

Epidemiological Characteristics of Superior Vena Cava Syndrome in Patients with Tunneled Catheters in a Civil Pension Hemodialysis Unit in the State of Chihuahua, Mexico.

David Sebastián Paz Berúmen ¹, Maria Fernanda Espinoza Figuera ^{2*}, Johana Paola Vázquez ³, Iván Enrique Herrera Carillo ⁴, Parish Pablo Martínez Pecina ⁵, Jesus Eduardo Adame Garza ⁶

¹Nephrologist, Department of Hemodialysis State Civil Pensions

²Resident Physician of epidemiology 2 years Work area: General Hospital Querétaro ISSSTE

³Medical nephrologist, Work area: Physician assigned to the nephrology area of the Civil Pensions Institute of the state of Chihuahua

⁴Nephrologist: Work area: Physician assigned to the nephrology area of the Civil Pensions Institute of the state of Chihuahua

⁵Resident physician of internal medicine for 2 years Work area: General Hospital B ISSSTE Cd. Juárez Chihuahua

⁶Resident physician of internal medicine for 2 years Work area: General Hospital B ISSSTE Cd. Juárez Chihuahua

Article Info

Received: October 18, 2022

Accepted: November 24, 2022

Published: December 28, 2022

***Corresponding author:** Maria Fernanda Espinoza Figuera, Resident Physician of epidemiology 2 years Work area: General Hospital Querétaro ISSSTE.

Citation: Paz Berúmen DS, Espinoza Figuera MF, Johana P Vázquez, Herrera Carillo IE, Martínez Pecina PP. (2022) "Epidemiological Characteristics of Superior Vena Cava Syndrome in Patients with Tunneled Catheters in a Civil Pension Hemodialysis Unit in the State of Chihuahua, Mexico". International Journal of Epidemiology and Public Health Research, 2(3). DOI: <http://doi.org/11.2022/1.1037>.

Copyright: © 2022 Maria Fernanda Espinoza Figuera. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Abstract:

Superior vena cava syndrome is an entity mainly associated with malignancy, although it is increasingly associated with central catheters and cardiac electronic devices. The incidence in hemodialysis units is not known. Although it is a relatively rare entity, during 2021 a high number of cases were registered that had not been observed in other years. Without being able to establish a direct cause, this study collects data for a year in our hemodialysis unit. An incidence of 7.8% was documented. affected patients had a 60% mortality from all causes, although direct causality with superior vena cava syndrome was not established in any case.

Introduction

Superior vena cava syndrome is a clinical entity that determines significant morbidity and mortality in the patient who suffers from it. It is estimated that in the United States alone it affects 15,000 patients annually [1]. Although the most common cause is of malignant origin in approximately 70% of cases [2], the incidence of this pathology is increasingly associated with the presence of catheters and other cardiac devices [3]. Although the most frequent symptoms are facial edema, non-pulsatile venous congestion, dyspnea and cough [2]; atypical presentations such as Bud-Chiari syndrome [4], upper gastrointestinal bleeding secondary to esophageal varices [5] [6] [7], chylothorax-chylopericardium [8], intracranial hypertension [9], or pulmonary embolism have been reported [10]. On the other hand, the presence of thrombosis associated with venous catheters and pacemakers can be a silent complication with a high prevalence and is potentially dangerous [2] [11].

Although the association of transvenous intracavitary electronic devices such as pacemakers with superior vena cava syndrome is a frequent complication of these, between 31-64% of patients with these devices develop some degree of obstruction [12], in hemodialysis few studies have been described in this regard and the real incidence is unknown. The present work aims to collect the epidemiological data of patients in our hemodialysis unit who are carriers of a tunneled catheter as vascular access and have developed superior vena cava syndrome during the year 2021.

Material and Methods

An analytical cross-sectional study was carried out, using the database of the State Civil Pensions hemodialysis unit in the city of Chihuahua. All patients admitted to the chronic hemodialysis program during the year 2021 and who have a tunneled catheter were included. While the exclusion criteria were patients with arteriovenous fistula, temporary catheters or who have been assigned to the temporary hemodialysis program.



