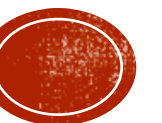




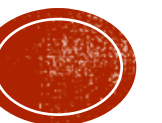
**DES ŒDÈMES BIEN
MYSTÉRIEUX
(OU LA BOITE À SURPRISE)**

Marco Lefebvre, R4 MIG, UdeS



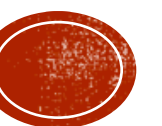
CONFLIT D'INTÉRÊTS

✦ Aucun



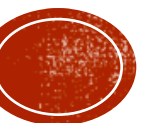
4 ÉTAPES DE L'ÉVOLUTION

- ✿ Présentation du cas
- ✿ Revue de littérature sur l'anomalie vasculaire diagnostiquée
- ✿ Évolution du cas clinique
- ✿ Questions



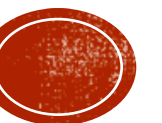
PRÉSENTATION INITIALE

- ✦ Septembre 2013
- ✦ 46 ans
- ✦ TVP iliaque gauche extensive idiopathique



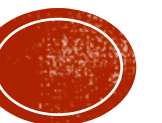
PRÉSENTATION INITIALE

- ✦ Septembre 2013
- ✦ 46 ans
- ✦ TVP iliaque gauche extensive idiopathique
- ✦ Alopécie
- ✦ Leucopénie profonde
- ✦ Photosensibilité



PRÉSENTATION INITIALE

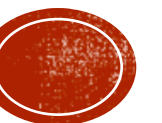
- ✦ Complément d'investigation :
- ✦ ANA 1:320 homogène
- ✦ DS-DNA+, C3 N, C4 bas
- ✦ LAC +, anti-B2-GP1 +, anti-cardiolipine très hauts titres



PRÉSENTATION INITIALE

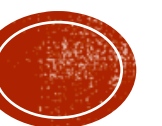
- ✦ Complément d'investigation :
- ✦ ANA 1:320 homogène
- ✦ DS-DNA+, C3 N, C4 bas
- ✦ LAC +, anti-B2-GP1 +, anti-cardiolipine très hauts titres

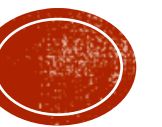
Dx LED \Rightarrow Coumadin, Plaquenil



SURPRISE NO.1

- ✿ 6 janvier 2015
- ✿ Lésions douloureuses bleutées sur les orteils bilatéralement

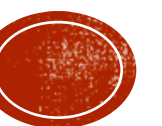




SURPRISE NO. 2

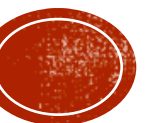
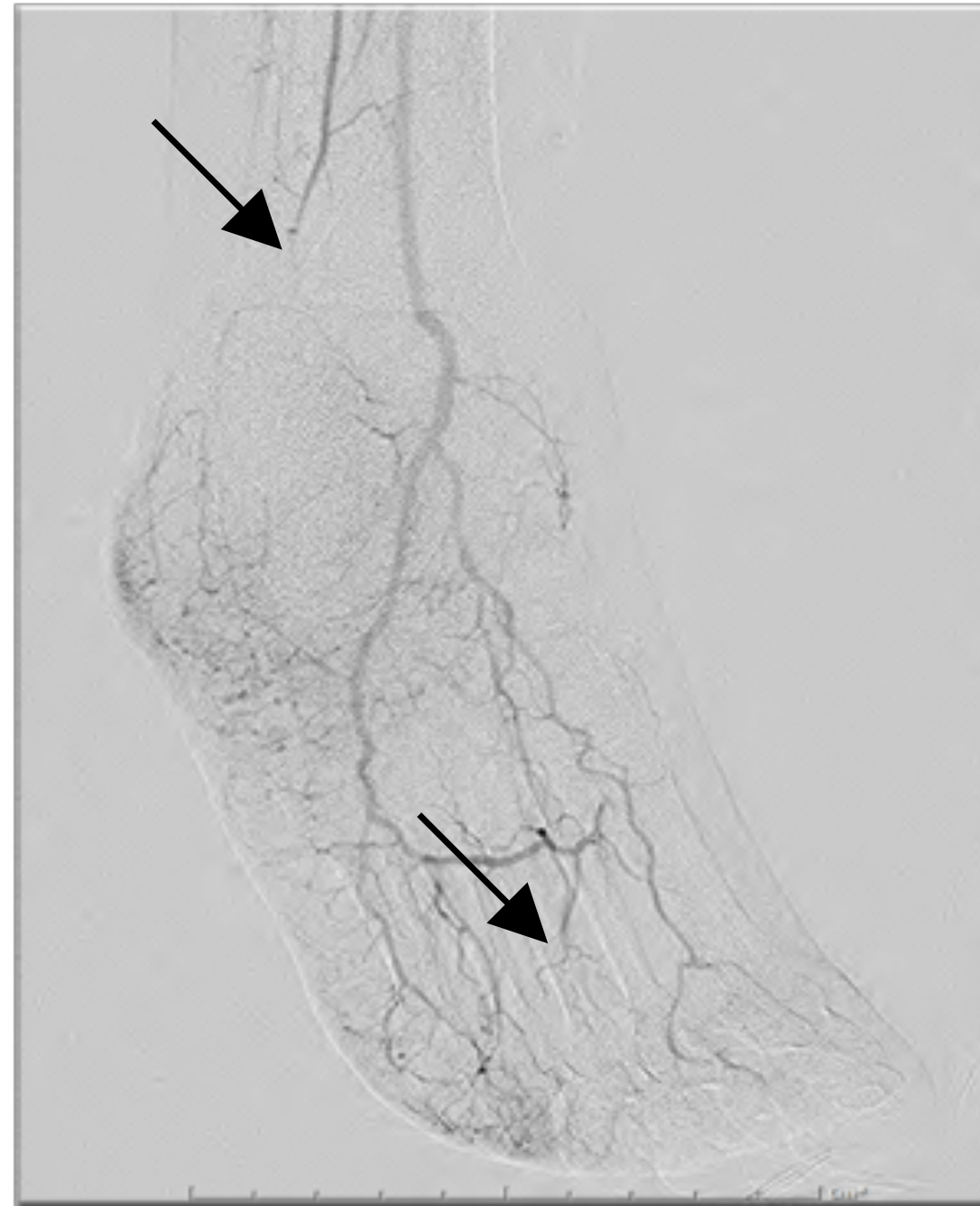
§ Février 2015

§ Aggravation marquée du
tableau avec début de
nécrose des orteils



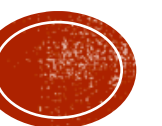
SURPRISE NO. 2

- § Février 2015
- § Aggravation marquée du tableau avec début de nécrose des orteils
- § Artériographie = vasculite petits moyens Vx
- § Prednisone 30mg débuté
- § Ajout d'ASA



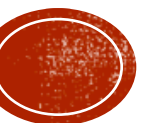
SURPRISE NO. 3

- * Février 2015
- * Ménométrorragies importantes, HB 55
- * Transfusions, Dépot Lupron
- * Fragmin ad résultat Bx endomètre



SURPRISE NO. 4

- ✿ 25 février 2015
 - ✿ Sous Coumadin,
Fragmin en pont et
ASA
 - ✿ Choc, douleur
abdominale, IRA
Creat ad 500



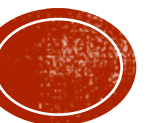
SURPRISE NO. 4

- ✦ 25 février 2015
- ✦ Sous Coumadin, Fragmin en pont et ASA
- ✦ Choc, douleur abdominale, IRA
Creat ad 500



ÉTIOLOGIE?

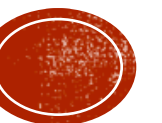
- ✿ Anticoagulation et antiplaquettaire + probable
- ✿ Vasculite abdominale ?
- ✿ Diathèse hémorragique ?
- ✿ Stabilité hémodynamique, IRA persistante
 - ✿ Pas d'angiographie durant l'épisode de soins



ÉTIOLOGIE?

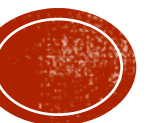
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- ✿ Diathèse hémorragique ?
- ✿ Stabilité hémodynamique, IRA persistante
- ✿ Pas d'angiographie durant l'épisode de soins

Pulse de solumédrol, cyclophosphamide po & coumadin



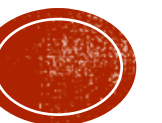
ÉVOLUTION

- ✿ Bonne évolution
- ✿ Résolution de la vasculite des membres inférieurs
- ✿ Résolution du tableau abdominal
- ✿ Pas de récurrence de saignement
- ✿ Reprise de l'anticoagulation per hospitalisation



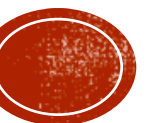
ÉVOLUTION

- ✿ 6 mois cyclophosphamide puis mycophénolate mofétil
- ✿ Coumadin INR thérapeutique stable
- ✿ Pas de resaignement
- ✿ Sevrage de prednisone en 6 mois



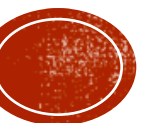
SURPRISE NO. 5 !! 1 AN PLUS TARD...

- ✿ Février 2016
- ✿ Œdème des MI à godet ad bassin
- ✿ Claudication veineuse
- ✿ Pas de prednisone x plusieurs mois



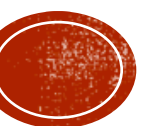
SURPRISE NO. 5 !!

- ✦ Février 2016
- ✦ Œdème des MI à godet ad bassin
- ✦ Claudication veineuse
- ✦ Pas de prednisone x plusieurs mois
- ✦ Doppler MI -
- ✦ Doppler abdo = veine cave dilatée et visualisation d'une structure semblant obstruer le flot à la jonction entre l'oreillette droite et la veine cave



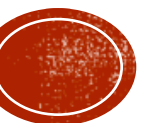
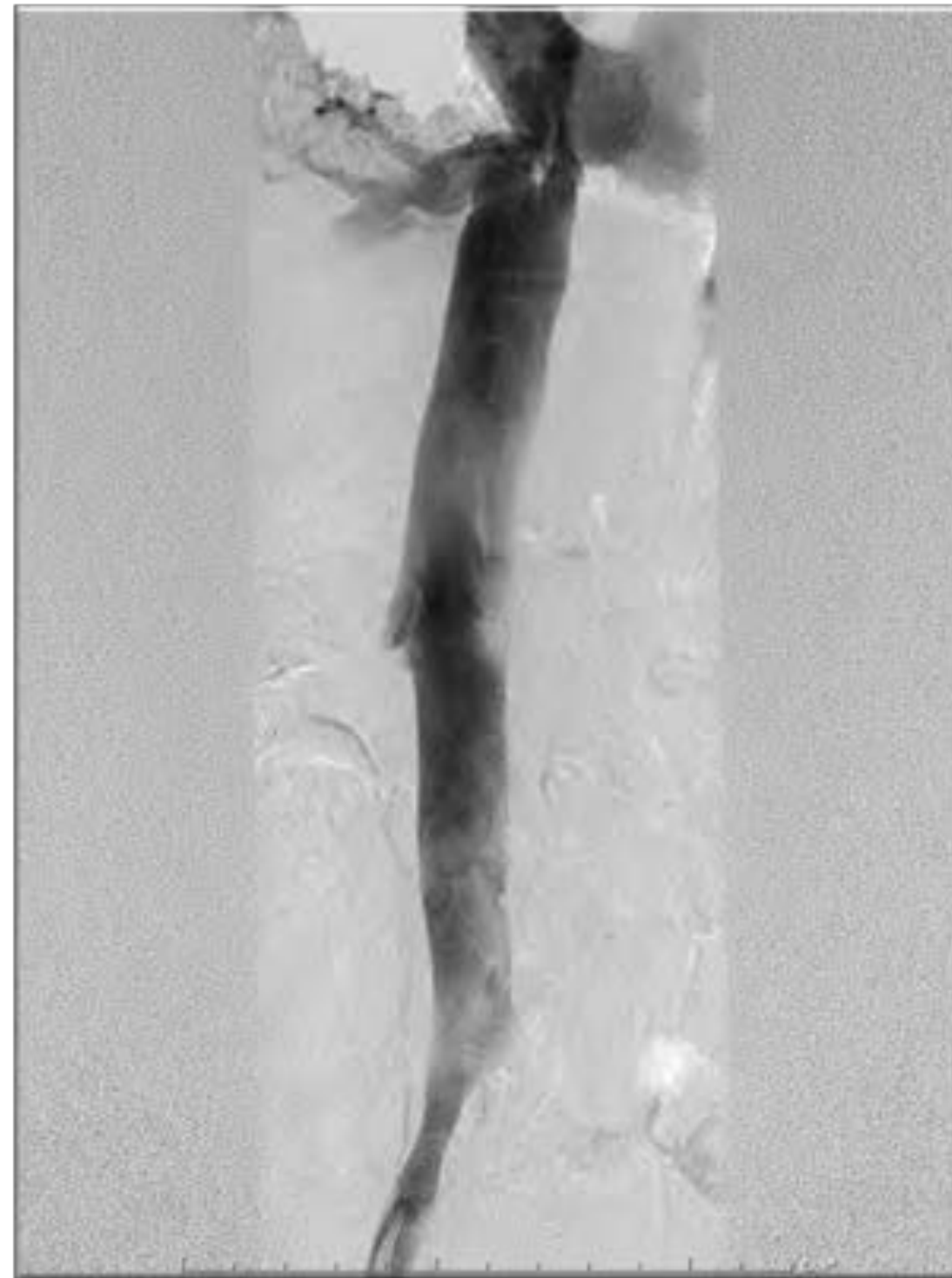
IRM T-A G+

- Lésion caractérisée par IRM = densité tissulaire (pas thrombus) intra-luminale
- Pas de compression extrinsèque; pas de signe de malignité



