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Persistent left superior vena cava: Two case reports and a review from nephrologists' perspective

Tuncay SAHUTOGLU,[1] Tamer SAKACI,[1] Ekrem KARA,[2] Elbis AHBAP,[1] Taner BASTURK,[1] Yener KOC,[1] Mustafa SEVINC,[1] Cuneyt AKGOL,[1] Arzu Ozdemir KAYARLAR,[1] Zuhal Atan UCAR,[1] Feyza Bayraktar CAGLAYAN,[1] Nuri Baris HASBAL,[1] Perin NAZIF,[1] Mahmoud ISLEEM,[1] Elif SAHUTOGLU,[3] Abdulkadir UNSAL[1]

[1]*Department of Nephrology, Sisli Hamidiye Etfal Educational and Research Hospital, Istanbul, Turkey;*
[2]*Faculty of Medicine, Department of Internal Medicine, Recep Tayyip Erdogan University, Rize, Turkey;*
[3]*Department of Chest Disease, Yedikule Chest Disease and Surgery Education and Research Hospital, Istanbul, Turkey*

Abstract

Thoracic venous anomalies without congenital heart anomalies are present in minority of the population, but they are frequent enough to be encountered while placing hemodialysis catheters through the jugular or subclavian veins. Persistent left superior vena cava is the most commonly seen anomaly and it is rarely noticed before the observation of an unusual course of hemodialysis catheter or guidewire on chest X-ray. We present two patients with previously unspotted persistent left superior vena cava and uncomplicated hemodialysis catheter insertions through the internal jugular veins with good catheter functions. Review of the relevant literature from a nephrologists' perspective with technical aspects is provided.

Key words: Hemodialysis catheter, persistent left superior vena cava, hemodialysis

INTRODUCTION

Hemodialysis is one of the three renal replacement therapy options, for which vascular access is sine qua non. In the order of preference, arteriovenous fistula (AVF), arteriovenous grafts, tunneled, and temporary hemodialysis catheters constitute the current means of vascular access.[1] Nonetheless, hemodialysis catheters are still very frequently used in some parts of the world, such as 51% and 75% of patients used catheters at dialysis initiation according to 2013 Turkish Society of Nephrology registry report and Xue et al. in the United States, respectively.[2,3] Hemodialysis catheters are typically inserted into one of the central veins, preferably the right internal jugular vein (IJV), assuming that the venous anatomy is normal. Thrombosis, venous stenosis, infections, and insertion related traumatic injuries are the well-known major

Correspondence to: T. Sahutoglu, MD, Department of Nephrology, Sisli Hamidiye Etfal Educational and Research Hospital, Halaskargazi Cad. Etfal Sk., 34371 Şişli/İstanbul, Turkey. E-mail: tu_cay83@yahoo.com