Patients who in the two months prior to the definitive or suspected diagnosis of superior vena cava syndrome had manifested bacteremia or had been diagnosed with a catheter-related infection were also excluded. Those patients who were known carriers of thrombophilia of some kind and previously received anticoagulants were also excluded. Obtaining a sample of 64 cases.

Suspected superior vena cava syndrome was defined as patients presenting facial and/or neck edema, plethora and/or cyanosis. And definitive to patients who had the necessary radiological studies to corroborate total or partial obstruction of the superior vena cava in the electronic file. Discarding patients who, due to lack of radiological studies, could not be classified as definitive. With the confirmed cases, demographic data were collected, as well as hemoglobin levels (Gr/dL), KT/V, vitamin D levels (ng/mL), parathormone (pg/mL), C-reactive protein (mg/L) at the time of diagnosis. Likewise, the information from the radiological studies was collected in order to determine the degree of obstruction of the superior vena cava, if there is dilation of the azygos system and inversion of the flow to it.

Results

A total of 65 patients with tunneled catheters assigned to the chronic hemodialysis program were found with an average age of 65.6 years with a standard deviation of 7.37, with a predominance of males in 60% of cases with a mortality of 60%. Of which, during the year 2021, seven met the definition of suspects, of which one of the cases was ruled out for presenting thrombophilia related to protein S deficiency and who previously took anticoagulants. When evaluating the electronic file, another patient was discarded for not having the radiological studies as they were still pending. Therefore, in the end, only 5 cases were considered in 64 patients with tunneled catheters (7.8%), as shown in Table 1.

Table 1: Incidence of superior vena cava syndrome in patients with tunneled catheters.

Síndrome de vena cava superior	Incidencia	IC 95%	
		Inferior	Superior
Sí	7.8	1.2	14.4

Table 1: Incidence of superior vena cava syndrome in patients with tunneled catheters, showing an incidence of 7.8% with a 95% confidence interval.

With respect to the epidemiological profile, 5 defined cases with superior vena cava syndrome were found, which shared the characteristics that all the cases were diabetics and with arterial hypertension, and the cause of their renal failure was diabetic nephropathy. With age ranges from 54 to 74 years and an average of 65.6 years. Regarding gender, 3 patients were male and 2 female patients. All had at least 6 months with the catheter installed. whose epidemiological characteristics of the cases are described in Table 2.

None of the patients had evidence of uremic syndrome or subdialysis. Hemoglobin figures above 13 gr/dl were not observed, although two of these cases showed an increase in it since they were at levels below 11 gr/dl at the time of diagnosis; however, an average level is presented. of 10.93 gr/dL. All

showed vitamin D deficiency with an incidence of 17.52% with a standard deviation of 7.06%. Four of the patients exhibited levels of uncontrolled secondary hyperparathyroidism with mean levels of 474.60. The levels of C-reactive protein according to the parameters of the laboratory that processed the samples, showed elevation in two cases (cut-off point 10 mg/l). as well as a time of 25.5 months in relation to the placement of the catheter. the clinical profile of the patients broken down is shown in Table 3.

Three patients required angioplasty as part of their treatment, while 2 of them underwent catheter removal and are on anticoagulant treatment with good evolution. With a mortality of 60% of cases as seen in table 4.

As for the tomographic findings, an obstruction of 90% or greater was identified in all cases, as well as significant dilatation of the azygos system in 3 of the cases and in one mild case. The radiological descriptions were not uniform, so only obstruction was reported in most cases, and dilatation of the azygos system was observed. Only in one case was a flow of the contrast medium into the azygos system reported.

Table 2: Epidemiological profile of cases with superior vena cava syndrome.