CAVOGRAPHIE

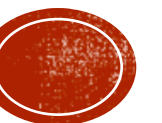
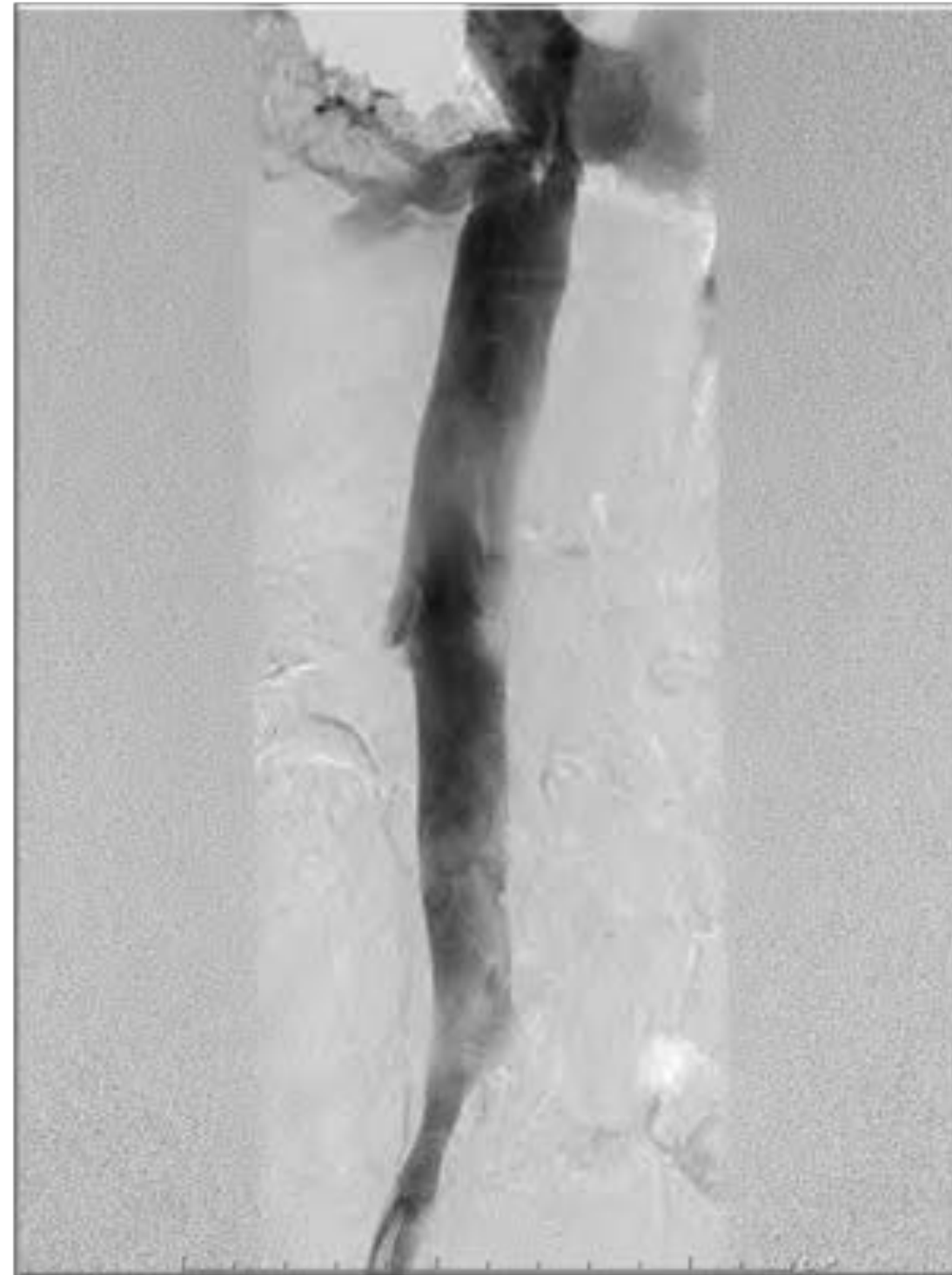
- ✦ Février 2016
- ✦ Membrane à la jonction cavo-cardiaque
- ✦ Reflux dans veine hépatique avec veine cave dilatée
(contribution aux hémorragies abdominales?)
- ✦ Gradient 13mmHg



CAVOGRAPHIE

- ✦ Février 2016
- ✦ Membrane à la jonction cavo-cardiaque
- ✦ Reflux dans veine hépatique avec veine cave dilatée (contribution aux hémorragies abdominales?)
- ✦ Gradient 13mmHg

- ✦ Angio-radiologiste : On dilate-tu ? Ca va bien aller !
- ✦ Interniste : Non ! Cette patiente est une bombe ! INR 3 !



REVENONS À NOTRE CAS CLINIQUE

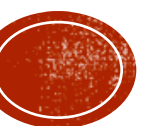
§ En résumé...

§ Femme de 49 ans

§ ATCD de LED et SAPL triple positif avec évènements thrombotiques anticoagulée

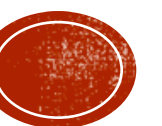
§ Se présente avec symptômes de congestion veineuse invalidants aux membres inférieurs

§ Et découverte d'une membrane sténosante au niveau de la veine cave inférieure avec un gradient de 13mmHg



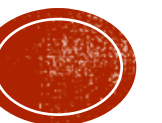
QUESTIONS ??

- ✿ Quelle est l'étiologie de cette sténose ?
- ✿ Quelle est l'approche thérapeutique optimale ?



QUESTIONS ??

- ✿ Quelle est l'étiologie de cette sténose ?
- ✿ Quelle est l'approche thérapeutique optimale ?



Topic Outline

[SUMMARY & RECOMMENDATIONS](#)

[INTRODUCTION](#)

[ETIOLOGY](#)

- [Myeloproliferative disorders](#)
- [Malignancy](#)
- [Infections and benign lesions of the liver](#)
- [Oral contraceptives and pregnancy](#)
- [Other hypercoagulable states](#)
- [Behçet's syndrome](#)
- [Membranous webs](#)
- [Miscellaneous](#)
- [Idiopathic](#)

Membranous webs — Membranous obstruction (partial or complete) of the inferior vena cava (MOVC) and/or the hepatic veins is an unusual but potentially treatable cause of the Budd-Chiari syndrome in the United States [64], but is much more common in South Africa, India, and Asia [65,66]. These web-like lesions, which usually are found just cephalad to the entrance of the right hepatic vein into the inferior vena cava, may be the result of a congenital anomaly. However, they are more often attributable to an acquired thrombotic process such as a myeloproliferative disease [67,68]. MOVC has also been associated with the subsequent development of hepatocellular carcinoma [69,70].

It is important to diagnose MOVC or so-called short-length hepatic vein stenoses [71] because of the availability of treatment options (eg, anticoagulation, angioplasty, or stenting) that are usually not germane to other causes of the Budd-Chiari syndrome.

Miscellaneous — Miscellaneous associations with the Budd-Chiari syndrome include systemic lupus erythematosus, mixed connective tissue disease, Crohn's



Etiology of the Budd-Chiari syndrome

Topic Outline

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[ETIOLOGY](#)

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membranous obstruction

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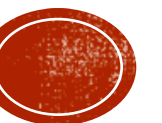
include systemic lupus erythematosus, mixed connective tissue disease, Crohn's

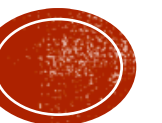
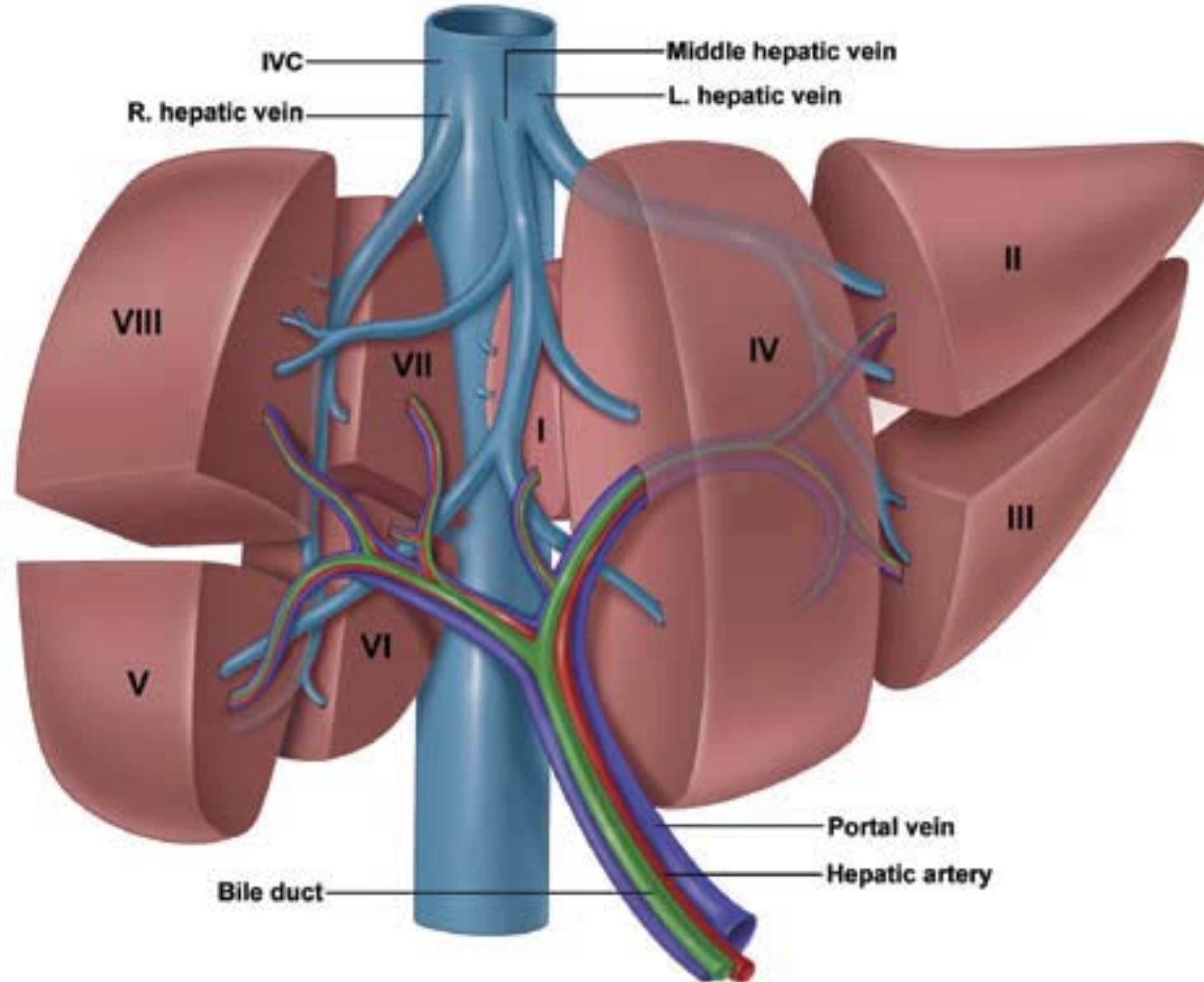


SYNDROME DE BUDD-CHIARI

- ✦ Budd Chiari : obstruction retour veineux hépatique de quelque pathologie que ce soit, excluant maladie cardiaque, péricardique ou maladie sinusoidale veineuse.
- ✦ Primaire : thrombose ou lésion sténosante
- ✦ Secondaire : compression extrinsèque

DeLeve LD, Hepatology.
2009;49(5):1729.



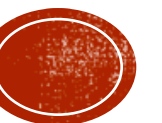


REVIEW

**Membranous obstruction of the inferior vena cava (obliterative
hepatocavopathy, Okuda)**

KUNIO OKUDA

Journal of Gastroenterology and Hepatology (2001) **16**, 1179–1183

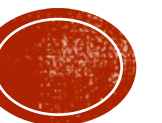


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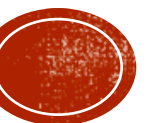
REVIEW

Membranous obstruction of the inferior vena cava (obliterative hepatocavopathy, Okuda)

KUNIO OKUDA

Journal of Gastroenterology and Hepatology (2001) 16, 1179–1183

- ✦ **Attention : Le MOIVC est différent du Budd-Chiari thrombotique aigu habituel...**
- ✦ **Processus thrombotique récurrent de la VCI à sa hauteur hépatique**
- ✦ **Le fardeau de thromboses des veines hépatiques est variable**
- ✦ **La présentation clinique est moins fulminante s'il n'y a pas de thrombose de veine hépatique aiguë (Budd-Chiari classique)**



