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CASE REPORT

Double superior vena cava: presentation of two cases and review of the literature

Christos Farazi-Chongouki[a,b], Ioannis Dalianoudis[a,c], Anestis Ninos[b], Pantelis Diamantopoulos[a,d], Dimitrios Filippou[a], Stefanos Pierrakakis[b] and Panagiotis Skandalakis[a]

[a]Department of Anatomy and Surgical Anatomy, Medical School, National and Kapodestrian University of Athens, Athens, Greece; [b]Department of Surgery, "Thriasio" General Hospital, Athens, Greece; [c]Department of Plastic Surgery, "Thriasio" General Hospital, Athens, Greece; [d]Department of Plastic Surgery, General Anticancer-Ongologic Hospital "Agios Savvas", Athens, Greece

ABSTRACT

Introduction: Various anomalies in the development of the great thoracic veins of the embryo can be incidentally discovered in the normal adult. Duplication of superior vena cava (SVC) is a rare abnormality, but the most common thoracic venous congenital anomaly.

Case reports-methods: We present two cases in the intensive care unit of our hospital, of asymptomatic patients who underwent an uneventful central line placement in the left subclavian vein. The track of the catheter, as shown in the X-ray, was misplaced to the left of the aorta and further investigation with computed tomography angiography confirmed a persistent left SVC. In both cases the vein drained into the coronary sinus and then to the left atrium. In the second case the echocardiography revealed a dilated coronary sinus.

Conclusions: Double SVC can be fortuitously discovered during catheter insertion, thoracic or cardiac imaging and surgery. In most cases it drains into the left atrium, through the coronary sinus. This entity is significant to the physician because of its importance in differential diagnosis as a cause of a widened mediastinum, as well as any difficulty that can occur in the placement of a central venous catheter or a pace maker.

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Left superior vena cava; coronary sinus; congenital venous anomalies; widened mediastinum

Introduction

Congenital abnormalities of superior vena cava (SVC) can present as incidental findings either in childhood, be related to underlying cardiac conditions or be syndromic in origin [1]. During the eight week of the embryological period, an oblique anastomosis is formed between the two anterior cardinal veins, which becomes the left brachiocephalic vein. Then the caudal part of the left anterior cardinal vein occludes. Right anterior cardinal vein and right common cardinal vein form SVC [2].

The SVC drains blood from the head and the upper limbs, into the right atrium. Normally, the right side is drained through the right brachiocephalic vein, whereas the left side through the left brachiocephalic vein. In some instances, a double SVC may be encountered. These vessels are referred to as the right and the left SVC [3].

Persistent SVC is a rare anomaly with a prevalence of 0.3–0.5% in general population. On the contrary, this percentage varies between 10–11% in patients with congenital heart disease [4]. In most cases a persistent left vena cava drains into the right atrium, through the coronary sinus [5]. We hereby present two cases of a double superior vena cava and a short review of the existing literature.

Case 1

A 62-year-old woman was admitted to the intensive care unit (ICU) of our hospital intubated, under sedation and mechanical ventilation, because of spontaneous subarachnoid hemorrhage. The patient underwent an uneventful central vein catheterization in her left subclavian vein. The X-ray revealed an abnormal position of the catheter, which appeared to descend into the aorta (Figure 1). A chest computed tomography was performed with 3D reconstruction and revealed the catheter tracking into a left superior vena cava with the tip inside the coronary sinus, which was confirmed with intravenous contrast (Figures 2–4). Further investigation with echocardiography did not reveal any other cardiac anomalies.

CONTACT Christos Farazi-Chongouki cfarazi@ yahoo.gr Department of Anatomy and Surgical Anatomy, Medical School, National and Kapodestrian University of Athens, Plateia Thoukididou 3, Alimos, Athens, Greece

Case 2

A 58-year-old woman was immediately intubated and transferred to the ICU of our hospital, after a pulmonary infection and hypoglycemic coma. A guided-wire central catheter was placed in the left subclavian vein. The X-ray showed an increased cardiac index and a misplacement of the catheter left to the aorta (Figure 5). A computed tomography angiography (CTA) revealed a left superior vena cava with an uncertain track. Because of an