Caso	1	2	3	4	5
Sexo	F	M	M	M	F
Edad	54	74	69	65	66
Diabetes	Si	Si	Si	Si	Si
Hipertensión	Si	Si	Si	Si	Si
Kt/v	1.36	1.28	1.44	1.29	1.6
Hemoglobina	10.27	10.2	11.5	11.1	11.6
Niveles de vit. D	13	21	26	19.6	8
Niveles de PTH	415	144	577	777	460
PCR	10	10	17	4.6	14
% de obstrucción	¿?	90	¿?	Mayor al 90	¿?
Dilatación del sistema ázigos	Si	No	Leve	Si	Si
Tiempo en meses desde la colocación del catéter y el desarrollo del síndrome	12	6	42	12	54
Tratamiento	Angioplastia	Anticoagulación	Anticoagulación	Angioplastia	Angioplastia
Estatus actual	Finado	Vivo	Vivo	Finado	Finado

Table 2: Epidemiological profile of the cases with superior vena cava syndrome: they found 5 defined cases with superior vena cava syndrome, which shared as characteristics that the total of the case are diabetics and with arterial hypertension, with an average of 65.6 years



Table 3: Clinical and biochemical profile of superior vena cava syndrome in patients with tunneled catheters.

Característica	Promedio	Desviación estándar
Edad	65.6	7.37
Hipertensión		
Diabetes		
Kt/v	1.39	0.13
Hemoglobina	10.93	0.67
Niveles de vit. D	17.52	7.06
Niveles de PTH	474.60	231.84
PCR	11.12	4.69
Tiempo en meses desde la colocación del catéter y el desarrollo del síndrome	25.20	21.38

Table 3: Clinical and biochemical profile of superior vena cava syndrome in patients with tunneled catheters: where it is observed that the average catheter placement and development of the syndrome is 25.20 months.

Table 4: Treatment profile and current status of superior vena cava syndrome in patients with tunneled catheters.

Característica	Prevalencia
Sexo	
Masculino	60.0%
Femenino	40.0%
Dilatación del sistema ázigos	
Sí	60.0%
Leve	20.0%
No	20.0%
Tratamiento	
Angioplastia	60.0%
Anticoagulación y retiro de cateter	40.0%
Estatus actual	
Finado	60.0%
Vivo	40.0%

Table 4.- Treatment profile and current status of superior vena cava syndrome in patients with tunneled catheters: a mortality rate of 60% can be observed, which is directly proportional to angioplasty.

Discussion

The superior vena cava provides venous drainage from the head, arms, and upper chest to the heart and contributes about 30% of cardiac venous return. The obstruction of this can be due to an extrinsic origin such as a tumor at the level of the anterior or middle mediastinum, an inflammatory process or an aneurysm of the aorta, although it can also occur due to an intrinsic occlusion by a thrombus. When the obstruction occurs, the venous return must be diverted through collateral routes towards the inferior vena cava or through the azygos system. This supposes an increase of the venous pressure. For example, a cervical venous pressure of 2 to 4 mm Hg is calculated, and can be increased to 20 or 40 mm Hg. This increased pressure leads to facial edema but can even be severe enough to cause cerebral edema [13].

It is this increase in venous pressure that can cause the clinical manifestations, which have been classified as neurological, facial, laryngeal-pharyngeal, and thoracic-upper extremities to facilitate their description [14].

Based on the venographic findings, the classification of obstruction in 4 degrees by Stanford and Doty was proposed in a series of cases of patients with superior vena cava syndrome of

malignant origin and that was related to survival [15].

Grade I: partial obstruction (more than 90%) of the SVC, azygos patent to the right atrium. Survival 9.3 months.

Grade II: almost complete obstruction (between 90 and 100%) of the SVC, with patency of the azygos system to the right atrium. Survival 6.6 months.

Grade III: almost complete obstruction (between 90 and 100%) of the SVC, with reversed azygos flow. Survival 2.2 months

Type IV: complete obstruction of the SVC and the azygos system. Average survival 12 months.

Yu proposed a severity scale according to symptoms in patients with CVS also of malignant origin [16].

With this same scale, he proposes a treatment algorithm. This algorithm, however, is not applicable to hemodialysis patients since it considers only malignant causes.

In exclusive hemodialysis patients, the reported presentation has been variable, from 2% [11] to 6.15% [17]. However, it is very likely that it is an underdiagnosed entity.