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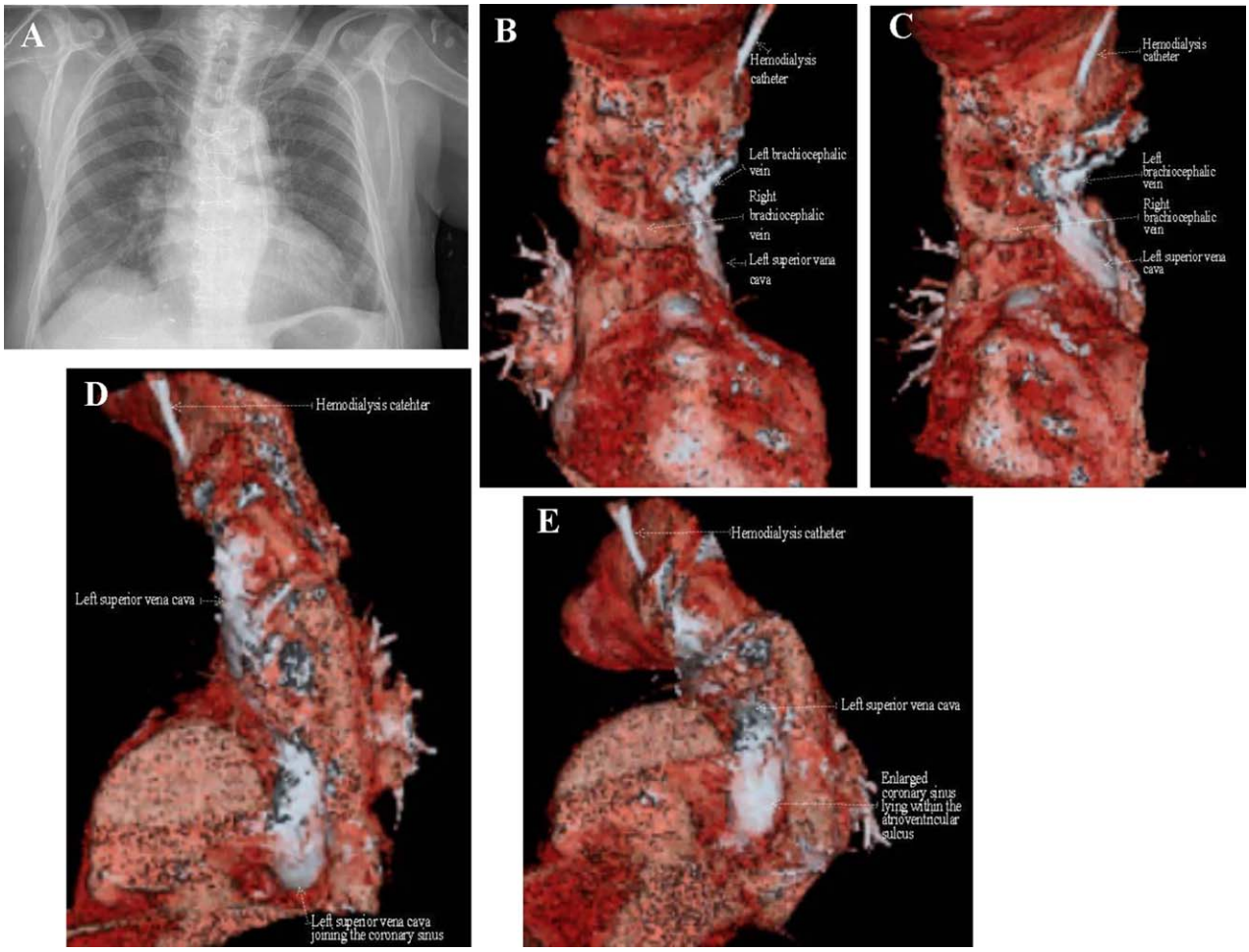

Figure 1 (A) Posteroanterior chest X-ray shows that the left internal jugular vein catheter is descenting straightly on the left parasternal border. (B and C) Anterior and anterolateral views of three-dimensional reconstruction of thorax computed tomography with contrast injection through the left arm show that the right brachiocephalic vein fuses with the left brachiocephalic vein to form the left superior vena cava, and the catheter is seen entering the left internal jugular vein. The right superior vena cava is absent. (D and E) Lateral and inferolateral views show that the left superior vena cava continues to drain into the coronary sinus, which is located within the atrioventricular sulcus and enlarged.

complications associated with hemodialysis catheters, whereas intrathoracic congenital major venous anomalies can be seen in a minority of patients and little is known about its catheter related consequences.[4] We herein present two patients with intrathoracic congenital venous anomalies and review the literature.

