renal pelvis and upper ureter are typically elongated and dilated in a “J” or fish-hook shape before they pass behind the IVC. However, ureteral obstruction is a frequent but not an inevitable complication [31].

### 3. Radiologic diagnosis

Intravenous urogram (IVU) and retrograde urography were commonly used to diagnose a circumcaval ureter [8,12] (Figs. 13 and 14). The IVU in the early stage of ureteral stenosis may only show a dilatation of the renal pelvis, calyces, and upper ureter above the site of obstruction. Typically, IVU can fail to visualize the portion of ureter that extends behind the IVC; a retrograde ureteropyelogram, however, may help demonstrate the typical fish-hook curve of the upper ureter towards the midline, with the retrocaval segment at the level of L3 or L4 [8,12,24,32,33].

A spiral computed tomography (CT) scan has been recently considered the tool of choice for the diagnosis of IVC abnormalities and circumcaval ureter [2,13,34–36]. In 1999, Pienkny et al. suggested that a three-dimensional volume-rendered CT scan in the excretory phase, combined with diuretic
renography, could be the radiologic evaluation of choice for patients with a suspected circumcaval ureter, as it is non-invasive and accurately determines the anatomic relationship of the IVC and ureter [37]. CT can also represent the procedure of choice to confirm an ultrasound diagnosis and avoid retrograde ureteropyelography [38–41]. Spiral CT, which can simultaneously outline the ureter and IVC free of respiratory mis-registration and motion artefacts, may produce overlapping images, and may represent the diagnostic modality of choice, because it enables the surgeon to avoid the possible complications and expenses of other invasive procedures [13].

More recently, however, Uthappa et al. [42] reported data that relate to the potential use of magnetic resonance imaging (MRI) to differentially diagnose a circumcaval ureter with equal effectiveness of a spiral CT scan but without radiation risk (see Chapter 3 in this supplement).

In terms of differential diagnosis, circumcaval ureter must be differentiated from retroperitoneal mass and retroperitoneal fibrosis. Typically, in the first case the IVU often reveals lateral deviation of the ureter from the compression of the mass. When retroperitoneal fibrosis occurs, the ureter is involved on both sides and the whole segment of the ureter is stiff in the IVU feature [24].

4. Personal case report

A 14-year-old Moroccan boy, normal 46, XY karyotype, was referred to the Department of Paediatrics for failure to thrive (height: 140.6 cm = −3 DS; weight = 32.3 kg), which was documented also by a total skeleton X-ray that showed a dorsal kyphosis and growth delay. He was the second-oldest of four siblings born from Moroccan parents; it has not been possible to establish a kinship with certainty.

During the hospitalisation he was submitted to various instrumental evaluations, among which an
echocardiogram and a cardiac MR, which documented the presence of a right aortic arc, with a right-positioned descending aorta that goes down progressively, anterior to the dorsal vertebral column bringing itself leftwards in correspondence of the diaphragmatic passage. The cardiac MR also showed a pervious oval foramen, with a diameter of 8 mm, and small ectasia of both atria.

Paediatric examination: hypertelorism, synophris with dark and thick eyebrows, lengthened filter, small mouth with thinned superior lip, micrognazia, hypoplasia of the dental enamel. Slightly flattened short nasal septum, with rounded off tip, ogival palate, thickened ear tents were also documented. The boy also suffered from ligamentous laxity, clynodattilia in the left hand, significant bilateral hallux valgus, and gait with inarotation of the limb, coupled with valgism of the hind foot. The ophthalmic visit found a bilateral iris coloboma.

The urological history underlined an intervention of bilateral orchipexy at the age of three; enuresis was still persistent at the time. The physical examination showed hypotrophy of the right testis compared with the contra lateral (testis ultrasound: right testicle = $23 \times 15 \times 11$ mm $[2.1 \text{cc}]$; left testicle = of $26 \times 15 \times 12$ mms $[2.7 \text{cc}]$; US presence of some small [1–2 mm] hyperechogenic area, possibly fibrotic, in the left testis).

