Cholangite sclérosante primitive diagnostic et prise en charge

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Rabat

FMC SMMAD 2018

Tanger 29 Nov2018

Objectifs pédagogiques

- Epidémiologie
- Ethiopathogénie
- Histoire naturelle
- Présentations cliniques
- Moyens diagnostiques
- Traitement
  - Médical
  - Endoscopique
  - TH
- surveillance

Incidence/Prévalence

- Europe du nord : 0.5 - 1.3 /100 000 /an
- Amérique du nord : 3.85 - 16.2 / 100,000/an
- Espagne, Singapore Japan : 0.022, 0.13 , 0.095 / 10,000

Prévalence CSP si RCH

Author	Year	Country	Number of patients	PSC %
Aitola[60]	1994	Finland	534	2%
Broome[61]	1994	Sweden	1274	2.3%
Lupinetti[63]	1980	USA	202	1%
Olsson[64]	1991	Sweden	1500	3.7%
Rasmussen[65]	1992	Denmark	305	3.6%
Schrumpf[66]	1982	Norway	336	5.4%
Shepherd[67]	1983	UK	681	2.4%
Hashimoto[68]	1993	Japan	163	3.1%
Bernstein[69]	2001	Canada	4454	Men: 3% Women: 1%
Tobias[71]	1983	South Africa	250	3.2%
Wewer[72]	1991	Denmark	396	0.75%

E Gizard ,Aliment Pharmacol Ther 2014

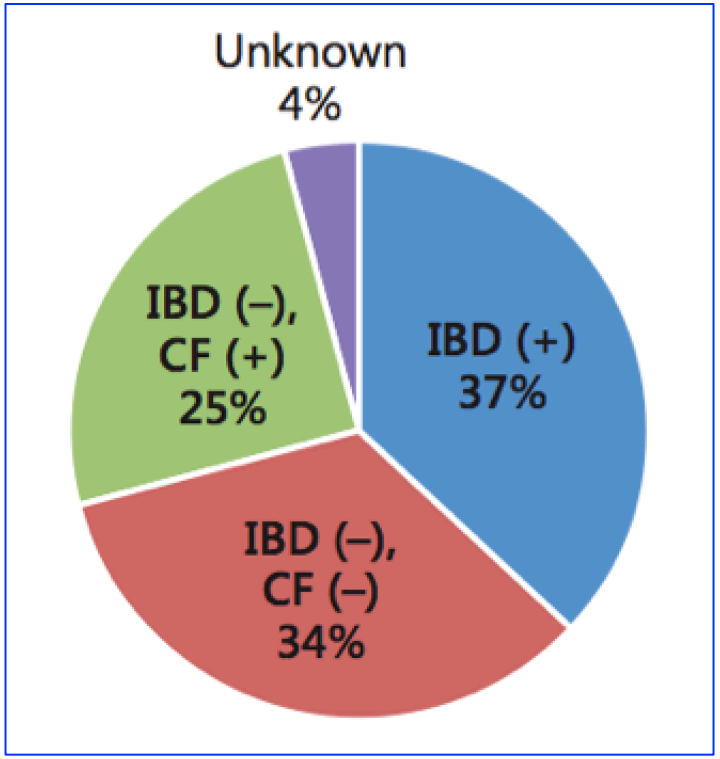
Facteurs de Risque de CSP si MICI résultats du registre suisse

- 2744 patients [1188 RCH 1556 Crohn)
- 57 avaient une CSP (48 RCH et 9 CROHN).
- Prévalence **4.04 vs. 0.58%, P<0.001**
- Facteurs de risque
  - Sexe masculin [OR 2.771, P=0.022],
  - Pancolite (OR 2.855, P=0.011)
  - Non fumeur (OR 9.253, P=0.030),
  - ATCD appendicectomie (OR 4.114, P=0.019)

M Fraga ,European Journal of Gastroenterology & Hepatology 2017

Prévalence de RCH si CSP

Pays	Prévalence
France	60 %
Espagne	44 %
Turquie	43-63 %
Iran	62 %
Inde	50 %
Japon	34-37 %



TH Karlsen,Journal of Hepatology 2017

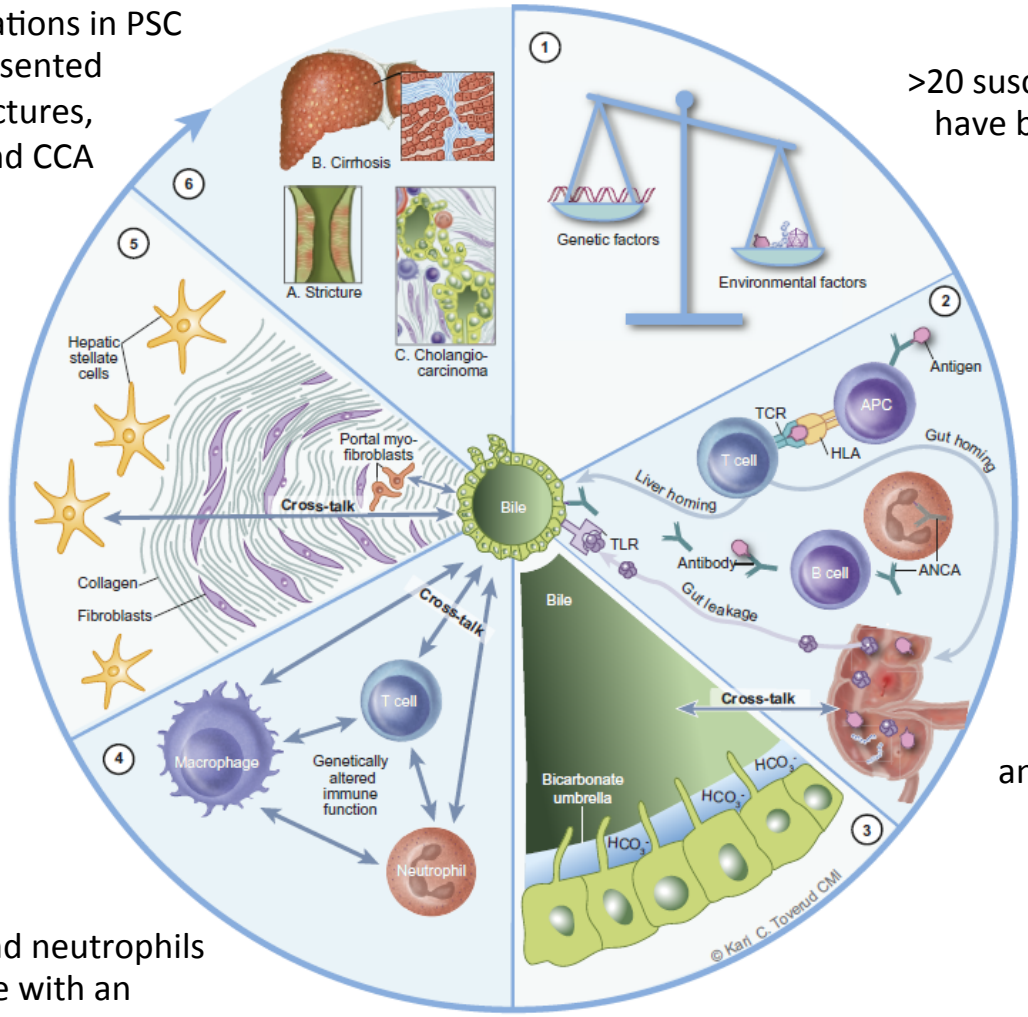
Iatsushi T, nflamm Intest Dis 2016

physiopathologie

Liver-related complications in PSC are mainly represented by bile duct strictures, liver cirrhosis and CCA

>20 susceptibility genes have been identified

Chronic injury + common fibrotic mechanisms involving hepatic stellate cells and portal myofibroblasts in an undefined cross-talk with cholangiocytes