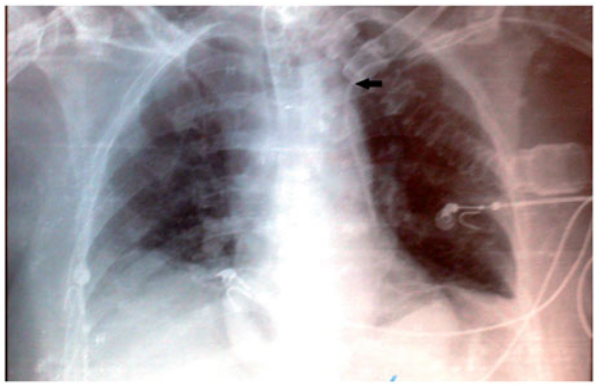

Figure 1. Central vein catheter tracking left to the aorta. Widened mediastinum. The arrow indicates the presence of the catheter.

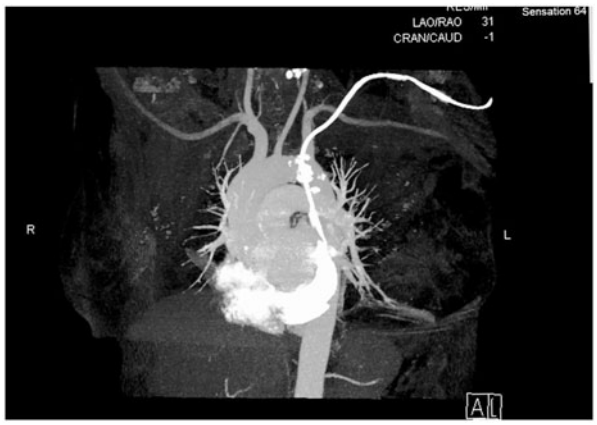

Figure 2. Computed tomography angiography (CTA). The central line catheter inside the left superior vena cava. Injection of a contrast media through the catheter reveals the tip inside the coronary sinus.

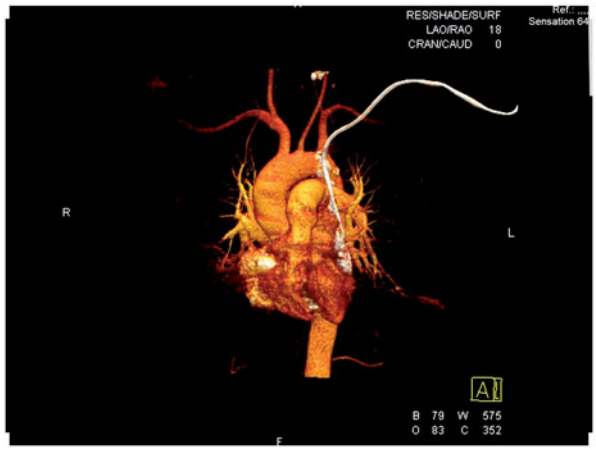

Figure 3. The CTA in Figure 2 depicts a more detailed view of the contrast media draining into the coronary sinus and the right atrium.

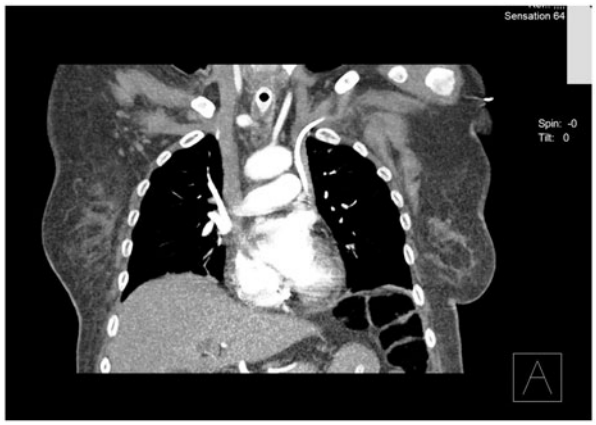

Figure 4. A coronal view. Absence of the left innominate vein. The central vein catheter inside the persistent left superior vena cava.