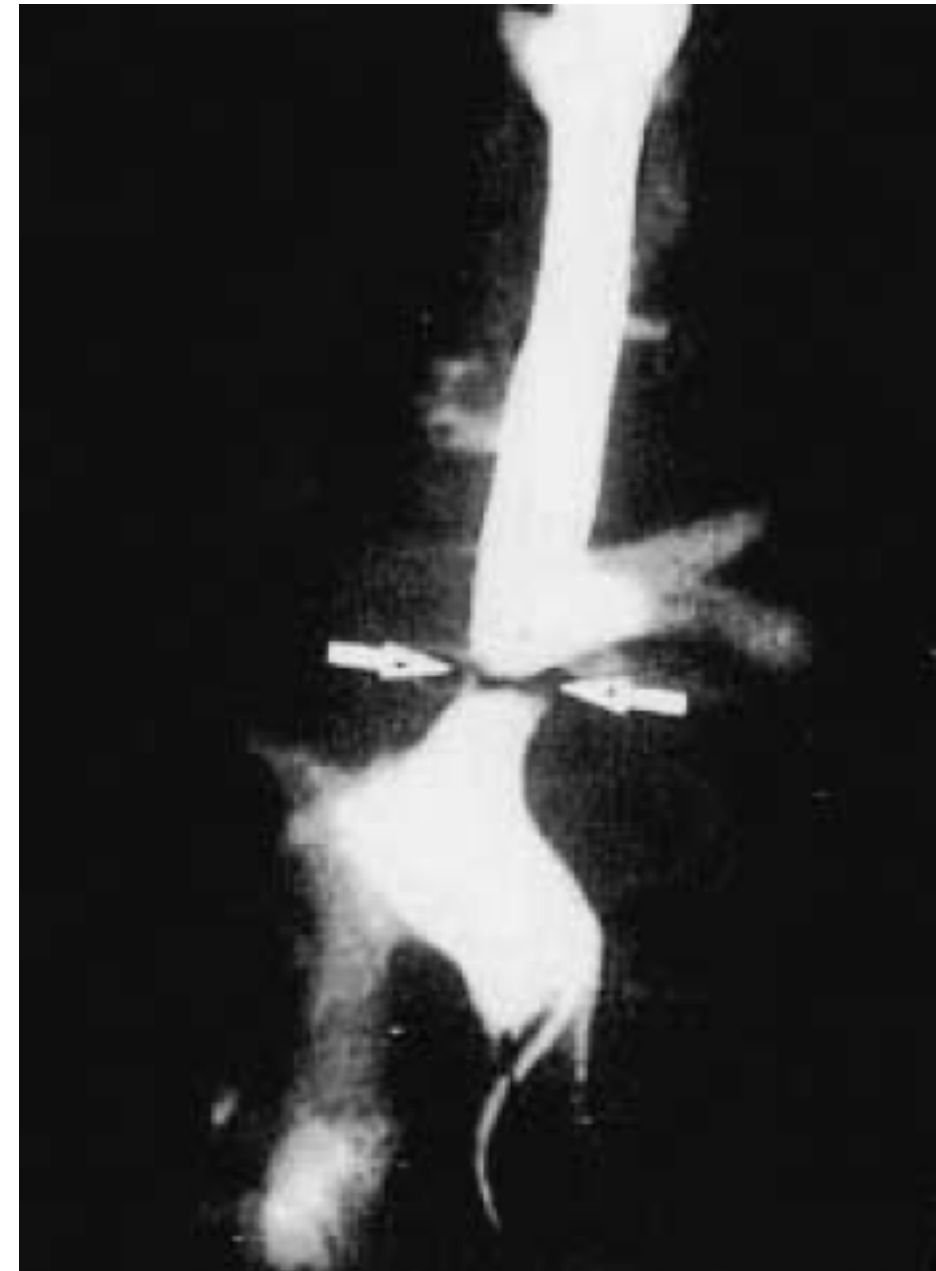
ÉTIOLOGIE DE LA MEMBRANE ?

- ✦ Hypothèse historique : coarctation veineuse congénitale.
- ✦ 17 autopsies (Kage, M Gastroenterology, 1997) MOIVC
 - ✦ Vieux thrombus organisés avec âges variables, certaines portions calcifiées dans les vieux thrombus
- ✦ 150 cas MOIVC revus (Hirooka, M Arch Surg, 1970)
 - ✦ Très grande variabilité d'épaisseur de membrane, de localisation anatomique, de thrombose de veines hépatiques (droite, moyenne, et gauche) ne militent pas en faveur d'une origine congénitale



ÉPIDÉMIOLOGIE - MOVC

- ✦ Pays de l'Est (Chine, Népal, Inde, Continent Africain)
 - ✦ MOIVC >> Budd-Chiari classique
 - ✦ Hypothèse infectieuse menant à des thrombus (milieu socio-sanitaires défavorisés, fièvre per-cavographie)
- ✦ Pays de l'Ouest
 - ✦ Budd-Chiari classique >> MOIVC
 - ✦ Étiologie prothrombotique plus prévalente probable



ÉTIOLOGIES DE BUDD-CHIARI (QUEST)

Annals of Internal Medicine

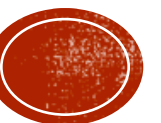
| ARTICLE

Etiology, Management, and Outcome of the Budd-Chiari Syndrome

Thrombophilia	
Inherited†	
Any inherited disorder	32/154 (21)
Factor V Leiden G1691A mutation	18/147 (12)
Prothrombin gene G20210A mutation	5/144 (3)
Protein C deficiency‡	5/117 (4)
Protein S deficiency‡§	3/108 (3)
Antithrombin deficiency‡	3/112 (3)
Acquired†	
Any acquired disorder	67/153 (44)
Antiphospholipid antibodies	37/150 (25)
Hyperhomocysteinemia	28/129 (22)
Paroxysmal nocturnal hemoglobinuria	15/77 (19)
Myeloproliferative disorder or JAK2 mutation¶	
Any myeloproliferative disorder	50/103 (49)
Polycythemia vera	27
Essential thrombocythemia	9
Idiopathic myelofibrosis	2
Unclassified	11
Occult**	1
JAK2 V617F mutation	35/121 (29)

163 patients

Hormonal factors (women only)†		35/93 (38)
Oral contraceptive use		31/93 (33)
Pregnancy within 3 mo before diagnosis		6/93 (6)
Systemic†		37/163 (23)
Connective tissue disease		10
Inflammatory bowel disease		8
Behçet disease		4
Sarcoidosis		2
Vasculitis		1
Dehydration		5
Other††		10



ÉTIOLOGIES DE BUDD-CHIARI (QUEST)

Annals of Internal Medicine

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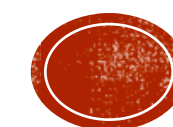


Clinical features and etiology of Budd–Chiari syndrome in Chinese patients: A single-center study

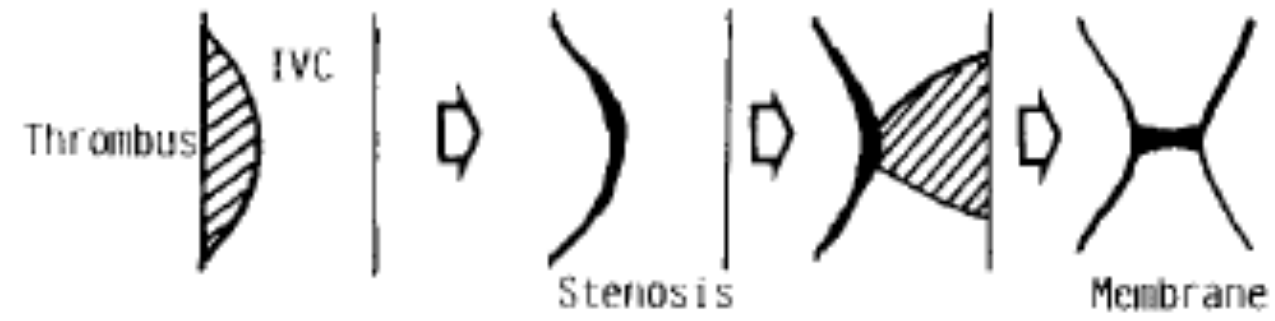
Delei Cheng,^{*} Hao Xu,^{*} Zhao-jun Lu,[§] Rong Hua,[†] Huan Qiu,[‡] Hongtao Du,[†] Xinjian Xu^{*} and Jing Zhang[§]

Table 4 Risk factors present in 145 patients with Budd–Chiari syndrome (patients could have more than one factor registered)

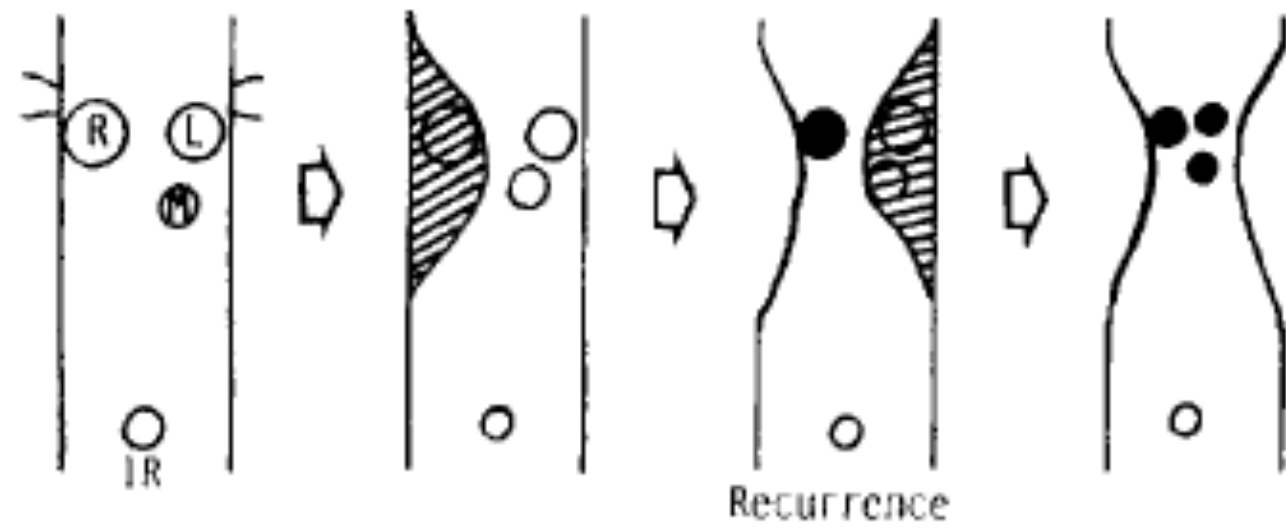
Risk factors	<i>n/n</i>	%	Risk factors	<i>n/n</i>	%
Thrombophilia			Systemic		
Myeloproliferative disorder [†]	5/105	5	Systemic lupus erythematosus	1/145	1
Polycythemia vera rubra	2/105	2	Ulcerative Colitis	1/145	1
Essential thrombocythemia	2/105	2	Phlebitis	1/145	1
JAK2 mutation	5/105	5	Ankylosing spondylitis	2/145	1
Factor V Leiden mutation	0/95	0	Hormonal factors (women only)		
Prothrombin G20210A mutation	0/95	0	Oral contraceptive use	0/55	0
Paroxysmal nocturnal hemoglobinuria	0/83	0	Pregnancy within 3 mo before diagnosis	2/55	4
Protein C deficiency [‡]	0/80	0	MO	89/145	61
Protein S deficiency [‡]	0/80	0	MOVC	6/145	4
Antithrombin deficiency [‡]	0/80	0	MOHV	23/145	16
Antiphospholipid antibodies	25/145	17	MOVC and hepatic vein involvement	60/145	41
Hyperhomocysteinemia [§]	30/145	21	Idiopathic	28	19



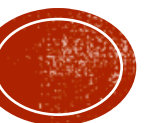
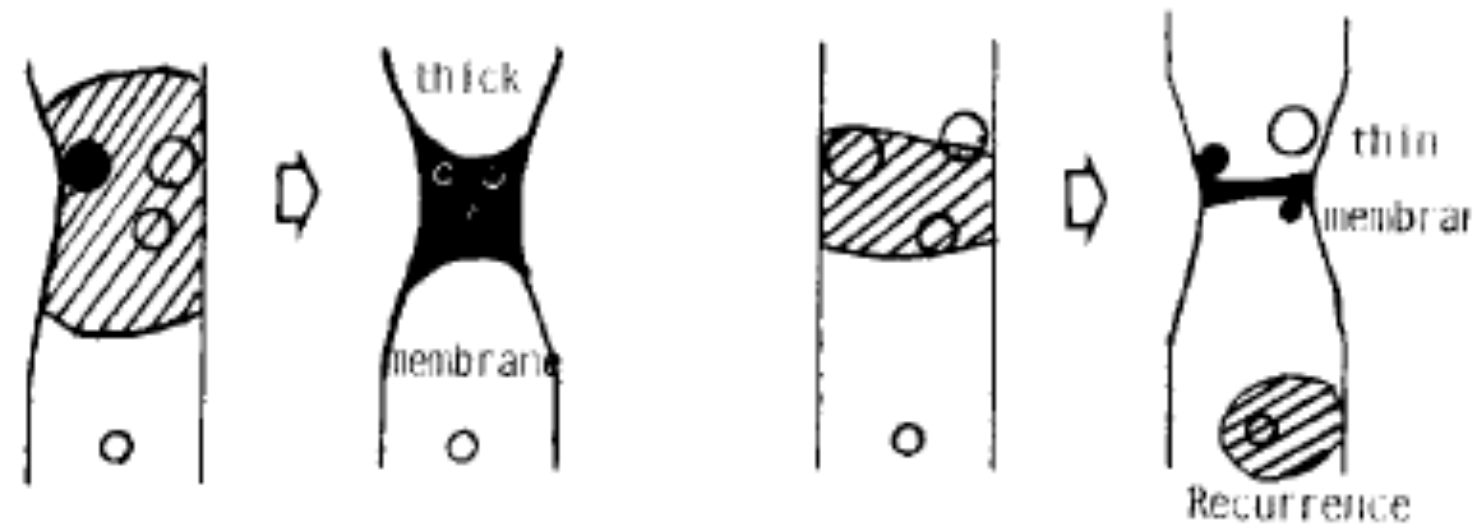
Stenosis
and
Membrane



Distal
Obstruction



VC obstruction



PRÉSENTATION CLINIQUE

Table 2 Symptoms in Hepatic Vein Thrombosis (HVT) and Obliterative Hepatocavopathy (Membranous Obstruction of the Vena Cava, MOVC)

	Rector et al ⁴³ (USA)		Hadengue et al ⁴² (France)	Okuda et al ¹⁹ (Japan)
	HVT (n = 10)	MOVC (n = 8)	HVT (n = 81)	Mostly MOVC (n = 157)
Ascites	90	38	83	31
Abdominal pain	70	50	72	23
Hepatomegaly	100	100	88	55
Dilated veins over body, trunk	0	50	0	27
Leg edema[[MR 09]]	40	88	33	32
Jaundice	4.6 mg/dL*	1.9 mg/dL*	33	6
Fever	31			
Encephalopathy	13			

*serum bilirubin



PRÉSENTATION CLINIQUE

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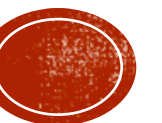
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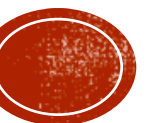
DIAGNOSTIC

- ✦ Échographie doppler
 - ✦ Sensible pour l'obstruction VCI et absence/renversement de flot dans les VH
- ✦ CT-SCAN C+ ou IRM gado +
 - ✦ Hétérogénéité ou non-visualisation des VH, collatérales intra-hépatiques ou obstruction VCI
- ✦ IRM gado +
 - ✦ Apporte la localisation anatomique



DIAGNOSTIC

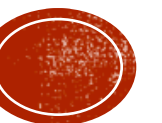
- ✦ Cavographie
 - ✦ Permet de mesurer le gradient de pression en mmHg
 - ✦ < 3 non-significatif
 - ✦ $3 - 6$ possiblement significatif
 - ✦ $6 - 9$ probablement significatif
 - ✦ > 9 significatif



NOTRE PATIENTE

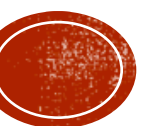


- ✦ Gradient VCI et OD 13mmH significatif



QUESTIONS

- ✿ Quelle est l'étiologie de cette sténose ?
- ✿ Quelle est l'approche clinique optimal ?