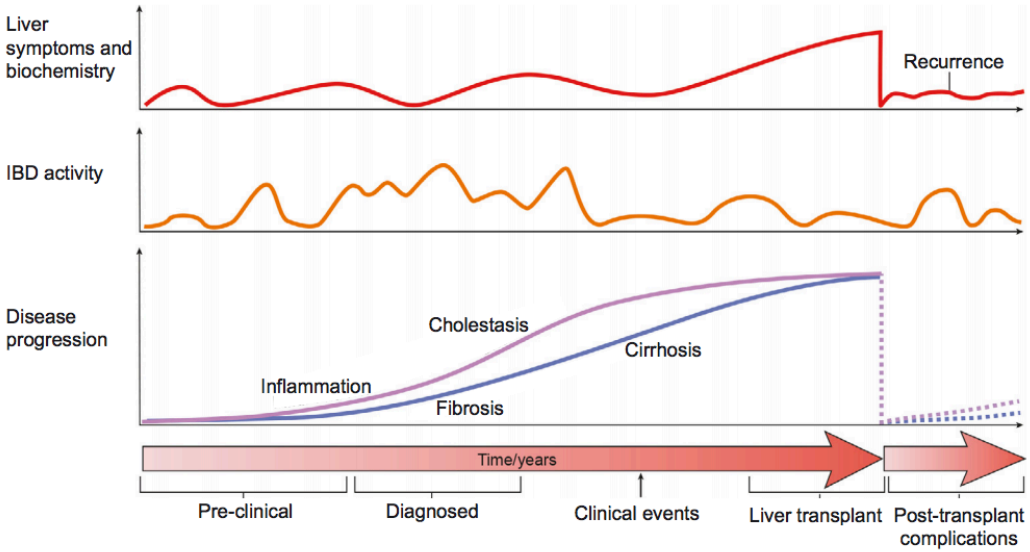
Role of gut-derived antigens remains unclear

T cells, macrophages and neutrophils are likely to engage with an activated cholangiocyte phenotype

Primary or secondary disturbances in bile homeostasis are thought to contribute to PSC

1. Karlsen TH, et al. J Hepatol 2017;67:1298–323;
ESGE/EASL CPG Endoscopy in PSC. J Hepatol 2017;66:1265–81

CSP : Histoire naturelle et causes de mortalité



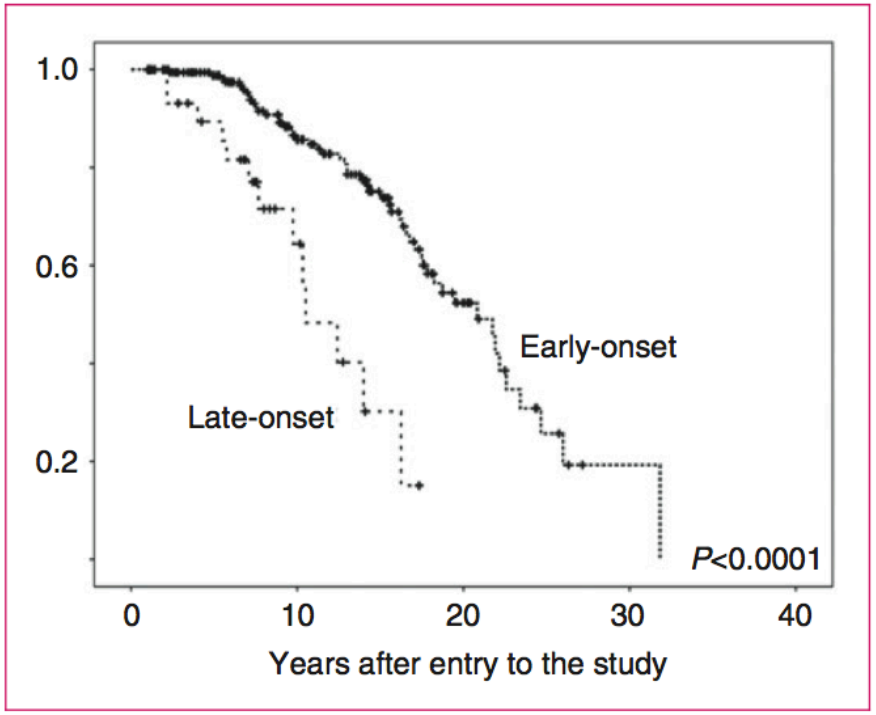
Causes de mortalité

- CCA (32%)
- IHC (15%)
- Complications de la TH (9%)
- CCR (8%)

TH Karlsen,Journal of Hepatology 2017

Impact of age at diagnosis on disease progression in patients with primary sclerosing cholangitis

Christian Rupp[1,2], Alexander Rössler[1], Taotao Zhou[1], Conrad Rauber[1],



Patients with later age at diagnosis
increased risk for progressive liver failure and biliary malignancies

United European Gastroenterology Journal 2018

Patients with large-duct primary sclerosing cholangitis and Crohn's disease have a better outcome than those with ulcerative colitis, or without IBD

J. Fevery*, W. Van Steenbergen*, J. Van Pelt*, W. Laleman*, I. Hoffman†, K. Geboes‡, S. Vermeire§ & F. Nevens*

Conclusions

The prevalence of PSC with concomitant Crohn's disease is relatively rare, but the outcome is more benign than PSC with UC or without IBD. Approximately one-fourth has small-duct PSC. In large-duct PSC/CD, liver disease is less aggressive and the outcome is much better. The outcome of PSC patients with UC resembled that of PSC without IBD.

Aliment Pharmacol Ther 2016; **43:** 612–620

Présentation typique

- Homme de 30- 40 ans
- Suivi pour MICI (RCH)
- cholestase +/- cytolyse

S Sirpal, Clinical and Experimental Gastroenterology 2017

Particularités des CSP associées à une MICI

- Survenue à un âge plus jeune
- Formes de passage plus fréquentes (CSP-HAI)
- Pas de parallélisme sévérité CSP/RCH.
- Colectomie: pas de modification de la CSP

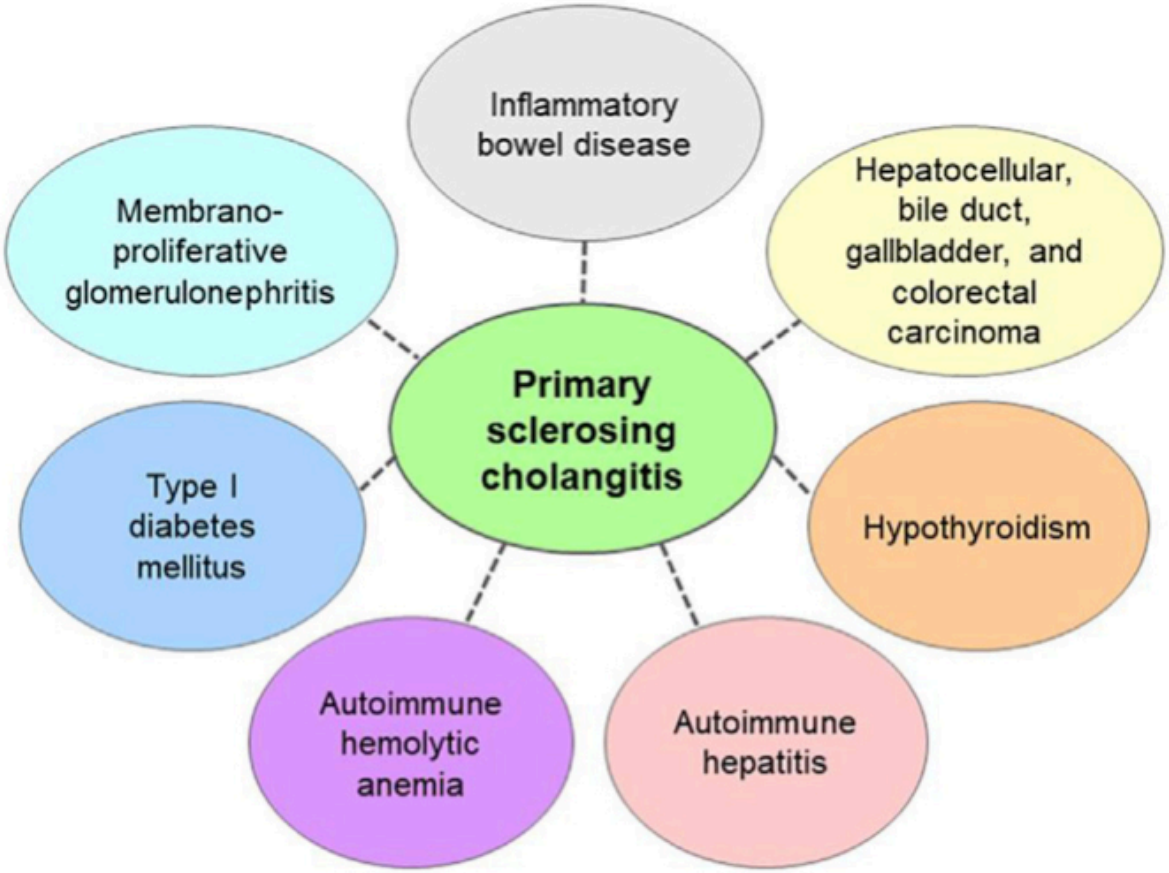
Autres modes de découverte

- Découverte fortuite de cholestase
- Prurit ,ictère ,asthénie
- angiocholite
- Cirrhose décompensée
- Cholangiocarcinome
- 25 % des cas : pathologies auto-immune associée