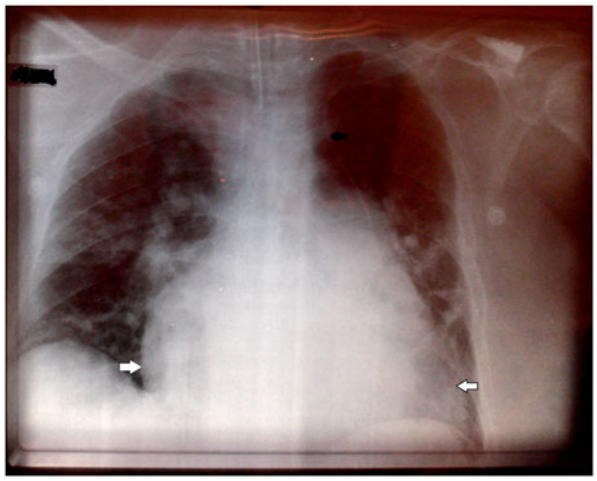

Figure 5. Widened mediastinum. Increased cardiac index (white arrows). The central vein catheter is misplaced to the left of the mediastinum (black arrow).

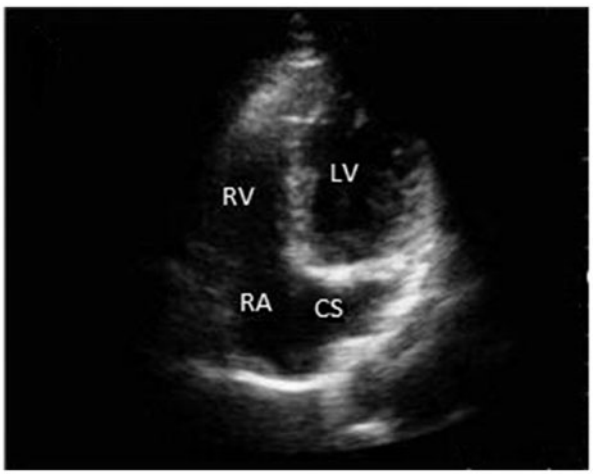

Figure 6. Echocardiography. Dilated coronary sinus (CS). RA: right atrium; RV: right ventricle; LV: left ventricle.

unexplained sinus tachycardia, a cardiac ultrasound was performed. The patient suffered from a serious pulmonary hypertension with a dilated coronary sinus (Figure 6). The patient's status deteriorated and no further intervention could be accomplished.

Discussion

The cardinal veins include the anterior cardinal vein (draining the cephalic portion of the body) and the posterior cardinal vein (draining the remainder of the body of the embryo). The anterior and posterior cardinal veins on each side join to