Original article

Inferior vena cava obstruction: long-term results of endovascular management

B.C. Srinivas^{1*}, P.V. Dattatreya², K.H. Srinivasa¹, Prabhavathi¹, C.N. Manjunath¹

Indian Heart Journal 6402 (2012) 162–169

Table 1

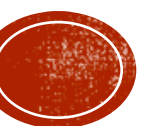
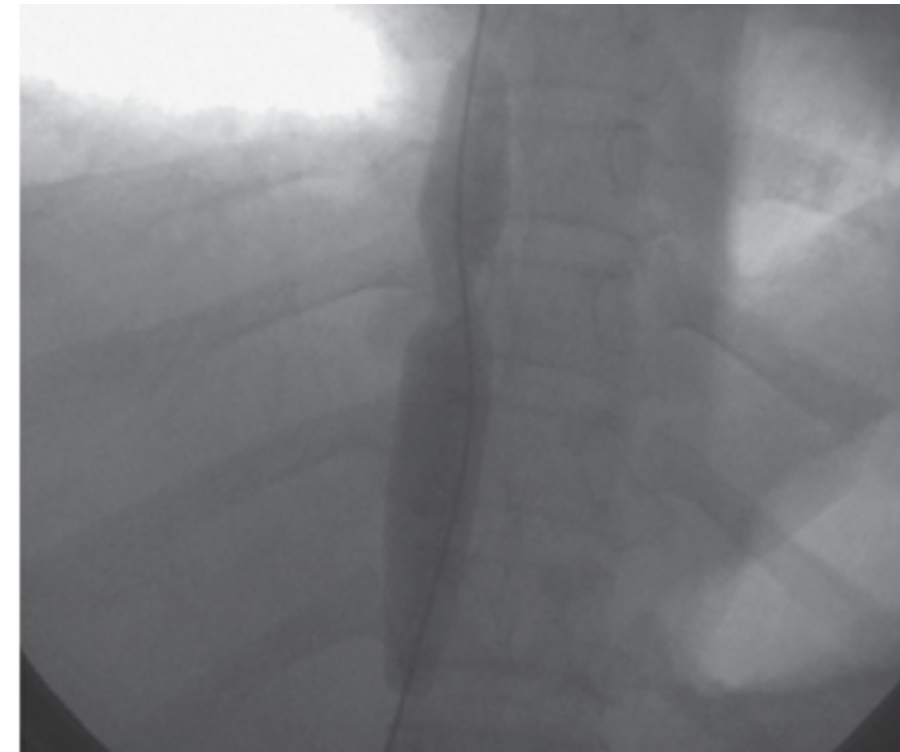
Baseline characteristics of patients with chronic inferior vena cava obstruction.

Age	35±6.1 yr
Sex (male/female)	7/5
Oedema of lower extremities	12/12 cases
Abdominal swelling and ascites	12/12 cases
Prominent veins on abdomen and back	12/12 cases
Hepatomegaly	10/12 cases
Hepatomegaly with nodules on liver (metastatic nodules from adenocarcinoma of stomach)	1/12 cases
Splenomegaly	4/12 cases
Impaired hepatic function	10/12 cases



TRAITEMENT

- ✦ Gradient 23 ± 5 mmHg à 4 ± 3 mmHg
- ✦ 7 angioplasties
- ✦ 5 stents
 - ✦ Obstruction sur long segment n=2
 - ✦ Resténose après angioplastie n=2
 - ✦ Compression extrinsèque de la VCI n=1
- ✦ Anticoagulation indéfinie chez tous



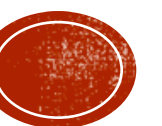
ÉVOLUTION CLINIQUE PÉRI- PROCÉDURE

- Chez tous les patients
 - Collatérales ont disparues
 - Résolution de l'œdème, ascite et douleur abdominale en 1 mois
 - Pas de saignement
- Amélioration bilan hépatique à 1 mois sauf 2 patients



LONG TERME

- ✿ Suivi clinique et échographique entre 15 mois et 14 ans
 - ✿ 1 cas de resténose 8 mois (POBA) -> stent
 - ✿ Autres : VCI perméable au suivi



Percutaneous balloon angioplasty of inferior vena cava in Budd–Chiari syndrome-R1

Tongguo Wu^a, Lexin Wang^{b,*}, Qiang Xiao^a, Bosong Wang^a, Shanying Li^a, Xiurong Li^a,
Jinying Zhang^a

^a*Department of Cardiology, The Affiliated Hospital of Taishan Medical College, Taishan, China*

^b*School of Biomedical Sciences, Charles Sturt University, Wagga Wagga, NSW 2678, Australia*

International Journal of Cardiology 83 (2002) 175–178

§ Étude descriptive chinoise

§ 42 patients

§ MOIVC 28 patients

§ Segmentaire 14 patients

§ 14/42 patients

§ Obstruction veine hépatique (gauche ou moyenne)

§ Majorité : ascites, hépatosplénomégalie, douleur abdominales et OMI



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§ Obstruction veine hépatique (gauche ou moyenne)

§ Majorité : ascites, hépatosplénomégalie, douleur abdominales et OMI

§ Procédure

§ Ballon Inoue 15-22mm 2-3x

§ Anticoagulation 1 semaine

§ Résultats

§ Angioplastie succès 97,6%

§ Stent 1/42 patient

§ Amélioration

§ Clinique chez tous 42/42

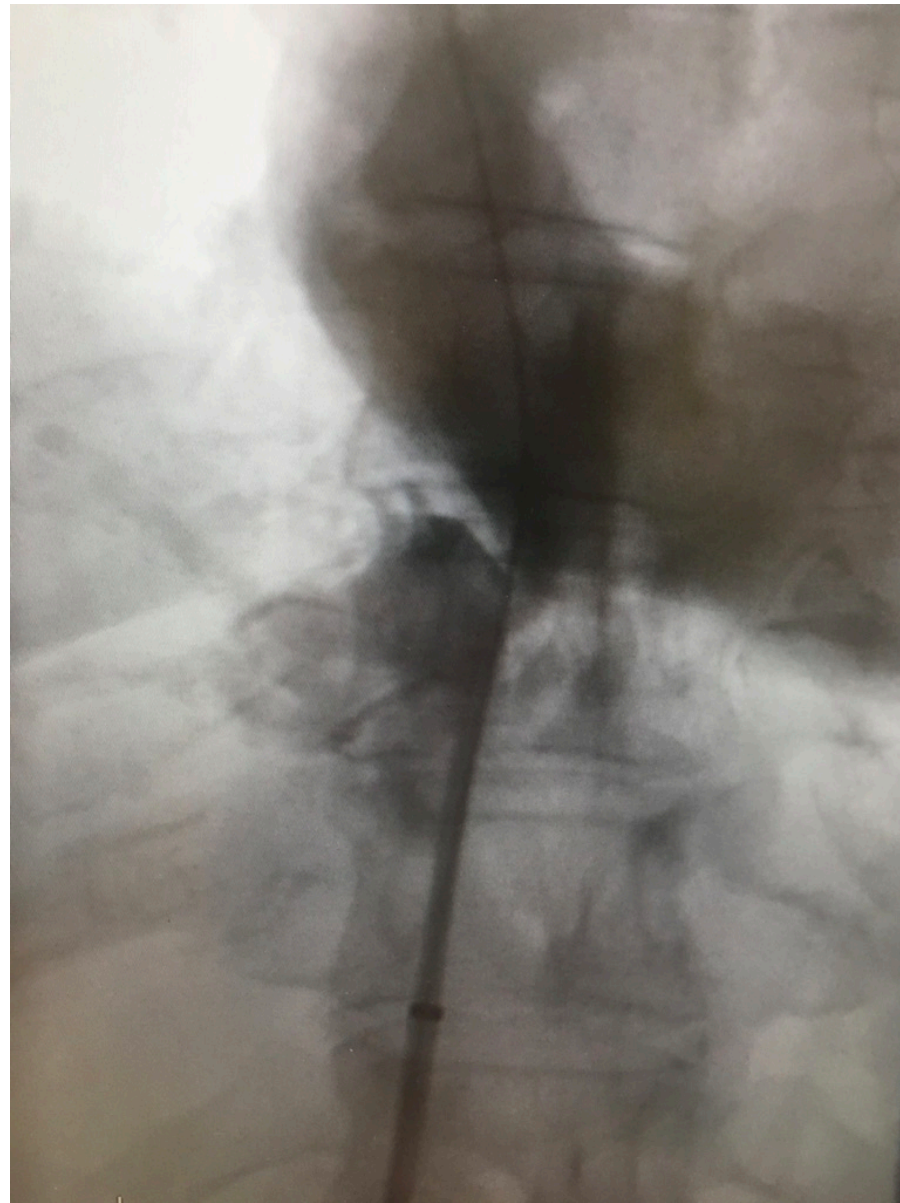
§ Persistance perturbation hépatique chez 3/12

§ FUP : 6 à 72 mois (Doppler)

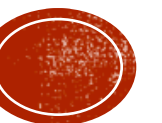
§ 1 patient resténose à 3 ans



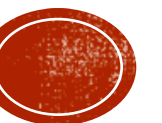
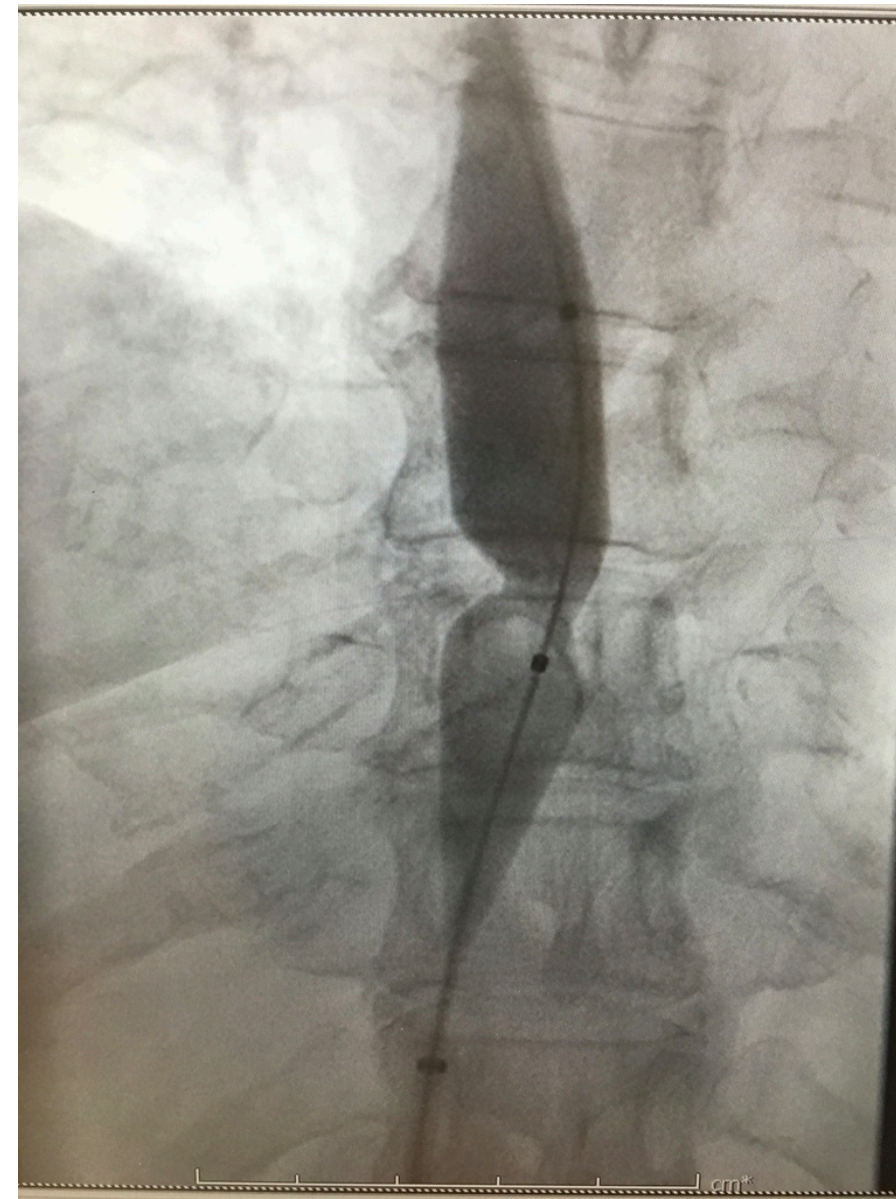
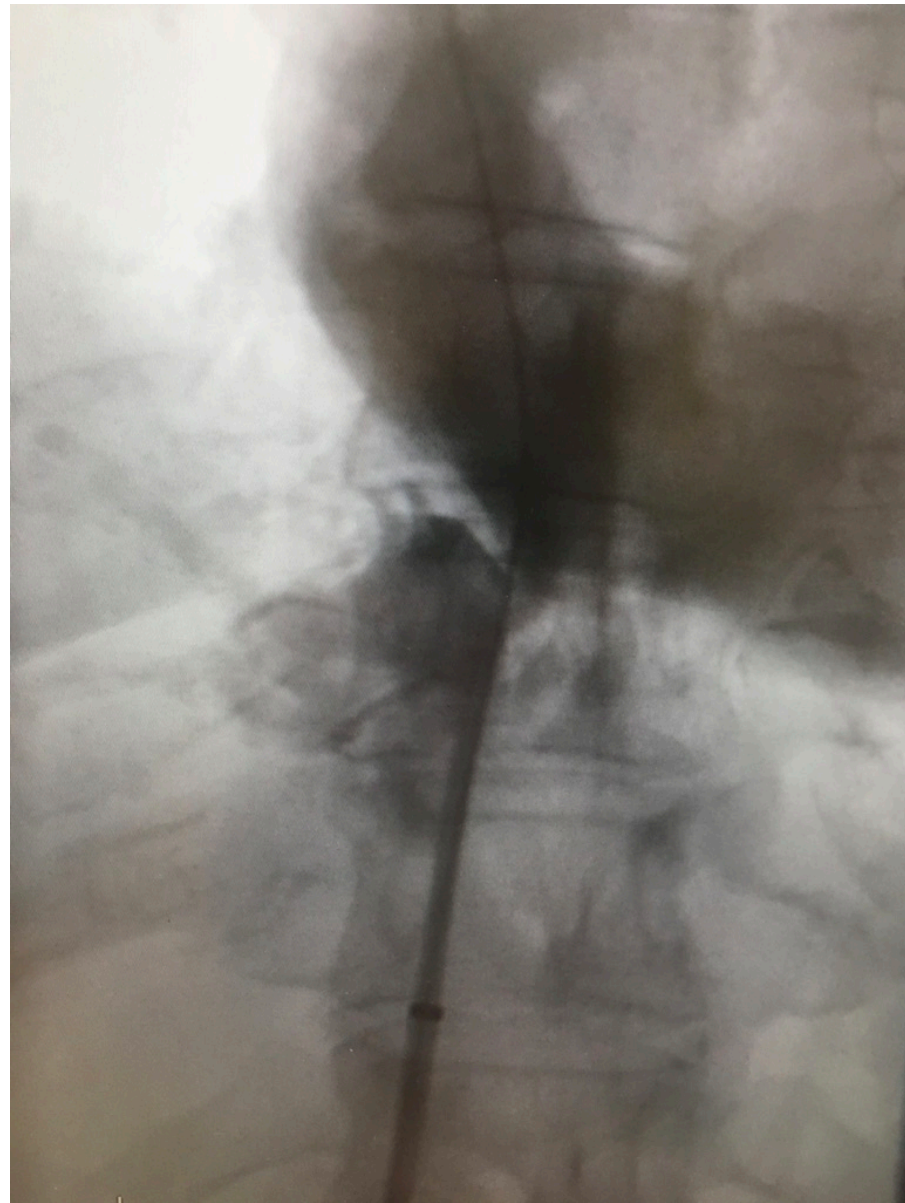
NOTRE PATIENTE