Tom H. Karlsen Journal of Hepatology 2017

Jessica K Dyson ,Lancet 2018

Pathologies pouvant être associées à la CSP



JH Tabibian ,Liver Research 2017

RCH avec CSP :Phénotype particulier

- Souvent pas d'atteinte rectale : 18-56 % vs 10 %
- Pancolite dans 60 % vs 50 %
- Atteinte colon droit fréquente
- iléite de reflux : 20–51%
- Fréquence des formes indéterminées
- Forme souvent bénigne
- Risque du cancer du colon élevé (10-15%)
- Cancer souvent du colon droit

Iatsushi T, nflamm Intest Dis 2016

M.Harbord ,Journal of Crohn's and Colitis, 2016

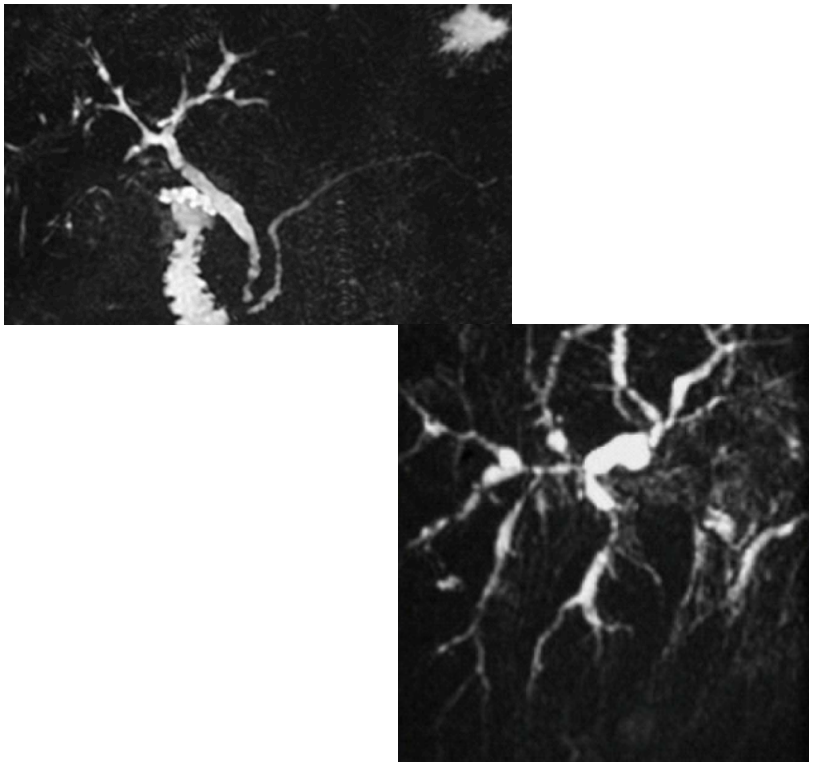
Moyens diagnostiques

- IRM +++
- CPRE
  - si dic douteux aprés IRM+PBF
  - Si biopsies ou cytologie necessaire
  - si Tx endoscopique indiqué

1. Weismüller TJ, et al. J Hepatol 2008;48 Suppl 1:S38–57;
ESGE/EASL CPG Endoscopy in PSC. J Hepatol 2017;66:1265–81

Recommendations on the Use of Magnetic Resonance Imaging in PSC-A Position Statement From the International PSC Study Group

MRCP should be the first diagnostic imaging modality in patients with suspected PSC. (1A)



If the initial MRI/MRCP at the time of establishing a PSC diagnosis has been performed without contrast media, a second MRI/MRCP including contrast media should be considered within 6 months of the diagnosis because of the higher risk of prevalent CCA when PSC is detected. (1C)

C. Schramm, Hepatology 2017

Prevalence of Sclerosing Cholangitis Detected by Magnetic Resonance Cholangiography in Patients With Long-term Inflammatory Bowel Disease

- 222 RCH et 100 Crohn surveillance à long terme (20 ans)
- 24 patients (7.5%) : lésions évoquant une CSP
- seulement 7 patients (2.2%) connu porteur de CSP
- Avec l'IRM dans le suivi ; on détecte 3 x plus de CSP asymptomatique
- 65 % des patients avaient une CSP sans anomalies biochimiques

A K Lunder Gastroenterology 2016

Anomalies biologiques associées

- NFS
- Transaminases GGT Bili PA
- EPP
- Ig G4
- Ac anti mitochondries
- Ac anti muscles lisses

Serologic finding	Prevalence
Hypergammaglobulinemia	30%
Increased serum immunoglobulin M (IgM) levels	40–50%
Atypical perinuclear antineutrophil cytoplasmic antibodies (P-ANCA)	30–80%
Human leukocyte antigen DRw52a	0–100% in various reports
Increased immunoglobulin G4 (IgG4)	9%

Chapman , LINICAL LIVER DISEASE, VOL 9, NO 5, MAY 2017

S Sirpal, Clinical and Experimental Gastroenterology 2017

CSP et Ig G4 positif

CSP +Augmentation Ig 4
fréquence : 9 %
Immunohisto : 23 %
CSP mauvais Pic
TH précoce
Iléite associée
Colectomie fréquente

U Navaneetham,Gastroenterology 2014

Diagnostic différentiel entre CSP et cholangiteIg G4

Feature	Primary sclerosing cholangitis	IgG4 cholangiopathy or autoimmune cholangiopathy
Age	Young to middle age	Older
Obstructive jaundice	Marked	Less severe
Biliary strictures	Band-like strictures with beaded and pruned tree appearance	Segmental strictures (distal CBD)
Associated autoimmune pancreatitis	No	Yes
IgG4-related diseases	No	Yes
IBD association	Yes	Rarely associated with type 2 AIP
Risk of cholangiocarcinoma	Yes	Rare (with limited data), can mimic

Ig G4 > 4 x N
IgG4/IgG1 > 0.24
PCR pour mesure de ce rapport

Hisham Al-Dhahab Can J Gastroenterol 2013

Jessica K Dyson ,Lancet 2018

Indications de la PBF

- Doute de CSP des petits canaux avec IRM normale
- Ig G4 élevée
- HAI associée

Fibroscan dans la CSP

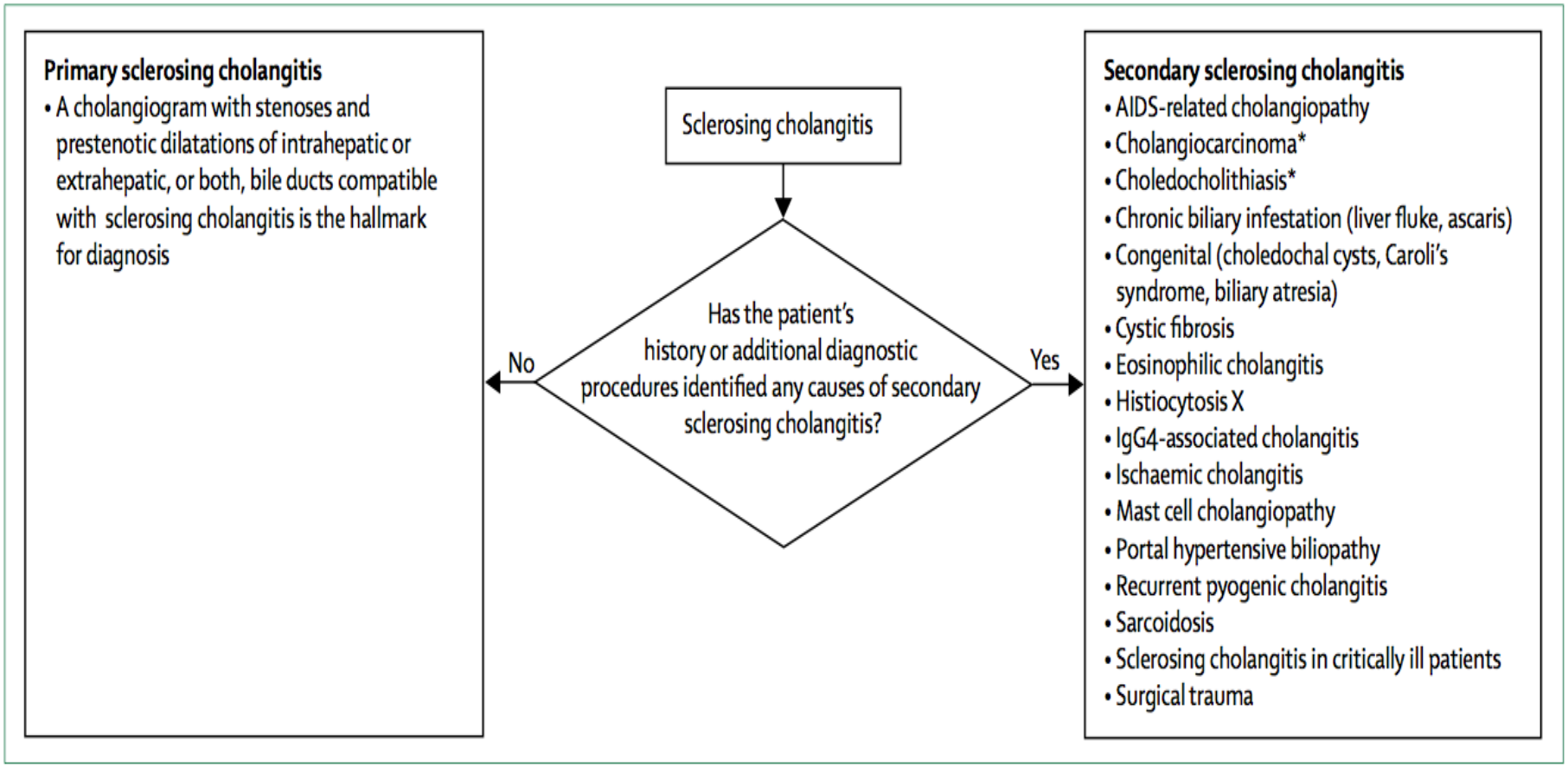
Stage	n	Cutoff (*kPa*)	AUROC
All patients (n = 66)			
≥F1	60	7.4	0.71 (0.69)
≥F2	32	8.6	0.84 (0.79)
≥F3	15	9.6	0.93 (0.90)
F4	9	14.4	0.95 (0.91)

