

# Secondary pelvic congestion syndrome: description and radiographic diagnosis

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WINER AG, CHAKIRYAN NH, MOONEY RP, VERGES D, GHANAAT M, ALLAEI A, ROBINSON L, ZINN H, LANG EK. Secondary pelvic congestion syndrome: description and radiographic diagnosis. *Can J Urol* 2014;21(4):7365-7368.

**Introduction:** Pelvic congestion syndrome (PCS) is a complex condition of the pelvic venous system leading to nonspecific pelvic pain that was initially described in females alone. The underlying abnormalities, though diverse, all result in increased pressure in the left gonadal vein which is transmitted retrograde into the pelvic venous system. Our primary aim was to describe our findings of secondary PCS as a distinct entity from primary PCS in that it has an identifiable vascular etiology and is gender nonspecific. We also aimed to assess the adequacy of late-arterial phase CT urography (CTU) as the initial imaging modality in diagnosing and evaluating secondary PCS.

**Materials and methods:** We retrospectively reviewed 59 patients with PCS, 36 males and 23 females ages 24 to 63, from 2000-2011. To maximize opacification, CTU images were taken in the late-arterial phase with a 35-50 second delay after contrast administration.

**Results:** Review of our cases revealed multiple etiologies for PCS, including: Nutcracker syndrome (19 cases), cirrhosis (17), retroaortic left renal vein (11), tumor thrombosis of the IVC (5), portal vein thrombosis (4), renal cell carcinoma with left renal vein thrombosis (2), and left kidney AVF (1). The most common symptom was unexplained chronic pelvic pain. The patients in our series had clearly identifiable vascular flow abnormalities leading to the development of PCS, and were therefore diagnosed as having secondary PCS. All cases were easily identified utilizing CTU to visualize and measure dilation of the left gonadal vein and pelvic varices. This modality also proved valuable in the identification and management of the various underlying causes of secondary PCS.

**Conclusion:** Secondary PCS is distinct from primary PCS in that it arises from clearly identifiable vascular flow abnormalities and occurs in both males and females. The diverse set of underlying etiologies, as well as the resulting congested varices, can be reliably and adequately visualized using CTU as the initial imaging modality.

**Key Words:** pelvic, congestion, syndrome, chronic pelvic pain

## Introduction

Pelvic congestion syndrome (PCS) is an often under- and misdiagnosed condition of the pelvic venous system. It most commonly manifests as unexplained chronic pelvic pain in women only. Many etiologies exist, but all result in increased pressure in the pelvic venous system, resulting in engorged, painful pelvic varices.

Primary PCS has been previously described, occurring solely in multiparous women.<sup>1-5</sup> The exact etiology remains unknown, however it is believed to be associated with increased estrogen levels and anatomic changes related to pregnancy, including increased intravascular volume and valvular incompetence leading to pelvic venous varicosities.

Another set of patients, that to date has not been well described, present with PCS due to an underlying vascular etiology resulting in either increased flow or outflow obstruction at the level of the left renal or left gonadal vein. The increased pressure is transmitted retrograde into the pelvic venous system, resulting in dilated veins and symptoms of PCS. This set will be referred to as secondary PCS.

Accepted for publication May 2014

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TABLE 1. Groupings and etiologies of secondary pelvic congestion syndrome cases encountered

Category	Etiology	Number of cases
High flow	Cirrhosis	17
	Portal vein thrombosis	4
	Left kidney AVF	1
Outflow obstruction	Nutcracker syndrome	19
	Retroaortic left renal vein	11
	RCC w/tumor thrombus of IVC	5
	RCC w/tumor thrombus of left renal vein	2

AVF = arteriovenous fistula; RCC = renal cell carcinoma; IVC = inferior vena cava

Current radiographic modalities being used to image PCS include ultrasound, CT angiogram (CTA), MR angiogram (MRA), and venography. Recent literature suggests ultrasound as the appropriate initial imaging test to work up PCS, and venography as the gold standard imaging.<sup>4,7</sup>

Our aim in this study is to identify and describe the various etiologies of secondary PCS and assess the adequacy of late-arterial phase CT urography (CTU) as the initial imaging modality in diagnosing and evaluating this condition.