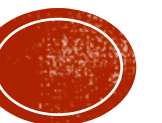
- ✦ Désanticoagulée avec pont Fragmin
- ✦ Gradient VCI et OD 13mmH très significatif



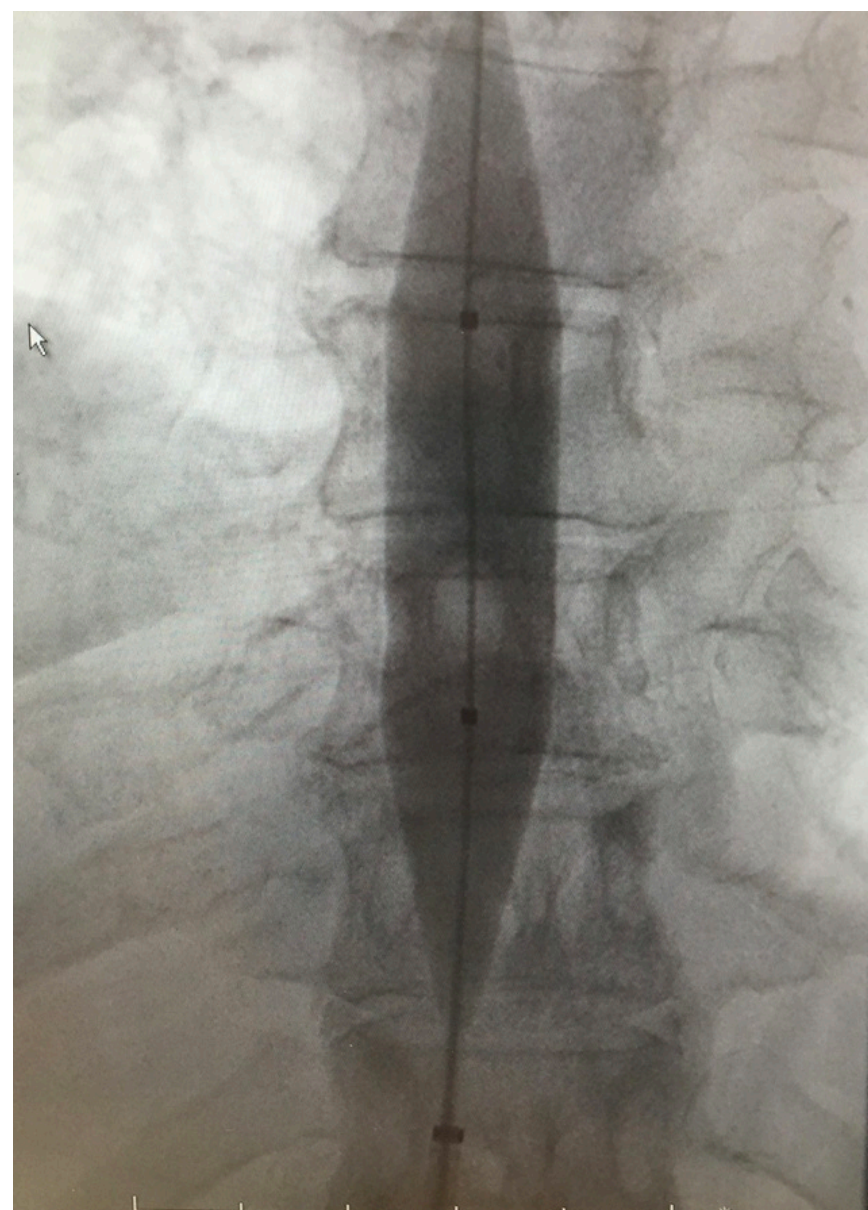
NOTRE PATIENTE



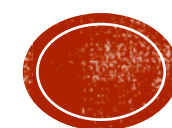
NOTRE PATIENTE



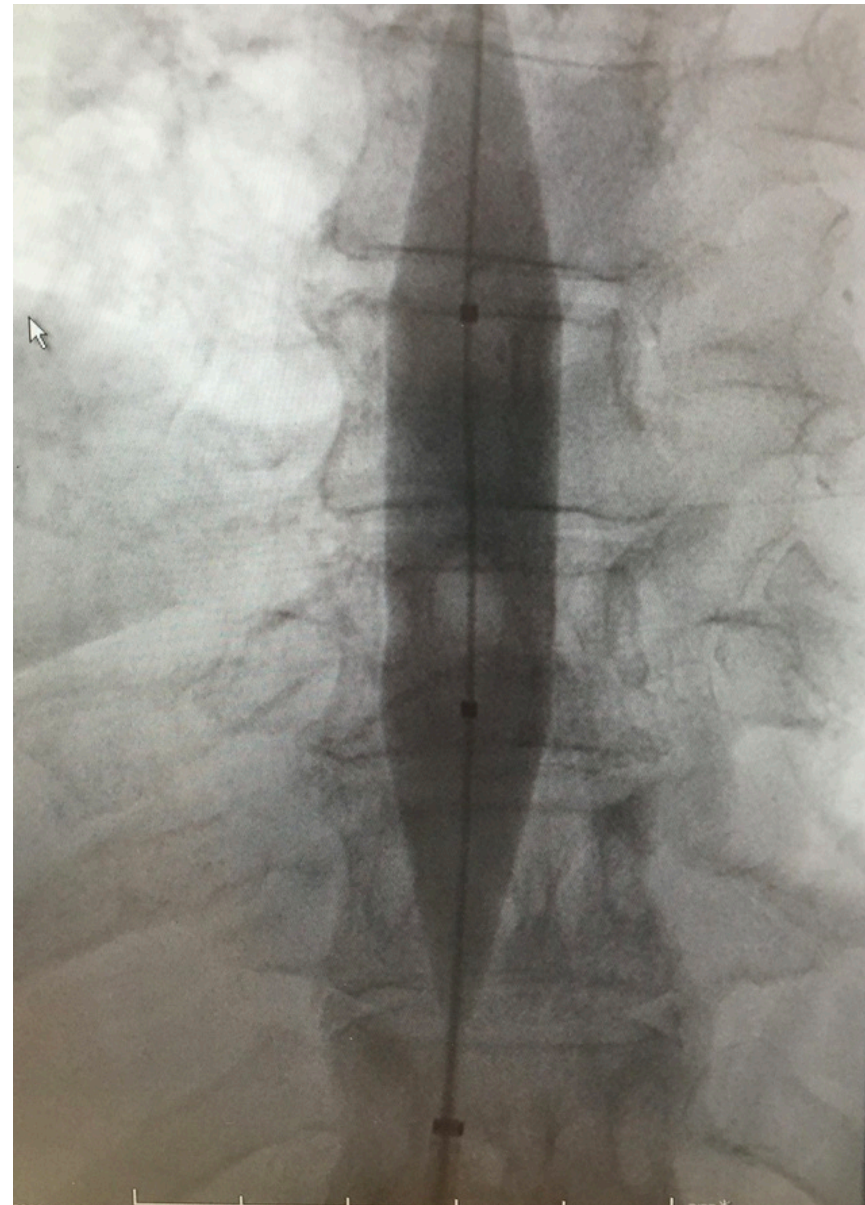
NOTRE PATIENTE



**SURPRISE
NO. 6 !!!**

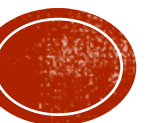


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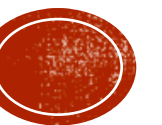
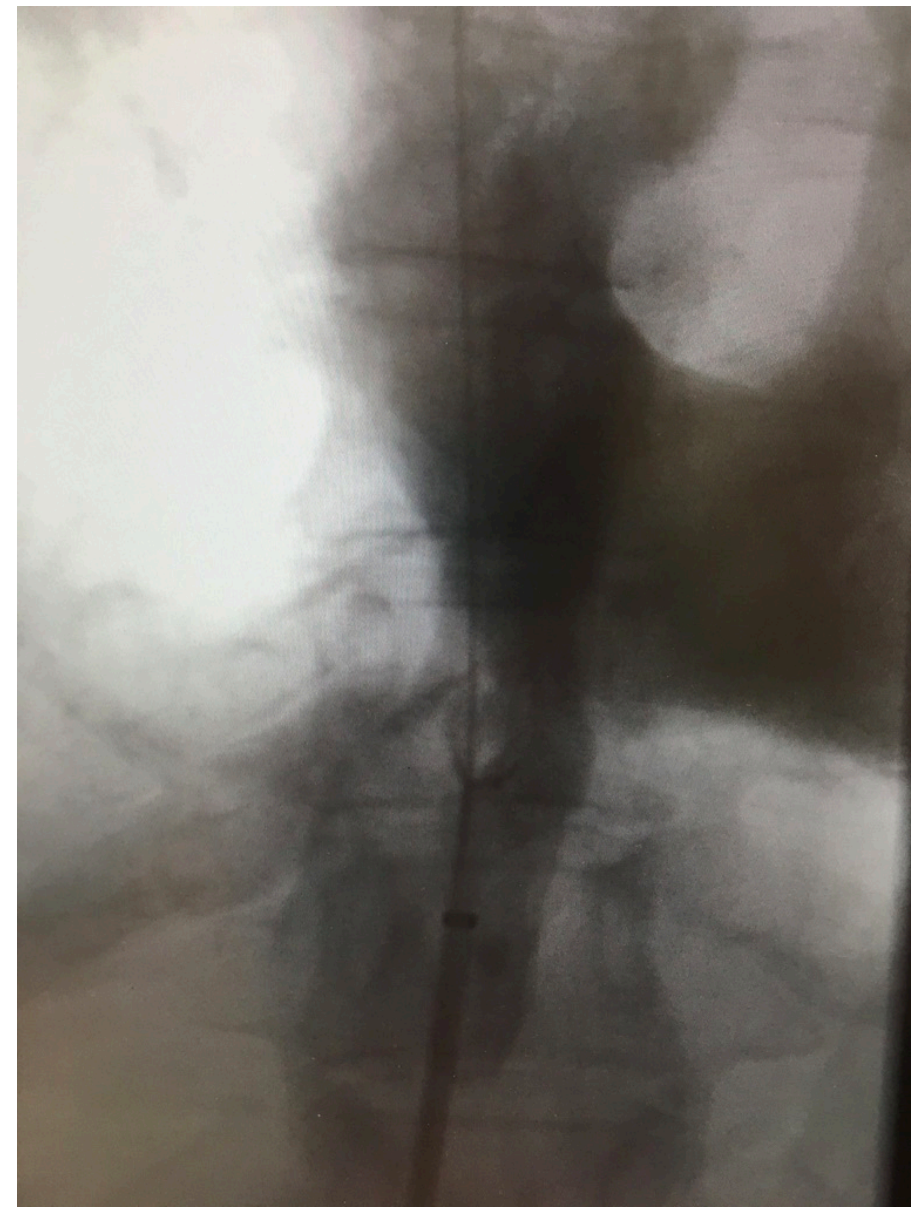
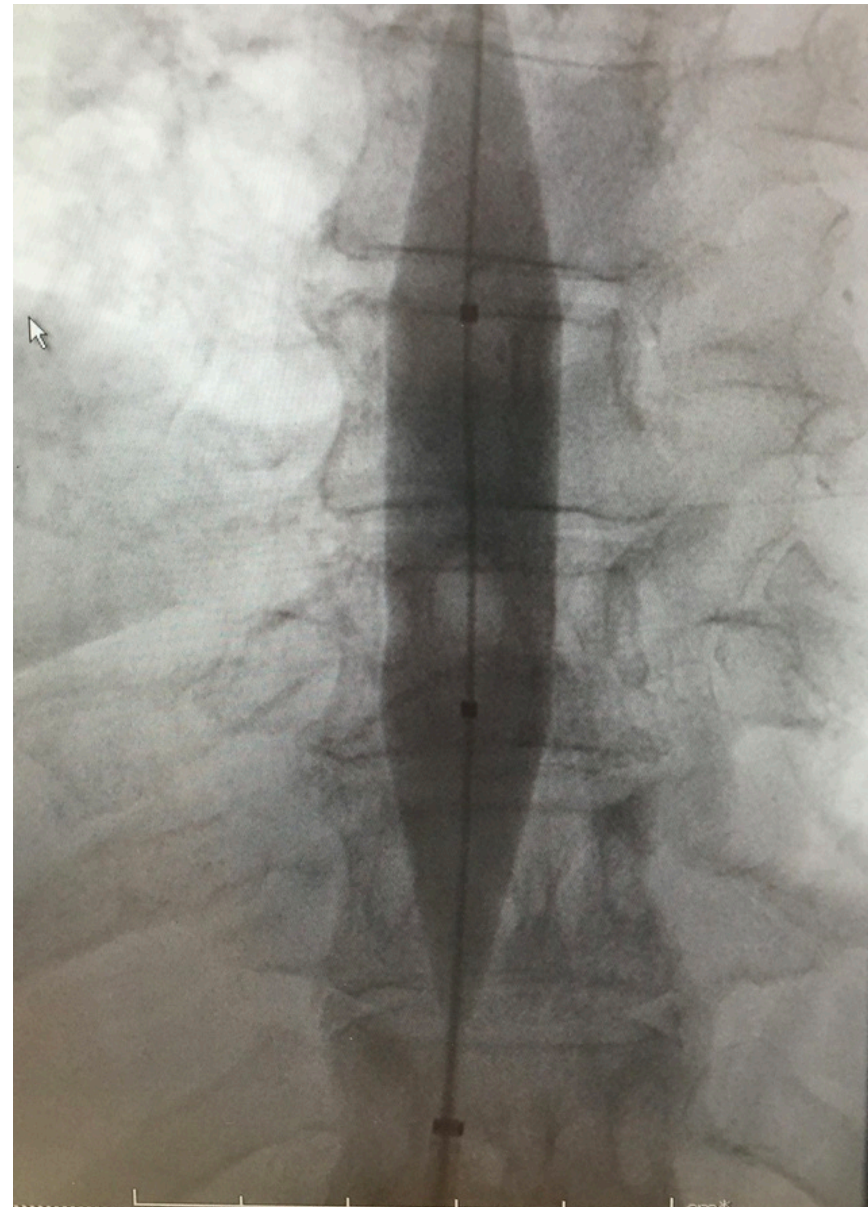