Case 1

An 80 year-old male patient with a history of diabetes mellitus, hypertension, coronary artery disease, and continuous ambulatory peritoneal dialysis for 7 months was admitted for acute peritonitis. Appropriate antibiotherapy and clinical management were started promptly, but the patient deteriorated and the peritoneal catheter was removed. Ultrasonography of the right IJV revealed chronic intraluminal thrombosis due to previous catheterization, therefore, the left IJV was used for a nontunneled catheter without any complication. Chest X-ray showed an unusual left parasternal downward course of the catheter, although we confirmed that the catheter was within the vein (dark red blood, blood gases) and in good functioning (Figure 1A). Hemodialysis was performed using this catheter with good blood flow (300–350 mL/min) for 3 months until the maturation of a newly created radiocephalic AVF. A thorax computed tomography (CT) with

contrast infusion into the left arm, which was obtained while an abdominal CT was ordered for the management of peritonitis, revealed a persistent left superior vena cava containing the hemodialysis catheter and absent right superior vena cava (SVC) (Figure 1B–D). Echocardiography confirmed the presence of persistent left superior vena cava (PLSVC) with identification of the left arm infused agitated saline passing through the coronary sinus into the right atrium. The patient was discharged following the resolution of peritonitis, and the hemodialysis catheter was removed after the AVF was ready to use.

Case 2

A 35-year-old male patient with a history of diabetes mellitus, hypertension, and chronic kidney disease was admitted for dyspnea and lower extremity edema. Past and current laboratory analyses and renal ultrasonography indicated that the patient was having end stage renal disease. Right IJV catheter was placed under ultrasound guidance without any complication, and chest X-ray revealed that the catheter was having an unusual course that crossing the mediastinum from right to left (Figure 2A). Blood gas analysis and dark red blood from the catheter confirmed that it was within the vein. A thorax CT was obtained to find out the vascular anomaly, and PLSVC with absent right SVC was identified (Figure 2B–D). The patient had residual renal function; therefore, contrast agent for CT was not given. Echocardiography confirmed the presence of PLSVC with identification of the left arm infused agitated saline passing through the coronary sinus to the right atrium. The hemodialysis catheter provided good blood flow (300–350 mL/min) for 2 months throughout the hemodialysis sessions until the maturation of his new left radiocephalic AVF. The patient was scheduled on ambulatory hemodialysis thrice a week.

DISCUSSION

We presented two cases with PLSVC and absent right SVC, and uncomplicated hemodialysis catheter insertions (both at right and left IJVs) with good catheter functions.

During normal embryonic development of the sinus venosus, which later forms the right atrium, the left anterior cardinal vein fuses with the right cardinal vein, thereby the SVC flourishes on the right side and the left common cardinal vein caudal to the innominate vein obliterates and leaves the oblique vein of the left atrium, coronary sinus and left superior intercostal veins behind.[5] Embryonic development of the sinus venosus with its tributaries and the three-dimensional (3D) nonenhanced CT reconstruction of the normal SVC can be seen in Figure 3. Alterations in fusions or obliterations of embryonic cardinal veins can result in a number of theoretically anticipatable variations in the venous anatomy, such as persistence of the left anterior cardinal vein results in PLSVC which is the most common thoracic venous anomaly.[4] Anomalies of the SVC have been classified into three major categories (duplicated SVC, left SVC, and right SVC), and left SVC is present in less than 0.5% of the general population, but it accompanies to 4% of patients with congenital heart anomalies.[4] Left SVC virtually always drains into the coronary sinus, when it is not a part of congenital heart anomalies.[4]

We have found 20 case reports/series to review regarding venous catheterization for hemodialysis in patients with PLSVC, when the entries "persistent left superior vena cava, hemodialysis" were used in PubMed (Table 1). None of those patients had accompanying congenital heart anomalies, and all cases were asymptomatically discovered during evaluation for vascular procedures. Chest X-ray, contrast venography, CT venography, saline agitated echocardiography, and digital subtraction angiography were the diagnostic tools. Contrast enhancement was used in most of the cases, and this may pose a risk of loss of residual renal function if present. Contrary to the common conception, our experience demonstrates that contrast agents may not be mandatory, as 3D reconstruction of a thorax CT with slicing off of perimediastinal tissues can provide with sufficient anatomical details of the SVC with its tributaries, and the hemodialysis catheter is intrinsically enhanced due to its markedly higher intensity than the background. Nontunneled and/or tunneled hemodialysis catheters were able to be inserted either through the right or left IJVs without any significant difficulties or complications, except for one patient with an absent right SVC who had right subclavian catheter insertion following failed multiple attempts of right IJV catheterization.[6] There were not any vascular or perivascular traumatic complications associated with the procedure of catheter insertions, and only one catheter related complication, which was arrhythmia, was reported.[7] Catheters provided good blood flow in all patients after successful catheter placement throughout the course of case presentations. The durations of catheter use for hemodialysis were not provided in all, but ranged from 4 weeks to 15 months where available.