Siegel and Kuker published a series of cases in which they found that only 38% of patients with superior vena cava obstruction reported symptoms compatible with superior vena cava syndrome, of these, only 7% had clinical signs suggestive of the syndrome. ; however, in our case we were able to observe a higher incidence since 90% of the cases dilation of the azygos system was observed; however, the radiological descriptions were not uniform, so only obstruction was reported in most cases, and observed the dilation of the azygos system, so it is necessary to increase the number of observations in future studies.

On the other hand, 62% of the cases did not report symptoms or suggestive clinical findings. Furthermore, when compared with patients with superior vena cava syndrome of neoplastic origin, complete occlusion was found in 85% of hemodialysis patients compared with 33% of patients of malignant origin; concluding that the behavior is more asymptomatic, although mortality was similar in both groups (31% vs 29% of patients on hemodialysis vs of malignant origin, respectively) [18].

In hemodialysis patients, the pathophysiology is related to endothelial damage caused by the catheter, development of microthrombi, and smooth muscle proliferation [19] [20], which can lead to stenosis of the vena cava. However, superior vena cava syndrome may present before stenosis due to thrombotic occlusion. The latter can be effectively treated by removing the catheter and administering anticoagulation; effectiveness has also been demonstrated with intravenous thrombolytic treatment. Although the effectiveness of this treatment decreases with the days after the formation of the thrombus, with the best results before 5 days [21] [22]. We were able to define that in the 40% where the catheter removal and the administration of anticoagulation were performed, survival was more than 90%, being the most viable alternative in our population, however the number of cases is too small to carry out a study. analysis that seeks to determine risk factors. However, it was notorious that patients with high degrees of obstruction and with consequent



dilation of the azygos system died.

Conclusion

Given the low frequency of this complication in hemodialysis units, a multicenter study is required to help determine risk factors and poor prognosis. In this study, it can be seen that most of the cases with dilation of the azygos system present unfavorable outcomes, although a direct relationship cannot be established with the data obtained given the low number of patients. It is also necessary to evaluate the impact that the presence of the syndrome may condition in the future, such as in mortality from any cause or in secondary morbidities.

It should be noted that the present work was motivated by an apparent increase in cases with superior vena cava syndrome, since in the data obtained from 2017-2020, and for which it was decided not to include in the present work, the case report was as low as 1 or none per year. So the question still remains: Is this entity usually underdiagnosed?

Acknowledgements: to the hemodialysis department

Funding sources: not required.