Corpechot C et al Gastroenterology 2014

Stage	n	AUROC	95% CI	Cutoff (kPa)	SE	95% CI	SP	95% CI
All patients	62							
≥ F1	57	0.63	0.44 to 0.82	6.6	64.9	52.5 to 77.3	60	17.1 to 100
≥ F2	27	0.91	0.82 to 0.99	8.8	81.5	66.8 to 96.1	88.6	78 to 99.1
≥ F3	20	0.95	0.89 to 1.0	9.6	90	76.9 to 100	90.5	81.6 to 99.4
F4	16	0.978	0.93–1.0	14.4	68.8	46 to 91.5	97.8	93.6 to 100

Ehlken H et al Plos One 2016

Diagnostic différentiel



J K Dyson, Lancet 2018

CSP :Critères diagnostiques

- Cholestase sans causes évidentes
- Aspect typique en IRM
- Pas de cholangites secondaires

Diagnosing PSC	Diagnostic approach
Classical large-duct PSC	Cholestatic serum liver tests (elevated ALP) GGT) and/or cholestatic symptoms (pruritus, jaundice, cholangitis), MRC, diagnostic ERC (occasionally)
Small-duct PSC (MRC/ERC normal)	Liver biopsy, IBD confirmed
PSC high IgG4	Serum IgG4, IgG4 staining of relevant biopsy
PSC with features of AIH (PSC-AIH overlap)	ANA, anti-SMA, anti-LKM, anti-SLA/LP, IgG, liver biopsy

TH Karlsen,Journal of Hepatology 2017

Traitement

Traitement du Prurit

Recommendations

1. Local skin treatment should be performed with emollients and/or antihistamines in patients with PSC and mild pruritus, to reduce symptoms. (Conditional recommendation, very low quality of evidence) (123,124)
2. Bile acid sequestrants such as cholestyramine should be taken (prescribed) in patients with PSC and moderate pruritus to reduce symptoms. Second-line treatment such as rifampin and naltrexone can be considered if cholestyramine is ineffective or poorly tolerated. (Conditional recommendation, very low quality of evidence) (124–126)

ACG 2015

Chercher ostéoporose et déficit en vit liposoluble

Patients with PSC should undergo bone mineral density (BMD) screening at diagnosis with dual energy X-ray absorption at diagnosis and repeated at 2- to 4-year intervals. (Conditional recommendation, moderate quality of evidence) (128)

Patients with advanced liver disease should be screened and monitored for fat-soluble vitamin deficiencies. (Conditional recommendation, moderate quality of evidence) (129)

ACG 2015

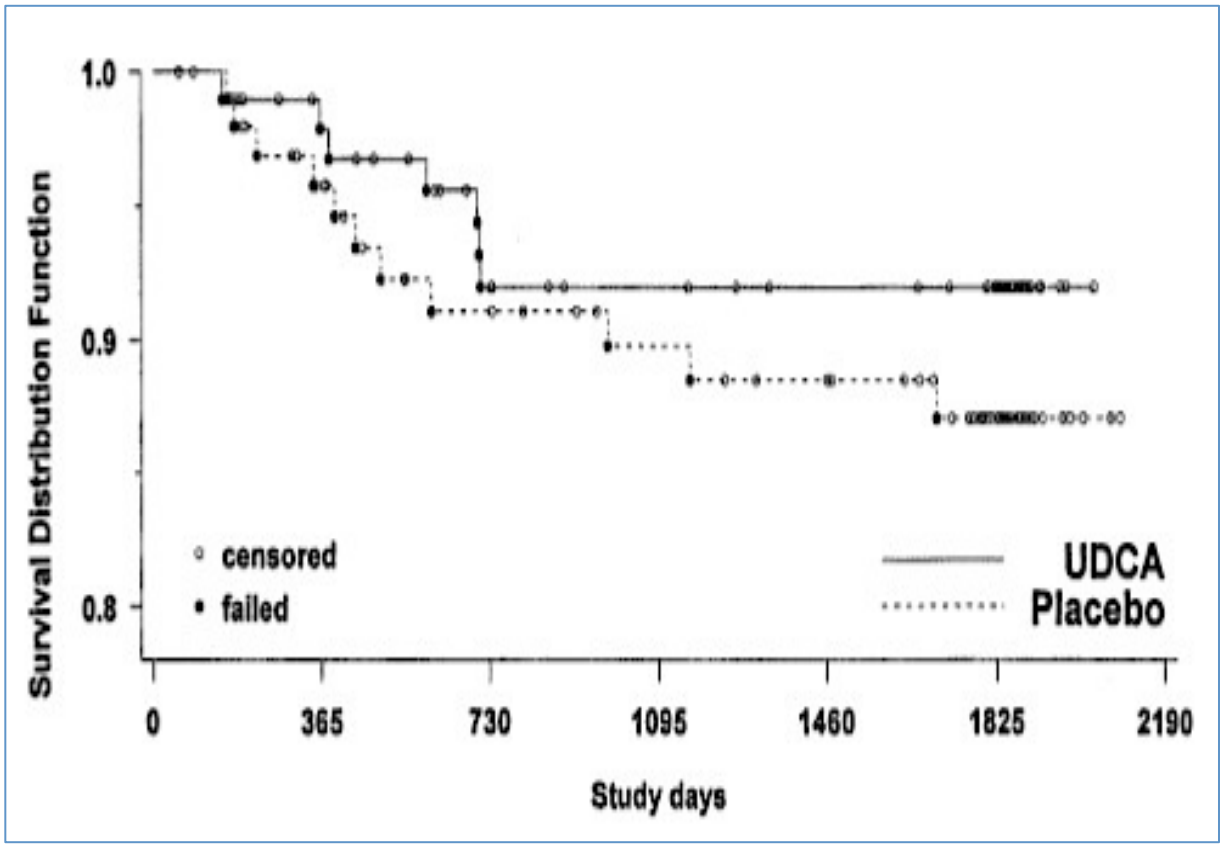
15. In patients with hepatic osteopenia, we suggest the use of calcium 1.0-1.5 g and vitamin D 1,000 IU daily for therapy (2C).

16. In patients with hepatic osteoporosis, we suggest the use of bisphosphonate therapy in addition to calcium and vitamin D supplementation (2C).