In everyday clinical practice, patients do not routinely undergo imaging for identification of congenital anomalies of the SVC before central venous catheter placements; such anomalies are recognized during an irrelevant imaging/vascular procedure or after identification of an

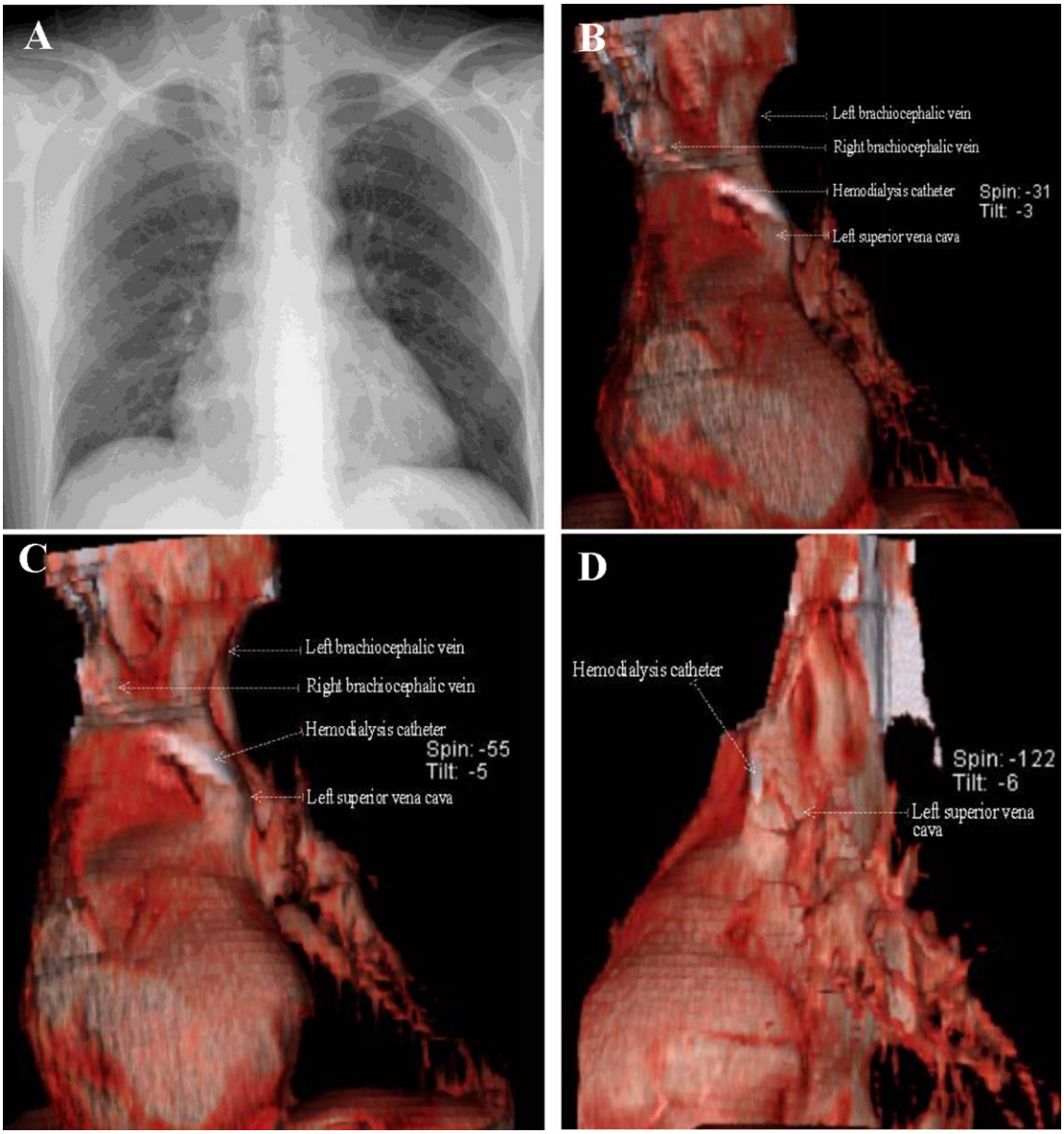

Figure 2 (A) Posteroanterior chest X-ray shows that the right internal jugular vein catheter is crossing the upper mediastinum from right to left. (B–D) Anterior, anterolateral, and lateral views of three-dimensional reconstruction of thorax computed tomography without contrast enhancement show that the right brachiocephalic vein fuses with the left brachiocephalic vein to form the left superior vena cava, and the catheter is spontaneously enhanced within the right brachiocephalic vein and the left superior vena cava; the right superior vena cava is absent.

unusual course of a catheter or guidewire on chest X-ray such as in previous reports and our cases. Furthermore, this issue was not addressed within the most recent guidelines of KDOQI for vascular access in hemodialysis.[1] Therefore, two questions are needed to be answered.

What should be done for a patient with known PLSVC before hemodialysis catheter insertion? In addition to compliance with the guidelines for vascular access, side preference may need to be made according the features of present anomaly, diameters of the right and left IJVs and the innominate veins, and the angles of venous confluences or courses. The previous reports and our two cases indicate that nontunneled or tunneled hemodialysis catheters may safely be inserted in patients with PLSVC, using