**SURPRISE
NO. 6 !!!**

&#\$%#@# !!

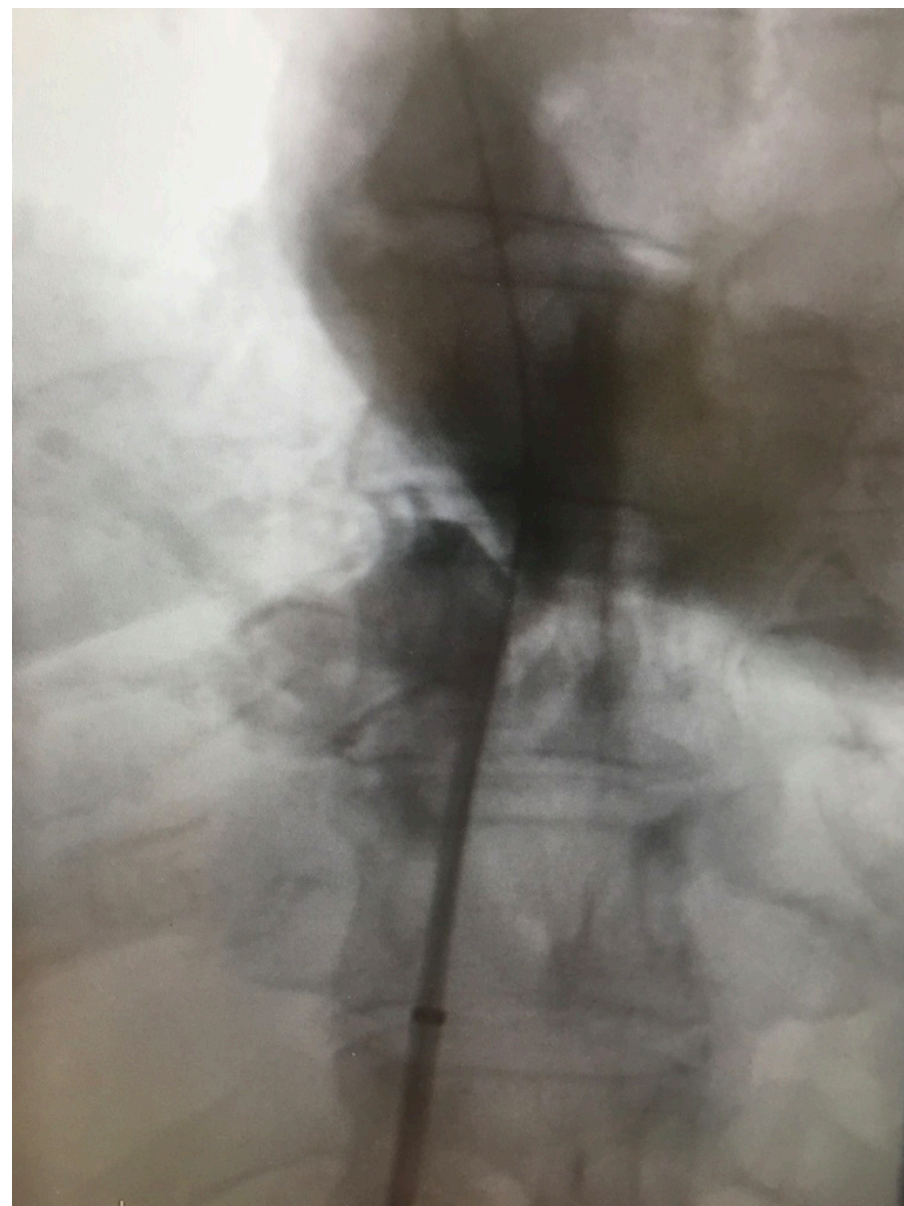


NOTRE PATIENTE



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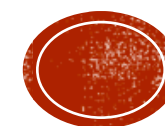
✦ Gradient 12-13mm-Hg à 3-4 mmHg !!



AVANT

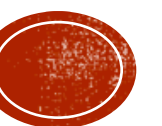


APRÈS



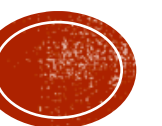
ÉVOLUTION CLINIQUE

- ✦ En 24-48 heures, amélioration clinique drastique
 - ✦ Résolution OMI, claudication veineuse
- ✦ Reprise de l'anticoagulation indéfinie
- ✦ Contrôle doppler VCI 12 mois post plastie



DIAGNOSTIC FINAL

- ✦ Obstruction membraneuse de la veine cave inférieure secondaire état prothrombotique sur syndrome anti-phospholipide triple positif



Transformation of Inferior Vena Caval Thrombosis to Membranous Obstruction in a Patient With the Lupus Anticoagulant

HIDETAKA TERABAYASHI, KUNIO OKUDA, FUMIO NOMURA,
KUNIHICO OHNISHI, and PAKMING WONG

First Department of Medicine and Second Department of Medicine, Chiba University School of Medicine, Chiba, Japan

§F 24 ans, bonne santé

§Douleurs abdominale x quelques semaines

§Doppler abdominal : thrombose partielle VCI
et VH droite

§Thrombolyse systémique et coumadin

§Cavographie, lésion membraneuse sténosante
de la VCI avec gradient 13mmHg

§Pas d'angioplastie à l'époque

§Anticorps lupique +

§Anticoagulation indéfinie



Membranous Obstruction of the Inferior Vena Cava and Extensive Bone Marrow Necrosis Associated with Catastrophic Antiphospholipid Syndrome

Alex C. Spyropoulos

Journal of Clinical Rheumatology • Volume 5, Number 5 • October 1999

§H 37 ans, Tb schizo-affectif

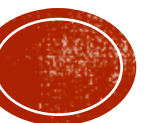
§S/p CCK 7 jours

§Douleur abdominale, OMI

§CIVD présumée : Plt 34, HTE
25%

§Transaminases 100-125.

§Doppler abdominal, VH, VCI
N



Membranous Obstruction of the Inferior Vena Cava and Extensive Bone Marrow Necrosis Associated with Catastrophic Antiphospholipid Syndrome

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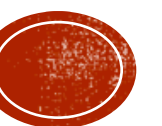
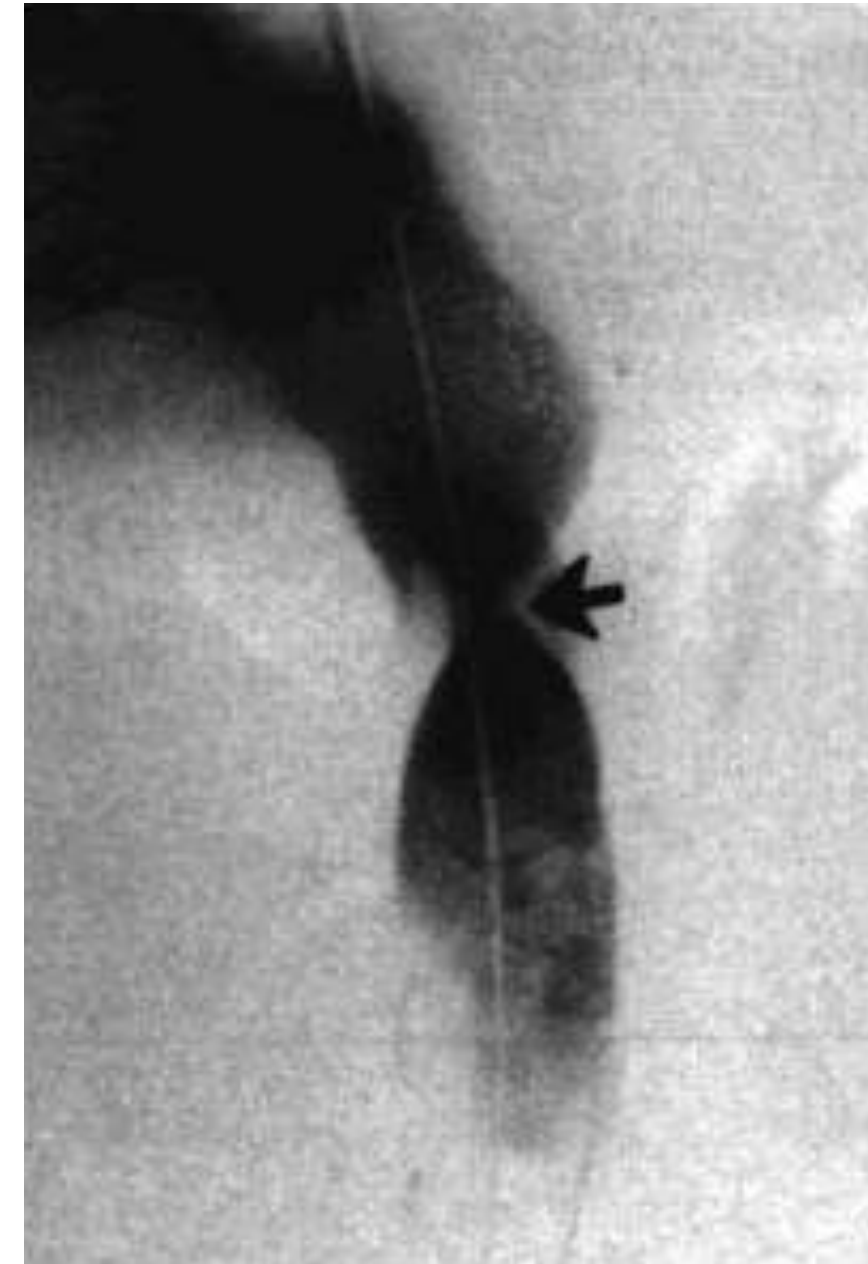
§Amélioration sous antibiothérapie