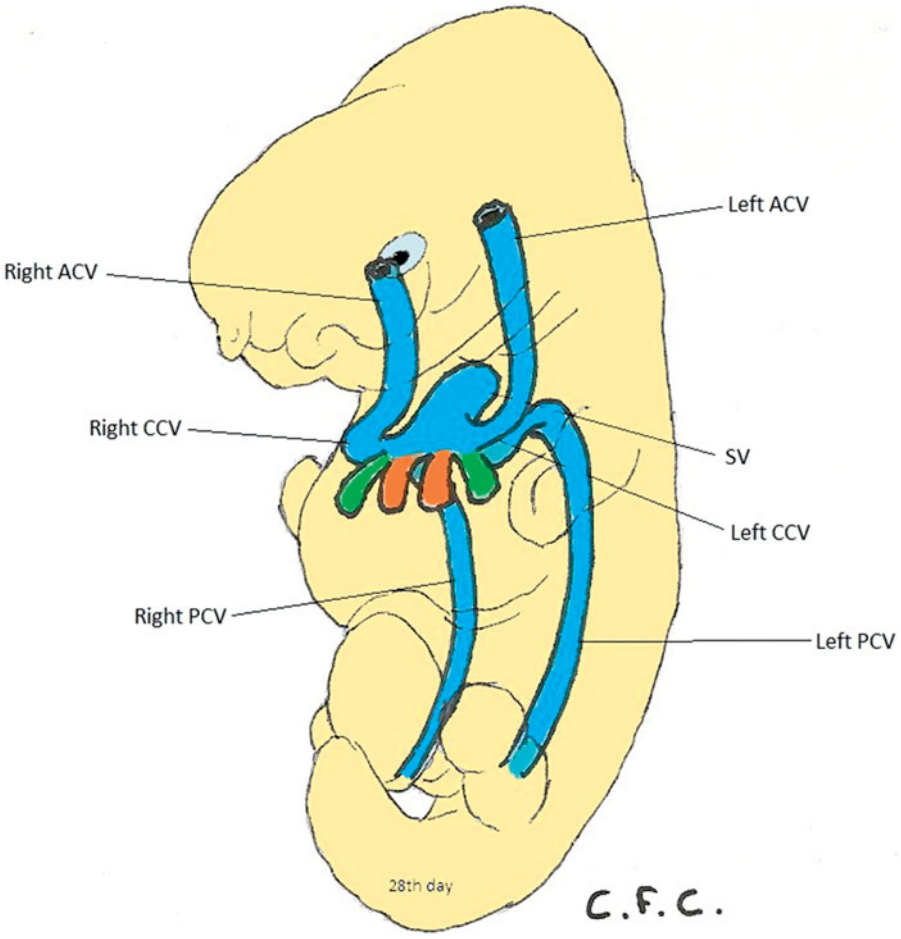

Figure 7. The cardinal veins on the day 28 of embryo . ACV: anterior cardinal vein; CCV: common cardinal vein; PCV: posterior cardinal vein; SV: sinus venosus.

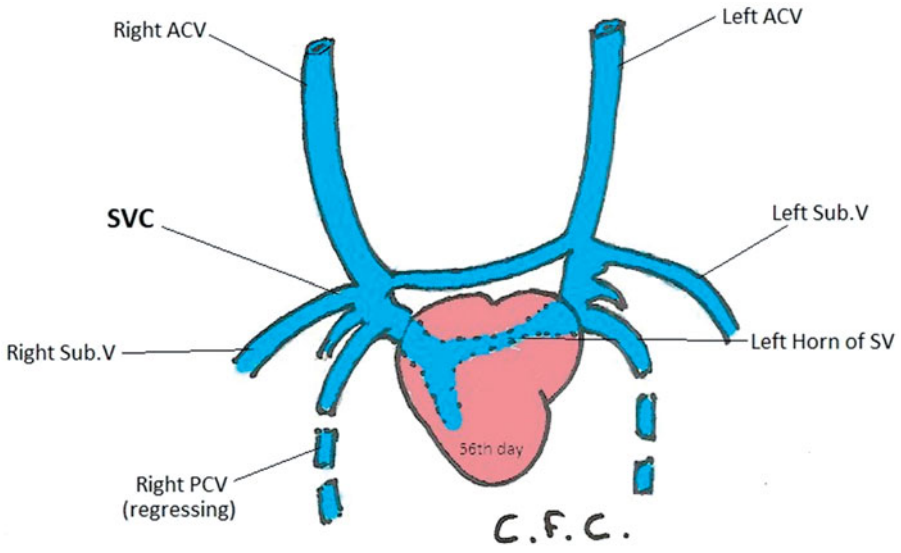

Figure 8. The rise of SVC. ACV: anterior cardinal vein; Sub.V: subclavian vein; PCV: posterior cardinal vein; SVC: superior vena cava; SV: sinus venosus.

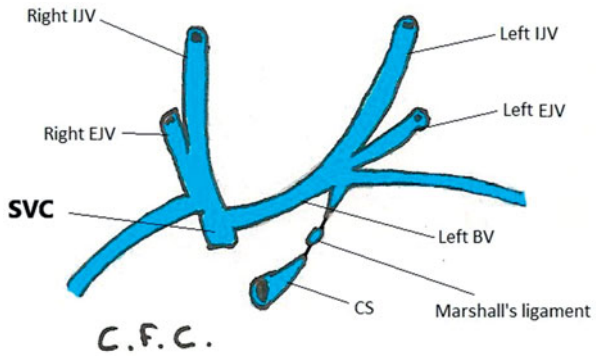

Figure 9. The final development of the venous system . IJV: internal jugular vein; EJV: external jugular vein; BV: brachiocephalic vein; CS: coronary sinus.

form the common cardinal vein before entering the sinus venosus (Figure 7).