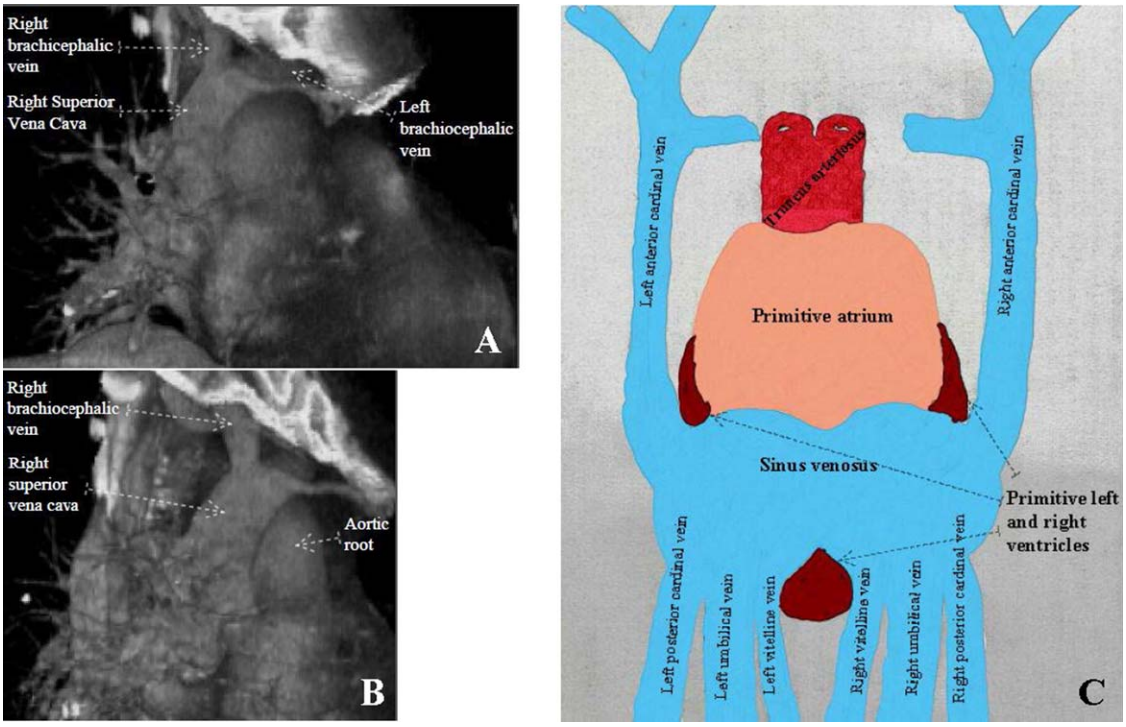

Figure 3 (A and B) Anterolateral (right) and posterolateral (right) views of three-dimensional reconstruction of nonenhanced thorax CT of a patient with normal superior vena cava anatomy. (C) Dorsal view of a representative illustration of the development of sinus venosus at approximately 24 days (the illustration was drawn by the authors as a representative based on the descriptions and figures in the Chapter 11 of Langman's Medical Embryology ninth edition, 2004).[5]

gentle and careful technique (Table 1). One of the technical details to be mentioned is that when an angled vein (innominate vein crossing the mediastinum from right to left or vice versa) is chosen for catheterization, semirigit dilators should be advanced gently and to a level not beyond the anticipated curve of the relevant innominate vein, to prevent venous injury or kink of the guidewire. However, in patients with absent right SVC, left IJV may be the preferred catheter insertion site as it can provide with straight catheter descending and avoid abutting of the catheter to the innominate vein. An important consideration is whether left sided catheter placement can cause coronary sinus thrombosis in patients with PLSVC. Coronary sinus thrombosis is a rare and potentially fatal entity that has been associated with endovascular procedures in most cases according to a review by Yeo et al.