§7 jours plus tard

§OMI ++, douleur abdo et lombaire, confusion, hypotension

§Bx moelle - nécrose 80%

§Scan abdo - lésion sténosante VCI à la hauteur hépatique



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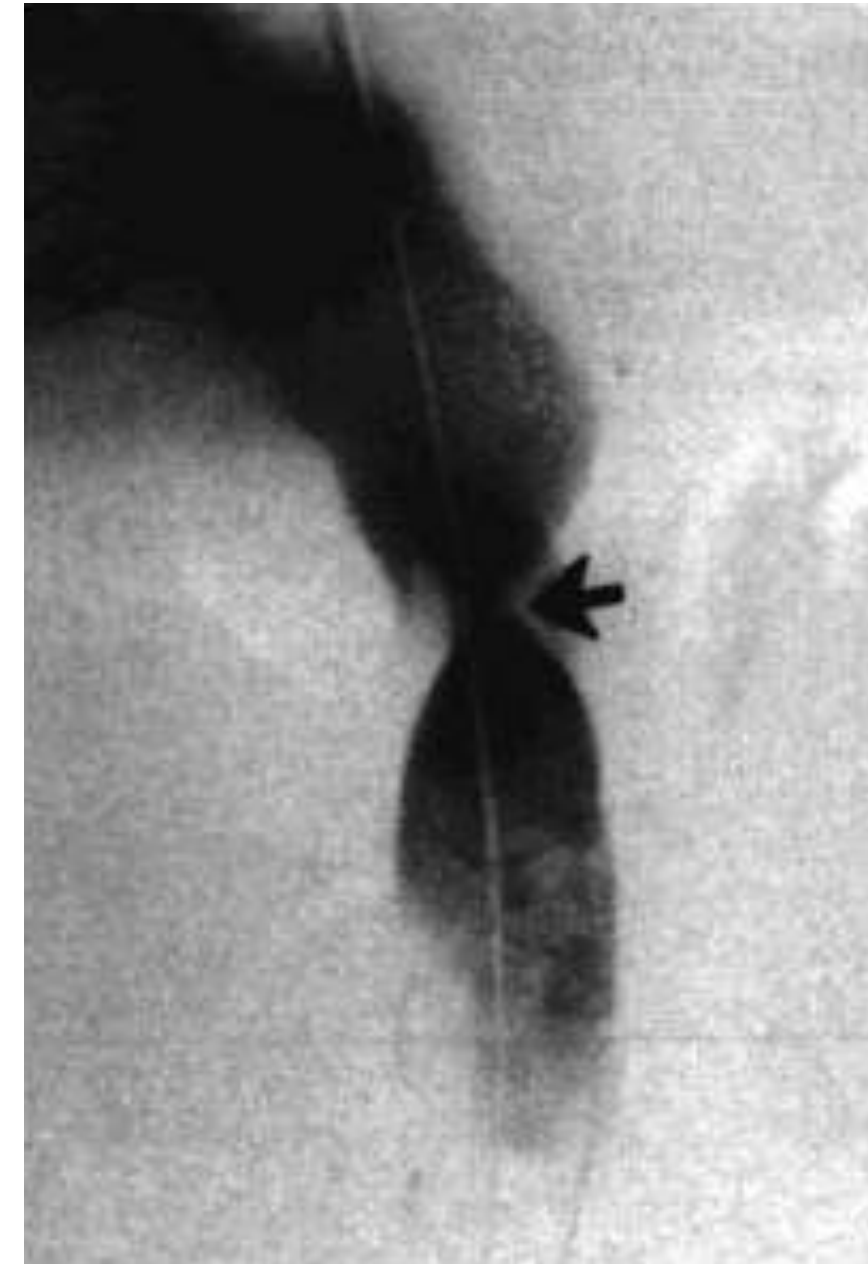
§Gradient 10mmHg

§Angioplastie ballon succès  2 mmHg

§Amélioration drastique confusion, œdème

§Fonction hématopoïétique a récupéré

§ATC indéfinie



Primary antiphospholipid antibody syndrome with membranous obstruction of the inferior vena cava—successful PTA and thrombolytic therapy

Nephrol Dial Transplant (1998) 13: 2641-2644

Vincenzo La Milia¹, Flavia Tentori¹, Claudio Pozzi¹, Giuseppe Taddei² and Francesco Locatelli¹

§H 42 ans, bonne santé

§Douleurs lombaires x 3 jours sévères

§Œdème à godet MID

§IRA non-oligurique

§Doppler veineux : thrombose extensive profonde MID et VCI ad sous veine hépatique

§Héparine IV

§Amélioration clinique et biochimique en 7 jours

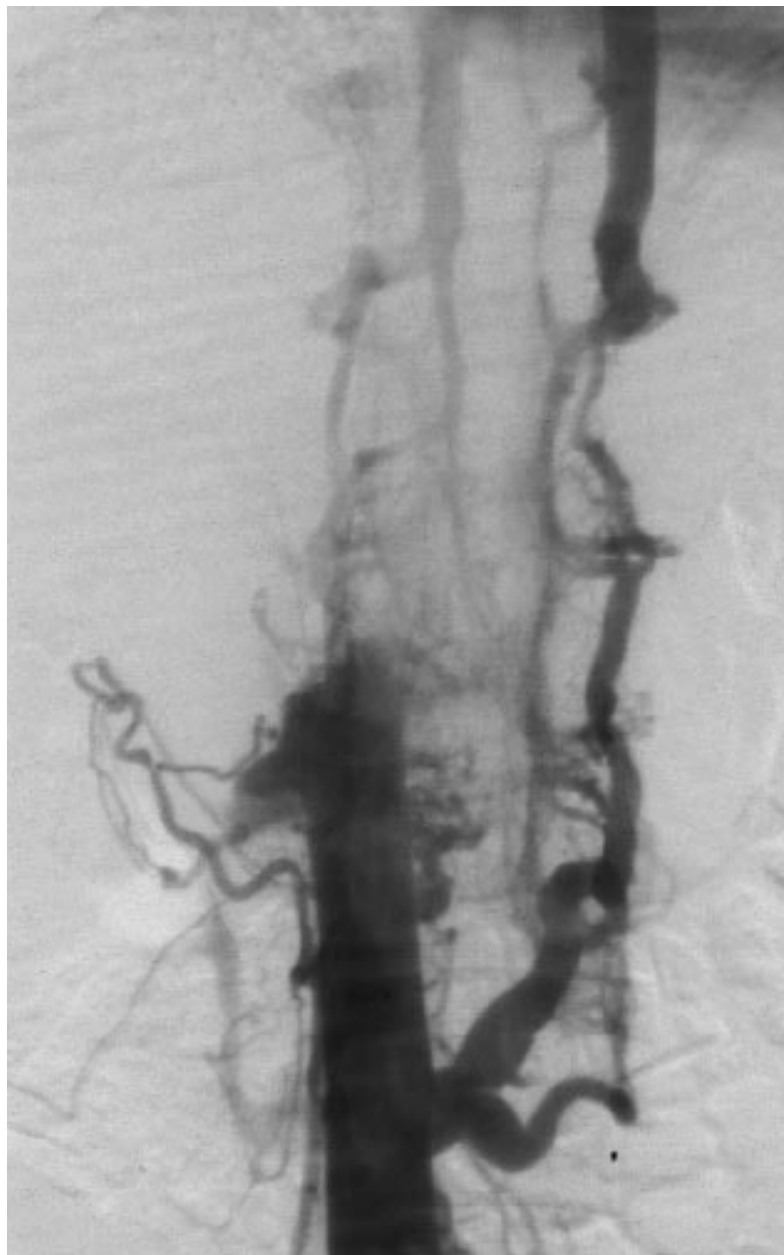
§SAPL double-positif hauts titres (Anti-B2 glycoprotéine 1 pas fait)



Primary antiphospholipid antibody syndrome with membranous obstruction of the inferior vena cava—successful PTA and thrombolytic therapy

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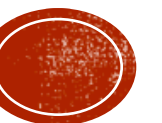
Vincenzo La Milia¹, Flavia Tentori¹, Claudio Pozzi¹, Giuseppe Taddei² and Francesco Locatelli¹



§Après 14 jours d'héparine IV

§Thrombose complète VCI

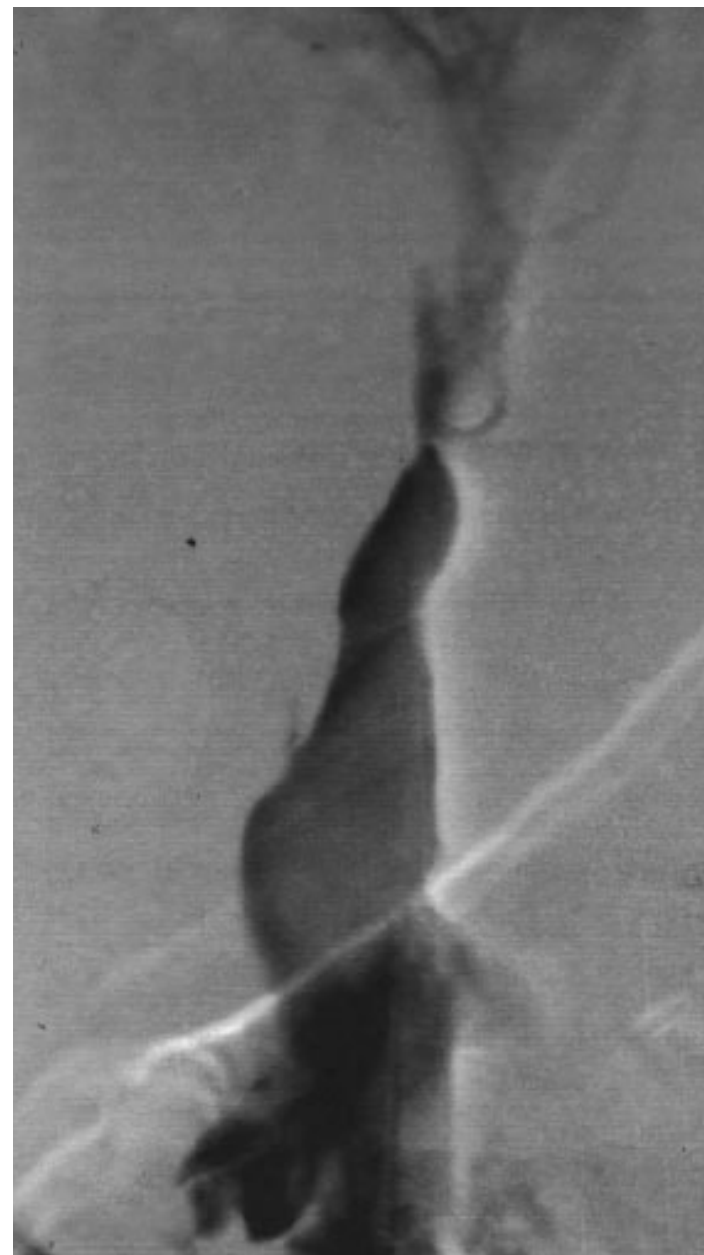
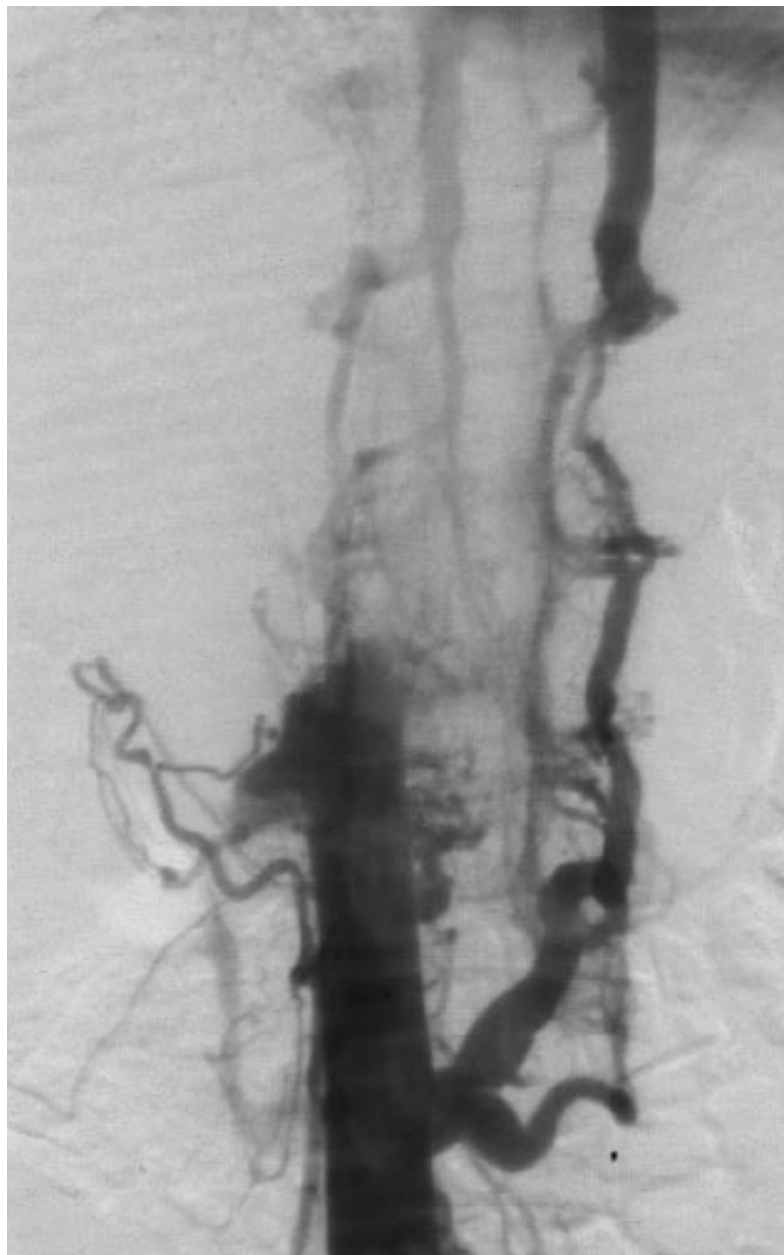
§Thrombolyse locale urokinase



Primary antiphospholipid antibody syndrome with membranous obstruction of the inferior vena cava—successful PTA and thrombolytic therapy

Nephrol Dial Transplant (1998) 13: 2641-2644

Vincenzo La Milia¹, Flavia Tentori¹, Claudio Pozzi¹, Giuseppe Taddei² and Francesco Locatelli¹