### Materials and methods

A retrospective review of charts from SUNY Downstate Medical Center, Brooklyn, NY and Tulane Health Science Center, New Orleans, LA from 2000 to 2011, revealed 59 patients with secondary PCS, 36 males and 23 females, aged 24 to 63. Informed consent for the imaging study was obtained from all patients. A separate file was created for extracted data to insure privacy. Triple or quadruple phase enhanced CTs were performed on each patient using GE MDCT 4; GE Lightspeed Plus (all Milwaukee, WI, USA) and Toshiba XP (Japan) generating late cortico-medullary phase (25-35 second delay) and parenchymal phase (35-50 second delay) images. Collateral flow or obstruction of the left renal vein and decompression flow via the gonadal vein into the pelvic veins was best demonstrated on late cortico-medullary or parenchymal phase contrast enhanced CTs.

Particular attention was paid to evaluating the underlying etiology of each patient's PCS, and analyzing the incidence of these etiologies amongst our population of secondary PCS patients.

### Results

Listed in Table 1 is an overview of our series of patients, which reveals multiple distinct etiologies for secondary

PCS that can be divided into two categories: high flow, and outflow obstruction. Underlying etiologies in the high flow group were cirrhosis (n = 17), portal vein thrombosis (n = 4), and AVF of the left kidney (n = 1). Outflow obstruction was the underlying etiology in patients with Nutcracker syndrome (n = 19), retroaortic left renal vein (n = 11), and renal cell carcinoma with tumor thrombus of left renal vein (n = 2) and of the IVC (n = 5).

Utilizing CTU, all of these cases were clearly identified as PCS by visualizing and measuring the dilated left gonadal vein, an indication of high pressure in the pelvic venous system. Each of the underlying

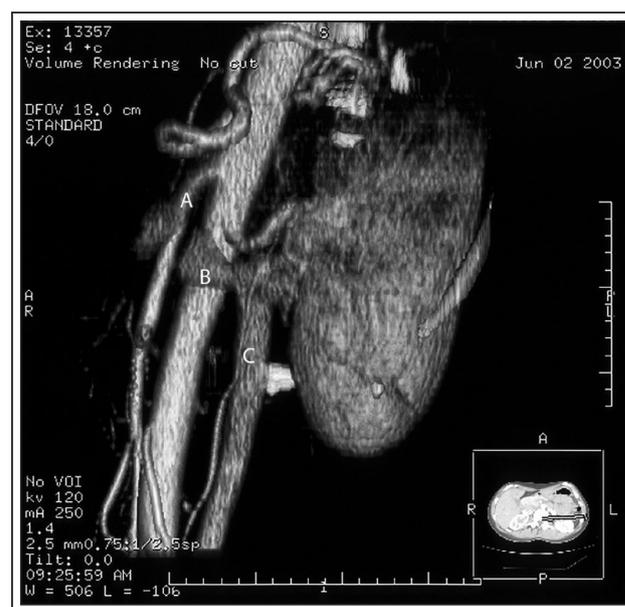


Figure 1. Volume rendered CTU demonstrating nutcracker syndrome in a case of secondary pelvic congestion syndrome. The superior mesenteric artery (A) is impinging on the left renal vein (B) resulting in a massively dilated left gonadal vein (C).

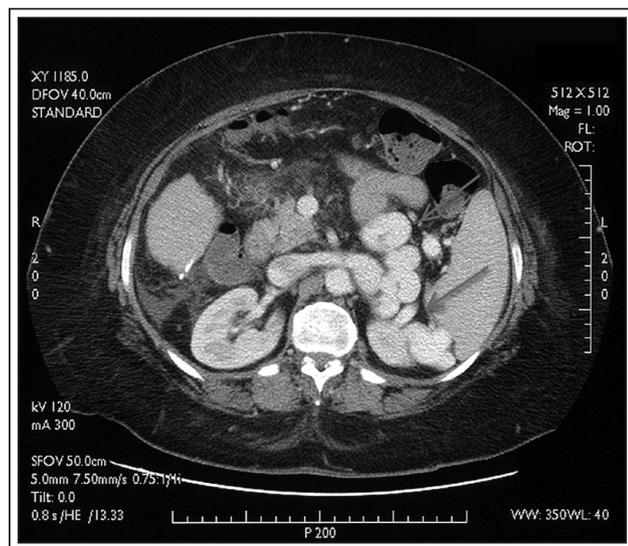
etiologies were also easily diagnosed and evaluated with CTU, as it can visualize the vascular disturbances that led to the formation of secondary PCS, Figures 1-4.