[8] PLSVC catheterization has not been reported as a cause of coronary sinus thrombosis, but keeping the catheter tip above the coronary sinus could be an important precaution. Dionisio et al. reported a case with PLSVC who developed clinically significant arrhythmia following catheter placement; hence they suggested that catheters should be removed immediately once PLSVC has been recognized.[7] However, arrhythmia can also occasionally occur in patients with normal venous anatomy of the thorax and it may be due to an inappropriate extension of the catheter tip into the right atrium.[9] In such cases, we attempt a slight withdrawal of the catheter, which has terminated arrhythmia almost in all patients. Furthermore, only one case among 22 (20 plus our 2 cases) patients developed arrhythmia (Table 1). Indeed, the catheter tip in Dionisio's case appears to have reached the right atrium through the coronary sinus, and withdrawal of the catheter to a sufficient level could be expected to terminate arrhythmia. Therefore, in patients who developed arrhythmia but are clinically stable, we suggest that a stepwise withdrawal of the catheter until arrhythmia ceases should be attempted first, instead of removal.

What should be done after successful insertion of a functional hemodialysis catheter in a patient with PLSVC? Once a functionally adequate catheter has been placed without any complication, chest X-ray should be obtained to see the course of the catheter and determine the level of the catheter tip. Significant anxiety may arise if an

Table 1 Anomalous, diagnostic, and outcome characteristics of case reports with persistent left superior vena cava and venous catheterization for hemodialysis