AASLD 2010

AUDC et CSP

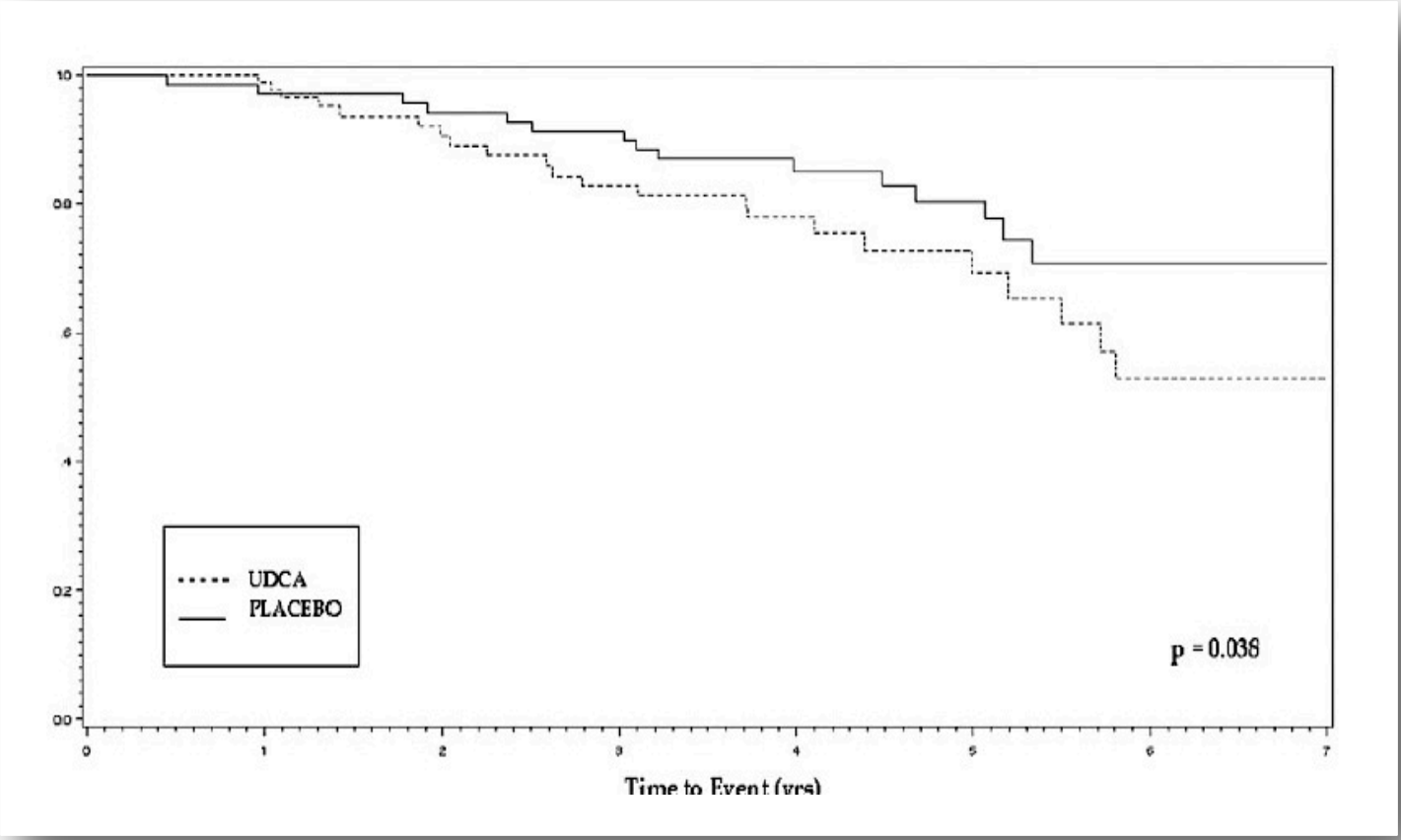
17-23 mg/kg/j



RTC avec 219 patients
tendance à améliorer la survie

Olsson et al. Gastroenterology 2005

AUDC forte dose
Effet délétère
AUDC 28-30 mg/kg/j *vs.* Placebo



Lindor et al. Hepatology 2009

URSO forte dose

- Risque plus élevé de
  - Cirrhose,VO
  - cholangiocarcinome voir de CCR
  - Transplantation hépatique

Lindor KD, Hepatology 2009

Eaton JE, Am J Gastroenterol 2011

AUDC et CSP: que fait-on ?

Recommendations:

28. In adult patients with PSC, we recommend against the use of UDCA as medical therapy (1A).

29. In adult patients with PSC and overlap syndrome, we recommend the use of corticosteroids and other immunosuppressive agents for medical therapy (1C).

AASLD recommendations 2010

Recommendations

1. The available data base shows that UDCA (15–20 mg/d) improves serum liver tests and surrogate markers of prognosis (I/B1), but does not reveal a proven benefit on survival (III/C2). The limited data base does not yet allow a specific recommendation for the general use of UDCA in PSC.

EASL recommendations 2009

CME

ACG Clinical Guideline: Primary Sclerosing Cholangitis

Keith D. Lindor, MD, FACG[1,2], Kris V. Kowdley, MD, FACG[3] and M. Edwyn Harrison, MD[2]

Recommendation

1. Ursodeoxycholic acid (UDCA) in doses >28 mg/kg/day should not be used for the management of patients with PSC. (Strong recommendation and high quality of evidence) (42)

Am J Gastroenterol 2015;

Que faire en pratique ?

Acide ursodésoxycholique (AUDC) : 15-20 mg/kg/j (AMM 2012). Les posologies très fortes 28-30 mg/kg/j sont contre-indiquées

Société Nationale Française de Gastro-Entérologie
SNFGE

2014 Olivier Chazouillères

Conseil de pratique

AFEF
ASSOCIATION FRANÇAISE POUR L'ETUDE DU FOIE

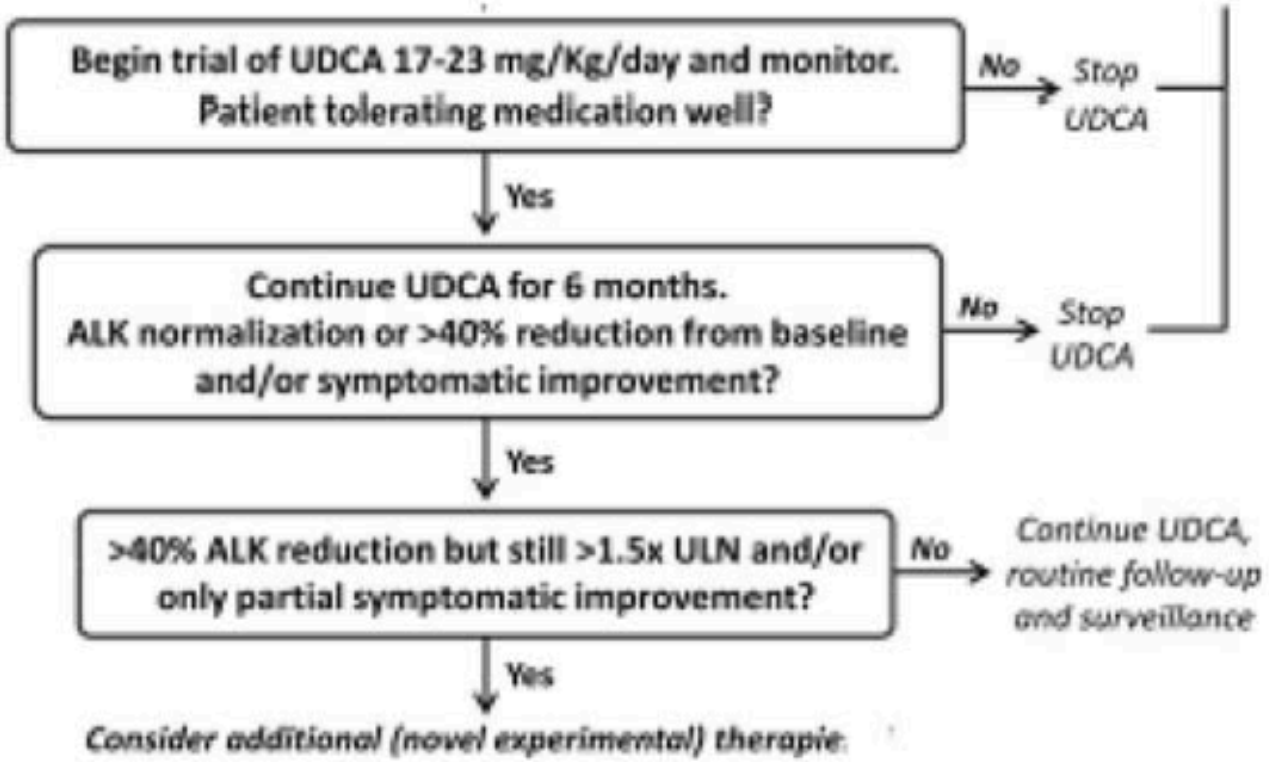
La plus part des experts proposent
une dose de 18-21 mg/kg