## Discussion

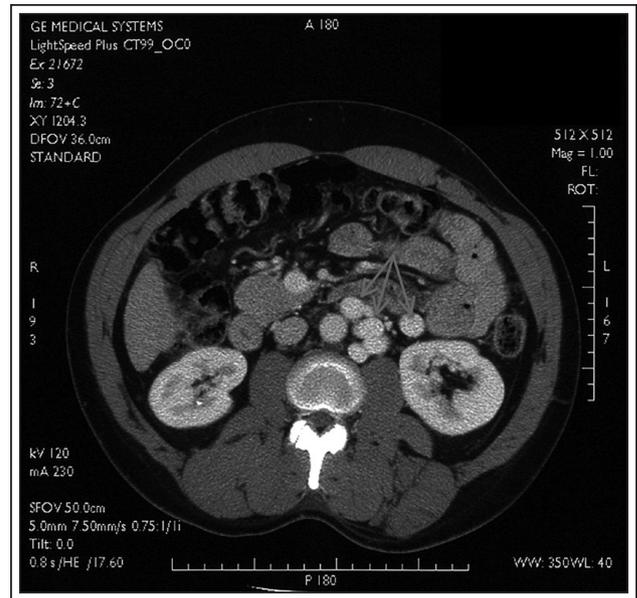
Our findings provide new insight into a unique subset of PCS patients that does not fit the classic description. We have demonstrated that secondary PCS, in contrast to primary PCS, is not gender specific given that the underlying vascular abnormalities resulting in secondary PCS are not gender specific themselves. Therefore, we feel that the diagnosis of secondary PCS should be considered in the differential diagnosis of a male patient presenting with unexplained chronic pelvic pain.

High flow secondary PCS is a result of increased flow rates in the left renal vein. The increased flow rate intensifies the pressure in left renal vein, which leads to retrograde flow in the left gonadal vein and the pelvic venous system, resulting in dilated painful pelvic varices. Our cases of high flow secondary PCS were overwhelmingly caused by cirrhosis (17/22, 77% of cases). Cirrhosis results in dramatically increased pressure in the portal venous system, which can result in portocaval shunting between the splenic vein and left renal vein, Figure 2. These splenorenal varices cause a significant elevation in the flow rate in the left renal vein, with pressures transmitting down the left gonadal vein and into the pelvic venous system, Figure 3.

Outflow obstruction secondary PCS occurs as a result of obstruction of the left renal vein. This obstruction

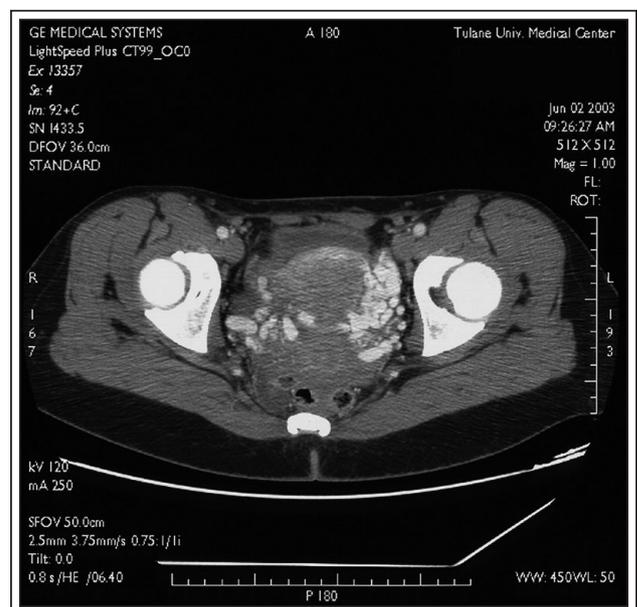


**Figure 2.** Massive splenorenal varices are seen as a result of cirrhosis and subsequent portal venous hypertension.



**Figure 3.** Multiple massively dilated left gonadal veins draining into the left renal vein as a result of increased pelvic venous pressure and resultant angiogenesis.