Authors, year	Study type	Number of patients	Additional anatomical variations	Diagnostic method	Type of catheter	Complication during catheter insertion	Catheter related complication	Catheter function	Duration of catheter use
Pautler et al.[10], 1999	Case report	1	None	Chest X-ray	Unknown (left IJV)	No	No	Good	Unknown
Kim et al.[11], 1999	Case report	1	None	Fluoroscopy	Nontunneled (left SCV)	No	No	Good	Unknown
Radovic et al.[12], 2002	Letter to the editor	1	None	Chest X-ray DSA	Nontunneled (left IJV)	No	No	Good	4 weeks
Dionisio et al.[7], 2003	Case report	1	Agenesis of a solitary kidney	Endocavitary electrocardiography Chest X-ray Venography Echocardiography Nuclear magnetic resonance	Unknown (left IJV)	Failed right IJV attempt Arterial puncture of the right carotid artery	Arrhythmia	Good	NA
Kuppusamy et al.[13], 2004	Case report	1	None	Chest X-ray DSA	Nontunneled (left IJV)	No	No	Good	Unknown
1. Wasse et al.[6], 2006	Case reports	2	Absence of RSVC	Venography	None	Failed right internal jugular catheter insertion	Failed right internal jugular catheter insertion	NA	NA
Stylianou et al.[14], 2007	Case report	1	Drainage of pulmonary veins to the posterior aspect of the LA	Chest X-ray Contrast enhanced CT Echocardiography	Nontunneled (left IJV)	None	None	Good	4 weeks
Jang et al.[15], 2009	Case report	1	None	Chest X-ray CT	Nontunneled (left IJV)	None	None	Good	Unknown
Orija et al.[16], 2009	Case report	1	None	Venography	Nontunneled (right IJV)	None	None	Good	Unknown
Parreira et al.[17], 2009	Case report	1	None	Chest X-ray Venography	Nontunneled (left IJV), tunneled (left IJV)	None	None	Good	Unknown
Sriramnaveen et al.[18], 2010	Case report	1	None	Chest X-ray CT	Nontunneled (left IJV)	None	None	Unknown	Unknown
Messina et al.[19], 2011	Case series	1	None	CT venography Digital venography	Tunneled (left IJV)	None	None	Good	15 months onward

Table 1 *Continued*

Authors, year	Study type	Number of patients	Additional anatomical variations	Diagnostic method	Type of catheter	Complication during catheter insertion	Catheter related complication	Catheter function	Duration of catheter use
Lim et al.[20], 2010	Case report	1	Aortic coarctation	Fluoroscopy DSA	Tunneled (left IJV)	None	None	Good	5 months
Kute et al.[21], 2011	Case report	1	None	Chest X-ray Echochardiography Venography CT	Tunneled (left IJV)	None	None	Good	2 months
Wong et al.[22], 2013	Case report	1	None	CT venography	Nontunneled (left IJV)	None	None	Good	Unknown
Kukavica et al.[23], 2014	Case report	1	None	CT angiography	None-tunneled (left IJV)	None	None	Good	4 months
Lui et al.[24], 2014	Case report	1	None	CT venography	Nontunneled (left IJV), tunneled (left IJV)	None	None	Good	6 months
Dubey et al.[25], 2014	Case report	1	Absent RSVC	Chest X-ray Venography Echocardiography	None-tunneled (right IJV)	None	None	Good	Unknown
Jaffer et al.[26], 2015	Case report	1	Horseshoe kidney	Contrast venography CT	None-tunneled (right IJV)	None	None	Good	Unknown

Acronyms: CT = computed tomography; DSA = digital subtraction angiography; IJV = internal jugular vein; LA = left atrium; NA = not applicable; RSVC = right superior vena cava; SCV = subclavian vein.

unusual course of the catheter has been recognized; in this case, if the patient is stable and there is no apparent complication, the initial approach should be to ensure intravenous catheterization by reviewing the steps of insertion carefully and checking the color and pressure of blood drawn from the catheter, simultaneous blood gases analyses from the catheter and peripheral artery. Further imaging by one or more options of contrast venography, CT angiography with contrast injection from both arms, and echocardiography can expose the underlying venous anomaly, where single left SVC and double SVC are the most common.[4] However, 3D reconstruction of thorax CT without contrast enhancement may also provide with satisfying anatomical details as it is demonstrated in Figures 1 and 2, if contrast agents are considered harmful in some patients. The available evidence and experience suggest that catheters should be used and managed as usual in patients with PLSVC, if there is no catheter kink or a major distortion in relevant veins, arrhythmia, pneumothorax, hemothorax, or any other significant complication.

CONCLUSIONS

Thoracic venous anomalies and PLSVC in particular are likely to be encountered at some occasions during the practice of interventional nephrology, despite their rarity. Unexpected course of a hemodialysis catheter while everything is normal should prompt the investigation for thoracic venous anomalies. Evidence suggests that tunneled or nontunneled hemodialysis catheters can be inserted in these patients with good catheter function and no major increase in risk of complications. Adherence to vascular access guidelines and judicious planning with gentle insertion of catheters can be expected to produce the best outcomes.

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