James H. Tabibian, MD, PhD, Ahmad H. Ali, MBBS, and Keith D. Lindor, MD

Gastroenterology & Hepatology Volume 14, Issue 5 May 2018

Algorithm for UDCA Use in PSC

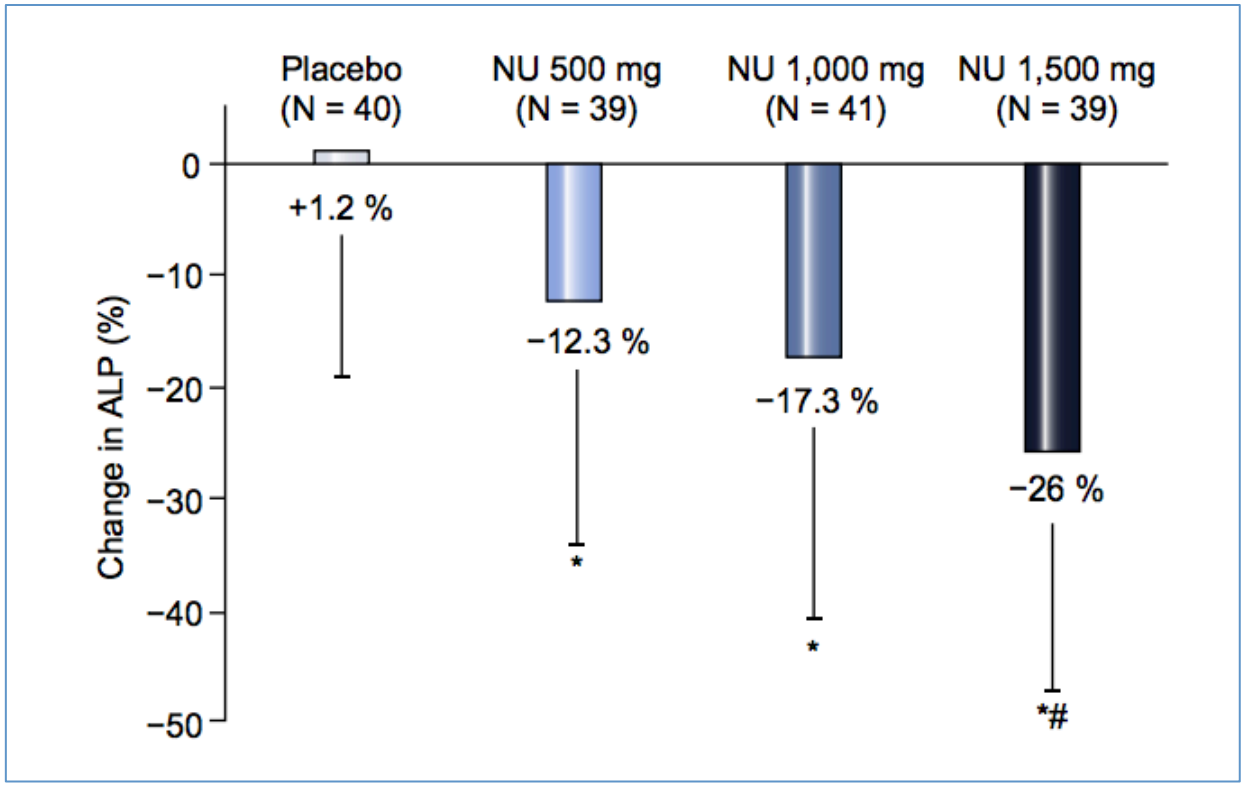
*Proposed by Tabichian and Lindor**



*Hepatology 2014;60:785

*nor*Ursodeoxycholic acid improves cholestasis in primary sclerosing cholangitis

Peter Fickert[1], Gideon M. Hirschfield[2], Gerald Denk[3], Hanns-Ulrich Marschall[4], Istvan Altorjay[5],



J HEPATOL 2017

Vancomycine dans le Tx la CSP

Agent(s)	Sample Size, n	Study Design	Summary of Main Findings
Vancomycin in pediatric patients[95,96,99,146,147]	19 (total of the 5 studies)	Open-label, pilot, and case series	• Improvement in GGT, ALT, C-reactive protein, symptoms, and cholangiographic findings
Vancomycin or metronidazole in adults[98]	8 vs 9 vs 9 vs 9 (low-dose vs high-dose vancomycin vs low-dose vs high-dose metronidazole)	Randomized, double-blind	• Improvement in ALP (primary endpoint) in the vancomycin groups • Decrease in bilirubin in the low-dose metronidazole group and trend toward significant decrease in the low-dose vancomycin group (P=.06) • Decrease in Mayo PSC risk score in the low-dose vancomycin group and low-dose metronidazole group
Vancomycin[97]	18 vs 11 (drug vs placebo)	Triple-blind, randomized, placebo-controlled	• Improvement in ALP, GGT, and symptoms

J H Tabibian ,Gastroenterology & Hepatology 2018

Damman JL, Rodriguez EA, Ali AA, et al. Review article: the evidence that vancomycin is a therapeutic option for primary sclerosing cholangitis. *Aliment Pharmacol Ther*. 2018;47:886-895.

Editorial: vancomycin – a promising option for the treatment of primary sclerosing cholangitis?

- AP&T Alimentary Pharmacology & Therapeutics - 2018

R. W. Chapman[1,2]

[1]*Nuffield Department of Medicine, Oxford University, Oxford, UK*

Statine et CSP

- Swedish, register-based cohort study of PSC patients with IBD (n=2914) diagnosed between 2005 and 2016
- Cox regression used to analyze associations between different drugs and: death, LTx, CCA, and bleeding oesophageal varices
- Statin exposure: 13.9% (n=404)

Hazard ratios (95% CIs)

Drug	All-cause mortality (n=2914)	Mortality and liver transplantation (n=2794)	Adverse liver events* (n=2740)
UDCA	1.04 (0.87, 1.25)	1.34 (1.12, 1.62)	3.10 (2.36, 4.07)
Statins	**0.68 (0.54, 0.88)**	**0.50 (0.28, 0.66)**	**0.53 (0.36, 0.80)**
NSAIDs	0.86 (0.72, 1.02)	0.82 (0.68, 0.99)	0.87 (0.68, 1.62)
ASA	0.99 (0.80, 1.21)	2.16 (1.72, 2.70)	3.35 (2.46, 4.55)
Antibiotics	1.70 (1.27, 2.29)	2.27 (1.70, 3.05)	3.03 (2.09, 4.41)
Antimycotics	2.78 (2.24, 3.44)	3.13 (2.48, 3.94)	1.74 (1.20, 2.51)
Metronidazole	1.27 (1.06, 1.53)	1.20 (0.99, 1.47)	1.58 (1.23, 2.03)
AZA/mercaptopurines	0.66 (0.52, 0.84)	0.65 (0.50, 0.83)	0.80 (0.60, 1.08)
Steroids	1.94 (1.60, 2.34)	2.14 (1.75, 2.60)	1.28 (1.00, 1.65)

Conclusions: statin use associated with decreased risk of death and LTx in PSC

*Liver-related death, LTx, CCA or variceal bleeding

Stokkeland K, et al. ILC 2018, #PS-128

fibrates + acide ursodeoxycholique
expérience Franco-espagniole

- 20 Patients en réponse biochimique incomplète à Urso (PA > 1.5 N)
- Elastometrie : 11kPa (≥F3)
- fibrates (fenofibrate 200mg/day or bezafibrate 400mg/day) / 6 mois
- Résultats :
  - Diminution du prurit
  - Réduction des PA de 41 %
  - L'arret des fibrates : augmentation des PA
  - Fibrates bien toléré
  - Augmentation de l'elasticité

Lemoinne S et al,Clin Res Hepatol Gastroenterol. 2018

Quand Tx endoscopique

Recommendations		
Traitement Endoscopique avec prelevement sur les stenoses suspectes **est suggéré chez les patients symptomatiques qui profiteront de ce Tx endoscopique**	elevé	faible

*Strength of recommendation, weak; quality of evidence, low
ESGE/EASL CPG Endoscopy in PSC. J Hepatol 2017;66:1265–81

CSP : Quel traitement endoscopique et chez qui ?

4. ESGE/EASL suggest that a dominant stricture at ERCP should be defined as a stenosis with a diameter of ⩽1.5 mm in the common bile duct and/or ⩽1.0 mm in an hepatic duct within 2 cm of the main hepatic confluence.
Weak recommendation, low quality evidence.

8. ESGE/EASL suggest that the choice between stenting and balloon dilation should be left to the endoscopist's discretion.
Weak recommendation, low quality evidence.

13. ESGE/EASL suggest that stents used for treating dominant stricture should be removed 1–2 weeks following insertion.
Weak recommendation, low quality evidence.