Abdomen ultrasound was positive for a small degree of right dilatation of the pelvis with right lumbar ureter of a diameter equal to 16 mm. The cystogram resulted positive for sphincter dyssnergia, but negative for vesico-ureteral reflux. The boy thus underwent IVU. Although the boy was completely asymptomatic, at 14 years and five months a right circumcaval ureter was diagnosed during an IVU (Figs. 15–17).

The paediatricians diagnosed a recessive autosomal syndrome called spondylocarpotarsal synostosis syndrome and planned a substitutive therapy with growth hormone.

Moreover, the boy had an intervention of right ureteroplasty with a right flank surgical access along the prolongation of the XI rib. After the right kidney was found, a lysis of adherences between the right circumcaval ureter and the IVC was performed. Subsequently, a dismembered pyeloplasty was done, after having uncrossed the ureter from the IVC. A pyeloureteral anastomosis was then completed using a PDS 6/0 running suture, thus, a D-J ureteral stent was placed. The boy had a regular postoperative hospital stay, with the removal of the indwelling catheter after a 24-hour period and the hospital discharge during the second postoperative day. The D-J ureteral stent was then removed at the 30-day postoperative visit. A three-month follow-up ultrasound evaluation showed mild hypotonia of the right pelvis without any sign of hydronephrosis. At 20-month follow-up, a renal scintigraphy documented a bilateral normal renal function, coupled with a minimal right-sided pyelecstasy.

5. Treatment

Various techniques for the management of circumcaval ureter have been reported.

In patients with minimal calicectasis and no subjective symptoms, surgical correction is not mandatory, but observation should be maintained. Therefore, circumcaval ureter has been defined a rare congenital anomaly that requires surgical correction in the symptomatic patients [33]. Conservative treatment and periodical examination
should be given to those patients without hydronephrosis, infection, and stone formation. A periodic examination has been also suggested for patients with mild hydronephrosis.
When either obstructive symptoms or kidney function caused by circumcaval ureter worsen, surgical correction is indicated to preserve renal functioning and to provide long-term symptomatic relief [5,43,44].

5.1. Surgical treatment

Open surgery is the standard treatment and is usually successful. Division of the dilated renal pelvis with transposition and reanastomosis was initially described by Harril [45] and has been the most popular form of treatment in patients with symptomatic hydronephrosis. Both flank and trans-abdominal approaches have been used for this procedure.

The choice of open surgery approach depended primarily on the severity of the hydronephrosis, impairment of kidney function, and the type of anomaly [8,24].

Most of the authors that suggested an ureteropelvis anastomosis also advocated Harril’s method [45] by which a section is made at the level of the pelvis just above the ureteropelvic junction. Char-
acteristic of this approach is that the abundant vascular supplies of the pelvis and proximal ureter may be kept, and postoperative stricture at the anastomotic site is less likely. Xiaodong et al. [24] underlined that when an 8F catheter could not pass through the circumcaval segment easily, the stenotic section of the ureter must be excised, and a dismembered pyeloplasty should be done. Puigvert et al. [46], for instance, supported the resection of the pelvis just above the ureteropelvic junction, with a subsequent ureteropyeloplasty, just leaving untouched the segment of the ureter previously allocated in a retrocaval fashion (Figs. 18–20).

A number of open surgical approaches have suggested the transection of the ureter with the uretero-ureteric anastomosis and repositioning in a normal anatomic relationship with the IVC [8,24]. These techniques suggested either the ureterotomy or the resection of the stenotic ureter at the site of the segment of ureter compressed by the IVC [47], distally and close to the bladder with

Fig. 17 – Post-voidal i.v. urogram feature showing a persisting right-sided hydronephrosis and dilatation of the proximal ureter in the 14-year-old boy with a type I circumcaval ureter (personal case).

Fig. 18 – Resection of the hydronephrotic pelvis throughout the ureteropyeloplasty according to Puigvert et al. (modified from Bissi and Rigatti. L’uretere circumcavale. Anatoma chirurgica – Diagnostica –Terapia. Eds Casa Editrice Ambrosiana. pp 1–149, 1977).