The right anterior and common cardinal veins normally give rise to the SVC. An anastomosis that forms between the right and left anterior cardinal veins become the left innominate (brachiocephalic) vein. The anterior cardinal veins regresses except for a small terminal portion that persist as superior intercostals veins (Figure 8) [6]. When the left anterior cardinal vein regresses, a ligament remains that joins the left superior intercostals vein with the coronary sinus. It is called the ligament of left SVC or Marshall's ligament (Figure 9). If the left

Table 1. Cases reported with double SVC (persistence left SVC).

Year	Author(s)	References
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1953	Odman P.	Acta radiol. 1953 Dec;40(6):554–60.
1953	Steinberg et al.	November 1953Volume 24, Issue 5, Pages 479–488.
1975	Stevens JS et al.	J Nucl Med. 1975 Jun;16(6):469.
1984	Lembo CM et al.	Angiology. 1984 Jan;35(1):58–62.
1986	Yoshida K.	Anat Anz. 1986;161(5):397–403.
1990	Mori C et al.	Jpn Heart J. 1990 Nov;31(6):881–8.
2000	Sarodia BD et al.	Respir Care. 2000 Apr;45(4):411–6.
2004	Waikar HD et al.	J Cardiothorac Vasc Anesth. 2004 Jun;18(3):332–5.
2005	Alhaj EK et al.	J Am Soc Echocardiogr. 2005 May;18(5):483–5.
2005	Singh B et al.	Clinical Anatomy Volume 18, Issue5, july 2005, Pages 366–369
2006	Albay S et al.	Morphologie. 2006 Mar;90(288):39–42.
2006	Ratliff HL et al.	Int J Cardiol. 2006 Nov 10;113(2):242–6. Epub 2005 Nov 28.
2008	Goyal SK et al.	Cardiovasc Ultrasound. 2008 Oct 10;6:50. doi: 10.1186/1476–7120-6-50.
2009	Miraldi F et al.	Int J Cardiol. 2009 Jan 9;131(2):e78–80. Epub 2007 Aug 10.
2010	Sharma OP et al.	Asian Journal of Medical Sciences Vol.1(1) 2010 p.18–19
2010	Ucar O et al.	Cardiovasc J Afr. 2010 May-Jun;21(3):164–6.
2011	Shenoy M et al.	J Clinic Case Reports 2012 1:102
2011	Iimura A et al.	Okajimas Folia Anat Jpn. 2011 May;88(1):37–42.
2012	Duymus M et al.	Pol J Radiol. 2012 Oct-Dec; 77(4):65–66.
2012	Middlebrooks E et al.	J Cardiovasc Comput Tomogr. 2012 Jul-Aug;6(4):289–91.
2013	Cooper CJ et al.	Am J Case Rep. 2013 Oct 2;14:395–7
2014	Enuh H et al.	The Internet Journal of Endovascular Medicine 2014 Volume 2 Number 1
2014	Mabry VL.	The Journal of the Association for Vascular Access. June 2014 Volume 19, Issue 2, Pages 84–85
2014	Malik A et al.	American Journal of Respiratory and Critical Care Medicine 2014;189:A6192
2014	Mneimneh S et al.	International Journal of Clinical Trials 2014 Nov;1(3):114–116
2015	Bolognesi M et al.	J Integr Cardiol, 2015,Volume 1(5):115–117
2015	Lee HJ et al.	Korean Journal of Critical Care Medicine 2015; 30(1):22–26.
2015	Morgan LG et al.	Hindawi Publishing Corporation Case Reports in Medicine Volume 2015, Article ID 198754
2015	Rossi UG et al.	J Vasc Access. 2015 Jul-Aug;16(4):265–8
2016	Bernardes MVAA et al.	J. vasc. bras. vol.15 no.2 Porto Alegre Apr./June 2016
2016	Lalenis C et al.	Journal of Medical Cases Volume 7, Number 7, July 2016, pages 253–257
2016	Sahuloglu T et al.	Hemodial Int. 2016 Jul;20(3):369–77.
2016	Sarsenov D et al.	J Breast Health. 2016 Oct; 12(4):177–179.
2016	Earl Choi Y et al.	Iran J Pediatr. 2016 May 15;26(3):e4692
2016	Rawal G et al.	J Clin Diagn Res. 2016 May;10(5):OD17-8
2017	Ricciardi B. et al.	J Vasc Access. 2017 Sep 11;18(5):e66–e69
2017	Bisoyi S. et al.	Ann Card Anaesth. 2017 Jan-Mar; 20(1):104–107.
2017	Nagasawa H et al.	Pediatr Cardiol. 2017 Nov 27
2017	Kemal HS et al.	Case Rep Cardiol. 2017;2017:9842524
2017	Rabinowitz EJ et al.	World J Pediatr Congenit Heart Surg. 2017 Jan 1:2150135117701377
2017	Pardinas Guitierrez MA	BMJ Case Rep. 2017 Jul 31;2017. pii: bcr-2017-220133
2017	Tyrak KW et al.	Cardiovasc J Afr. 2017 May 23;28(3):e1–e4
2017	Sundhu et al.	Cureus. 2017 Jun; 9(6):e1311.
2017	Park SY et al.	Medicine (Baltimore). 2017 May; 96(19):e6803.
2017	Rio PP et al.	Clin Case Rep. 2017 May; 5(5):587–590.
2017	Jazayeri MA et al.	Cureus. 2017 Feb; 9(2):e1057.
2017	Shafiepour DS et al.	Anesthesiology 9 2017, Vol.127, 566
2017	Sonawane NB et al.	A A Case Rep. 2017 Jun 15;8(12):330–333.
2017	Blair SG et al.	Am Surg. 2017 Mar 1;83(3):76–77.
2017	Deshpande R et al.	Anesthesiology 7 2017, Vol.127, 165
2017	Screiber C et al.	Ann Thorac Surg. 2017 Feb;103(2):e161–e162.
2017	Mandolfo S et al.	J Vasc Access 2017; 18(3):e30 – e30
2018	Demeyere M et al.	Diagn Interv Imaging. 2018 Jan;99(1):47–48