EASL 2017

Résultats du traitement endoscopique

- 171 PSC suivi à long terme (moy de 20 ans)
  - Traitement endoscopique répété
  - Survie sans TH après la première endoscopie
    - à 5 ans : 81%
    - à 10 ans : 52%
- 6 % des sténoses dominantes étaient des CCA

Gotthard DN,Gastrointest Endosc 2010

dilation vs. dilation + prothese

- Etudes retrospectives :
  - Amelioration de la Survie sans TH avec dilatation
  - Complications plus fréquente avec prothese

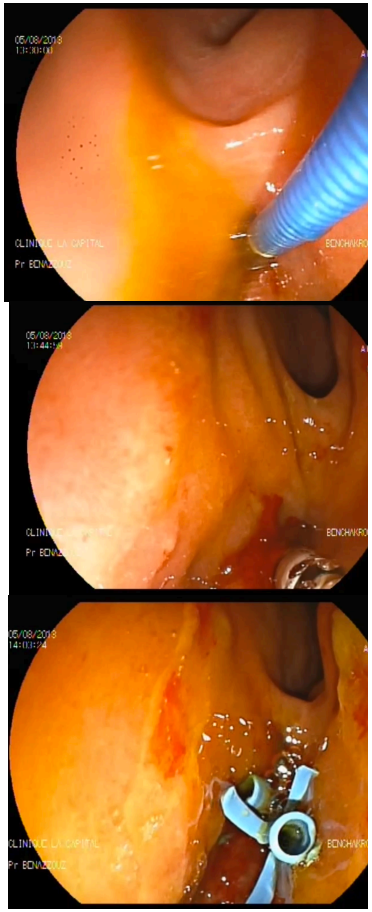
- Etude DILSTENT dilation vs prothèse a été arrêtée
  - Pas de diffrence : echec initial et recidive
  - Plus de complications pancreatite et angiocholite , (42%) vs. (10%)

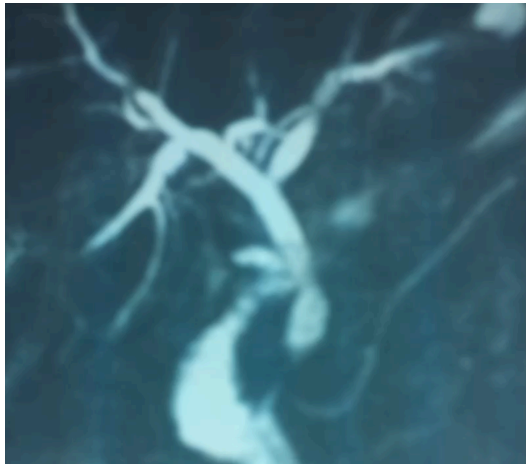
*≤2 weeks

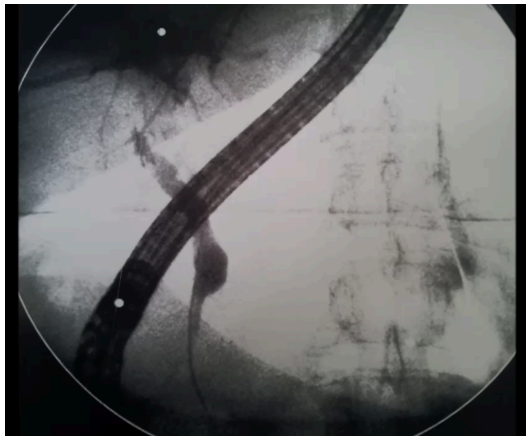
1. Ponsioen C, et al. J Hepatol 2017;66:S1–2;
ESGE/EASL CPG Endoscopy in PSC. J Hepatol 2017;66:1265–81

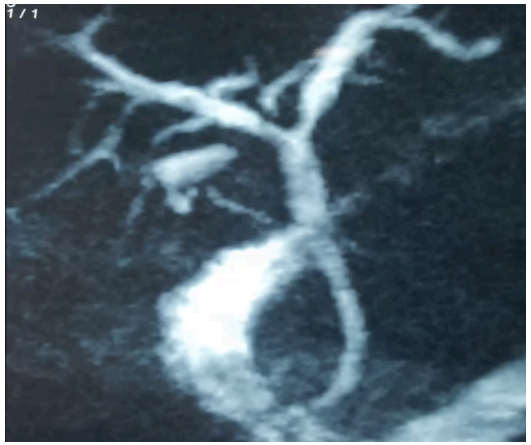
Cas clinique

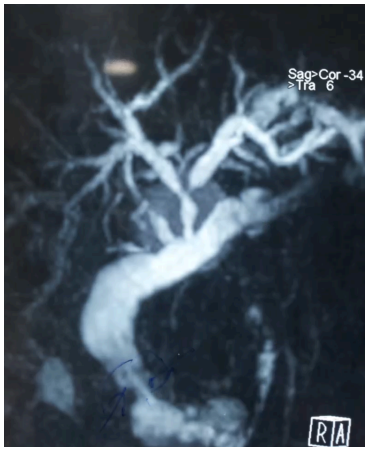
- BN ,55 ans
- ATCD d'angiocholite
- Pas de trouble de transit
- Consulte pour subictère
- Biologie : cytolyse et cholestase
- Echo : DVBIH et VBP
- IRM : 2 sténoses avec 2 petits calculs VBP
- Ig 4 sérique : N
- Tx endoscopique : SE + dilatation et extraction des calculs
- 8 mois :Récidive de la sténose :Stent 10Fr
- 13mois plus tar: récidive : 2 stents 7Fr
- Evolution à 5 ans : bonne (après 5 CPRE)
- Sous Urso
- colo initiale et à 5 ans : RAS











Indications de la TH

- Ictère prolongé avec bilirubinémie > 100 µmol/l
- Episodes répétés et sévères d'angiocholites
- Cirrhose décompensée (Child-Pugh > 9 ou MELD > 15).
- Cholangiocarcinome hilaire < 3 cm sans atteinte ganglionnaire et inclus dans un protocole très strict de radio-chimiothérapie pré-TH

LIVER TRANSPLANTATION

Recommendations

1. Liver transplantation, when possible, is recommended over medical therapy or surgical drainage in PSC patients with decompensated cirrhosis, to prolong survival. (Strong recommendation, moderate quality of evidence) (94–96)
2. Patients should be referred for liver transplantation when their Model for End-Stage Liver Disease (MELD) score exceeds 14. (Conditional recommendation, moderate quality of evidence) (97)

ACG 2015

TH pour CSP
évolution à long terme

- 45 TH pour CSP
- 40 % Récidive de la CSP (recul moyen 30 mois)
  - 24.5% à 3 ans
  - 39.3% à 5 ans
  - 45.8% à 6 ans
- Survie du greffon
  - 56.3% à 5 ans après récidive
  - 21,9 % à 10 ans après récidive

Y Ueda ,Transplant Direct. 2017

CSP et risque de cholangio-carcinome(CCA) et Cancer colorectal

- Prévalence de néoplasie si CSP 13–14%
  - CCA (11%)
  - CCR (9%).
- Le risque de CC augmente si
  - Ancienneté de la RCH >15 ans
  - Pancolite
  - indépendamment de la sévérité de la maladie
- Le risque de CCR est
  - multiplié par 5 si CSP
  - multiplié par 9 si MICI avec CSP

Fang Y ,Austin J Gastroenterol 2015

RCH/CSP :AUDC et prévention du CCR

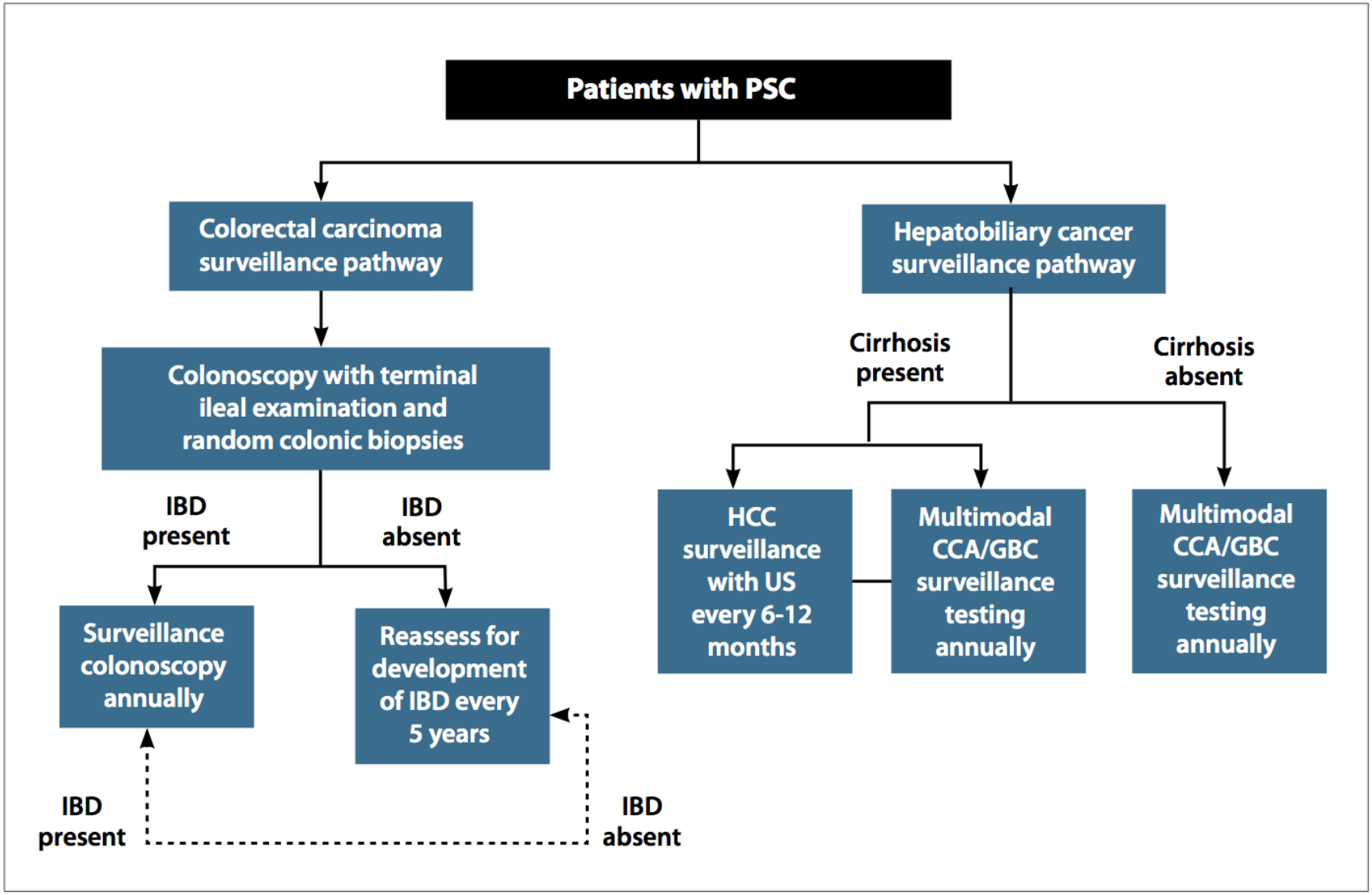
Study type	Is UDCA chemopreventive?
RCT	Yes
Retrospective	No
RCT	No
RCT	No-high dose UDCA
Prospective	No-short term; yes-long term