Fig. 19 – Puigvert’s technique. After transposition of the ureter, a ureteropyeloplasty was performed (modified from Bissi and Rigatti. L’uretere circumcavale. Anatoma chirurgica – Diagnostica –Terapia. Eds Casa Editrice Ambrosiana. pp 1–149, 1977).
a concomitant ureterocystostomy [48], or proximally to the ureteropelvic junction [49].

Other surgical options include nephrectomy for the non-functioning kidney, because of severe hydronephrosis and infection [8, 24].

In 1957 Goodwin et al. [50] proposed a surgical approach aimed to divide and re-anastomose the IVC successfully after relocating the ureter anteriorly (Figs. 21–23). This technique had great operative risks and potentially severe complications. Supporters underlined that it was a good choice of treatment, particularly when there was absence or disease and malfunction of the contralateral kidney.

Xiaodong et al. [24] also suggested to use a so-called “vena cava supporter” (Fig. 24) mainly for cases with lumen stenosis caused only by torsion and compression by the IVC. Adhesive tissue around the circumcaval ureter was well dissected, and the torsion of the ureter was corrected. A chemical or metal tool, as well as surrounding soft tissue such
as the greater psoas muscle, were then placed between the IVC and the ureter.

Major criticisms of any kind of open surgery are that it may require a large skin incision and cause significant postoperative pain; moreover, the convalescence might be complicated by wound pain and infection.

5.2. Laparoscopic repair of the circumcaval ureter

The open ureteroureterostomy remained the gold standard surgical approach to treat the circumcaval ureter for many years [51]. In the past 10 years, however, technological breakthroughs coupled with the intensive growth of minimally invasive laparoscopic procedures have almost replaced open surgery [52,53].

After that a laparoscopic dismembered pyeloplasty for ureteropelvic junction stenosis was reported by Schuessler et al. in 1993 [54], laparoscopic transposition and reanastomosis of a circumcaval ureter were also performed [55–64]. The laparoscopic dismembered pyeloplasty rapidly demonstrated the obvious advantage of minimal disfigurement and morbidity to the patients (see Chapter 4 in this supplement).

Baba et al. [55] reported a dismembered pyeloplasty under transperitoneal laparoscopy. These authors decided to mobilize the entire ascending colon by incising the right paracolic gutter around the inferior margin of the cecum, which provided the best exposure of the retroperitoneal area to the right of the left renal artery above and the inferior mesenteric artery below. The IVC was easily dissected by retracting the ascending colon toward the midline of the abdomen with the operating table tilted to keep the patient in the half-recumbent position. With blunt dissection and gentle traction, the postcaval segment of the right ureter was safely dissected with instruments inserted through two 11-mm trocars placed in the midclavicular line. A 5-mm port placed in the middle axillary line was used for dissection and to lift the extrarenal pelvis and the upper third of the ureter. The difficulty of reapproximating the ureter and the renal pelvis accounted for almost one-third of the operative time. The renogram parameters obtained at two-month follow-up showed marked resolution of the obstruction [55].

A transperitoneal laparoscopic approach was also used by others [56,57,59]. Matsuda et al. [56], for instance, reported that in their 38-year-old patient a part of the ureter, which was about 3 cm long and located behind the IVC, had to be removed because it looked narrow and dysplastic. An end-to-end reanastomosis of the upper and lower ends of the ureter was then performed laparoscopically. At the end of the procedure the retroperitoneum was incised laterally to the ascending colon was completely closed. Bleeding during the procedure was less than 30 ml; the postoperative period was uneventful and the patient complained of minimal post-surgical pain and received no analgesics.