§Après 4 jours de thrombolyse

§Cavographie : lésion sténosante supra-rénal avec un gradient de 10mmHg

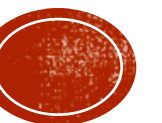
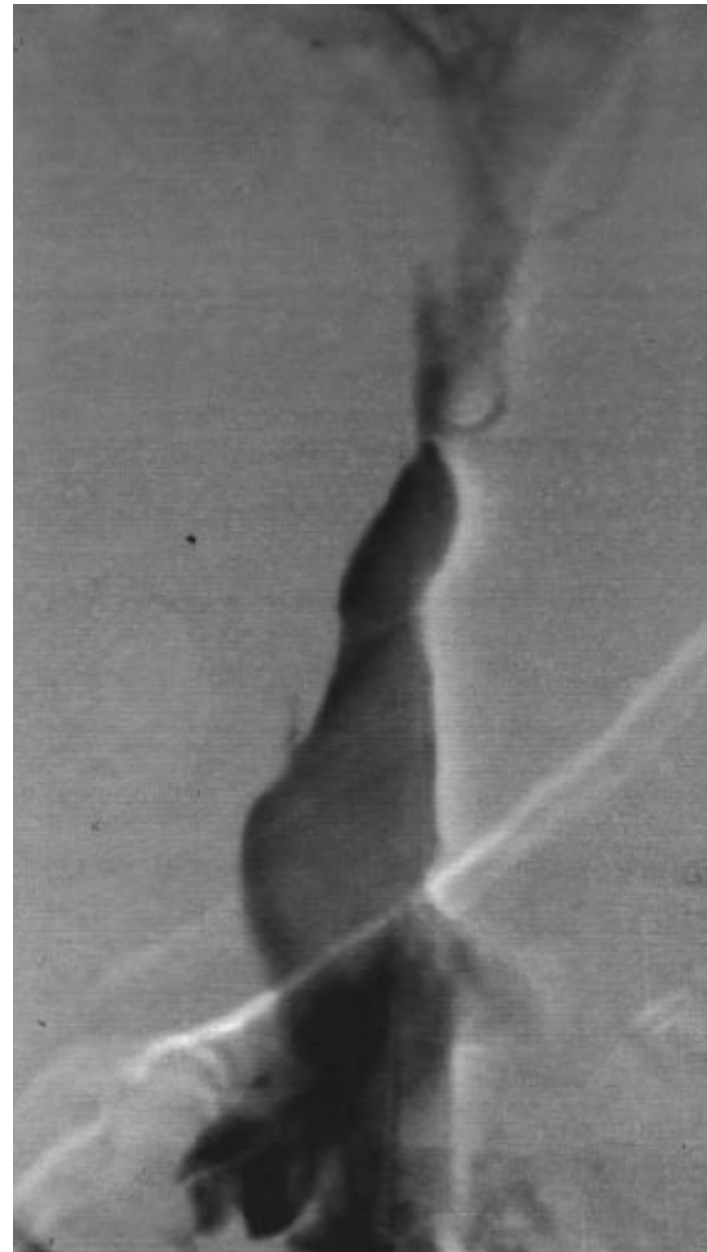
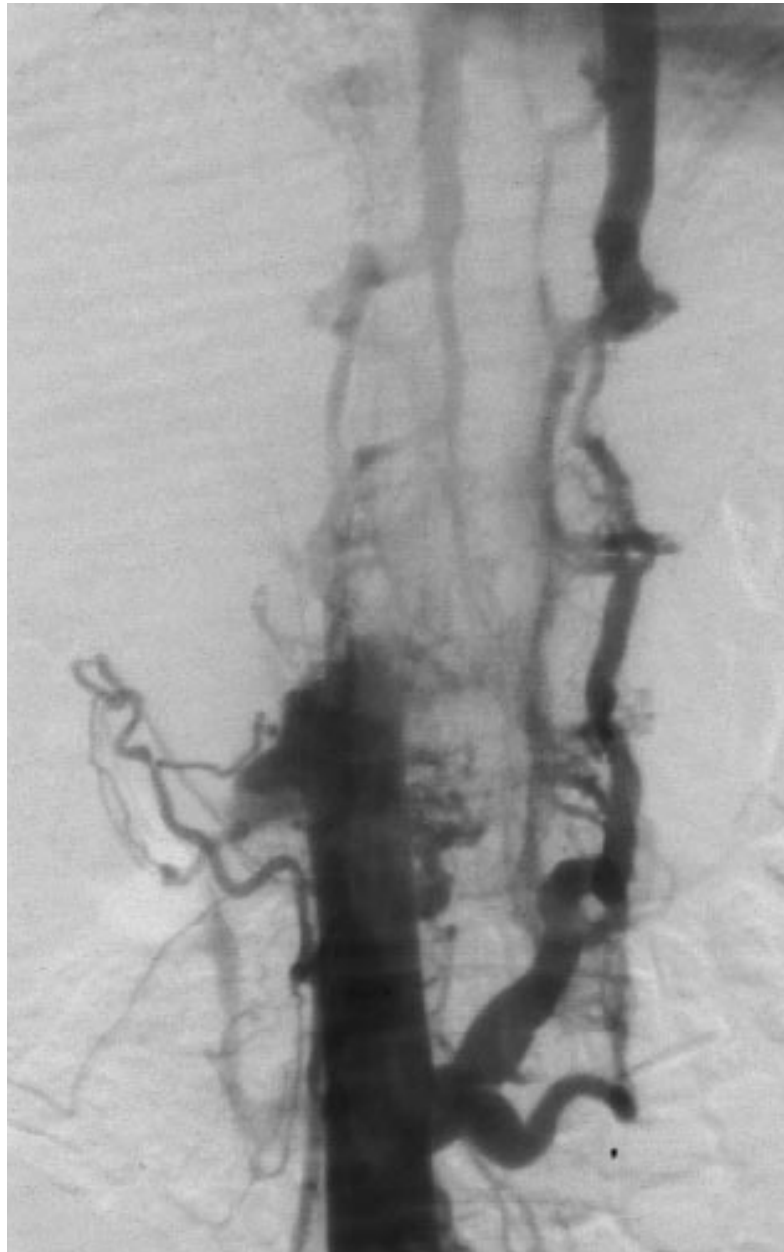
§Angioplastie ballon 15mm



Primary antiphospholipid antibody syndrome with membranous obstruction of the inferior vena cava—successful PTA and thrombolytic therapy

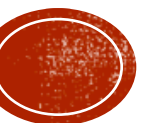
Nephrol Dial Transplant (1998) 13: 2641-2644

Vincenzo La Milia¹, Flavia Tentori¹, Claudio Pozzi¹, Giuseppe Taddei² and Francesco Locatelli¹



CONCLUSION

- ✦ L'obstruction membraneuse de la veine cave inférieure (MOIVC) serait due en grande partie à un état protrombotique à l'Ouest
- ✦ La présentation clinique est moins fulminante qu'un syndrome de Budd-Chiari classique et dépend de la présence ou non de thromboses de veines hépatiques.
- ✦ La cavographie permet d'évaluer l'impact hémodynamique de la sténose.
- ✦ L'angioplastie par ballon est hautement sécuritaire et suffisante sauf exception.
- ✦ L'amélioration clinique est rapide et drastique.
- ✦ La resténose ne semble pas fréquente.
- ✦ Le rôle de l'anticoagulation indéfinie reste à éclaircir.



MÉDIAGRAPHIE 1/2

- Cheng, Delei, Hao Xu, Zhao-Jun Lu, Rong Hua, Huan Qiu, Hongtao Du, Xinjian Xu, et Jing Zhang. 2013. « Clinical Features and Etiology of Budd-Chiari Syndrome in Chinese Patients: A Single-Center Study ». *Journal of Gastroenterology and Hepatology* 28 (6): 1061–67. doi: 10.1111/jgh.12140.
- Darwish Murad, Sarwa, Aurelie Plessier, Manuel Hernandez-Guerra, Federica Fabris, Chundamannil E. Eapen, Matthias J. Bahr, Jonel Trebicka, et al. 2009. « Etiology, Management, and Outcome of the Budd-Chiari Syndrome ». *Annals of Internal Medicine* 151 (3): 167–75.
- DeLeve, Laurie D., Dominique-Charles Valla, Guadalupe Garcia-Tsao, et American Association for the Study Liver Diseases. 2009. « Vascular Disorders of the Liver ». *Hepatology (Baltimore, Md.)* 49 (5): 1729–64. doi:10.1002/hep.22772.
- « Etiology of the Budd-Chiari syndrome - UpToDate ». 2016. Consulté le novembre 17. https://www.uptodate.com/contents/etiology-of-the-budd-chiari-syndrome?source=search_result&search=etiology%20of%20budd%20chiari&selectedTitle=1~129.
- Kage, M., M. Arakawa, M. Kojiro, et K. Okuda. 1992. « Histopathology of Membranous Obstruction of the Inferior Vena Cava in the Budd-Chiari Syndrome ». *Gastroenterology* 102 (6): 2081–90.
- Kimura, C., S. Matsuda, H. Koie, et M. Hirooka. 1972. « Membranous Obstruction of the Hepatic Portion of the Inferior Vena Cava: Clinical Study of Nine Cases ». *Surgery* 72 (4): 551–59.
- La Milia, V., F. Tentori, C. Pozzi, G. Taddei, et F. Locatelli. 1998. « Primary Antiphospholipid Antibody Syndrome with Membranous Obstruction of the Inferior Vena Cava--Successful PTA and Thrombolytic Therapy ». *Nephrology, Dialysis, Transplantation: Official Publication of the European Dialysis and Transplant Association - European Renal Association* 13 (10): 2641–44.



MÉDIAGRAPHIE 2/2

- Okuda, K. 2001. « Membranous Obstruction of the Inferior Vena Cava (Obliterative Hepatocavopathy, Okuda) ». *Journal of Gastroenterology and Hepatology* 16 (11): 1179-83.
- Okuda, Kunio. 2002. « Inferior Vena Cava Thrombosis at Its Hepatic Portion (Obliterative Hepatocavopathy) ». *Seminars in Liver Disease* 22 (1): 15-26. doi:10.1055/s-2002-23203.
- Spyropoulos, A. C. 1999. « Membranous Obstruction of the Inferior Vena Cava and Extensive Bone Marrow Necrosis Associated with Catastrophic Antiphospholipid Syndrome ». *Journal of Clinical Rheumatology: Practical Reports on Rheumatic & Musculoskeletal Diseases* 5 (5): 279-84.
- Srinivas, B.C., P.V. Dattatreya, K.H. Srinivasa, Prabhavathi, et C.N. Manjunath. 2012. « Inferior vena cava obstruction: long-term results of endovascular management ». *Indian Heart Journal* 64 (2): 162-69. doi:10.1016/S0019-4832(12)60054-6.
- Terabayashi, H., K. Okuda, F. Nomura, K. Ohnishi, et P. Wong. 1986. « Transformation of Inferior Vena Caval Thrombosis to Membranous Obstruction in a Patient with the Lupus Anticoagulant ». *Gastroenterology* 91 (1): 219-24.
- Wu, Tongguo, Lexin Wang, Qiang Xiao, Bosong Wang, Shanying Li, Xiurong Li, et Jinying Zhang. 2002. « Percutaneous Balloon Angioplasty of Inferior Vena Cava in Budd-Chiari Syndrome-R1 ». *International Journal of Cardiology* 83 (2): 175-78.
- Zhou, Pengli, Jianzhuang Ren, Xinwei Han, Gang Wu, Wenguang Zhang, Pengxu Ding, et Yonghua Bi. 2014. « Initial Imaging Analysis of Budd-Chiari Syndrome in Henan Province of China: Most Cases Have Combined Inferior Vena Cava and Hepatic Veins Involvement ». *PLOS ONE* 9 (1): e85135. doi:10.1371/journal.pone.0085135.

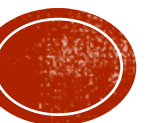


PÉRIODE DE QUESTIONS



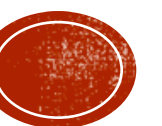
LUPUS

It's NEVER lupus.



ANNEXE 1 : TECHNIQUES

- § VCI mesure 20-22mm,
- § Inoue idéal 1:1 à 1.1:1 (ballon/VCI)
- § 1-2 ATM, 3 secondes



Lupus Anticoagulant-Hypoprothrombinemia Syndrome

Report of 8 Cases and Review of the Literature

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TABLE 2. Epidemiology and Associated Diseases in 74 Patients With LAHS

Characteristic	No. (%)
Epidemiology	
Women	43 (58)
Age at diagnosis, yr, mean (range)	22.7 (2–76)
Age <15 yr	43 (58)
Associated disease*	
Autoimmune disease	41 (55)
SLE	28 (38)
Lupus-like syndrome	5 (7)
Primary antiphospholipid syndrome	5 (7)
Discoid lupus	1 (1)
Celiac disease	1 (1)
Autoimmune hepatitis	1 (1)
Infectious disease	25 (33)
Viral	23 (31)
Bacterial	2 (3)
Lymphoma	2 (3)
Drugs (phenytoin and quinidine)	2 (3)
No underlying disease	7 (9)

*Total number of patients is more than 74 because 3 patients had both an autoimmune disease and an acute infection.

TABLE 4. Laboratory Features of LAHS

Feature	n*	Value
PT and factor II level		
PT, median (range)	49	1.7 N (1–3.2 N)
Factor II level, median (range)	63	11% (1%–40%)
Antiprothrombin antibodies, positivity, n (%)	33	29 (88%)
Antiprothrombin antibodies, titer, median (range)	10	5.5 N (2–34 N)
aPTT and antiphospholipid antibodies		
aPTT, median (range)	65	2 N (1.5–4 N)
Lupus anticoagulant, positivity, n (%)	74	74 (100%)
aCL, positivity, n (%)	47	33 (70%)
aCL IgG, titer, median (range)	14	6 N (1.3–366 N)
Anti-β2GP1, positivity, n (%)	11	8 (73%)
Anti-β2GP1 IgG, titer, median (range)	5	12 N (1.5–66)
Thrombocytopenia, n (%)	63	15 (24%)

Abbreviations: N = number times the upper value of the normal range.

*Number of patients with data available.