References

1. K. B. Q. S. A. K. R. S. W. D. C. M. Tamir Friedman, «Malignant Venous Obstruction: Superior Vena Cava Syndrome and Beyond,» *Semin Intervent Radiol*, vol. 34, p. 398-408, 2017.
2. I. N. K. R. R. A. R. B. Abdul HussainAzizi, «Superior Vena Cava Syndrome,» *JACC: Cardiovasc Interv*, vol. 13, n° 24, pp. 2896-2910, 2020.
3. B. J. C. ., A. H. L. ., E. S. ., M. S. Peter Zimetbaum, «Lead-Related Venous Obstruction in Patients With Implanted Cardiac Devices: JACC Review Topic of the Week,» *J Am Coll Cardiol*, vol. 25, n° 79(3), pp. 299-308, 2022.
4. S. S. A. Z. L. P. S. C. G. M. Gabriel Stefan, «Budd-Chiari syndrome: An unusual complication of an internal jugular tunneled dialysis catheter,» *J Vasc Access*, n° DOI: 10.1177/11297298211050187, p. online, 1 diciembre 2021.
5. S. S. a. A. S. Hassam Ali, «Recurrent Hematemesis From Downhill Esophageal Varices: A Therapeutic Challenge for Gastroenterologists,» *Cureus*, vol. 13, n° 3, p. e13840, 2021.
6. J. P. R. H. V. A. L. M. L. V. S. R. F. S. S. A. Pablo V Uceda, «Management of Superior Vena Cava Occlusion Causing Bleeding "Downhill" Esophageal Varices,» *J Endovasc Ther*, vol. 28, n° 3, pp. 469-473, 2021.
7. C. T. S. N. R. M. J. A. C. G. A. R. G. J. M. A.-V. J. L. M. G. Guillermo Ontanilla Clavijo, «Downhill varices: an uncommon cause of upper gastrointestinal bleeding,» *Rev Esp Enferm Dig*, vol. 108, n° 7, pp. 440-442, 2016.
8. I. B. J. F. T. J. James Livesay, «Chylothorax and Chylopericardium: A Complication of Long-Term Central Venous Catheter Use,» *Case Rep Pulmonol*, vol. 2019, n° 2019, p. online, 2019.
9. J. L. B. P. H.-S. K. S.-W. C. S.-H. K. Hyon-Jo Kwon, «Intracranial Venous Hypertension Induced by Superior Vena Cava Syndrome Mimicking Cavernous Dural Arteriovenous Fistula,» *World Neurosurg*, vol. epub2018, n° 2018, pp. 265-268, 2018.
10. P. B. T. B. B. K. Sritika Thapa, «Hemodialysis catheter-associated superior vena cava syndrome and pulmonary embolism: a case report and review of the literature,» *BMC Res Notes*, vol. 23, n° 9, p. 233, 2016.
11. M. D. S. A. S. S. A. N. K. F. C. Martin Garcia-Nicoletti, «Silent and dangerous: catheter-associated right atrial thrombus (CRAT) in children on chronic haemodialysis,» *Pediatr Nephrol*, vol. 36, n° 5, pp. 1245-1254, 2021.
12. D. J. S. J. B. T. M. K. M. A. D. M. L. S. a. P. Z. Andrew H. Locke, «Lead-associated Superior Vena Cava Syndrome,» *J Innov Card Rhythm Manag.*, vol. 12, n° 4 doi: 10.19102/icrm.2021.120404, pp. 4459-4465, 2021.
13. F. C. D. J. Y. Lynn D. wilson, «Superior Vena Cava Syndrome with Malignant Causes,» *N Engl J Med*, vol. 356, n° 1, pp. 1862-9, 2007.
14. K. B. Q. S. A. K. R. S. W. a. D. C. M. Tamir Friedman, «Malignant Venous Obstruction: Superior Vena Cava Syndrome and Beyond,» *Semin Intervent Radiol.*, vol. 34, n° 4, pp. 398-408, 2017.
15. W. D. D. B. Stanford, «The Role of Venography and Surgery in the Management of Patients with Superior Vena Cava Obstruction.,» *The Annals of Thoracic Surger*, vol. 41, n° 2, pp. 158-163, 1986.
16. L. D. W. a. F. C. D. James B. Yu, «Superior Vena Cava Syndrome—A Proposed Classification System and Algorithm for Management,» *journal of thoracic oncology*, vol. 814, p. 811, 2008.
17. F. G. P. M. P. O. J. T. S. G. B. A. Ana Isabel Morales García}, «Incidencia de síndrome de vena cava superior en el mayor centro de hemodiálisis de la provincia de Granada en un año,» *j. dialis*, vol. 36, n° 2, p. 56, 2015.
18. R. K. Yoel Siegel, «Superior Vena Cava Obstruction in Hemodialysis Patients: Symptoms, Clinical Presentation and Outcomes Compared to Other Etiologies,» *Ther Apher Dial*, vol. 20, n° 4, pp. 390-393, 2016.
19. B. M. P. N. J. H. Anil K Agarwal, «Central vein stenosis: a nephrologist's perspective,» *Semin Dial*, vol. 20, n° 1, pp. 53-62, 2007.
20. A. K. Agarwal, «Central Vein Stenosis,» *Am J Kidney Dis*, vol. 61, n° 6, pp. 1001-1015, 2013.
21. J. W. O. R. A. G. J. R. Y. J. R. B. W. F. R. B H Gray, «Safety and efficacy of thrombolytic therapy for superior vena cava syndrome,» *Chest*, vol. 99, n° 1, pp. 54-59, 1991.
22. T. K. a. Z. I. Jie Cui, «Catheter-directed Thrombolysis in Acute Superior Vena Cava Syndrome Caused by Central Venous Catheters,» *Semin Dial*, vol. 28, n° 5, pp. 548-551, 2015.