More recently, Ameda et al. [62] reported data that related to the case of a 20-year-old woman who suffered from a circumcaval ureter that was discovered as she was being treated for urolithiasis. After a spontaneous stone discharge, right hydrenephrosis remained and a diuretic renography with 99mTc-DTPA showed no definite obstruction. This woman was transabdominally laparoscopically approached. Under general anaesthesia the patient was placed in a modified flank position with the right leg slightly elevated, which eased intraoperative manipulation of the ureteral catheter. Open trocar placement was done at the umbilicus (12 mm), then the second port (10 mm) at the midclavicular line below the costal margin, the third port (10 mm) midway between the anterosuperior iliac spine and the umbilicus, and the fourth port (5 mm) was placed on the anterior axillary line at the same level of the umbilicus (Fig. 25). The ascending colon was reflected medially to expose the retroperitoneum. After its identification, the right ureter was divided just distal to the crossing of the IVC. Therefore, the ureter was repositioned to lie anterior to the IVC itself and then the stenotic portion of about 2 cm was transacted. Tension-free anastomosis with interrupted 4-0 absorbable sutures was performed with an intracorporeal suturing technique. Total surgical time was 7.5 h.

Fig. 24 – Inferior vena cava supporter (modified from Xiaodong Z et al. Diagnosis and treatment of retrocaval ureter. Eur Urol 1990;18:207–10).
with estimated blood loss less than 20 ml. The post-operative course was uneventful without any complications.

Salomon et al. [60] in 1999 reported the first case of purely retroperitoneal laparoscopic repair of a circumcaval ureter, which suggested that in their patient the retroperitoneal laparoscopy represented the more direct approach to the urinary tract. The operative time was significantly shorter than with the previously reported transabdominal laparoscopic approach [56,57]. Salomon hypothesized that the shorter time was obtained because dissection of the retroperitoneal space was not hindered by intraabdominal organs [60]. Mugiya et al. [61] confirmed that the retroperitoneoscopy treatment could be superior to the conventional transabdominal approach to perform the laparoscopic transposition and reanastomosis of a circumcaval ureter. They also reported the use of an automatic suture device [65] to retroperitoneoscopically correct a circumcaval ureter [61].

The main limiting factor for both the transabdominal and the retroperitoneal laparoscopic repair of the circumcaval ureter was the intracorporeal anastomosis of the ureter that significantly increased the surgical time [55,56,60]. Mugiya et al. [61] first used an automatic suture device to reduce the surgical time. More recently, Tobias-Machado et al. [66] reported a case of retroperitoneoscopy surgery coupled with extracorporeal uretero-ureteral anastomosis for treating circumcaval ureter in a 24-year-old woman who complained of recurring lumbar colic pain associated with repeated episodes of acute pyelonephritis on the same side. After having observed the ureteral point with the least tension for exteriorization, the ureter was sectioned at that site. The ureteral stumps were thus exteriorized through the incision of the 12-mm port, after having enlarged the skin incision to 20 mm. The proximal and distal ureteral margins were resected and a double running spatulate suture (namely, posteriorly first and anteriorly subsequently) was performed. Total surgical time was 130 minutes; the end-to-end anastomosis took 40 minutes.

6. Conclusions

The circumcaval ureter is a rare congenital anomaly caused by an error in the embryogenic development of the IVC. It is usually associated with upper urinary tract hydronephrosis, and patients present in their third to fourth decade of life with right flank pain and discomfort. Patients may also complain of urinary tract infections, haematuria, or fever.

Intravenous urogram and retrograde urography have been commonly used to diagnose a circumcaval ureter. However, a spiral CT scan has been recently considered the tool of choice for the diagnosis of IVC abnormalities and circumcaval ureter. Even more recently, the potential use of the MRI to differentially diagnose a circumcaval ureter was suggested to have equal effectiveness of a spiral CT scan but without radiation risk.

Comparisons between historical reports about open surgery and laparoscopic transposition and reanastomosis of a circumcaval ureter have clearly shown the advantages of minimally invasive approaches, less intraoperative bleeding, specifically a shorter post-operative hospital stay, reduced postoperative pain, earlier return to daily activities, and a significant superior aesthetics effect, while preserving therapeutic efficacy.

References

Retrocaval ureter: a review of the literature