innominate vein fails to develop, the left anterior cardinal vein persists and continues to drain the left brachiocephalic veins. In this situation, it becomes the left SVC. The persistent SVC usually drains into the sinus venosus, which ultimately becomes the coronary sinus [7].

The clinical significance of left SVC varies and depends on the existence or lack of symptoms and other congenital heart disease. It can affect the clinicians in imaging diagnosis, central venous catheterization and surgery. Since McCotter first reported three cases in 1916 [8], there were several cases published (Table 1). Steinberg et al. suggested a classification of persistence left superior vena cava into three groups: (1) bilateral superior vena cava without congenital cardiac anomalies, (2) bilateral superior vena cava with associated congenital cardiac anomalies and (3) absence of the right superior vena cava [8].

In our case reports, the central line catheter was seen on the left side of the mediastinum, which was also widened. To an inexperienced physician this can lead to false conclusions, even to the assumption of a major complication. The stability of the patients allowed further investigation with CT scan. In both cases the left innominate vein was absent. Webb et al. reported that double SVC in most cases was associated with regression of the left innominate vein [9]. Widened mediastinum, can be incidentally found in adults and duplication of SVC should be considered as a differential diagnosis [10].

When a dilated coronary sinus is revealed in echocardiography, it may imply the presence of a persistent SVC [11]. Cooper et al. suggested that the gold standard for imaging of persistent left SVC is invasive angiography [12]. However, Ucar et al. proposed 3D reconstructions of CT images for better visualization, as it was performed in our first case [13].

Conclusions

Duplication of SVC is a rather rare entity but extremely important to the awareness of any physician. It can be suspected in several cases when imaging findings contradict the clinical presentation of the patient. Thus, in asymptomatic patients, a widened mediastinum, a dilated coronary sinus or a misplacement of a left internal jugular or subclavian venous catheter in the X-ray, may raise the suspicion of the presence of a left superior vena cava. In that case, there might be technical difficulty in placement of a catheter from the left side of the patient. Further investigation with CTA imaging and echocardiography is essential for diagnosis.

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Consent for publication

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ORCID

Christos Farazi-Chongouki http://orcid.org/0000-0001-7646-167X
Ioannis Dalianoudis http://orcid.org/0000-0003-3658-205X
Pantelis Diamantopoulos http://orcid.org/0000-0003-2823-5496
Dimitrios Filippou http://orcid.org/0000-0001-5410-3046

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