leads to a dramatic rise in pressure in the left renal vein, the left gonadal vein, and ultimately the pelvic venous system, Figure 4. The two most common etiologies in our series were Nutcracker syndrome (19 cases) and retroaortic left renal vein (11 cases). Retroaortic left renal vein is a congenital phenomenon while nutcracker



**Figure 4.** Dilated pelvic varices demonstrated in a case of secondary pelvic congestion syndrome.

syndrome can be either congenital or acquired. Nutcracker syndrome is an anatomic abnormality that involves the compression of the left renal vein between the superior mesenteric artery and the abdominal aorta.<sup>8</sup> This phenomenon can lead to the formation of varices in the presence of competent valves, Figure 1. Dellavedova et al suggested this diagnosis should be considered in patients presenting with chronic pelvic/flank pain and microscopic hematuria.<sup>9</sup>

Another cause of outflow obstruction secondary PCS was renal cell carcinoma with tumor thrombus of the IVC or left renal vein. Although a less frequent cause of secondary PCS in our cohort, the implications may be clinically relevant. Specifically, the possibility of renal cell carcinoma presenting with symptoms of PCS, secondary to tumor thrombus causing increased pressure in the pelvic veins. Thus, chronic pelvic pain as a result of secondary PCS may be the initial symptom leading to the diagnosis of renal cell carcinoma.

A notable finding encountered in our series was that of secondary PCS presenting with hematuria as a result of increased pressure in the left renal vein. This finding introduces secondary PCS as a possible diagnosis for the patient presenting with hematuria and chronic pelvic pain.

CTU was able to reliably detect dilation of the left gonadal vein, an indication of high pressure in the pelvic venous system, with the patient in a supine position. CTU also proved very useful in evaluating the various underlying vascular etiologies that contributed to the development of secondary PCS, Figures 1-3. Although less expensive and more readily available than CTU, we feel that ultrasound has a very limited capacity for evaluation of the underlying vascular etiologies of patients with secondary PCS, and therefore is not recommended. Also, in order to properly visualize the dilated pelvic veins on ultrasound, the patient would have to be upright to allow for filling of the varices. Additionally, the most reliable approach is either transrectal or transvaginal, which when combined with an upright positioning would make a reliable ultrasound diagnosis extremely uncomfortable and technically challenging.

Walling et al reported that women with chronic pelvic pain related to PCS report a higher incidence of depression and anxiety.<sup>10</sup> Their psychological comorbidities may be exacerbated by being subjected to a multitude of diagnostic tests, the frustration of chronic unexplained pain, and the fear of severe underlying disease. It is not unlikely that this psychological toll also affects patients with secondary PCS, specifically men who have gone undiagnosed due to the gender-specificity of the current PCS definition.

There are several limitations of the present study worth discussing. First and foremost, this was a strictly observational retrospective study. In addition, our analysis was limited by the small sample size of our cohort due to the rarity of the syndrome we aimed to describe. Finally, significant baseline information were not readily available for many of the patients in our study, which may have aided in the generalizability of our findings.

## Conclusion

We believe secondary PCS to be a distinct entity from primary PCS, as it is caused by an identifiable underlying vascular abnormality resulting in increased pressure and painful varices in the pelvic venous system. We have demonstrated that secondary PCS is not gender specific, as many cases have been identified in men and women across a wide spectrum of underlying etiologies. Additionally, we feel that CTU is an appropriate initial imaging modality for the workup of secondary PCS over other imaging techniques previously described. In light of these findings, secondary PCS should be considered in one's differential diagnosis when evaluating either a male or female patient presenting with unexplained chronic pelvic pain, with CTU being the most appropriate initial imaging study. □

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