Méta-analyse 177 CCR/763 MICI/CSP
pas d'effet préventif : OR = 0.81, 95% CI: 0.41-1.61.
Effet préventif contre néoplasie avancé

Singh S,Inflamm Bowel Dis 2013;

W Rosy,World J Gastroenterol 2014

Surveillance



J H Tababian, Gastroenterology & Hepatology 2018

surveillance Endoscopique si CSP/RCH

Recommendations	Strength of recommendation	Quality of evidence
Ileocolonoscopiede dépistage au moment du Dic de la CSP	elevé	elevé
Si MICI coloileoscopie annuelle	elevé	faible
Si pas de MICI coloileo à 5 ans ou chaque fois qu'il y a des symptomes évoquant MICI	faible	faible
Ileocolonoscopie avec biopsies dans les 4 quadrants au niveau du colon et de l'iléon	elevé	faible
Ileocolonoscopie avec chromoendoscopie avec biopsies dirigées si lesions dysplasiques	elevé	faible

ESGE/EASL CPG Endoscopy in PSC. J Hepatol 2017;66:1265–81

CCA sur CSP : diagnostic

ESGE/EASL 2017

17. EASL/ESGE recommend that cholangiocarcinoma (CCA) should be suspected in any patient with worsening cholestasis, weight loss, raised serum CA19-9, and/or new or progressive dominant stricture, particularly with an associated enhancing mass lesion.
Strong recommendation, moderate quality evidence.

18. A raised serum CA19-9 may support the diagnosis of CCA, but has a poor specificity.
Weak recommendation, low quality evidence.

19. ESGE/EASL recommend ductal sampling (brush cytology, endobiliary biopsies) as part of the initial investigation for the diagnosis and staging of suspected CCA in patients with PSC.
Strong recommendation, high quality evidence.

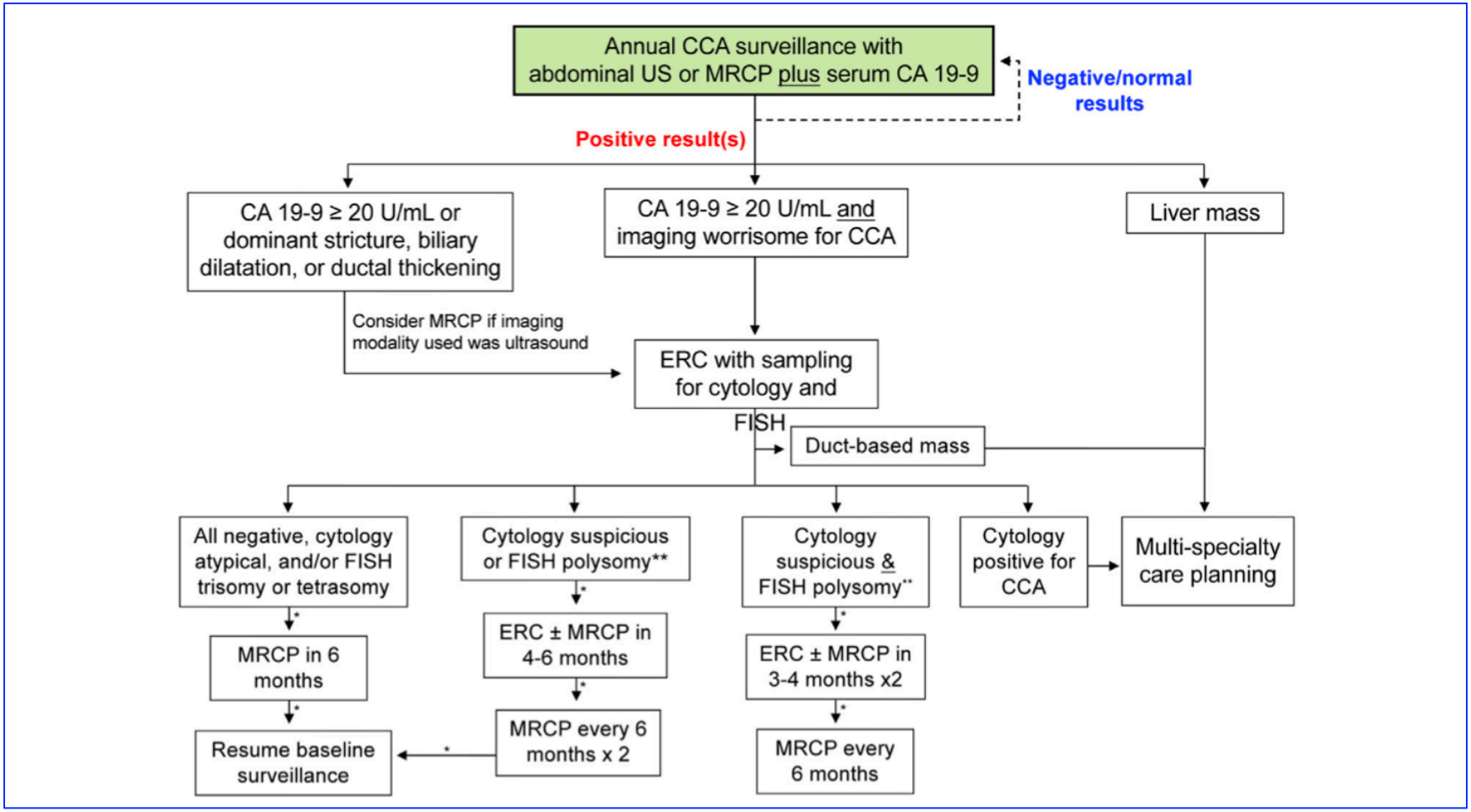
IRM et dépistage de CCA

The use of MRI/MRCP to screen for biliary cancers among asymptomatic patients with PSC should be an individualized decision. There is no quality evidence supporting or refuting CCA screening. However, many experts in the field of PSC recommend regular CCA screening with MRI/MRCP. (1C)

Recommendations on the Use of Magnetic Resonance Imaging in PSC-A Position Statement From the International PSC Study Group

Hepatology 2017

Modalités pratique de surveillance pour CCA



JH Tabibian ,Liver Research 2017

En pratique comment surveiller

- Tous les 6 mois:
  - Examen clinique
  - Tests hépatiques simples, CA 19-9
- Tous les ans:
  - Imagerie du foie et des voies biliaires (échographie « experte » ou mieux IRM hépatique et biliaire) avec examen attentif de la vésicule biliaire
  - tout polype doit faire discuter une cholecystectomie dont l'indication est formelle si taille > 8 mm
  - coloscopie avec biopsies (si MICI associée) dès le diagnostic de CSP
  - Elastométrie (?)
  - Dosage sérique vitamine D
  - Tous les 4 ans: ostéodensitométrie,
  - Si Plq < 150 000 : endoscopie digestive haute.